THE RECOGNITION OF WORDS IN PURE ALEXIA AND HEMIANOPIC ALEXIA: A NEUROPSYCHOLOGICAL STUDY OF 6 PATIENTS

By

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Al mio Piccolo

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”Discovery consists of seeing what everybody has seen and thinking what nobody has thought.” Albert Szent-Gyrgyi

Chapter 1

General introduction

1.1 Aim of the thesis

This thesis investigates the ability to read. Reading is a central process between vision and language that involves the ability to recognize letter strings in a rapid and parallel fashion. The earliest writing systems date back to 4000-3000 BC when Sumerian cuneiform writing was developed. Reading is also a skill and a difficult one since several years of learning are required in order to master it fully.

Reading can be selectively impaired after brain damage and this results in an acquired dyslexic disorder. Acquired dyslexia is characterized by a difficulty in reading that is not caused by a deficit in intelligence or by a low educational level. It differs from developmental dyslexia because the latter is a specific disorder in learning to read, while acquired dyslexia occurs in previously competent readers.

The present study investigated the performance of some patients with an acquired reading disorder that impaired the early stages of the visual processing of words, a disorder called peripheral dyslexia (Shallice and Warrington, 1980). There was the opportunity to study 6 patients with a deficit in that early visual stage of word processing, namely 5 patients with pure alexia (2 Italian and 3 English patients) and 1 patient with hemianopic alexia. In a classical model of word reading such as the 3-route model (Morton and Patterson, 1980) the visual components (the orthographic analysis and the visual input lexicon) are not carefully defined and it is not clear which stages we are likely to carry out when we see a word. The study of patients with an impairment in the visual components of word reading can help to identify those stages, especially if the deficit varies across them.

In pure alexia patients have lost the ability to recognize words in a rapid and automatic way. Some theories claim that this syndrome arises from a single type of functional origin (Farah and Wallace, 1991; Sekuler and Behrmann, 1996; Behrmann, Nelson and Sekuler, 1998), the most common version being of a generalised problem of lower-level visual processing which leads through cascade-type consequences to difficulties at higher word-form levels (Behrmann, Plaut and Nelson, 1998). Other theories claim that the deficit is at a higher-level, in the visual word-form unit which is the orthographic representation of the word (Warrington and Shallice, 1980; Warrington and Langdon, 2002).

However, if the deficit in pure alexia can vary qualitatively (Patterson and Kay, 1982; Price and Humphreys, 1992), therefore in some (maybe the purest) cases it will be possible to identify the visual subcomponents of the word recognition process that have been selectively damaged. From this viewpoint, we studied whether pure alexia is caused from a neuropsychological point of view by one type of deficit, common to all patients (Behrmann, Plaut and Nelson, 1998) or by qualitatively different forms of deficit (Price and Humphreys, 1992; Patterson and Kay, 1982). The Italian pure alexic patients have been extensively studied with several tasks; in particular we focused on two different abilities, namely the ability to process single letters and the ability to conjoin them together into syllables and words. As regards the second capacity, most current computational models of visual word recognition tend to underestimate the role of sublexical processing (Coltheart, Rastle, Perry and Ziegler, 2001; Plaut, McClelland, Seidenberg and Patterson, 1996; McClelland and Rumelhart, 1981) although there is evidence indicating an important role in word recognition (Grainger and Whitney, 2004; Carreiras, Vergara and Barber (in press); Dehaene, Cohen, Sigman and Vinckier, (2005)). We investigated whether there is a sublexical level that mediates the letter and the lexical levels and if this integrative process of letters into units can be selectively damaged in patients with an acquired reading disorder.

The results of this study provide support to the hypothesis that pure alexia can result from an impairment at different levels within the visual components of the word recognition process. This leads to the hypothesis that in models of normal reading (such as the 3-route model by Morton and Patterson, 1980) we do not have an ”orthographic analysis” component directly connected to a ”visual input component” (whose role is not properly defined), but a hierarchical series of more carefully defined units, which integrate the information from lower-level units. This would occur in the word-form system (Warrington and Shallice, 1980; Shallice, Warrington and McCarthy, 1983) which would have different units hierarchically organized. This hypothesis is consistent with the model proposed by Dehaene et al. (2005) of word recognition inspired by neurophysiological models of object recognition, where local combination detectors become sensitive to increasingly larger fragments of words.

Another group of English patients with pure alexia has been studied. The aim was firstly to test our hypothesis of the different patterns of functional deficits and secondly to assess whether the same methodology used in Italian could be replicated in another language such as English. Italian has a transparent orthography with consistent grapheme-phoneme correspondences, while English has a deep orthography containing many inconsistencies and complexities. This results in differences in the acquisition of the language as well as in the reading speed, Italian being easier to learn and quicker to read (Paulesu, McCrory, Fazio, Menoncello, Brunswick, Cappa, Cotelli, Cossu, Corte, Lorusso, Pesenti, Gallagher, Perani, Price, Frith and Frith, 2000). For this reason it is likely that some differences between Italian and English pure alexic patients are also caused by the differences in the two orthographies. More specifically, some discrepancies between the two groups can derive from two sources. First, from the relation between orthography and phonology that is ambiguous in English and highly regular in Italian; second, from the use of a serial reading strategy which can lead to activate multiple alternative words. For instance, a serial procedure that is applied to read an English word such as *island* can make one believe that the word is for example *islam*: a patient can make some conduites d'approche, or cannot recognize the word at all, or can recognize it immediately but generally reading is likely to proceed less efficiently. This type of errors where the pronunciation of a letter has not been over-ruled by the presence of a subsequent letter has been frequently reported in literature by patients using a serial procedure in English (Patterson and Kay, 1982; Newcombe and Marshall, 1985). Differently, a serial procedure applied in Italian can rely on the consistent relation between orthography and phonology and therefore reading can proceed more efficiently. This would be true independently from the type of letter-by-letter reading strategy (with letter-names or letter-sounds) used by the patient. We can hypothesize that if we had Italian and English pure alexic patients with the same degree of severity and a set of words which are very similar in terms of orthography and meaning across the two orthographies (e.g. naso-nose; zebra-zebra; treno-train; rosa-rose; distanza-distance; natura-nature; pigiama-pajamas), we expected that English are slower and maybe more error prone than Italian patients.

Finally, we studied an unusual case of hemianopic alexia which shows some symptoms of neglect dyslexia and a confabulatory perception of letters in word reading. AT is a rare case of acquired dyslexia where words are perceived systematically longer. This case study is interesting because it might shed light on the influence that visual field defects have on perceptual phenomena like cross-over (Marshall and Halligan, 1989), completion (Warrington, 1962; 1965) and confabulations (Chatterjee, 1995).

1.2 Methodology

In the present work the cognitive neuropsychological approach has been used to study reading. Reading represents an illustrative example of how cognitive neuropsychology principles have been applied successfully to comprehend complex cognitive functions (Marshall and Newcombe, 1966; 1973).

By studying people with selective disorders of cognition such as acquired dyslexia we can make inferences about the nature and the structure of that cognitive function, in this case reading. In fact reading involves different components, such as visual, semantic and phonological that are handled by different cognitive modules. Brain damage may result in the selective loss of certain modules, while leaving others intact. The contrast between intact and damaged aspects of reading may differ between individuals, producing different patterns of reading disorder. The pattern of single and double dissociations (Shallice, 1988) brings to the identification of the components necessary to read. In this way we can built models of normal reading functioning by studying patients with brain lesions.

Our experimental investigation used standardized tests as well as tasks developed ad hoc and run on a PC with the E-prime program. Most of the tasks have been translated into English so that Italian and English patients could be compared on the same type of material, with the same experimental procedures and also with the same criteria.

The performance of the Italian patients has been compared to a group of control subjects matched for age and educational level. As regards the English patients, the key contrast is the direct comparison between them and the

Italian group.

1.3 Outline of the thesis

The thesis is organized in the following way. In Chapter 2 models of word recognition are described starting historically from the first accounts of the 19th century. On the basis of models of normal reading functioning, the description of the different types of acquired dyslexia is provided. The cross-linguistic study of reading in English and Italian follows. Finally a brief description of the computational models of reading is taken into account to explore the implications of specific assumptions on the processes underlying reading. This allows to investigate more carefully how written words are coded.

In Chapter 3 we get to the heart of the theoretical study of acquired dyslexia, by discussing hemianopic alexia and pure alexia. A description of each syndrome is given. In particular as regards pure alexia, the different types of theoretical accounts have been discussed together with the neuroanatomical localization studies that have been carried out since the 19th century. The brain imaging research on pure alexia (and on the ability to read more generally) is also described.

Chapter 4 concerns the experimental study carried out on 2 Italian pure alexic patients, FC and LDS. The performance of FC and LDS has been investigated with many tasks mainly focusing on letter processing and orthographic integration. The aim was to investigate whether pure alexia can be caused by different functional deficits.

In Chapter 5 additional cases of English patients with pure alexia have been described and studied. This opportunity allowed me to explore whether the results achieved could be confirmed by another group of patients. Moreover the study of pure alexia in another language allows one to consider whether variations of performance between Italian and English patients could be caused by the differences in the orthographies.

Chapter 6 concerns the study of an interesting case of hemianopic alexia. AT is slow but accurate on word reading and makes consistent errors only with briefly presented words, adding extra letters at the end of the words. The rarity of the symptoms simultaneously present in AT (neglect dyslexia, a kind of completion phenomena, confabulation) has attracted much attention.

Finally in Chapter 7 the conclusions of the present investigation are drawn.

Chapter 2

Reading and dyslexia

2.1 Models of word reading

Reading is a visual process that allows us to recognize a letter string rapidly and effortlessly. This skill takes many years to learn and comprises different functional components which operate at least partially independently one from another. We learn to identify squiggles on a printed page as different letters of the alphabet, to built an abstract visual representation that is invariant to size, position and shape and then to match it to both semantic and phonological representations.

Visual word recognition is a remarkable process. In an unfamiliar script like Arabic or Chinese the characters appear meaningless and differences between characters are less evident. By contrast in our own language words leap out at us within a fraction of a second, carrying with them their meaning and sound. This is a rapid and effortless perceptual mechanism that acts ignoring large differences in visual form (e.g. A and a) while considering to small details such as distinction between 'e' and 'c'.

Models of word recognition are attempts to characterize the mental processes that allow a reader to identify, comprehend and pronounce written words. They try to decompose the act of word recognition into its component parts and describe the working of those parts. The components range from visual processing at an early stage to semantic and phonological processing at later stages.

2.1.1 Wernicke - Lichtheim

The first models of written language date back to 19th century. At that time the idea that language could be divided into different sub-systems in different anatomical areas was spreading with neurologists such as Dax (1836), Broca (1861) and Wernicke (1874). The first model of spoken and written language was developed by Wernicke (1874) in the attempt to capture and predict the wide variety of aphasic language deficits (an English translation of his work was published by De Bleser and Luzzatti in 1989). Wernicke (1874) assumed the existence of 4 language centres, a centre for the production of spoken words, a centre for writing words, the third one for the comprehension of words and the last one for optic word images. The latter comprised optic (graphemic) memory images of words and its lesion was supposed to result in cortical alexia. The sensory and motor representations of words were considered to be largely independent from each other, but together they constituted the word concept.

This elementary model was further elaborated by Lichtheim in 1885 and after a series of changes, the final version was made by Wernicke after 1886. Wernicke adopted Grashey's (1885) view that the unit of reading and writing was the letter rather than the word (it is noteworthy that on their model the processing unit of spoken language was the word rather than the individual letter). The center for writing was dependent on reading letters, so that there could be no agraphia without alexia and 2 forms of acquired dyslexia were predicted: pure alexia and alexia with agraphia.

2.1.2 Déjerine

The interest in the selective disorder of reading commenced in the late 19th century with the fundamental contribution of Déjerine (1891, 1892). Déjerine (1891) first reported a patient with alexia and agraphia in absence of other language impairment as the result of a stroke shown at autopsy in the left angular gyrus. A year later Déjerine (1892) presented the now famous case of Oscar C to the Biological Society in Paris, recounting how the highly educated textile merchant found himself completely unable to read after suffering a cerebrovascular accident in 1887. The patient was able to write, thus Déjerine called this disorder ”alexia without agraphia” or ”pure alexia”. His lesion involved the left occipital lobe and the splenium of the corpus callosum. Shortly before death, the patient developed agraphia; at autopsy a more recent infarction of the angular gyrus was noted.

Déjerine (1892) argued that a specialized system develops in skilled readers in the left angular gyrus representing the stored visual images of words. These images in the angular gyrus are necessary both to read as well as to write. On this account, the patient Oscar presented a disconnection of the angular gyrus, which stores the representation of visual words, from both visual cortices (see the Chapter 3 for more detailed description).

As conclusion, on the basis of his observations, Déjerine (1892) claimed that word and letter recognition occurs in the angular gyrus, which he held to be the image store of written words used for reading as well as for writing. However, although the explanation of pure alexia in terms of a disconnection syndrome remains credible to this day (see Chapter 3, Section 3.3.7), the hypothesis of a visual memory center used for reading and writing words did not obtain late support. Cases of patients who were able to read but not to write, namely with acquired agraphia, have been described in literature. For instance, patient JC (Bub and Kertesz, 1982) with deep agraphia showed an adequate comprehension of simple written sentences while writing of dictated sentences and spontaneous writing of sentences were both impaired. Patient PR (Shallice, 1981a) with phonological agraphia was totally unable to write nonwords, while his ability to read them was much superior.

2.1.3 Marshall and Newcombe

After the seminal contribution of Déjerine (1891, 1892), the study of acquired reading disorders languished for decades, during which the relatively few investigations that were reported focused primarily on the anatomic underpinnings of the disorders. The study of acquired dyslexia was revitalized by the elegant and detailed investigations by Marshall and Newcombe (1966, 1973). In their seminal work they launched the cognitive neuropsychological approach to alexia.

The interpretation of reading disorders given by classical aphasiology and by Déjerine (1982) could not explain even qualitatively aspects of normal and dyslexic behavior such as frequency effects, the performance with nonlexical stimuli or visual errors (Luzzatti, 2003)

Marshall and Newcombe (1973) demonstrated that the pattern of reading errors exhibited by dyslexic subjects is a kind of Ariadne's thread which leads to the identification of distinctly different and reproducible types of reading deficits. For the first time Marshall and Newcombe (1973) investigated systematically the patterns of errors made by dyslexic patients, relating them to those made by normal subjects in specific conditions (tachistoscopic presentation) and by children learning to read in order to built the functional architecture of our normal reading ability.

Marshall and Newcombe's (1973) paper produced a dramatic change. Within the neurological tradition little attention was paid to psycholinguistic properties of words such as imageability, frequency, part of speech, word length. Marshall and Newcombe applied the cognitive neuropsychological methods to the study of reading. As it is rare that patients are totally unable to read, they

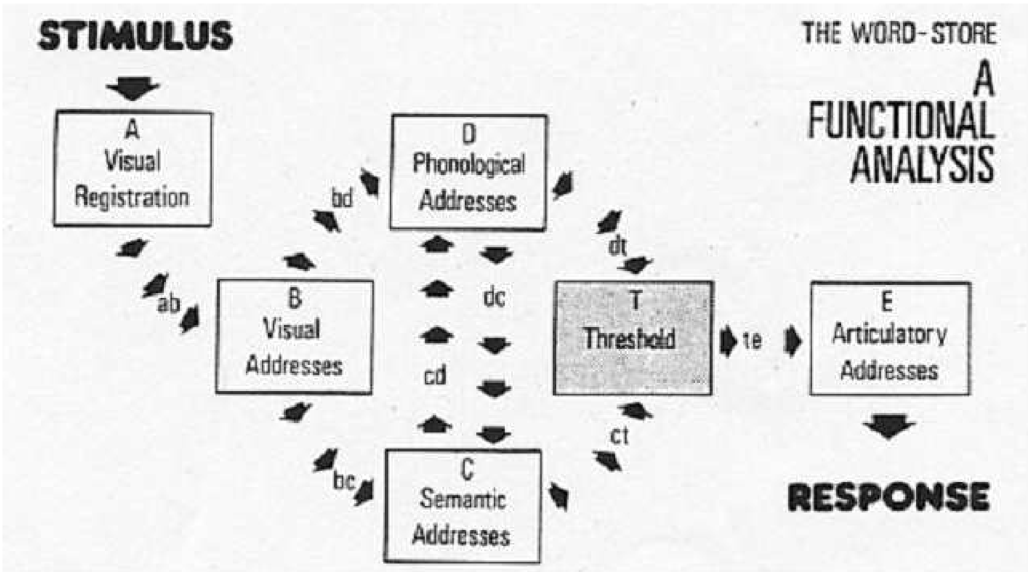


Figure 2.1: Marshall and Newcombe's (1973) two-route model of the reading process.

analyzed the nature of errors that may be highly consistent in a patient and yet surprisingly different across cases. Errors were studied from a qualitative point of view together with the orthographic variables that may affect reading ability (e.g. length, regularity and grammatical class) in order to identify the impaired component of the reading system.

On the bases of their data, Marshall and Newcombe (1973) concluded that the meaning of written words could be accessed by two distinct and separate procedures. The first was a ”direct” route which transmits the visual (orthographic) information to the semantic system. The second one was an ”indirect” route, assumed to map the visual (orthographic) output to a phonological system which in turn activates the semantic system (see Fig. 2.1).

This second route, hereafter termed ”print-to sound”, contains a set of rules for performing grapheme-phoneme correspondences on subcomponents of the input, thereby converting written spelling to sounds. This was their famous two-route model which has provided the conceptual framework that has motivated most subsequent studies of acquired dyslexias.

2.1.4 The 3-route model: Morton and Patterson (1980)

Contemporary reading models have two main differences compared with Déjerine's model (Luzzatti, 2003). First, the visual representation of letters and words, identified by Déjerine in the angular gyrus, has been substituted with two separate and independent orthographic representations for reading and for writing (orthographic input and output lexicons). Second, on the basis of the observations made by Marshall and Newcombe (1966, 1973), two distinct reading pathways, phonological and semantic, have been demonstrated.

Some researchers have indicated three routes to pronunciation of written stimuli, adding a third route to the semantic and non-lexical phonological routes because in direct dyslexia (see Paragraph 2.2) irregular words could be read and neither of the other two routes could have achieved that. Patients like WLP (Schwartz, Saffran and Marin, 1980) showed the pattern of good reading of irregular words in the face of apparent lack of comprehension of the same words.

Morton and Patterson (1980) developed a model (a version of the *logogen* model re-drawn (see Paragraph 2.4.1 in this Chapter)) which became very popular, where in addition to an assembled nonlexical route to phonology, there are 2 lexical routes: the semantically mediated route and a nonsemantic lexical route based on connections from a word's orthography to its phonology that bypasses semantics. This is also called the "direct" lexical route.

More specifically (see Fig. 2.2), the first route that is lexical-semantic involves the activation of a stored entry in the visual word form system and the subsequent access to semantic information and ultimately activation of the

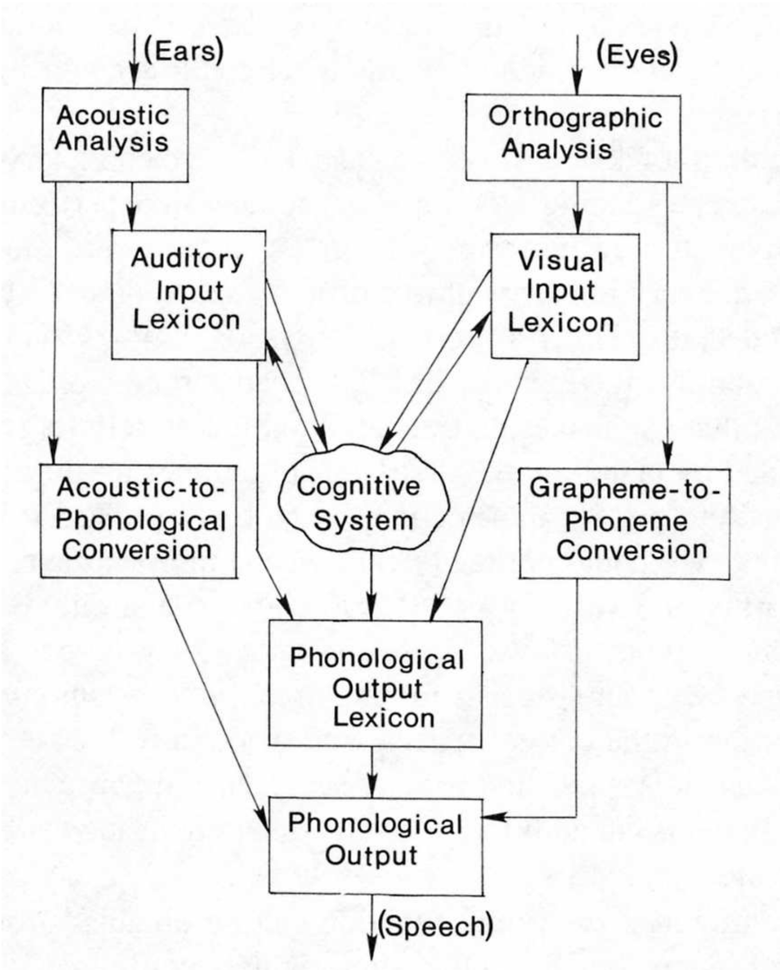


Figure 2.2: Morton and Patterson's (1980) model of the stages of processing single words presented auditorily or visually.

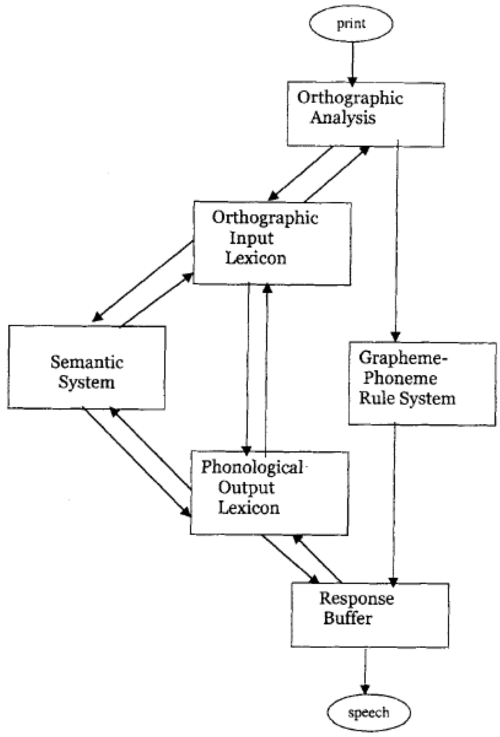


Figure 2.3: Basic architecture of the "dual-route cascaded model" of visual word recognition and reading aloud (from Coltheart, Rastle, Perry and Ziegler, (2001)). As shown, the model consists of three routes.

stored sound of the word at the level of the phonologic output lexicon. The second route involves the nonlexical grapheme-to-phoneme conversion with no access to any stored information about words. It allows reading of regular words and nonwords. The third lexical-nonsemantic route is assumed to involve the activation of the visual word form system and the phonologic output lexicon without any intervening activation of semantic information.

A variation of the 3-route model is the called strangely "dual route-model" (Coltheart, Curtis, Atkins and Haller, 1993), where in actual fact written language can be processed along 3 routes: the lexical semantic route, the lexical nonsemantic route and the grapheme-to-phoneme route (see Fig. 2.3 from Coltheart, Rastle, Perry and Ziegler, (2001)).

2.1.5 The extended 2-route model: Shallice, Warrington and McCarthy (1983)

Data supporting a putative separate ”direct” lexical mechanism met some critics. Shallice, Warrington and McCarthy (1983) argued that the performance of some patients such as WLP (Schwartz et al., 1980) could be accounted for in another way. Instead of hypothesizing a third, direct route, they argued that reading could be mediated by a phonological mechanism that is also lexical. On their account, the visual word form system parses letter strings into multiple units of different size, thus for example a letter string may be parsed into its constituent graphemes, consonant clusters, subsyllabic units, syllables and even morphemes (see Fig. 2.4). Information is transmitted to the phonological system where stored print-to-sound correspondences for these multiple units may be utilized to generate the appropriate phonology.

On this multiple-levels approach two reading routes are available to read: a sematic route and a ”broad” phonological route. In the latter, different levels of units are used, particularly subsyllabic units and also syllables (Shallice and McCarthy, 1985). Sub-syllabic or, at most, syllabic correspondences would be sufficient for the reading of the mildly irregular words, but very irregular words would require morphemic correspondences (Shallice, 1988).

Shallice and colleagues (Shallice et al., 1983, Shallice and McCarthy, 1985; Shallice, 1988) claimed that in neurological disease, correspondences based on larger units are more vulnerable than those based on smaller units; a progressive disease, such as that suffered by EM (Shallice and Warrington, 1980) and WLP (Schwartz et al., 1980) would increasingly restrict the range of correspondences available to the patient.

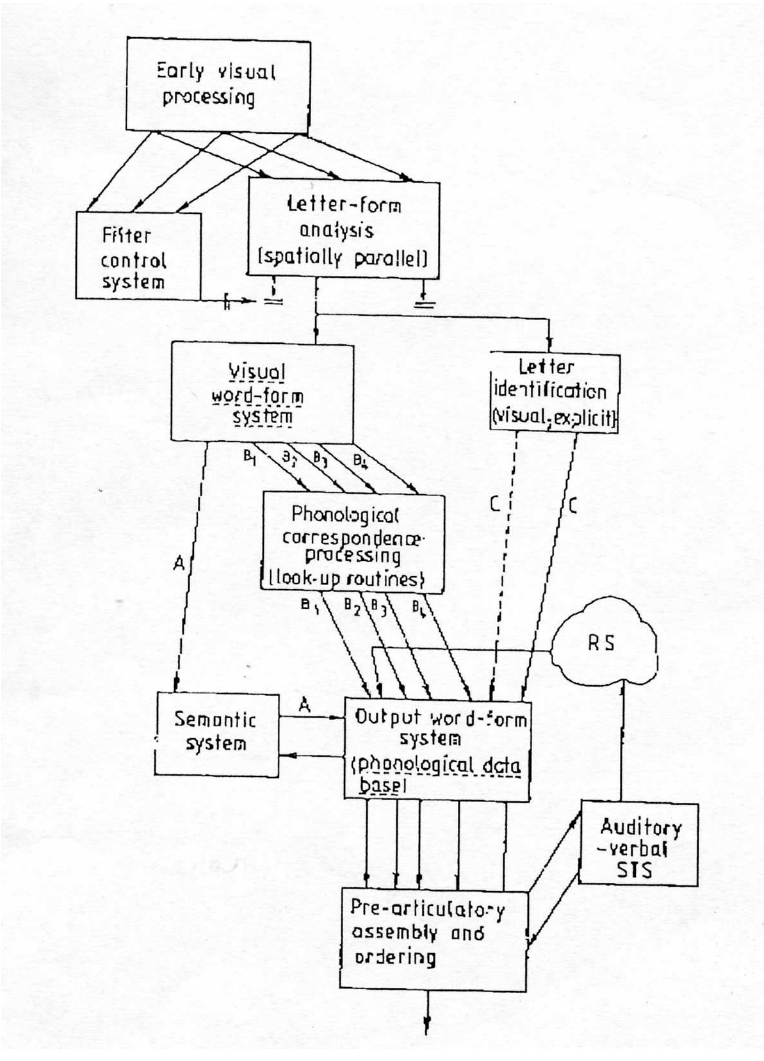


Figure 2.4: Model of the word-form multiple-levels approach to the reading process (Shallice, Warrington and McCarthy (1983)). A = the semantic route, B = the phonological route and C = the compensatory strategy route; B1-B4 represent difefrent levels of operation of the multiple level system and the division within subsystem.

2.2 Acquired reading disorders

Normal reading is a complex componential skill that is susceptible to particular patterns of breakdown after damage in relative isolation from other deficits. Because reading encompasses visual processing as well as linguistic analysis, deficits in any of these processes could give rise to an acquired dyslexia.

As shown before, acquired reading disorders have been extensively studied for a long time. For this reason reading (together with writing) represents one of the major contributions of neuropsychology to the study of cognitive functions. The neuropsychological approach to the study of reading ability allowed one to investigate the nature of the reading disorder and also to identify the different components of the reading system. In this chapter the features of the different types of acquired dyslexia are described and how reading disorders can be related to a model of reading will be examined.

2.2.1 Peripheral dyslexias

A useful starting point in the discussion of dyslexia is the distinction offered by Shallice and Warrington (1980) between ”peripheral” and ”central” dyslexias. The former are conditions characterized by a deficit in the processing of the visual aspects of the stimulus that interferes with the matching of a familiar word to its stored orthographic representation or ”visual word-form”. Central dyslexias, in contrast, are attributable to an impairment of the ”deeper” or ”higher” reading functions by which visual word-forms mediate access to meaning or speech production mechanisms. The different types of peripheral dyslexia are described below.

Pure alexia

Pure alexia is the most prototypical peripheral dyslexia that usually follows after damage in the left temporo-occipital region. These patients typically show intact production and comprehension of oral language, can even write normally, but they show a serial processing of letters to read in contrast to the parallel process observed in normal readers. The hallmark of the disorder is a ”word length effect”: using the slow and inefficient letter-by-letter procedure, pure alexics typically exhibit significant effects of word length, requiring more time to read long, as compared to short, words. The compensatory procedures used by such patients to read can be different (see Fig. 3.1 in Chapter 3): patients can circumvent damaged orthographic processing by letter naming (and then probably by using the knowledge of spelling in a ’reversed spelling process’ (Warrington and Shallice, 1980; Shallice, 1988) otherwise can use a sounding-out procedure based on learned associations between letters and sounds. The English patient MS studied by Newcombe and Marshall (1985) shows clearly as the ’letter-sound reading’ is possible as a procedure which assigns one phoneme to each letter of the input word (see also Patterson and Kay, 1982).

As said before, it was first described by Déjerine (1982) who gave a remarkable well-detailed anatomo-clinical description of his case (see Bub, Arguin and Lecours, (1993) for a detailed description of Déjerine’s interpretation). More recently it has been studied from a neuropsychological point of view to identify the components necessary to read. Warrington and Shallice (1980) and Patterson and Kay (1982) in their innovative work identified the word-form as a crucial component of the reading process which is damaged or inaccessible to pure alexic patients. This will be discussed in detail in Chapter 3.

In contrast to the central dyslexias, performance is typically not influenced by linguistic factors such as parts of speech (e.g. noun versus functor), the extent to which the referent of the word is concrete (e.g. table) or abstract (e.g. destiny), although this has occasionally been questioned (e.g. Behrmann, Plaut and Nelson, 1998).

Neglect dyslexia

Neglect dyslexia is characterized by a failure to identify letters on one side of a word. The affected side of a word is typically contralateral to the lesion, so when damage is in the right hemisphere, the difficulty usually appears in the identification of the left, initial part of the word. Errors are typically substitutions (word >lord), deletions (woman >man) and additions (hair >chair). One of the hallmarks of the disorder is the maintenance of target word length in the response, that is the range of letters omitted and added are -1, +1 (Kinsbourne and Warrington, 1962).

The performance of patients with neglect dyslexia is often influenced by the nature of the letter string: thus, patients may fail to report the initial letters in nonwords but read the real words correctly (Behrmann, Moscovitch, Black and Mozer, 1990). This lexical effect suggests the lexical representation may be partially accessed, therefore neglect dyslexia might not be attributable to a failure to register letter information but might reflect an attentional impairment at higher level of representation.

Although neglect dyslexia is generally seen in the context of the neglect syndrome, it has occasionally been observed in isolation (Patterson and Wilson, 1990) or even in the context of neglect of the opposite side of space (Costello, and Warrington, 1987).

In 1987 Ellis, Flude and Young investigated in detail the patient VB with neglect dyslexia. In text reading she often read only the right half of lines and in single-word reading she made errors which affected the initial letters (e.g. river >liver). Neglect errors in both words and nonwords typically involved the substitution of initial letters (rather than deletion or addition), resulting in errors of the same length as the target words.

Ellis et al. (1987) argued that in neglect dyslexia information about letter position is preserved but information about letter identity is lost. For instance, when train is read as chain, the visual analysis of letters provides the output -

(1), - (2), a (3), i (4), n (5) that indicates that the position has been coded, but not the identity. However, the existence of errors slightly different in length from the target suggest that the position of a letter in a word is not coded simply as its ordinal position in a left-to-right scan (Shallice, 1988).

Caramazza and Hillis (1990) proposed that neglect dyslexia reflects an impairment at one or more of three levels of representation involved in the early stages of visual word recognition. It has been argued that 3 kinds of spatial representations are needed for visual processing and therefore also for reading: representations at the retinocentric level, at the stimulus-centered level and at the word-centred level (Subbiah and Caramazza, 2000; Haywood and Coltheart, 2000). This idea is motivated by the fact that changes in word location and orientation do not affect patients in the same way.

Attentional dyslexia

Attentional dyslexia is a disorder of the visual attentional control associated with a damage to the left parietal lobe (Shallice and Warrington, 1977; Price and Humphreys, 1993; Warrington, Cipolotti and McNeil, 1993) where interference occurs when more than one item in a visual category must be identified. Patients with attentional dyslexia can have difficulties in identifying the letters in a word, even though the patient can read the word correctly. The deficit affects the reading of words in sentences -since there are many words visible at the same time- but not the reading of a single word displayed in isolation.

These patients make reading and migration errors: for instance, when several words appear simultaneously, letters from one word would migrate to the corresponding position in a second word. Thus, WIN FED would be read as FIN FED. These letter migration errors have also been observed with normal subjects under conditions of brief masked exposure of multiple words.

Interpretation of this deficit in attentional dyslexia begins with the idea that when we process an object the flow of information must be controlled by a filter that protects the ongoing analysis of that object from the activation of other competing, irrelevant objects (Triesman and Souter, 1995). Damage to this filter impairs selection, so that processing of a certain object is contaminated by other elements active at the same time. Shallice (1988) argues that the dyslexic errors occur because the filter control mechanism does not prevent letters outside the target word from activating units at the word-form level. The outcome is that elements of the target word are replaced by competing letters and the target is misidentified. This attentional control system operates by modulating an attentional window that is the sequence of parts of the letter string to be admitted to the word-form system; in attentional dyslexia it would be attenuated and would reduce the output of letter-level analyses.

Visual dyslexia

Visual dyslexia was described for the first time by Marshall and Newcombe (1973) as one the three acquired dyslexias following brain damage (i.e. visual dyslexia, surface dyslexia, deep dyslexia). References to visual dyslexia in the next 20 years were far less frequent than to surface or deep dyslexia. Where they were made, they tended to be less than positive: in 1988 Shallice for instance, claimed that visual dyslexia did not have a distinct status as a reading disorder. However, despite the little regard that visual dyslexia received in the eighties, the term has been used again more recently.

Lambon Ralph and Ellis (1997) have reported a case of visual dyslexia AB, that for some aspects is similar to AT, one of the patients described by Marshall and Newcombe (1973). Unlike AT, AB had a suspected form of dementia; her impairment suggested the presence of a central semantic deficit which it has been argued was not responsible for the large number of visual errors AB made in reading. AB showed a severe letter agnosia; on word naming she correctly read 57% of the words presented in different sets and she did not show a length effect. Over 90% of her reading errors could be classified as visual errors on the basis that they shared at least half the letters of the target word. However, AB's reading accuracy was affected by age of acquisition, imageability and frequency but not by letter length, similarly to AT (Marshall and Newcombe, 1973). However, AB was argued by the authors to show some right neglect symptoms: his reading errors tended to affect the ends of words more than their beginnings (without being morphological in nature). Overall, AB's problems appeared to not be purely visual or orthographic, as she showed phonological and semantic deficits and her performance was affected by a left-right gradient typically associated with right neglect dyslexia.

Cuetos and Ellis (1999) described a new case of acquired dyslexia which showed a large proportion of visual errors in reading. His letter processing was good in cross-case matching, but not innaming. In reading single words aloud, SC showed effects of imageability, frequency and also of word length. He read 45% of words correctly and 75.5% of the errors could be classified as visual errors, while only 1 error (out of 233 errors) bore a semantic relationship to the target. SC as well as AB (Lambon Ralph and Ellis, 1997) showed clear signs of semantic impairment in addition to their undoubted visual problems. The authors suggest that both semantic as well as visual-orthographic impairments may be necessary before a patient would misread such a high proportion of errors and make such a large proportion of visual errors (in the absence of semantic or phonological errors). Claiming that a semantic deficit in addition to a visuo-orthographic impairment might be necessary to have visual errors might appear as a surprise. However, Ellis and colleagues (Lambon Ralph and Ellis, 1997; Cuetos and Ellis, 1999) noted that their patients made a high percentage of reading errors generally (AB: 57% of correct words; SC: 45% of correct words), while the patients reported by Marshall and Newcombe (1973) were able to read words much more successfully (JL's last retest: 90% of correct words; AT: 85% and 87% of correct words from different subsets). According to them (Lambon Ralph and Ellis, 1997; Cuetos and Ellis, 1999), a combination of visual and semantic deficit may be necessary to induce such high levels of visual errors.

It can be hypothesized that the visual address or the orthographic representation is rarely damaged alone. It can be found damaged in association with for instance a left-right spatial gradient of attention, giving rise to neglect dyslexia, as the case reported by Warrington (1991). Alternatively, the orthographic representation can be damaged in association with the presence of a LBL reading strategy, giving rise to pure alexia. The fact that visual dyslexia is not contaminated by the use of the LBL reading strategy makes the syndrome more transparent and therefore worth investigating (Shallice and Rosazza, submitted).

2.2.2 Central dyslexias

In this section we describe the clinical features and conceptual basis of the major types of central dyslexia, including ”phonologic”, ”surface” and ”deep” dyslexia.

Phonologic dyslexia

Phonologic dyslexia was first described by Derouesne and Beauvois (1979) and it is characterized by a selective deficit in the procedure mediating the translation from print to sound.

Phonological dyslexia is a relatively mild disorder in which reading of real words may be only slightly impaired. Some patients with this disorder read all different types of words with equal facility (Funnel, 1983; Bub, Black, Howell and Kertesz, 1987; Friedman and Kohn, 1990), whereas other patients are relatively impaired in the reading of functors (Patterson, 1982; Glosser and Friedman, 1990).

Unlike patients with surface dyslexia described below, the regularity of print-to-sound correspondences is not a problem for them: these patients typically pronounce irregular words such as colonel and words with standard print-to-sound correspondences such as administer with equal facility. Most errors in response to real words appear to have a visual basis, often involving the substitution of visually similar real words (e.g. topple read as ”table”).

The striking and theoretically relevant aspect of the performance of phonologic dyslexics is a substantial impairment in the oral reading of nonword letter strings. Coslett (1991) and Derouesne and Beauvois (1979) have described patients with this disorder who read more than 90 percent of real words of all types yet correctly pronounce only 10 percent of nonwords. Most errors in nonword reading involve the substitution of a visually similar real word (e.g. phope read as ”phone”) or the incorrect application of print-to-sound correspondences (e.g. stime read as ”stim”).

Phonologic dyslexia has been observed in association with lesions in a number of sites in the dominant perisilvian cortex and, on occasion, with lesions of the right hemisphere (e.g. Patterson, 1982). Damage to the superior temporal lobe and in particular to the angular and supramarginal gyri is found in most, but not all, patients with this disorder.

Surface dyslexia

Surface dyslexia was first described by Marshall and Newcombe in their 1973 paper. Their patients JC and TS were able to read 50% of the words correctly and ”the vast majority of errors” were described as ”partial failures of grapheme-phoneme conversion”. They interpreted surface dyslexia as resulting from the semantic route being unavailable, leaving the patient ”no option other than attempting to read via putative grapheme-phoneme correspondence rules”. Patients with surface dyslexia are unable to access semantics by means of a direct lexical procedure: they can access to the word’s meaning only after the phonologic form has been derived. Thus, when presented the word ”listen”, a patient described by Marshall and Newcombe (1973) responded "Liston" and then added "that's the boxer".

Surface dyslexia is characterized by the inability to read words with "irregular" or exceptional print-to-sound correspondences. Patients with surface dyslexia are unable to read aloud words such as yacht, island and borough, the pronunciation of which cannot be derived by a phonologic strategy. In contrast these patients read words containing regular correspondences (e.g. state, hand, mint) as well as nonwords (e.g. blape) quite well.

In the context of a dual-route model of reading, the difficulty to process irregular words provides evidence that the impairment is in the mechanism(s) mediating lexical reading (see Fig. 2.3). Similarly, the preserved ability to read regular words and nonwords provides compelling support for the claim that the procedure by which pronunciations are computed (by the application of print-to-sound correspondences) is at least relatively preserved.

Noting that there is substantial variability in the performance of surface dyslexics with respect to reading latencies as well as accuracy, Shallice and McCarthy (1985) suggested that the syndrome of surface dyslexia can be fractionated. Type 1 surface dyslexia, they suggested, is characterized by relatively effortless and accurate reading of nonwords and regular words with poor performance of irregular words only. In the context of a dual-route model, in Type 1 surface dyslexia the deficit is at any point of the semantic route, i.e. either at the input lexicon or at the semantic system or at the phonological output lexicon (Shallice 1988). Type 2 surface dyslexia, in contrast, is characterized by slow, effortful reading; although these patients read irregular words less well than regular words and nonwords, they make errors with all the types of stimuli. These findings brought Shallice (1988) to reconsider whether patients actually use the grapheme-phoneme conversion. In fact if the patients with surface dyslexia used the grapheme-to-phoneme route, a different type of error should be expected. For instance, as Marcel (1980) noticed, only 25% of the errors that the original surface dyslexic patients JC and ST (Marshall and Newcombe, 1973) made were nonwords. Moreover, as Shallice (1988) reported, the errors that are consistently quoted as exemplars of surface dyslexia do not fit with the notion of the application of grapheme-phoneme correspondence rules (e.g. incense >increase or barge >bargain: where does the r in increase come from? And the -ain in bargain?). Finally as Henderson (1982) noticed, the surface dyslexic patient ROG studied by Shallice and Warrington (1980) was very slow at reading. Therefore Shallice (1988) concluded that surface dyslexia might arise from a deficit at the word-form system and that reading is a compensatory strategy adopted by the patients.

