



Converging evidence for the role of occipital regions in orthographic processing: a case of developmental surface dyslexia

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Abstract

Recently, there have been several reports focusing on the neural basis for word recognition. Two different views have emerged: one emphasizing the role of the left angular gyrus in recognizing printed words, and the second view suggesting that visual word processing activates the left extrastriate cortex. This paper describes the case of EBON, a 14-year-old girl with an extensive early (most likely congenital) brain lesion in the left occipital lobe. She demonstrates a clear pattern of developmental surface dyslexia in that she is more successful at reading and spelling regular words than irregular words and makes frequent regularisation errors. Thus, EBON is the first case reported with the potential to establish converging evidence for the role of extrastriate regions in the left hemisphere in the acquisition of orthographic representations. © 2000 Elsevier Science Ltd. All rights reserved.

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1. Introduction

Considerable research has focused on the impact of phonological processing in reading acquisition, as the core deficit in developmental dyslexia, and as a remedial treatment of reading disability (for a review, see [56]). In brief, this research has produced an abundance of evidence which shows that problems in tasks that require the processing of phonological information are causally linked to word recognition difficulties in children and a persistent deficit in adults with reading disabilities [2,3,15,18,22,30,32,39, 40,47,57,58,65].

Although developmental dyslexia, in most cases, is associated with phonological processing deficits, there are several cases reported with a different pattern of word recognition difficulties [1,7,11,23,24]. This pattern corresponds to a deficit in the direct or orthographic

route for word decoding, that is, a preserved ability to read aloud regular words and nonwords (i.e., using grapheme–phoneme translation skills), but with an impairment in reading irregular words. This type of word decoding disability is referred to as developmental surface dyslexia. More recently, at least five different group studies have confirmed that some dyslexic children fail to develop adequate orthographic representations, despite their relative skill in reading regular words and nonwords [6,8,33,50,59]. Thus, some individuals with reading disabilities seem to have deficits in orthographic processing, that is, the ability to represent visual properties of words (including sequences and letter positions in words).

As a result of this research on developmental surface dyslexia, attention is now turning to two important questions. First, what are the basic cognitive deficits associated with developmental surface dyslexia? Second, what structures in the brain are involved in the development of orthographic representations? So far, research focusing on cognitive deficits associated with developmental surface dyslexia have not been success-

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ful, suggesting no relation between any form of visual capacities and orthographic word decoding skill [7,16,24]. Recently, however, Samuelsson et al. [50] have reported a double dissociation between the magnitude of perceptual priming (i.e., implicit memory for visual and auditory information) and subtypes of reading disability. Priming refers to the facilitation of task performance by previously studied materials without intentional recollection [61]. Samuelsson et al. [50] found that a surface subgroup of poor readers produced more auditory compared to visual priming, whereas a phonological subgroup of poor readers showed the opposite pattern. Their conclusion was that visual and auditory priming might be seen as indicators of the strength with which visual and auditory information is implicitly extracted. According to this conclusion, visual and auditory forms of perceptual priming might constitute basic cognitive functions associated with the ability to represent either orthographic features in written words or spectral and temporal features of spoken words (see also [9,51,52,61]). To be more specific, Tulving and Schacter [61] suggest that different forms of perceptual priming reflect activations of pre-semantic visual or auditory word recognition systems, and that the main prerequisite for the occurrence of perceptual priming is that implicit tests of retention match either visual or auditory representations acquired during a single study episode. Thus, it might be hypothesised that the primary problem associated with developmental surface dyslexia lies in the visual word recognition system, hence the inability to represent orthographic structures of whole words.

The claims about a visual word recognition system mediating visual priming as a potential cognitive correlate to developmental surface dyslexia are also in line with recent work by Petersen and colleagues about the neural basis for word recognition ([42–44]; see also [45,46] for similar findings using fMRI). They suggest that orthographic word decoding activates primary extrastriate areas in the left hemisphere (see also [55]). With regard to this direct route for word decoding, Carr and Posner [5] have suggested that a left-medial prestriate region (called “Prestriate visual word form system” or “Prestriate VWFS”) seems to be specific to the encoding of orthography (see also [13,35]). Thus, these PET and fMRI studies provide further support for the hypothesis that a deficit in the visual word recognition system is one potential underlying the cognitive deficit associated with developmental surface dyslexia.

