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Alexia and quadrant-amblyopia: Reading disability after a minor visual field deficit

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Abstract

Reading difficulties caused by hemianopia are well described. We present a study of alexia in a patient (NT) with a milder visual field deficit. The patient had suffered a cerebral haemorrhage causing damage to the left occipital cortex and underlying white matter. NT's text reading was slow and prone to error, but recognition of single letters was preserved. Single word reading was accurate, but slower than normal. On perimetric testing NT initially showed an upper right quadrantanopia, but by attending covertly to this quadrant he could achieve luminance detection except in a small scotoma above the reading line. A whole report experiment showed that letter perception was severely compromised in the quadrant, consistent with cerebral amblyopia. On follow-up testing one and a half year post stroke, a clear spontaneous recovery had occurred, reflected in improved text reading with close to normal eye movements. Still, subtle reading difficulties and oculo-motor abnormalities remained. Overall, the study shows how amblyopia in one quadrant can lead to a characteristic form of alexia.

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Keywords: Cerebral amblyopia; Eye movements; Hemianopic alexia; Quadrantanopia; Reading; TVA

1. Introduction

Damage to posterior areas in the left hemisphere of the brain can result in reading disability (alexia) for various reasons. In the classical syndrome of alexia without agraphia (pure alexia), single word recognition is impaired, and in most cases the recognition of single letters is also compromised. Many patients with this syndrome resort to a compensating spelling strategy in reading (letter-by-letter reading) resulting in a pronounced effect of word length on response latency.

Pure alexia is almost invariably accompanied by a homonymous visual field defect affecting either the entire right hemifield or parts thereof (Damasio & Damasio, 1983). Such visual field cuts may in themselves lead to reading problems. Even when recognition of words is normal in the intact part of the visual field, a very high proportion of patients with visual field defects read text more slowly than controls (Kerkhoff, 1999). This condition has been termed hemianopic alexia (Willbrand, 1907), although it can also occur after smaller field defects (e.g., quadrantanopia). Generally, patients with hemianopic alexia have disproportionate problems in reading text compared to single words, and this has been attributed to deficient planning and execution of eye movements during reading (Zihl, 1995). In patients with alexia due to right sided hemianopia, eye movements are characterised by shorter and more frequent saccades to the right, prolonged fixation times and an increase in the number of regressive saccades (to the left). There are reports of patients with right hemianopia but no alexia (e.g., Binder & Mohr, 1992; Leff et al., 2001), and the presence and degree of reading disorder seems to be at least partially dictated by the degree of sparing of foveal vision (Zihl, 1995), and is also related to lesion localization. A lesion affecting either the left primary visual cortex or its geniculostriate afferents, but sparing both the left occipitotemporal junction and its connections to the right occipital lobe, will lead to hemianopic alexia if right foveal vision is compromised (Leff et al., 2001).

There are multiple similarities between (right sided) hemianopic alexia and pure alexia. Both patient groups have homonymous visual field defects, and both disorders give rise to word-length effects in reading, although of differing magnitude. Eye movements during reading also seem to be disturbed in both disorders (Behrmann, Shomstein, Black, & Barton, 2001; Zihl, 1995).
However, in patients with alexia due to right-sided hemianopia, single letter recognition is preserved. Typically these patients read short single words normally, and show an increase in reaction times only with longer words. As pointed out by Leff et al. (2001), care should be taken to distinguish between hemianopic and pure alexia, and one should also strive to tease apart the contribution made by the visual field defect from word or letter recognition deficits in individual patients. This is especially important because specific rehabilitation techniques exist to ameliorate reading deficits relating to hemianopia (Kerkhoff, Münssinger, Eberle-Strauss, & Stögerer, 1992; Zihl, 2000).

Visual defects that spare some luminance sensitivity while impairing shape processing in parts of the visual field, cerebral amblyopia, may also cause reading disturbance (Kerkhoff, 1999). Cerebral amblyopia is a less known field deficit, which does not lead to general problems in visual exploration and is often overlooked in the clinic. If at all aware of the condition, amblyopic patients experience a stimulus in the affected region as “foggy” and are often unable to describe its form (Fahle, 2002). Reading difficulties after cerebral amblyopia have typically been studied as a minor part of broader investigations on hemianopic alexia (e.g., Kerkhoff, 1999; Zihl, 1995), in which they have been grouped under the general heading hemiamblyopia. The term hemiamblyopia indicates that the whole hemifield is affected, which leaves the effect of minor types of amblyopia unclear. In general, detailed individual descriptions of amblyopic alexia are lacking.

We present a case study of alexia in relation to cerebral amblyopia confined to the upper right quadrant. To our knowledge, reading disability after this visual field defect has not previously been described in detail. Using a combination of alexia assessment, psychophysical testing of vision, and eye movement recordings, we provide a comprehensive account of the reading disability and its relation to basic visual processes. The study draws attention to a type of alexia that is easily overlooked in the clinic.