The anatomic correlate of surface dyslexia has not been well established. In fact the syndrome has been reported more frequently in the context of dementia (Shallice, Warrington and McCarthy, 1983; Warrington, 1975; Patterson and Hodges, 1992). Accordingly, surface dyslexia in demended patients is sometimes termed ”semantic dyslexia”. Many of these patients have exhibited brain atrophy most prominent in the temporal lobes (Breedin, Saffran and Coslett, 1994).

Direct dyslexia (Reading without meaning)

The study of Schwarz, Saffran and Marin (1980) provided strong evidence in support of the hypothesis of a third route to read in addition to the semantic and sublexical route (see the model of Morton and Patterson (1980) in Fig. 2.2). They reported a patient (WLP) who exhibited a profound loss of semantics in the context of dementia. Her performance was of particular interest because she was unable to comprehend words he read. This would not show anything if a high percent of words she was able to read was not irregular. Thus for example, when asked to sort written words into their appropriate semantic categories, she correctly classified only 7 of 20 names; critically WLP correctly read aloud 18 of these animal names, including irregular words such as hyena and leopard.

The same basic phenomenon, that is the ability to read aloud regular and irregular words that the patient does not understand has subsequently been reported by a number of investigators (Friedman, Ferguson, Robinson and Sunderland, 1992; Raymer and Berndt, 1994; McCarthy and Warrington, 1986). The pattern of performance exhibited by WLP and similar patients is of considerable theoretical interest. The fact that these patients can read irregular words which they do not comprehend suggests that reading may be mediated by a third mechanism, a third route that is lexical but not semantic (see Morton and Patterson, 1980). This mechanism was assumed to be lexically based, involving the activation of an entry in the visual word-form system and the "direct" activation of an entry in the phonologic output lexicon, with no intervening activation of the semantic system.

An alternative hypothesis was proposed by Shallice and colleagues (Shallice et al., 1983, Shallice and McCarthy, 1985; Shallice, 1988). These investigators attempted to explain reading without semantics within the context of a dual-route model by proposing that the phonologic reading procedure employs not only grapheme-to-phoneme correspondences but also correspondences based on larger units including syllables and morphemes. On this account WLP and similar patients are assumed to compute the pronunciation of irregular words they cannot understand by relying on the multiple levels of print-to-sound correspondences available in the phonologic system.

Deep dyslexia

Deep dyslexia is the other major reading disorder described by Marshall and Newcombe in their 1973 paper.

The hallmark of the syndrome are semantic errors: a deep dyslexic patient may read cat >mice and chair >table. These patients also produce a variety of other types of reading errors, including visual errors in which the response bears a clear visual similarity to the target (e.g. life >wife) and morphologic errors, in which a prefix or suffix is added, deleted or substituted (e.g. truth >true; invite >invitation).

Additional hallmarks of the syndrome include a greater success in reading words of high as compared to low imageability. Thus, words such as chair, table and ceiling are read more successfully by deep dyslexics than words such as peace, fate and destiny.

Also characteristic of the syndrome is a part-of-speech effect, such that nouns are read more reliably than modifiers (adjectives and adverbs) which are read in turn more accurately than verbs. Deep dyslexics manifest particular difficulty in the reading of functors like pronouns and prepositions (Saffran and Marin, 1977).

Finally all deep dyslexics exhibit a substantial impairment in the reading of nonwords. When presented with letter strings such as ”flig” or ”churt”, deep dyslexics are typically unable to employ print-to-sound correspondences to derive phonology and ”flig” is read as ”flag”.

Several accounts have been proposed to explain the deficit in deep dyslexia. Most investigators agree that multiple processing deficits must be hypothesized to account for the full range of symptoms. First the striking impaired performance in reading nonwords suggests that the print-to-sound conversion procedure is disrupted. Second, the presence of semantic errors and the effect of imageability (a variable usually thought to influence processing at the level of semantics) has been interpreted by many investigators as evidence that the patients also suffer from a semantic impairment (Morton and Patterson, 1980; Shallice and Warrington, 1980; Nolan and Caramazza, 1982). However, it should be noted that some deep dyslexic patients perform well on comprehension test with words they are unable to read. Semantic errors in these patients have been attributed to a deficit in or access to representations in the output phonological lexicon (Shallice and Warrington, 1980). Last, the production of visual errors has been interpreted by some to suggest that these patients suffer from an impairment in the visual word-form system.

Other investigators (Coltheart, 1980; Saffran, Bogyo, Schwartz and Marin, 1980) have argued that in deep dyslexia reading is mediated by the right hemisphere that is not normally used in reading. According to them, performance in deep dyslexia does not result from the operation of a normal system, within which certain components have been damaged, but depends on subsystems in the right hemisphere that are not normally used. The strongest argument for this hypothesis stems from the analogies between deep dyslexics and split-brain patients. Split-brain patients may be able to match printed words presented to the right hemisphere with an appropriate object (Zaidel and Peters, 1981), but unable to derive sound from the words presented to the right hemisphere; thus they are unable to determine if a word presented to the right hemisphere rhymes with an auditorially presented words. Another evidence is that deep dyslexia arises from large and deep left hemisphere lesions that affect much of the left-hemisphere areas, classically associated with language (Coltheart, 1980).

Although deep dyslexia has occasionally been associated with posterior lesions, this disorder is typically encountered in association with large perisylvian lesions extending into the frontal lobe (Coltheart, Patterson and Marshall, 1980).

2.3 Influences of cross-linguistic differences on reading

It has been shown that learning to read English is harder and possibly qualitatively different from learning to read consistent orthographies such as Italian, Spanish or Turkish (Goswami, Gombert, De Barrera, 1998; Ziegler, Perry, Ma-Wyatt, Ladner and Schulte-Korne, 2003). It is commonly accepted that the main reason for the delay of English-speaking children lies in the irregularity (inconsistency) of the writing system (e.g. Frith, Wimmer and Landerl, 1998). In English the relation between letters and sounds is often ambiguous: some letters or letter clusters can be pronounced in more than one way and some sounds can be spelled in more than one way (Ziegler, Stone, and Jacobs, 1997).

Recently, a cross-linguistic study compared word and nonword reading performance after the first year of reading instruction across 13 orthographies (Seymour, Aro and Erskine, 2003). They showed that word and nonword accuracy was only about 40% for English children at the end of grade 1. In contrast, word reading accuracy in most other European orthographies was close to ceiling with the exception of French and Danish. Fundamental linguistic differences in syllabic complexity and orthographic depth are thought to be responsible for the results.

Not only children but also adults show the influence of orthographic depth on word recognition. The regularity of the orthography-to-phonology correspondence interacts with frequency in normal English adults (Balota and Ferraro, 1993; Seidenberg, Waters, Barnes and Tanenhaus, 1984) and the lexical status of the stimulus (i.e. being a high-frequency word, a low-frequency or a nonword) affects reading speed more in languages with deep rather than shallow orthographies (Frost, Katz and Bentin, 1987). This happens because where the mapping between orthography to phonology is not transparent, readers must map the whole orthographic string onto a lexical representation to name words aloud.

In Italian, it has been shown that lexical stress influences word reading only if words are low in frequency (Colombo, 1992; Colombo, Fonti and Cappa, 2004). Moreover the stress consistency is predictive of reading speed and accuracy in low frequency words (Burani and Arduino, 2004). This interaction of stress consistency and frequency is equivalent to the frequency by regularity interaction in English (Seidenberg et al., 1984). In the cross-linguistic study reported by Paulesu et al. (2000), the behavioural data showed that Italian students read words and nonwords more quickly than English students, suggesting that reading in Italian can proceed more efficiently because of the consistent mapping between individual letters and their corresponding sounds.

The impact of an inconsistent orthography with respect to a transparent one has been studied also in developmental dyslexia. There is now overwhelming evidence in favour of a specific phonological deficit in dyslexia. However, a number of studies comparing the reading disorder in various countries has suggested that the differences in reading difficulties between regular and irregular languages are due to differences in orthographic consistency (Landerl, Wimmer and Frith, 1997; Ziegler, Perry, Ma-Wyatt, Ladner and Schulte-Korne, 2003).

Finally, neuroimaging studies have been used to explore which brain regions support reading and how this network of regions varies for different languages. An influential study investigated reading processing in Italian and English students (Paulesu et al., 2000). It concluded that there is a common multi-component system which supports reading of both Italian and English orthographies but that the contribution of each component varied significantly for the two languages. The Italian readers activate an area (the left superior temporal regions) involved in the phonological processing, whereas English ones activate areas (left posterior inferior temporal gyrus and anterior inferior frontal gyrus) more associated with word retrieval during both the reading and naming task. Thus cultural factors, as reflected in orthographic systems, can powerfully shape the cognitive and brain systems that underpins the language function.

Studies on Chinese reading have produced similar results in terms of a distributed network of brain areas with regions common to both logographic and alphabetic languages as well as brain regions specialized in processing logographic Chinese (Tan, Liu, Perfetti, Spinks, Fox, Gao, 2001). Reading a word in

Chinese is associated with greater activity in specific regions in the right hemisphere and in the left middle frontal gyrus, that is supposed to coordinate and integrate various information about written characters in verbal and spatial working memory (Siok, Perfetti and Tan, 2004). As in English word reading, the inferior prefrontal cortex is active in processing Chinese characters.

In conclusion, in a consistent language (shallow orthography) readers are encouraged to use the phonological pathway because the mapping between letters and sounds is relatively direct and unambiguous. In contrast, in an inconsistent language (deep orthography), readers should be reluctant to use phonological pathways because of the less systematic mapping between spelling and sound. Instead, they should rely to a greater extent on the lexical pathway. This cross-linguistic differences have been studied in cognitive psychology, developmental dyslexia and with brain imaging, but rarely applied in the study of acquired dyslexia. In the context of pure alexia where a serial reading procedure is applied, one could expect a greater difficulty in reading English words than Italian words. In fact in English the pronunciation of a letter or a letter cluster is ambiguous and context-dependent. When a word is read serially, the pronunciation of a letter can depend on the presence of subsequent letter(s), not necessarily adjacent, for instance ’mi’ is pronounced /mi/ in *mill*, but /mai/ in *mile*. English can have multiple phonological realizations and it is often necessary to go to the end of the word to know how to pronounce a combination of letters. This type of errors has been frequently reported in literature by patients using a serial procedure in English. For instance, the (English) pure alexic patients reported by Patterson and Kay (1982) made mistakes in putting into words letters correctly identified, as they pronounced silent letters (knowledge >/knouledg/; castle >/kastli/) or did not follow the ”final *e* rule” (e.g. while >will). The patient MS (Newcombe and Marshall, 1985) was able to read only by segmenting a word into single letters which were then transcoded into phonological forms, e.g. phrase >/pə’hə’ræsi/; treat >/tə’ri’ æt/. His pattern of performance was called ”letter-sound reading”.

In pure alexia a serial strategy which brings to the correct pronunciation of a word can occur through a spelling procedure (Warrington and Shallice, 1980), a ”letter-sound reading” (Newcombe and Marshall, 1985; Shallice, 1988) or a visual recognition of letters and letter clusters (Hanley and Kay, 1992). In any case, it is possible to speculate that the integration of letters will be easier in Italian than in English due to the lower degree of inconsistency between orthography and phonology.

2.4 Computational models of reading

Up to this point we have focused on information-processing approach based on flowcharts models to account for how we recognize visual words (and non-words). However flowcharts are never specific enough. In this section we examine some computational models that represent a theory in the form of a computer program which does so using exactly the procedures which, according to the cognitive theory, are used by human beings when they are carrying out that cognitive (reading) task.

All the models considered here take as input a visual representation of the word and output desired information such as meaning or sound.

A complete description of each connectionist model is beyond the scope of the current work. Here we take into account the principles of each model that we consider relevant to understand how we recognize a letter string. For instance, whether there are one or more processing routes, or how processing proceeds from letters to the word.

Fundamentally different types of reading models have been proposed.

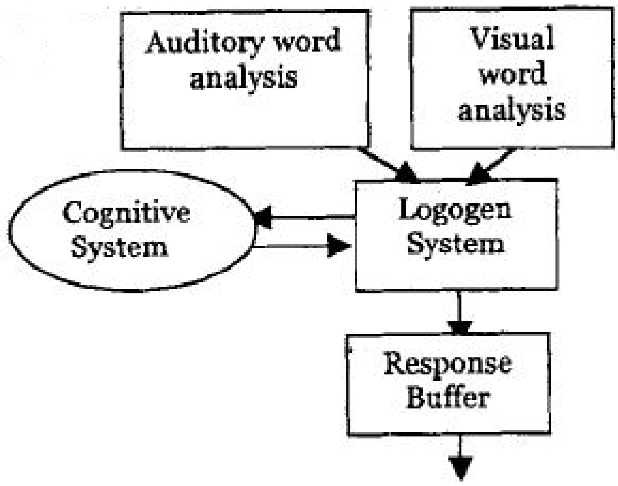


Figure 2.5: The original version of the *logogen* model (Morton, 1969).

2.4.1 Logogen model -Morton (1969, 1979)

Morton's original logogen model (1969), from the Greek words logos = "word" and genus = "birth", is the foundation for all activation models of lexical access. According to Morton (1969; 1979), each word has its own logogen, that is an evidence detector for a word, "the device which makes a word available" (Morton, 1979, p112). A logogen accumulates evidence until the amount of evidence reaches a threshold. When this happens, the word is recognized (see Fig. 2.5). The threshold processing, where activation is only passed on to the later modules after a threshold is reached in the earlier module, is still used as for instance in the DRC Model (Coltheart, Rastle, Perry and Ziegler, 2001) and it is an alternative to the cascaded processing used for instance in the IAC Model (McClelland and Rumelhart,1981, 1982).

Context and stimulus information increase a logogen's resting activation: word frequency for instances, changes the logogen's threshold. On this view, high frequency words have a low threshold and need less activation to fire than low frequency words. In this model activations but not inhibitions are considered; moreover in the original model (see Fig. 2.5) a single logogen carried out all language tasks for a particular word, regardless of modality. It has undergone considerable revision since its original formulation: it is noteworthy that the evolution of the logogen model was entirely data-driven.

2.4.2 IAC Model -McClelland and Rumelhart (1981, 1982)

One of the earliest and most influential connectionist models of reading is the interactive activation cascaded (IAC) model of McClelland and Rumelhart (1981; Rumelhart and McClelland, 1982). Interactive means that the activation at higher levels feeds back to lower levels to provide additional support (so that activation of the word nodes can affect what happens to the letter nodes). Cascaded means that activation propagates from lower to higher levels immediately and continuously without waiting until processing at lower levels is complete.

The model has 3 representational levels: a visual feature level, a letter level and an orthographic word level (see Fig. 2.6). The IAC is a non-learning model that means that the architecture of this model was specified by its creators and localist: features, letters and words are represented by individual nodes.

When a particular letter in a particular position is activated, it sends activation to all words which incorporate it and inhibition to others. Differently from the logogen model, in IAC there are both excitatory and inhibitory connections within and between levels.

This model was designed to account for recognition of 4-letter upper case words; however, many aspects have more general applicability. The model has been evaluated only against data from a forced-choice tachistoscopic recognition task and the model's performance was shown to fit rather well the performance of human readers.

Interesting for our view is that words are processed in parallel, by a set

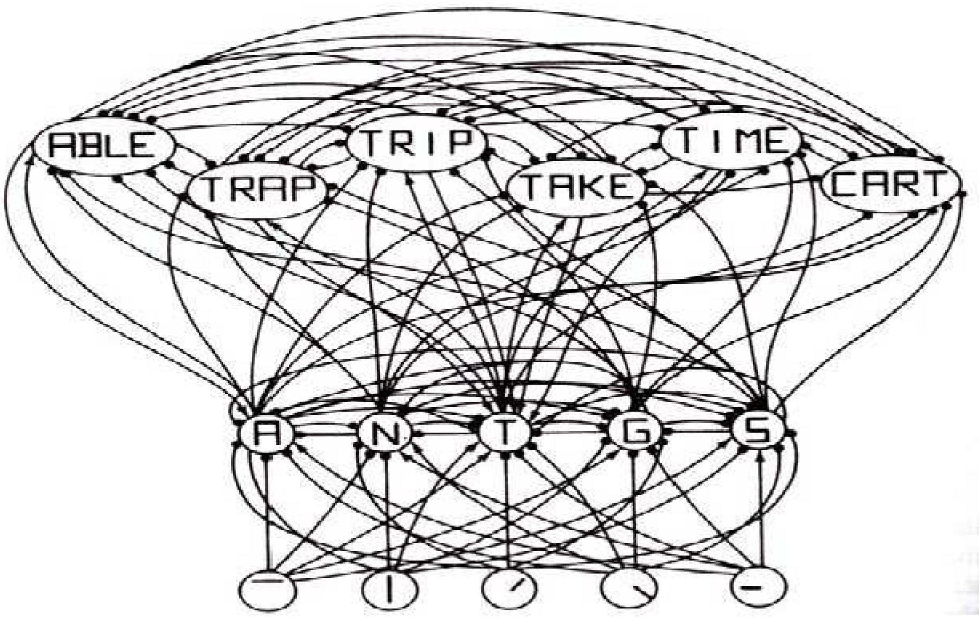


Figure 2.6: The interactive activation cascaded (IAC) model by McClelland and Rumelhart (1981; 1982).

of position-specific detectors. Visual information is first processed in terms of letter features (e.g. horizontal, vertical and oblique lines, curves and circles); then information is passed to a letter level where the features are combined to form letters. Finally the letter information is then passed upwards to the word level where letters are combined to form words.

It is worth noting that detectors are feature- and letter- position specific: this way of coding letter position information necessitates a large number of duplications of the alphabet to represent each letter in each possible position. Moreover letters are directly connected to words, without any intermediate level.

2.4.3 PDP Model (Plaut, McClelland, Seidenberg and Patterson, 1996)

Parallel distributed processing (PDP) models differ from information processing accounts in that they do not incorporate word-specific representations (e.g. visual word forms or output phonological representations). Subjects are assumed to learn how written words map onto spoken words through repeated exposure to familiar and unfamiliar words. Learning of word pronunciations is achieved by means of the development of a mapping between letters and sounds generated on the basis of experience with many different letter strings. The probabilistic mapping between letters and sounds is assumed to provide the means by which both familiar and unfamiliar words are pronounced (see Fig. 2.6).

The model developed by Plaut, McClelland, Seidenberg and Patterson (1996) is a PDP connectionist model. The critical feature of PDP models by comparison with dual-route framework (see below) is that there is a single uniform procedure for computing phonological representations from orthographic representation, that is applicable to regular as well as irregular words and nonwords; there is no route involving grapheme-phoneme correspondence rules.

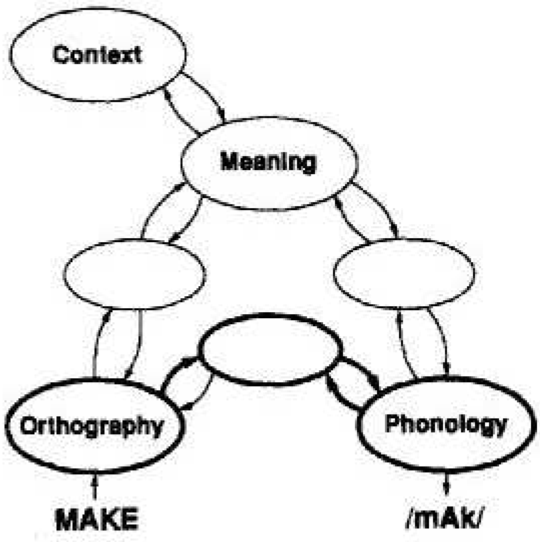


Figure 2.7: PDP Triangle model (Seidenberg and McClelland, 1989).

The model comprises 3 levels of units (grapheme units, hidden units and phonemes units) and it has been trained on 2998 monosyllabic words with back-propagation.

After training, the network read regular and exception words flawlessly, as well as nonwords. Interestingly, the network which implemented only 1 mechanism did not segregate itself over the course of the training into separate mechanisms for pronouncing exception words and nonwords. Therefore Plaut et al. (1996) showed that a one-route procedure can read both exception words and nonwords very well with appropriate RTs and typical effects such as frequency and regularity.

Important for our perspective is how words are coded in subunits. To convert a letter string into an activity pattern over the grapheme units, the string is parsed into 3 positions: onset consonant cluster, vowel and final coda consonant cluster. Letters (and letter clusters) are assigned to 1 of these 3 positions.

Moreover, this model not only accommodates many of the classic findings in the literature on normal reading, but it has also been ”lesioned” in an attempt to reproduce the patterns of reading impairment characteristic of surface dyslexia (Plaut et al., 1996; but see Coltheart 2001 for a different view). More generally, the PDP models have been used also to reproduce deep dyslexia (Plaut and Shallice, 1993).

2.4.4 DRC Model - Coltheart, Rastle, Perry and Ziegler (2001)

An alternative computational account of reading has been developed by Coltheart and colleagues (Coltheart, Rastle, Perry and Ziegler, 2001). The dual-route cascaded model represents a computationally instantiated version of the dual-route theory presented in Fig. 2.8.

This model consists of 2 routes, the lexical route and the nonlexical route. Each route is composed of a number of interacting layers. These layers contain different subset of units: visual feature units, letter level units and phoneme units. Importantly, layers have a position-specific coding. According to this model, the initial stage in reading consists of visual feature analysis of the letter string’s individual components. The sets of features extracted correspond to a letter and this process allows an abstract letter identification. Note that this has nothing to do with phonology. The abstract letter units system feeds the GPC rules as well as sends input to an orthographic lexicon, which contains a distinct unit for each of the words.

This model is modular with a functional partition (as opposed to the interactive of PDP models) and have localized representation (as opposed to distributed). Moreover the model assumes parallel activation in the lexical route, while the nonlexical route is assumed to occur serially, from left-to-right, across a grapheme string.

This model was able to simulate the data reported from cognitive psychology in terms of RTs, accuracy and of the effects of lexicality, frequency and

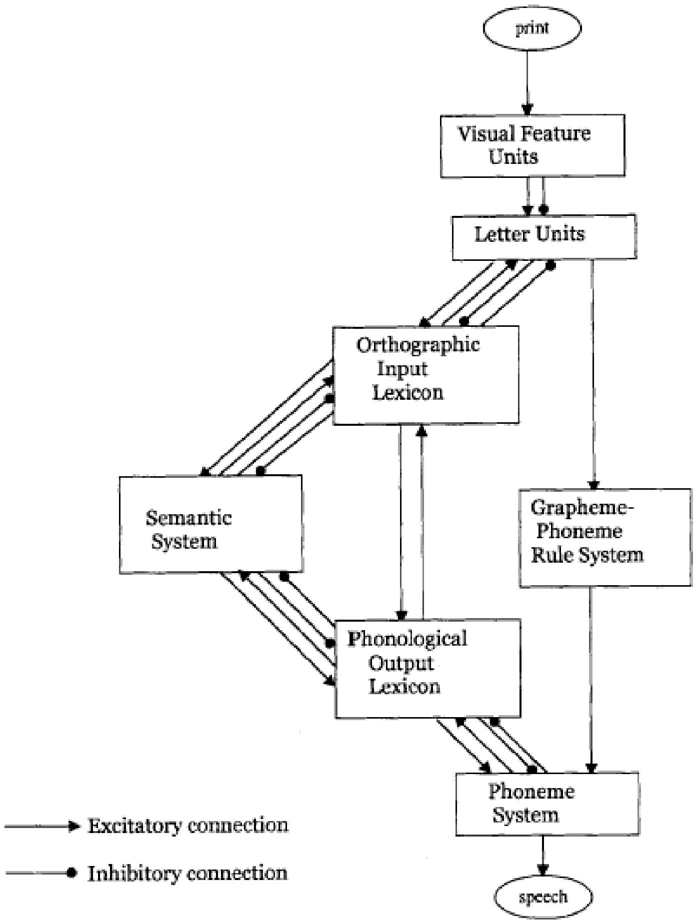


Figure 2.8: The dual-route cascaded model of visual word recognition (DRC) by Coltheart, Rastle, Perry and Ziegler, (2001).

regularity.

The main point for our perspective is that this model incorporates a lexical route as well as a nonlexical route by which the pronunciation of graphemes is computed on the basis of position-specific correspondence rules. The letter level is directly connected to the orthographic lexicon.

2.4.5 CDP -Zorzi, Houghton and Butterworth (1998)

Zorzi, Houghton and Butterworth (1998) developed a new computational model of reading. It is a connectionist dual-process (CDP) model, that maintains the uniform computational style of the PDP models but makes a clear distinction between lexical and sublexical processing in reading. The model comprises an input orthographic layer and a phonological output layer (see Fig. 2.9) and the processing is parallel also in the nonlexical route.

In the model the orthographic representation is strictly position specific, it is slot based and equivalent to that used in McClelland and Rumelhart (1981) for the letter detector level. Letter position in monosyllabic words is coded relative to the orthographic onset (i.e. letters preceding the vowel letter) and orthographic rime (or word body, i.e. all letters from the vowel onward). The first 3 positions are for the (orthographic) onset representation and the onset slots are filled from the first slot onward. The orthographic rime, which has a maximum length of 5 letters is represented on the following 5 slots.

The dual process model was able to simulate the latencies for regular and irregular words as well as the effects of lexicality and regularity and the performance of dyslexic patients. In particular they showed that the phonological assembly process can be implemented by a network which extracts the regularities in the spelling-sound mapping (for English) from training data containing many exception words. On this account knowledge about spelling-sound regularities can be acquired and easily used if it is separated from knowledge of the pronunciation of known words.

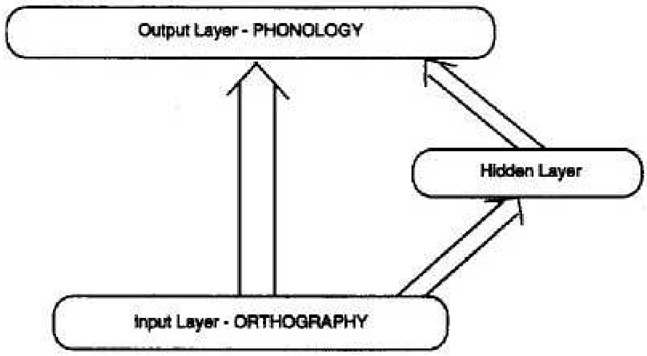


Figure 2.9: The connectionist dual-process (CDP) model by Zorzi, Houghton and Butterworth (1998). Architecture of the model with the hidden layer pathway.

Important for our perspective is that this model mainly focuses on the interaction between orthography and phonology and that the connections go directly from single letters to single phonemes.

2.5 How written words are coded?

Some models of word recognition have been expressed in the form of computer programs which aim to simulate aspects of human word recognition. The most recent models (Plaut et al., (1996), Coltheart et al. (2001), Zorzi et al. (1998) here described were able to simulate the data reported from cognitive psychology in terms of RTs, accuracy and effects of lexicality, frequency and regularity, although they adopted different assumptions. However, as we know, the fact that models are adequate does not guarantee that they are correct.

Although computational models have played a key role in the study of reading, most of them have focused on the interactions between orthography, phonology and semantics, ignoring the more peripheral stage of visual word recognition. Most current computational models (e.g. Coltheart et al., 2001; McClelland and Rumelhart, 1981) assumes that abstract letter representations are directly connected to word form representations, ignoring the possible role of sublexical units such as syllables. A further limit is that these models often presuppose a case- and location- invariant representation (Dehaene, et al., 2005). The main problem is that the processing from a single letter to the word identification has been underestimated. Therefore since most of the models did not focus on the early stage of word recognition, they do not make predictions about the reading patterns which result from a deficit at the different levels of this early stage.

Therefore, we can hypothesize which stages we are likely to carry out when we see a word. When a word such as 'albero' is presented, it is first analysed in terms of contours, shape and single letters by the visual areas (ranging from V1 to V4) which compute increasingly abstract representations. The first stage specific to orthographic material we experience is of letter processing. Then letters are integrated into subunits like syllables and morphemes. Finally the word unit is carried out.

Different types of evidence suggest the existence of these stages. The existence of the letter-level processing is provided by various studies. First, it is unlikely that reading is based directly on visual features or shape information: any time we see a word written in a new font we should not be able to read it. Second, the mixed effect, revealed more than 25 years ago (McClelland, 1976), where a word like 'aLbErO' can be read with relative ease (although more slowly than the normal presentation). An analysis at the single abstract letter identity level must be carried out in order to read the word. Third, migration errors that can occur in attentional dyslexia as well as with normal subjects under certain conditions of brief presentation (Davis and Coltheart, 2002), indicating that letters are coded separately. Fourth, experimental psychology studies using priming methods have consistently shown that information concerning the identity and the position of individual letters is already coded in the early phase of processing (Peressotti and Grainger, 1995). Humphreys, Evett and Quinlan (1990), for example have shown that letter-string primes facilitate the identification of the target word when prime and target share common letters. This priming effect can occur with only 1 letter and it varies with the position of that letter. For instance, the target report was facilitated when primes and targets had the first letter in common, but not when the letter was the second one or the third one. In fact the effect is more robust for end letters than middle letters. Fifth, an imaging study with normal subjects showed that the most posterior part of the visual word form area (VWFA), a brain region particularly responsive to visual word recognition, was bilaterally activated by letter-form processing and insensitive to the word-level (Dehaene, Jobert, Naccache, Ciuciu, Poline, Le Bihan and Cohen, 2004).

As mentioned before, the integrative process of letters into sublexical units has been largely underestimated. However there is evidence indicating an intermediate stage comprising bigrams, syllables and morphemes before accessing the word form. First, Grainger and Whitney described a scheme called 'open bigrams' (Schoonbaert and Grainger, 2004; Grainger and Whitney, 2004; Whitney, 2001). This scheme, that has not been implemented in a computational model, comprises 5 layers: a retinal level, a feature level, a letter level and then a bigram level before the final, word level. Therefore letter and word levels are not directly connected. On this account, word coding is based on ordered letter pairs, the bigrams, which do not contain precise information about letter position: for example the word 'take' is represented by activation of units representing TA TK, TE, AK, AE and KE[1].

This model can account for effects of similarity and of letter transposition shown in experimental studies of priming. For example in studies of masked-priming, 'garden' is identified more rapidly when preceded by a masked prime that respects the relative positions as 'grdn', compared to the control condition 'pmts' or to the condition where the order is changed as 'gdrn'. Although this scheme has some problems (for instance it fails to assign a unique code to each word) it takes into account the issue of how integration of letters can occur.

Second, there is evidence in favour of syllabic processing in reading words at least in languages with clear syllabic boundaries such as Italian, French or Spanish (Ferrand, Segui and Grainger, 1996; Carreiras, Alvarez and de Vega, 1993). In a study with ERPs, Carreiras et al. (in press) found that when in bisyllabic words and pseudowords there was a match between syllable boundaries and colour boundaries, different evoked responses emerged compared to when there was a mismatch between the syllable boundaries and the colour boundaries. In particular, the ERP effect of colour-syllable congruency for both words and pseudowords was very early, namely in the P200 time window. Lexicality effects showed up at the N400 component. This suggests that: 1. at least in languages with clear syllabic boundaries, the syllable has a role in word processing, not only for its phonological but also for its visual nature; 2. generally, we might not process bisyllabic words as a whole.

Finally, in a recent model put forward by Dehaene et al. (2005), it was attempted to explain how words are coded by solving problems of location and case invariance (see Fig. 2.10). Their model is inspired from neurophysiological models of invariant object recognition and proposes a hierarchy of local combination detectors sensitive to increasingly larger fragments of words. More

[1]It is noteworthy that in these calculations open bigrams are limited to a maximum of two intervening letters, therefore the word 'garden' is represented only by GA, GR, GD, AR, AD, AE, RD, RE, RN, DE, DN and EN.

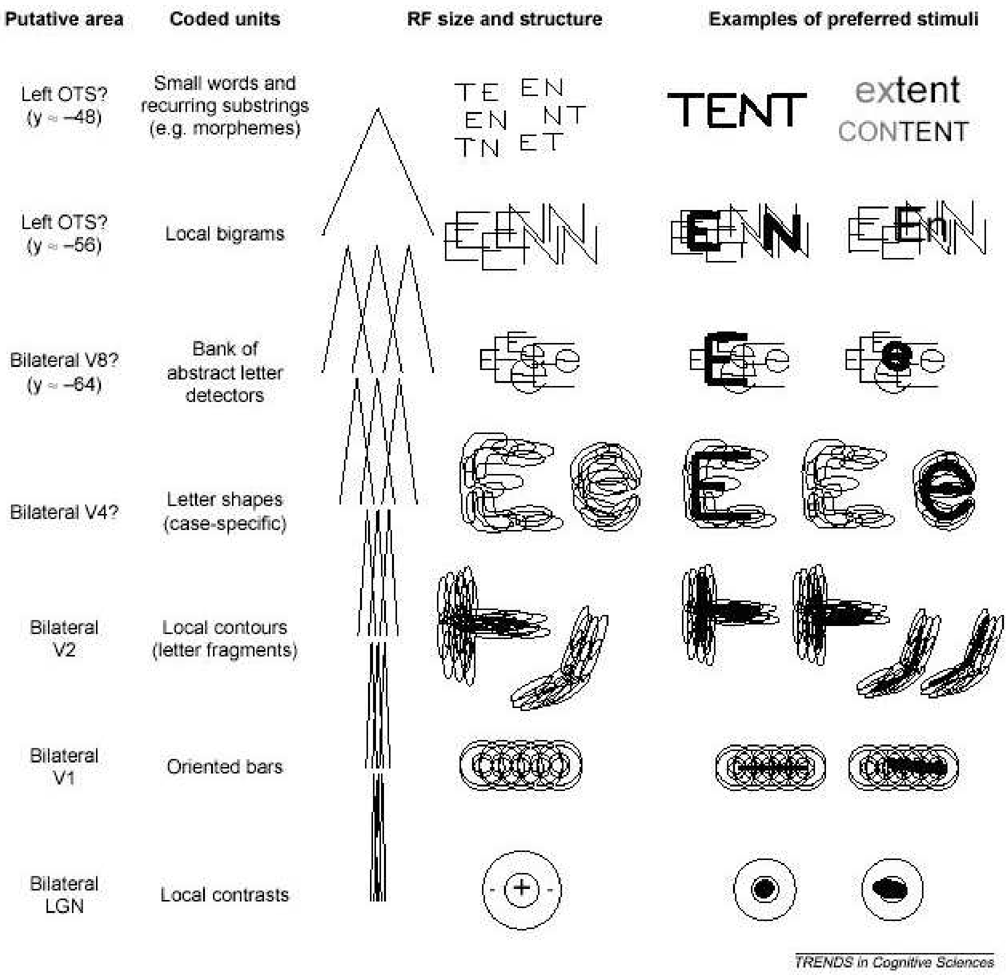


Figure 2.10: Model of invariant word recognition by a hierarchy of local combination detectors (Dehaene, Cohen, Sigman and Vinckier, 2005). This model is inspired from neuropsysiological models of invariant object recognition. Each neuron is assumed to pool activity from a subset of neurons at the immediately lower level, thus lead to an increasing complexity, invariance and size of the receptive field at each stage.

specifically, by assuming to pool activity from a subset of neurons at the immediate lower level, they propose that at first there are neurons which respond to local contrasts, then to oriented bars and then to local contours (fragments). At the next stage, combinations of fragments can be used to recognize a letter shape in a specific case; abstract letter identities is then recognized at the next stage by pooling activity from letter shapes detectors. The subsequent stage comprises neurons sensitive to local combination of bigrams, as it is hypothesized by the model of Grainger and Whitney, (2004); finally there are neurons sensitive to ordered combinations of bigrams, morphemes and small words. Therefore as occurs in object recognition, an integrative process is postulated for word recognition through a hierarchy of local combination detectors.

In the present work we attempted to study the early stage of word coding by focusing both on the letter level and on the integrative process that brings to the identification of syllables and words.

Chapter 3

Pure alexia and hemianopic alexia

3.1 Introduction

The term alexia is derived from Greek and literally means ”without word” or ”not word” (the prefix ”a-” means without or not and ”lexis” means word). The term alexia denotes the presence of an acquired reading disorder that prevents the rapid and effortless recognition of written language. Hemianopic alexia, as well as pure alexia are different forms of peripheral alexia, where the deficit prevents the patient from matching a word to its visual lexical representation (Shallice and Warrington, 1980). In pure alexia patients lose the ability to identify letters rapidly and in parallel; the brain damage often causes right homonymous hemianopia (RHH) but this visual defect is not critical for the inability to read. Instead, in hemianopic alexia the deficit is strictly caused by the presence of the visual field deficit which slows text reading (Leff, Crewes, Plant, Scott, Kennard and Wise, 2001). In hemianopic alexia word recognition is considered to be intact, but a parafoveal field loss leads to a disruption of the oculomotor reading pattern during text reading (Zihl, 1995). Naturally, both the visual fields can be damaged and the degree of parafoveal visual sparing contributes significantly to the observed reading impairment.

In order to understand the difference between pure alexia and hemianopic alexia, it is worthwhile considering how words are processed when presented in the LVF and RVF. Due to the routing of optic fibers at the optic chiasm, a word presented in the left visual field (LVF) is initially projected to the visual areas in the right hemisphere (RH) and then processed in the LH via the corpus callosum. By contrast, a word presented in the right visual field (RVF) is directly projected to the left hemisphere (LH). This direct connection explains why the number of letters in a word has a greater impact on recognition speed and accuracy in the LVF than in the RVF (Ellis, Young and Anderson, 1988; Lavidor, Hayes, Shillcock and Ellis, 2004). The asymmetry of the length effect reflects the LH superiority for language with efficient parallel processing of letters and the RH non-parallel processing (Ellis, 2004).

In alexia, a lesion affecting either the left primary visual cortex or its geniculostriate afferents, but sparing the interhemispheric connections, will cause hemianopic alexia if right foveal (and/or parafoveal) vision is compromised. In reading the role of the interhemispheric connections through the corpus callosum is to convey visual information about letters displayed in the LVF from the right visual cortex to the language areas in the LH. As a consequence, an isolated callosal lesion will induce pure alexia restricted to the LVF, namely left hemialexia, as called by Cohen and colleagues (Cohen, Martinaud, Lemer, Lehricy, Samson, Obadia, Slachevsky and Dehaene, 2003; Molko, Cohen, Mangin, Chochon, Lehéricy, Le Bihan and Dehaene, 2002). Patients with left hemialexia can read words presented in the RVF but not in the LVF. By contrast, in patients with hemianopic alexia the callosal route is vital to reading, as it provides the only visual input to the language areas (Cohen et al., 2003). Reading will be possible with words presented in the LVF but not in the RVF. For these patients an additional callosal lesion would be sufficient to yield pure alexia. In fact a patient with hemianopic alexia cannot read words presented in the RVF, (depending on the degree of visual field damage), but he or she will be able to read words presented in the LVF, with some degree of length effect similar to the moderate length effect observed in normal subjects reading LVF words (Cohen et al., 2003). Differently from hemianopic alexia (and left hemialexia), in pure alexia the lesion has compromised both intrahemispheric and interhemispheric visual pathways, therefore patients are only able to read words in either visual field, slowly and laboriously.

3.2 Hemianopic alexia

While varieties of acquired dyslexia have attracted most attention in the literature, only few reports deal with reading impairments caused by visual deficits. Difficulties at the level of visual processing can interfere with reading because disrupted or missing text information impairs access to the representation of words. It was Wilbrand who in 1907 coined the term ”hemianopic alexia” and suggested that parafoveal field loss was the main cause of this reading disorder. Time has proven him right, the parafoveal visual defect represents the most common cause of impaired reading (Zihl, 1989, 1995), underscoring the crucial importance of visual processing in addition to lexical, semantic, phonological and motor processing components. In hemianopic alexia the parafoveal field loss results in a slowed text and word reading. The visual field defect can be both in the LVF and RVF, depending on where the brain damage occurred. The slowness in text and word reading naturally depends on the extent of the visual defect.

The visual field comprises three regions as we look straight ahead: foveal, parafoveal and peripheral. Acuity is very good in the fovea, not so good in the parafovea and even poorer in the periphery. It is commonly agreed that parafoveal visual field plays a crucial role for both text recognition and guidance of eye movements in reading. Both the foveal and the parafoveal visual field regions act as a common 'perceptual window' and provide the basis for the so-called perceptual span (word recognition/reading span) which is defined as the field of useful vision during an eye fixation (McConkie and Rayner, 1975). Its extent, which depends on the characters and the difficulty of the text, is in general in the range of 8 degrees (Ikeda and Saida, 1978; Rayner and Bertera, 1979). Readers of left-to right orthographies acquire more information from the right side of fixation than from the left, indicating that the perceptual window (span) is asymmetric; it extends far more to the right (up to 15 characters) than to the left (three or four letters) (McConkie and Rayner, 1975, 1976; Rayner and Bertera, 1979). Interestingly, for scripts read from right-to-left the preference of the eye is to land to the right of the center of words (Deutsch and Rayner, 1999). Despite the fact that on-going linguistic processing modulates reading eye movements, the location where the eyes initially land in words appears to be determined mainly by low-lovel visuo-motor factors (Nazir, Ben-Boutayab, Decoppet, Deutsch and Frost, 2004; Rayner, 1998).

Since for scripts read from left-to-right the perceptual window is smaller on the left, it is plausible to expect differential effects of left versus right hemianopia on reading performance and such differences have been repeatedly reported (De Luca, Spinelli, Zoccolotti, 1996).

Some of the most important work done recently on hemianopic alexia is by Zihl (1995). Zihl (1995) investigated the role of the parafoveal field loss in 50 patients with hemianopic alexia and studied how this loss impaired reading. As expected, Zihl (1995) found that the degree of reading impairment depended on the extent of visual field sparing, ranging from 1 to 5 degrees. Moreover, patients with right hemianopia were more impaired than patients with left hemianopia loss. Eye movement reading patterns confirmed this observation. Left-sided field loss mainly impairs return eye movements to the beginning of a line, while right-sided field loss was associated with prolonged fixation times, reduced amplitudes of saccades to the right and many regressive saccades. This result suggested that the perceptual window (the reading span) is altered: its spatial size is reduced, while its temporal extent is increased.