Taken together, there is some evidence that a left-medial prestriate region is involved in orthographic word decoding, and that implicit memory (i.e., repetition priming) for visually presented words might be one potential cognitive factor associated with develop-

mental surface dyslexia. However, can anything be said about the relation between regions in the occipital lobe and implicit memory for visually presented words? Keane et al. [28] have reported a case (LH) with bilateral occipital-lobe lesions who displayed impaired implicit memory for visually presented words. This finding provides evidence that impaired visual priming can be linked to a damage in the occipital regions. In another study, Schacter et al. [53] found that visual priming (word-stem completion) was associated with a blood flow decrease in extrastriate occipital cortex (bilateral; see also [38]). In addition, Carlesimo et al. [4] found impaired visual priming in a case (AM) with an extensive lesion involving the infero-medial areas of the left temporal and occipital lobes. There were also strong indications that AM displayed a pattern of impaired word recognition referred to as acquired surface dyslexia, that is, she clearly exhibited intact phonological decoding skills, while her orthographic route for decoding was impaired. Moreover, studies of Alzheimer's disease patients, with relatively spared occipital circuits, have revealed normal visual priming on implicit tasks (e.g., identification of briefly presented words; [21]), as well as a preserved ability in word recognition [14]. These studies support the view that visual priming of words involves occipital areas and that there might be a relation between a pattern of surface dyslexia and impairment in visual priming.

The PET work by Petersen and his colleagues [42–44] was conducted on adult normal readers (see also [45,46]), and the cases reported by Keane et al. [28] and Carlesimo et al. [4] were both on elderly people with brain lesions acquired long after their acquisition of reading skills (i.e., with damages to the normal reading system). However, to study the involvement of left occipital regions in the acquisition of reading skills and to explore whether left prestriate regions might be associated with problems in the acquisition of orthographic decoding skills (i.e., developmental surface dyslexia), neural evidence from children is needed. The aim of the research reported here was to document a case of developmental surface dyslexia with an early brain lesion in the left occipital lobe, and thus, to provide some evidence that these brain structures might be crucial in the acquisition of orthographic representations. To my knowledge, there is no such case reported in the literature (see also [5]).

2. Case report

EBON is a right-handed girl and her age was 14 years at the time of the commencement of the study. EBON has no family history of neurological disease, and no incidence of dyslexia has been recorded in

other members of her family. EBON is a native Swedish speaker. The course of pregnancy was normal, with a normal full-term birth and a birthweight of 3.9 kg. She had good neonatal status, adequate weight gain and normal motor development. Her speech production and speech comprehension appeared to be normal. Thus, EBON's birth and early milestones were quite normal. No hearing problems were evident on testing when she started school. However, her vision has been tested on several occasions at early preschool age due mainly to squinting. This problem disappeared gradually and without the need of any corrections, and thus, she did not report any current or past difficulties with her eyes whilst reading. EBON started school at the age of 7 years and 4 months (which is normal in Sweden), and things went well with her early acquisition of reading and writing skills, and she received conventional reading instructions in primary school (i.e., a combination of whole language and phonics instructions). In fact, the first signs of problems with

reading and writing acquisition were reported by her teacher in the second grade (i.e., 8 years of age).

By the age of 8 years (10/6/91) EBON was admitted to Norrköping Hospital in Sweden following an epileptic seizure (which lasted for over 45 min) and standard EEG showed epilepticform activity in the left hemisphere, with the main EEG abnormalities located to occipital regions. Cerebral computer tomography (CT) at that time indicated a large cerebro-spinal fluid filled cavity comprising the medial and dorsal areas (Brodmann's area 17, 18, and 19) of the left occipital lobe (see Fig. 1). There were no other brain abnormalities. Thus, EBON's lesion was not diagnosed at birth, but it is likely that her occipital lesion is in fact congenital. This assumption is mainly based on the fact that the lesion is of significant size and magnitude, and EBON had no reported head trauma prior to its discovery that could have caused such damage. After this episode, EEG recordings have been normal when repeated after one and three years, and no recurring

