Neglect dysgraphia

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SUMMARY A phonological dysgraphic syndrome is documented in a left handed man with a right-hemisphere lesion. His spelling was significantly affected by word length but neither word frequency nor the orthographical irregularity or word class proved to be relevant variables. Words were spelled equally efficiently forwards as backwards. A clear gradient of letter errors was shown to exist with letters on the left being mis-spelled more often than letters on the right of a word, irrespective of word length. These findings are discussed in terms of current models of spelling and their relevance to theories of unilateral neglect.

Unilateral neglect refers to the failure to respond to or report information from the side contralateral to a patient's lesion. As early as 1914, Poppelreuter¹ described patients with unilateral visual neglect and further case studies were also reported by Scheller and Seidemann.² The syndrome may vary in degree from a total neglect of stimuli to one side of the body (usually the left) patients acting as if both personal and extrapersonal space did not exist,³ to milder cases where the disorder is only observed under special testing conditions. Neglect is not necessarily restricted to the visual modality but can be observed with tactual and auditory stimuli.⁴⁵

Neuropsychological theories of neglect tend to fall into one of two groups. The first may be termed the "stimulus-bound" theories whereby neglect is seen as related to faulty input; as either a disturbance, interruption, extinction or lack of synthesis of sensory information conveyed to one hemisphere (for example⁶⁻⁸). The second group of theories evoke some kind of central internal representation such as body schema or a spatial model which underlies the organisation of perceptions and actions. Unilateral impairment of performance is thus held to occur at a higher level of organisation than the sensorimotor level and directs the search for information (for example³⁹¹⁰). Recent work by Bisiach and his colleagues has expanded and strengthened this latter view of neglect. They described two patients who failed to report details from the left side of the

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Received 25 May 1983. Accepted 20 June 1983 Piazza del Duomo in Milan, when asked to imagine it from the front of the Cathedral on the opposite side of the square and also when required to reverse the perspective and describe the square from the front doors of the Cathedral.¹¹ It is difficult to see how an input-output mechanism can account for such one-sided omissions from verbal descriptions of imagined scenes. Indeed they argue from their experimental data that two assumptions are required to explain the syndrome of unilateral neglect; the first necessitates the view that "the nervous system cannot conceivably be programmed to gather information without the recruitment of an inner model..." and the second assumes that "any unmonitored failure in performing the programme implies a breakdown of the model itself".^{12, p.614}

The interaction of visual neglect on the reading process was noted by Gilliatt and Pratt13 and documented more extensively by Kinsbourne and Warrington.¹⁴ Their six patients with right hemisphere lesions made paralexic errors limited to the beginning of words both in normal word reading and tachistoscopic presentation to the intact right visual field. In their patients "neglect dyslexia" was not associated with dysphasia or dysgraphia and was interpreted as an abnormality in the distribution of attention affecting both words and other visual forms. Although it has been noted that unilateral neglect can distort script,¹⁵ to our knowledge, no comparable investigation of the interaction of neglect and spelling (oral or written) has been described.

Our understanding of those dysgraphias which are independent of faulty spatial or praxic skills derives primarily from recent investigations of acquired dyslexic syndromes. It is now well-established that there are at least two reading processes: one which extracts word-meaning directly (semantic route) and one in which the phonological analysis of a word precedes processing for meaning (phonological route).16-18 Recently, individual case studies of dysgraphic patients have provided evidence for two dissociable processes in spelling which parallel these alexic syndromes. The first is illustrated by Beauvois and Dérouesné's¹⁹ patient RG who showed no impairment of the phoneme-grapheme conversion rules, being able to spell regular words and nonsense syllables quite efficiently, but whose spelling was severely affected by the orthographical irregularity and ambiguity of a word. The converse is illustrated by Shallice's²⁰ patient PR who was unable to spell nonsense words or syllables, and yet, with the exception of function words, was able to spell both regular and irregular words at a superior level. Shallice²⁰ concludes that these two cases of phonological and orthographical/lexical dysgraphic syndromes provide strong support for spelling syndromes analagous to phonological and direct route reading.