3.3 Theoretical accounts of pure alexia

Pure alexia is a form of acquired dyslexia in which patients do not appear to be able to read in the sense of fast, automatic word recognition. As noted in Chapter 2, pure alexic patients use a slow LBL procedure to read and this feature characterizes the syndrome: the time taken to read increases with word length. The effect of word length on reading times varies greatly from one case to another, but a typical patient might require 3 or 4 seconds to read 3-letter words and reading times often increase by 2-3 seconds (or more) for every additional letter (Bowers, Bub and Arguin, 1996). Generally the deficit occurs in the absence of impaired language and even without impaired writing or spelling. It is noteworthy that patients typically have relatively intact visuo-perceptual abilities and can recognize objects accurately at a gross level: this is not a trivial aspect since a word is nothing else than an object, although a special class of objects.

Spelling often is intact in these patients, as well as comprehension, speech, production and writing yet they struggle to read what they have written.

Typically the lesion associated with pure alexia is in the left occipito-temporal lobe, sometimes accompanied by damage to callosal fibers in the splenium of the corpus callosum or forceps major. In most pure alexic patients the lesion results in a dense right visual field defect.

3.3.1 Where is the deficit? Different classes of theories

Different accounts have been proposed so far in the attempt to explain the specific functional source of the deficit, from those providing an explanation in terms of impairment to more peripheral processes (Farah and Wallace 1991; Sekuler and Behrmann 1996; Behrmann, Nelson and Sekuler 1998, Behrmann, Plaut and Nelson 1998; Rapp and Caramazza 1991; Price and Humphreys 1992), to those giving a somewhat more central account of the locus of impairment (Warrington and Shallice, 1980; Patterson and Kay, 1982; Arguin, Bub and Bowers, 1998). However the term ”impairment to peripheral processes” is imprecise and ambiguous because it might have different meanings. It could indicate a general perceptual deficit involving object processing but also an impairment that is more peripheral compared with the word level, such as a difficulty at letter level (Behrmann and Shallice, 1995). If there is a slowed letter activation, the deficit can be seen at an intermediate level between peripheral early processing and central, word processing. In this view the subdivision of the theoretical accounts in ”peripheral versus more central deficit” reflects a contrast that is too simple.

It would be more appropriate to divide theories into those claiming the deficit to be at a general perceptual level and those claiming the deficit to be at a more central level, possibly involving letter and word processing (e.g. Patterson and Kay, 1982; Kay and Hanley, 1996).

3.3.2 The deficit at a general perceptual level

A number of studies have claimed that the fundamental impairment in LBL reading occurs early in processing, prior to the activation of an orthographic representation. In this context the deficit appears not to be specific to orthographic material, but extends to other kinds of perceptual processing, namely to the processing of objects.

Behrmann and colleagues argued for a general perceptual problem. Sekuler and Behrmann (1996) studied 4 patients (MA, TU, DS and MW) to investigate whether their reading problems derived from a more general non-orthographic perceptual deficit. The patients (whose neuropsychological profiles were not reported in detail) showed a word length effect which varied in severity, as indicated by the different slopes of 1293, 541, 101 and 93ms/letter, respectively. The patients performed significantly more poorly than the control subjects on the Perceptual Fluency task (Ekstrom, French and Harman, 1976) which comprises 3 perceptual speed tests, namely the Finding As, the Number comparison and the Identical picture test. The first experiment was created to explore how the configuration of a nonorthographic object affects pure alexics' abilities to integrate parts. More specifically, they explored how patients' performance is affected by the number of parts (4, 5, 6) in an object and by the perceptual characteristics of the stimuli (good and poor configurations). In a second experiment they investigated how perceptual cues affect patients' abilities to integrate objects.

Although the aim of the study was interesting, the choice of the material was less convincing because stimuli were too "artificial". For example, in the second experiment, subjects made same/different judgements on the number of bumps that appeared at the end of two overlapping bars with one bar partially occluding the other. However, the results showed that patients are able to form a unified percept but only when the figure is continuous or symmetrical. Overall, the problem does not seem to be one of integrating parts per se; rather the difficulty manifests itself under impoverished perceptual conditions in which there is less support from organizational cues for representing the display. According to the authors, the results imply that pure alexia is most likely to arise from a general, nonorthographic deficit and that the nature of the disorder is revealed when the perceptual context lacks strong perceptual cues.

To further test their hypothesis Behrmann, Nelson and Sekuler (1998) administered some tasks to 6 patients (EL, MA, DS, MW, DK and IS). The patient EL who performed well on the Benton facial recognition test as well as on the VOSP showed a weak performance on the Perceptual Fluency test (Ekstrom, French and Harman, 1976). EL was given 255 of the 260 pictures from the Snodgrass and Vanderwart's set (1980). She made just a few mistakes (the difference is not significant relative to controls), although there is a significant difference in identification times between EL and the two control subjects. There is a great increase in EL's RTs both as familiarity decreases and visual complexity increases.

As another experiment, a selected subset of 40 low and 40 high-complexity items taken from the Snodgrass and Vanderwart set was presented to the other 5 LBL readers. Patients made just a few mistakes (no difference in errors between patients and controls), but there was a significant difference in RT between the two groups, with the patients' RT being on average 595 ms slower than the control subjects. Visual complexity did not affected RT in all the analyses. However it has not been controlled whether slowed RTs can be due to the visual field defects (hemianopsia or upper quadrant hemianopsia) shown by most of the patients. According to the authors, these findings challenge the traditional view that pure alexia is a 'pure' deficit and suggest that pure alexia emerge from a disorder to a more general-purpose cognitive mechanism. They argued that since reading is a relatively new cognitive ability, it is likely that it is mediated by a neural substrate that subserves other visuoperceptual functions and that this general purpose system has recently been recruited to mediate the processing of alphanumeric symbols.

In 1998 Behrmann, Plaut and Nelson developed a theoretical account of letter-by-letter reading (LBL) which tries to reconcile the experimental findings of both impaired letter processing and of lexical/semantic effects on reading performance. They assume that there is a general perceptual deficit that prevents the identification of words and they adopted the Interactive Activation Model of letter and word perception as a framework (McClelland and Rumelhart, 1981; Rumelhart and McClelland, 1982) to explain both normal reading as well as LBL reading. They suggest that LBL reading is the result of the remaining capacity in the normal reading system, which is bilaterally distributed, cascaded and interactive. In a cascade system, partial information about a stimulus can be passed on to higher levels in the system and since the system is also interactive, activation on the superior level will feed back to preceding levels. Thus, in LBL readers the reading system is unchanged from its premorbid state, and the same reading system functions and is governed by the same computational principles for normal readers and for alexic patients.

On their account, the deficit in pure alexia lies either at the feature level or between the feature level and the letter level. Nonetheless, the weak activation propagates in a cascaded and interactive way to higher-levels to engage lexical and semantic representations which in turn provide top-down support for letter and word identification. This lower-level deficit does not suffice for identification of the word and results in sequential processing of letters. The strength of the top-down support is held to be related to the length of time to read the word: in patients taking longer to process words with more letters, lexical/semantic activation has more time to accumulate and influence the degraded letter activation output. The straightforward predictions are first, that high-frequency or high-imageability words are likely to be read more quickly than those with low-frequency or low-imageability. Secondly, that there should be an interaction between frequency/imageability and word length. Thirdly, since partial word information is passed on to the lexical or semantic level, this information might be sufficient to allow the patient to make lexical or semantic judgements about stimuli they cannot explicitly identify.

In support of their position they presented a review of 57 published cases of LBL reading in which they documented the existence of an early deficit and simultaneously a higher-level processing in word reading. It is noteworthy that by "early deficit" Behrmann et al. (1998) meant a deficit in letter processing, that could be specific for the orthographic material and not necessarily involve object processing. In any case, a problem at letter level was observed in almost all the patients reviewed. In fact although some of these patients were accurate on single letter naming, the time to name the letters was often abnormally long. They assume that there is strong evidence for a peripheral deficit that affects letter processing and brings the patients to make both overt and covert gaze shifts to enhance letter activation. As far as concerned the evidence for lexical and semantic effects, only a few patients had been tested on the higher-order variables, mainly on word naming and on tasks of implicit reading. Patients usually are tested with different tasks or different variables are taken into account, thus it is difficult to make comparisons.

However, across a population of 57 subjects, only 13 subjects did not show any lexical or semantic effects on any of the measures. On this account the severity of the deficit may play a crucial role in explaining the differences among LBL readers of the extent of the lexical/semantic effects. There may be an inverted-U function relating the severity of impairment to the strength of higher-level effects observed in the LBL readers. When the visual input is well processed, top-down support is largely unnecessary and when the deficit is too severe higher-order representations are not strongly engaged. It is only in the middle range, when the orthographic input is still sufficiently intact that the later lexical effects become apparent. However as they themselves note, this account may be too simple since the correlation between the degree of peripheral impairment and reading performance is not linear (Hanley and Kay, 1996).

As described, the conclusions of their work are: 1. with their unitary account of LBL reading they have reconciled both a deficit in letter processing and the presence of lexical/semantic effects observed on reading performance of pure alexic patients; 2. the letter processing deficit is attributable to a general perceptual deficit that is common to all the patients; 3. in the context of a cascaded, interactive system it is possible to account for normal reading and LBL reading. More specifically, the sequential LBL reading shown by the patients arises from the residual function of the normal reading system that probably involves both the left and the right hemispheres.

Chialant and Caramazza (1998) have taken a position that appears close to the accounts of a peripheral perceptual impairment. They describe a patient MJ and show that she is a pure LBL reader, with a reading deficit not related to her visual field cut. She takes about 4sec to read a 6-letter word and 7.5sec to read a 9-letter word. In object naming she was assessed on the Snodgrass and Vanderwart set of pictures (1980) in different conditions: in free vision, at 250msec of exposure duration and at 100msec. In most of the tasks her performance has been compared to one normal control and to a hemianopic patient. MJ's performance was flawless in the unlimited exposure duration condition and still good (84 percent accuracy) at 100ms of exposure duration. This result indicate that she does not have evident perceptual impairments. Her abilities in single letter processing are intact, while her visual span is reduced, under presentation of linguistic as well as nonorthographic stimuli. In fact when MJ was tested on a line orientation detection task, she performed less accurately than the normal control, but similarly to the hemianopic patient: her performance was affected by both the absolute and the relative position of the items in visual space.

Chialant and Caramazza (1998) found no evidence of lexical or semantic access in tasks of brief presentation of words. MJ was able to extract some visual information from briefly presented words but her above-chance performance on a semantic categorization task was more likely to reflect a sophisticated ability to guess than a true lexical-semantic access.

The authors interpreted this pattern of results as indicating that LBL reading (at least in MJ) results from damage to prelexical or perceptual processing mechanisms. However, a possible account in terms of a more general, perceptual deficit is not strongly supported by the results. MJ's performance was very good on object naming also when the object presentation made the task more difficult. Moreover the fact that MJ's performance on the line orientation detection task was more impaired than the hemianopic control might suggest that the right visual defect (RHH) has a critical role, independently from the pure alexic deficit.

3.3.3 The deficit at a central, lexical level

Warrington and Shallice (1980) introduced the concept of the visual word-form unit where orthographic categorization occurs. After a first stage of visual feature processing, the letter string is grouped into integrated perceptual units, that are different levels of the visual word-form system (Shallice and McCarthy, 1985). These representations are held to be utilized by both phonological and semantic reading routes (see Fig. 3.1). The word-form system is responsible for parsing letter strings into recognizable units which can range in different sizes as graphemes, syllables, morphemes and words (letters being the default option, in case the system fails to find larger units).

Warrington and Shallice (1980) held this system to be damaged in pure alexia. In fact the authors reported the case of two pure alexic patients, RAV and JDC, both with temporo-parietal lesions. Their perceptual skills appeared entirely normal: they did not have difficulties in interpreting complex pictures and the recognition of objects in unconventional views was preserved. The patients, tested on visual short-term memory tasks, showed relatively intact visual span of apprehension for letters and numbers; moreover their selective attention was normal. However the capacity for whole word reading in the two patients was either absent or so impaired as to prevent normal reading.

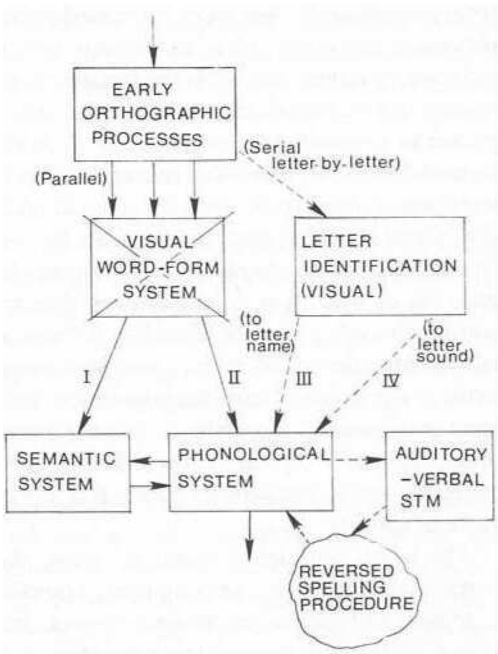


Figure 3.1: The word-form system (Warrington and Shallice, 1980).

Two techniques were employed to maximize the possibility to use whole word reading, by attempting to suppress a LBL reading strategy. Script reading and tachistoscopic presentation of words, displayed for 100 and 200ms. The results showed that in the absence of visual-perceptual deficits, the reading of whole words was markedly impaired for both RAV and JDC.

Patients described as being able to read only ”letter-by-letter” were already reported by Kinsbourne and Warrington in 1962 in the context of simulanagnosia. The authors noticed that the capacity to perceive single forms was preserved in 4 patients, while their ability in recognize 2 or more forms simultaneously presented was impaired. In this case the reading impairment, also called ”spelling dyslexia”, was caused by a limitation in perceptual processing.

As regards the impairment of RAV and JDC, Warrington and Shallice (1980) hypothesized 3 different alternatives to account for the functional basis of the phenomenon. The first hypothesis was that LBL-reading was due to a general simultanagnosia, that is the failure in either perceptual processing (Kinsbourne and Warrington, 1962) or comprehension (Wolpert, 1924) of the whole visual display as an entity. However complex picture interpretation was relatively unaffected in both the patients, indicating that LBL reading can be selectively impaired in simultanagnosia and pure alexia. A second possibility was that the LBL reading strategy resulted from a failure in the transmission of information from the RH to the LH, the traditional disconnection account (Déjerine, 1892). However, patients had a good letter span, so the explanation of their word reading difficulties in terms of the disconnection theory seems most implausible. Thus, Warrington and Shallice (1980) hypothesized that the deficit is at the visual word-form system and that their patients no longer perceive higher-level perceptual units in written words. In this view, the word-form must be reached prior to phonological and semantic analyses. These authors also held that the letter-by-letter (LBL) reading strategy involves the

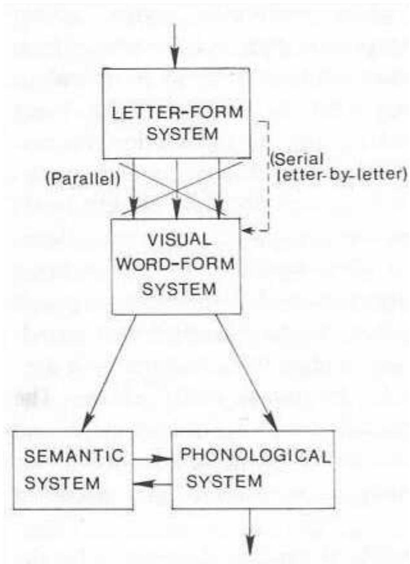


Figure 3.2: Patterson and Kay, 1982.

spelling system operating in reverse (from letters to word): reading is considered to be possible only after naming each letter either covertly or explicitly.

As far as the functional origin of pure alexia is concerned, Patterson and Kay (1982) used Warrington and Shallice's model (1980) to explain pure alexia but proposed an alternative to their account of the LBL reading strategy. They suggested that pure alexia arose from a disconnection of letter-form analysis from the word-form system which they held to result in an impairment in parallel letter identification. On this approach patients are able to access the word form system although in a different way from normal readers, by means of serial rather than parallel letter identification (see Fig. 3.2).

More specifically, Patterson and Kay (1982) divided their 4 cases of pure alexia in two types: Type 1 patients (CH and MW) had difficulty in identifying letters, while Type 2 (TP and KC) suffered from a lexical level impairment in addition to a letter level deficit. However, using Shallice's model (1981b) they held that the deficit for all the patients lies between the letter-form analysis and the word-form system, but in addition there is a deficit in the word-form system for Type 2 patients. However, if all the patients often misidentify letters (see p. 429-430), it remains unclear why they should have sustained a disconnection deficit (instead of a letter processing impairment) and so whether the letter-form level was indeed intact. Finally, in contrast to Warrington and Shallice (1980) who suggested the use of reverse spelling as a compensatory strategy in reading, Hanley and Kay (1992) hypothesized that during reading the letter names are converted into visual images and these representations access the visual word form system. Therefore no phonologically based strategy is involved.

3.3.4 Other theories

Rapp and Caramazza in 1991 argued for an impairment in visuospatial attention to explain their patient's pure alexia, where the deficit impairs the distribution of attention across all spatial locations following a left-to-right gradient of processing. They proposed a reading model where three distinct levels of representation are involved: a retino-centric feature level, a stimulus-centered level and a word centered level. Their patient's deficit was interpreted as a perceptual prelexical impairment at the retino-centric and stimulus centered levels of representation, with a left-to-right processing difficulty affecting both levels. Because of this gradient of impairment, attention was supposed to be allocated sequentially to each location, with the result of a LBL reading pattern.

A few years later, Behrmann and Shallice (1995) conducted an exhaustive investigation of the pure alexic patient DS to explore whether Rapp and Caramazza's hypothesis (1991) could account for their case. DS was a 34-year-old female with an upper quadrant hemianopsia following an ischaemic stroke in the left occipital lobe. She was a mild LBL reader: she took 1.3 sec to read a 5-letter word and about a 1.6sec to red a 7-letter word. On a word reading task she showed a frequency effect, but not an effect of imageability. She was assessed on several, in-depth tasks of spatial and letter processing. The results showed that DS was able to distribute attention across multiple locations and that her performance was unaffected by the absolute or spatial location of the letters. Instead, on tasks of letter processing, she showed a slow or reduced letter activation which disrupted the rapid and efficient processing of single letters. The main problem is a non-spatial, visual impairment that affects the activation of individual letters. As a consequence, the patient's deficit was held to arise in the processing of single letters, independently from spatial location. An impairment of visuospatial attention did not therefore appear to be a valid explanation for DS's deficit and more generally, a characteristic of pure alexia.

Arguin, Fiset and Bub (2002) also suggested that LBL reading is caused by an impairment in letter encoding. This explanation could appear similar to the position held by Behrmann et al. (1998), but in this case the deficit is not a general perceptual one. According to the position of Arguin et al. (2002), the residual capacity of parallel letter processing that contributes to overt word recognition provides uncertain information about letter identities to the level of lexical-orthographic representations: this results in an activation of several possible words compatible with the degraded letter input. The parallel letter analysis is incapable of providing unambiguous information on word identity and it is held to result in sequential letter analysis for overt word recognition. In one experiment they examined the effect of letter confusability (visual similarity among letters) in their patient IH in relation to the sequential letter analysis, i.e. the word length effect. Letter confusability is defined as the shape similarity between a particular target letter and the remaining letters of the alphabet, with confusability values determined from empirical letter confusion matrices obtained on previous studies with normal subjects (Gilmore, Hersh, Caramazza and Griffin, 1979; Loomis, 1982; Van der Heijden, Malhas and Van den Roovaart, 1984). The results showed independent effects for the two factors (letter confusability and word length), suggesting that letter confusability has no impact on the modulation of the magnitude of the word length effect. Arguin and Bub (2005) replicated these observations with another 3 LBL readers. They found that for all the 4 patients the facilitatory effect of increased N size (the number of orthographic neighbours of the target word) on word recognition did not occur with high letter-confusability words. A facilitatory N size effect requires parallel letter processing; therefore the residual capacity for parallel letter processing is blocked by letter similarity. Thus they argue that the residual ability for parallel letter processing is blocked by letter similarity, which implies a deficit of letter identification.

3.3.5 One or multiple causes in pure alexia?

One of the issues that is central to understanding pure alexia is whether there are one or multiple sources of deficits, namely whether patients so classified can have different functional deficits. However, just a few authors have dealt with this aspect directly.

It appears that only Price and Humphreys (1992) took a clear position on this issue and it was against the hypothesis of a single basic type of impairment (as for example claimed by Behrmann et al., 1998). In fact Price and Humphreys (1992) suggested that the deficit could be at any level of visual processing which would prevent normal access to lexical, semantic and phonological knowledge. They claimed that their 2 patients had different functional deficits from one another and that the abnormally large word-length effects were not necessarily a consequence of the same compensatory reading strategy.

Price and Humphreys (1992) hypothesized that the patient EW had a deficit in parallel feature processing, which limited her ability to access lexical or semantic information from words she was unable to read. This type of deficit would account for the effects of word length irrespective of exposure duration and for the deterioration of reading as the exposure duration was reduced, as she requires abnormally long exposure durations to discriminate the features of both letter and number strings and her performance is affected by the visual discriminability of the stimulus. In a subitisation task, where patients are asked to report the number of dots in visual displays presented for brief exposure durations, her ability appeared intact, namely her performance did not change as a function of the number of dots. However her performance did not even change across exposure duration: this is unexpected, since her deficit is supposed to be in parallel processing. According to the authors, EW would have a deficit in parallel processing (but surprisingly not in the subitisation task), while her ability to direct attention would be intact. Moreover Price and Humphreys (1992) hypothesized that EW shows an abnormally strong effect of word length because 1) her poor parallel feature processing results in longer words taking more time to reach the threshold for identification than short words; 2) there is implicit serial processing, when EW rapidly scans her attention over subword units in order to construct a visual representation of the word; 3) there is explicit serial processing, when EW names individual letters making up the word.

By contrast, the other patient HT did not have impairment in parallel feature processing. In fact his reading accuracy was unaffected by word length at exposure durations of less than a second, indicating that his performance was not affected when increased demands were placed on parallel letter identification. However, according to the authors, HT has a deficit in orienting attention because at short durations he performed weakly and he was more impaired with irregular configurations that required him to count the dots, thus indicating an impairment in scanning attention covertly. So according to the authors, when HT fails to engage his attention appropriately, he also fails to access lexical, semantic or phonological information. Moreover Price and Humphreys

(1992) hypothesized that HT uses an explicit serial processing strategy on the words he fails to read from parallel letter identification. Such failures occur when HT fails to engage attention appropriately within a word. However it is less clear why his reading performance did not worsen when exposure durations decreased if his impairment was in orienting attention appropriately.

As a result, Price and Humphreys (1992) hypothesized that these 2 cases of pure alexia were caused by different functional deficit and that different compensatory strategies could be used by the patients, although the results were not so clear-cut.

Another position which included the possible existence of different functional deficits was held by Patterson and Kay (1982) and Hanley and Kay (1992). As discussed before, Patterson and Kay (1982) studied 4 pure alexic patients, whose reading speed was generally slow: the quickest patient (TP) needed over 7 s to identify a 3- to 4- letter word and 19.5 s to identify a 9- to 10-letter word, whereas the slowest patient (CH) needed 16.9 s with 3- to 4- letter words and 97.3 s to identify a 9- to 10- letter word. The authors claim that the impairment shown by their patients is specific to reading and cannot be due to a general simultanagnosia. Despite the use of multiple techniques, no evidence was obtained for the hypothesis that comprehension of a word could occur in the absence of the LBL analysis required for oral reading. However, on more detailed investigation major qualitative differences between their impairments emerged.

The systematic study of the pattern of reading errors made by the patients revealed that all the 4 LBL readers tend to make 'clear letter misidentification' mistakes e.g. men: "h,e,n... hen". More interestingly, the error category 'letters right word wrong' e.g. head: "h,e,a,d... heed" divides the patients into 2 groups, MW and CH are Type 1 LBL readers and TP and KC Type 2. MW and CH produce many incorrect reading responses because of inaccurate letter recognition, but they rarely make errors once they have identified all the letters in a word. TP and KC, on the other hand, will sometimes read the word incorrectly, even when they have successfully identified all the letters in a word. Using the model developed by Warrington and Shallice (1980) they held that the impairment for all their patients lays to the connections between the letter-form analysis system and the word-form system, but in addition there was a deficit at the word-form system for Type 2 patients. However, as it will be discussed in Chapter 4, it is not clear whether the letter-level (including letter-form) was intact in all the 4 patients (see also Hanley and Kay, 1992).

By contrast, as described before, the Behrmann et al. model (1998) implicitly claims that pure alexia arises from a single type of functional origin, a generalised problem of lower-level orthographic processing which leads through cascade-type consequences to difficulties at higher word-form levels. As reported in their article, Behrmann et al. (1998) implicitly assume that a serial LBL reading process occurs as a consequence of the low-level deficit: ”This weak activation does not suffice for explicit identification of the word [] and the system must resort to sequential processing to enhance the activation of individual letters” (page 14) and ”The slope of the reading function in our account is determined by the severity of the lower-level deficit” (page 42-43).

Although Behrmann et al. (1998) have presented an account of pure alexia which reconciles the impairment in early visual processing with lexical and semantic effects on reading, their model gives only a partial explanation of the variability of performance that patients show. The explanations put forward to account for the variability across patients are i) methodological: patients are studied with different tasks, for different aims. The sparse data may be exaggerating the difference between patients. This may be a plausible and interesting possibility to take into account, although it is unlikely that it could explain the whole variability by itself. ii) The severity of the deficit which might determine the extent of the lexical/semantic effects: when the visual input is well processed, top-down support is largely unnecessary and when the deficit is too severe, higher-ordered representations are not strongly engaged. It is only in the middle range, when the orthographic input is still sufficiently intact that the later lexical effects become apparent. However, as they themselves noticed, in their account the slope of the reading function is determined by the severity of the lower-level deficit, but the correlation between the degree of peripheral impairment and reading performance is far from linear (Hanley and Kay, 1996). Thus, this explanation does not appear to be exhaustive. iii) The strategies that patients might employ in compensating for their peripheral impairment. Particular strategies can diminish and even eliminate lexical/semantic effects. For instance the patient JWC (Coslett et al., 1993) appeared to be able to use two distinct strategies, a serial LBL strategy and a "whole-word" strategy according to the task he was asked to do. Although Behrmann et al. (1998) considered these as possible explanations for the variability observed in LBL readers, they conclude with the claim of the unitary account of LBL reading: "a deficit in letter processing (perhaps attributable to an even more fundamental perceptual impairment) is common to all LBL readers" (page 45-46).

Having taken into account the different theoretical positions on the existence of one or multiple functional causes of pure alexia, it appears clear that this issue is still highly controversial. Several accounts have been provided to explain the functional deficit of pure alexia; moreover it remains possible that pure alexia cannot be reduced to a unique form, by studying it more carefully.

3.3.6 Role of the right hemisphere in reading

The role of the right hemisphere in affective processing of language and in the pragmatics of discourse is well accepted (Bryden and Ley, 1983). The right hemisphere appears to be specialized for the expression and perception of prosody and emotional content. One important and controversial issue regarding reading concerns the putative reading capacity of the right hemisphere (RH). In recent years several lines of evidence have suggested that the right hemisphere possesses the capacity to read. One strong line of evidence comes from the performance of a patient who underwent a left hemispherectomy at age 15 for treatment of seizures (Patterson, Vargha-Khadem and Polkey, 1989). After the operation the patient was able to read approximately 30 percent of single words and exhibited an effect of part of speech; however she was probably unable to use print-to-sound conversion. The performance of some split-brain patients is also consistent with the claim that the right hemisphere is literate (see deep dyslexia in Section 2.2.2). Another line of evidence comes from patients with pure alexia and optic aphasia (Coslett and Saffran, 1989a; 1989b).

The first extensive report of covert reading in pure alexia has been provided by Shallice and Saffran (1986). Their patient, ML, was able to carry out lexical decision and semantic categorization on words that were presented too briefly to support LBL reading. Notable features of ML's performance include the following: above-chance but far from perfect performance on lexical decision, with error rates dependent on word frequency and the similarity of nonwords to words; performance ranging from 70 to 94 percent correct on binary choice categorization tasks, where the categories included countries (in vs out of Europe), occupations (author vs politician) and objects (living vs nonliving). ML was also able to locate places, corresponding to names rapidly presented on a map of Britain. In contrast he was rarely able to report explicitly a word presented briefly. Another notable feature of ML's tacit reading behavior was a lack of sensitivity to the appropriateness of affixes (e.g. windowing, strongs, galloply).

A few years later, Coslett and Saffran (1989a) reported the data from 4 pure alexic patients who performed well above chance on a number of lexical decision and semantic categorization tasks with briefly presented words that they could not explicitly identify. Three of the patients who regained the ability to identify rapidly presented words explicitly exhibited a pattern of performance consistent with the right-hemisphere reading hypothesis: nouns read better than functors, words of high better than words of low imageability. More specifically Coslett and Saffran (Coslett and Saffran 1989a; Coslett and Saffran 1993; Saffran and Coslett, 1998) investigated the contradictory behavior that some pure alexic patients show on tasks of letter identification and of word processing under condition of brief word presentation.

According to the authors, letter identification poses a significant problem for LBL readers, but this impairment is difficult to reconcile with the evidence that some patients are capable of processing printed words presented rapidly, although they are generally unable to report them. Coslett and Saffran (1989a, 1993, 1998) claim that the intact right hemisphere (RH) possesses the capacity to read, but the damaged left hemisphere (LH) can only process information transmitted by the RH slowly and serially. Thus, LBL reading is a compensatory strategy adopted by the LH, which supports explicit identification of printed words. The covert reading shown under rapid presentation conditions is assumed to be the product of a different reading system in the RH, which does not interface directly with LH mechanisms for language production.

In general, all these data together are consistent with the hypothesis that the right hemisphere is not word-blind but may support the reading of some types of words. The full extent of this reading capacity and whether it is relevant to normal reading, however, remains unclear.

3.3.7 Neuroanatomical localization

The anatomic basis of pure alexia has been extensively investigated. The systematic analysis of reading disorders commenced with the work of a few neurologists in the late 19th century as reported in chapter 1. Neurologists such as

Charcot and Wernicke noted that reading could be impaired selectively by cerebral lesions. The fundamental contribution in this clinical-pathological tradition was the work of the neurologist Jules Déjerine. In 1891 Déjerine described a patient with an impairment in reading and writing which he termed ”alexia with agraphia” following an infarction in the left parietal lobe.

The year after, Déjerine (1892) described a second patient, a 68-year-old man, referred as Monsieur C, who abruptly lost the ability to read after a cerebrovascular accident. According to Déjerine, Monsieur C’s reading impairment was total: ”The patient did not recognize a single letter, not a word, except his own name on occasion” (Bub, Arguin and Lecours, 1993). However, he could still write and spell without error and did not have difficulties in identifying other type of visual stimuli, including numbers. Afterwards Monsieur C suffered another stroke that left him with impaired spoken language and a total agraphia. He died a short time after his second episode. At postmortem, the patient’s brain showed gross infarction of the left occipital lobe and the splenium of the corpus callosum (Déjerine, 1892). The damage produced by the more recent cerebrovascular event was confined to the posterior and inferior part of the left parietal lobe, the angular gyrus (see Fig. 3.3).

Déjerine designated the disorder as ”alexia without agraphia” and from the analysis of this important case, together with the other one with ”alexia and agraphia” he enunciated the initial theoretical anatomic basis of pure alexia. Déjerine hypothesized that the angular gyrus contained a center that was crucial for reading and writing, a center where the ”optical image of letters” would be located. For Monsieur C, the left occipital damage must have disconnected the ”center of the optical images for words” from both visual cortices. The damage to the visual pathways from both the LH and RH, on their way to the angular gyrus would affect reading but not writing, since the angular gyrus itself could still arouse the optic image of letters necessary to guide the movements of writing (see Fig.3.4).

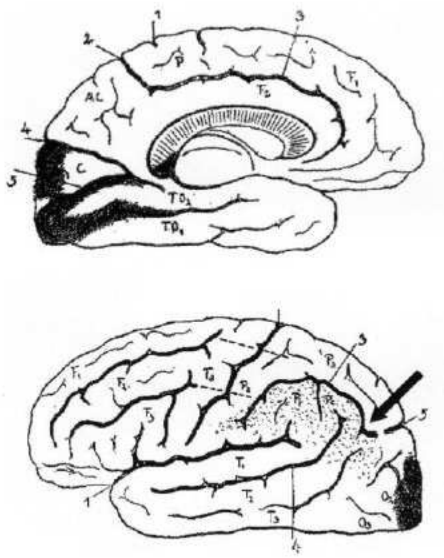


Figure 3.3: Déjerine's original lesion drawing of Monsieur C (1892).

Déjerine explicitly stated that he did not think the tiny lesion of the splenium of the corpus callosum should be considered to play an important functional role in the syndrome. The damage to the white matter tract from the left occipital lobe was sufficiently great to disrupt the communication of both visual cortices to the angular gyrus without requiring any additional speculation on the effect of the splenial damage.

The explanation of pure alexia put forward by Déjerine in terms of a disconnection syndrome remains credible to this day. For instance, in one of the cases we have studied (patient FC, see Chapter 4), we argue, has a disconnection syndrome, as his paraventricular white matter lesion has compromised both intrahemispheric and interhemispheric visual pathways. However, the idea of a ”visual memory center for words” in the angular gyrus has not been confirmed; an area particular responsive to visually presented words, if anything, is in the occipito-temporal gyrus (Cohen, Dehaene and colleagues, 2000, 2002, 2003). The importance and the influence of Déjerine's work is due to the details

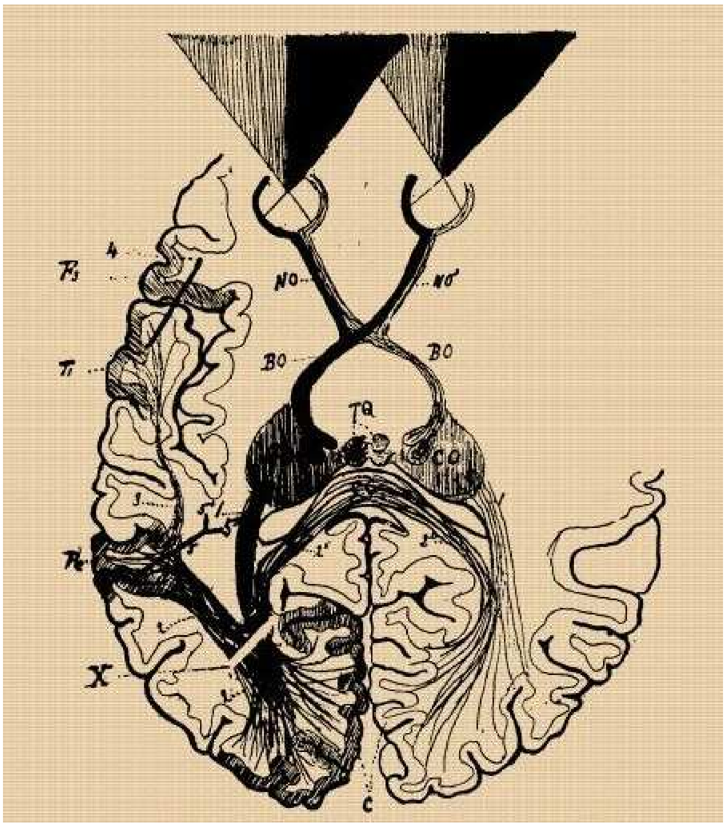
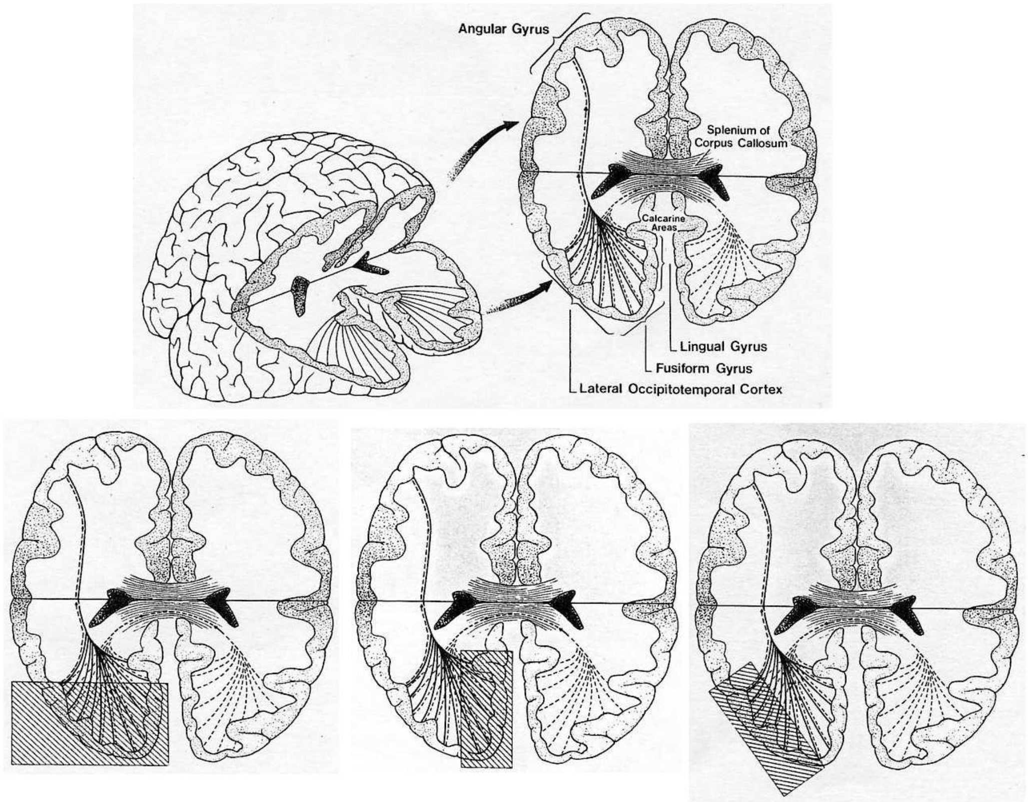


Figure 3.4: Déjerine's (1892) account of pure alexia. The disconnecting lesion is marked with an X and was hypothesized by Déjerine to produce pure alexia.

of the behavioral evidence and the force of the neuroanatomical arguments.

The emphasis on the splenium as a major factor of pure alexia has been attributed to Geschwind (1965). Another important contribution to the study of pure alexia was given by Greenblatt (1976, 1983, 1990). In 1990 Greenblatt reported two cases of pure alexia. The first case, a 55-year-old man developed pure alexia after a dominant occipital lobectomy following a glioblastoma in the left posterior parieto-occipital area. Although the patient recovered some reading abilities, this process was never efficient. Case 2 was a 19-year-old male who had a more restricted, occipital lobectomy for an occipital vascular malformation. Greenblatt (1990) argued that the patient recovered efficient reading after 15 months (although it is not clear how normal his reading was). Unlike Case 1, the surgical lesion removed a dorsal and medial occipital region, sparing some inferolateral cortex of the left occipitotemporal area. The observation of these cases brought Greenblatt (1990) to hypothesize the existence of two separable syndromes: a "hololobectomy syndrome" (Case 1) and a "medial lobectomy syndrome" (Case 2).

According to Greenblatt (1990), Déjerine's (1892) original model of reading assigned critical roles to the lingual and fusiform gyri in the occipital lobes on both sides (see Fig. 3.4): visual signals arriving in the right calcarine cortex are transmitted to the adjacent right lingual and fusiform gyri. From there, the signals go through the ventral splenium to the homologous left lingual and fusiform gyri following "Flechsig's rule", according to which information is transferred across the corpus callosum only between homologous association areas (Flechsig, 1901). As reported by Bub, Arguin and Lecours (1993), Déjerine argued that "the damage must be located in such a way as to disrupt the connections of both occipital lobes to the angular gyrus" (p. 552). But the possibility that reading could eventually recover quite well in the medial occipital lobectomy syndrome (Case 2) was not entirely consistent with Déjerine's account. Therefore some alternative pathway and/or a larger cortical area (with



Hololobectomy syndrome | Medial lobectomy syndrome | Lateral lobectomy syndrome

Figure 3.5: Greenblatt (1990): different types of syndromes (see text for lesion details).

also the temporal region) must have been involved.

Thus, Greenblatt proposed that the visual input arriving in the right occipital cortex is transmitted to the right occipito-*temporal* cortex (see Fig. 3.5, superior part); then visual signals cross to the homologous association areas in the LH through the ventral splenium (dashed lines in Fig.3.5). Here RH input is joined by the input arriving from the left calcarine area (solid lines in Fig. 3.5). The combined signals are transmitted to the left angular gyrus from occipito-*temporal* cortex, probably via the vertical occipital fasciculus. In Case 1, where the medial and lateral aspects of the ventral occipito-temporal cortex were removed, the visual input from the RH had very little association cortex with which to connect in the LH. The patient's partial recovery of reading can probably be attributed to some remaining function in the left ventral occipital lobe, but it was insufficient to mediate normal, automatic reading.

In Case 2, by contrast, a large portion of the left ventrolateral occipito-temporal association cortex remained intact. On this account, Greenblatt (1990) predicted a third syndrome of alexia, after a left ventrolateral occipital lobectomy. Such patients should show considerable recovery of reading if the original lesion and the surgical removal are limited to the lateral aspects of the LH. Finally, the disconnection syndrome predicted by Déjerine's model has been named by Greenblatt (1976, 1983) as ”subangular alexia”, where a pure subcortical lesion disconnects the left angular gyrus from the entire left ventral occipital cortex. The work done by Greenblatt (1990) is important because it defines more carefully the anatomical pathways necessary to read, although he does not give his opinion on a more theoretical model of word recognition.