2. Case report

2.1. Medical background

NT is a right-handed man (Edinburgh Handedness LQ = +100; Oldfield, 1971) who in 2002 was 20 years old. He had no previous history of neurologic, psychiatric, or cardiovascular disease. In October 2002 he presented with a generalised epileptic seizure. Acute CT scan showed an intracerebral haemorrhage posteriorly in the left hemisphere with intrusion to the ventricle system. After evacuation of the

Fig. 1. T1-weighted MR scan of NT’s lesion. The lesion is shown in coronal, sagittal, and transversal views, as well as rendered in three dimension (produced using MIRCen software: Rorden & Brett, 2001).
haematoma, NT complained of headaches and double vision as well as memory impairment and difficulties in reading and writing. No visual field defect was noted in neurological examinations. A CT scan in December 2002 (2 months post injury) showed substance loss in the left occipital lobe lateral to the posterior horn of the lateral ventricle, which was dilated. Both temporal horns were slightly dilated, but there was no remaining haematoma. An MR scan in May 2004 localized the lesion to cortical areas in the basal occipital lobe with significant damage to the underlying white matter. The medial surface of the occipital lobe was intact, and primary visual cortex was in all probability spared (see Fig. 1).

NT gave informed written consent according to the Helsinki Declaration to participate in the study, and approval was given by ethical committees in Copenhagen (project no.: KF 01-116/02).

2.2. General neuropsychological assessment

NT’s medical records state that 2 months post injury he showed psychomotor slowness (as measured by the Trail Making Test), a deficit in verbal memory (word-list learning), and severe reading difficulties, especially in text reading. All other test results were within the normal range.

We conducted further assessment between February and May 2003 (4–7 months post-injury; see summary of test results in Table 1). At this time, NT was neurologically stable and the only remaining complaint was of reading difficulties. NT’s complaints were focused on his diminished ability to read fast and to read out loud. Premorbidly he had read a lot, both classical literature and scientific texts. He also enjoyed reading aloud to friends and family, and his family confirmed that he was talented in this. NT had also been a skilled reader of English.

Neuropsychological assessments revealed that performance on the Trail Making Test (A and B) was now normal, while speed on the Symbol Digit Modalities Test (SDMT) was below normal. Naming, verbal understanding, and verbal abstraction were at or above average level. There was no deficit in non-verbal memory, but the results in a word-list learning test were below normal both at learning and retention. Results in the Danish Adult Reading Test (Danish version of NART) was in the upper part of the normal range (+1S.D.) compared to Danish norms, indicating premorbid IQ in the average to superior range. Basic visual object recognition and pattern integration was intact. NT had no difficulties in visuo-constructive tasks, including copying, but was slightly hesitant when drawing from memory. His written production was flawless. In summary, although his performance was subnormal on one test of psychomotor speed and one verbal memory test, NT’s only neuropsychological complaint at this point was his reading difficulties.

2.3. Clinical testing of reading skills

In addition to the general neuropsychological assessment we explored NT’s reading performance, aiming to clarify the nature of his possible alexia. For two of the tests, a lexical decision and a text reading task, we obtained control data from a group of age- and education-matched participants (n = 5; mean age = 23 years, all undergraduate university students). To statistically analyze NT’s performance compared to this control group, we used a test devised by Crawford and Howell (1998) that has proven highly robust for evaluating single-case results against control groups of limited size (Crawford, Garthwaite, Azzalini, Howell, & Laws, 2006). Scores deviating more than 2.34S.D. from the control group reached significance on this test (i.e., were classified as pathological).

### Table 1

<table>
<thead>
<tr>
<th>Visual perception</th>
<th>VOSP</th>
<th>Shape detection (Screening) 20/20</th>
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<tbody>
<tr>
<td></td>
<td></td>
<td>Shape detection (letters) 20/20</td>
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<tr>
<td></td>
<td></td>
<td>Dot counting 10/10</td>
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<tr>
<td></td>
<td></td>
<td>Poppelreuter overlapping figures 14/14</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Street completion test 13/20</td>
</tr>
</tbody>
</table>

| Visuo-constructive                       | Rey’s complex figure, copying 36/36 |
|                                          | Copying test (cross, star, cube, house) 10/10 |
|                                          | Block design (Danish version) 12/12 |
|                                          | Block design time 16 s               |
|                                          | Drawing from memory 6/6              |

| General attention                        | Digit span, forward 5–6            |
|                                          | Digit span, backwards 4–5          |
|                                          | Mental subtraction (100 – 7) 38 s   |

| Stroop                                   | Words 20 s                        |
|                                          | Colors 28 s                       |
|                                          | Interference 37 s                 |

| Psychomotor speed                        | Trail making test A 18 s           |
|                                          | Trail making test B 73 s           |
|                                          | SDMT 39*                          |

| Memory                                   | Rey’s complex figure, recall (3 min retention) 23 |
|                                          | 10 word list (A.M. Busche) Learning 10/10 |
|                                          | Errors 20*                         |
|                                          | Retention 6/10*                    |

| Language                                 | Boston naming—split half 28/30 |
|                                          | WAIS similarities 21/26         |
|                                          | Semantic fluency (animals) 22   |
|                                          | Phonological fluency (5-words) 19|

| Writing                                  | Picture description 1/1          |
|                                          | Dictation 13/14                 |
|                                          | Numbers/arithmetic 10/10        |
|                                          | Oral spelling 5/5               |
|                                          | Naming to spelling 15/15        |

| NART (Danish version)                    | 33/50                           |

All results are reported as raw scores. Results below average (compared to Danish norms) are marked out with an *.
2.3.2. Word reading