The present case-study of a left-handed patient with a right-hemisphere lesion and phonological dysgraphia provides further support for lexical (orthographic) processes in spelling. In addition, the patient showed a predominance of errors at the beginning of words in oral spelling, which we will argue is a unilateral neglect phenomenon operating on a cognitive skill.

Case report

ORF (born 19 August 1932), a left-handed gentleman farmer, sustained his first stroke in November 1981, a year prior to the present investigation. He was admitted as an emergency to the National Hospital, Queen Square (20 November 1981) for investigation of a left hemiplegia of recent onset and increasing confusion and drowsiness. The CT scan showed a haematoma of mixed density indicative of repeated haemorrhage, most dense in the right parietal cortex and extending deeply into the capsular region. There was considerable surrounding oedema and compression of the right ventricle with associated midline shift (fig 1). Angiography at this time indicated contralateral displacement of the anterior cerebral artery and distortion and stretching of the temporo-parietal branches of the middle cerebral artery; there was medial displacement of the Sylvian point and the thalamostriate group of vessels and in addition there was evidence of segmental spasm of the supraclinoid area of the internal carotid. A right craniotomy to evacuate the haematoma was performed on 20 November 1981. The patient made a remarkably good recovery such that two months later he was discharged. His left homonymous hemianopia was reduced to an inferior quadratic field defect and his dense left hemiplegia had improved so that he could perform most activities of daily living, being able to walk unaided, without a stick. He continued to have loss of sensation to light touch, vibration and position sense and weakness of all arm and leg movements on the left side of his body. There was some ataxia. He had an expressive aphasia, characterised by literal paraphasic errors, anomia and marked difficulties with repetition (conduction aphasia). A detailed analysis of his speech production at this time has been reported.²¹ He obtained a verbal IQ of 99 and a nonverbal IQ of 92 on the WAIS (2 December 1981). In addition he showed features of phonological dyslexia together with a mild "neglect dyslexia". There was no other unequivocal evidence of unilateral visual neglect. At this stage he was severely dysgraphic.

One month later he was readmitted to hospital (4 February 1982) for investigation of a rapid deterioration in his motor functions and expressive speech. The CT scan again showed a large right parietal intracerebral haematoma extending into the cortex with surrounding oedema which improved considerably over the next month (16 March 1982) (fig 2). Angiography showed vascular displacements compatible with a posterior temporal haematoma. On this occasion he was treated conservatively with dexamethasone. His recovery was much slower than previously despite extensive rehabilitation efforts. His intellectual level had deteriorated somewhat since the recovery from his first stroke such that he obtained a verbal IQ of 91 and a performance IQ of 89 on the WAIS (16 March 1982). However his pattern of scores was identical on the two occasions. He had a mild "fluent" dysphasia characterised by hesitations and circumlocutory phrases. There was

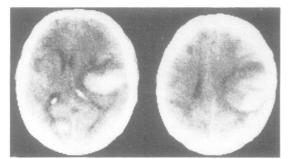


Fig 1 Pre-operative CT scan, 20 November 1981

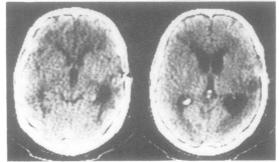


Fig 2 Post-operative CT scan, 16 March 1982

a marked nominal dysphasia, but there were now only minor errors in his speech production; these occurred when he attempted to repeat low frequency multisyllabic words. Once again there was evidence of mild phonological dyslexia with occasional paralexic errors affecting the beginning of words. He continued to be severely dysgraphic.

On neurological examination, four months after his second discharge from hospital (15 September 1982), there was a dense left hemianopia and slight left facial weakness. The upper limbs showed a moderate contracture of the left hand and an increase in tone in the remainder of his arm. There was no movement of the arm except at the shoulder. Tone was again increased in the lower limb on the left and there was distal weakness. Joint position sense was absent on the left and there was no two-point discrimination in the fingers of that hand. Light touch was subjectively impaired down the whole of the left side of the body and face.