The case of pure alexia FC, we have investigated (see Chapter 4), might fit with the medial lobectomy syndrome here described by Greenblatt (1990), because the lateral occipito-temporal cortex was intact. Moreover FC is a mild LBL reader and the quickest among the 5 patients here studied although he never did recover finally. However as reported before, in our case a further white matter lesion was crucial in the resulting pure alexia.

Ajax, Schenkenberg and Kosteljanetz in 1977 described a new case of alexia without agraphia, documenting it with remarkable anatomic details. The 59-year-old patient had a right homonymous hemianopia; his language functions and cognitive abilities were intact, as well as the capacity to name objects. Single letter naming was usually spared; however, reading words of one syllable was difficult and polysyllabics were impossible for him to read. He showed difficulties with arithmetic tasks, presented visually and orally. His lesion involved an extensive infarction of major elements of the left occipital lobe, the inferior one third of the left forceps major and the contiguous splenium of the corpus callosum. The entire left lingual gyrus and the posterior half of the fusiform gyrus were also involved, as well as the subjacent white matter. In addition there was involvement of the posterior part of the left parahippocampal gyrus and the major portion of the left hippocampus. The left angular and supramarginal gyri were uninvolved.

According to Ajax et al. (1977) in this patient the left occipital lobe was largely destroyed, but only one third of the splenium was involved, therefore the disconnection of the left angular gyrus from the intact right calcarine cortex was limited. Since the patient retained some reading abilities, they assumed that the unaffected splenium or anterior callosal fibers were of some importance in the transmission and recognition of orthographic stimuli. On this account, even destruction of inferior elements of the left peristriate cortex alone appears capable of producing alexia without agraphia.

An important contribution to localization in pure alexia was the study reported by Damasio and Damasio (1983). In this study 16 patients identified on a series of standardized reading tests as having alexia without agraphia, underwent CT scanning and careful lesion analysis as well as neuropsychological and neuroophthalmological evaluation for colour vision and naming, memory, visual agnosia and optic ataxia. The lesion is associated with different

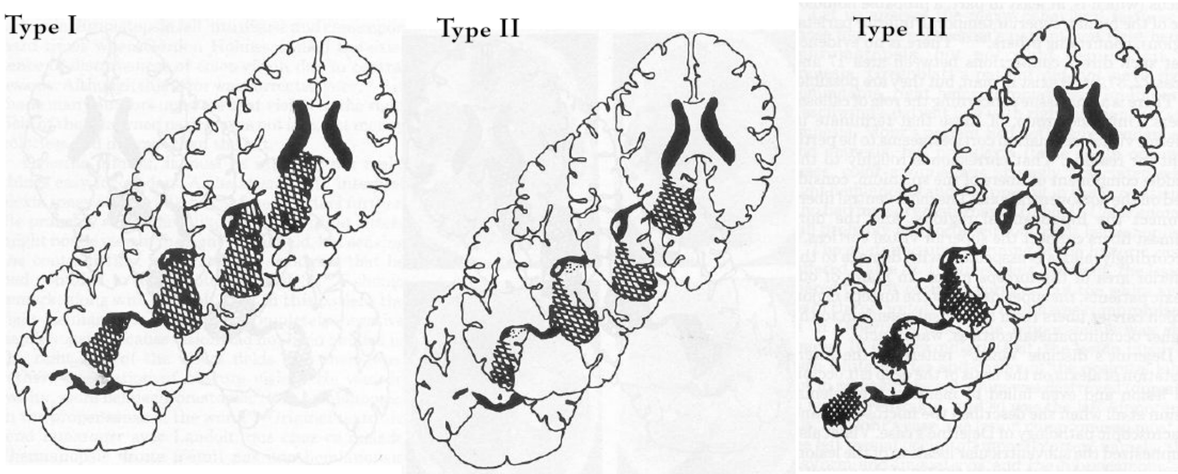


Figure 3.6: Damasio and Damasio (1983): three different types of pure alexia (see text for lesion details).

alexic syndromes were classified into 3 types. Type I (see Fig. 3.6), found in patients with RHH and colour anomia, involved extensive infarction of the posterior cerebral artery territory and included paraventricular damage of the white matter of the occipitotemporal junction, of the left half of the splenium and forceps major and of the inferior and superior medial-occipital cortex. In Type II (see Fig. 3.6), associated with RHH without colour anomia or verbal amnesia, paraventricual white matter damage interrupting interhemispheric pathways was noted in conjunction with damage to optic radiations, to the calcarine region or to both. Type III (see Fig. 3.6), alexia with an upper quadrant hemianopsia and lower acromatopsia (colour vision loss) demonstrated no colour anomia or verbal amnesia. The inferior optic radiation and inferior visual association cortex were involved, but paraventricular white matter damage was the critical lesion. Extension into the adjacent association cortex and splenium could occur but was not essential. The authors concluded that the crucial anatomic substrate for pure alexia was in the paraventricular white

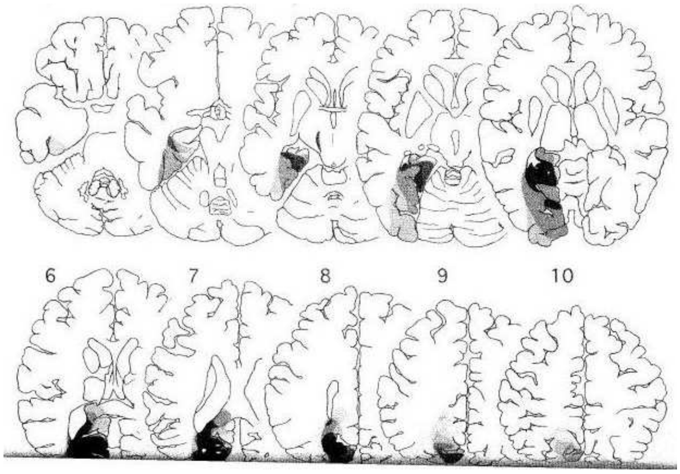


Figure 3.7: Binder and Mohr (1992): global alexia group (see text for lesion details).

matter region of the left occipital lobe. Such a lesion caused disconnection of transcallosal fibers from the right occipital region and of fibers from the left visual association cortex traveling to the left tempoparietal language area.

Binder and Mohr (1992) have made a further important contribution to the clinical-pathological study of pure alexia. They examined the lesion sites of 17 patients with left posterior cerebral artery infarctions and divided them into 3 groups: those with normal reading ability; ”global” alexics who were unable to read at all; and spelling alexics or LBL readers. The patients with preserved reading had lesions in the medial and ventral occipital lobe, involving lingual and posterior fusiform gyri, white matter beneath the occipital horn, but sparing dorsal white matter pathways and the ventral temporal lobe. Critically, the ventral temporal lobe was intact as well as the dorsal white matter pathways running over the occipital horn of the lateral ventricle.

Global and permanent alexia occurred only with additional injury to the splenium and forceps major, to the white matter above the occipital horn of the lateral ventricle and to the ventral temporal lobe (see Fig. 3.7). In these cases

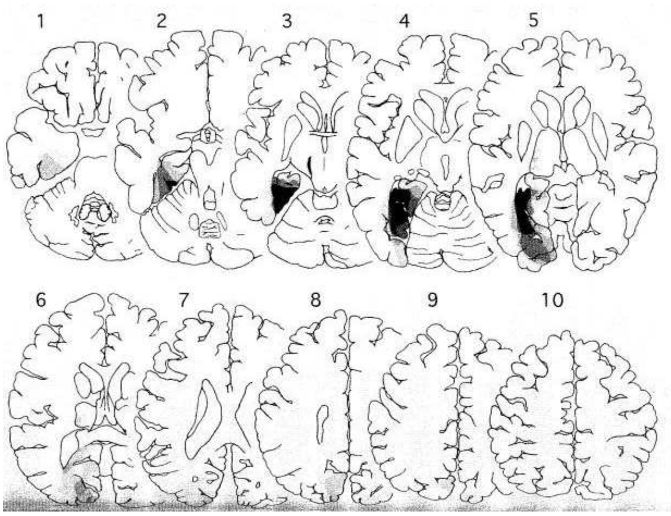


Figure 3.8: Binder and Mohr (1992): spelling dyslexia group (see text for lesion details).

it is reasonable to assume that the lesion blocked the transmission of visual information from the RH to the LH, including letter information. The patients identified as patients with spelling dyslexia had large lesions of the ventral temporal lobe, in particular both the cortex and white matter of the inferior temporal and anterior fusiform gyrus, in addition to the lesions in the medial and ventral occipital lobe (see Fig. 3.8).

These data suggest that the callosal pathways mediating reading follow a course over the top of the occipital horn and have little connection with the ventromedial occipital region. Binder and Mohr (1992) claim that if any callosal projections run inferomedial to the occipital ventricle, their data show that these are of little importance for reading, since this white matter lesion was commonly involved in patients in whom reading was normal. However, the authors acknowledge that there are some patients reported in literature whose lesion appeared to be in the ventral occipito-temporal region. For instance the case reported by Greenblatt (1973) concerned a patient with dyslexia from a

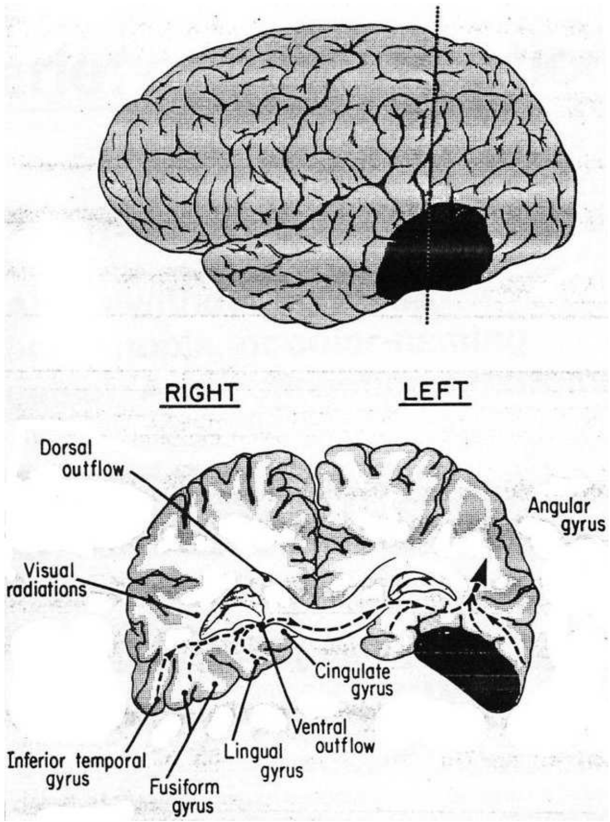


Figure 3.9: Artist's postsurgery reconstruction of meningioma compressing the inferior surface of the occipitotemporal junction. The left visual association cortex and inferior splenial outflow from right visual association cortex are distorted (Vincent, Sadowsky, Saunders and Reeves, 1977)

ventral occipital glioma. In another influential report by Vincent, Sadowsky, Saunders and Reeves (1977), dyslexia was associated with a meningioma producing compression in the ventral occipito-temporal region.

This patient and others with similar placed lesions (Rosati, De Bastiani, Aiello and Agnetti, 1984; Caffarra, 1987) had relatively mild reading deficits. Therefore the authors hypothesize that similar mild reading deficit might have resulted from lesions of the lateral occipital lobe (Greenblatt, 1990) and of the lateral occipito-temporal region, thus suggesting that reading pathways can be partially injured in a variety of locations on the lateral and ventral brain surface. Also our case FC (see Chapter 4) appears to have damage in the ventrolateral occipital region (in addition to the white matter lesion), suggesting that this area might be important for the callosal projections coming from the RH.

In conclusion, it is evident that the studies reported in literature on the anatomical basis of pure alexia and on the anatomo-functional pathways involved in reading have not arrived at conclusive results yet. From the study of his case, Déjerine (1892) hypothesized that the critical site in pure alexia was a lesion which disconnected the angular gyrus from the visual input of both the hemispheres. According to Damasio and Damasio (1983), the crucial lesion is in the paraventricular white matter region of the left occipital lobe. Here the damage can compromise both interhemispheric and intrahemispheric visual pathways. Finally Binder and Mohr (1992) indicated the cortical and subcortical region in the ventral temporal lobe as a critical lesion.

Compared to the more recent imaging findings, the anatomical studies carried out by Déjerine (1892) and by Damasio and Damasio (1983) have considered the white matter in the occipital and temporal regions as more critical than the correspondent cortical areas in pure alexia. The more recent studies by Greenblatt (1990) and Binder and Mohr (1992) gave equal importance to them. As will be described later, Cohen and Dehaene (2004) have shown that also purely cortical regions can result in pure alexia. However, it is worth considering that some discrepancies may derive from methodological differences in the reconstruction of the lesion: from the post-mortem studies, to the use of CT scans in the studies of Damasio and Damasio (1983) and Binder and Mohr (1992), to the use of magnetic resonance imaging in the more recent research. These very different tools may be sufficient to account for some important differences.

3.3.8 Imaging studies

The brain imaging research has highlighted functional brain activity during the normal (and pathological) process of reading, providing new insight into the neural circuitry of language. Word recognition is a field that has attracted much attention in cognitive neuroscience and many studies have been carried out to understand the functional architecture of our ability to read and its neural bases.

The first groundbreaking attempt to apply a functional brain imaging technique to the cognitive domain was made by Petersen, Fox and Posner (1988). By using positron emission tomography (PET), they examined functional activation during processing of words presented in auditory and visual modes to 17 normal right-handed volunteers. A further study with PET was designed to explore orthographic effects more carefully (Petersen, Fox, Snyder and Raichle, 1990). Four different sets of stimuli were used, including words, pseudowords, unpronounceable consonant letter strings and false font strings. Using the subtractive technique, lateral occipital (extrastriate) activation was noted with all 4 sets of stimuli. In addition, real words and pseudowords produced a left medial occipital extrastriate response. Subtraction of the real and pseudowords conditions showed no significant difference in the occipital region, but a significant activation in the left frontal cortex was seen in the real word relative to the pseudoword condition. The region activated was similar to that activated in the semantic processing tasks, suggesting that the lateral prefrontal region on the left is involved in semantic processing of single words.

As mentioned before, the ability to read is an issue particularly investigated in different conditions: in normal subjects, in children learning to read (Turkeltaub, Gareau, Flowers, Zeffiro, and Eden, 2003), in developmental dyslexia (Paulesu et al., 2001; McCrory, Mechelli, Frith and Price, 2003) and in acquired dyslexia (Cohen et al., 2003; Price, Gorno-Tempini, Graham, Biggio, Mechelli, Patterson and Noppeney, 2003). We are not going to consider all the studies reported on reading, rather we will take into account only those that are relevant for visual word recognition, in particular to identify the crucial area in pure alexia and to understand whether there are specific neural mechanisms which have become specialized for reading. In this view, two positions are critical: the Cohen, Dehaene and colleagues' account which conflicts with the Price and colleagues' position.

The position of Cohen, Dehaene and colleagues

In functional imaging studies Cohen, Dehaene and their colleagues have localized a region of the visual cortex that is particularly responsive to visual words. They have recently taken a position close to the original Warrington and Shallice's (1980) one, by presenting evidence for the existence of a word-form system. Reading is a recent cultural invention, for which the brain has not evolved specifically innate cerebral mechanisms. Nevertheless, it is possible to lose selectively the ability to recognize letter strings. This suggests that i) there is some specialization for reading in the visual system, namely in the visual word form area (VWFA), a cortical region within the left occipitotemporal sulcus bordering the fusiform gyrus (see Fig.3.10); ii) that this ability can be damaged following occipito-temporal lesions (McCandliss, Cohen and Dehaene, 2003). In the VWFA there is a form of functional specialization: it becomes specifically tuned to the recurring properties of a writing system. From this point of view, the rise of perceptual expertise in word recognition provides a remarkable example of how specialization processes within the visual system can accommodate a novel cultural invention.

Cohen and Dehaene (2004) hypothesized that three different forms of specialization can be distinguished. 1. Functional specialization: the visual system has become attuned to the requirements of reading in a given script. For instance some visual neurons fired identically to ”a” and ”A”, thus indicating

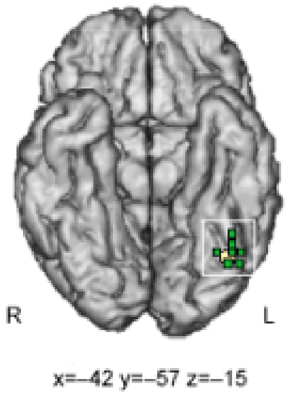


Figure 3.10: Peak of the Visual Word Form Area (VWFA), identified in individual subjects (green squares) and in group analyses (yellow circle) by Cohen, Dehaene and colleagues, 2001; 2003).

abstract case invariance. This hypothesis does not imply any punctuate localization of such processes. It could be logically possible that case-invariant neurons are presented throughout the visual system, without any particular localization. 2. Reproducible localization: Such neurons tend to be grouped together in some fixed regions of the visual cortex. This hypothesis only claims that neurons engaged in the reading process are not localized randomly. The same neurons could be involved in other processes such as face, object or colour processing. 3. Regional selectivity: There are regions of cortex devoted solely to word reading. With those distinctions in mind, they propose that there is functional specialization for reading in the brain of literate subjects and that there is reproducible localization of the neural circuits that are attuned to the reading process. They do not believe that there is complete regional selectivity for word recognition: even the voxels that respond optimally to words tend to be also activated by other stimuli such as pictures.

In neuroimaging studies they demonstrated reliable activation for words when compared to stimuli that control for visual stimulation in the VWFA such as chequerboards. Cohen, Dehaene and colleagues (Cohen, Dehaene, Naccache, Lehricy, Dehaene-Lambertz, Hnaff and Michel, 2000; Cohen, Lehricy, Chochon, Lemer, Rivaud and Dehaene, 2002; Cohen et al., 2003) demonstrated that the VWFA was activated by words irrespective of the visual hemifield in which words were presented, thus realizing a location-invariant process. Furthermore the VWFA showed invariance for typographic case (Dehaene, Naccache, Cohen, Le Bihan, Mangin, Poline and Riviere, 2001), responded only to written stimuli and was generally insensitive to factors that influence lexical access, that is words or pseudowords. Hence the VWFA is considered prelexical (Dehaene, Le Clec'H, Poline, Le Bihan, and Cohen, 2002).

Molko et al. (2002) used diffusion tensor imaging to study the brain connectivity in a patient with alexia in his left visual field (left hemialexia) following a posterior callosal lesion. Results demonstrated the abnormal anatomical disconnection of the VWFA from the right visual regions, thus supporting the idea that this cortical brain area is crucial for orthographic processing. In a recent fMRI study Cohen, Dehaene and their colleagues (Cohen et al., 2003) further developed their initial model of word perception, by comparing data from normal subjects with those of 6 peripheral dyslexics, namely 2 LBL readers, 2 left hemialexic patients, 1 patient with right hemianopic alexia and 1 patient with global alexia (see Fig. 3.11). The VWFA was argued to overlap with the critical lesion site for pure alexia, as identified in their patients (see Fig. 3.12); its destruction as well as the loss of input to or output from it was held to result in pure alexia. This would account for the explanation of pure alexia resulting from white matter damage, as Déjerine (1892) and Damasio and Damasio (1983) argued.

In addition, Cohen et al. (2003) hypothesized that the right-hemispheric homologous region (R-VWFA) could assume some of the functional properties normally specific to the VWFA. In fact the patient F., a typical LBL reader,

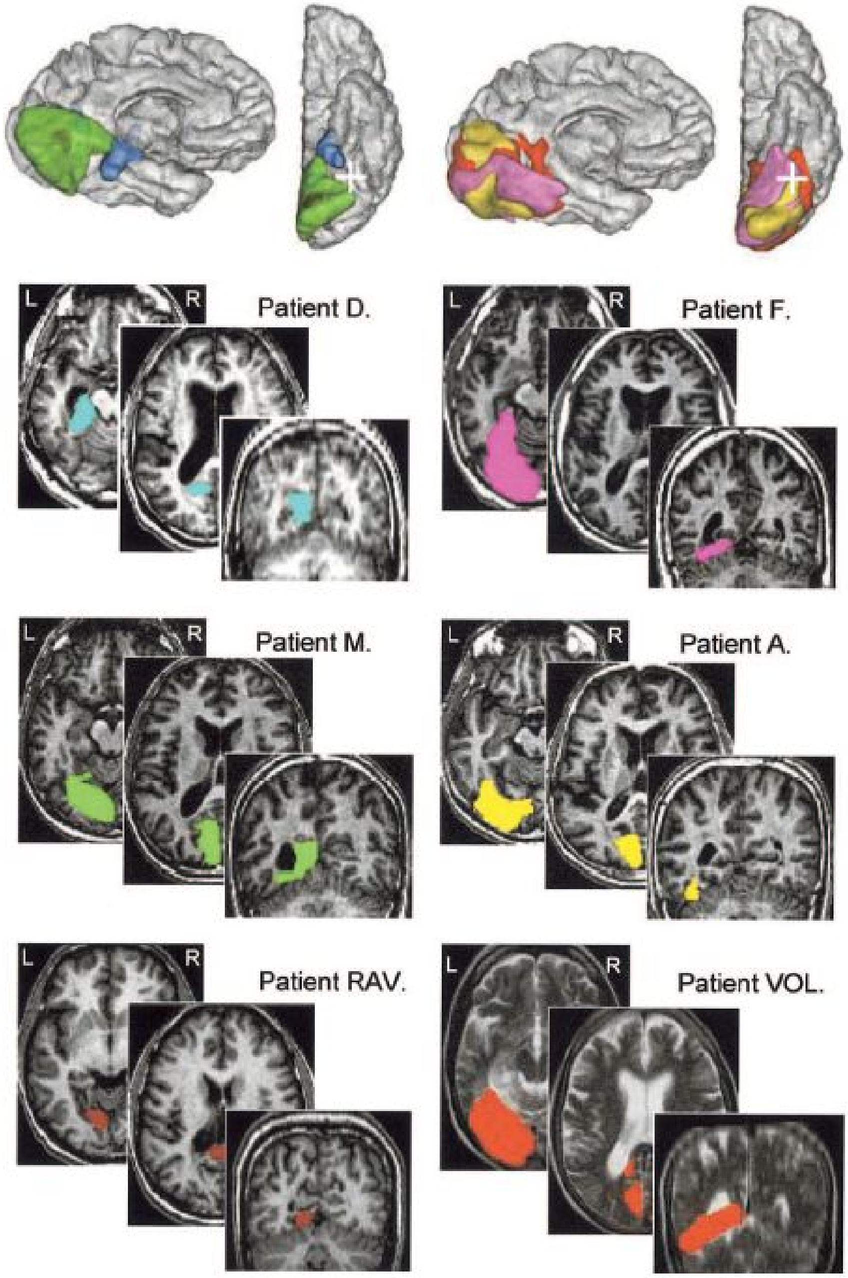


Figure 3.11: Reconstruction of the lesion of 6 patients in Talairac space, compared with the average normal location of the VWFA (white cross). Note that patient D has left hemialexia and patient M has right hemianopic alexia and their lesions did not affect the critical VWFA. Patients A and F are LBL readers and patient VOL has global alexia; for all of them the VWFA is damaged. The additional callosal lesion maybe responsible for the left hemialexia in patient D and for the lack of LBL reading abilities in patient VOL. (Cohen, Dehaene and colleagues, 2003).

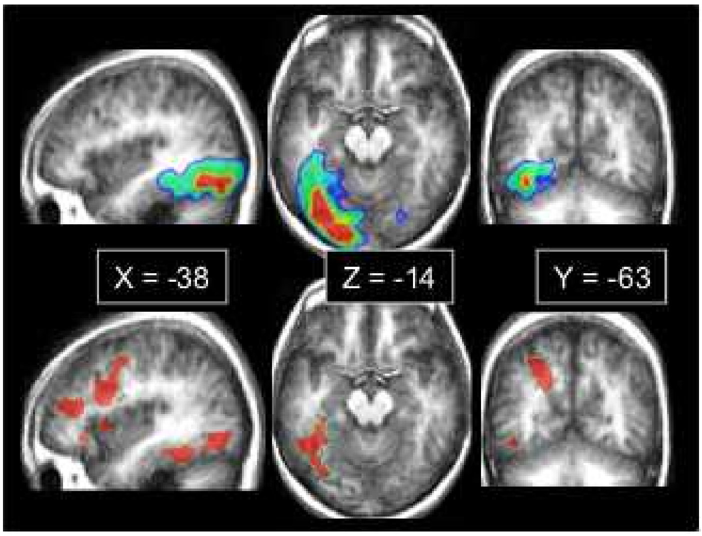


Figure 3.12: Correlation between the lesion site associated with pure alexia (top) and the left occipito-temporal activation (which includes VWFA) during normal reading (bottom), Cohen, Dehaene and colleagues, 2003)).

showed a pattern of activation in the R-VWFA normally specific to the VWFA itself, i.e. stronger activation for alphabetic strings than for chequerboards. However, a residual activation was present in the VWFA, when comparing alphabetic stimuli with chequerboards, likely because the lesion in patient F overlapped with the normal region of the VWFA but spared the dorsal bank of the calcarine sulcus. This residual activation could be due to a partially spared left-hemispheric pathway leading from V1 to the VWFA which could contribute or even be essential to LBL reading. A similar finding was reported by the study of Cohen et al. (2004) where a young patient (CZ) with LBL reading was presented. CZ's VWFA was anatomically spared but deafferented of all visual input, following the surgical resection of the occipito-temporal regions. Using fMRI, they observed an activation in the R-VWFA when alphabetic stimuli was compared with chequerboards. Surprisingly, the VWFA was activated as well:

they argued that this residual activation probably resulted from top-down influences reaching the VWFA through preserved long-distances and U-shaped association fibers.

Overall, according to Cohen, Dehaene and colleagues (Cohen et al., 2003; 2004), the pattern of activation observed in patients with pure alexia (as well as right hemianopic alexia and left hemialexia) compared to normal controls suggests that, in LBL reading, alphabetic symbols are identified in the R-VWFA, then serially transferred to the LH and here word identity is recovered through an effortful verbal working memory process. The abnormal strong activations in a left frontoparietal network related to verbal working memory in the patient F (Cohen et al., 2003) gives support to this hypothesis.

In order to try to investigate the specific components involved in LBL reading, Cohen et al. (2004) attempted to identify the interhemispheric communication which occurs in normal reading and in pure alexia. In normal reading, when a word is displayed in the LVF, the projections go from the right visual cortex such as V4 to the VWFA through the splenium of the corpus callosum. Severing those connections yields alexia in the LVF (left hemialexia). In pure alexia both the intrahemispheric and interhemispheric visual pathways going towards the VWFA are damaged. However, the patients are still able to read. How is it possible? i) Cohen et al. (2004) proposed that during LBL reading the R-VWFA communicates the outcome of letter identification to the LH language system through a pathway whose anatomical substrate is not precisely defined. Severing those connections should prevent LBL reading in alexic patients. The pathway might comprise heterotopic callosal connections which project from the R-VWFA to the language areas such as Broca and Wernicke. Such connections (reported by Di Virgilio and Clarke (1997)) could be involved in the transfer of letter identities across hemispheres. ii) It has been shown that semantic information can be transferred from the RH to the LH through the anterior half of the corpus callosum (Cohen and Dehaene, 1996; Gazzaniga, 2000). On this account, it is possible that the strategic transfer of letter identities across hemispheres during LBL reading can be carried out by a variety of more or less anterior anatomical pathways and thus adapting to a variety of callosal lesions.

Finally in a study which used the subliminal masked priming method, Dehaene et al. (2004) separated letter-level from whole-word codes by preceding target words with an unrelated prime, a repeated prime or an anagram made of the same letters. Moreover they evaluated the invariance of those codes by changing the case and the retinal location of primes and targets. The results showed that the coding units in the VWFA vary across the location. In fact in the more posterior part there is a location-specific priming: activation was reduced only when the same letters were repeated at the same retinal location. Thus, those regions may comprise ”letter detectors” tuned to the presence of a given letter at a specific retinal location (Peressotti and Grainger, 1995). Instead, more anteriorly, activation was reduced whenever the same word was presented twice, even when shifted by one letter location. This suggested that location invariance was achieved in this region. The results of this study indicate that an invariant binding of letters into words is achieved unconsciously through a series of increasingly invariant stages in the left occipito-temporal pathways.

The position of Cathy Price and colleagues

Price and colleagues have extensively used functional imaging to study language functions and in particular the ability to read. Their work has mainly focused on 4 issues: 1. the attempt to segregate the brain areas that are responsible for different reading processes; 2. the lack of specific brain areas dedicated to the recognition of visual words; 3. the model of reading which can account for pure alexia; 4. the use of brain imaging techniques with brain-damaged patients as a powerful method to study reading.

We will start from the first issue. In several imaging studies Price and colleagues (Price et al., 2003) have shown that reading activates a widely distributed set of areas in the occipito-temporal region, in the posterior temporal region, in the precentral and inferior frontal gyri. These regions include areas that sustain orthographic, semantic and phonological processing. Starting from the areas that are involved in the reading process (but not *only* in the reading process), Price et al. (2003) showed that the pattern of neural activation for reading aloud relative to rest (with eyes closed) comprises: bilateral activations in the occipital lobe, in the superior temporal sulci (Wernicke's area included), in the left fusiform gyrus, inferior frontal region (Broca's area) and in the motor cortices (see Fig.3.13). Compared to the historical model of reading (Déjerine, 1892), according to Price and colleagues (Price et al., 2003; Price and Friston, 2002) there is no activation of the angular gyrus (whose lesion do not typically impair reading comprehension) while the fusiform gyrus is activated (Cohen, Dehaene and colleagues, 2000, 2002, 2003).

Price et al. (2003) also attempted to segregate reading from sensorimotor processing. In one of their tasks, they compared reading[1] to a baseline condition where controls were instructed to silently articulate ”OK” in response to seeing strings of false fonts (visual stimuli that resemble letters). The false fonts control for the visual input, while saying ”OK” partially controls for articulation. The activation, left lateralized, was observed in the fusiform gyrus, corresponding to the VWFA (Cohen et al., 2000; 2002; 2003) and in the frontal regions (left anterior ventrolateral frontal and premotor cortex), likely associated to phonological as well as semantic processing. According to them, the reading system divides neatly into areas involved in speech production and areas involved in the lexical and semantic aspects of reading. The speech production areas include dorsal and ventral regions of the posterior superior temporal

[1]It is not clear which kind of reading since in the text it is reported that subjects are instructed to silently articulate the sound of written words, but in the figure legend they write ”reading aloud”.

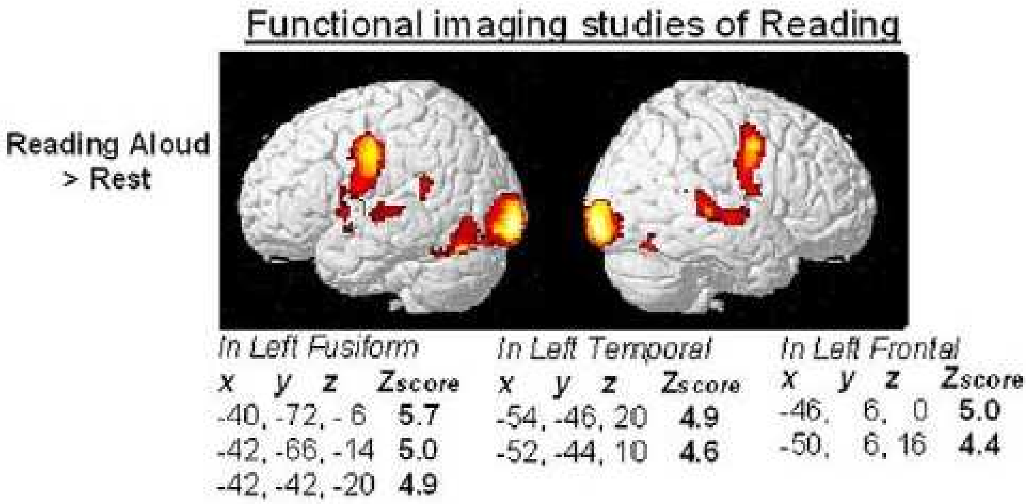


Figure 3.13: Functional imaging study of reading by Price et al., (2003). Activation for reading aloud relative to rest.

gyrus, precentral areas of the sensorimotor cortex and the anterior insulae. In contrast, the reading areas when speech production is controlled include the left fusiform gyrus and the left inferior frontal cortex (Fig.3.14). In their view the left fusiform regions are not simply involved in visual processing, but play an important role also in perceptual categorization (posterior midfusiform) and semantic processing (anterior midfusiform), that are not specific to reading. However, this is not evidenced by this study.

Another attempt was to segregate the sublexical, lexical and semantic routes in reading. Functional imaging studies have compared activation for reading familiar words relative to pseudowords with the expectation that reading familiar words will increase activation in the lexical/semantic areas, whereas reading pseudowords will increase activation in areas involved in sublexical orthographic and phonological conversion. However the results about a dissociation between the lexical and sublexical processes are weak and not clear-cut.

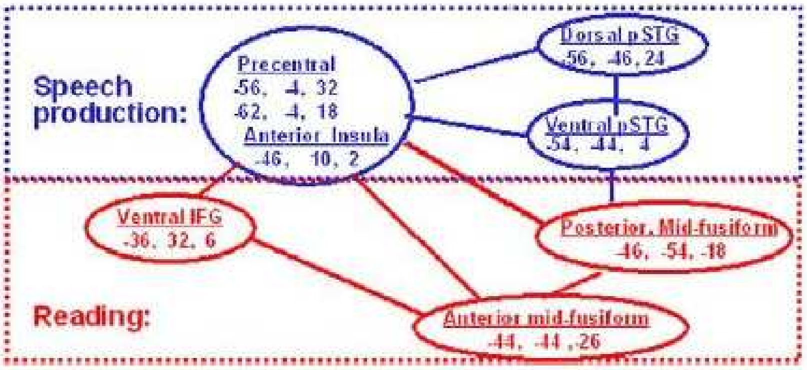


Figure 3.14: Connections between speech production (blue components) and reading when speech production is controlled (red components) (Price et al., 2003).

As pointed out by Price et al. (2003) "However, there is no strong or consistent evidence that illustrates a division in lexical and sublexical routes from orthography to phonology during reading, although this may be because both familiar and pseudowords implicitly activate all possible reading strategies." (p. 35). They go on: "With respect to the cognitive models of reading, the functional imaging data could be consistent only with the distinction between phonological (posterior inferior frontal and superior temporal gyri) and semantic (left anterior fusiform, ventral frontal, angular gyrus[2] and middle temporal areas) processes". However this is consistent with different models of reading, namely with the dual-route theory (Coltheart et al., 2001), with the model of the word-form multiple-levels approach (Shallice and McCarthy, 1985) as well as with the triangle model (Plaut et al., 1996, see Chapter 2).

The second issue that Price and colleagues emphasize concerns the non-existence of a neural area specific to visual word processing. Although Cohen, Dehaene and colleagues argue that the activation of the left occipitotemporal

[2]They say (p.35): "The angular gyrus responds during semantic decisions but not during reading aloud, which is inconsistent with a role in visual word form processing."

cortex is greater for written words than nonword stimuli, the comparison of written words to pictures has produced inconsistent results (Price and Devlin, 2004). According to Price and colleagues reading gives rise to a distributed neural activation that is also engaged in the processing of objects; as a consequence pure alexia should not be so pure. According to them a direct comparison of object naming versus reading showed that occipito-temporal regions were activated to a greater extent by object naming than by reading (Price and Devlin, 2004; Price and Mechelli, 2005), suggesting a lack of support for the presence of left occipito-temporal neural populations dedicated to orthographic processing (but see Shallice (1988; 2003) as regards resource artifacts).

However, Cohen and Dehaene (2004) argue that the same neurons in the VWFA which respond optimally to words could also be involved in other types of processing and thus activated by other stimuli such as pictures or objects (reproducible localization, see previous paragraph). In fact the overlap between activations in words, objects and even faces does not preclude the study of the functional contribution of the VWFA to reading per se. The issue of the nature of the contribution of a given cortical sector to reading (e.g. does it code for single letters or for syllables and morphemes? How local invariance is achieved?) can be addressed completely independently from the issue of whether neurons in this area also contribute to object and face recognition.

The third issue is about the account of pure alexia. Price and Devlin (2004) argued that it is well established that lesions to the left occipito-temporal cortex can result in pure alexia, therefore it should be explained why object recognition is less impaired than reading. They assume that neither reading-specific processes, nor specific areas are necessary to explain pure alexia. For instance, there is no evidence that processes such as location or case invariance are specific to reading. Location invariance is generally considered as a basic property of visual object recognition (Marr, 1982) while case invariance is a special version of object invariance, that is recognizing two visual stimuli as the same despite differences in their surface characteristics.

They claim that reading and object recognition share a number of processing mechanisms: one of these processes might be crucial for reading, but less critical for object recognition which could use other processing mechanisms. For instance, Hasson et al. (2002) associated the left occipito-temporal area with center-biased representation and pointed out that written words are dependent on such center-biased recognition. The same center-biased visual processes can also be involved in object identification, but in addition object identification can proceed successfully from the periphery-biased representations (e.g. in the right occipital cortex). Hence, damage to center-biased representation will impair reading more than object naming, although object naming may also be affected. This account does not require reading-specific processing in the left occipital cortex, nor the assumption of stored visual words forms.

Thus, Price and Devlin (2004) offer an alternative account of pure alexia which posits different dependencies for reading and object recognition on shared processing mechanisms. However, they do not give any example and this position which, although logically possible, is not supported by any evidence as regards pure alexia. By contrast, it is possible that only a subpart of the areas involved in object processing is involved in word processing (Cohen and Dehaene and colleagues).

The fourth issue that is important for Price and coll.'s position (2003) is the use of functional imaging in the study of patients with acquired dyslexia. They have pointed out that the patient-based approach in lesion studies of dyslexic patients has strong limitations: for instance in specifying the precise location of any of the reading areas identified or in indicating how different reading strategies might be implemented. Moreover it cannot provide good spatial resolution or establish that an area is not involved in a given function. However, it is evident that functional neuroimaging studies on normal subjects also have limitations: for instances a significant activation in response to one task does not indicate that the activation is necessary for correct performance. For this reason Price and coll. (2003) propose that the limitations of both approaches can partly be overcome by functional imaging studies of patients with acquired dyslexia. In fact, if one wants to determine which areas are necessary for reading, it is possible to start to consider whether a lesion to each area results in a reading deficit. This is the lesion-deficit approach. Conversely, imaging can be used to guide that approach because the critical factor is whether the lesion involves an area that activates during normal reading. The double approach can be used to determine whether more than one neural system can enable reading. For example, if a patient with a lesion to the reading system is still able to read, then functional imaging may reveal whether the individual's successful reading results from activation in atypical reading areas. Furthermore, the observation of abnormal patterns of activation can then guide the investigation of individual variability in normal subjects.

From this viewpoint, activation studies in brain-injured individuals are still in their infancy, but may become very important as a means of testing reserve function and mechanisms of recovery.

Chapter 4

Neuropsychological study of 2 pure alexic patients: FC and LDS

4.1 Aim of the study

As explained in Chapter 3, different accounts have been proposed so far in the attempt to explain the functional origin of pure alexia. Some theories claim that this disorder is caused by a single source of deficit (e.g. Behrmann, Plaut and Nelson, 1998), while others suggest that the deficit may vary across patients (e.g. Price and Humphreys, 1992; Patterson and Kay, 1982; Hanley and Kay, 1996).

In this investigation we follow the general perspective of Price and Humphreys (1992), and Kay and colleagues (Patterson and Kay, 1982; Hanley and Kay, 1996) and describe two new cases of pure alexia, FC and LDS in order to study whether the functional origins of their impairments differ. On the perspective of multiple different forms of deficits, Kay and colleagues have proposed a model (taken from Warrington and Shallice, 1980) which considers both the ability to process single letters and to put them together in a word. In their model they assume a letter-form analysis stage connected to a word-form system: however 1. it is not clear whether their patients are able to process letter rapidly and accurately from that level; 2. the word-form system is supposed to work only on words.

By contrast, Price and Humphreys (1992) argued that one patient, EW, had a deficit in parallel processing and the other one, HT engaged his attention too far to the left of the stimulus. Thus, their deficits appeared to involve a rather general perceptual problem which prevents normal access to the lexical and semantic representations. However, their study, as described in Chapter 3, mainly focused on visual span, on the discrimination of features in letter and number strings and on the ability to scan attention on a subitising task and it did not yield to clear-cut results. Moreover the ability to read per se was not fully investigated.

In our study of pure alexic patients we aimed to investigate various cognitive processes, but in particular two components that are involved in visual word recognition: the ability to process letters and to integrate them together in syllables and words. As discussed in the last paragraph of Chapter 2, these are crucial points in the processing of words and represent a step forward in the study of pure alexia, since the relation between these two skills (as well as the intermediate level such as syllables) has never been carefully studied, although the syllable level can be more easily studied in languages such as Italian with clear syllabic boundaries.

It is worth emphasizing that letter processing and letter integration are two serial processes, where the former takes place before the latter. This observation has important methodological consequences. A deficit at the letter level will result in a less efficient integration ability, even if this process is not itself damaged. Thus, a patient with slowed letter identification can show at most a strong dissociation if his capacity in conjoining letters is good (because performance on relevant tasks will still be below average). The careful study of our two alexic patients has brought interesting and clear-cut results.

Our patients LDS and FC show a close quantitative similarity in the characteristics of their overt reading; however, their LBL reading problems, we argue, arise from damage to different components of the reading process (Rosazza, Appollonio, Isella and Shallice, submitted). LDS has a lower-level deficit in letter processing, whereas FC has a higher-level deficit in integrating information across letters. These results present difficulties for a unitary account of pure alexia which hypothesizes a single functional deficit at a low level for all the patients manifesting the symptoms (Behrmann et al. 1998, Farah and Wallace 1991, Saffran and Coslett, 1998) and instead imply that letter-by-letter reading can occur after different components of the reading process are selectively damaged.