2.3.2.1. Nouns. NT was presented with a reading task consisting of 52 concrete nouns of three, five, or seven letters. Half the words were high-frequency (>20 per million, frequency ratings from Bergenholz, 1992). Words were presented in 36-point Times New Roman (white letters on black background) one at a time, centrally on a computer screen. NT was asked to read the words as quickly and accurately as possible, and the initiation of a verbal response terminated the presentation of the words. Errors were noted by the experimenter. NT made no reading errors in this task.

2.3.2.2. Lexical decision. NT was presented with a lexical decision task consisting of 39 nouns and 39 non-words that were three, five, or seven letters long. The non-words were constructed by changing one or two letters in the nouns (one letter in three-letter words, two letters in five- and seven-letter words). All non-words were pronounceable, but meaningless. Half the nouns were concrete (as listed by Paivio, Yuille, & Madigan, 1968) and half were high frequent (see above). The words were presented for 250 ms on a computer screen, in the same format as in the word reading test. NT was told that he might not be able to read the words given the short exposure time, and that he should try to derive a feeling of the word and then decide whether it was familiar or not. Responses were made by pressing buttons on a response box, the left button for real words and the right button for non-words. NT made three errors in this task, all of which he spontaneously corrected. The test was also given to five control participants, who on average made 5.4 errors (S.D. = 2.2). Thus NT’s error rate clearly fell within the normal range.

2.3.2.3. Word regularity and non-word reading. On the regularity-subtest of the PALPA (Test 35, Kay, Lesser, & Coltheart, 1992; Danish version: Lønnberg & Hallas-Møller, 2005) NT misread one regular and one irregular word out of 30 words. NT was also presented with the homophone decision task from PALPA (Test 28), which comprises 60 pairs of words and non-words that are to be read out loud followed by a judgement of whether the two words sound the same. In this test NT read aloud a total of 40 regular words, 40 irregular words and 40 non-words. NT made four errors in this test: three non-words and one irregular word were misread.

2.3.3. Number reading

When presented with single digits and common symbols (e.g. ?, 5, &), NT named all digits and symbols rapidly and correctly. With multi-digit numbers presented the same way, NT showed no difficulties in reading two- and three-digit numbers. When asked to read multi-digit numbers (>3 digits) out loud, NT had difficulties in naming hundreds, thousands, and upwards. He could correctly name the digits comprising the multi-digit number, but failed to attach the right label to them. For instance he named 45,679 as “forty-five, six, seven, nine”. Thus he seemed to apply a “digit-by-digit” strategy in this test when reading numbers with more than three digits.

Number identification was further investigated by administering the number reading tasks devised by Cohen and Dehaene (1995), where single and two digit numbers are read several times with different and increasingly complex task demands. In the eight administered tests, comprising 320 digits all together, NT made three reading errors (0.9%). Two of these errors were made in the most complex task. In comparison, the pure alexic patients studied by Cohen and Dehaene (1995) made three to seven reading errors even on the easiest task (reading single digits), and on average 27% reading errors all together.

2.3.4. Text reading

We presented NT with four texts of varying complexity: two Danish texts, one a literary text (from a novel) and one from an undergraduate textbook in biology, and two English texts; one literary and one scientific. NT was asked to read the text from the book, while errors were noted by the experimenter. NT was asked to read the texts out loud at his own pace.

In general, NT’s reading of all four texts was slow and effortful. He frequently hesitated before pronouncing a word, and his prosody was without respect to sentence construction and punctionations. In all, NT made 48 reading errors on the four texts that, taken together, consisted of 1153 words. In comparison, five control participants on average made 12.8 errors (S.D. = 3.6). The difference between NT and control subjects was highly significant (one-tailed p < 0.0005, Crawford & Howell’s test).

See Table 2 for a summary of NT’s reading errors. He did not produce any clear-cut semantic errors, but apart from this...
he made a varying number of other paraphasias (visual, derivational, regularisation, function word substitutions; as defined by Coltheart, 1987) and also a few unclassifiable errors (for instance, he mispronounced the e in the, every time the preceded a vowel), but notably no migration errors. There was no clearcut effect of word class in NT’s reading errors; verbs, nouns, function words and other words were misread.

2.3.5. Summary of reading assessment

Of the 162 single word NT read out loud in our tests, he made only three errors (1.9%). He also made three errors of 78 presented words and non-words in a lexical decision task, which was normal compared to a control group. Comparatively, when reading text, NT produced 48 errors on 1153 words (4.2%).