At this time he obtained a verbal IO of 101 on the WAIS (15 September 1982) and his age-scaled scores were as follows: Arithmetic 12, Similarities 13, Digit Span 2, Voc abulary 13. His recognition memory for words was close to the 50th percentile for his age (40/50).²² His ability to name objects from their description was adequate (13/ 15)²³ although there was evidence of mild nominal difficulties on a graded-difficulty naming test (18/30).²⁴. His speech production was much improved and he was able to repeat 27/30 low frequency polysyllabic words. He obtained a reading IQ equivalent of 111 on the NART²⁵ but made a few paralexic errors affecting the beginning of words suggesting that his reading was slightly below his optimal level. His reading of four-letter nonsense words was impaired (3/10), but his spelling level was very poor indeed and he was only able to score at just above the 10th percentile on a graded difficulty test.²⁶ His pattern of errors was extremely patchy such that he succeeded in spelling words such as "scissors" and "journey" but made errors on simpler words such as "folk" and "skirt". Preliminary observations suggested that his errors were predominantly affecting the letters at the beginning of words. ORF himself described attempting to spell like reading off an image in which the letters on the right side were clearer than those on the left. It was these observations that led to the following, more detailed analysis of his spelling skills.

EXPERIMENTAL INVESTIGATIONS

Procedures

ORF was unable to use his preferred left hand to write and attempts to use his right hand were extremely slow and hard to decipher. Consequently only his oral spelling was tested. Individual words were repeated if requested and homophones were presented in a sentence context. No time limits were imposed. Although ORF did not have difficulties in naming letters of the alphabet when they were presented singly in a random order, occasional hesitations were noted when spelling words aloud. He was therefore given a visual display which he could use if he wished to facilitate naming the target letter. He would often spontaneously spell the stimulus word backwards, making it clear to the examiner that this had been his intention. If he did not specify in which direction he was spelling a word,

Table 1 Effect of word frequency, regularity and length

$\begin{array}{c ccccccccccccccccccccccccccccccccccc$		3-letter		4-letter		5-letter	
A or AA		R		R		R	N = 20
		(N = 40)	(N = 40)	(N = 30)	(N = 30)	(N = 30)	(N = 50)
		75%	80%	30%	60%	20%	20%
Low frequency (N = 80) 60% 65% 30% 30% 20% 104	Low freque $(N = 80)$	ency 60%	65%	30%	30%	20%	10%

I.... Irregular words

the examiner always asked him to indicate the direction, regardless of whether he had, in fact, spelled the word forwards or backwards.

All the following investigations were carried out on the same day, nine months after his second stroke (25 November 1982).

Test I: Effect of word regularity, frequency and length

Specially constructed lists were devised in order to investigate the effect of word regularity, length and frequency. The test stimuli consisted of a pool of 120 A or AA words and 80 words varying in frequency between 1 and 50 per million.²⁷ There were 80 three-letter, 60 four-letter and 60 five-letter words of which half in each group were orthographically irregular or ambiguous. The words were presented in a pseudo-random order. The percentage correct for each of these variables is shown in table I.

There was a highly significant effect of word length ($\chi^2 = 35.509$; p < 0.001), but the effects of word regularity ($\chi^2 = 0.726$) and word frequency ($\chi^2 = 1.305$) were not significant. In addition to scoring each word correct or incorrect, an error analysis taking letter position into account was computed. Summing across frequency and word regularity, the percentage correct for each letter position is given in fig 3.

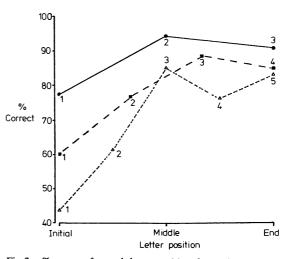


Fig 3 % correct for each letter position for each word length (\bigcirc \bigcirc 3-letter words, \bigcirc \bigcirc 4-letter words, \triangle \bigcirc \frown \frown 5-letter words)