4.2 FC: case study

FC is an 85-year-old man with 8 years of schooling (starting with normal day school for 5 years and after at evening school) who worked as a draughtsman and mechanic. Despite his low educational level, he read books and newspapers extensively, being very interested in politics. In April 2001 he suffered an ischaemic stroke in the left occipital lobe, confirmed by MRI scans and neurological examinations revealed a right homonymous hemianopia. MRI examination revealed a stabilized left occipital lobe and subcortical focal ischaemic lesion (see Figure 4.1).

His left inferior and medial occipital lesion involves the occipital pole (area 17), the lingual gyrus (area 18) with anterior and medial extension to the posterior parahippocampal gyrus and the occipital fusiform gyrus encompassing part of the collateral gyrus. Multiple, diffuse foci of chronic ischaemic encephalopathy are also present, mainly in the periventricular white matter and in the basal ganglia. More specifically, there is a primary white matter lesion

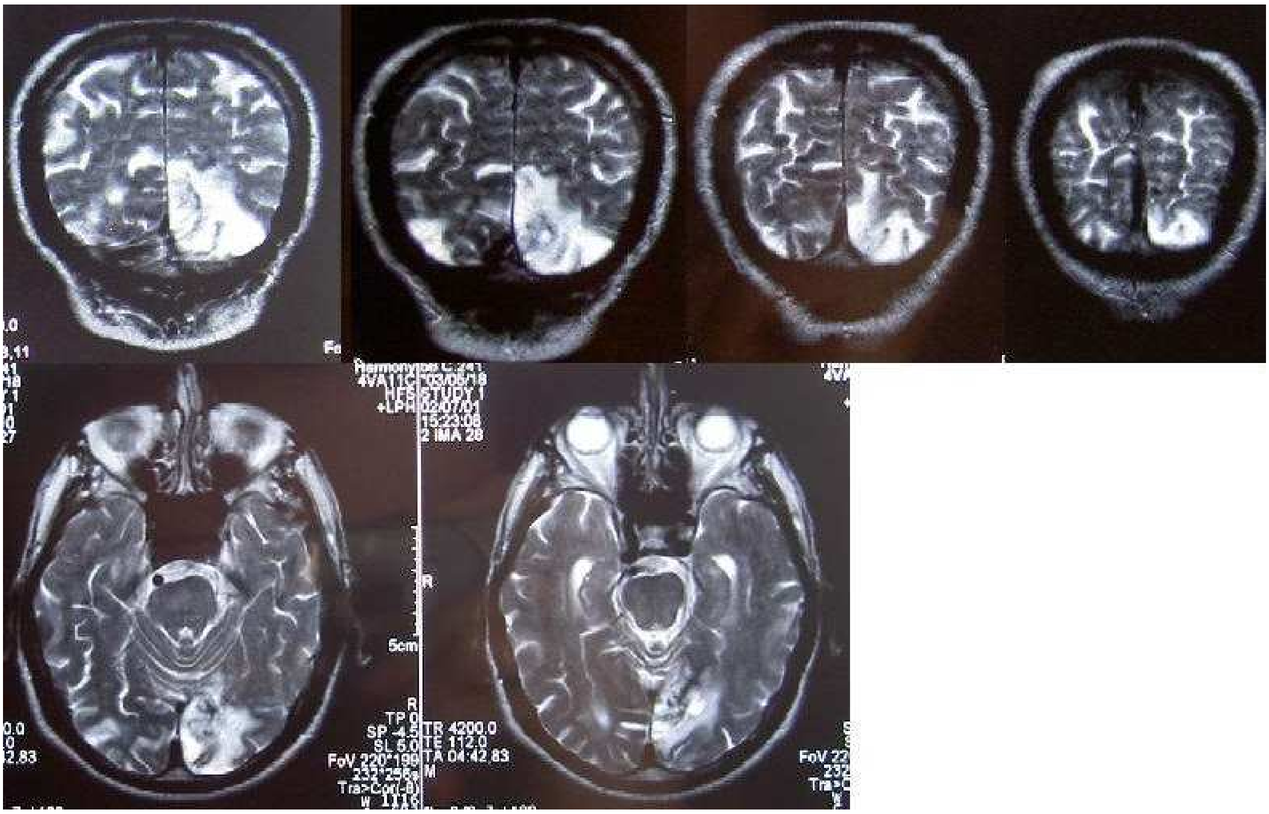


Figure 4.1: MRI scans of FC. The coronal and axial sections show the left posterior occipital lesion which involves the occipital pole, the lingual gyrus, the occipital fusiform gyrus and the underlying paraventricular white matter in the left hemisphere. A smaller right occipital lesion is also present in the lateral occipital gyrus. The corpus callosum is intact.

that involves the ventro-medial paraventricular white matter beneath and the medial-posterior side of the occipital horn. The area over the occipital horn is likely to be intact. The splenium of the corpus callosum is spared. A small right occipital lesion is also present in the lateral occipital gyrus (areas 18 and 19) and possibly extending into the posterior occipito-temporal sulcus bordering the occipital fusiform gyrus (area 19). A further MRI examination carried out in March 2004 confirmed the previous lesions and revealed a new one in the cortical and subcortical left parietal area involving the supramarginal gyrus and the angular gyrus. Three different neuroradiologists confirmed that the 2001 MRI examination did not show the left parietal lesion. The lesion did not cause any evident symptoms since the subsequent neurological examinations as well as his neuropsychological assessment repeated in 2004 did not show any change. In particular no change was apparent in his reading performance. FC received a reading therapy for some months after his stroke and before this experimental study began. At the time of testing FC (from February 2002) is able to write normally and to read letters and digits. When he is asked to read words, he uses a letter-by-letter strategy and his reading performance is stable. He is alert, well oriented in time and place, and cooperative. He did not show other language, apraxic or memory deficits; his main complaint is his inability to read newspaper and books.

4.2.1 FC: neuropsychological assessment

A general assessment revealed that FC was of normal intelligence (see Table 4.1) performing well on the WAIS (Wechsler, 1986). He had good language abilities except for reading, with spontaneous speech being normal. His memory as assessed by the Story Recall (Spinnler and Tognoni, 1987) indicated a performance within the average range (see Table 1). After the second MRI examination, FC has also been tested with the ideomotor apraxia test (De Renzi, Motti and Nichelli, 1980): his performance was completely normal.

When assessed on standardized perceptual tests (see Table 4.2), FC achieved normal scores on all the subsets of the VOSP (Warrington and James, 1991) except on Silhouettes, although his performance was impaired on the Object decision and the Association match of the BORB (Riddoch and Humphreys, 1993). In another perceptual test, the CORVIST (James, Plant and Warrington, 2001), he scored well on all the subsets except those tapping colours. FC had slight colour agnosic or anomic difficulties on the colour naming task (7/10) included in the AAT (Luzzatti, Willmes and De Blaser, 1996). Brown was named as orange, looking at orange he said: "It is not brown, not orange, I don't know" and violet was not recognized. However, his performance on the Ishihara test was flawless (25/25). In a colour-object matching test FC was asked to provide the appropriate colour of 71 items: 20 animals, 14 fruits, 12 vegetables and 22 nonliving things. His performance was within the average (63/71, 89%; mean among 4 matched controls (C1, C2, C5, C6): 62; sd: 5.48). FC was given the Benton Faces Recognition Test (Benton, Silvan, Hamsher, Varney, Spreen, 1992) where his performance was borderline (see Table 2). The patient scored 18/38 on a Famous Faces Test: his accuracy was 18/38 and he made 9 anomic errors; his performance is not greatly worse than one of these two matched controls C1 and C2 (32/38 and in addition 1 anomic error; 20/38 and in addition 12 anomic errors, respectively). A series of tasks tapping general abilities (Raven and verbal fluency) and arithmetic-related skills (digit span and arithmetic subsets of the WAIS) in addition to tests of ideomotor apraxia (see above) was carried out again after his second MRI examination to reveal any difficulty associated with his left parietal lesion. The results did not show any significant change (see Table 4.1).

	FC	LDS
MMSE	–	29
Token test ∼	32 (31.05± 2.93)	33 (33.6± 2.11)
Verbal Fluency (FAS)∼	16; 13# (22.5± 10.11)	28 (30.19± 9.64)
Raven Progressive Coloured Matrices ∼	25; 27# (21.50± 6.95)	19.5* (26.75± 5.13)
WAIS ∼ $		
Information	11	10
Digit span	8; 7#	7
Vocabulary	9	11
Arithmetic	9; 8#	5*
Comprehension	–	13
Similarities	11	13
Picture Completion	12	8
Picture Arrangement	–	9
Block Design	9	6*
Object Assembly	5*	6*
Digit symbol	–	7*
Story recall ∼	7.7 (10.86±3.17)	13.7 (12.37±3.73)
Rey figure copy	11** (32.44±3.5)	
Attentional matrices ∼	30*(39.68±8.03)	53 (50.41±8.78)
Test of ideomotor apraxia &	68#	

Table 4.1: The performance of the patients on general neuropsychological assessment. In brackets mean and sd prorated by age are given. *score <1sd below normal mean; ** score <2sd below normal mean; ∼ the scores are prorated by age; $ mean=10 and sd=3 for each subset according to the Italian correction (WAIS-R Contributo alla Taratura Italiana; age: 55-64); & cut off: 53; borderline: 54-62. # the score refers to retesting in 2004, carried out after the last MRI examination.

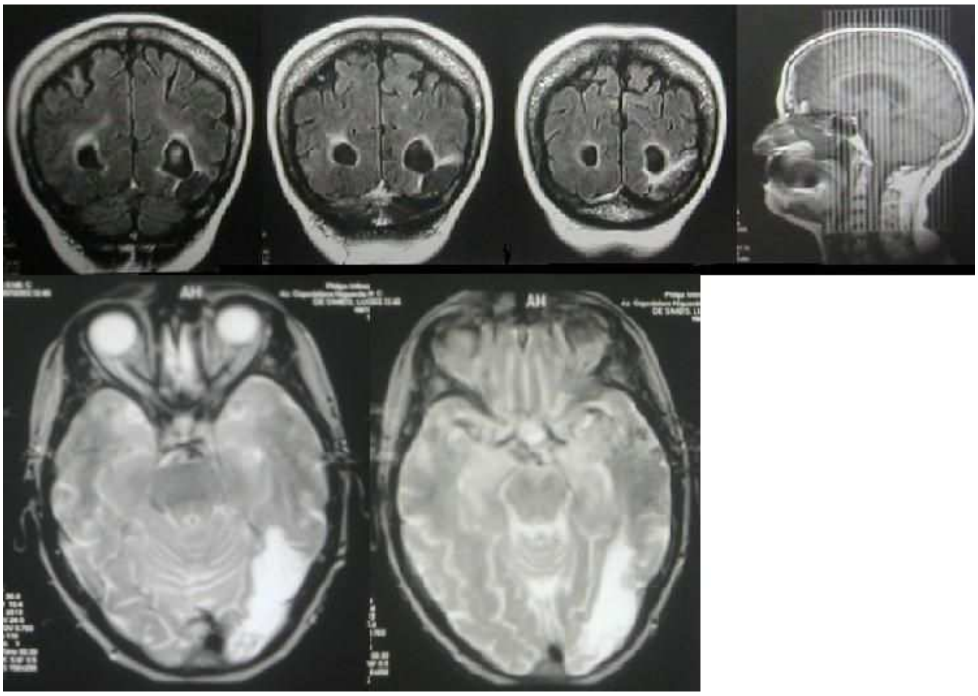


Figure 4.2: MRI scans of LDS. The coronal and axial sections show a lesion in the left occipito-temporal lobe which involves the occipito-temporal fusiform gyrus and include the occipito-temporal sulcus. The corpus callosum is intact.

4.3 LDS: case study

LDS is a 56-year-old right-handed woman with 13 years of schooling who had worked as an agent for a pharmaceutical company. In February 1998 she noted the sudden onset of an inability to read. This was due to an ischaemic stroke. A CT scan (February 1998) demonstrated an infarction in the left posterior temporal lobe and a small right posterior frontal subcortical ischaemic area. The result was confirmed by an MRI carried out in June 2000 (see Figure 1b), which indicated a lesion in the left occipito-temporal part of the inferior temporal gyrus and in the left temporo-occipital fusiform gyrus encompassing the occipito-temporal sulcus (area 37).

The lesion also involves the occipital fusiform gyrus (area 19) with no apparent involvement of the subcortical white matter. The corpus callosum is intact. Another lesion involves the subcortical white matter of both hemispheres in the fronto-central and posterior frontal regions and in the right hemisphere the subcortical white matter of the precentral gyrus. She had a right homonymous hemianopia. LDS is clinically a 'pure alexic patient': she is able to write normally and to read letters and digits, but she uses a letter-by-letter strategy to read words. She received a reading therapy from Prof. Basso before this experimental study began; at the time of testing her reading performance is stable. She is alert, well oriented in time and place and cooperative.

4.3.1 LDS: neuropsychological assessment

When tested in April 2002, she performed within normal limits on the Vocabulary, Digit Span and Similarities subsets of the WAIS (Wechsler, 1986), but had weak performance on the Arithmetic subset. She also performed weakly on the WAIS Performance IQ subsets (Block Design, Object Assembly and Digit Symbol). She showed impaired performance on the Rey Figure copy test. These problems may be related to her right frontal lesion. Her spontaneous speech was fluent and well formed, as well as her comprehension and memory abilities (see the standardized tests in Table 4.1). On visual perceptual tasks her performance was normal, for the VOSP (Warrington and James, 1991) on all the subsets except for the Silhouettes task and for the BORB (Riddoch and Humphreys, 1993) again all the subsets, except for the Object decision test (see Table 4.2). Moreover she had no difficulties with the CORVIST test (James, Plant and Warrington, 2001). Her performance was flawless on the colour naming test (10/10) included in the AAT (Luzzatti, Willmes and De Blaser, 1996) while she made a few errors on the same colour-object matching task as administered to FC (LDS: 63/71, 89%; C8, C9, C10, C11 mean: 68.5, sd: 0.06). Finally her performance was normal on the Benton Faces Recognition Test (Benton et al., 1992). When LDS was given a Famous Face Test her performance (19/38, 50%) was severely affected by anomic errors (15 out of 19 total errors, 79%) such as: Hitler >"he has died, he was the Nazi leader". The performance of her matched controls C10 and C11 was better (36/38 and 38/38).

4.4 Control group

Control subjects participated in the study: seven (from C1 to C7) were matched to FC for age (mean age: 84.4 years, sd: 3.4) with 4 men and 3 women. The comparison is a conservative one as all the controls except one had a higher education level (mean schooling: 11,7, sd: 2,4, almost 4 years more than FC). The other eight controls (from C8 to C16) were matched to LDS for age (mean age = 58.75 years, sd = 2.71), gender and education level (C10 had 8 years of schooling, the other controls 13 years of schooling). A key contrast will be of the direct comparison between FC and LDS' performance. However as they are different in age, interpretations must also be supported by comparisons of each with their controls.

4.5 Naming abilities

Naming abilities of both patients were initially assessed with 48 figures from the Snodgrass and Vanderwart set. FC's accuracy was 38/48 (79%): he identified 12/18 (67%) living things and 26/30 (87%) non living ones with no significant difference between the two groups. LDS's performance was slightly impaired (37/48; 77%): she correctly identified 11/18 (61%) living items and 26/30 (86%) non living things.

On another session the patients were given 266 pictures taken from the Lotto, Dell'Acqua and Job set (2001). FC scored 231/266 (87%), in particular

	FC	LDS
CORVIST		
Symbol acuity test	24/24	36/36
Shape discrimination test	8/8	7/8
Size discrimination test	1/2	2/2
Shape detection test	8/8	8/8
Hue discrimination test	1/4	4/4
Word reading test	Normal, but slow 7/8	Normal, but slow 7/8
Fragmented numbers test	7/8	8/8
Face perception test	5/8	8/8
BORB		
Minimal features	–	22/25
Foreshortened view	–	24/25
Association match	25/30*	27/30
Item Match	31/32	30/32
Object Decision (A&B hard version)	20/32**& 25/32§	23/32**& 22/32§
Object Decision (A&B easy version)	26/32*& 26/32**§	28/32& 26/32**§
VOSP •		
Screening test	20/20	19/20
Incomplete Letters	17/20	17/20
Silhouettes	11/30#	15/30#
Object Decision	20/20	18/20
Progressive Silhouettes	10	6
Dot Counting	10/10	10/10
Position Discrimination	19/20	19/20
Number Location	10/10	10/10
Cube Analysis	10/10	10/10
Colour Naming	7/10	10/10
Ishihara's test	25/25	
Colour-object matching test	63/71	63/71
Benton test&	41	42
Famous faces naming test	18/38 (9 A) ∼	19/38**(15 A) ∼

Table 4.2: The performance of the patients on visual perception tasks; & the scores are prorated by age and education; •The scores are scaled by age; # score below 5%cut off score; §the two measures reported in a cell are based on different pictures and therefore have different control values; *score <1sd below normal mean; ** score <2sd below normal mean; ∼ A means anomic errors

his performance was 75/95 (79%) on living things and 154/171 (91%) on non-living things. Two controls (C1 and C2) matched to FC scored 244/266 (92%) and 210/266 (79%), respectively on the same naming task. As evident, FC's performance is well between the normal range.

LDS correctly named 226 out of 266 pictures (85%) taken from Lotto, et al. set (2001): 71/95 (75%) were living items and 154/171 (90%) were non-living ones. The two controls C10 and C11 matched to LDS were presented with the same naming task. They scored 225/266 (85%) and 260/266 (98%), respectively. LDS's performance is the same as one of her matched controls.

These results indicate that the ability of FC and LDS to identify pictures of objects is rather accurate (87% and 85% of correct responses, respectively) and is not different from their matched controls at a gross level.

4.6 Reading and writing

4.6.1 Prose reading

Patients and control subjects were presented with a text to read taken from a battery of 325 words (Nuove prove di lettura MT per la scuola media inferiore, by Cornoldi and Colpo, 1995) used to assess reading abilities in young students with possible developmental dyslexia. The text comprised 325 words. Subjects were asked to read at their own speed. Text reading was recorded with a microphone connected to a PC via a cool edit program for each patient and control subject. FC took 9 min 49 sec to read the text and made 2 errors. LDS took 10 min 38 sec to read and made 5 errors. Control subjects C1 and C2 matched to FC, took 3 min and 2 min 38 sec and made 6 and 0 errors, respectively. C8 and C9, matched to LDS, took 3 min 7 sec and 2 min 21 sec and made 1 and 0 errors, respectively.

4.6.2 Word reading

A total of 120 words, 30 each of 4, 6, 8 and 10 letters in length were presented in random order on a PC monitor using an E-prime program. Words were divided into abstract and concrete and each of the 2 subgroups was matched for frequency. Words were printed in lower case 30-point Arial font and the microphone connected to the PC started measuring at the beginning of the vocal response. Therefore we measured the voice onset time because it is the best measure to use when patients are moderately slow and because it is usually employed as reported in the literature (Bowers and Arguin, 1996; Behrmann, Plaut and Nelson 1998; Behrmann, Nelson and Sekuler, 1998; Lambon Ralph, Hesketh and Sage, 2004; Arguin and Bub, 2005). Patients were asked to report the word when they were sure, avoiding conduites d'approche; answers with stuttering have been removed as well as trials with breaths close to the answer. Moreover if the microphone did not "hear" the voice, the E-prime program did not move to the next trial. These criteria were applied in all experiments.

By performing an analysis of variance, a clear word length effect emerged for both FC ($F(1, 113) = 41.14$ $p < .0001$) and LDS ($F(1, 115) = 34.3$ $p < .0001$), whose increments in reading speed per letter were 90ms and 160ms, respectively (see Figure 4.3). They showed neither effects of imageability nor frequency. They each made 1 error. Control subjects (from C1 to C9, C12, C13 and C14) showed little increment in reading speed (ms) per letter (-11 <B textless 10) with the effect of word length being completely insignificant. C1, C6, C7, C8, C12, C13 made no errors, C2, C3, C4, C5, C9 only 1 error each.

FC was asked to read the same set of word in 2004 after his last MRI examination. His performance, if anything, showed a slight improvement. In fact despite an evident word length effect ($F(1, 106) = 27.27$ $p < .0001$), the increment in reading speed per letter was slightly less than before, namely 70ms. He made 2 errors and 3 conduites d'approche (e.g. meta >me.. meta; fantasia >fanta.. fantasia; termometro >ter.. termore.. termometro). The results

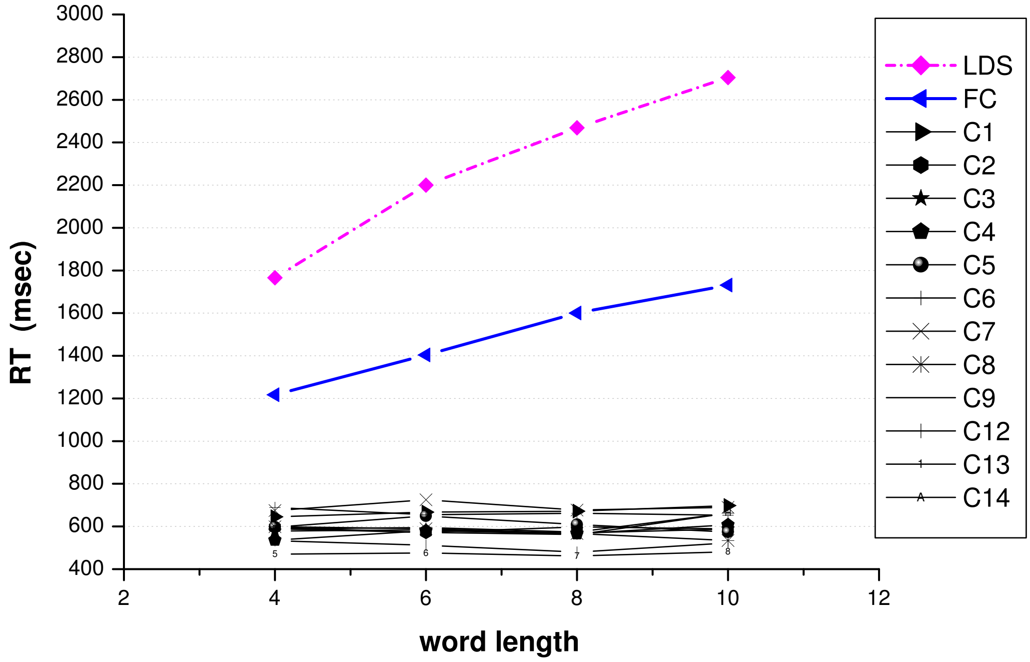


Figure 4.3: Word reading. RTs (ms) of FC, LDS and their matched control subjects as a function of word length. The voice onset time has been measured and errors and conduites d'approche were removed from the analysis.

indicated that FC was quicker than LDS. Qualitatively their performance is that of relatively mild LBL readers since they are accurate and not so slow at word reading. In fact regarding the word reading speed, FC and LDS take an average of 1,6 sec and 2,5 sec respectively to name 8-letter words. If their performance is compared to the 4 patients described by Patterson and Kay (1982) and the 7 reported in Behrmann et al. (1998) one sees that only 3 patients out of the 11 mentioned showed a similar reading pattern, taking about 1,3 sec, 2,3 sec and 2,3 sec on average to read 7-letter words.

4.6.3 Writing

Both the patients were asked to write to dictation 3 prose passages from the MT battery (Gruppo MT, prove di lettura per la scuola media inferiore), a reading test for teenagers with probable developmental dyslexia. The passages had a total of 101 words. Patients were also asked to write 50 words of 4, 5, 6, 7 and 8 letters in length. We assessed the accuracy (not writing speed). FC made 7 mistakes writing the prose passages, 6 of which are grammatical errors (e.g. un attimo (an instant) >un'attimo (a instant); l'integrit (integrity) >lintegrit; ha cominciato (has begun) >a cominciato) and 1 is semantic (viaggiare (to travel) >guardare (to look)). The grammatical errors could well be due to his relatively low education level. His performance was fairly good with the 50 words to write, since he made 4 mistakes (e.g. ciliegia (cherry) >cigliegia). LDS made just 1 mistake (inurbarsi (to move into urban areas) >inerbarsi) and 2/50 errors with single words (e.g. carriola (wheelbarrow) >cariola, a common error made also by FC).

4.7 Implicit reading

The aim of these tasks was to test patients' ability to extract lexical and semantic information from briefly presented words, in order to investigate if they show implicit processing of words. As discussed in Chapter 3, there are patients who are able to access lexical and semantic information of visually presented words which they are unable to identify explicitly and, in some instances, claim not to have even seen (Shallice and Saffran, 1986; Coslett and Saffran, 1989a; 1993; Saffran and Coslett, 1998). In semantic categorization tasks the performance of some dyslexic patients was significantly above chance level even when the items were presented at exposures too brief for overt reading and recognition.

4.7.1 Semantic categorization task: 1 word

In these experiments words were presented too briefly to support LBL reading and patients were encouraged to guess by using a different reading strategy. We were interested in the correct categorization percentage of words not read and in the type of mistakes patients make. In this task, 30 words of 4-8 letters in length were displayed individually for 250ms in the central position of the monitor by means of E-prime. Subjects had to carry out a binary classification task deciding if the word was a name of for instance 'a town or an animal'. Six different versions of this task were created and each patient was presented with 3 or 4 of them in different sessions: 1) towns or animals; 2) buildings or vegetables; 3) Italian or foreign towns in script [1]; 4) fruit or animals in capital letters. Patients were discouraged from attempting to read letter by letter and were asked to try to extract any information they could from words that were displayed too quickly to be read.

Results showed that (see Table 4.3) when FC was not able to read the word, he did not even have any idea of what the word was e.g. Bologna >'foreign, Bellinzona'; Verona >'foreign, Vienna (Vien)'. When they guessed the meaning of a word, the number of words correctly classified by both FC and LDS

[1] Differently from the other version which comprised 30 words, this version consisted of 40 words, 20 Italian and 20 foreign cities

	Tot	Read W	CC not read W	Incorrect	Refused to guess	Correct guessing
FC	138	80	20	11	27	20/31 (64%)
LDS	97	49	29	18	1	29/47 (62%)

Table 4.3: Semantic categorization task: 1 word. "Tot": total of words presented in the task; "Read words": total of words that patients were able to read (at 250ms); "CC not read W": total of correctly classified not read words; "Incorrect": incorrect classified words; "Refused to guess": number of "I don't know" answers, which have not been included in the analysis of correct guessing; "Correct guessing": finally the number of correctly classified words out of the total of guessed words, both correct and incorrect (with percentage).

did not differ significantly from chance (Binomial, FC: $p = .15$, N=31; LDS: $p = .14$, N=47). Although he was continuously told to try to guess, at first he often gave the target name he thought to be correct, and secondly the category e.g. pecora (sheep) (town vs animals) >'pesce (fish)... animal'; teatro (theatre) (building vs vegetables) >'tetto (route)... building'. He was essentially unable not to use a LBL reading strategy. The visual mistakes LDS made also revealed her difficulty in suppressing LBL reading (e.g. Damasco (Damascus) >Italian, Domodossola; Ginevra (Geneva) >Italian, Genova; Monaco >Italian, Mantova; Bologna >foreign, Barcelona).

4.7.2 Semantic categorization task: 3 words

In this task patients were presented with lists of 3 words, where one was a target and the other two were abstract words (e.g. cipolla (onion) agio (comfort) odio (hate)). Words (from 4 to 8 letters) were presented successively, at the same central spatial location, for too short a time for patients to be clearly identified (250 ms). The task, which comprised 30 triplets, had again a binary classification procedure where patients were instructed to indicate whether the target word, which could occupy any of the three positions, was for example a building or a vegetable. Different versions were created: 1) a town or an

	Tot	Read W	CC not read W	Incorrect	Refused to guess	Correct guessing
FC	119	31	5	4	79	5/9 (55%)
LDS	119	25	37	35	22	37/72 (51%)

Table 4.4: Semantic categorization task: 3 words. ”Tot”: total of words presented in the task; ”Read words”: total of words that patients were able to read (at 250ms); ”CC not read W”: total of correctly classified not read words; ”Incorrect”: incorrect classified words; ”Refused to guess”: number of ”I don’t know” answers, which have not been included in the analysis of correct guessing; ”Correct guessing”: finally the number of correct classified words out of the total of guessed words, both correct and incorrect (with percentage).

animal; 2) a building or a vegetable; 3) an Italian or a foreign town; 4) a fruit or an animal; 5) a vehicle or a cloth. In the third task (Italian vs foreign towns) we put the all the initial letters of the list words in capital letters (e.g. Vizio (Vice), Merito (Merit), Parigi (Paris)) in order to have town target words more easily recognizable as a word form[2]. Patients were always encouraged to derive just a feeling that the word could belong to one of the two given categories and it was hoped that the two distractor words would help patients not to rely just on the few letters identified. Some triplets were removed because patients were distracted on that trial.

Results are shown in Table 4.4. If read words are left out, it appears clear that FC was unable to guess: he just relied on the few letters he could identify to guess the target word (e.g. fruit vs animals: pena (pain) sforzo (effort) pesca (peach) >pecora (sheep); gloria (glory) lealt (loyalty) prugna (plum) >leone (lion)). He also gave a lot of ”I don’t know” answers. Also LDS carried on using the first letters she was able to read to guess the word (e.g. vehicles vs clothes: patto (agreement) slitta (sledge) lode (praise) >’cloth, loden’ (overcoat); towns vs animals: Coraggio (bravery) Pisa Scelta (choice) >’animal, scoiattolo’ (squirrel)).

[2] Differently from the other version which comprised 30 triplets, this version consisted of 40 triplets

	Tot	Read W	CC not read W	Incorrect	Refused to guess	Correct guessing
FC	144	89	6	7	16	6/13 (46%)
LDS	144	54	17	21	4	17/38 (45%)

Table 4.5: Semantic categorization task: Chialant and Caramazza's task. Patients were asked to read aloud, guessing if unsure. "Tot": total of words presented in the task; "Read words": total of words that patients were able to read; "CC not read W": total of correctly classified not read words; "Incorrect": incorrect classified words; "Refused to guess": number of "I don't know" answers, which have not been included in the analysis of correct guessing; "Correct guessing": finally the number of correct classified words out of the total of guessed words, both correct and incorrect (with percentage).

4.7.3 Semantic categorization task: Chialant and Caramazza's task

Patients were given another word categorization task, one created by Chialant and Caramazza in 1998. Words were displayed individually for 250ms and centrally on the screen. Patients were presented with 144 words divided into 3 sublists, each consisting of 48 words. Within each list there were words belonging to three different categories, for example list 1 consisted of 16 names of cities, 16 of body parts and 16 of household items. Patients were asked to read aloud each word, guessing if unsure. For cases in which they produced a "don't know" response, they were asked to choose the one to which they thought the word might belong from 3 different categories. If they read a word correctly, the trial was discarded, as were cases in which they produced an incorrect response.

Results are reported in the Table 4.5.

FC often refused to guess: even if he was constantly encouraged to guess, he carried on telling me: "I have no idea what I read" or "It's nonsense to say a name by chance, so nothing". LDS made several mistakes and generally she was unable to guess the word. Some of her incorrect responses were orthographically very similar and possibly semantically related to their targets: e.g. oliera (oil cruet)>oliva (olive); mestolo (ladle) >minestra (soup); tovaglia (tablecloth) >tavolo (table); fornello (cooker) >formaggio (cheese). Others errors LDS made shared only the first part of the word with the target (e.g. bocca (mouth) >bomba (bomb); spalla (shoulder) >spada (sword); gomito (elbow) >gommone (rubber dinghy)), suggesting again that she tended to rely on the few words seen to guess the word.

As a conclusion of all the 3 semantic categorization tasks, the results indicate that both the patients were unable to show implicit reading at least on the experimental paradigms used. Unlike the finding of Shallice and Saffran (1986) and Coslett and Saffran (1989a, 1993, 1998), neither FC nor LDS were able to guess the semantic domain of the word and instead they both relied on the few letters they had decoded to attempt to identify the target word. Other investigators have already reported a lack of covert reading in the tasks they used with pure alexic patients (Behrmann and Shallice, 1995; Patterson and Kay, 1982). The patient MJ (Chialant and Caramazza, 1998) for instance was able to extract some visual information from briefly presented words but her above-chance performance on a semantic categorization task was more likely to reflect a sophisticated ability to guess than a true lexical-semantic access.

It is conceivable that some other manipulations (e.g. the use of very short exposure durations, but see Shallice and Saffran (1986) who used longer durations (2 seconds) to test their patient ML and implicit reading was evident in tasks of lexical decision and semantic categorization) might give a positive result. However there is currently no evidence that implicit access to semantics was widely available to our patients.

4.8 The visual span

The visual span in the apprehension task investigates patients' ability to identify more than one stimulus at a particular exposure duration. Failure to report more than one stimulus from brief tachistoscopic presentation could occur for a number of reasons. Patients may have a deficit either in visual short term memory, or in earlier processing stages such as in parallel processing of features or in orienting visual attention to briefly presented stimuli. Following Behrmann and Shallice (1995), patients were presented with 3-digit and 3-letter arrays briefly displayed on a PC screen. A vertical presentation in addition to a horizontal one was introduced to assess whether patients' performance was affected by the presence of a hemianopia that might affect the third horizontal position.

Materials and Procedure

A total of 48 triplets of digits (e.g. 7 2 5) and 48 triplets of letters (e.g. A L D) were displayed horizontally on half of the trials and vertically on the other half for 3 different exposure durations: 100, 150 and 250ms. Stimuli were presented in an ABBA design, so that patients were presented 24 triplets of letters vertically, 24 digits horizontally, 24 digits vertically and 24 letters horizontally. A fixation point (a cross) appeared in the centre of the screen for 1 sec and immediately after each triplet was displayed with one of the 3 different exposure durations selected at random. Horizontal triplets were presented on the left of the cross to avoid the visual field defect; the final item (letter or digit) displayed horizontally and the last one displayed vertically occupied the same absolute location, corresponding to the fixation point. Participants (FC, C1 and C2; LDS, C8, C9 and C11) were asked to report all the three items.

Results

As shown in Table 4.6, FC's performance was significantly worse than C1 and C2 with letters (Wilcoxon test, $z = -3.2$, $p < .001$; Wilcoxon test, $z = 3.58$, $p < .0001$, respectively), as well as with digits (Wilcoxon test, $z = -3.77$, $p < .0001$;

	Performance								
		Horizontally				Vertically			
		100	150	250	Avarege	100	150	250	Average
FC	Letter	83	83	92	86	75	75	79	76
	Digit	83	83	96	87	75	87	100	87
C1	Letter	100	100	100	100	87	96	87	90
	Digit	100	100	100	100	96	96	100	97
C2	Letter	100	100	96	99	87	96	87	90
	Digit	96	100	100	99	83	83	100	89
LDS	Letter	62	46	62	57	58	50	71	60
	Digit	71	62	71	68	58	71	67	65
C8	Letter	100	100	100	100	100	100	100	100
	Digit	100	100	100	100	100	100	100	100
C9	Letter	100	100	100	100	83	91	92	89
	Digit	100	100	100	100	100	100	100	100
C11	Letter	100	100	100	100	100	87	100	96
	Digit	100	100	100	100	100	100	100	100

Table 4.6: Visual span. Percentage of letters and digits reported by patients and their matched control subjects at each exposure duration (100ms, 150ms and 250ms). The three letters and digits were presented at the left of fixation (horizontally) and just below the fixation (vertically).

Wilcoxon test, z =-2.32, $p < .05$, respectively). LDS also performed significantly worse than her matched controls C8, C9 and C11 with all the stimuli. However her performance is more impaired than FC's with both letters (Wilcoxon test, z = -4.4, $p < .0001$) and digits (Wilcoxon test, z = -4.6, $p < .0001$). Both patients showed the control pattern of being better with letters than with digits, although this was only a trend in FC (FC: F (1, 91) = 2.996, p = .087; LDS: F (1, 91) = 4.34, $p < .05$).

Unlike HR (Rapp and Caramazza 1991), both FC and LDS behaved in a qualitatively equivalent fashion with vertical and horizontal stimuli. This finding rules out an explanation of a deficit related to neglect or hemianopia. They do not show visual extinction. Overall, the pattern of patients' performance during this brief tachistoscopic presentation of stimuli indicated that LDS at least has a restricted visual span, and confirmed that LDS is much more impaired in processing letters than FC.

4.9 The ability to orient attention

The aim of this task is to investigate whether patients are able to orient and to move their attention in space adequately on a non-reading task. We used a typical visual search paradigm (Treisman and Gelade, 1980) where the detection of a target such as a red X among green distractors is easy when colour (or shape) alone can be used to discriminate the target from distractors - a X search, whereas detecting the red X among green Xs and red Os - a conjunction search is more difficult. In the first case the time taken to detect a feature is typically independent of the number of distractors, as the target ”pops out”, whereas in the second case the time taken to detect a conjunction of features increases linearly as the number of distractors increases. On Treisman and Gelade's (1980) theoretical perspective, the search for targets defined by a unique feature can be parallel, whereas search for a conjunction of features is serial. This paradigm was applied by Rapp and Caramazza (1991) who used displays of just 2, 4 and 6 items. In a feature search task they found a display effect for their LBL reading patient HR but not for control subjects. However, as Behrmann and Shallice (1995) pointed out, Rapp and Caramazza (1991) did not investigate whether HR's ability to detect a target reflected the left-right gradient observed in reading. Behrmann and Shallice's (1995) patient DS was presented with an analogous feature search task but did not show a display size effect and her RTs for right-sided targets relative to the left targets were not different from normal. We used this type of paradigm to investigate whether our two patients were able to distribute their attention in parallel across displays.

Materials and procedure

In the feature task, 1, 5, 15 and 30 circles (O) appeared on a PC screen: on half of the trials a red O was present among blue Os and on the other half only blue Os were present. The conjunction search task was the same except that the target was a red O and distractors were red Xs and blue Os. In both tasks the red circle was equally present in the leftmost part, in the middle and in the rightmost part of the screen in order to assess whether RTs are influenced by the target position. Patients had to perform a yes/no detection task and were instructed to press one key for present trials and another for absent trials as quickly as possible. The experimental design 4X2X2 with number of distractors, feature/conjunction, present/absent was implemented by means of an E-prime program for a total of 144 trials. Participants (FC, C1 and C2; LDS, C8 and C9) were given 9 practice trials; the display remained on the screen until a response was given. FC and LDS each did the experiment twice.

Results

Only the target present trials were analysed. FC made 7 mistakes (7/288) and LDS made 3 (3/288). The analysis of variance for each single subject (with feature/conjunction and number of distractors as factors) indicated that all the control subjects showed a significant display size effect in the conjunction task, as shown in Figures 4.4 (C1: $F(1, 30) = 10.4$, $p < .005$; C2: $F(1, 32) = 6.98$, $p < .05$; C8: $F(1, 34) = 16.85$ $p < .0001$; C9: $F(1, 32) = 29.3$, $p < .0001$). However there was no significant effect in the feature task, as expected from Treisman and Gelade's position.

The patients showed the same pattern of performance. Thus FC showed a display size effect in the conjunction task ($F(1, 66) = 24.9$, $p < .0001$), but not in the feature task ($F(1, 69) = 1.4$, $p = .24$). However his performance in the conjunction task was much slower than that of his matched control subjects. LDS showed no effect of display size in the feature task ($F(1, 66) = .45$, $p = .5$), but the effect was evident in the conjunction task ($F(1, 65) = 46.3$, $p < .0001$) where she performed very slowly compared to her matched control subjects. Neither patient showed an effect of target position in the conjunction task, when it was entered as a factor in the analysis of variance with number of distractors (FC: $p > .34$; LDS: $p > .57$). However, there was an effect of position for LDS in the feature task ($F(2, 66) = 8.5$, $p < .001$), but no interaction with the number of distractors ($F(2, 66) = .15$, $p = .86$).

These findings indicate that generally LDS was slower than her matched controls on the conjunction task and that on the feature task she tended to process the rightmost part of the screen more slowly, independent of the number of distractors. In fact, her visual search performance was not affected by an increase in the display size up of to 30 distractors, except that in the feature task she was slower with rightmost targets. This may be because of mild neglect of the right field. With respect to the two previous investigations, our experiment showed the performances of our two patients on a visual feature search task and on a conjunction search task which both comprise a wider range of distractors.

