Single Case Report

Dissociating spatial attention from neglect dyslexia: A single case study

Margaret J. Moore* and Nele Demeyere
University of Oxford, Department of Experimental Psychology, Radcliffe Observatory Quarter, Oxford, United Kingdom

Abstract

Word-centred neglect dyslexia is generally thought to be caused by a visuospatial neglect-like attentional deficit which impacts orientation-canonical representations of visual stimuli. However, the relationship between word-centred neglect dyslexia and more general attentional processes is not well described. Here, we investigated the impact of attentional load manipulations within a case of word-centred neglect dyslexia.

This study presents data from a single case, Patient CD, who exhibited ipsilesional word-centred neglect dyslexia in conjunction with severe, contralesional allocentric neglect. CD demonstrated an intact ability to name all letters in visually presented words, but committed neglect dyslexia errors when subsequently asked to read the same word as a whole. The severity of patient CD’s neglect dyslexia was not found to be impacted by attentional manipulations. We found no effect of exposure time or visual crowding on the frequency of neglect dyslexia errors. This absence of an apparent, right-lateralised perceptual deficit, comorbid left-lateralised object-centred neglect, and insensitivity to attentional load manipulations suggests that the deficit underlying word-centred neglect dyslexia is not related to broad visuo-spatial attention. These findings suggest that neglect dyslexia and domain-general visuospatial neglect may not be as related as previously asserted.

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Introduction

Neglect dyslexia is a cluster of acquired dyslexia impairments characterized by consistently lateralized reading errors. While some forms of neglect dyslexia seem to be best understood as consequences of domain-general visuospatial neglect impairment, this characterisation may not apply to all forms of neglect dyslexia. Overall, the underlying causes of word-level neglect dyslexia are not yet clearly understood.

Neglect dyslexia is a highly heterogenous condition with many patients exhibiting qualitatively different reading error patterns. Hillis and Caramazza (1995) propose a multi-tiered model of neglect dyslexia in which patients can exhibit

* Corresponding author.
E-mail address: Margaret.moore@psy.ox.ac.uk (M.J. Moore).
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impairment within three distinct reference frames: retinocentric, stimulus-centred, or word-level (Figure One). In retinocentric neglect dyslexia, patients commit full-word and letter omission errors which are mediated by each stimulus’ location within egocentric space (Beshcen, Cisari, Cubelli, & Della Sala, 2014; Hillis & Caramazza, 1995). This impairment pattern is generally content-unspecific with patients exhibiting similarly lateralised omission errors when reading number strings or general visuospatial attention tasks (Beshcen et al., 2014). The occurrence of retinocentric neglect dyslexia has been associated with cases of particularly severe egocentric visuospatial neglect (Beshcen et al., 2014; Moore, Shalev, Gillebert, & Demeyere, 2020). Retinocentric neglect dyslexia is therefore best understood as a peripheral deficit resulting from disruption of early visual feature processing rather than an independent cognitive impairment.

A second type of neglect dyslexia is characterized by a stimulus-centred error pattern in which the prevalence of reading errors is unaffected by egocentric location but is instead modulated by allocentric (viewer-independent, stimulus-centred) spatial-attentional biases. Patients with stimulus-centred neglect dyslexia commit consistently lateralised letter omission, addition, and substitution errors, regardless of the word location in the visual space (Hillis & Caramazza, 1995). For example, the neglect dyslexia patient VB was found to misread the right letters of horizontal words presented on both the right and left sides of egocentric space, but exhibited no impairment when reading vertical words (Ellis, Flude, & Young, 1987; Hillis & Caramazza, 1995). This stimulus-centred neglect dyslexia impairment is thought to impact a level of processing in which spatial information is encoded in an allocentric reference frame, rather than an egocentric frame of reference (Hillis & Caramazza, 1995). While this stimulus-centred reading error pattern can occur in conjunction with retinocentric impairment, previous research has illustrated that these two forms of reading errors are behaviourally dissociated (Beshcen et al., 2014; Moore et al., 2020; Ptak, Di Pietro, & Schnider, 2012).

Many cases of stimulus-centred neglect dyslexia are best understood as the reading analogue of allocentric visual neglect, where impairment occurs within an object-centred spatial reference frame (Hillis & Caramazza, 1995; Ota, Fuji, Suzuki, Fukatsu, & Yamadori, 2001). This framing is in line with content-unspecific errors in stimulus-centred neglect dyslexia (e.g., similar patterns when reading numbers or lists of meaningless shapes Arduino, Daini, & Silveri, 2005; Ellis et al., 1987). However, neglect dyslexia has also been identified in patients exhibiting no visuospatial neglect impairment (Beshcen et al., 2014; Lee et al., 2008; Moore & Demeyere, 2017). For example, Friedmann and Nachman-Katz (2004) identified a neurologically healthy boy who systematically misread the left portions of individual words, while exhibiting no symptoms of visuospatial neglect on non-reading tasks. Similarly, Moore and Demeyere (2018) identified a right neglect dyslexic patient who was not found to exhibit any signs of egocentric (self-centred) or allocentric (object-centred) visual neglect when tested on a standardised cancellation task. Therefore, this categorisation of stimulus-centred neglect dyslexia as the reading analogue of allocentric visual neglect may not extend to patients exhibiting a more strictly defined word-centred neglect dyslexia.