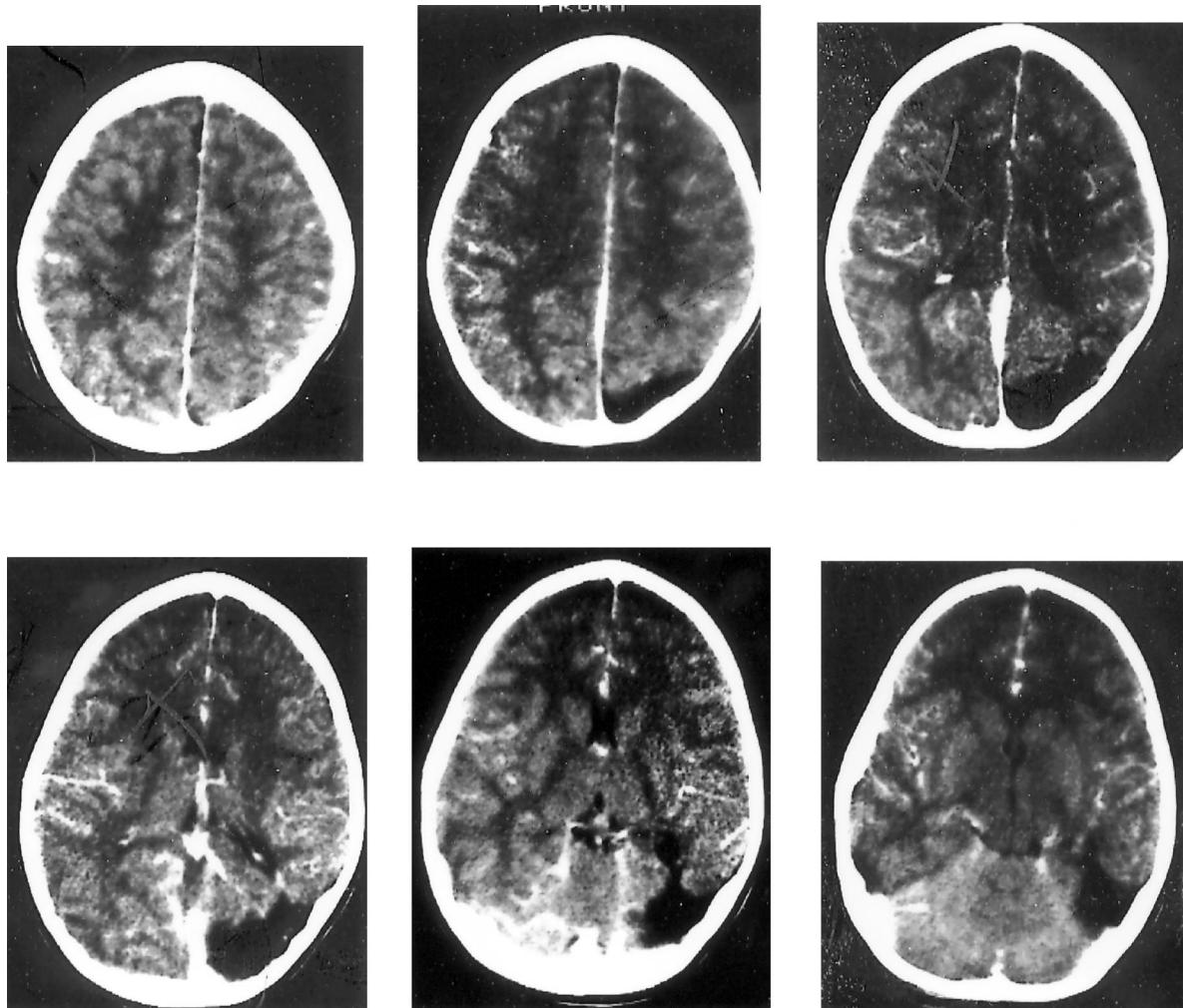


Fig. 1. From the upper left to the lower right: six layers of increasing depth illustrating a cerebrospinal fluid filled cavity comprising dorsal and medial regions (Brodmann's area 17, 18, and 19) of the left occipital lobe.

epileptic seizures have been reported. EBON received antiepileptic therapy (i.e., Valproate) shortly after this episode, and has been subjected to this therapy up to 14 years of age. With regard to this valproate monotherapy, it is important to note that there are several studies demonstrating none, or modest cognitive effects attributed to this treatment among adults [29,48], as well as among children and adolescents [19,34,63]. Finally, a psychologist also found that EBON had a visual field defect causing a loss of vision for stimuli falling to the upper right quadrant of the visual field (outside an angle size of 50°).

3. Some initial evidence for developmental surface dyslexia at 12 years of age

By the age of 12 years (i.e., grade 6), EBON was examined for her reading and writing difficulties at Ekhaga Rehabilitation Centre in Linköping. Administration of the Wechsler Intelligence Scale for Children revealed that EBON had a full scale IQ of 95, with a verbal IQ of 97 and a performance IQ of 93. At this time, her performance on a word decoding task was approximately 4 years behind her chronological age, and thus, she demonstrated a clear IQ–reading discrepancy. Furthermore, in two additional word decoding tasks, she made 11 errors out of 39 items (28% incorrect answers) in a test measuring orthographic skills. The task was to indicate which word (in a pair of words with identical phonological structure) was correctly spelled (see [37]). On the other hand, when the task was to read pairs of nonwords and to indicate which word sounded like a real word (see [37]), she made only two incorrect answers (or 5%). These test performances suggest that EBON has poor word decoding skills, and to be more specific, that her primary problem is with the acquisition of orthographic decoding skills. This pattern of word decoding deficits was further reinforced by a closer look at her spelling. In free writing (she was asked to write down a short story about a boy stealing candy), she misspelled 11 out of 15 irregular words (i.e., low degree of correspondence between sound and spelling), with all errors being phonologically correct. These misspelled words were highly frequent and word length varied between four and seven letters. No other words were misspelled (the complete story was 120 words in length). In addition, she was almost perfect in her reading of single nonwords (she read 75 out of 80 nonwords varying between two and nine letters in length), and she could easily repeat grammatically complex sentences up to 14 words in length (i.e., a short-term memory index). Finally, when she was asked to read single words aloud, EBON read high frequency short words using a direct or orthographic decoding strategy (cf. words

like milk, table, cat, hair; varying between three and five letters in length in Swedish), but as soon as the words became slightly less frequent and five letters or more in length, she started to use a phonological decoding strategy.