In a similar fashion to the findings of Behrmann and Shallice (1995), FC and LDS did not show a display size effect in a feature task. This result indicates

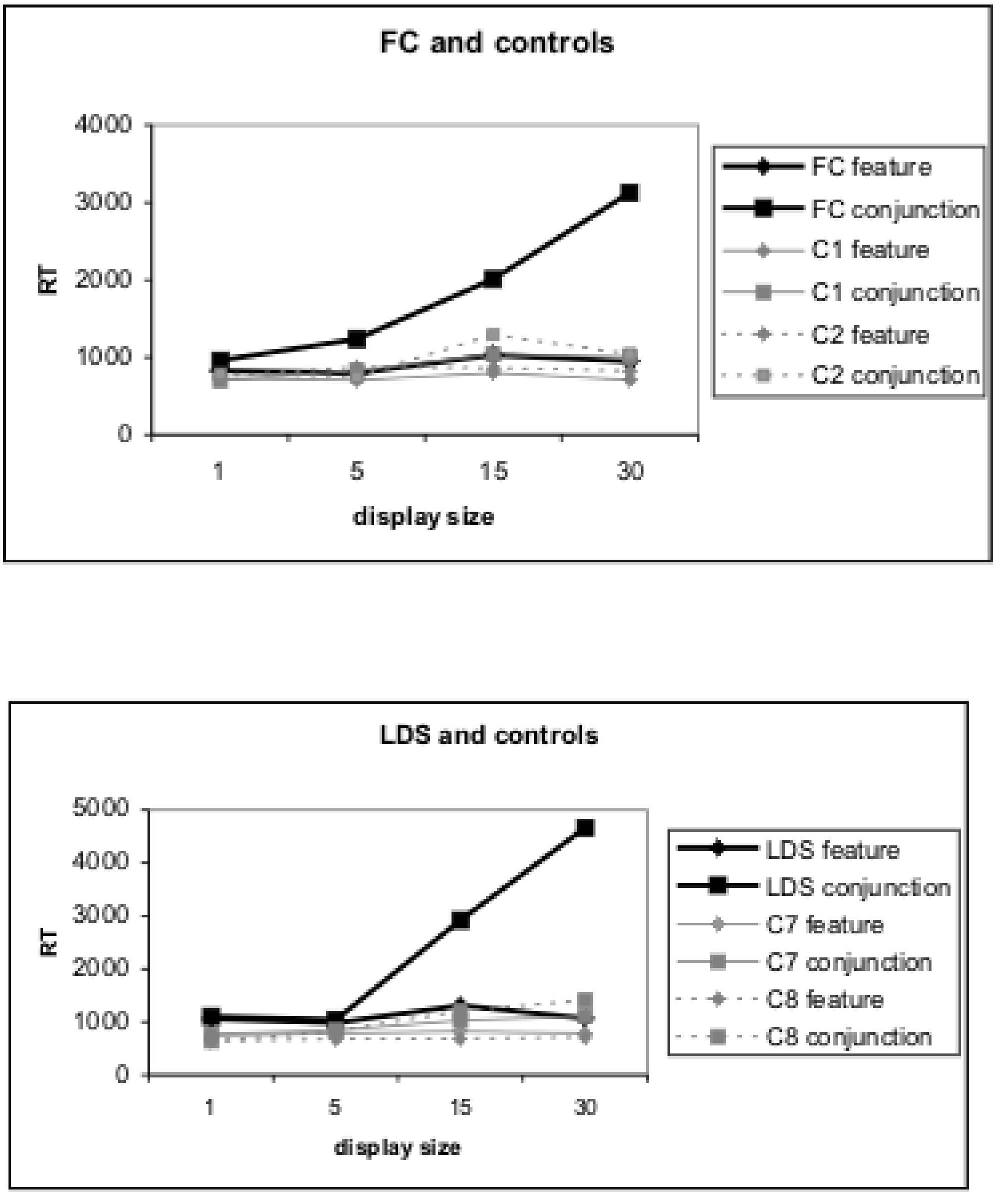


Figure 4.4: Treisman attentional task. RTs for FC and his matched control subjects on visual search as a function of display size for the feature and the conjunction tasks (first figure). Treisman attentional task. RTs for LDS and her matched control subjects on visual search as a function of display size for the feature and the conjunction tasks (second figure).

that their ability to spread their attention to detect a target that is in one of many spatial positions and that differs from distractors by a single feature is intact and the process is parallel.

The performance observed in FC and LDS differed markedly from that reported by Rapp and Caramazza (1991): there is no effect of display size in the feature task even with 30 distractors. Disturbances in the ability to distribute attention in space when letter analysis is not critical are not a necessary part of pure alexia. However the patients were much slower in the more serial conjunction task by comparison to their matched controls. This may derive from a possibly voluntary focusing of attention on the parts in more difficult visual tasks. In tasks such as reading that are demanding for FC and LDS their focus of attention in space may be narrower and this may result in a serial, slower processing of information (Ladavas et al., 1997).

4.10 Letter processing ability

In this section we assessed patients' ability at letter processing, in terms of speed and accuracy. This can be considered a core stage to access orthographic representation.

4.10.1 Letter naming

In this task we investigated the time taken to name each single letter. We were interested to see whether patients performed within the normal range.

Materials and Procedure

All the 21 letters of the Italian alphabet were presented 10 times each in a random order on a computer screen by means of an e-prime program for a total of 210 stimuli. Letters were printed in upper-case, in Arial font size 30, displayed in black on a white background. Patients and 14 controls (from C1 to C7 matched to FC and from C8 to C14 matched to LDS) were asked to name each letter; the patients performed the task 3 times. Mean and median RTs were measured for each participant (starting from the beginning of the vocal response).

Results

As shown in Fig. 4.5, FC's performance was well within the normal range ($z = 0.903$). By contrast, LDS was significantly slower in identifying letters than her matched controls ($z = 6.49$). Finally a direct comparison between FC and LDS showed a significant difference by considering the RT (Wilcoxon test, $z = -17.85$, $p < .0001$). Regarding FC, the letters that resulted in the slowest RT were H and I, with RTs of 166ms and 126ms above the average, respectively. However, this RT difference is well within the range of the difference shown by the controls matched to FC for some letters: letter Q being 190ms above the average (C2); letter V being 100ms, 140ms and 140ms above the average (C4, C5 and C7, respectively); letter O being 110ms above the average (C6). By contrast LDS was particularly slower with Q (500ms more than the average) and H (250ms more than the average) and became confused 4 times with Q, which she read as H. Unlike FC and his controls, the RT difference for LDS of 506ms and 264ms was much greater than the difference showed by her matched controls (maximum of 100ms above the average for letter V (C8, C10 and C12) and O (C11)). LDS is therefore qualitatively worse than her controls for particular letters. This is not the case for FC.

4.10.2 Digit naming

This task was carried out with digits instead of letters, for the same purpose as the previous one. Digits from 0 to 9 were presented 10 times in a randomised order using e-prime. Patients and 14 control subjects (FC, LDS, and from C1 to C14) were asked to name each digit written in black (Arial 30) on a white background. The patients performed the task twice. Mean and median RTs were measured for each participant.

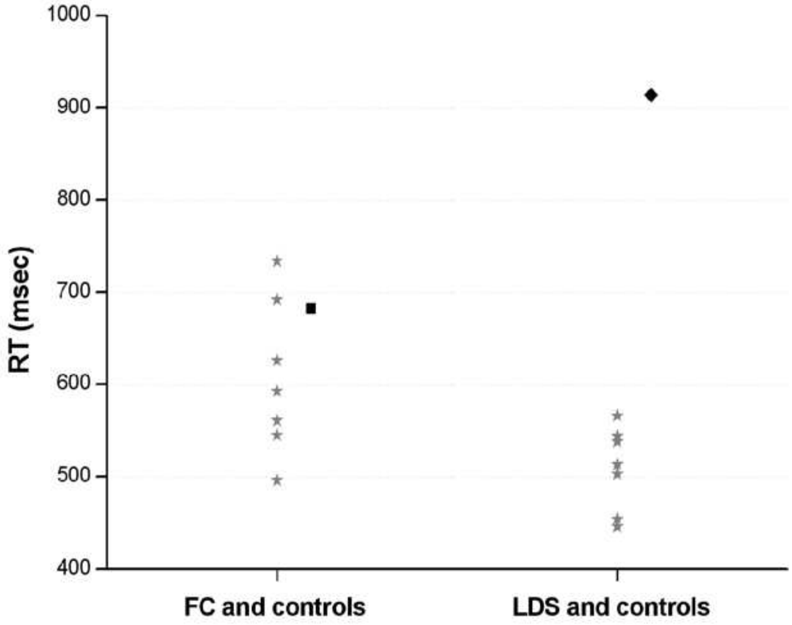


Figure 4.5: Letter naming task. RTs for FC and LDS and their relative matched controls.

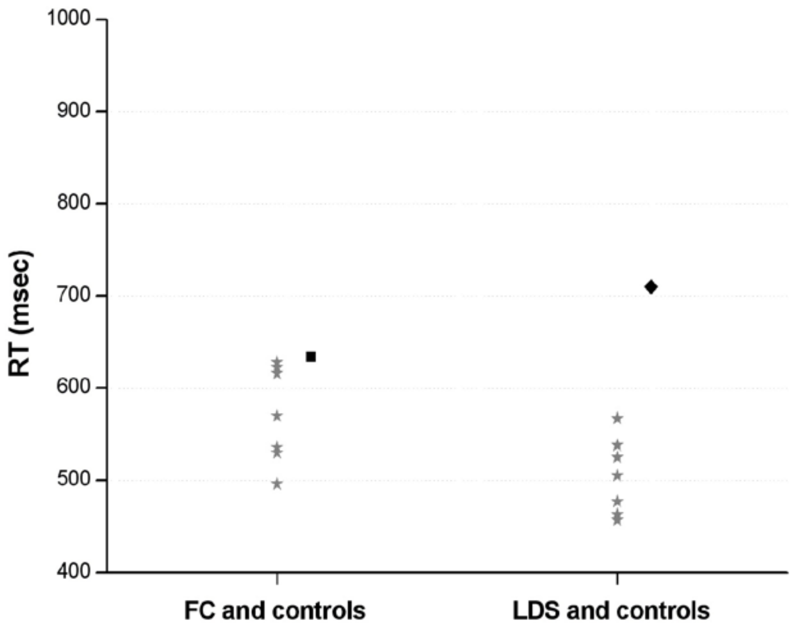


Figure 4.6: Digit naming task. RTs for FC and LDS and their relative matched controls.

Results

As shown in Fig. 4.6, FC's performance was within the range of the other 7 matched controls (z = 1.198). By contrast LDS's performance was worse than her matched controls (z = 2.54). The difference between the performance of FC and LDS is still significant (Wilcoxon test, z = -9.1, p <.0001).

4.10.3 Rapid letter recognition

In this task, taken from Warrington and Langdon (2002), strings of 6 letters were presented serially in the same spatial location for brief exposure durations. Participants are asked to say if a target letter verbally given prior to the string is present or absent in the 6-letter string. This experiment is assumed to assess the early stage of letter processing, when prior to identification a letter, letter-form or structural description are activated. Normal performance would indicate that apperceptive stage processing for letters (letter-form categorization) is adequate and that damage is likely to be at higher levels.

Materials and Procedure

Strings of 6 letters were presented serially, in the same spatial position without an interval between successive letters and with 3 different exposure durations (60, 80, 100 ms). Prior to the presentation of each letter string, a letter was named (e.g. 'L'). Patients and controls (FC, from C1 to C7; LDS, C8, C9, C12, C13, C14) had been instructed to say whether that letter was present or absent in the subsequent string (e.g. T S L A C V). Participants were tested in 9 blocks of 10 strings, 3 blocks at each of the three exposure durations in a Latin square design. Stimuli were printed in upper-case letters (Arial 30) and appeared black on a white background in the centre of the PC screen using the E-prime program. The target letter could occupy any of the positions 2-5 in the 6 item strings, and there were an equal number of 'Yes' and 'No' responses.

Results

As reported in Table 4.7, FC's accuracy was very good (93%), very comparable to his controls. In contrast LDS's performance was impaired with respect to all her matched control subjects C8, C9, C12, C13 and C14 (Chi2 (1) = 7.1, $p < .007$; Chi2 (1) = 13.4, $p < .0001$; Chi2 (1) = 10.98, $p < .001$; Chi2 (1) = 16.2, $p < .0001$, Chi2 (1) = 7.1, $p < .008$, respectively). In z-scores LDS's performance is -7.69 below the control mean. A direct comparison between FC and LDS showed that he performed significantly better (Chi2 (1) = 4.3, $p < .05$). The finding indicates that FC appears to have preserved letter-form activation and LDS has impairment at the level of letter- form processing.

	Performance			
	60	80	100	Total score
FC	25/29	28/30	30/30	83/89 (93%)
C1	27/30	29/30	29/30	85/90 (94%)
C2	25/30	29/30	28/30	82/90 (91%)
C3	27/30	24/30	30/30	81/90 (90%)
C4	30/30	29/30	29/30	88/90 (98%)
C5	29/30	29/30	30/30	88/90 (98%)
C6	30/30	30/30	24/30	84/90 (93%)
C7	24/30	28/30	29/30	81/90 (90%)
LDS	21/30	28/30	26/30	75/90 (83%)
C8	29/30	29/30	28/30	86/90 (95%)
C9	30/30	30/30	29/30	89/90 (99%)
C12	28/30	30/30	30/30	88/90 (98%)
C13	30/30	29/29	30/30	89/89 (100%)
C14	30/30	28/30	29/30	87/90 (97%)

Table 4.7: Rapid letter recognition task. Number of correct responses for each patient and control subject at different exposure durations (ms). Controls from C1 to C2 are matched to FC; C8, C9, C12 and C14 are matched to LDS.

4.10.4 Task with 13 digits and 2 letters

The previous task presumably tapped letter-form activation. To assess whether letter identification is slowed in pure alexic patients, Behrmann and Shallice (1995) presented their patient DS with 2 letters to report. The first and second letter appeared in the same spatial location but with different temporal intervals between them. The idea was that if single letter identification is slowed, than additional time will be required to process the first letter satisfactorily; this in turn will interfere with the processing of the second letter presented later. Thus, when the two letters are presented in a temporally adjacent fashion, the task is more difficult than if there is a gap between them. With respect to the previous experiment, this task requires not only letter-form activation, but also letter identification under conditions where stimuli are briefly presented and must be processed very quickly.

Materials and Procedure

A trial consisted of a string of 15 symbols (2 letters and 13 digits), which were displayed individually for 100ms under conditions of rapid serial visual presentation (RSVP). The 60 trials were divided into 3 conditions: when the 2 letters appeared either near to each other with no digit between them (e.g. 3942PL785134096), with 3 digits between them (e.g. 2045T327A069735) and with 7 digits between them (136G0754236D198). Letters were not presented in either the first or the final three positions of the string to avoid primacy and recency effects and there were no letters O and I because of possible interference with digits 0 and 1. Letters were all capitals, in Arial font size 30, as were the digits; the stimuli were displayed centred in black on a white background using the software E-prime. Patients and 13 controls (FC, from C1 to C7; LDS, C8, C9, C12, C13, C14, C15) were asked to report both the letters. This task was given 3 times to each patient (for a total of about 180 trials), but just once to control subjects.

Results

	Performance			
	Size gap	++	+-	-+
FC	0	(29/59) 49	(10/59) 17	(15/59) 25
	3	(42/61) 69	(12/61) 20	(6/61) 10
	7	(42/57) 74	(7/57) 12	(5/57) 9
From C1 to C7	0	Mean: 52% sd:23	Mean: 17% sd:16	Mean: 30%, sd:9
	3	Mean: 63% sd:27	Mean : 26% sd:24	Mean: 6% ,sd:5
	7	Mean: 69% sd:10	Mean: 11%, sd:8	Mean: 16%, sd:3
LDS	0	(5/60) 8	(26/60) 43	(19/60) 32
	3	(14/60) 23	(20/60) 33	(16/60) 27
	7	(7/59) 12	(13/59) 22	(35/59) 59
C8 C9; from C12 to C15	0	Mean: 78% sd: 18	Mean: 5% sd: 5	Mean: 15% sd:97
	3	Mean: 82% sd: 24	Mean: 16% sd: 22	Mean: 3% sd: 4
	7	Mean 91% sd: 9	Mean 2% sd: 3	Mean 6% sd: 6

Table 4.8: Task with 13 digits and 2 letters. Percentage of times in which patients and controls reported both letters correctly (++), just the first letter (+-) and just the second letter (-+). The control subjects' percentage is in italic script: C1 and C2 are matched to FC; C3, C4, C7, C8 and C9 are matched to LDS.

As shown in Table 4.8, FC's performance is clearly normal given the comparison with his 7 matched controls in the 3 conditions: with no digit, with 3 digits and with 7 digits between the two letters. Although there is some variability among controls, FC was roughly at the mean of normal controls of his age. This finding suggests that single letter identification operates at normal speed in FC. In contrast, LDS performed very much worse than her controls at all size gaps, even when the interval between the two letters increased. Again in this task FC had no difficulties in letter processing while LDS was slower in identifying letters; her performance indicates a letter identification deficit.

4.11 Summary of the letter and digit experiments

The results from the experiments on letter processing abilities suggested that FC had no difficulty in single letter recognition and in single letter identification (letter and digit naming; rapid letter recognition) and he processed letters very rapidly (13 digits and 2 letters). In contrast, LDS was slower in rapid letter recognition and much more impaired than FC in letter processing (letter naming; 13 digits and 2 letters). Overall, LDS has clear letter identification problems and FC is essentially normal in letter processing when compared with controls of his age. The difference between their performance is significant in the letter naming task (Wilcoxon test, $z = -17{,}85$, $p < .0001$), in the rapid letter recognition ($Chi2 = 4.3$, $p < .05$) and in the 13 digits and 2 letter task (Mc Nemar, $p < .0001$).

4.12 Orthographic integration ability

Processing individual letters is a prerequisite for being able to read a particular writing system like the alphabetic one efficiently. As discussed in Chapter 2 (last paragraph), when we see a word, single letter units are activated in parallel. The subsequent step consists in conjoining letters together to read the letter string. This integration process can proceed by different stages: letters are first conjoined in sublexical parts like bigrams, syllables and morphemes and then these visual percepts lead to the identification of the word. As mentioned, this visual integrative process which goes through sublexical levels has received less attention.

In this section we investigated the patients' ability to integrate letters in syllables and words. This ability is supposed to come into play once letter-form units have been accessed, thus revealing the serial nature of these abilities (letter processing and letter integration). The aim is to assess whether there is evidence of the sublexical processing and whether it can be selectively damaged in pure alexia.

4.12.1 Cumulative and successive presentation of words

This task, taken from Warrington and Langdon (2002) was designed to investigate whether purely serial processing of letters was all that was used by letter-by-letter readers. Two types of presentations were used. In the cumulative condition a letter was added one at a time to the letter string until the word was complete (e.g. G GE GEL GELA GELAT GELATO); in the successive condition each letter appeared one at a time in the appropriate spatial position but the preceding letter was removed when a letter was presented. In these two different presentation conditions there is a critical difference in the processing of letters in their spatial relation to other letters which are simultaneously present. Thus single letters appeared in the correct spatial position in the successive condition, but not in the cumulative condition where letters are added one at a time and spatial letter integration is possible with nearby letters. The LBL reader ROC studied by Warrington and Langdon (2002) showed a very similar type of performance on the cumulative and successive presentations: his accuracy was about 10% at 200ms exposure duration per letter, about 50% at 300ms and about 80% at 500ms in both conditions.

Materials and Procedure

A pool of 80 mid-frequency 6-letter words was selected and presented in 2 different conditions as described above. The word stimuli were presented individually on a PC screen on the left of a central fixation point using the E-prime program. Stimuli were black on a white screen printed in capitals (Arial 30). Words appeared for 100, 200, 300 or 500ms exposure duration per letter, so that words lasted for 600, 1200, 1800 or 3000ms. Four blocks of 20 words were presented in 4 increasing exposure durations in an ABBA BAAB design, so that the first 20 words displayed in the cumulative condition at each exposure duration were followed by 2 successive presentation blocks and so on.

The task was given 3 times to both FC and LDS and twice to C3, C4, C5, C6, C7, C9, C12, C13 and C14.

Results

Control subjects performed perfectly on the cumulative condition. This was very easy for competent readers, while in the successive condition they made errors at the brief time durations which they found rather difficult. As shown in Figures 4.7 and 4.8, the cumulative condition was easier than the successive one for the patients too. If we consider the successive condition, it is evident that LDS' performance was highly impaired; FC's performance is better than hers but not significantly in this condition. By contrast in the cumulative condition when letters were added one at a time until the word was complete, LDS' performance improved strongly and was much better than that of FC especially for short exposures: the difference in performance between LDS and FC was significant at 100ms of exposure duration (McNemar, $p < .002$), as well as at 200ms (McNemar, $p < .01$). In other words, FC's performance improved in a rather similar fashion in the cumulative and successive presentations as exposure duration increased, while LDS performed very differently in the two conditions.

It is worth noting that FC gave some unusual answers in the successive condition with longer exposure durations, when he could identify all the letters. For example with the word GIGLIO (lily) (at 500 ms per letter), he correctly spelled each letter, but he said ”GIUGLIO” (which does not exist); MOGLIE (wife) (500 ms) $>$”M..O..G..L..I..E .. I don't know the word!”; CODICE (code) (500 ms) $>$”C..O..D..I..C..E ..I don't know!”; LUMACA (snail) (500 ms) $>$”L..U..M..A..C..A .. MUCCA!; ESTATE (summer) (300 ms) $>$”E..S..T..A..T..E .. I don't know the word!; SANGUE (blood) (300ms) $>$”S..A..N..G..U..E .. I don't know the word!” These answers reveal that although FC is able to recognize the single letters of a word, he often fails to put them together correctly to read the word. Words were considered correct if they were either reported or

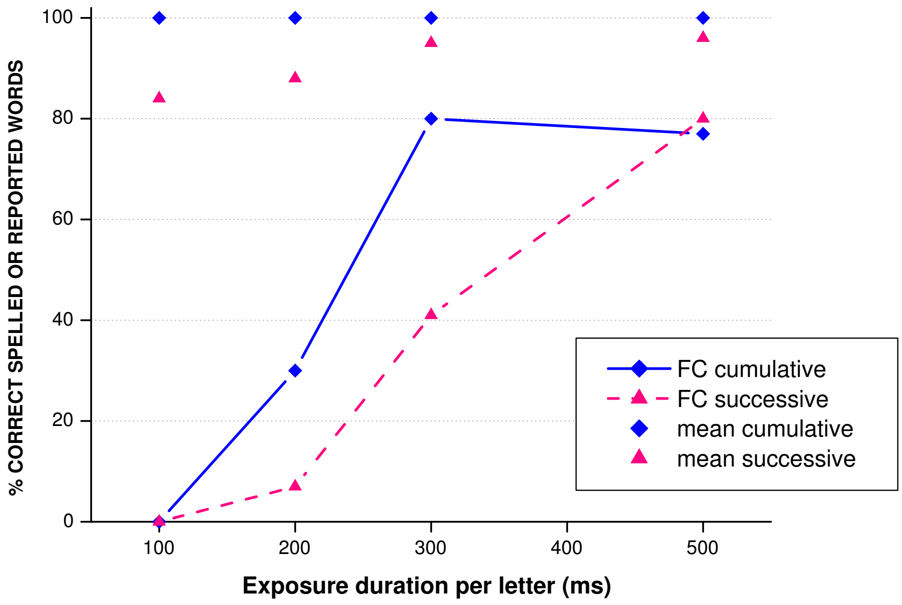


Figure 4.7: Cumulative and successive task. Performance of FC and his matched controls (on average) on the cumulative and successive presentation of words. Words were considered correct if they were either reported or spelled correctly.

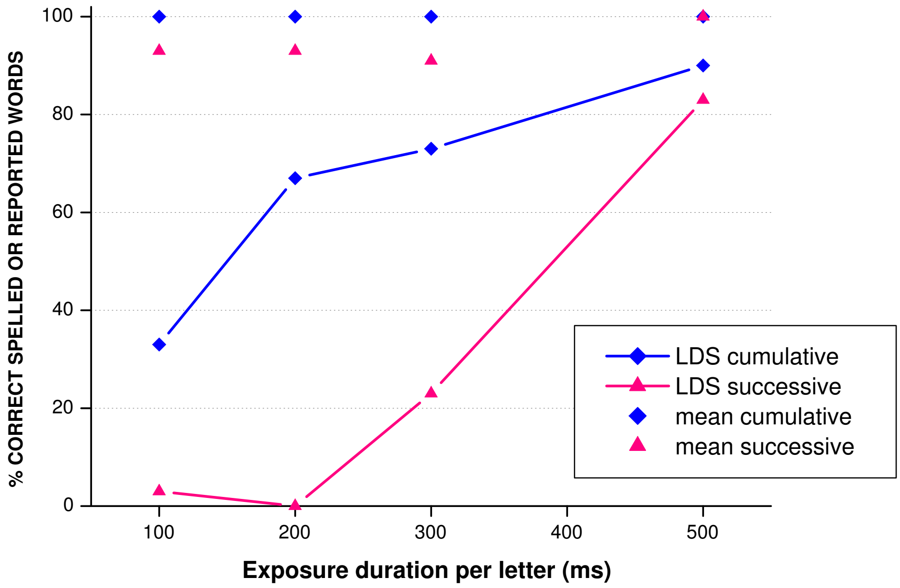


Figure 4.8: Cumulative and successive task. Performance of LDS and his matched controls (on average) on the cumulative and successive presentation of words. Words were considered correct if they were either reported or spelled correctly.

spelled correctly. Similar answers have already been reported in pure alexia: for instance by Patterson and Kay (1982). FC's performance was similar to the performance of the patient ROC (Warrington and Langdon, 2002) since the difference between the 2 conditions was not significant. However the authors claimed that the deficit of ROC was at the word-form system, with intact letter identification abilities. In this experiment, the performance and the type of errors made by FC provides further evidence that FC was not able to make effective use of any spatially parallel processing over different letter positions in order to read and integrate information about letters, while LDS was able to do so.

4.12.2 Syllable task

In this experiment our aim was to obtain further evidence as to whether the patients were able to integrate letters present simultaneously. We aimed to assess whether they could use information from syllabic structure, an intermediate level between letters and words. Evidence in favor of syllabic processing in reading words has been mostly obtained in languages such as Italian, French and Spanish with clear syllabic boundaries (e.g. French: Ferrand, Segui and Grainger, 1996; Ferrand and New, 2003; Spanish: Alvarez, Carreiras and Taft, 2001; Carreiras, Alvarez and de Vega, 1993; Carreiras and Perea, 2002). Moreover, in the study of Carreiras, Vergara and Barber (in press) colour changes coinciding with syllable boundaries lead to different evoked responses from those which did not so coincide: the manipulation of the sublexical information (i.e. of the syllabic unit) resulted in early ERP effects for both words and pseudowords in the time window of the P200. The onset latency of these effects was earlier than that of lexical variables which modulated the N400 component. This study suggests visual organization of syllabic structure as well as phonological. It is important to bear in mind that the fact that syllables are phonological units in speech does not necessarily imply that any syllabic effects observed in visual word recognition experiments are due to the activation of phonological codes (Taft, 1979 1987; Rouibah and Taft, 2001; Alvarez, Carreiras and Perea, 2004).

In the present experiment we investigated the ability of the patients to integrate letters into orthographic syllables, by comparing different conditions: when patients and controls were presumed to be able to process letters together (in orthographic syllables) compared to ones where this was more difficult, with nonsyllables (in condition A). The condition A has also been compared to the condition B (which comprised only syllable) where the ability to integrate letters into syllables was explicitly required.

Materials and procedure

A total of 80 triplets of letters was created and an ABBA design was used. In condition A, half of the triplets were orthographic syllables (e.g. por, man, dre, gri) and the other half nonsyllables, namely not orthographically plausible syllable (e.g. tsa, dsi, igf, mhi); items were randomly presented. Patients were asked to report the triplets as 3 single letters. In condition B, all the triplets were orthographic syllables, patients were told that the 3 letters were pronounceable triplets and they were asked to report the triplet as a syllable. The criterion according to which triplets have been divided into orthographic syllables (see Table A.1 in Appendix A) and nonsyllables (see Table A.2 in Appendix A) is consistent with the database of the orthographic syllables in written Italian (Stella and Job, 2001).

The 3 letters were presented for 165ms on a PC screen using the E-prime program; at the beginning of each trial a central fixation point appeared for 1000ms and following a 150ms ISI, the triplet was presented centrally in the screen in lower case letters, 35-point Arial font. The task has been presented to the patients and to the controls (C1, C2, C7, C8, C9, C13 and C14) and the instruction was to report the triplets as 3 single letters in condition A and as a

	Condition A			Condition B	
	Letters	Syllables	Nonsyllables	Letters	Syllables
FC	187/234 80%	20/38 53%	16/40 40%	199/237 84%	46/79 58%
LDS	178/231 77%	18/38 47%	11/39 28%	213/240 89%	53/80 66%

Table 4.9: Syllable task. Performance of FC and LDS on the syllable task where subjects were asked to report the triplet as 3 single letters in condition A and as a syllable in condition B. The table shows the number of letters and triplets reported in both the conditions.

syllable in condition B.

Results

All the 7 control subjects performed the task perfectly. FC as well as LDS were distracted in one trial, so the trial was excluded from each respective analyses. One triplet (scr), considered as a syllable, was not consistent with the database of orthographic syllables in written Italian (Stella and Job, 2001), therefore it was excluded from the analysis.

The answer was considered as an ordinal variable (which can be 0, 1, 2 or 3) and the Mann Whitney test was used to analyze the data. As shown in Table 4.9, FC did not show a significant difference between conditions A and B in terms of the number of letters reported (z = -1.63, p = .10) and of the syllables reported (z = -.695, p = .49; Chi2[3] = .25, p = .381). In addition, there was no significant difference for FC between the performance on syllables and nonsyllables in condition A (z = -1.02, p = .31). These results indicate that his performance was not sensitive to the presence of pronounceable units. By contrast, LDS showed a significant improvement between conditions A and B in terms of the number of letters (z = -3.84, $p < .0001$) and the number of syllables (z = -2,03, $p < .05$; Chi2[3] = 3.83, $p < .05$). This suggests that the explicit request to conjoin letters into syllables enhances her ability to put them together by

[3]The data have been analysed also with Chi2, by using the number of triplets reported by the patient as dependent variable.

inducing a broader attentional window. Moreover there is a significant difference between LDS's performance on syllables and nonsyllables in condition A ($z = -2.05$, $p < .05$) and a trend for the number of triplets reported ($Chi2^3 = 3.3$, $p = .057$).

Some triplets which were nonsyllables from an orthographic point of view such as *tsa, dsi, mhi, qem* and *qer* could be ambiguous from a phonological point of view (this is not the case for: *etm, qco* and *plc*). A triplet such as *tsa* can be pronounced as /tsa/ (which is not a phonological legal syllable in Italian) but also as /ça/, which is a phonological legal syllable in Italian. In that case, the assimilation in /ça/ is not automatic and this phonological syllable has a different orthographic representation, namely it is not written as *tsa*, but *za* (as in pizza, cozza, tazza). Another example is *vho* which could be pronounced as /vho/ (that is not a phonological legal syllable in Italian) but also as /vo/, which is a phonological syllable but with a different orthographic representation (as in volo, voce). However the fact that some "orthographic nonsyllables" could be pronounced as a phonological syllable might have influenced the ability to report the 3 letters.

Therefore it was assessed whether the number of letters reported by FC and LDS in the category "orthographic nonsyllables" was influenced by the possibility of being a phonological syllable. As a post-doc, the nonsyllables were divided into two groups: those which cannot be phonological syllables in Italian (like *bqo*, *etm* and *stc*, see Tables A.1 in Appendix A) and those which *might* be phonological syllables (although the assimilation, when possible, is not even automatic), but in that case they would have a different orthographic representation (like like *dsi*, *mhi* and *tsa*, see Table A.2 in Appendix A). The Mann Whitney test was used and the results showed that the ability to report the 3 letters of the orthographic nonsyllable was not influenced by the possibility of being a phonological syllable either for FC ($z = -.914$, $p = .420$) or LDS ($z = -.1.53$, $p = .185$).

This result fits with the previous finding that FC does not benefit from a possible integration of letters, while LDS can integrate letters into orthographic syllables. This capacity improves her letter identification and so in part compensates for her letter identification problems. These results show that although the performance of the patients is more impaired than that of their matched controls', LDS retains the capacity to conjoin letters together, and that her ability is significantly more effective than FC's even though LDS is slower than FC in word reading and in letter naming.

4.12.3 Summary of the orthographic integration ability

FC who is quick and accurate with single letters has difficulty to conjoin letters into syllables and words (cumulative and successive task; syllable task). On the contrary, having letters presented together in a typical arrangement facilitates LDS' performance considerably; moreover LDS is able to use information from the syllabic structure, an intermediate level between letters and words. Despite the fact that LDS has a deficit at letter processing, as shown in the previous section, she has a significantly better performance than FC in the cumulative presentation at the shortest exposure durations (McNemar, $p<.002$; $p<.01$); moreover in the syllable task, if it is considered the number of letters reported in condition B subtracted by the number of letters reported in condition A, we see that LDS reported more letters (35) than FC (12), although LDS is slower than FC in letter processing.

We can draw an important conclusion from this section: the difference between the performance of FC and LDS fits with the hypothesis of two qualitatively different forms of impairments giving rise to the letter-by-letter reading pattern typical of pure alexia.

4.13 Visual Imagery of words

An impairment in the visual identification of letters or words might suggest an impairment of mental representation of the same stimuli in pure alexia (Bartolomeo, Bachoud-Lvi, Chokron and Degos, 2002; Bartolomeo, 2002). In the few cases of pure alexia where identification of letters and words was difficult and imagery of letters was assessed, visual imagery has often been found intact (Behrmann, Moscovitch and Winocur, 1994; Goldenberg, 1992; Perri, Bartolomeo and Silveri, 1996). However, visual mental imagery for letters and words was found to be impaired in the pure alexic patient VSB (Bartolomeo et al., 2002). Recent cases of patients showing double dissociations between perception and imagery suggest that from an anatomical point of view occipital damage is neither necessary nor sufficient to produce imagery deficits (Bartolomeo, 2002; Goldenberg, 1998). However, extensive left temporal damage often accompanies imagery deficits. In this experiment we investigated whether the perception deficit shown by the patients for words also extended to the visual mental imagery.

Materials and procedure

Subjects were presented a list of 39 words auditorily, which were of 4 to 9 letters in length, matched for frequency. Words were divided into 3 groups, according to how they are written in lower-case: physically higher at the beginning compared to at the end, physically higher at the end compared to at the beginning or with physically equal size beginning and end ('beginning' and 'end' refer to the first and the last 2 letters of words). Examples of words which are higher at the beginning are: chiesa, clero, lupo; examples of higher words at the end are: erede, usignolo, patata and examples of 'similar words' are: cratere, zebra, ragno. Controls and patients (FC, C1, C2, C3, C4 and C7; LDS, C8, C9, C12 and C14) listened to the words and had to revisualise the visual form of a given word from memory, focusing in particular to the beginning and to the end. The task was to report whether the beginning and the end of the word were equal, or the beginning was higher or the end was higher. Six words were presented orally as practice trials. Subjects' answers were recorded with a microphone connected to a PC via a Cool Edit program. At the end of the task, subjects were asked to say whether they relied on the visual word form to accomplish the task.

Results

At the end of the task all the subjects said that they thought about the visual word form to answer. As shown in Table 4.10, FC's performance was comparable to his matched control subjects in terms of the percentage of correct answers and of the time taken to answer (z = 0,31). Although LDS was able to answer well above chance, her answers were significantly slower than her matched controls (z = 2,9). A direct comparison between FC and LDS indicated a pattern of performance not significantly different (Chi2 (1) = 3.03, p = .076) regarding the number of mistakes made, but significantly different (t (23) = -2.94, p <.01) regarding the time taken to accomplish the task. The mistakes made by LDS included 1 word with a physically higher beginning (abisso), 7 with equal size beginning and end (sedia, capra, brivido, zebra, vetro, ragno, tromba) and 1 word with physically higher end (custode).

These results suggest that FC can easily process the word form information in a visual mental imagery task. On the other hand, while LDS is able to perform the visual mental imagery task with words better than chance, she is slower and makes more errors than her matched controls. Although not tested, on the basis of these imagery, behavioural and also anatomical data (Bartolomeo, 2002; Goldgenberg, 1998), we could expect that LDS is slower also with single letters.

	Performance	Mean RT and sd	Errors	Autocorrections
FC	35/38 (92%)	5.7 (1.9)	3	4
C1	32/35 (91%)	5.3 (2.1)	3	2
C2	32/37 (86%)	6.8 (3.7)	5	1
C3	37/38 (97%)	3.9 (2.5)	1	0
C4	35/39 (90%)	4.1 (1.4)	4	0
C7	33/38 (87%)	4.9 (2.3)	5	1
LDS	28/37 (76%)	7.3 (2.4)	9	0
C8	38/38 (100%)	3.4 (1)	0	1
C9	37/39 (94%)	3.7 (1.5)	2	3
C12	36/38 (95%)	3.3 (1.6)	2	1
C14	36/39 (92%)	3.4 (1.15)	3	1

Table 4.10: Visual imagery task for words. Performance of patients and their matched controls. Words which resulted in either doubt or unclear answers for patients and controls were eliminated; answers with autocorrections were eliminated from the analyses of RT.

4.14 The use of LBL reading strategy: via spelling system?

According to Warrington and Shallice (1980), LBL reading is mediated by a strategy of letter naming, where reading is possible after naming each letter either either covertly or explicitly. Therefore a simultaneous articulatory suppression task, which would be expected to prevent other verbal tasks, should disrupt reading. We used a task developed by Warrington and Langdon (2002) to assess whether patients are able to read words during a simultaneous articulatory task. This is a critical experiment to investigate whether LBL reading is dependent, at least in our patients, on the viability of the letter naming strategy.

Materials and Procedure

Thirty-two mid-frequency and high-frequency 5-letter words were selected

	Condition	Mid freq words	High freq words
FC	no suppr	16/31 (52%)	28/32 (87%)
	with suppr	20/32 (62%)	29/31 (93%)
LDS	no suppr	12/16 (75%)	9/16 (56%)
	with suppr	12/16 (75%)	9/16 (56%)

Table 4.11: Reading with a simultaneous articulatory task. Proportion (and percentage) of correctly read words in the condition where the articulatory suppression was required and when it was not required. Words were presented for 300.

and presented in blocks of 8 in an ABBA design. Words were displayed in capital letters, in Arial font size 30 on a PC screen via e-prime. In the suppression condition when a ! appeared on the screen, patients were instructed to count from 1 to 10; while they were doing this a word was presented for 300ms and patients carried on counting until after the offset of the word. In the condition without suppression, patients were presented with words displayed for 300ms and were just asked to read them aloud. Mid- frequency words were presented in the same session as high- frequency words. The task has been presented twice to FC.

Results

Control subjects showed perfect performance in both conditions, since the task was generally easy for them. Surprisingly, patients also showed a good performance in the condition with the simultaneous articulatory task (table 4.11): LDS performed exactly the same with and without the articulatory task. Her errors are visual in nature (e.g. furia (fury) >furto (theft); gonna (skirt) >gomma (rubber); genio (genius) >gemma (bud)). FC had an even slightly better performance with the simultaneous suppression, although it is far from significant. He said, he concentrated more when the task was more difficult.

Since our LBL readers were able to read aloud during the concomitant articulatory task, these finding provide direct evidence that the LBL reading strategy adopted by our patients is not mediated by a spelling procedure. It is noteworthy that spelling is not commonly used in Italian, therefore this result may also be due to the type of language spoken.

4.15 Conclusions

In our investigation we present two cases of pure alexia who show normal language abilities (preserved writing, speech, comprehension), generally spared visuo-perceptual and visuo-spatial functions and normal memory skills but who are unable to read normally. They are mild LBL readers, showing a length effect in word reading, impaired performance under tachistoscopic presentation and difficulties with script (Warrington and Shallice, 1980). In this study we investigated whether there is a single primary source of deficit common to both patients as expected on the position of Behrmann, Plaut and Nelson (1998) or whether the two patients have different functional deficits from a neuropsychological point-of-view both giving rise to the overall pattern of performance characteristic of pure alexia (Price and Humphreys, 1992).

At a gross level the reading performance of FC and LDS had similar characteristics: they both performed accurately with single letters and digits, their reading speeds were similar and neither patient was able to access semantic representations from briefly presented words. However, on more detailed investigation major qualitative differences between their impairments emerged. Moreover there was a clear double dissociation between the patterns of deficits shown by the two patients. Of course FC and LDS differ considerably in age and educational level. However this alone cannot explain the difference in pattern, as the contrasts they show with each other also occur when their performance is compared with the appropriate 7 controls.

In the first set of experiments we assessed the abilities of the patients to identify and process letters independently of each other. FC performed well on letter recognition (rapid letter recognition) and on letter identification (letter naming) and he was able to process single letters very rapidly and accurately in a RSVP task (13 digits and 2 letters). Overall in such tasks he was generally as accurate and fast as his 7 matched control subjects. By contrast, LDS had difficulties in a task which required rapid letter identification; she was very slow at naming letters and to lesser extent also digits and unlike FC had especial difficulties with particular letters. She was impaired when letter identification was required at speed using the RSVP task. In these tasks LDS performed worse than both FC and her own matched controls.

In a complementary set of experiments we investigated the patients' ability in orthographic integration tasks which comes into play once letter-form units have been accessed. LDS' reading who had slowed letter activation was facilitated when groups of letters were presented together in space; she performed significantly better with cumulative compared with successive presentation of the letters in a word. In addition, it was evident from the syllable task that her reading performance improved if supra-letter units were simultaneously available (see Osswald, Humphreys and Olson (2002) for a related phenomenon). By contrast, FC was unable to use spatial information to form the letters he was correctly identifying individually into familiar groups; he gained no advantage in the cumulative over the successive presentation condition. In addition, he was not able to conjoin letters into supra-letter visual units, as he showed no superiority in identifying the constituent letters of syllables compared with random letters (syllable task).

Finally in a task requiring visual mental imagery of words, FC was able to perform as well as controls in assessing the visual forms of words. LDS was significantly slower than FC and somewhat less accurate. This suggests that for FC the visual word form system is intact but that this is not the case for

LDS.

What inferences can one draw about the nature of the underlying system and the damage to it that would produce the patterns of performance arising from the lesions in the two different patients? If one looks at the double dissociations in more detail, the tasks where FC performed better than LDS give rise to a classical dissociation in the sense of Shallice (1988) in that FC's performance is as good on the letter processing tasks and the imagery task as that of the matched controls. By contrast, where LDS performs much better than FC (orthographic integration tasks), one has a strong dissociation in that she still performed worse than her matched controls. Bullinaria and Chater (1995) and Bullinaria (1999) analysed the conditions under which two lesions to either the hidden units or the connections of a three-layer feed-forward connectionist net could give rise to complementary strong double dissociations. In general they found that they did not do so, but if the input-output mapping of the two tasks and the two different lesions were "made to measure" (Bullinaria, 1999 p 61) complementary strong double dissociations could in fact occur. However these theorists never described a situation where two complimentary lesions to comparable levels of a feed-forward net gave rise to a classical dissociation as well as a complementary strong one. Thus we take the observed pattern as providing powerful evidence that the impairments of the two patients are two different parts of a complex whole. Moreover given that in the reading system letter identification occurs before letter integration, this implies that impaired letter processing will result in a less efficient input to the letter integration process, even if this process is not itself damaged. With this organization of the system, one could never observe two classical dissociations. Complementary classical and strong dissociations are the most powerful combinations that could be observed.