Word-centred neglect dyslexia is characterised by reading errors which impact letters in a specific location within individual words, regardless of how these words are presented in space (Fig. 1). For example, patient NG (Caramazza & Hillis, 1990) presented with visuo-spatial neglect, but made content-specific errors when reading. NG systematically misread the terminal letters of individual words, regardless of whether these words were presented normally, vertically, or in mirror-reflected orientation. Patient AB (Moore & Demeyere, 2018) was found to exhibit a similar word-centred error pattern impacting the terminal letters of individual words in the absence of egocentric and allocentric visuospatial neglect. Hillis and Caramazza (1995) suggested that this pattern of impairment is best understood as a form of visual neglect occurring at the level of object-based stored spatial representations, producing consistently lateralised biases within stored representations of orientation-canonical stimuli. This characterisation asserts that word-centred neglect dyslexia is best understood as a symptom of a third tier of domain-general neglect impairment, at the canonical object level, rather than a reading-specific impairment (Caramazza & Hillis, 1990; Hillis & Caramazza, 1995).

However, there is only very limited evidence for rotation-insensitive neglect impairments in orientation-canonical stimuli (Buxbaum, Coslett, Montgomery, & Farah, 1996; Driver, Baylis, Goodrich, & Rafal, 1994). Several previous studies have attempted to identify rotation-insensitive neglect, but have employed stimuli which would not clearly have stored, intrinsically coded “left” and “right” sides (Cubelli & Speri, 2001; Driver et al., 1994; Driver & Halligan, 1991). Savazzi, Mancini, Veronesi, and Posteraro (2009) attempted to control for this issue by inducing specific “canonical” representation of line bisection stimuli, and identified a single patient exhibiting bisection bias reversal within reflected stimuli. However, more recent evidence has strongly

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**Fig. 1** – The three levels of neglect dyslexia reading impairment as proposed by Hillis and Caramazza (1995).
suggested that line bisection is not a reliable method for quantifying spatial-attentional biases, given that performance on this task is frequently confounded by comorbid motor or perceptual impairments (Ferber & Karnath, 2001; Sperber & Karnath, 2016). It therefore remains unclear whether word-centred neglect dyslexia can really be considered an analogue of object-based representational visuospatial neglect.

The relationship between word-centred neglect dyslexia and domain-general visuospatial neglect and attention can be investigated by manipulating factors which modulate the occurrence and severity of behavioural impairments within these conditions. Several qualitative differences between word-centred neglect dyslexia and visuospatial neglect have been documented. Patient NG exhibited a preserved ability to name each letter in visually presented word stimuli, but committed neglect dyslexia reading errors when asked to read each word as a whole (Caramazza & Hills, 1990). This demonstrates that patient NG was consciously aware of “neglected” stimuli, but simply did not employ this information when reading. This pattern of impairment is not clearly analogous to visuospatial neglect, as patients with visuospatial neglect are markedly unable to consciously report stimuli presented within the neglected side of space (Marshall & Halligan, 1988; Parton, Malhotra, & Husain, 2004). This awareness of neglected stimuli within neglect dyslexia is also evidenced by neglect dyslexia patients’ tendency to preserve word length when misreading individual words (Vallar et al., 2010). A meta-analysis of neglect dyslexia cases studies found that the most common form of neglect dyslexia reading errors was substitution rather than omission (Vallar et al., 2010). These findings suggest that patients with neglect dyslexia may demonstrate a tacit awareness of “neglected” letters or a preserved ability to encode the number of graphemes independently of their identity (Ellis et al., 1987; Vallar et al., 2010). These findings demonstrate that some attributes of word-centred neglect dyslexia are not characteristic of visuospatial neglect, suggesting that these two impairments may not be as related as previously asserted.

Only a relatively small number of word-centred neglect dyslexia patients have been identified, and none of these patients have completed experiments designed to investigate the effects of attentional manipulations on their reading impairments (Caramazza & Hills, 1990; Haywood & Coltheart, 2000; Moore & Demeyere, 2018). It therefore remains unclear whether word-centred neglect dyslexia is best understood as a form of representational neglect or as an impairment which may be unrelated to attention and overall visuo-spatial neglect. The purpose of the current study was to investigate the impact of attentional load manipulations within word-centred neglect dyslexia, in order to clarify this condition’s relationship with the neglect syndrome.