Thus, on the basis of these assessments, there are several findings suggesting that EBON does not perform like a “typical” developmental dyslexic (i.e., with clear deficits in phonological processing). Arguments against this include the fact that she reads nonwords without any problems, is successful in using phonological strategies in reading, has no problem in spelling regular words, and finally, performs almost perfectly on a sentence repetition task. Instead, the pattern of reading and writing difficulties displayed by EBON suggests that her main problem is acquiring or representing the orthographic features of written words, a failure which sometimes forces her to treat words as novel unfamiliar letter strings.

4. More systematic evidence for developmental surface dyslexia at 14 years of age

4.1. Statistics and comparison group

To determine the pattern of reading and writing difficulties displayed by EBON at 14 years of age (i.e., in grade 8), I decided to compare her performance on a number of different tests with a control group of 15 age-matched right-handed girls without any documented problems of reading and writing (cf. [6,7]). The control children have been given the same set of tests (excluding the reading comprehension test and the word chain tests described below) under the same testing conditions as EBON. All tests were performed in Swedish. Means and standard deviations derived from the control group were then used for a statistical evaluation. The procedure adopted was to convert the raw scores obtained for EBON into standard scores, and only z -scores less than -1.64 (or above 1.64) were accepted as significant. In the results presented below, there are some variables with a mean close to 100% in the control group (i.e., ceiling effects). These ceiling effects are all present in conditions where a pure phonological strategy is appropriate to solve the task, that is, in those cases where there is a perfect correspondence between phonemes and graphemes (i.e., the phonological choice task, nonword reading, and to read and spell regular words). In fact, it is almost impossible, at least in Swedish, to select regular words or pronounceable nonwords to prevent ceiling effects as long as the 14-year-old girls possess normal skills in phonological decoding. Thus, some contrasts between EBON and the control group are not meaningful when evaluated statistically. However, these ceiling effects do

not interfere with the purpose, which is to establish a specific pattern of reading and writing difficulties displayed by EBON.

4.2. Reading comprehension test

To provide empirical evidence that EBON has reading difficulties, a standardised reading comprehension test was administered (a test used by the International Association for the Evaluation of Educational Achievement, 1990). Norms are available for children in grade 8 (almost 4000 children), all with Swedish as their native language. This test is made up of 10 different text passages followed by 3–5 multiple choice questions. The type of text in each passage ranged from pure prose to facts embedded in tables and figures, and the number of words in each passage varied between 10–700 words. The task was to read one text passage at a time and to answer the questions linked to each passage. There was a total of 40 multiple choice questions and the testing time was restricted to 40 min.

As can be seen in Table 1, this test placed her below the 10th percentile for reading comprehension ($z = -1.83$, $P < 0.05$). Her problem was not primarily to comprehend the text, but to read all the passages within the time restriction (10 out of 40 questions at the end of the test were unanswered).

Table 1
Mean scores for EBON and controls (standard deviation in parentheses)

Variable	EBON	Controls	<i>z</i> -value
Reading comprehension	20	31 (6) ^a	-1.83*
Word chains (unrelated words)	15	40 (11) ^b	-2.27**
Word chains (related words)	22	51 (12) ^b	-2.41**
Orthographic choice (/40)	32	38.8 (1.0)	-6.80**
Orthographic choice RT	2.69	1.13 (0.27)	5.77**
Phonological choice (/40)	39	36.8 (2.2)	1.00
Phonological choice RT	3.87	2.71 (0.74)	-1.56
Nonword reading (/72)	69	69.7 (1.3)	-0.53
Nonword reading RT	2.10	2.0 (0.28)	0.36
Spoonerisms (/30)	27	28.0 (3.1)	-0.32
Spoonerisms RT	7.40	4.47 (3.2)	0.91
Word reading			
High frequency regular (/18)	18	18 (0)	-
High frequency irregular (/18)	13	17.5 (0.52)	-8.65**
Low frequency regular (/18)	16	17.0 (1.1)	-0.90
Low frequency irregular (/18)	11	14.6 (2.0)	-1.80*
Spelling			
High frequency regular (/8)	7	8 (0)	-
High frequency irregular (/8)	6	7.5 (0.74)	-2.02**
Low frequency regular (/8)	8	7.7 (0.59)	0.51
Low frequency irregular (/8)	1	5.0 (1.3)	-3.07**

^a Mean score based on 4000 age-matched controls.

^b Mean score based on 50 age-matched controls.

* $P < 0.05$.