Unlike the case for NT’s single word reading, his error rate for text reading was significantly abnormal compared to the control group. Analyzing the errors he produced in text reading, we could not identify a pattern consistent with any of the central alexias. Also we observed no migration errors — the hallmark feature of attentional alexia — in reading.

Due to instrument error, reaction times in single word reading were not measured accurately. An effect of word length on response latency can therefore not be ruled out, but recorded RTs were below the level commonly reported in pure alexia, where reaction times are usually several seconds (e.g., Behrmann, Plaut, & Nelson, 1998; see also Section 4.3.2 and General Discussion). NT was capable of identifying single letters and digits correctly. In number reading, NT had problems in reading multi-digit numbers consisting of more than three digits. Although number reading skills are rarely reported in studies of alexic patients, it has been noted that patients with hemianopic alexia have difficulties reading multi-digit numbers (Zihl, 1995) and numbers with embedded zeroes (Kerkhoff, 1999), which perhaps corresponds to NT’s difficulties in this task.

Overall, NT’s pattern of reading disability seemed similar to hemianopic alexia: single word reading was accurate, whereas text reading was non-fluent and prone to error. However standard neurological examination (i.e., confrontation testing) had not revealed a visual field deficit. Neither had NT at any point during recovery complained of restricted vision. Still, we hypothesized that a more subtle deficit in visual function might underlie his reading problems. We therefore included a target detection task to explore NT’s attentional function in different parts of the visual field.

2.3.6. Target detection

NT was presented with a computerized visual reaction time test (Pedersen, 1993) in which double-lined target crosses appear in random positions on a background of static distractor single-line crosses. The target remains on screen until the subject responds. The subject is instructed to maintain central fixation and respond as quickly as possible when a target appears. The subject does not have to indicate where the target appears. If there is no response within 3 s the target disappears, and reaction time is recorded as 3000 ms. In the first trial, NT produced very high reaction times (RT) in the upper right quadrant, while RTs were relatively equal, and much lower, in the other quadrants of the visual field (see Table 3). In a second run of the test, we asked NT to pay special attention to this quadrant, while maintaining central fixation. This improved his mean RT, but not to the level observed in the other visual quadrants. When instructed to fixate above and to the right of the central fixation point, RT was still slower in the upper right quadrant. Surprisingly this test suggested a visual field deficit in the upper right quadrant, though with some remaining visual sensitivity, which was missed in the neurological examination. However the design of the test did not allow us to differentiate detection rate and reaction time, and we followed up with stricter psychophysical testing of NT’s visual field.

### Table 3

<table>
<thead>
<tr>
<th>Visual field</th>
<th>Central fixation</th>
<th>Central fixation</th>
<th>Fixation in upper right quadrant</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>standard (50 trials)</td>
<td>Attention to upper right quadrant (50 trials)</td>
<td>Fixation in upper right quadrant (50 trials)</td>
</tr>
<tr>
<td>Upper right</td>
<td>2200</td>
<td>1100</td>
<td>750</td>
</tr>
<tr>
<td>Lower right</td>
<td>310</td>
<td>330</td>
<td>290</td>
</tr>
<tr>
<td>Upper left</td>
<td>200</td>
<td>270</td>
<td>340</td>
</tr>
<tr>
<td>Lower left</td>
<td>330</td>
<td>330</td>
<td>340</td>
</tr>
</tbody>
</table>

Reaction times are given in ms.

3. Experimental testing: Phase 1

3.1. Perimetry

To investigate NT’s visual field in detail, we used a perimetry program developed by Kasten, West, Behrens-Baumaan, and Sabel (1998) and Kasten, Gothe, Brunsfahl, and Sabel (1999) to test for luminance sensitivity. Two hundred and fifty small white dots were flashed on a black background for 150 ms each, at random time points and positions across the visual field. Central fixation was controlled by a color detection task at the fixation cross (and monitored by video camera in one of the test runs). In the first perimetry conducted where testing was monocular, NT completely overlooked stimulation in the upper right quadrant, suggesting a simple field cut in this area (see Fig. 2a). However, since neurological examinations had not revealed any quadrantamotopsia, and the target detection task described above showed at least some preserved visual sensitivity, we hypothesized that the visual field deficit was partial rather than absolute: cerebral amblyopia. We tested this possibility by instructing NT to direct his attention covertly, still with central fixation, to the impaired quadrant. This dramatically changed his performance (see Fig. 2b). The observed threshold shift in luminance detection with attention suggested that visual input was merely attenuated in most of the quadrant.

To assess the reliability of this finding, we conducted an additional binocular perimetry, still instructing NT to attend covertly to the quadrant. NT again detected most of the stimuli in the quadrant. However stimuli in a horizontal area starting roughly 1 visual degree above and 2 degrees to the right of fixation remained undetected (see Fig. 2c). A similar finding was made in two binocular perimetry 9 months later (see Section 4.1), suggesting that a parafoveal scotoma accompanied the general amblyopia in the quadrant. Although stimuli in the same general area were missed, there was some variability from test to test: on the three binocular repetitions of the test, NT missed 8, 9, and 12 stimuli in the quadrant, respectively. Three locations were missed in all repetitions (marked out on Fig. 2c): two locations were missed twice, and misses at other locations were not replicated. Therefore, the exact location of the scotoma is not entirely clear. However it should be noted that in all repetitions of the test, NT missed stimuli only when they were more than 1 visual degree above the horizontal meridian. That is, stimulus points corresponding to the reading line were spared in all three runs of the test. In addition, stimuli in the central 1–2 visual degrees were always detected in the binocular perimetry, indicating foveal sparing.