2. Methods

2.1. Case report

Patient CD was a 77 year-old man with no higher education (total ten years of education). Patient CD was admitted to the John Radcliffe Hospital’s hyper acute stroke unit in May 2019 and was diagnosed with a right middle cerebral artery stroke. Patient CD had a complex medical history including constrictive pericoronitis, pulmonary hypertension, heart failure, nystagmus, and a previous right-hemisphere middle cerebral artery stroke 2 years earlier. Following CD’s most recent cerebrovascular accident, he presented with left-sided weakness and a facial droop but remained able to walk independently with the support of a stick. Patient CD reported some reading difficulties which were new following his most recent stroke. CD completed this investigation on three sequential days, beginning 13 days after admission. CD remained alert and attentive throughout the duration of testing. The research followed approved ethical procedures (REC reference 18/SC/0550) and CD provided written informed consent.

A routine clinical CT scan (5 mm slice thickness, 32 slices) which was taken on the day of admission was retrieved in line with the study protocol. This scan demonstrated a right hemisphere ischaemic infarct centred in the basal ganglia. The location of this lesion was quantified by manually delineating the infarct on the native space scan using MRIcon (Rorden, 2007). This lesion mask was smoothed at 5 mm full width at half maximum in the z-direction and binarized using a .5 threshold. Both scan and lesion mask were reoriented to the anterior commissure and warped into 2 x 2 x 2 mm stereotaxic space using Statistical Parametric Mapping 12 and Clinical Toolbox functions (Brett, Leff, Rorden, & Ashburner, 2001; Rorden, Bonilha, Fridriksson, Bender, & Karnath, 2012). This normalised lesion mask was then compared to the Harvard–Oxford cortical and Johns Hopkins University white matter atlases to formally quantify the anatomy impacted. CD’s lesion was found to primarily affect the right frontal orbital cortex (fraction = 21.45%) and insular cortex (15.05%) as well as the right uncinate fasciculus (78.7%) and right external capsule (18.78%). Additional areas were also more minimally impacted (<4%). See Fig. 2 and Table 1 for patient CD’s clinical CT scan, lesion mask, and corresponding descriptive statistics.

2.2. Materials and procedure

Patient CD completed the Oxford Cognitive Screen (OCS) as a component of standardised post-stroke assessment. The OCS is a brief, stroke-specific cognitive screen, designed to provide a concise, comprehensive, and multi-domain summary of post-stroke cognitive impairments (Demeyere et al., 2016; Demeyere, Riddoch, Slavkova, Bickerton, & Humphreys, 2015). The OCS evaluates five domains of cognition: language, memory, attention, praxis, and number processing (see www.ocs-test.org).

2.3. Neglect tests

The OCS hearts cancellation task is a validated and highly sensitive to visuospatial neglect, with 94.12% sensitivity compared to the Behavioural Inattention Test’s star cancellation test (Demeyere et al., 2015). This cancellation task is able to reliably differentiate between allocentric and egocentric visuospatial neglect (Bickerton, Samson, Williamson, & W
In this task, patients are presented with a search matrix including 50 complete, 50 left-gap, and 50 right-gap heart line drawings pseudo-randomly distributed across a full landscape orientation A4 page. Patients are instructed to search through this array and cross off all complete drawings of hearts whilst ignoring incomplete, distractor stimuli. Patients are allowed three minutes to complete this task. Egocentric neglect is scored by subtracting the number of correctly identified targets on the left side of the page from those correctly identified on the right. Allocentric neglect is identified by subtracting the number of right-gap false positive responses from the number of left-gap false positives.

CD also completed a series of simple drawing tasks in which he was asked to draw an analogue clock from memory (e.g., Nasreddine et al., 2005) and was asked to copy a drawing of a simple scene. Finally, CD completed a computerised line bisection task. In this task a series of 100 lines ranging in length (7 cm, 9 cm, or 12 cm) were presented at one of 9 grid locations on a tablet computer screen. Patient CD was instructed to mark the centre of each line with a stylus and the distance between the actual and reported line centres was recorded in cm.

### Table 1 – Full descriptive statistics for CD’s lesion mask. All ROIs are defined according to the Harvard–Oxford and Johns Hopkin’s University Atlases. Fraction is the proportion of the ROI damaged, extension is the proportion of the lesion within each specific ROI. All impacted areas are within the right hemisphere.