** $P < 0.01$.

4.3. Word chain tests

Two word decoding tests were given in which EBON was asked to segment letter strings into their constituent words [27]. Three words were combined on each trial in both tests (all words were concrete nouns), but the words were semantically unrelated in the first test (e.g., "housecatfood"), whereas the words were semantically related in the second test (e.g., "horsepigcow"). There were 80 chains in each test and EBON was allowed to work with each test for only 3 min (i.e., it was impossible to complete all items). The theoretical rationale for these tests was to use the fact that mean performance differs between the two tests such that the number of words correctly segmented is, on average, 30% higher when there is a semantic relation between the words (51 vs 39 segmented words on average in a comparison group of 50 age-matched students). The hypothesis was that EBON should not display this difference between the tests due to her strong reliance on phonological decoding.

EBON's performance shows clear word decoding deficits, and the tests placed her below the 10th percentile (see Table 1). Significant differences were obtained for both semantically unrelated word chains ($z = -2.27$, $P < 0.05$), and semantically related chains ($z = -2.41$, $P < 0.01$). However, EBON completed more word chains when the words were semantically related (i.e., a difference between 22 and 15 correctly segmented word chains). One explanation for this difference might be that most of the words embedded in each chain were highly frequent words, which increased the possibility for EBON to use a direct route word decoding.

4.4. Orthographic and phonological choice tasks

To determine EBON's word decoding skills in more detail, two different tasks were adopted with similar procedures used by Olson et al. [37]. The first task was designed so that EBON would have to recognize words on a visual basis (using her orthographic representations). Two printed letter strings were arranged horizontally on the computer screen (Geneva font, 24-point), and her task was to decide which letter string was a correctly spelled word. There were no exception words with irregular spelling patterns among the real words (i.e., it was possible to read all words with rather simple grapheme–phoneme rules), and all non-words were pseudohomophones. EBON responded by pressing the right or left button on a response console. There were five practice and 40 experimental items in the test, and both accuracy and reaction times were recorded.

In the next task, also designed after Olson et al. [37], the rationale was to force EBON to read

using phonological decoding. In the test, pairs of nonwords (five practice and 40 experimental items) were presented on the computer screen and the task was to decide which word sounded exactly like a real word. The task was identical to the orthographic choice task in all other aspects.

The scores for EBON and means and standard deviations for the control group are shown in Table 1. As predicted, EBON was less accurate (32 vs 38.8), and slower (2.69 vs 1.18 s) than the control group on the orthographic choice task. On the other hand, her performance on the phonological choice task was even better than that of the control group (39 vs 36.8), and the difference in reaction time was not significant, even if she was somewhat slower in her phonological decisions compared to the control group (3.87 vs 2.71).

These results indicate that EBON is impaired relative to age-matched normal controls on tests measuring orthographic decoding skills, whereas her ability to decode via a sublexical procedure seems rather intact.

4.5. Nonword reading task

To further dissociate between orthographic and phonological decoding skills, a nonword reading task was constructed with two types of nonwords. The test consisted of 36 nonwords with no real word neighbors, and 36 nonwords with at least two or more orthographical neighbors (i.e., it should be possible to create at least two real words by changing one letter in the nonword). The nonwords were presented on a computer and they were printed in lower-case letters (Geneva font, 24-point). The more specific rationale for the two types of nonwords was to study whether EBON read nonwords by applying grapheme–phoneme correspondence rules only, or whether she could benefit from orthographically similar real word neighbors in her nonword reading. If her performance (time and accuracy) on nonword with real word neighbors did not differ from her performance on nonwords with no neighbors, then the test would provide further support for a deficit in representing orthographic information.

As expected from previous results, EBON had no problem with nonwords. She read 69/72 (96%) nonwords correctly, and there was no significant difference between EBON and the control group in speed (i.e., 2.1 vs 2.0 s). In addition, the difference between the time to read nonwords with and without orthographic neighbors was on average 0.01 s faster for EBON and 0.175 s ($SD=0.10$) faster in the control group ($z=-1.65$, $P < 0.05$). This effect is in line with a deficit in the representation and processing of orthographic information.

4.6. The spoonerism task

This task is assumed to measure phonological processing skills without contamination by reading and writing experience [41,62]. In the test, the experimenter read out two words with the instruction to exchange the first sound of the first word with the first sound of the second word. Thus, the task is to perform a transposition of the initial sounds of two words and then to repeat the words back aloud. Both speed and accuracy are recorded.

In this task, EBON scored 27 out of 30 (90%), which is comparable with the controls (28.0). The average time taken to spoonerise the words was somewhat longer for EBON (7.40 s) compared to the controls (4.47 s). However, this difference was not significant.

4.7. Reading aloud regular and irregular words

The single most important criterion used to establish developmental surface dyslexia is to determine whether the ability to read irregular words aloud correctly is selectively impaired relative to the ability to read aloud regular words [7,11]. One further criterion is that the performance on irregular word reading should be more vulnerable with a decrease in word frequency, that is, an interaction between type of word (regular vs irregular) and word frequency (high and low frequency words). Although Swedish has a rather shallow orthography (compared to English), there are many infrequent spelling/sound correspondences [31]. For example, the /ʃ/ sound in Swedish which can be spelled in the following ways; sj, skj, sch, ch, sh, g, k, si, or ti (i.e., the Swedish words for beige/beige/, cart/kärra/, regime/regim/, cortége/kortäge/ are examples of high and low frequency irregular words used in the test), and the /j/ sound which is spelled as j, g, gj, hj, lj, or dj (i.e., organ/orgel/, basket/korg/, in return/gengåld/, gallows/galge/ are additional examples of high and low frequency words deviating from standard letter-sound relationships). Furthermore, vowels such as /o/ and /e/ are quite randomly spelled /o/ or /å/ and /e/ or /ä/, respectively, and there are also many exceptions to the principles for doubling consonants in Swedish.