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<td>330</td>
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</table>

Reaction times are given in ms.
In our experiment, five letters were flashed on an imaginary semi-circle (radius: 24 visual degrees) centred on fixation, such that two letters were displayed within the normal range: the span was 3.7 letters, compared to the control mean of 4.3 (S.D. = 0.43, one-tailed \( p < 0.01 \)). In the left side, NT's processing rates were (from top to bottom): 0.4, 3.0, 6.1, 2.3, and 1.2 letters/s. Note that NT's processing rate was especially high (6.1) at the middle position, which contrasts strongly with his poor processing rate (0.27) at this location in the right side. NT's total processing speed (the sum of the individual processing rates) was about equal for left and right side displays in the right side NT made only one, three, and seven correct reports, respectively, from the upper three positions. In comparison, control participants on average made 28.4 correct reports (S.D. = 10.6) from the three individual locations where they performed poorest: significantly better than NT's average of 3.7 correct reports (one-tailed \( p < 0.05 \)). Crawford & Howell's test). Only one control participant had less than 20 correct reports from a location (namely, 13 correct reports), and none showed anything like NT's near-zero performance in a large region of the visual field.

Overall, the whole report experiment showed that visual shape recognition was severely deficient (amblyopic) in the upper right quadrant and directly to the right of fixation. NT's visual processing speed and visual span in the intact (left side) parts of the visual field were relatively low, but not significantly reduced compared to the control group mean. His visual perception threshold was significantly elevated, but the impairment was moderate: a group of 12 healthy subjects of mean age 55 years, who were tested in a parallel study (Habekost & Rostrup, 2006), also had a perception threshold at 25 ms on average in this task.

To sum up, the investigations so far suggested that NT's reading difficulties were related to his visual field defect in the upper right quadrant. Although there was no large field cut, we hypothesized that the reading deficit was qualitatively similar to alexia after hemianopia, and wanted to pursue this idea by recording
4. Experimental testing: Phase 2

In April-June 2004 (18–20 months post insult) we conducted a follow-up study to NT’s reading problems. At this time NT reported some improvement in text reading, but compared to his premorbid reading abilities he still felt impaired, and was uncertain about pursuing a theoretical education. NT’s persisting difficulties were confirmed in our testing, which now included eye movement recordings during text reading as well as improved testing of single letter and single word recognition by more sensitive psychophysical investigations.

4.1. Re-assessment of visual field deficit

Two new rounds of perimetry confirmed the pattern observed the year before: NT was able to detect luminance in most of the upper quadrant, given covert attention to this area, but a small scotoma remained above and to the right of fixation (see Section 3.1 for details).

4.2. Re-assessment of single letter processing

Although NT’s recognition of single letters had previously been found normal in a paper and pencil test, it was possible that more subtle deficits were present. We therefore conducted a psychophysical examination of NT’s visual processing rate for single letters, and compared the results to an age- and education-matched control group (n = 5, mean age = 25 years, all were undergraduate university students). In this experiment we presented a single letter at the center of fixation, followed by a pattern mask displayed for 500 ms. Exposure durations in this task were very brief (below 60 ms), which should provide extra sensitive testing. As previously described, the results were analyzed using Bundesen’s TVA model. This time, since there was only one stimulus in the display, the capacity of visual short-term memory (for multiple objects) was not estimated, and only a single processing rate was computed. As before, the visual perception threshold was also computed. In the control group, the visual threshold was on average 10 ms (S.D. = 2 ms), and the mean processing rate was 95 letters/s (S.D. = 20 s⁻¹). Both NT’s visual threshold (13 ms) and processing rate (84 letters/s) were within normal range on this demanding task (Crawford & Howell’s test). The correlation between NT’s reaction times and word lengths (S.D. = 43 ms). NT’s deviation was significant at p = 0.003 (one-tailed Crawford & Howell’s test). The correlation between NT’s reaction times and word lengths was highly significant (Pearson’s r = 0.49; one-tailed p < 0.0005), mainly driven by his extra slow reading of words with nine or more letters (although note that NT’s mean reaction time for words with less than nine letters was still significantly longer than the controls’; p = 0.003, one-tailed Crawford & Howell’s test). By linear regression, we estimated the slope of NT’s word-length effect to be 57.3 ms per extra letter. Only one control participant had a significant word-length effect (Pearson’s r = 0.28, one-tailed p = 0.006), which was considerably smaller than NT’s (slope: 15.4 ms per extra letter). See Fig. 5 for reaction time plots and Table 4 for estimated word-length effects in NT and control participants. Table 4 also includes comparable data from a single word reading experiment by Leff et al. (2001).