<table>
<thead>
<tr>
<th>ROI Name</th>
<th>n Voxels</th>
<th>n Voxels Significant</th>
<th>Fraction</th>
<th>Extension</th>
</tr>
</thead>
<tbody>
<tr>
<td>Uncinate fasciculus</td>
<td>616</td>
<td>485</td>
<td>.787</td>
<td>.038</td>
</tr>
<tr>
<td>External Capsule</td>
<td>8083</td>
<td>2379</td>
<td>.94</td>
<td>.188</td>
</tr>
<tr>
<td>Orbitofrontal Cortex</td>
<td>14,694</td>
<td>2705</td>
<td>.184</td>
<td>.214</td>
</tr>
<tr>
<td>Insular Cortex</td>
<td>11,505</td>
<td>1907</td>
<td>.66</td>
<td>.151</td>
</tr>
<tr>
<td>Parahippocampal Gyrus (Anterior Division)</td>
<td>8163</td>
<td>426</td>
<td>.65</td>
<td>.094</td>
</tr>
<tr>
<td>Internal Capsule Anterior Limb</td>
<td>4119</td>
<td>174</td>
<td>.042</td>
<td>.014</td>
</tr>
<tr>
<td>Planum Polare</td>
<td>4361</td>
<td>177</td>
<td>.041</td>
<td>.014</td>
</tr>
<tr>
<td>Anterior Corona Radiata</td>
<td>8756</td>
<td>136</td>
<td>.016</td>
<td>.011</td>
</tr>
<tr>
<td>Subcallosal Cortex</td>
<td>5482</td>
<td>63</td>
<td>.011</td>
<td>.005</td>
</tr>
<tr>
<td>Internal Capsule (Retrolinear Part)</td>
<td>3286</td>
<td>15</td>
<td>.005</td>
<td>.001</td>
</tr>
<tr>
<td>Temporal Pole</td>
<td>23,987</td>
<td>101</td>
<td>.004</td>
<td>.008</td>
</tr>
<tr>
<td>Fornix (Cres) Stria Terminalis</td>
<td>1891</td>
<td>6</td>
<td>.003</td>
<td>.000</td>
</tr>
<tr>
<td>Internal Capsule (Posterior Limb)</td>
<td>4648</td>
<td>11</td>
<td>.002</td>
<td>.001</td>
</tr>
</tbody>
</table>

### 2.4. Reading tasks

In each of this investigation’s reading tasks, a series of written stimuli were presented sequentially and centrally on a Windows Surface touch-screen tablet computer. This presentation program was written using MATLAB’s Psychtoolbox (Brainard, 1997). Each stimulus was displayed in all capital letters in size 23 Arial font at the centre of the screen, and remained present for an unlimited duration, until a response was made by the participant. Given a viewing distance of approximately 50 cm, the viewing angle for the horizontal and vertical stimuli was between 2 and 12°, depending on stimulus length. Responses were recorded as correct or incorrect by the examiner through a tap on the left or right side of the screen. Audio recordings of patient responses were collected and were subsequently reviewed to confirm and code error types. Neglect dyslexic reading errors were classified based on the criteria set by Ellis et al. (1987).

These reading tasks consisted of real words, pronounceable pseudowords, number strings, and unpronounceable letter strings. For transparency, the full list of reading stimuli are included in the supplementary materials made available on Open Science Framework (https://osf.io/27n85/). All stimuli types were presented in separate experimental blocks to avoid
potentially confounding effects of intermixed presentation. Real word stimuli were selected to include many roots with multiple derivational affixes in order to increase the probability of neglect dyslexia reading errors (Riddoch, 1990; Vallar et al., 2010). The unpronounceable strings used in this investigation consisted of consonants only. See Table 2 for an overview of the stimuli and manipulations included in each of the 8 reading tasks and the order of administration. No part of the study procedures or analyses was pre-registered prior to the research being conducted. We report all data exclusions (if any), all inclusion/exclusion criteria, whether inclusion/exclusion criteria were established prior to data analysis, and all measures in the study.

3. Results

3.1. Cognitive screening and general neglect testing

CD exhibited significant impairment in the orientation (3/4), reading (8/15), number writing (2/3, 15,200 written as 50,200), calculation (2/4), cancellation task (Total = 23, Egocentric Asymmetry = 13, Allocentric = 15), praxis (8/12), and verbal memory (1/4) subtests of the Oxford Cognitive Screen on admission. The OCS cancellation task was repeated on five separate days (twice before beginning reading testing and on each day of reading testing) (Fig. 3). Notably, CD was found to exhibit severe left allocentric neglect on each of these testing occasions (on average 15.8 left allocentric errors). Fig. 4 presents the cancellation task CD completed immediately before beginning reading assessment. Though CD had also exhibited significant left egocentric neglect immediately after the stroke, this impairment was no longer present when the reading-specific tests were conducted 13 days later.

CD exhibited no apparent signs of visuospatial neglect when completing the clock drawing task (on day 3), but omitted the leftmost portion of the ground line when completing the scene copy task (Fig. 3). CD demonstrated a slight bias towards the right side of space demonstrating left inattention (M = .22 cm, SD = .633, left inattention) when completing the line bisection task. This finding is in line with impairment on line bisection tasks reflecting allocentric, rather than egocentric neglect impairment (Mennemeier, Chatterjee, & Heilman, 1994).