Can one be more specific about the problems of the two patients? Consider first the model of Patterson and Kay (1982) which they derived from Shallice

(1981b). They argue that different forms of pure alexia exist -one (Type 1) with a lesion to the connection between the letter-form analysis and the word-form system and the other (Type 2) with in addition a lesion to the word-form system too. Hanley and Kay (1996) took a similar approach except that they appear to move the Type 1 impairment earlier in the system and assume it involves letter-form analysis system itself, but they retain the assumption that Type 2 has an additional impairment to the word form system.

In some respects this accounts fits well with the two contrasting patterns observed in our patients. However there are certain problems. First, FC has problems when integrating letters into syllables as well as into words. This however can be dealt with by a more complex take on how the word form system operates (see Shallice and McCarthy, 1985). On this approach it involves units of subword size such as consonant clusters, syllable onset and rime and syllables themselves, as well as words. The second problem is that unlike the patients Hanley and Kay (1996) discussed -PD, DS (Behrmann and Shallice 1995) and WL (Reuter-Lorenz and Brunn, 1990), the letter processing abilities of FC are intact. This would be compatible with the Patterson and Kay (1982) disconnection account , but not on the Hanley - Kay modification (1996), where a primary letter processing problem is assumed. The third problem relates to the loci of the two impairments. First, assume an isolable subsystem interpretation of the double dissociation and that there is a serial relation between the two subsystems (as there is between letter recognition and letter integration). If we assume in addition that all the tasks require the same set of input systems, but differ in where their processing requirements diverge from the standard track, then one can order the loci of the two impairments. The patient, who is completely spared on some tasks (e.g. letter processing), must have a locus of impairment later in the system than that of the patient who is impaired on all tasks by comparison with normal controls (e.g. letter processing and orthographic integration). This means that the locus of LDS's impairment must precede that of FC. However, this clashes with the anatomy of the two lesions.

In the light of these problems for the account given by Kay and colleagues, there seem to be at least two ways in which the double dissociation shown by FC and LDS can be explained. If one considers the development of the word-form model (Shallice and McCarthy, 1985; Dehaene et al., 2004), the initial stage of processing involves activation in both hemispheres of case-specific letter-form units in particular retinal locations. Then it can be assumed that in the word-form system, information across letters that are simultaneously present can be integrated to produce the activation of larger and larger orthographic units. As the subunits of the representation of a word like 'albero' (tree) are built, they can be utilised by the phonological reading route and the ultimate orthographic representation can be used by the semantic route. In addition we assume that an attentional control system (Ladavas, Shallice and Zanella, 1997) controls what portion of the letter string is admitted to the word-form system. As far as FC is concerned, his preserved visual imagery results would imply that the visual word-form system is undamaged which means it must be disconnected from earlier systems. The site of his lesion fits reasonably well with a disconnection of input to the VWFA from V1 of the left hemisphere as well as from fibres crossing the splenium from the right hemisphere. On this approach he would use the secondary letter identification system (see Fig. 5). It is most plausible that the input to such an intact letter identification system would be derived from outputs of letter-form systems in the right hemisphere.

Thus Dehaene et al. (2004) have shown that the more posterior parts of what they consider anatomically to be the VWFA are bilaterally activated by letter-form processing which is specific to a particular retinal location. This corresponds to the operation of our letter-form units. By contrast, more anterior parts of the anatomically-defined VWFA which were found to be activated

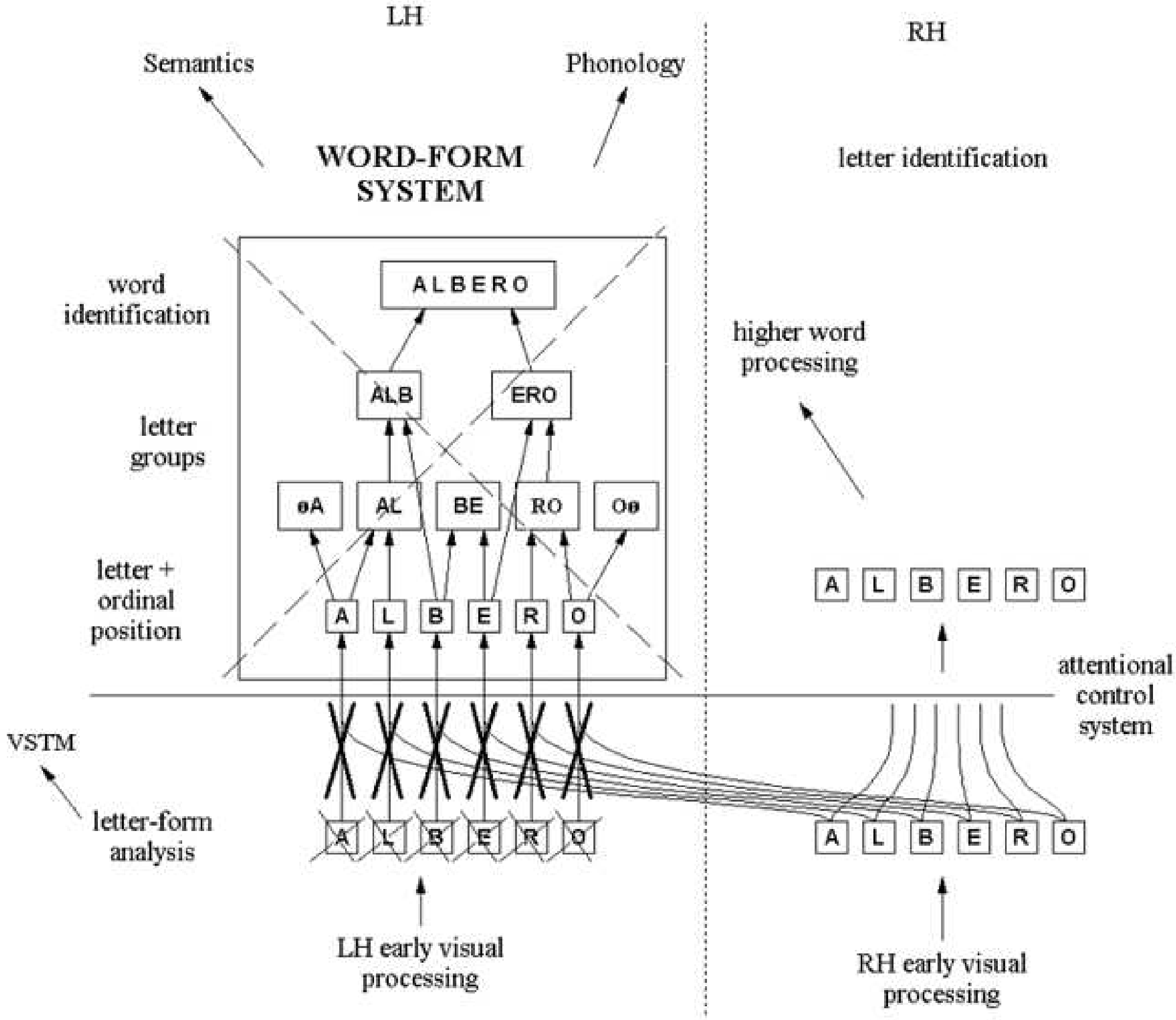


Figure 4.9: The explanation of the double dissociation. FC (solid line) would have an impairment in accessing the word-form system: it is assumed to be intact but disconnected from its visual input. By contrast LDS (dashed line) would have a partially damaged word-form system, which would still be used.

by more orthographically complex and less retinally-specific strings are lateralised to the left hemisphere. In contrast to FC, LDS would have a partially damaged left hemisphere visual word-form system, but would still use it for tasks involving letters either because of strategic functional-fixedness, typical of letter by letter readers (see Coslett and Saffran, 1993) or because her back-up letter identification system is less effective. It would be natural to assume, and this would fit with the anatomy of her lesion, that the letter-level aspects of the word-form system are also impaired. The lesion of LDS is where Cohen and colleagues (2000, 2002, 2003) consider the VWFA to be. However her partial ability to carry out the imagery task, albeit more slowly and with more mistakes than controls, together with the findings of her partial sparing of performance on the supra-letter tasks, suggest that her VWFA is not totally damaged.

A second possibility, for which however there is no direct supporting evidence, is to assume that the formation of larger units - at above the level of the individual letter - requires the functioning of a separable spatial integration system, and that it is this system that FC has lost. He would thus be able to process only a single letter at a time. On this perspective LDS will again have sustained partial damage to the word-form system but would continue to use it.

The primary aim of our study was to investigate whether these two cases of pure alexia could be explained by a single functional cause. The findings indicate that the nature of the deficit in pure alexia differs qualitatively across patients and that there is not just a single basic type of impairment common to all LBL readers. FC and LDS show complementary classical and strong dissociations. We interpreted this in terms of LDS having a lower-level deficit in letter processing, while FC having a higher level deficit in grouping together the letters that he can correctly identify.

As secondary aspects, we investigated whether patients showed implicit reading in 3 different types of semantic categorization tasks. Results showed that both of them had difficulties in suppressing the LBL reading strategy and they were unable to extract semantic information from briefly presented words. Moreover, as regards the LBL reading strategy used by FC and LDS, the results indicated that both of the patients, who had intact spelling abilities, were able to read words without using the spelling procedure.

Chapter 5

English pure alexic patients

5.1 Introduction

The results of the study on the Italian LBL readers provided strong evidence that in Italians pure alexia is a syndrome that can be caused by different functional deficits. It is argued that FC and LDS present two different forms of pure alexia: LDS has a lower-level deficit in letter processing, while FC has a higher level deficit in grouping together the letters that he can correctly identify.

The study of pure alexia in another language such as English has to take into account the cross-linguistic aspects of reading associated to the orthography, as already said in Chapter 2. English is a language with an inconsistent orthography and this difference with respect to Italian is likely to complicate the performance of dyslexic patients who use a serial strategy to read.

I had the opportunity to test our hypotheses at Professor Matt Lambon Ralph's lab on a new group of pure alexic patients in another language, namely English. Our hypotheses concerned the possibility to have different types of functional deficits with the procedure and the criteria used to investigate the Italian alexic patients. More specifically, from the study of our 2 Italian cases, at least 3 different points need to be considered to understand pure alexia: 1. letter processing abilities; 2. the orthographic integration of letters; 3. the LBL reading strategy.

Moreover the the effects that different orthographies have on the reading performance of patients with pure alexia have been considered. Italian has a transparent orthography in which the relations between orthography and phonology are highly regular. In English the relation between letters and sounds is often equivocal: some letters or letter clusters can be pronounced in more than one way (e.g. the vowel 'a' in hand, ball and span has 3 different pronunciations) and some sounds can be spelled in more than one way (Ziegler, Stone, and Jacobs, 1997).

Moreover, the fact that pure alexic patients read through a serial procedure might therefore be less disrupting in Italian than in English because the mapping between orthography and phonology is more consistent and can lead to the identification of the word more successfully. For instance, in English if the word *put* is read serially, one could start to pronounce /p/, then /pə/ and only at the end when you see the *t* you could change /pə/ into /pu/ to read "put". Two of the four alexic patients studied by Patterson and Kay (1982) had difficulties in assigning pronunciations to words, for instance made regularisations (ache >"aitch"), pronounced silent letters (sword >/sword/), violated the "final *e* rule" (e.g. mile >"mill") and had vowel digraph problems (duel >"dull"). By contrast, Italian is a transparent language where a serial reading strategy can take advantage of the consistent mapping between orthography and phonology and this type of mistake in more difficult to occur.

5.2 Case description

As mentioned, 3 new cases of pure alexia have been investigated: AT2, SC and TS. AT2 and SC are two LBL readers about seventy who suffered a stroke. TS is a younger LBL reader, about fifty, who underwent a surgical resection of a tumor in 2003. All of them can speak, understand and perceive relatively normally objects except when reading. Each of them showed further deficits: AT2 and SC have spelling problems and SC has some kind of frontal, perseverative behavior and SC and TS show some anomic difficulties. However the main symptom is an acquired reading disorder.

Case AT2

AT2 is a 69-year-old man with 10 years of schooling. He suffered from dysfluency (i.e. stutter) when he was younger, but the disorder disappeared as he grew up. AT2 left school at 15 years of age to join the army after which he worked in the mines for the majority of his working life. AT2's main interest for over 40 years was reading scriptures, in particular the bible. In December 2003 he suffered a cerebral vascular accident and a CT scan showed an infarct in the occipito-parietal region (see Fig. 5.1), probably affecting also the temporal area. His speech was affected and he suffered a right homonymous hemianopia. A short time after his lesion his speech rapidly improved (he could speak about religion or the jobs he used to do), his visual field defect changed to an upper right quadrantanopia and he stated that reading was his main difficulty. At the moment AT2 is an independent and self-sufficient person in everyday life. He has good auditory comprehension and speaks in well constructed sentences. His reading difficulties remain.

He has an acquired spelling problem, which is not associated with his premorbid stutter, though may be compounded by his relatively low educational achievement level. AT2 showed typical surface dysgraphia type errors in written spelling (e.g. 'yacht' >yot) with a score of 6/24 on PALPA length test (Kay et al., 1992). He was not able to spell nonwords to dictation (10/24), he was better at spelling regular words such as 'cat' (8/20), than irregular ones such as 'egg' (1/20; PALPA 44, Kay et al., 1992) and he shows a frequency effect scoring 8/10 on the high frequency words compared to 4/10 on the low frequency words

(PALPA 40; Kay et al., 1992).

Repetition of words is preserved (PALPA 9: 77/80) while his performance is worse with nonwords (PALPA 9: 50/80). His short-term memory is relatively intact; his performance is within the normal range on the digit span (forward: 6; backward: 4) as well as on the Corsi blocks (5).

When assessments of semantics used picture or written material, AT2 had some difficulties. For example he scored 44/52 on the 3 picture Palm and Pyramids Test (Howard and Patterson, 1992) which involves making a decision about the relationship of one picture (e.g. the Pyramid) to two others e.g. a pine tree or a palm tree). The Warrington auditory synonym test (Warrington, McKenna and Orpwood 1998) involves listening to a target word and then two related words to decide which is closest in meaning (for example: 'marquee' - 'tent/palace'). He had an impaired performance on this test with a score of 28/50. However, he was able to carry out the less demanding word-to-picture match in both auditory and written versions (PALPA 47 and 48; Kay et al., 1992) for which he scored 37/40 and 37/40 respectively. On both versions of this assessment, he was required to look at 5 pictures, one of which was the target, the others being a close semantic distractor, a distant semantic distractor, a visual distractor and an unrelated distractor (for example, Target: carrot; Distractors: cabbage, lemon, saw, chisel). He then read the target word and selected it or he listened to the target word and selected it. These assessments of semantics would suggest that AT2 did have some mild problems making semantic judgements but these only became apparent when the test materials were more challenging and in his everyday language he showed no evidence of semantic problems.

AT2 was no better at naming to definition (33/52) than naming to picture (38/52) suggesting that, on this assessment, he showed no signs of optic aphasia.

His perceptual abilities have been assessed with the VOSP, where he achieved

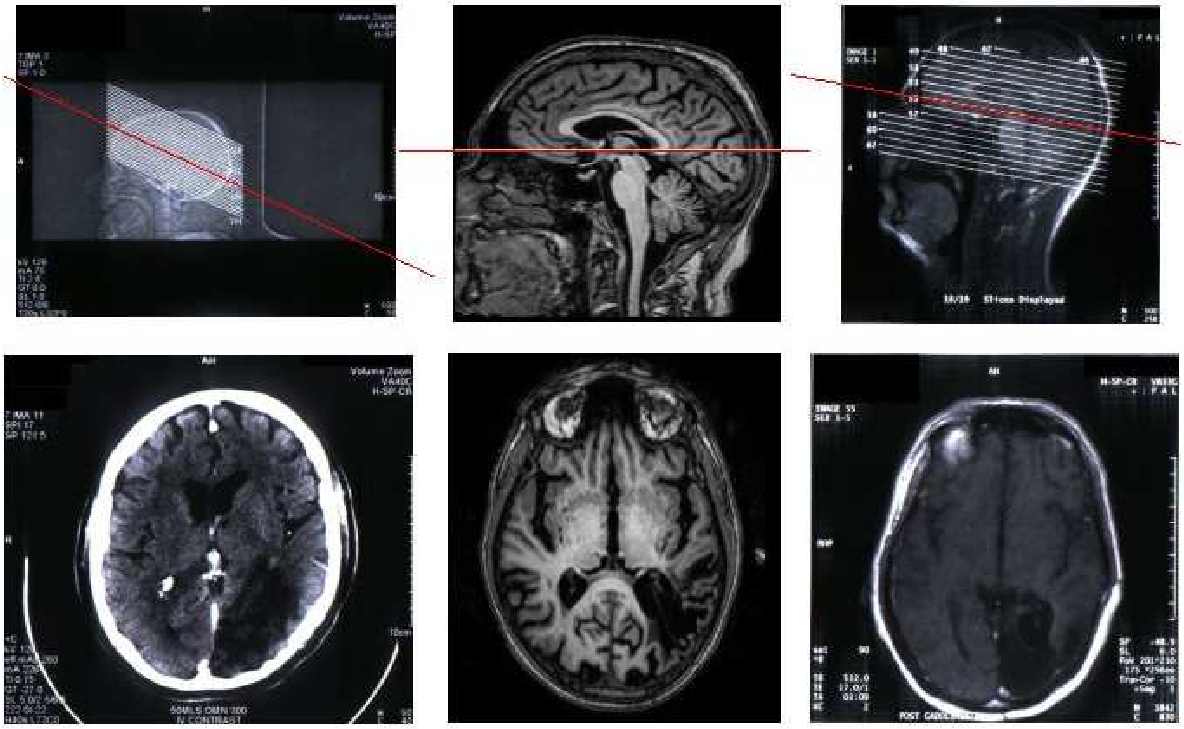


Figure 5.1: Scans of AT2, SC and TS, respectively. The scans of AT2 and SC show a lesion in the left occipito-parietal area; the lesion probably affected also the temporal area in AT2 and in SC; the third scan, of TS, shows a lesion in the occipito-temporal area.

normal scores except on the Silhouettes subset. AT2 showed a good performance with colours (8/11), while he had a weak performance with famous faces (11/92 with 4 anomic errors), although it should be checked with normal controls matched for age and schooling. Finally he can copy pictures well.

In short, AT2 can speak and understand well, and has intact general perceptual abilities. He has some spelling problems and some semantic difficulties. His main problem is in reading: AT2 is a slow LBL reader who tends to mistake visually similar letters.

Case SC

SC is a 76-year-old man with 10 years of schooling. He was the owner of a florist where he used to work with his wife. He suffered 2 strokes: the first one in the left occipito-parietal area, affecting also the temporal region (Fig. 5.1), resulting in a rather large lesion and the second in the right fronto-parietal area. He has a right homonymous hemianopia.

After the strokes SC showed severe deficits in speaking, in reading and also in understanding. SC has improved consistently since then; at the time of testing, SC had problems with speech due to his anomic difficulties, problems with phonology and writing (e.g. he tends to make regularisations like ”fox” >”focks”). In general, SC shows perseverative behaviour which is most likely due to his frontal lesion: for instances he tends to say a word repeatedly, sometimes if it is wrong he cannot break away from it. His main complaint is the inability to read.

His performance was normal on the Raven test matched for age (raw score: 25; mean: 23.03; sd: 4.62); when tested on the VOSP, the results showed very good performance except for the Number location subset, where he scored 4/10 (cut off 7). His performance on colour naming showed anomic errors (5/11 with 6 anomic errors).

Case TS

TS is a 52-year-old man with 10 years of schooling who worked as a trainee chef and then as a sales representative until he was 21, then he ran his own business selling motorcycles for 27 years. In 2003 he had a resection for a brain tumor in the left occipito-temporal area (Fig. 5.1). He has a right homonymous hemianopia. At the time of testing TS is almost totally self sufficient. He works voluntarily, twice a week at a hospital. TS has very good language production with excellent sentence structure. He does not have any problems in comprehension and his spelling abilities are intact, as indicated by the Naida Graham spelling task where his performance was within the normal range. He performed within normal limits on the Raven test matched for age (raw score: 22, mean: 26.75, sd: 5.13).

His performance was normal on all the subsets of the VOSP except for the Silhouettes test because of his anomic difficulties (he scored 11/30, making 12 anomic errors; cut off: 15). He showed severe difficulties with colours (6/11), for instance blue was named as red, gray as green, pink ”I don’t know!”.

5.3 Naming abilities

The 260 pictures of the Snodgrass and Vanderwart set (1980) were presented to all the 3 patients, in order to have a rough idea of possible perceptual deficits in object processing. Pictures were presented in an untimed condition on a PC using E-prime program. Patients were asked to name each object and in case they could not think of the name, they were asked to give a detailed description of it, so that one could understand they have recognized the object.

Results

Results are reported in Table 5.1. AT2, who was the slowest LBL reader, showed a good performance at object identification (83% accuracy). He showed some difficulties in recognizing living items compared to nonliving things and the living effect was significant (Chi 2 = 56,8; $p < .0001$). Examples of his errors are: lobster >locust; rhino >hippo; zebra >”horse, maybe a donkey”; orange >”a planet, a drawing”; strawberry >”some kind of pear”; ant >spider; bee>scorpion. However, as is widely-known the living items tend to be less familiar than the nonliving ones, therefore the comparison between the two groups remains only at a gross level since living and nonliving entities should be matched for (at least) familiarity. In this task, patients were presented with the whole picture naming test to see whether they showed difficulties in object processing, beyond the living and nonliving comparison.

Both SC and TS had an impaired performance, 68% and 64% accuracy respectively. They made several anomic errors, namely 35% and 66% of the total number of errors, respectively. An error was considered anomic was the patients did not name the target but gave a number of details sufficiently high to indicated that they recognized the picture (e.g. TS: ashtray >"Cigarette holder... You put cigarettes in it"; TS: pumpkin >"Vegetable, it is also used during Halloween"; TS: trafficlight >"In the street, it tells you when to stop and when to go"; SC: anchor >"You put it into the water and then you can't move"). By contrast, identification errors included semantic paraphasic mistakes (e.g. TS: scissors >knife; cannon >gun), visual errors (SC: nail >razor) the "I don't know" answers and all the object descriptions not sufficiently detailed (e.g. TS: carrot >"You eat them...").

If we consider the number of anomic errors made by SC and TS in addition to their correct responses, (columns SC+A and TS+A in Table 5.1) we see that SC's performance improves from 68% to 79% and TS's from 64% to 88%. As a conclusion, the ability to identify objects seems relatively intact for all the 3 patients (AT2's accuracy was 83%).

5.4 Experimental investigation

The experimental investigation focused on different aspects of the visual recognition of words, starting with a documentation of the patients' reading skill on a prose passage and on a word reading task. Then the patients' ability to process single letters was assessed, as well as their capacity to put them together to read. The last section is about the use of the LBL reading strategy. The aim is to consider the possible strategies that patients can use in order to read, not only based on spelling (Warrington and Shallice, 1980; Warrington and Langdon, 2002), but also on the visual image of the word (Patterson and Kay, 1982; Hanley and Kay, 1992).

		N	AT2 (%)	SC (%)	SC+A (%)	TS (%)	TS+A (%)
LIVING	Animal	38	29 (76)	32 (84)	32 (84)	23 (61)	30 (79)
	Birds	8	6 (75)	5 (63)	7 (88)	5 (63)	7 (88)
	Insect	8	2 (25)	3 (38)	3 (38)	4 (50)	5 (63)
	Fruit	11	4 (36)	4 (36)	4 (36)	4 (36)	9 (82)
	Veg	14	3 (21)	5 (36)	6 (43)	5 (36)	12 (86)
	Natural	7	6 (86)	6 (86)	6 (86)	4 (57)	6 (86)
	TOT	86	50 (58)	55 (64)	58 (67)	45 (52)	69 (80)
NLIVING	Objects	135	132 (98)	95 (70)	119 (88)	94 (70)	124 (92)
	Tools	16	16 (100)	9 (56)	9 (56)	9 (56)	15 (94)
	TOT	151	148 (98)	104 (69)	128 (85)	103 (68)	139 (92)
	Body parts	13	11 (85)	11 (85)	12 (92)	11 (85)	12 (92)
	Musical instruments	10	7 (70)	7 (70)	9 (80)	7 (70)	8 (90)
TOT		260	216 (83)	177 (68)	206 (79)	166 (64)	229 (88)

Table 5.1: Object naming (from the set of Snodgrass and Vanderwart (1980)). Number of pictures presented and correctly identified by each patient (with relative percentage). The columns SC+A (%) and TS+A (%) indicate the number of pictures correctly identified (with relative percentage) by SC and TS when also the anomic errors (A) were included. AT2 did not make anomic errors.

Patients have been studied with the same experimental procedures used for the Italian patients, namely the experiments were run with the same PC and with the E-prime program (except when differently specified); moreover a digital recorder was also used to keep record of the answers which have been also checked by an English colleague. English controls have not been included in the study yet; the key contrast is the direct comparison between English and Italian patients.

5.4.1 Documentation of reading skills

The ability to read has been investigated in all the 3 patients with a prose passage and with a word reading task.

Prose reading

Patients were presented with a prose passage taken from a famous English tourist guide (the Lonely Planet) describing the Italian climate. The text comprised 4 sentences for a total of 88 words. Text reading was recorded with a digital recorder and then analysed with a Cool Edit program. Normal readers take less than 1 minute to read the passage.

AT2 was very slow, taking more than 16 min to read it. He often did not realize his mistakes (e.g. situated >sighted; regarded >recorded; orientation: oriental) and a few times he was prompted.

SC took about 10 minutes to read it. He made 10 mistakes and sometimes he was prompted.

TS was quicker at reading the passage: he took about 5 min. He made 1 mistake and was never prompted.

Word reading

A pool of 120 words, 40 each of 3, 5 and 7 letters in length was selected. Words were matched for imageability and frequency (imageability and frequency ratings was taken from the MRC database, frequency from the Kucera-Francis written frequency) and appeared in 28-point Arial font to the left of a fixation point on a laptop screen using the E-prime program. As for the Italian patients The patients were asked to read the word when they were sure.

RTs were measured with the digital recorder and then analysed with a Cool-Edit program: just before a word appeared, a sound like a ”biip” was generated and the voice onset time was measured (from the ”biip” to the beginning of the vocal response), as for the Italian patients. In this task the digital recorder RTs were used (instead of the e-prime RTs) because the patients tended to make some conduites d’approche in normal reading, therefore the answers could be measured and analysed more carefully.

In order to be sure that the RTs with the digital recorder and with the E-prime recording (used with the Italian patients) measured the same latency, both the measures were employed in the letter naming task for all the 3 patients. The correlation between the 2 measures was 0.99, 0.97 and 0.96 for AT2, SC and TS, respectively, the slope was 0.98, 1 and 0.95, respectively and the intercept was 631ms, 683ms and 688ms, respectively. This indicates that the two procedures measure the same latency reliably and that the digital recording was generally 667ms slower than the E-prime recording. Thus the digital RTs have been converted in the E-prime RTs so that all the 5 patients could be compared.

Results

Although the patients were asked to read the word only when they were sure, they made some conduites d’approche. This type of answer has been removed from the analysis, so that the Italian and the English patients could be compared with the same procedure (the voice onset time) without conduites

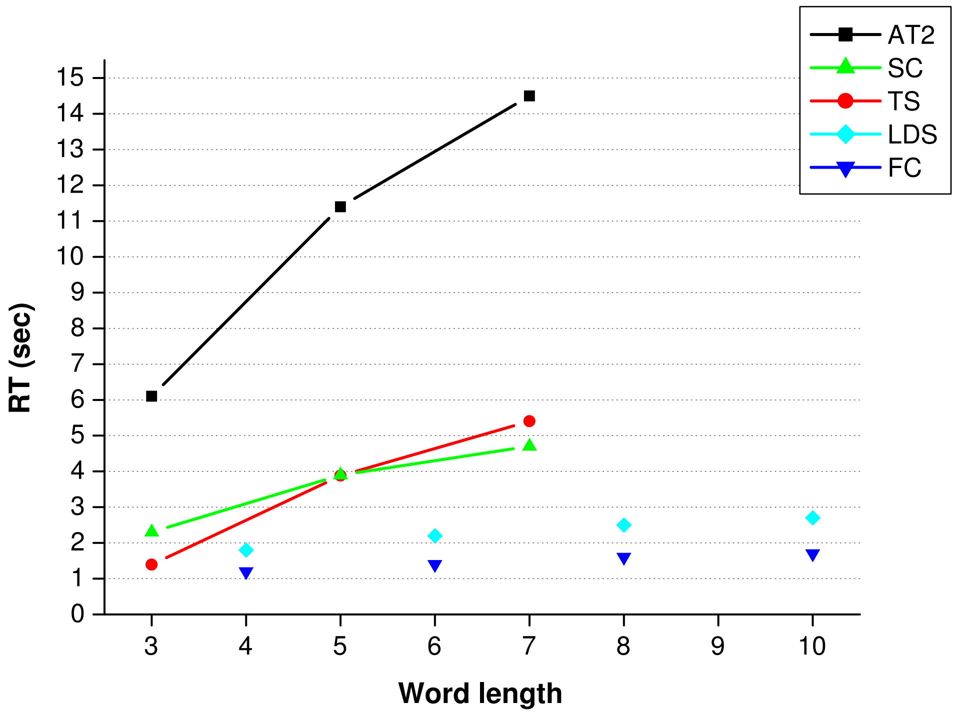


Figure 5.2: Word reading. RTs (sec) of AT2, SC and TS and the Italian pure alexic patients (FC and LDS) as a function of word length. The voice onset time has been measured and errors and conduites d'approche were removed from the analysis.

d'approche.

By performing an ANOVA with frequency, word length and imageability, a significant word length effect emerged for all the 3 patients (see Figure 5.2). AT2 ($F(1, 82) = 12.74$, $p < .001$) had a reading speed of 2.1 sec per letter, SC ($F(1, 69) = 4.59$, $p < .05$) showed an increment of 0.6 sec and TS ($F(1, 105) = 53.98$, $p < .0001$) showed an increment of 1sec per letter.

AT2, the slowest reader, made 25 mistakes and 7 conduites d'approche, which have been removed; he did not show any effect of frequency or imageability on RTs. On this task he was often able to identify the letters correctly, without being able to conjoin them together in an automatic way (e.g. ink >"'i' 'n' 'k'.. /aink/.. ink!" (45sec); paint >"'p' 'a' 'i' 'n' 't'.. paint?"; pat >"'p' 'a' 't'... pat"; vehicle >"/vi/../aitch/.. vehicle (40sec). Generally, AT2's mistakes reveled the difficulty to assign a correct sound to an ambiguous letter or group of letters: e.g. red >/rid/; net >/nit/; war >"were"; earth >"ears"; fur >"far"; group >"grope". Moreover some mistakes show that during the serial reading, letter integration which proceed by assembling phonemes which overrule the pronunciations assigned to earlier parses does not occur automatically, for instance: bedroom >/bi'drom/.. /bi'droom/.. bedroom!"; delight >"/del/../deli/.. delicate?"

In this task SC made 25 conduites d'approche (5 with 3-letter words, 12 with 5-letter words and 10 with 7-letter words), 13 errors and 7 times he pronounced the word correctly under his breath, but without recognizing it or without being sure of it (e.g. bet: "/bet/, /bit/? /Bet/? Not sure!"; essence >éssence.. essènce.. éssence? essènce? I don't know!"; peace >"/peis/, peace, /peis/, peace? Not sure"). We considered correct only words SC recognized as right and without conduites d'approche: on this criteria, we excluded 45 words. He showed an effect of imageability ($F(1, 69) = 4.7$, $p < .05$) and an effect of frequency ($F(1, 69) = 7.6$, $p < .007$). Many errors and conduites d'approche revealed the difficulty to assign the correct sound to some letters and letter cluster, e.g. anger >anʤier.. /enʤier/.. 'a' 'n' 'g'.. anger!"; cause >"/kau/.. kuis.. cause"; grace

$>$”grass? Grace!”; merit $>$”/merít.. /marait/.. merit!”. Other errors suggested that the serial reading strategy might pose a further problem for the recognition of words in English (e.g. student $>$”/stədnt/.. ’s’ ’t’ ’u’ ’d’.. /stiu/... student!”; value $>$/v/.. /vel/.. value!”). Finally other errors were more visual in nature, e.g. blame $>$”plane.. plaam, pleem.. Oh B! Blame!”; century $>$country; olive $>$”alive”; machine $>$”matching”.

This type of errors is very similar to that made by the two pure alexic patients TP and KC described by Patterson and Kay (1982) who made mistakes in assigning pronunciations to words (e.g. pin $>$”pine”; back $>$”bake”; group $>$”grope”)

TS made only 3 mistakes and 4 conduites d’approche (e.g. anger $>$”angel.. no anger”; officer $>$”office.. officer!”) which have been removed from the analysis. He showed an effect of imageability (F (1, 105) =13.77, $p < .0001$). Generally he was pretty sure of the word, without having hesitations.

As is shown in Figure 5.2, all the English patients (especially AT2) are slower that the Italian ones. From the point of view of the latencies, TS and SC look similar to each other at reading words; however TS hardly has hesitations while SC made several conduites d’approche. Since the instructions were the same for all the Italian and English patients (namely, they were asked to read the word when they were sure), in principle we could compare patients’ performance by including also the conduites d’approche made by all the English patients patients (see Figure 5.3). In that case RTs extends significantly especially for SC.

5.4.2 Letter processing ability

After having assessed the patients’ abilities on general tasks and on reading tasks, we investigated how much their slowness in reading can be traced back to a letter processing level.

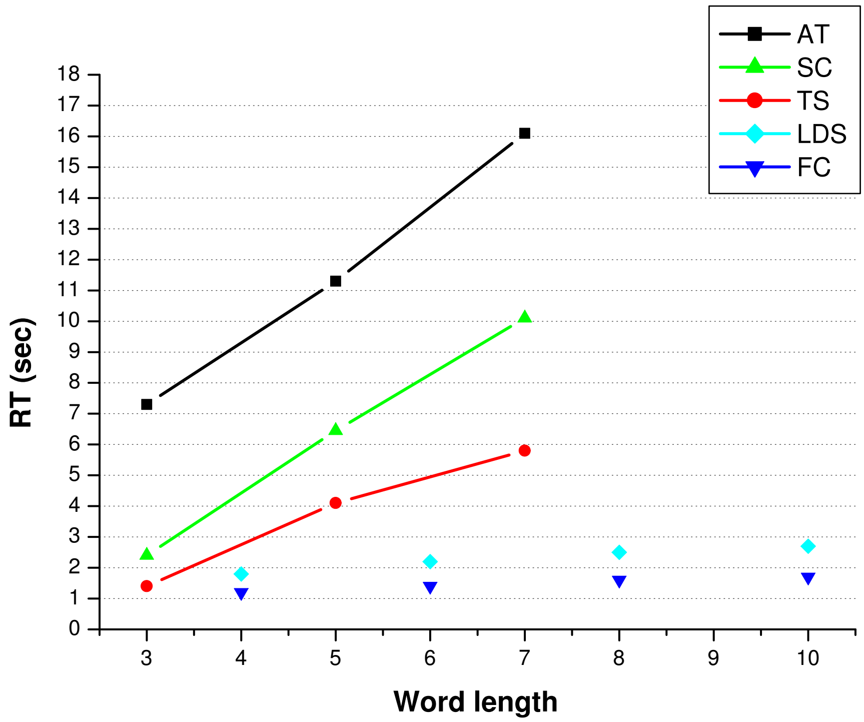


Figure 5.3: Word reading. RTs (sec) which include also conduites d'approche for AT2, SC and TS and the Italian pure alexic patients (FC and LDS) as a function of word length. Voice onset time was measured and conduites d'approche were included in the analysis.

Letter naming

In this task we assessed the time taken to name each single letter of the English alphabet. We were interested to see whether the patients were generally slow and whether they showed any difficulty with particular letters.

All the 26 letters of the English alphabet have been presented 10 times each on a computer screen via e-prime program in random order. The 260 letters were printed in uppercase Arial font size 30, displayed in black on a white background at the left of a fixation point. RTs were recorded with e-prime, so that the results with all the pure alexic patients (Italian and English) were easily comparable.

Results

Results (see 5.4 showed that AT2 was very slow at letter naming, taking generally 1,5sec to identify a single letter. He made 13 mistakes, 7 of which were with the letter G. AT2 was significantly slower with the letter J, more than 2sd above the mean.

Regarding SC, he appeared slow at letter naming too, taking 820msec on average. He made 12 errors, 3 with letter G, 2 with J and 2 with D.

Patient TS was the quickest at letter naming: he took generally 720msec to identify the single letters (with e-prime RTs). He made 3 mistakes (2 with letter I) and letter K resulted in the slowest RTs, almost 2sd above the mean.

At a letter level there is no reason to assume that English is more difficult than Italian; therefore English controls are not yet available, the English patients were compared to the Italian controls. In particular AT2 and SC who are 69 years old with 10 years of schooling can be compared to FC's controls (mean age: 84.4; mean schooling: 11,7), while TS who is 52 years old with 10 years of schooling can be compared to LDS's controls (mean age: 58.7; mean schooling: almost 13 years). According to this provisional control group, all the 3 English patients appear slower at letter naming (AT: $z = 10.31$; TS: $z = 4.63$; SC: $z = 2.21$).

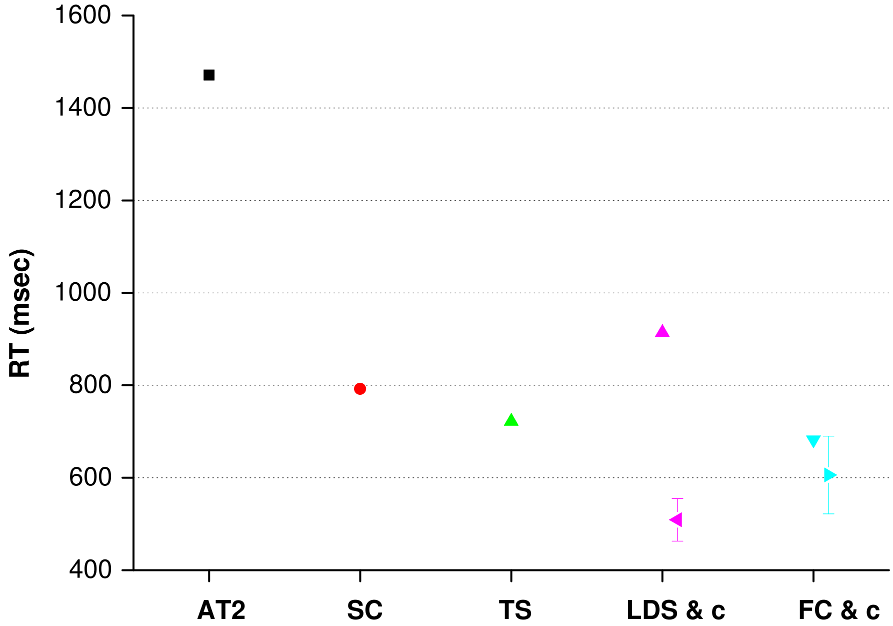


Figure 5.4: Letter naming task. RTs (in mesc) for AT2, SC and TS and the two Italian pure alexic patients FC and LDS with the means and standard deviations of their matched controls.

Rapid letter recognition

This task, taken from Warrington and Langdon (2002), aimed to assess the time taken to recognize a letter, a stage that occurs before letter identification.

Materials and Procedure

As for the Italian patients (see Paragraph 3.10.3), strings of 6 letters were presented serially in the same spatial position with the same procedure used before. Letters which belong to the English but not Italian alphabet have been added (e.g. X, K, W). The same durations were first used (60, 80, 100ms); then longer exposure durations were chosen (150, 200 and 250 ms) because the accuracy of 2 patients was very low, in particular AT2 was at chance (see below). These new durations are the same used by Warrington and Langdon (2002). Moreover since patients were not always accurate at letter naming, a letter was named and also drawn e.g. L prior to the presentation of each letter string, to be sure that patients did not become confused. Patients were asked to say whether the letter was present or absent in the subsequent string (e.g. T K J L G M). Patients were tested with 30 trials per duration, for a total of 90 trials.

Results

With the shorter durations (60, 80, 100ms) used with the Italian LBL readers, AT2 was almost at chance level (57%), scoring 51/90 (15/30, 19/30, 17/30, respectively). TS, the quickest patient on word reading and letter naming, showed a better performance, but still impaired: 60/90 (67%) of accuracy (20/30, 18/30, 22/30, respectively).

Results with longer durations (in Table 5.2) showed that AT2's performance was still impaired (67%), while SC and TS were quicker and more accurate at recognizing letters (90% and 89%).

If we compare the English patients' performance to the Italians', we see that FC scored 93% (mean of his matched controls: 93%) and LDS scored 83% (mean of her matched controls: 98%) with the shorter exposure durations. This might

	Performance			
	150	200	250	Total score
AT2	15/30	23/30	22/30	60/90 (67%)
SC	23/29	28/29	28/30	79/88 (90%)
TS	27/30	27/30	26/30	80/90 (89%)

Table 5.2: Rapid letter recognition task with the English patients. Number of correct responses for each patient at different exposure durations (150ms, 200ms and 250ms).

suggest that the 3 English patients are more impaired than the 2 Italians at letter recognition level (but not because of differences in the orthography).

Summary of the letter processing abilities

The results of this section indicate that all the patients appear slow at letter naming, especially patient AT2. This slowness seems to be due to a letter recognition deficit. In fact in the rapid letter recognition task both SC and TS show a good performance (although not at ceiling), but the exposure durations are slower that those used with the Italian patients. These findings suggest that all the patients start to have a deficit at letter level which starts to be evident in letter recognition.

5.4.3 The visual span

As was done for the Italian LBL readers, visual span was investigated to assess patients' ability to identify more than one stimulus at a particular exposure duration.