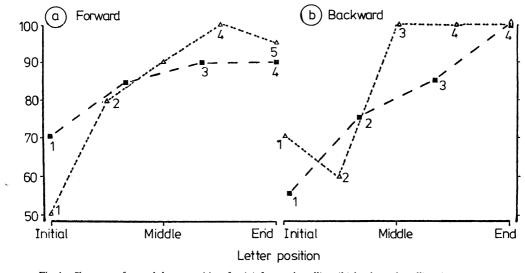


Fig 4 % correct for each letter position for (a) forward spelling (b) backward spelling (\blacksquare — 4-letter words, \triangle — — — — \triangle 5-letter words)

For all word lengths there was a highly significant trend on the Jonckheere test²⁸ for errors to decrease towards the end (3-letter words z = 2.505, p < 0.02; 4-letter words z =3.518, p < 0.001; 5-letter words z = 5.025, p < 0.002). Further analysis showed that there was a significant difference between the first and second letter position for the three-, four- and five-letter words. (Sign test p < 0.01, p <0.02, p < 0.05 respectively). In the case of the four- and five-letter words there was also a significant difference between the second and third letter positions (p < 0.05, p <0.001 respectively).

Test II: Letter order: forwards and backwards spelling

It could be argued that ORF's increased error rate for letters at the beginning of words in all these lists might be due to his tendency to spell words backwards and his shortened auditory span of attention meant that letters at the beginning of words were recalled last. In order to control for this possibility, two matched lists of 20 four- and 20 five-letter A or AA words were constructed. One list of four- and five-letter words was required to be spelled forwards and the other lists were required to be spelled backwards. Each pair of lists at each word length was matched for phonemic ambiguity and for letter frequency. This latter constraint was necessary in order to control for ORF's difficulty when naming particular letters. An ABBA design was used.

There was no significant difference in the number of words correctly spelled in the two conditions; he spelled 15 of the 40 words correctly in the forwards condition and 11 out of 40 words in the backwards condition ($\chi^2 = 0.513$). As before, the number of errors for each letter position for each of the four conditions was noted and the results are shown in fig 4.

There was a significant trend on the Jonckheere test²⁸ for

errors to decrease from the beginning to the end of a word in three of the four conditions, (4-letter words spelled forwards, z = 1.616, p < 0.2; 5-letter words spelled forwards, z = 4.027, p < 0.0002; 4-letter words spelled backwards, z = 3.401, p < 0.001; 5-letter words spelled backwards, z = 3.954, p < 0.0002).

Tiest III: Nonsense syllable spelling

His ability to convert a spoken letter sound (for instance a as in cat) into its alphabet name (A as in tame) was extremely poor (14/25). Similarly he had great difficulty spelling aloud both two-letter nonsense words (VC or CV) and three-letter nonsense words (CVC). The number of words and letters correct regardless of position for each condition is given in table 2. This result may in part be confounded by his very poor auditory span. However, this argument cannot explain his inability to sound out words slowly, for example "k ... ae ... t' as in cat. He totally failed to segment five three-phoneme words into their constituent sounds in spite of his ability to understand the requirements of the task and to recall the target word. In contrast to his poor performance at the individual phoneme level, he was able to provide rhymes to seven out of 10 real words.

Та	bl	le	2
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Nonsense words	No. correct	Letters correct	
$2-\text{letter} \\ (N = 10)$	0/10	9/20	
$\begin{array}{l} 3-\text{letter} \\ (N = 10) \end{array}$	0/10	12/30	

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Test IV: Effect of word class

There were no significant differences in his ability to spell four-letter high frequency concrete nouns (6/10), abstract nouns (4/10) or function words (7/10) (not significant on Fisher's exact probability test).

Discussion

This left-handed man with a right-hemisphere lesion provided us with a unique opportunity to investigate a rare dysgraphic syndrome. ORF's ability to spell real words contrasted markedly with his almost total inability to spell nonsense syllables and identify single sounds. His spelling was not affected by the orthographical irregularity or ambiguity of a word but there was an effect of word length and frequency. However, the most striking feature of his performance related to his tendency to make errors at the beginning rather than the end of words, the reverse of the normal pattern in children.29 This pattern of errors occurred in all word lists attempted, irrespective of word length or whether the words were spelled in a forwards or backwards direction. These observations are relevant both to current models of spelling and to theories of unilateral neglect.