3.2. Initial reading

First, CD read a series of 100 normally presented words under unlimited exposure time. In this condition CD read 70/100 words correctly, committed 21 neglect dyslexia reading errors, and made 9 other reading errors [5 no response, 3 visual errors (non-lateralised letter omissions or switches — e.g., MILLSTONE read as “milestone”), and 1 unintelligible response]. Of CD’s right neglect dyslexia reading errors, 11 involved letter substitutions (e.g., THRASHING read as “thrasher”) and 10 involved letter omissions (e.g., REMISSION read as “remiss”). All but two of these omissions and substitutions (90.5%) impacted the terminal letters of words. This distribution of reading error types is consistent with previous case studies of word-centred neglect dyslexia patients (Caramazza & Hillis, 1990; Moore & Demeyere, 2018).

3.3. Experiment one: reference frame manipulations

Next, a series of manipulations was conducted to determine whether CD’s neglect dyslexia represented a retino-centric, stimulus-centred or a word-centred reading deficit. CD’s proportion of right neglect dyslexia reading errors was not found.

<table>
<thead>
<tr>
<th>Table 2 – Details of the various assessments employed in this investigation. E.T. = Exposure Time.</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Reading Tests</strong></td>
</tr>
<tr>
<td>Reading Baseline</td>
</tr>
<tr>
<td>Initial 100 3–9 Unlimited 1</td>
</tr>
<tr>
<td>Orientation Manipulations</td>
</tr>
<tr>
<td>Vertical 75 3–10 Unlimited 1</td>
</tr>
<tr>
<td>Reflected 56 3–8 Unlimited 1</td>
</tr>
<tr>
<td>Content Manipulations</td>
</tr>
<tr>
<td>Pseudowords 44 3–9 Unlimited 2</td>
</tr>
<tr>
<td>Letter Strings 43 3–5 Unlimited 2</td>
</tr>
<tr>
<td>Attentional Manipulations</td>
</tr>
<tr>
<td>Location 100 5 500 msec 1</td>
</tr>
<tr>
<td>Reduced Exposure 73 3–9 400 msec 2</td>
</tr>
<tr>
<td>Spacing Manipulations</td>
</tr>
<tr>
<td>OCS Hearts Cancellation 1,2,3</td>
</tr>
<tr>
<td>Clock Drawing/Figure Copy 3</td>
</tr>
<tr>
<td>Line Bisection 2,3</td>
</tr>
</tbody>
</table>
to be significantly different for stimuli presented on the right (10/50) or left (11/50) side of space ($X^2(1) = .605, p = .437$). CD then read a series of 75 vertically presented words and 56 reflected words, presented in separate blocks. In the vertical condition, CD read 24/75 words correctly and committed 34 neglect dyslexia errors ($N_{mission} = 18$, $N_{substitution} = 15$, $N_{addition} = 1$). All 34 neglect dyslexia errors impacted the terminal letters of individual words. Of the 17 other errors, 7 were no responses, 8 were unintelligible responses, and 2 were non-lateralised visual errors.

In the reflected condition, CD read 20/56 words correctly and committed 29 neglect dyslexia errors (20 substitution, 9 omission). All of these lateralised errors impacted the terminal letters of words, even though these usually right-lateralised letters were now presented in the left side of egocentric space. CD also committed 5 letter-by-letter and 2 non-lateralised visual errors in the reflected reading conditions. Overall, this error pattern strongly suggests that CD’s neglect dyslexia is best characterised as a word-centred reading impairment (Caramazza & Hillis, 1990; Hillis & Caramazza, 1995).

### 3.4. Experiment two: content manipulation

Next, a series of content manipulations was conducted to investigate whether CD’s neglect dyslexia represented a domain-general visual feature processing impairment or a content-specific deficit. CD was presented with a series of 43 unpronounceable letter strings and 44 pseudowords. CD read 39/43 letter strings correctly. Of the four errors committed in this condition, 3 were non-lateralised visual errors and 1 was a left-lateralised letter substitution error (LRMXT read as “A-R-M-X-T”). In the pseudoword reading condition, CD read 22/44 pseudowords correctly. Of the 22 errors committed, 4 were letter-by-letter errors, 9 were non-lateralised visual, and 9 were right-lateralised neglect dyslexia errors (7 substitutions, 2 omissions). A chi-squared test was then performed to investigate the relationship between neglect dyslexia prevalence and stimulus type. Overall, there was a significant difference in the prevalence of neglect dyslexia errors when reading letter strings and pseudowords ($p = .005$) and between letter string and real words ($p = .003$), there was no statistically significant difference between the prevalence of neglect dyslexia errors when reading real words and pseudowords ($p = 1.00$).