4.3.2. Speed of single word reading

NT and controls were presented with a computerized reading task consisting of 80 words of varying lengths (2–12 letters) chosen from the PALPA battery (subtest 31). Words were presented centrally on a computer screen, and reaction times were measured with a voice-key. Responses terminated the presentation of the word. Errors were registered by the experimenter. NT made no error in this task, and neither did any of the controls, but NT’s reaction times were higher: on average they were 729 ms, compared to the control mean of 491 ms (S.D. = 43 ms). NT’s deviation was significant at p = 0.003 (one-tailed Crawford & Howell’s test). The correlation between NT’s reaction times and word lengths was highly significant (Pearson’s r = 0.49; one-tailed p < 0.0005), mainly driven by his extra slow reading of words with nine or more letters (although note that NT’s mean reaction time for words with less than nine letters was still significantly longer than the controls’; p = 0.003, one-tailed Crawford & Howell’s test). By linear regression, we estimated the slope of NT’s word-length effect to be 57.3 ms per extra letter. Only one control participant had a significant word-length effect (Pearson’s r = 0.28, one-tailed p = 0.006), which was considerably smaller than NT’s (slope: 15.4 ms per extra letter). See Fig. 5 for reaction time plots and Table 4 for estimated word-length effects in NT and control participants. Table 4 also includes comparable data from a single word reading experiment by Leff et al. (2001).

3 Due to errors of administration, one control subject received a different version of the lexical decision task, and two control subjects were tested using a different set of words in the speeded reading task. The data from these participants were not included in the analysis. They were replaced by other age- and education-matched controls.

4 The experiment only included one word with 12 letters, and NT’s very slow response to this word may be considered an outlier.
Comparable data from a single word reading experiment by Leff et al. (2001) of six normal subjects (N1–N6), four patients with hemianopic alexia (HA1–HA4), and one patient with pure alexia (PA1).

<table>
<thead>
<tr>
<th>Word-length effects</th>
<th>Slope</th>
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<tbody>
<tr>
<td>Estimated increase in reaction time (ms) per letter for NT and five controls</td>
<td></td>
</tr>
<tr>
<td>Control 1</td>
<td>−0.6</td>
</tr>
<tr>
<td>Control 2</td>
<td>14.4</td>
</tr>
<tr>
<td>Control 3</td>
<td>13.4</td>
</tr>
<tr>
<td>Control 4</td>
<td>5.4</td>
</tr>
<tr>
<td>Control 5</td>
<td>0.7</td>
</tr>
<tr>
<td>NT</td>
<td>57.7</td>
</tr>
</tbody>
</table>

Table 4

| NT's saccades were of this type, compared to 1.7% on average in the controls (S.D. = 3.3%). Actual "micro-saccades" (i.e., <5.5 visual degrees, scored as non-directional by the recording system) made up 12.4% of NT's eye movements, compared to an average of 6.0% (S.D. = 3.4%) in the control group. NT's deviation from the control group on this parameter did not reach significance (one-tailed p = 0.08, Crawford & Howell's test). Still, it is noteworthy that nearly 20% of NT's saccades, with accompanying fixations, were spent on non-progressive scanning of the text (vertical or micro-saccades), which is more than double the average amount used by control participants.

Overall, NT's eye movements during text reading were surprisingly close to normal. Contrary to prediction, his saccade length, fixation duration, and frequency of minor regressions were all within the normal range. Still, NT's value of 23% fell within the normal range, actually better than the control group. NT's deviation from the control group on this parameter did not reach significance (one-tailed p = 0.08, Crawford & Howell's test).

Another abnormality concerned the frequency of vertical saccades. 5.5% of NT's saccades were of this type, compared to 1.7% on average in the controls (S.D. = 1.3), a significant deviation (one-tailed p = 0.03, Crawford & Howell's test). These vertical saccades were typically small in NT, their mean amplitude was 0.88 visual degrees (S.D. = 0.33). Actual "micro-saccades" (i.e., <5.5 visual degrees, scored as non-directional by the recording system) made up 12.4% of NT's eye movements, compared to an average of 6.0% (S.D. = 3.4%) in the control group. NT's deviation from the control group on this parameter did not reach significance (one-tailed p = 0.08, Crawford & Howell's test). Still, it is noteworthy that nearly 20% of NT's saccades, with accompanying fixations, were spent on non-progressive scanning of the text (vertical or micro-saccades), which is more than double the average amount used by control participants.

Eye movements were measured using the Eyelink II system (SR Research Ltd., Canada), which uses two video cameras mounted on a headband to register pupil position in each eye, 250 or 500 times per second (depending on individual calibration). Head movements are corrected for by a third camera on the headband. After calibration, the Eyelink system supplies accurate information on the amplitude and direction of saccades, as well as the duration and position of fixations. Participants were seated in a dimly lit room about 50 cm from a computer screen. We calibrated eye position using a 9-point grid, and drift correction was performed before the onset of each page. The tests to be read were shown for 60 s per page. The participants were instructed to read out the text at their normal speed, while the experimenter noted any errors. The tests were chosen from Danish secondary school books, and were thus on a level that should be relatively easy for both NT and the control subjects. Each page consisted of 10–13 lines of about 60–70 characters, and recordings from two texts (12 pages) were made.