In this task, EBON was asked to read aloud 36 regular and 36 irregular words. The 72 words were presented one at a time and in random order. The pronunciations of the regular words were all predictable by grapheme-to-phoneme conversion rules, whereas the pronunciation of irregular words was not. The 36 words of each type were also divided into low and high frequency words. Regular and irregular words were matched for frequency, number of letters, morphemes, and syllables.

The words were presented on a computer screen and the letters were printed in lower-case, Geneva font, 24-

point. EBON read 33/36 (92%) regular words and 24/36 (67%) irregular words correctly (see Table 1). The errors were all phonologically correct. The number of irregular words read correctly differed significantly between EBON and the controls (27 vs 32; $z = -2.27$, $P < 0.01$). However, the detrimental effect of low frequency words was about the same for EBON and controls. When the irregular words were considered separately, the control subjects read both high and low-frequency words significantly better than EBON. Thus, EBON's ability to read irregular words was dramatically reduced, suggesting that EBON's main problem is to establish visual representations of words in her visual input lexicon.

4.8. Spelling test

To determine whether EBON employed the same strategy for reading and spelling, a spelling to dictation task was administered. The stimuli were 32 words selected from the list of 72 words used in the reading aloud task. There were 16 regular and 16 irregular words in the test, and each type of word was divided into high and low frequency words. The words were read out loud to EBON, one at a time in random order, and she was asked to write them down.

The results of the spelling test are shown in Table 1. EBON spelled 15/16 (94%) regular words and 7/16 (44%) irregular words correctly. The difference between EBON and the control group was statistically significant for irregular words, $z = -3.64$, $P < 0.01$. When the irregular words were considered separately, the control group spelled both high and low-frequency words significantly better than EBON. In addition, the errors made by EBON when spelling irregular words could all be classified as phonologically correct, that is, characterised by a phonetic spelling strategy (the same pattern of errors was made by the controls). Thus, EBON's spelling was typical of that which has been found to be associated with developmental surface dyslexia [12,24].

4.9. Implicit and explicit memory for visually and auditorily presented words

These two task procedures were designed to measure both explicit (i.e., episodic memory) and implicit (i.e., repetition priming) memory for visually and auditorily presented words (see [50] for more details). In the auditory modality, 32 target words and 16 distractor words were randomly assigned to four lists of 12 words in each with the distractor words placed at positions 1, 6, 7, and 12 in each list. All words were regular nouns (four to six letters in length) frequently used in written Swedish. The lists were recorded at normal conversational level by a male speaker with a speed of

one word every fifth second. The same number of target and distractor words were used in the visual modality. The words used in each modality were matched on the number of letters, syllables, and frequency.

At the study phase, EBON heard one list at a time and her task was to free recall (i.e., explicit memory) as much as possible after each list of 12 words (with a delay of 15 s). This procedure was repeated for all four lists in the auditory modality. EBON then completed a distractor task for approximately 5 min (the orthographic choice task described above). After the distraction task, EBON was given an auditory identification test in which all 32 target words from the study phase together with 16 new distractor words (these should reduce the risk that EBON becomes aware of the fact that some words are repeated) were masked by white noise. EBON was instructed to verbally repeat each word with the first word that came to mind (i.e., implicit memory). No references to the study phase were made, and the level of white noise was set to make sure that the proportion of non-primed completion varied between 25 and 30%.

In the visual modality, EBON was told to read one word at a time at her own pace (each word from a list of 12 words was presented only once) and that a new word would appear on the computer screen as soon as she pressed a button. She was also instructed to verbally recall as many words as possible after reading a list of 12 words (with 15 s delay). Thereafter, EBON was engaged in a distractor test for 5 min (the phonological choice task). Implicit memory for visually presented words was measured by a stem completion test with the instruction to complete two-letter stems verbally (these were presented on the computer screen) by the first word that came to mind.

To evaluate the priming effects, baseline performances (i.e., completion rate for the stem completion test and the auditory identification test without having been exposed to the target words previously) for each test were selected from 40 children randomly selected from three different schools (ranging from 10 to 14 years of age). Baseline performances were 0.08 for the word stem completion test, and 0.31 for the auditory identification test.