### 3.5. Experiment three: attentional manipulations

A series of attentional manipulations was conducted to investigate whether the severity of word-centred neglect dyslexia is modulated by the same factors which modulate the severity of visuospatial neglect impairment. First, CD read a series of 100 horizontally presented real words under unlimited exposure time. Half of these words were presented under normal letter – spacing condition (visual angle approximately 7°) and half were presented under a wide spacing condition (visual angle approximately 11.5°). Overall, CD read 57/100 words correctly and committed 43 reading errors [1 unintelligible, 5 generalised visual, 5 letter-by-letter, and 32 neglect dyslexia (25 substitution, 5 omission, and 2 addition)]. CD’s proportion of neglect dyslexia reading errors was not found to be significantly different between the normal and wide spaced reading condition [26/51 normal vs 29/49 spaced, $X^2(1) = .318$, $p = .573$].

Next, CD read a series of 73 real words presented under reduced exposure time (500 msec). CD was found to read 40/73 words correctly and to commit 1 unintelligible, 12 no response, 1 non-lateralised visual, and 19 (10 substitution, 9 omission) neglect dyslexia errors. The proportion of neglect dyslexia errors committed was not found to be significantly different between the unlimited exposure and limited exposure conditions [$X^2(1) = .350, p = .554$].

### 3.6. Letter-by-letter spelling

When shown a real word reading stimulus, CD frequently responded that he did not know what the word was. In cases where this occurred, CD was asked if he could see the individual letters within words. CD demonstrated a preserved ability to name all individual letters within words. However, when asked to read the word as a whole, CD consistently committed neglect dyslexia reading errors. For example, when shown the word RADIANS, CD reported that he did not
recognise the word and was therefore prompted to name all the letters. CD correctly reported all letters within this word (“R-A-D-I-A-N-S”), but when asked to read the word as a whole he read RADIANS as “radish”. Similarly, CD misread LEARNED as “legend”, CLOVER as “cloves”, and EVERY as “even” immediately after successfully naming all the letters within individual words. CD committed this error pattern 35 times in this investigation’s real-word, unlimited exposure conditions. Audio files of these reading errors are available on the Open Science Framework (https://osf.io/27n85/).

4. Discussion

The purpose of the present study was to investigate the relationship between word-specific neglect dyslexia and more general aspects of spatial attention. Patient CD was found to exhibit right word-centred neglect dyslexia in conjunction with left allocentric visuospatial neglect following a right hemisphere MCA stroke. Notably, this is the first documented case of word-centred neglect dyslexia occurring in conjunction with oppositely lateralised visuospatial neglect. In this study, we investigated the impact of attentional load manipulations within word-centred neglect dyslexia, in order to further clarify this condition’s relationship with broad levels of attention. CD’s neglect dyslexia was found to be specific to stimuli which he was required to “read as a whole” rather than to simply name all letters. The severity of CD’s reading impairment was not found to be modulated by attentional factors which have been found to modulate the severity of visuospatial neglect, suggesting that CD’s reading impairment may not be accurately characterised as a neglect deficit. The data collected in this investigation offer a unique insight into the cognitive mechanisms subserving domain-general and reading-specific visuospatial perception.

CD’s stroke was found to result in ischemic lesions to areas within the right hemisphere middle cerebral artery vascular territory, primarily the orbito-frontal and insular cortex as well as the right external capsule. Neglect dyslexia is generally associated with lesions impacting the lingual and fusiform gyri (Moore & Demeyere, 2018), but neglect dyslexia patients exhibiting more frontal lesions have been identified (Friedmann & Nachman-Katz, 2004; Vallar et al., 2010) and a previous study by Moore and Demeyere (2018) documented a case of neglect dyslexia in a patient with transient ischemic attack with no visible lesions. Cases of ipsilesional neglect impairment, like the one described here, have been previously documented, though this impairment seems comparatively rare (Cubelli, Nichelli, Bonito, De Tanti, & Inzaghi, 1991; Kim et al., 1999; Kwon & Heilman, 1991). Kim et al. (1999) identified five cases of ipsilesional neglect and found that ipsilesional neglect impairments were associated with frontal-subcortical lesion patterns, similar to that of patient CD. Left allocentric neglect is traditionally associated with more posterior lesions impacting the angular, middle temporal, and middle occipital gyri (Chechlacz, Rotstein, & Humphreys, 2012, 2010; Hills & Caramazza, 1991; Medina et al., 2008), but previous research has demonstrated that damage to a surprisingly wide range of neural correlates can result in visuospatial neglect impairment (e.g., Bird, 2006; Hildebrandt, Spang, & Ebke, 2002; Karnath, Himmelbach, & Rorden, 2002; Karnath & Rorden, 2012; Mort et al., 2003). CD’s lesion data demonstrates that ipsilesional word-centred neglect dyslexia can occur, but data from additional patients is required before any firm conclusions about the neural correlates of word-centred neglect dyslexia can be drawn.