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Overall, NT's eye movements during text reading were surprisingly close to normal. Contrary to prediction, his saccade length, fixation duration, and frequency of minor regressions were all within the normal range. Still, it also reported being less hesitant to make eye movements into the affected field. Still his reading speed and error rate was significantly poorer than the control group. NT's deviation from the control group on this parameter did not reach significance (one-tailed p = 0.08, Crawford & Howell's test).

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Overall, NT's eye movements during text reading were surprisingly close to normal. Contrary to prediction, his saccade length, fixation duration, and frequency of minor regressions were all within the normal range. Still, it also reported being less hesitant to make eye movements into the affected field. Still his reading speed and error rate was significantly poorer than the control participants, corresponding to an increased number of large regressive sweeps and non-progressive (vertical or micro-saccades) saccades. It seems that NT's reading alternated between a normal oculo-motor pattern and frequent interruptions during reading. She was replaced by another control subject with the same age and level of education.

The data from one control subject was excluded because of excessive blinking (30% of all fixations; 10–20 times more than the other participants) and several interruptions during reading. She was replaced by another control subject with the same age and level of education.

Fig. 5. Voice-key recorded reaction times for single words of varying length. Mean reaction times are plotted as a function of word length. NT's mean reaction times are shown as filled circles connected by a bold line. The mean reaction times of five control participants are plotted as open circles, connected by thin lines for each participant.
was caused by this subtle sensory deficit, and represents a mild cerebral amblyopia. We propose that NT’s reading disability be modulated by spatial attention, and fits the characteristics of form and luminance in the upper right quadrant. The deficit could be considered a cause rather than an effect of NT’s reading disability. It is hard to rule this possibility out completely, but it seems unlikely for two reasons: First, NT’s eye movements were normal on the main parameters of saccade length and fixation duration. If oculo-motor control was compromised by the brain damage, performance would be much slower, and the effect would be much larger than what was observed.

On a speeded test of single word reading, NT was about 250 ms slower than the controls on average. The delay in reaction time was more pronounced with long words, and there was a significant word-length effect. NT’s performance on this test might suggest that his text reading difficulties were caused by a pure alexic deficit resulting in letter-by-letter reading. However, the increase in reaction time per letter was only about 50 ms, which is much smaller than what is typically observed in patients with pure alexia (see discussion below). Also, NT did not have hemianopic alexia, and need not be explained by a pure alexic deficit. Alternatively, the minor abnormalities of eye movements found during text reading (regressive sweeps, micro- and vertical saccades) were a cause rather than an effect of NT’s reading disability. It is hard to rule this possibility out completely, but it seems unlikely for two reasons: First, NT’s eye movements were normal on the main parameters of saccade length and fixation duration. If oculo-motor control was compromised by the brain damage,
one would expect a more general disturbance. Second, deficits in saccade planning and control are typically related to more anterior damage than NT’s, specifically the fronto eye fields and adjacent prefrontal areas (Pierrot-Deseilligny, Ploner, Muri, Gaymard, & Rivaud-Pechoux, 2002; Rivaud, Muri, Gaymard, Vemersch, & Pierrot-Deseilligny, 1994).

Instead, general deficits in attentional capacity could be relevant. In the initial neuropsychological assessment NT’s performance on the Symbol Digit Modalities Test was below average, indicating reduced psychomotor speed. However difficulties with scanning the test page might also have produced NT’s low score on this test. Also in the first phase of our investigation, NT’s perception threshold was moderately elevated in a whole report task, whereas his visual processing speed and visual span were in the lower normal range. One year later NT’s visual processing speed and perception threshold was found to be normal when we tested it using a demanding whole report task with very brief stimulation. This was the case even though alphabetic stimuli were used. Thus, whereas it cannot be ruled out that a slight reduction of attentional capacity contributed to NT’s reading disability in the first phase of our investigation, this cannot explain the persisting reading difficulties found in the second phase. Another possibility is a deficit in sustained attention, which might disturb NT’s reading of longer texts. NT was not tested explicitly for this, but our perimetry task was similar to the “watchkeeping” tests traditionally used to assess vigilance (Robertson & Manly, 1999): a monotonous detection task, where stimuli occur at irregular intervals over a period of many minutes. Except for one of the initial monocular perimetry, NT practically always detected stimuli outside the impaired quadrant, and even for this region he could concentrate sufficiently to detect most stimuli. The quadrant-amblyopia was accompanied by a parafoveal scotoma starting about 1–2 degrees above and to the right of fixation. The scotoma thus encroached on the central 5 degrees of vision, which makes it potentially relevant to reading ability (Zihl, 1995). However the scotoma did not affect the area directly to the right of fixation (the reading line), but was located at least 1 visual degree higher. During text reading this scotoma should only block out vision in one or two above fixation, and thus cannot explain NT’s alexia.