Although ORF was spelling at a much lower overall level than Shallice's²⁰ patient PR, it is evident that his residual spelling is being mediated by lexical processes. They both appear to be spelling from an "inner screen" which has direct semantic access. This was perhaps particularly clear in ORF who was able to spell equally efficiently backwards and forwards. In both patients, there was a marked discrepancy between their ability to spell real words and their inability to achieve simple phonemegrapheme correspondences, the cardinal feature of a phonological dysgraphic and thus providing support for the two-route model of spelling.³⁰³¹ Moreover neither patient was able to segment a spoken word into its constituent phonemes. The two patients differ insofar as ORF did not show any features of auditory/verbal agnosia, nor did he have any greater difficulty in spelling function words in comparison to other word classes. This suggests that a selective deficit with spelling function words (a feature also described by Bub and Kertesz³²) is not a necessary component of this syndrome but may be an instance of a category-specific spelling deficit.

The most striking feature of ORF's performance was his tendency to make more errors at the beginning of words. Letter position effects occurred equally for 3-, 4- and 5-letter words. There appears to be evidence of some kind of gradient from left to right, rather than a stepwise function. This gradient appears to be more apparent across the beginning than the end letters of a word. Clearly these observations are in line with Kinsbourne's³³ notion of gradients of attention. But our data suggest that this abnormal distribution of attention is likely to arise from a central mechanism rather than faulty sensory input.

Kinsbourne and Warrington¹⁴ discussed "neglect dyslexia" in similar terms, namely an abnormal distribution of attention. The paralexic errors of their patients almost invariably affected the beginnings of words, the error responses being alternative real words. Moreover, it has now been observed that this type of error occurs as frequently with short as with long words and that the target word-length is usually maintained. (A corpus of such errors from a single case study, JAF, is included in the appendix). These latter observations also suggest that neglect dyslexia, like neglect dysgraphia, arises from a faulty distribution of attention to the central representation of a word.

Whereas the arguments against neglect dyslexia being interpreted in terms of faulty sensory input must still be regarded as tentative, the observation of the neglect dysgraphic syndrome described in the present case study, as with cases of "neglect imagery"¹¹ compels an interpretation in terms of a faulty distribution of attention in cognitive processing.

We are grateful to Dr P Rudge for permission to investigate ORF, a patient under his care and to report our findings. We also thank Drs R McCarthy and T Shallice for their help and advice in the preparation of this manuscript.

Appendix

The reading skills of JAF a 63-year-old secretary who had sustained bilateral occipital lobe infarctions (for further details see³⁴) were assessed on July 1981. She obtained an IQ equivalent of 102 on the Schonell Graded Word Reading Test.³⁵ A *complete* corpus of her reading errors from (1) the Schonell GWRT, (2) 24 concrete nouns, (3) 24 abstract nouns, (4) 52 function words, (5) 20 words written in script + 20 control words (Total 240) is given below:

Stimulus	Response	Stimulus	Response
to	unto	hence	sense
it	lit	though	bough
if	life	beggar	vinegar
for	car	hammer	summer
red	tape	decent	recent
cap	gap	finger	ginger
her	mother	bright	right
are	fare	canary	granary
bun	gun	enigma	stigma
	-	-	-

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his	this	anyone	someone
tar	for	anybody	nobody
let	wet	example	sample
rum	drum	nothing	bathing
lag	wag	service	rice
sip	zip	genuine	guarantee
cog	dog	rescind	insend*
tug	rug	homonym	synonym
hen	when	windmill	sawmill
lip	zip	inferior	interior
rid	kid	appeared	disappeared
unto	into	smoulder	shoulder
seige	besiege	nourished	flourished
clock	block	terrestrial	celestial
train	rain	preliminary	preparatory
gnome	home	institution	investitution*
light	fighting	susceptible	acceptable
aside	hide	preferential	deferential
*These were the only two neologisms recorded			

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