Patient CD’s reading error pattern aligns closely with that of a word-centred neglect dyslexia deficit as defined by Hillis and Caramazza (1995). CD was found to commit reliable letter omission and substitution errors impacting the terminal letters of individual words, regardless of how these words were presented in space. This reading error pattern has been documented in a series of previous case studies (Caramazza & Hillis, 1990; Haywood & Coltheart, 2000; Moore & Demeyere, 2018) but this is the first case in which word-centred neglect has been found to co-occur with oppositely lateralised visuospatial neglect impairment. Preliminary evidence for a dissociation between word-centred neglect dyslexia and the neglect syndrome was provided by patient AB, who was admitted following a transient ischemic attack and exhibited a similar pattern of right word-centred neglect dyslexia in the absence of visuospatial neglect (Moore & Demeyere, 2018). Patient CD’s novel impairment conjunction builds upon these previous findings by providing further evidence suggesting that word-centred neglect dyslexia can be dissociated from domain-general visuospatial neglect, not just in occurrence but also in lateralisation.

Theories which characterise neglect dyslexia as a peripheral impairment caused by domain-general neglect have difficulty accounting for CD’s co-occurring left allocentric neglect and right word-centred neglect dyslexia. Driver and Pouget (2000) suggested that object-level neglect impairments can be best understood as sort of “relative” egocentric neglect which is mediated by retinal position on an egocentric gradient of inattention. However, it is not clear how a right-lateralised neglect dyslexia impairment could be expected to arise from a left-lateralised domain-general neglect impairment. Riddoch (1990) proposed that neglect dyslexia which appears to occur independently of neglect may be best understood as a form of stimulus-density sensitive impairment in which a sub-clinical neglect deficit is exacerbated to the point of significance by the increased visual crowding associated with reading. However, it remains unclear how increasing visual crowding could be expected to change the lateralisation of a domain-general neglect impairment. CD’s right neglect dyslexia therefore seems best understood as an ipsilesional neglect dyslexia impairment which operates independently of domain-general neglect impairment.

If the same three-level perceptual system was responsible for subserving perception of all visual stimuli, a patient with right word-centred neglect dyslexia would be expected to exhibit right allocentric neglect when completing the cancellation task, as the impairment underlying reading difficulties would also underlie perception of the heart line drawings. However, patient CD was found to exhibit a consistent, severe left-lateralised allocentric deficit when completing the cancellation task. CD represents the first documented case of word-centred neglect dyslexia co-occurring with oppositely lateralised visuospatial neglect. Though previous reports have
included lateralised reading errors in conjunction with oppositely lateralised visuospatial neglect (Costello & Warrington, 1987; Moore et al., 2020), the current study is the first to confirm that the neglect dyslexic deficit occurs within a word-centred reference frame. CD’s novel impairment conjunction would not be expected if the impairment underlying word-centred neglect dyslexia also played a role in domain-general spatial attention and perception. Importantly, these findings strongly suggest that a neglect-like impairment at the level of spatially canonical internal representations does not necessarily represent damage to a domain-general system.

Further support for this conclusion is provided by CD’s performance on the attentional manipulation experiments. Word-centred neglect dyslexia is thought to be analogous to both retino-centric and stimulus-centred neglect impairment, but simply within a separate reference frame. However, CD's performance on this investigation's attentional manipulations suggests that the severity of word-centred neglect is not modulated by the same factors which modulate the severity of retino-centric and object-level neglect impairments. First, reducing exposure time was not found to exacerbate CD’s reading impairment. Previous studies have found that reducing exposure in reading tasks appears to exacerbate the severity of domain-general neglect impairments. For example, Ellis et al. (1987) assessed patient VB, a patient exhibiting both domain-general neglect and retino-centric neglect dyslexia, and found that VB’s frequency of neglect dyslexia errors rose from 6%-8%–15% when exposure time for individual words was limited.

Second, the reduction of visual crowding was not found to significantly impact the severity of CD’s reading impairment. It is well established that the severity of visuospatial neglect can be exacerbated by increasing visual crowding and density within cancellation tasks (Husain & Kennard, 1997; Kartsounis & Findley, 1994; Parton et al., 2004). It therefore follows that the effects of domain-general neglect on word reading would be expected to be ameliorated by reducing the visual density of word stimuli. This pattern was not observed within patient CD. Finally, visuospatial neglect is characterised by a marked inability to consciously report stimuli presented within neglected space (Ellis & Young, 2013; Marshall & Halligan, 1988). However, CD was able to name individual letters within words, even in cases where he committed a neglect dyslexia reading errors. Considered cumulatively, this evidence suggests that CD’s reading error pattern may not be best characterised as a visuospatial neglect related deficit.