Given that amblyopia is likely to be a causal factor behind NT’s reading disability, what might be the actual mechanism? As demonstrated in our psychophysical experiments, NT’s visual processing in the upper right quadrant was very ineffective. The threshold for luminance detection was elevated, and letter recognition was severely compromised. However, apart from a small scotoma, information pick-up was not zero in the area. NT was occasionally successful at discriminating letter forms in the quadrant, and with spatial attention directed to the area he could detect small flashes of light. Still, NT’s perceptual deficit should impair access to more specific characteristics of the word at the next fixation. Continuous priming of word identity in parafoveal vision, up to 14–15 characters to the right of fixation (Rayner, 1998), is important to facilitate effective reading. Deficits in both luminance and form perception might impair this function. General reductions of luminance contrast can strongly affect reading speed (Legge, Rubin, & Luebker, 1987). If NT’s amblyopia effectively produced a similar reduction in the (perceived) contrast of text in the impaired region, impaired reading should result. Another possibility is that NT had a specific difficulty with form perception in the upper right quadrant in addition to his deficit in luminance perception. NT’s whole report performance is compatible with this hypothesis, but the observed deficit in letter discrimination could also be caused by a subjective reduction of luminance contrast in the region. Thus our results do not allow us to specify whether NT’s deficit in luminance perception can account for all his reading difficulties, or if impaired form perception was an independent factor behind his alexia.

Another relevant mechanism for NT’s alexia is that word-length information acquired parfoveally is used to compute where to look next (Rayner, 1998). Interestingly, NT’s rightward saccades into the affected field were of normal amplitude, at least in the second phase of our investigation. One explanation, originally suggested by DeLuca, Spinelli, and Zoccolotti (1996) for hemianopic alexia, is that NT adopted a strategy of making pre-determined saccades into the impaired field without regard for text characteristics. This should result in random “hit-or-miss” saccades, where fixations may or may not land at an optimal position to permit word identification. However, not even patients with a full hemianopia follow this random motor pattern (McDonald, Spitsyna, Shillcock, Wise, & Leff, 2006), but use at least some information from the text to guide eye movements. This information can either be derived “top-down” (i.e., context driven) or “bottom-up” (i.e., based on non-linguistic information from the parafoveal area such as word size). Given that NT’s text scanning was more normal than reported for hemianopic alexia (e.g., McDonald et al., 2006; Zihl, 1995) we suggest that he was able to derive more “bottom-up” information from the impaired field than these patients, due to his less severe visual deficit.

NT had a basal occipital lesion that evidently spared primary visual cortex, but affected extra- striate visual areas. This damage pattern is consistent with his amblyopic symptoms (Fable, 2002). In addition to the cortical damage, the lateral ventricle was severely dilated, which should affect the function of the posterior white matter tracts. Given the greatly enlarged size of the ventricle, it is in fact surprising that ND did not present more severe visual problems than amblyopia and a small scotoma, but it seems that most visual fibres were merely displaced rather than completely cut off. Concerning his oculo-motor behaviour, it is relevant to note that both the posterior thalamus and occipito-parietal areas were intact. Based on a large group study, Zihl (1995) suggested that spontaneous occulo-motor compensation for hemianopic alexia is related to sparing of these regions. NT’s marked improvement in the second phase of our investigation is consistent with this hypothesis.

An established treatment exists for hemianopic alexia: in systematic oculo-motor training, the patient is instructed to make longer saccades into the blind field (Kerkhoff et al., 1992; Zihl, 2000). It is possible that NT’s gradual improvement of reading ability was mediated by a spontaneous use of this strategy. Indeed, at the second testing phase he reported being less hesitant to make saccades into the affected part of the visual field. The recording of eye movements at that time confirmed that the mean
amplitude of saccades, as well as the fixation duration, was in the normal range. Still, NT made more non-progressive saccades (with accompanying fixations) than controls, and larger regres-
sions were also more frequent. Therefore our study suggests that while oculo-motor compensation can alleviate hemianoptic alexia, a normal length of saccades and fixations is no guarantee for unimpaired reading.

6. Conclusion

We have described the subtle reading disturbance of a patient with a left basal occipital lesion. Using a combination of alexia assessment, eye movement recordings, and psychophysical test-
ing of vision, we have shown how amblyopia in one quadrant can lead to a mild form of hemianoptic alexia. The study should alert clinicians to the possibility of this disturbance in patients with left posterior lesions but no obvious field cuts.

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References


ment of language processing in aphasia*. Hoe: Lawrence Elbaum Associates.


hagen: Center for Brain Injury.


ogy: Cognition, Memory, & Suppl., 76*, 1–25.

Reitan, R. M. (1971). *Scandicraft version 1.03*. Hillerød, Denmark: COG-
Nifo.


Perrot-Dessinguy, C., Ploner, C. J., Muri, R. M., Gaymard, B., & Pierrott-