Table 2
Mean proportions of explicit memory for visually and auditorily presented words and visual and auditory priming (i.e., the relative advantage for previously visually and auditorily presented words)

Presentation mode	Explicit memory		Priming	
	EBON	Controls	EBON	Controls
Visually presented words	0.35	0.36 (0.09)	0.07	0.15 (0.06)
Auditorily presented words	0.42	0.37 (0.06)	0.16	0.16 (0.07)

The magnitudes of visual and auditory priming for EBON and the control group are displayed in Table 2. EBON exhibited more auditory priming compared with visual priming (16% vs 7%). Furthermore, the amount of auditory priming resembled that of the control group (16% for EBON and 16% for the control group), whereas visual priming displayed by EBON was lower compared with the control group (7% for EBON and 15% for the control group). Although the difference in visual priming was not significant ($z = -1.35$, $P > 0.05$), this pattern of visual and auditory priming effects might indicate that implicit memory for visually presented words is one potential cause to deficits in representing orthographic structures of words as a clue to their identification (cf. [50]). In fact, the range of visual priming obtained in the control group (proportions varying between 0.08 and 0.33) clearly indicate that there is no overlap in the distribution of priming effects between EBON and the control group.

EBON's recall performance (i.e., explicit memory) was similar to that of the control group. In fact, her recall performance for visually presented words was almost identical with the mean of the control group (35% vs 36%), and her recall for auditorily presented words was slightly better than for the control group (42% vs 37%; $z = 0.83$, $P > 0.05$).

4.10. Wisconsin card sorting test (WCST)

This test is normally used to examine executive functions and cognitive flexibility. The more specific rationale for me to include this test was to make sure that EBON's strong reliance on sublexical procedures for both reading and spelling is not due to the possibility that she is cognitively arrested in one particular learning strategy. Thus, it is suggested that if EBON has no difficulties in shifting her strategy along the three different dimensions (i.e., form, number, and colour), then it might be, at least to some extent, possible to exclude that her reliance on phonological processing is not primarily caused by an inability to shift strategy in a cognitive task in general. Her performance in this task was normal for her age and she completed all six categories.

4.11. Visual analysis tasks

Because EBON has had some problems with squinting as a child and with a visual field defect causing a loss of vision for stimuli falling to the right quadrant of the visual field (outside an angle size of 50°), I decided to test her on two different visual tasks. It should be noted, however, that EBON had no specific problems with those parts of WISC measuring visuo-spatial abilities at 12 years of age.

In the first visual task, EBON was presented with a string of six letters, one at a time [26]. In half of the items the strings were composed of identical letters, whereas the remaining strings contained one deviating letter located at various positions. A total number of 64 strings were presented, and the task was to detect strings with odd letters by pressing a "yes" or "no" button when a uniform set of letters was presented. Except for somewhat slower reaction times (0.90 s for EBON and 0.81 for the control group), no problems could be detected in this task. Her accuracy was 92%.

In the second visual task, visual perception was measured using overlapping stimuli (BORB; [49]). In these tests, IT was presented with overlapping letters (two or three letters), geometrical forms (two or three different forms), and line drawing of objects (only two overlapping objects). In these five different tasks, EBON's performance was perfect and she ran through all items easily and without hesitation.

5. General conclusions

The results of this study replicate previous findings that some developmental dyslexics exhibit significant problems with the acquisition of orthographic representations [1,6–8,11,23,24,33,50,59]. Assessment of EBON indicated that her phonological decoding skills were appropriate for her chronological age in most conditions, which strongly supports that her main problem is to acquire orthographic representations and to progress to the orthographic stage of reading [20,54]. This pattern of reading and writing difficulties was present at 12 years of age, and replicated two years later.

The findings also provide unique and convergent support for the localization of a visual word recognition system to the left extrastriate cortex (cf. [42–46]). Most of the neuropsychological evidence concerning the neural substrate of orthographic decoding skills comes either from normal adult readers observed with different functional neuroimaging technology, or from behavioural studies of patients with acquired surface dyslexia. To my knowledge, EBON is the first case reported with the potential to establish a relation between deficits in the acquisition of orthographic representations or developmental surface dyslexia and a lesion in the extrastriate regions in the left hemisphere. Therefore, any hypothesis about developmental issues and the involvement of prestriate regions on reading and writing performances by developing readers have not yet been directly examined. Clearly, if an extrastriate visual word recognition system constitutes a specifically orthographic coding mechanism in adult readers, then it might also be crucial for the acquisition of orthographic skills.