We suggest it is possible that CD’s reading error pattern may represent a subtle executive function impairment rather than a strictly perceptual, neglect-related impairment. Under this hypothesis, a lack of inhibitory control might lead to CD completing the words with a more prepotent response. CD demonstrated a preserved ability to perceive and report individual letters, but only considered information from the left-most portions of words when reading as a whole. This impairment pattern could be in line with inference errors committed by children guessing words which they have not yet acquired lexical representations for and thus be related to a regulation deficit. For example, it is possible that CD simply lacked the self-regulation necessary to inhibit responding to presented reading stimuli until all relevant information had been processed. If this were the case, CD would be expected to respond based on information within the first-processed letters while ignoring any information presented in latter letters, resulting in apparently neglect dyslexic reading impairment. It seems possible that CD’s reading performance may have been impacted by executive deficits, especially given that CD’s lesion impacted basal ganglia white matter tracts which have been found to be significantly associated with executive functioning (Hua et al., 2014; Vataja et al., 2003). Further research on ipsi- sional neglect dyslexia could investigate this hypothesis in more detail by including executive tasks such as the Hayling test (Bielak, Mansueti, Strauss, & Dixon, 2006) which measure prepotent response suppression.

However, single case studies are inherently better at demonstrating what neuropsychological deficits are not rather than what they are. CD’s impairment pattern alone should not be interpreted as strong evidence for or against the characterisation of word-centred neglect dyslexia as an inhibition deficit. We present this alternate explanation of neglect dyslexia primarily to suggest a direction for future research rather than to draw strong theoretical inferences from the presented patient. Additional research in group studies is needed before any firm conclusions about the underlying causes of word-centred neglect dyslexia can be drawn.

4.1. Limitations

The investigators were only able to access and assess patient CD for the duration of his hospitalisation, creating a very limited time-window for his reading error pattern to be documented. In addition, at a time so shortly after stroke, CD fatigued easily and could not sustain lengthy testing conditions. This investigation dealt with these time constraints by limiting the number of words read in each condition. This adjustment does not decrease the theoretical significance of the documented qualitative neglect dyslexia reading error pattern, but instead only lessens the statistical power of comparisons between conditions in which CD only read a small number of stimuli. Additionally, some stimuli were presented twice in the reading experiments. These repeated stimuli were intentionally included in multiple orientation conditions in order to facilitate a direct comparison between CD’s exact responses for the same stimuli in multiple different topographical presentations. While carry-over effects cannot be entirely ruled out, their effects should be minimal as each stimulus was presented a maximum of three times throughout three separate days of testing. Finally, there was no data available detailing CD’s pre-morbid cognitive abilities. It is unclear whether the cognitive impairments observed in CD were directly related to his most recent stroke or whether they were residual impairments from his previous neurological events.

Previous case studies have revealed that neglect dyslexia is a highly heterogeneous condition. While some patients, have been found to exhibit neglect dyslexia impairment patterns which are similar to CD, the conclusions of this investigation cannot be generalised to all cases of neglect dyslexia (Vallar
et al., 2010). There is a lack of data from standardised assessments of substantive samples of neglect dyslexia patients, precluding valid comparisons about the condition as a whole. Specifically, comparatively few detailed studies of word-centred neglect dyslexia have been conducted, making it difficult to draw generalisable conclusions. It remains unclear whether the attentional manipulation effects documented in CD can be identified in a wider sample of word-centred neglect dyslexia patients. Additional research is needed to confirm whether some cases of word-centred neglect dyslexia may be related to executive impairment rather than to visuospatial impairment.

4.2. Conclusion

Patient CD was found to exhibit ipsilesional right word-centred neglect dyslexia in conjunction with left allocentric visuospatial neglect, illustrating a dissociation between these conditions. The severity of CD’s reading impairment was not found to be modulated by attentional factors which have been found to modulate the severity of visuospatial neglect, suggesting that CD’s reading impairment may not be accurately characterised as a neglect-like deficit. These data, considered in conjunction with CD’s preserved ability to perceive and correctly identify individual letters implies that his reading impairment is not related to visuo-spatial attentional function and instead we suggest it may be related to a self-regulation executive dysfunction. However, additional research is needed to conclusively determine whether ipsilesional word-centred neglect dyslexia is, in actuality, not a neglect-related reading impairment.

Credit author statement

MM conceived the study, collected data, performed analysis, and drafted manuscript. ND supervised study design and analysis and aided in manuscript preparation.

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Open practices

The study in this article earned Open Materials and Open Data badges for transparent practices. Materials and data for the study are available at https://osf.io/27n85/.

Declaration of Competing Interest

All authors declare that there are no conflicts of interest in this paper.

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