There are several PET and fMRI experiments car-

ried out with the purpose of identifying brain regions related to the processing of single printed words. In general, these studies have provided support for two different neurologically based theories of reading single words. One view is the classical Geschwind–Wernicke model which assumes that the left angular gyrus is strongly involved in recognizing printed words (cf. [25,36]), whereas the other view, proposed by Petersen and colleagues, suggests that visual word processing mainly activates the left medial extrastriate cortex [42–44]; see also [45,46]. With regard to these two views, EBON's reading and writing profile tends to provide support for the proposal of a visual word recognition system in the left extrastriate cortex. From a developmental perspective, EBON's results also support the hypothesis that extrastriate regions might be involved in the acquisition of orthographic representations, and consequently, even if she has successfully reached the alphabetic stage in learning to read (i.e., her phonological skills were intact at 12 and 14 years of age) she clearly failed to progress to the orthographic stage. Thus, EBON's pattern of reading and writing difficulties clearly provides support for dual-route models of reading, that is, a distinction between lexical and sublexical procedures for reading single words aloud [12]. EBON not only replicates previous cases suggesting that phonological decoding skills can be developed independently of orthographic skills (cf. [1,6,7]), the relation between her deficits in orthographic processing and the localization of her brain lesion also suggests a close association between a particular cognitive operation with a particular brain region (cf. [43,46]).

With respect to underlying cognitive deficits, I speculate that implicit memory or repetition priming for visually presented words might constitute one potential basic cognitive deficit associated with developmental surface dyslexia (cf. [50]). Within memory research, there are several findings supporting the existence of perceptual representation systems (PRS), and that different forms of priming reflect activation of pre-semantic visual and auditory word recognition systems [51,52,61]. Theoretically, this research is in line with the view proposed by Petersen et al. [42–44] and Pugh et al. [45,46] that a visual word recognition system represents orthographic features of written words. Anatomically, the occipital lobes are assumed to be involved in repetition priming of visually presented words [4,28,53], and thus, there is at least some overlap between the brain regions assumed to be activated in reading printed words and implicit memory for words. Although EBON exhibited more auditory than visual priming, this effect did not reach significance, and thus, the present findings are suggestive rather than conclusive and clearly need to be replicated. Nevertheless, further research using this perceptual priming paradigm has the potential to gain further

insights into cognitive deficits underlying subtypes of reading difficulties.

Although the present study provides developmental evidence for an association between left extrastriate damage and an impairment of the visual word recognition system, it does not follow that left extrastriate lesions must always be present in cases of surface dyslexia. Instead, surface dyslexia seems to occur with different patterns of impairment to orthographic processing, possibly mediated by a wide range of lesion sites. First of all, models for lexical processing are composed of a number of subsystems which may not be functioning in a pattern of surface dyslexia. For example, Coltheart and Funnell [10] identified seven different subsystems with the potential to mediate different locus of impairment to orthographic processing. Second, most cases with acquired lesions resulting in surface dyslexia have been described with quite extensive lesions, including temporal lobe structures as well as frontal, parietal and occipital structures [64].

This work is based on two underlying assumptions that should be made more explicit. One assumption is that an early/congenital brain lesion and its particular localization is associated with an arrest at the alphabetic–phonological stage in reading and spelling. However, even if this assumption seems to be valid, I can not fully ignore the possibility that EBON's problems in orthographic processing might also be the result of a more linguistically based inability of using patterns of higher order orthographic regularities. That is, it is never possible to completely rule out the possibility that EBON might have displayed the same pattern of word decoding deficits without her brain lesion. In more general terms, we have to live with the fact that it is impossible to disentangle the developmental outcome in cases of dyslexic children with a particular brain lesion from cases with similar developmental outcomes without any obvious signs of neurological, sensory, physical, emotional, educational or cultural influences. It might be argued, however, that the possibility that EBON's pattern of reading difficulties might have been present even without her brain lesion is somewhat reduced by the fact that (a) previous findings suggest a priori a relation between her particular lesion and deficits in several tasks of interest, and (b) her specific pattern of reading disability clearly deviates from the phenotypic performance profile of children with reading disabilities (cf. [17,58]).

Finally, one further assumption underlying the findings presented here is that the developmental outcome of a congenital lesion to some extent resembles the association between isolated cortical regions and components of word decoding skills observed with different functional neuroimaging technology using normal adult readers. However, the question of structural plasticity and potential redistribution of functions

following early brain lesions should be taken into consideration. Two general principles seem to resemble what we know about language recovery in general after brain lesions acquired in infancy (cf. [60]). First, the prognosis for recovery of language is better for lesions acquired at a young age, and second, the size of the lesion also seems to be related to recovery. That is, an extensive brain lesion involving classical language regions acquired after approximately 10 years of age is generally associated with a modest recovery of language functions. With regard to reading, however, the onset of reading acquisition is relatively delayed and a more systematic exposure to written language in school does not normally occur until 7 years of age. Things went well with regard to reading and spelling until EBON reached the age of 8 years, and thus, it might be argued that the pressure to relocate functions specific to reading acquisition was even more delayed due to her skills in phonological processing.

In summary, the results show that left occipital regions might be involved in the acquisition of orthographic representations which is assumed to be one important prerequisite to progress to the final orthographic stage of reading. I also speculate that poor implicit memory, mediated by a pre-semantic visual word recognition system, might be one potential underlying cognitive deficit associated with developmental surface dyslexia, that is, an impairment in extracting physical features about the visual form and structure embedded in written language.

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