Materials and Procedure

The same procedure used with the Italian patients has been performed with the English ones (see Paragraph 3.8). The only difference is the exposure duration which has been extended (from 100, 150 and 250ms): 250, 350 and 400ms

		Performance							
		Horizontally				Vertically			
		250	350	400	Average	250	350	400	Average
AT2	Letter	38	46	50	44	50	54	46	50
	Digit	67	63	54	61	57	63	58	59
SC	Letter	48	42	46	45	58	58	63	60
	Digit	67	63	63	64	71	67	67	68
TS	Letter	67	71	75	71	75	88	83	82
	Digit	75	67	71	71	83	96	100	93

Table 5.3: Visual span with the English patients. Percentage of letters and digits reported by patients and their matched control subjects at each exposure duration (250ms, 350ms and 400ms). The three letters and digits were presented at the left of fixation (horizontally) and just below the fixation (vertically).

because they were slower at letter identification.

Results

For each participant individually an analysis of variance was carried out with 3 factors: Horizontal/Vertical, Letters/Digits and Exposure durations (250, 350 and 400). The results are shown in Table 5.3.

As shown in Table 5.3, AT2 showed a significant effect of letters versus digits ($F(1, 90) = 13.3$, $p < .0001$). SC showed a significant effect of type of stimuli ($F(1,89) = 25.13$, $p < .0001$), of type of presentation ($F(1, 89) = 12.12$, $p < .001$) and a trend for the interaction between the 2 factors ($F(1, 89) = 3.77$, $p < .055$). TS showed a significant effect of horizontal vs vertical ($F(1, 91) = 19.2$, $p < .0001$).

5.4.4 Orthographic integration ability

As was done for the Italian patients, we investigated the ability of the patients to conjoin letters, once they have been recognized. However, while it was possible to replicate completely the methodology used for the Italian patients in tasks of letter processing here we need to consider a few points. First, the possible effects of the inconsistency of the English language on letter integration; second the fact that reading in English relies more on the lexical route and therefore a serial reading strategy can be particularly disrupting in English; third, English does not have clear syllabic boundaries in the recognition of either spoken words and in written words. English readers can segment words according to orthographic sublexical units that do not necessarily correspond to phonological syllabic units (Taft, 1979; 1992). Therefore the syllable level is not worth studying.

We investigated the patients' ability to integrate letters with the cumulative and successive task, as described in Chapter 4. It is worth noting that the successive condition leads to the use of a serial LBL reading strategy to read the word. By contrast, the cumulative condition allows one to integrate letters in subparts also with the help of the lexical route which comes into play at the end when the word is complete.

Methods

A total of 80 mid-frequency 6-letter words were used: it was the set created originally by Warrington and Langdon (2002). The task is the same used with the Italian alexics (see Chapter 4). As in the rapid letter recognition task and the visual span, longer exposure durations per letter were chosen (200, 300, 500 and 700ms), since English patients were slower than Italian ones (previously we used: 100, 200, 300, 500ms). The same durations were used for all the patients in order to compare their performance. An ABBA + BAAB design was created (with the same set of words presented once in the cumulative condition and in another session in the successive condition). The task was presented twice; the second time words were exchanged with different durations with respect to the first time, so that the set of words was presented at different durations in a randomized order. The ABBA + BAAB design was maintained.

Results

As shown in Table 5.5, AT2 showed a similar performance in both the presentations, with a little, not significant improvement in the cumulative condition. Only at 200ms of exposure duration per letter did he read a number of words significantly higher in the cumulative than in the successive condition (McNemar, $p < .05$).

As regards SC, the different performance across the cumulative and successive presentations is more marked and significant at 300ms (McNemar test, $p < .05$), at 500ms (McNemar test, $p < .05$) and 700ms (McNemar test, $p < .05$). With respect to the performance of AT2 on the cumulative condition, SC's one is significantly better (McNemar test, $p < .001$). These results suggest that SC maintains some ability to integrate letters into words in order to read, while AT2 is impaired.

TS showed a better performance on this task in both the presentations and he is almost at ceiling when exposure durations are longer than 400ms. The improvement on the cumulative condition compared with the successive one is marked and significant at 200ms (McNemar test, $p < .05$) as well as at 300ms (McNemar test, $p < .0001$). However, since the control subject used by Warrington and Langdon showed a significantly better performance than TS's on the cumulative task at 200ms of exposure duration (Chi2 = 1.88, $p < .242$), we can assume that TS' integration abilities are better than the other two patients AT2 (McNemar test, $p < .0001$) and SC (McNemar test, $p < .0001$), but they are still impaired.

The type of errors made by the patients is closely associated to the inconsistency of the English orthography that makes word reading difficult strike (at 300ms with cum) >"str/i/k"; bright (at 200ms with succ) >"/bri/... bridge"; moreover the use of a serial strategy to read seems to lead to incorrect responses. For instance SC made several errors where silent letters were pronounced: island (at 300ms and 700ms with the successive presentation (succ)) >"I..S..L..A.. Islam?" (twice); listen (at 300ms with succ) >"list...listen??"; knight (at 200ms

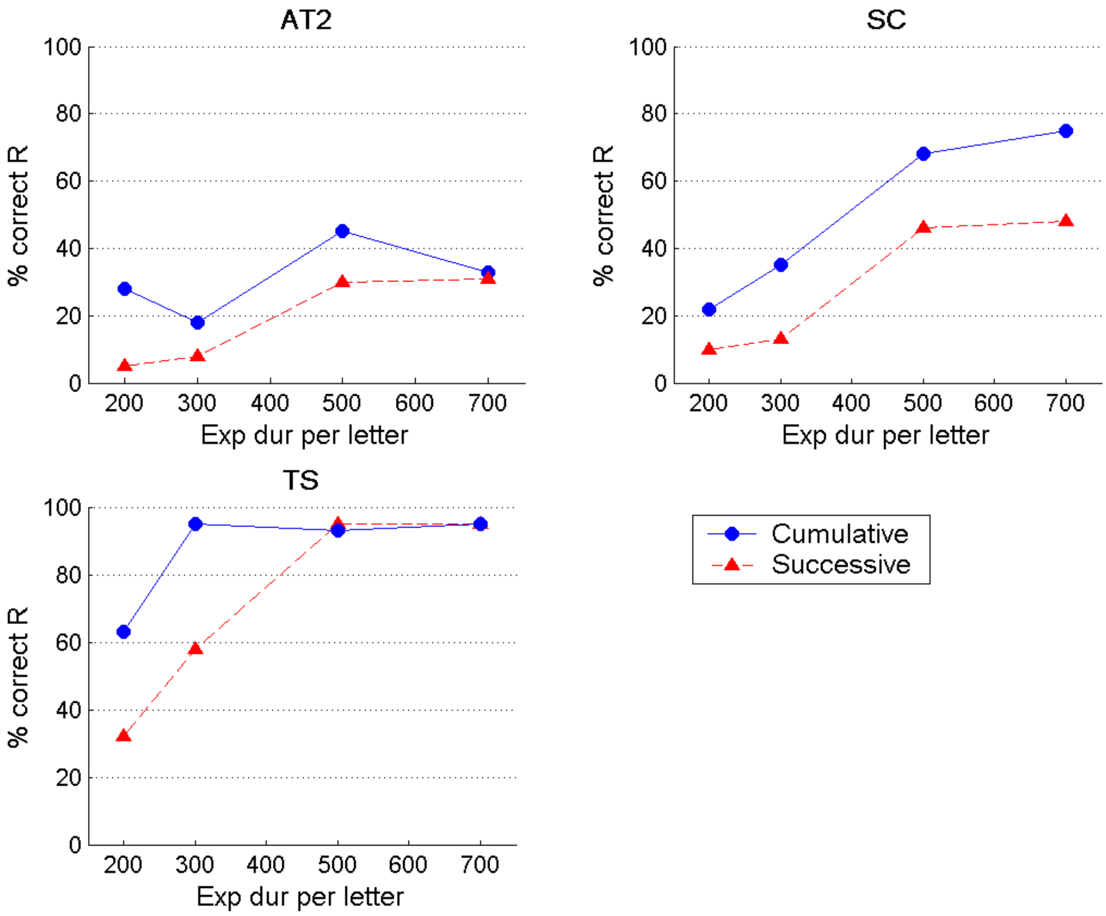


Figure 5.5: Cumulative and successive task. Performance of AT2, SC and TS on the cumulative and successive presentation of words.

with succ and at 500ms with cum) >”king” twice.

These errors show the tendency to translate from orthography to sound on a letter-sound-to-letter-sound basis without allowing successive letters to over-rule earlier parses. These mistakes are reminiscent of those made by the two pure alexic patients TP and KC (Patterson and Kay, 1982) and by surface dyslexic patients (Marshall and Newcombe, 1973).

AT2 made similar errors (e.g. island (at 300ms with cum) >”islam?”; weight (at 700ms with succ) >”white?”) and others which suggested a lack of ability to integrate the letters correctly identified (e.g. almost (at 700ms with cum) >”A..L..M..O..S..T.. I don’t know”; around (at 700 with succ) >”A..R..O..U..N..D.. I don’t know”; single (at 200ms with cum) >”signal”).

TS made some mistakes more visual in nature: e.g. minute (at 500ms with succ >”music”; caught (at 200ms with cum) >”cough”; almost (at 200ms with cum) >”almond” and bridge (at 200ms with cum) >”bride”; stream (at 300ms with cum) >”strength”.

Summary of the orthographic integration ability

The cumulative and successive task was administered to investigate whether patients could integrate letters. As with the Italian patients, we looked at the difference between the 2 conditions, to see whether they could benefit in the cumulative condition when letters were one close to the other one with respect to the successive presentation.

As regards AT2, from the observation of his errors and conduit-d’approaches made on the word reading task, it appeared that he had a weak letter integration ability. This has been confirmed by the small difference between the cumulative and successive task and by the type of errors made. As regards SC, on the cumulative and successive task the difference between the 2 conditions was mild, although significant at 300, 500 and 700ms. His errors reveal the difficulty to read an inconsistent language such as English.

Finally, TS showed a better facilitation in the cumulative presentation compared to the successive one at short durations, although his ability to integrate letters was not intact. Some of his errors were more visual in nature and this might suggest that he tended to read through the lexical route. This different type of strategy used to read may explain why TS is better than SC on this task as well as on word reading. Another explanation is that TS is the only patient whose spelling abilities are intact. However, we will see in the next section if spelling abilities are crucial for reading.

As a result, data from this section suggest that: 1. all the patients have a further deficit in integrating letters; 2. this ability is not equally damaged in all the 3 patients, but it is very weak in AT2, much more intact in TS and for SC it is in-between; 3. from the type of errors made it appears clear that English makes reading more difficult and that different types of strategy can be used, as suggested by the results of the next paragraph.

5.4.5 The use of LBL reading

According to Warrington and Shallice (1980), LBL reading is mediated by a strategy of letter naming via spelling system; therefore a simultaneous articulatory suppression task, which by definition prevents other verbal tasks, should disrupt reading. We replicated a task used by Warrington and Langdon (2002) to assess whether patients are able to read words during a simultaneous articulatory task. This is a critical experiment to investigate whether LBL reading is dependent, at least in our patients, on the viability of the letter naming strategy. An alternative hypothesis, given by Hanley and Kay (1992), is that not a phonological but a visual strategy can be used by the patients to read. In fact Hanley and Kay (1992) noticed that most of the errors made by their patient PD in a word reading task was visual in nature, while those made in a spelling task were more phonological. It is argued that it is unlikely that the reading

	No suppression	With suppression
AT2 at 2500ms	12/32 (38%)	7/28 (25%)
SC at 2500ms	27/32 (84%)	15/29 (52%)
TS at 2500ms	30/32 (94%)	22/31 (71%)
AT2 at 3500ms	18/30 (60%)	17/33 (51%)

Table 5.4: Articulatory suppression task with the English patients. Proportion (and percentage) of words correctly read in the condition where the simultaneous articulatory suppression was required and when it was not required.

strategy involves the spelling system to read. Moreover also our Italian patients were able to read also when the spelling system could not be used (see Chapter 4, paragraph 4.14).

Materials and Procedure

Thirty-two 5-letter words were selected and presented to the English patients with the same procedures used for the Italian patients (see Chapter 4, paragraph 4.14). The only difference is the exposure duration that here is 2500 (the same duration used for patient ROC by Warrington and Langdon (2002)). Since AT2 was the slowest LBL reader, the same version of the task was presented but with a longer duration (3500ms per word)[1].

Results

Some items were removed during the concomitant articulatory task because the patients stop counting too early (specifically 4 items with AT2 at 2500, 3 items with SC and 1 with TS); one item was removed from the condition with no suppression with AT2 at 3500.

All the 3 patients were able to read words in both the conditions, with a better performance when reading was without articulatory suppression (Table 5.4). The performance was significantly better for SC (Chi2= 7.56, $p<.006$) and TS (Chi2=5.67, $p<.019$), but not for AT2 with either the exposure durations

[1]This version of the task comprised 1 more word to read with the suppression task (tot=31) and 1 less word to read without the concomitant task (tot=33)

(p=.224 at 2500ms and p=.336 at 3500ms). It is noteworthy that AT2 and SC had spelling problems (see their case description), therefore they might have adopted different strategies to read. However, TS's spelling abilities were intact.

The main finding is that all of them did read a number of words presented with the concomitant verbal task. If we leave out the performance of AT2 at 2500ms, we see that the patients were able to read more than the 50% of words with the articulatory suppression.

The nature of their errors is mainly visual. AT2: horse >house; visit >vast; sharp >shape; clean >clan; TS: brief >bright; chair >Charlie; SC: dream >bride; route >round. SC made also some mistakes which could be both visual and spelling e.g. movie >move; smell >small; ideal >idea.

The performance of our patients on this task is very different from ROC's, the patient studied by Warrington and Langdon (2002). ROC read 15/16 (94%) of words correctly without suppression, but only 3/16 (19%) with the concomitant verbal task. The authors concluded from this performance that LBL reading is a strategy that involves explicit letter naming. The fact that our patients could report more than 50% of words even when explicit naming was prevented suggests that at least another strategy was available.

From the nature of the errors that the English patients made, which was mainly visual, we can hypothesize that the LBL reading strategy can be carried out with a mixture of verbal and visual strategies. The effective use of one or of both of them can depend on an individual's remaining intact abilities. Moreover the fact that patients make some visual errors might suggest that the visual word-form system can accept input from a serial sequence of letter names (Patterson and Kay, 1982; Hanley and Kay, 1992), without hypothesizing an alternative route.

5.5 Discussion

A new group of LBL readers has been investigated to test our hypotheses of the different functional causes in pure alexia in English. Patients were tested with the same procedures used for the Italian patients and with the same criteria to valuate their performance. In particular, our experimental investigation mainly focused on 3 specific points: the ability to process single letters, to put them together and the LBL reading strategy. Moreover the effects that different orthographies might have on the reading performance of our patients have been considered.

The 3 cases of English LBL readers showed some difficulties with language (spelling for AT2 and spelling and speaking for SC), had relatively intact visuo-perceptual abilities, normal memory skills but they were unable to read normally. Generally they were rather slow at word naming (especially compared to the Italian patients), showing a strong length effect and they had a restricted visual span.

The results concerning the 3 points showed that all the patients were slow at letter processing, especially AT2. The deficit started at letter recognition for all of them. The second point concerned the patients' ability to conjoin letters together. Results showed that all the patients were generally weak in integrating letters, although this ability was differently distributed across the patients. AT2 had a weak ability; TS showed a better capacity to put letters together into words and SC showed an integrative ability between AT2 and SC. The type of conduites d'approche made especially by AT2 and SC provide support to this hypothesis (e.g. AT2 read ink >”’i’ ’n’ ’k’.. /aink/.. ink!” (45sec); paint >”’p’ ’a’ ’i’ ’n’ ’t’.. paint?”; SC read student >”/stədnt/.. ’s’ ’t’ ’u’ ’d’.. /stiu/... student!”; value >/v/.. /vel/.. value!”). Finally as a third point, the LBL reading strategy seems not to be reliant only on the spelling system and on the use of letter names, but also on the visual aspects of letters and letter clusters, as suggested by Hanley and Kay (1992). It is possible that the LBL reading strategy can be carried out by a mixture of phonological and visual strategies and that the effective use of one or both of them can depend on an individual's remaining intact abilities.

Moreover it has been considered whether the differences in the reading performance and in letter integration between the English and Italian patients can be due to differences in the two orthographies. As mentioned in the introduction and in Chapter 2 (Paragraph 3), English is an inconsistent language and a letter or a cluster of letters can have multiple phonological realizations. Some errors made by our English patients show the difficulty to assign the right pronunciation to words or parts of words (for instance strike >"stri/i/k"; earth >"ears"), as also noticed by Patterson and Kay (1982). Since Italian is a transparent language it does not have this type of difficulty and some types of errors can never occur (as the final *e* rule in "fine") because not-adjacent dependencies are not present in the language.

In addition, the serial strategy used in pure alexia prevents the whole-word reading and this might be highly problematic for reading English where the pronunciation of letters is ambiguous, context-dependent and can vary according to the last letter (e.g. the final *e* rule). Therefore the serial reading strategy is likely to lead to a less efficient reading or to the pronunciation of a wrong word. For instance the conduite d'approche made by AT2: bedroom >"/bi'drom/.. /bi'droom/.. bedroom!" suggests that the patient selects a smaller unit ('be') and lexicalises it. Therefore letter integration which should proceed by assembling letters and, when necessary, by over-ruling the previous assignment of pronunciations to letters (as for instance in the final *e* rule) does not occur automatically. In other cases the serial strategy leads to the pronunciation of a wrong word, e.g. island >"i..s..l..a.. islam!" or knight >"king", listen >"list... listen?", being based on a strategy of letter-sound-by-letter-sound. By contrast, in Italian, readers tend to rely on the sublexical route (Paulesu et al., 2000) because it can take advantage of the consistent mapping between orthography and phonology.

Overall, for these reasons we claim that letter integration is more difficult in English than in Italian. If the two English pure alexic patients SC and TS are compared to the Italian LDS, we see that on the letter naming task the two English pure alexic patients SC and TS are not slower than the Italian LDS (792ms, 722ms and 914ms, respectively). However on the word reading task, using a similar set of words varying for frequency and imageability, LDS took half of the time than TS and SC with 6-letter words (without including the conduites d'approche). These results would support the hypothesis that the difference at least between LDS and TS (who have a similar degree of severity) is due to a difference in the ability to integrate letters, caused by the different types of orthographies.

An alternative hypothesis is that the difference in performance between our English and Italian patients is affected by a difference in severity in terms of the systems impaired. The English patients might have suffered from a more severe deficit in integrating letters, beyond the difference between the orthographies; moreover the English patients are less pure than the Italian ones, with two of them (AT2 and SC) showing spelling difficulties and generally the lesions of the 3 English patients are larger than the lesions of both the Italian patients.

A prediction which can be made is that if we have Italian and English pure alexic patients with the same degree of severity and a set of words which are very similar in terms of orthography and meaning across the two orthographies (e.g. naso-nose; zebra-zebra; treno-train; rosa-rose; distanza-distance; natura-nature; pigiama-pajamas), we could expect that English are slower and maybe more error prone than Italian patients.

Overall, we argue that in pure alexia finding qualitative distinctions beyond the letter level is more difficult in English than in Italian.

Chapter 6

Right hemianopic alexia: confabulation at the end of the words. A case study

6.1 Introduction

In this chapter an interesting case of acquired dyslexia has been investigated. AT has a right upper quadrant hemianopsia following an ischaemic stroke in the left temporo-occipital area which also affected the VWFA. However AT is not a pure alexic patient: he is slow at reading because of the visual field defect, but he can read words presented in his intact visual field accurately, thus showing right hemianopic alexia.

AT is an unusual, interesting case because when words are presented briefly, he tends to perceive them as longer than their objective length. Errors tend to involve only last letters, with omissions and additions of extra-letters. Some examples of his errors are: ospite (guest) >ospedale (hospital), giacca (coat) >giardino (garden), pugno (fist) >pugnale (dagger). This pathological ”elongation” is not evident when he has time to read, or in bisection tasks, or in writing and drawing.

This disorder has not been properly defined before. In fact AT has hemianopic alexia with some symptoms of neglect dyslexia and letter position dyslexia. However, the features of his deficit do not fit either of these definitions exhaustively, as will be shown soon. Moreover, this unusual ”elongation” disorder may be better described in terms of confabulation than completion, although again, it is difficult to define it since it is so unusual. A description of each of these syndromes will be given.

6.2 Hemianopic alexia

As described in Chapter 3, hemianopic alexia is a reading disorder caused by a parafoveal visual field loss (Wilbrand, 1907). It is characterized by slowed reading. Hemianopic alexia has attracted much less attention than other acquired disorders and it has been described less frequently. The crucial point in this reading disorder is that word recognition is intact, but the visual field defect results in a disruption of the oculomotor reading pattern during text reading (Zihl, 1985). Therefore if words are flashed in the intact visual field, words are recognized accurately and rapidly.

6.3 Neglect and neglect dyslexia

A reading disorder that is often observed in patients with visual neglect is neglect dyslexia. As discussed in Chapter 2, in this disorder, reading is compromised by frequent errors affecting the contralesional portion of either single words. However, neglect dyslexia can occur in the absence of neglect symptoms (Patterson and Wilson, 1990) and neglect dyslexia for one side of space has even been reported in association with neglect for nonverbal materials for the opposite side of space (Costello and Warrington, 1987).

As already described in Chapter 2, one of the hallmarks of neglect dyslexia is the maintenance of target word length in the response, that is, the range of letters omitted and added are -1, +1. However, Ladavas, Shallice and Zanella (1997) described 4 Italian patients with severe neglect dyslexia for words and nonwords which did not maintain the stimulus word length. Typically their errors, also called ”Missing-Left variety”, were: elicottero (helicopter) >tero; pomodoro (tomato) >doro; caguro (kangaroo) >uro. This type of error where the incorrect response is much shorter than the target can be associated to the language used. In fact, in Italian, readers rely more on the sublexical route (see Chapter 2) and this allows them to subdivide the letter strings into subparts. By contrast, as discussed in Chapter 2, in English words are likely to be read more as a whole through the lexical route and this prevents subdivisions in letter clusters.

Right neglect dyslexia is an uncommon syndrome as compared to left neglect dyslexia; in right neglect dyslexia errors occur at the end of the words.

Since the first brief clinical description of this syndrome reported in 1957 by Warrington and Zangwill, no further example of this type of reading deficit has been documented until 1990, when Caramazza and Hillis reported the case of NG. NG was a 77-year-old woman who after a stroke in the left parietal area showed right-sided neglect with objects and words. The patient made reading and spelling errors only on the right half of words, regardless of length. Importantly, she made the same pattern of errors regardless the arrangement of stimuli in reading (horizontal, vertical or mirror-reversed words).

On the basis of these results, Caramazza and Hillis (1990) argued that the deficit was in the processing of the right half of an abstract, orientation-invariant representation and that order information is coded spatially in a word-centred coordinate system.

Shortly after, Warrington (1991) described the patient RYT who showed right neglect dyslexia without spatial neglect after a left parietal infarct. His errors affected the right side of the word, with a gradient of accuracy that did not change across word length: the beginning of any word was read with 100% accuracy, the middle portion with 70% accuracy and the last part with 15% accuracy. RYT maintained the target word length in the neglect error response. Moreover qualitatively similar responses have been reported with tachistoscopic presentation and in free vision.

According to Warrington (1991), the spatial basis of the errors suggests that wrong target selection follows from an abnormal distribution of attention in the access procedures. There is evidence that the attentional mechanisms operating on the stored representations of written words are characterized by a left-right gradient.

6.4 Letter position dyslexia

The term ”positional dyslexia” was first introduced by Katz and Sevush (1989). The authors described two patients, JM and LS with left-hemisphere lesions (JM also with right upper quandrantopsia) whose positional reading difficulty could not be attributed to either neglect or extinction. Their reading errors occurred at the beginning of words, both in horizontal as well as vertical presentation. Examples of their errors are: cast >fast; port >sort; light >night; acquire >require. Katz and Sevush (1989) hypothesized that positional dyslexia was caused by selective damage to the activation of specific letter position nodes. They suggested that since initial letters may automatically draw attention, the patients' reading errors at the beginning of words occurred because attention was no longer automatically drawn to initial letters.

Patterson and Wilson (1990) reported an in-depth case of the patient TB who showed a reading pattern very similar to that described by Katz and Sevush, (1989). TB had a posterior left hemisphere lesion with a right homonymous hemianopia and his reading errors mainly involved the initial letter (safe

>cafe). His reading impairment could not be attributable to visual problems since the left-hemisphere lesion and his visual field defect would predict, if anything, visual problems at the end of words.

The patient showed some letter recognition difficulties; he was particularly likely to misidentify the initial character in random strings of letters (rose >nose). By striking contrast, he identified letters in positions from 2 to N of words and of strings with good accuracy. Although the majority of his word-reading errors maintained target length (67%) a certain number of his reading errors (19%) involved additions of letters (e.g. tout >”stout” or ”trout”; fall >shall). According to the authors it is possible that precise information about word length was no longer maintained.

TB showed one feature of attentional dyslexia (better performance on letters right flanked by numbers than by other letters), but not other features (errors reflecting intrusions of letters from another word) and he was able to read the letters in the other positions correctly except for position one. The deficit also affected numbers, which tended not to be reported when presented in the first position.

TB’s identification of the first letter in a string was not improved when the word (for instance, land) was preceded by a letter or a number (2land, xland or bland): importantly, TB was asked to ignore the first item when reading the word. These results indicate that it was the first character in the array which TB was identifying that is more vulnerable. Patterson and Wilson (1990) hypothesized that the patient may have some general deficit in the control of attention.

In 2001 Friedmann and Gvion described two Hebrew-speaking acquired dyslexic patients with occipito-parietal lesions. These patients suffered from a letter position deficit: they were able to identify the letters in a string, but failed to attach them to their positions. Hebrew is a language that is written from right to left, where vowels are usually not represented in the orthography and many words comprise only consonant letters. Moreover most of the letter sequences can be read in more than one way; as a result, when a letter erroneously change position in a sequence, there are many possible ways to read the new sequence. These properties of the Hebrew orthography make a selective deficit of letter migration within words easier to detect in Hebrew.

The patients BS and PY studied by Friedmann and Gvion (2001) did not have language problems; their reading pattern did not show effects of regularity, or imageability and neither semantic nor regularization errors were made. There was an effect of syntactic class (although function words were read better than verbs). The big effect was evident on a reading task with migratable and non-migratable words: both the patients showed a percentage of correct words significantly lower when words had an anagram, with respect to when letters could not migrate to form another word. Most of their errors were within-word migrations of middle letters (board >broad), not only in reading tasks, but also in lexical decision, same-different decision and letter location. The deficit seemed to be specific for orthographic material.

The patients showed a deficit in letter position encoding: letter identification was intact but location information within words was lost. Similarly to the "classical attentional dyslexia", it has been hypothesized that these patients suffered from an attentional deficit that prevented them from locating letters within words. According to Friedmann and Gvion (2001) the impairment was likely to be at the visual analysis of orthographic input (Ellis and Young, 1988), possibly in its letter location function.

As a conclusion, from these few reported cases of positional dyslexia, it is possible to argue that this disorder occurs after damage in the posterior regions of the left hemisphere and it affects reading by preventing the patients from assigning letters to their correct positions within a word. In the patients described by Katz and Sevush (1989) and by Patterson and Wilson (1990) the deficit was specific to the initial letters of the words. In the 2 patients reported by Friedmann and Gvion (2001) errors occurred in the central positions of the words. A common explanation for all these cases is an attentional deficit which prevents the correct matching between letter identity and position within words.

6.5 Completion phenomena

Poppelreuter (1917) was the first to describe the completion phenomenon. When a figure was exposed on the tachistoscopic screen so that it overlapped the blind part of the visual field, some patients with RHH still reported seeing the whole figure. "The paradox is that the patient apparently sees with his blind field". He interpreted it as a means of compensating for their hemianopia. However, not all the patients showed this phenomenon.

Fuchs (1920) confirmed the observation of Poppelreuter and further claimed that the effect could be elicited only with simple, regular and symmetrical figures. Meaningful drawings (like a dog or a face) were never completed even though symmetrical. Letters, words, and lines were likewise never completed. Completion (of whole figures) may be indicative of some residual vision in the blind parts of the field.

Warrington (1962) studied 20 patients with complete (right and left) HH by presenting geometrical forms tachistoscopically. The results indicated that 11 patients showed 60% or more completion, and 9 showed 30% or less completion. The complete responses decreased with the increase of exposure time. The content of the completed perception was determined by visual stimuli that fall within the intact visual field. This tendency to complete forms across the damaged field was associated with the presence of parietal lobe lesion of either hemisphere. None of the 5 patients with occipital lesions showed completions.

Warrington (1965) studied the completion phenomenon in 26 patients with a left visual field defect in relation to the nature of the stimuli used. While the highest incidence of complete response occurred with simple geometrical forms, a number of complete responses occurred with more complex objects which were familiar (like an elephant, a dog and a cat). Warrington (1965) stressed the importance of expecting a whole figure from a familiar stimulus.

As a conclusion, it emerges that hemianopic patients with occipital damage rarely exhibit pathological completion of partial shapes while having veridical perception of whole figures (Warrington, 1962; Hornak, 1995; Marcel, 1997). Rather, completion appears to be associated with patients with parietal damage with or without hemianopia, in whom disorders such as unilateral neglect and extinction may be important contributing factors (Warrington, 1962; Sergent, 1988). Finally, Walker and Mattingley (1997) question the pathological completion phenomena on methodological and theoretical grounds. They argue that since they are mostly associated to parietal lobe damage and concomitant attentional disorders, these phenomena may reflect unawareness of visual loss, rather than active filling-in.

6.6 Cross-over and confabulation in left-sided neglect

Unilateral neglect is a disorder in which patients do not report, respond to or orient to stimuli presented contralaterally to the lesioned hemisphere. When patients with left-sided neglect are asked to mark the midpoint of horizontally oriented lines, they tend to mis-bisect horizontal lines to the right. However with short lines many patients bisected to the left of the mid-point, thus demonstrating right-sided neglect (Halligan and Marshall, 1988; Marshall and Halligan, 1989; Tegner and Levander, 1991). Cross-over can hardly be explained by defective representation or attention for the contralesional space or by hyperattention for the ipsilateral space, because these accounts predict ipsilesional rather than contralesional deviation in line bisection.

In 1995 Chatterjee reported the study of 5 patients with left-sided neglect who exhibited this paradoxical behaviour. To understand this cross-over phenomenon, Chatterjee (1995) tested the patients on a single-word reading task, with the idea that the way in which patients read short words could provide insight into how they bisected short lines. He found that when patients were asked to read short words, additional letters were reported on the left side of the word: patients making larger cross-over errors on line bisections were likely to confabulate and to report extra letters on the left of the word. However, it was mainly 2 patients who produced overestimation with mainly 2-letter words and the number of letters added was very small (on average less than 1 letter).

Chatterjee (1995) hypothesized that these patients with left-sided neglect perceive short lines as longer and when trying to read a short word they extended it. However this confabulation or elongation occurred with constraints: the spatial extent of confabulation is influenced by the length of the target word and does not extend leftward indefinitely.

Chatterjee (1995) explained the productive nature of these errors in terms of a failure of normal inhibitory processes. According to his hypothesis, a region of disinhibition at the left edge of a rightwardly restricted attentional window may lead to a delusional elongation of objects on the left side.

This phenomenon could be called completion or confabulation. As discussed above, completion usually refers to the completion of a simple geometrical visual pattern over a blind region. It is associated with parietal damage and spatial neglect (Warrington, 1962). Chatterjee (1995) argued that the addition of letters to the left of short words is not completion in this precise sense, but rather patients seem to be confabulating letters.

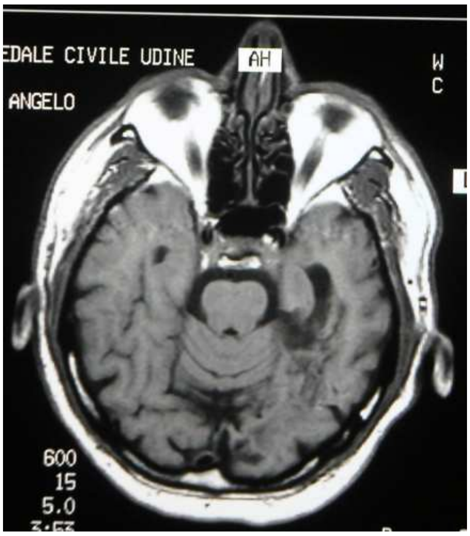


Figure 6.1: MRI scan of AT. This axial section shows a lesion in the left occipito-temporal lobe which involves the occipito-temporal fusiform gyrus and the occipital gyrus.

6.7 AT: case description

AT is a 80-year old, left-handed professional Italian man with a high-school education who had an ischaemic stroke in July 2000 following an operation on his left carotid. A SPECT scan (27-02-02) showed a lesion in the left occipital region and minor lesions in the left parietal and temporal lobes as well as in the left basal ganglia. A MRI scan (February 2002) showed a left infero-posterior temporal lesion that corresponds to the temporo-occipital gyrus and to the occipital gyrus between the collateral sulcus medially and the occipito-temporal sulcus laterally (Fig. 6.1). The inferior temporal gyrus is intact laterally. An atrophic enlargement of the temporal horn of the lateral ventricle is evident. The last CT scan (4-3-02) showed a low-density in the left temporo-occipital area with stable characteristics of a past ischaemic stroke.

He has a right upper quadrant hemianopsia with macular sparing (Fig. 6.2). AT is an intelligent man of vast knowledge; he is not a LBL reader, although

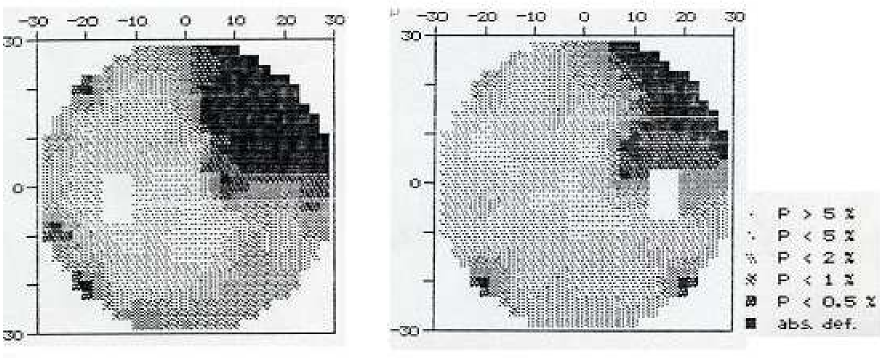


Figure 6.2: Octopus static perimetry: visual perimeter of AT displaying right upper quadrant hemianopsia. On the left there is AT's left eye, on the right his right eye.

he has difficulties in reading.

6.8 Neuropsychological assessment

The investigation of AT's reading started in June 2002. At the time of testing his language skills were spared with no difficulties in comprehending, speaking, writing, but only in reading. A general assessment showed normal processing (categorical fluency, verbal fluency and Raven Progressive Matrices) and his premorbid intelligence functions assessed with the T.I.B, an Italian short intelligence test, revealed a good intellectual level, well within the normal range (total IQ=118, Verbal IQ=119 and Performance IQ=121). His short-term memory was preserved (digit span forward and backwards, Corsi's test) and he performed well on the story recall.

His perceptual abilities were examined with the VOSP: he had difficulties with Incomplete Letters (15/20) and Silhouette subsets (8/30) and when presented with the BORB he showed an impaired performance only on the Object decision tasks (A hard version 18/32; A easy version 23/32; B easy version

24/32). He showed spared performance on the Benton face test. There was no evidence of spatial neglect: his performance was normal on the Bells test (Gauthier, Dehaut and Joanette, 1989), on the letter cancellation test (Diller and Weinberg, 1977) and on a bisection task where 50 lines 1.82, 3.62, 7.23, 12.65 and 18,08 cm long were presented on separate sheets in pseudo-random order.

6.8.1 Naming abilities

AT showed some difficulties in a naming task (Capitani et al., 1993), where he scored 56/80. He was presented with another naming test assessing 3 different modalities: he scored 24/30 with the visual modality, 25/30 with naming to definition and 18/30 in the tactile modality.

His naming abilities have been further investigated with the Lotto et al. set (2001): AT's performance was very impaired (171/250, 68%) especially with living items (42/83, 51%) with respect to nonliving (129/167, 77%). A logistic regression was performed with living versus nonliving together with name frequency, concept familiarity, typicality, age of acquisition and visual complexity: the outcome indicated a significant living effect (W=20,3; $p < 0.00001$), and of age of acquisition (W=11,6; $p < 0.0007$). He failed to recognize almost all vegetables (4/14 he said: "I don't know vegetables and gardening stuff!"), many fruits (11/20; e.g. Lemon >type of pear; orange >"I don't know, a vegetable?"; strawberry >Indian fig; acorn >mushroom) and many animals (mammals: 15/21; birds: 9/17, e.g. Sea-gull >eagle; owl >swan; hippopotamus >rhino; zebra >"I don't like these animals, I don't know"; rhino >pig).

6.9 Experimental investigation

The aim of this experimental part was to investigate AT's unusual reading disorder. AT tends to elongate the words he sees: these are characteristics that might be associated with neglect, but AT does not have neglect.

In order to assess his performance, AT has been compared to the controls subjects matched to the Italian pure alexic patient FC for age and education. In fact AT is 80-year old man (FC is 86-year old man), the mean age of the controls was 84.4 years and all of them except one had a high education level (mean schooling: 11,7, sd: 2,4), as AT. In particular, AT has been compared to the 2 controls C1 and C2, matched for gender, age (81 and 87 years), and educational level (13 years of schooling). AT has also been compared to the Italian pure alexic patients FC and LDS.

6.9.1 Documentation of writing and reading skills

Writing

As for FC and LDS, AT was asked to write 2 long passages with a total of 62 words. His performance was flawless. When asked to write the 50 words, he performed perfectly.

Prose reading

As for the Italian and English LBL readers, AT was presented with a text to read taken from a battery (Nuove prove di lettura MT per la scuola media inferiore) by Cornoldi and Colpo (1995) used to assess reading abilities in young students with possible developmental dyslexia. The text comprised 325 words. AT was asked to read at his own speed. Text reading was recorded with a microphone connected to a PC via a cool edit program for each patient and control subject. AT took 3 min and 55 sec to read the text and made 2 errors. Control subjects C1 and C2 matched to AT and FC, took 3 min and 2 min 38 sec and made 6 and 0 errors, respectively.

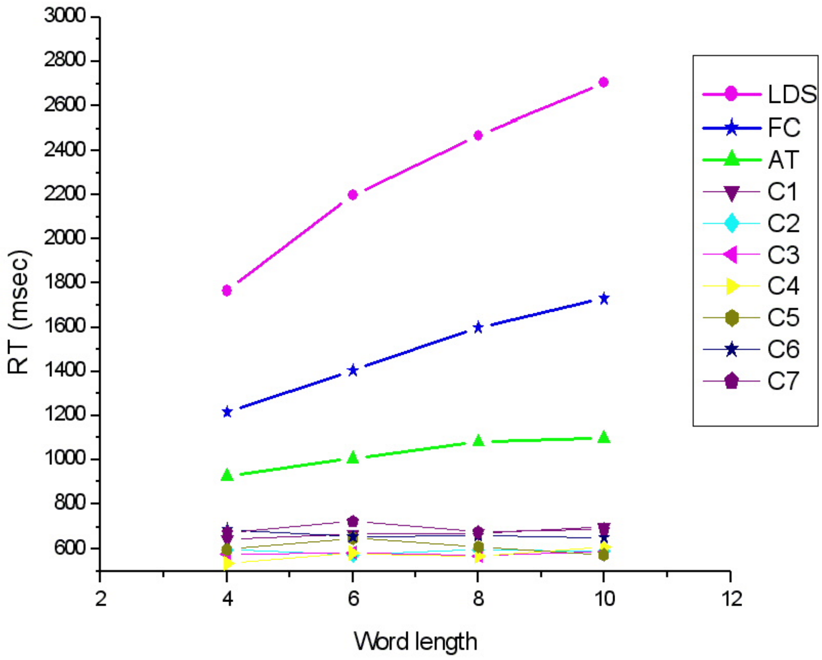


Figure 6.3: Word reading with AT. RTs (ms) of AT, FC, their matched control subjects and LDS as a function of word length.

Word reading

The same list of words given to FC and LDS was given to AT. The list comprised 120 words, 30 each of 4, 6, 8 and 10 letters in length, presented in random order on a PC monitor using an E-prime program. Words were divided into abstract and concrete and each of the 2 subgroups was matched for frequency. The same experimental procedures used with FC and LDS have been utilized for AT.

Results

Differently from FC and LDS, AT did not show a length effect (p = .27), probably for the large standard deviations, or a lexical effect, or an effect of imageability (Fig. 6.3); the increment in reading speed per letter was 29.9msec.

AT made 2 mistakes (ghiacciaio (glacier) >ghiaccio.. ghiacciaio (ice... glacier); cacciatore (hunter) >cacciagio.. gio.. cacciatore). Control subjects showed little increment in reading speed (msec) per letter (-12 <B <23) with the effect of word length being completely insignificant.

Word reading in the LVF and RVF

In this task it has been assessed whether AT's slowness in word and text reading is due to his visual field defect. If this is the case, AT has hemianiopic alexia and he is expected to read the words shown in his intact visual field.

Methods

A set of 60 words was selected: 20 words of 3, 4, 5 letters in length matched for frequency. Words were presented singly in the RVF and LVF, very quickly, for 170ms to prevent ocular movements and in a random order so that patients could not fixate where the word appeared (Ellis, 2003; Whitney and Lavidor, 2004).

Patients stayed at a distance of 50cm from the PC screen and have to fixate a central cross. Words were placed at the left or at the right of the cross, in particular the final letter in the LVF (and the first one in the RVF) was at a distance of 1,1cm away from fixation, which is 1 degree of visual angle. Word width ranged from 1.7 to 5.1 of visual angle (Cohen et al., 2003). Words were presented once in the LVF and once in the RVF in different sessions, for a total of 120 stimuli. There was a short practice of 10 trials. The task was presented to AT, to 2 matched controls (C1 and C2), twice to FC and LDS.

Results

The results showed that AT was able to read the words presented in the LVF, exactly as his matched controls C1 and C2 (see Table 6.1). By contrast, when words were presented in the RVF his performance worsened drastically. FC and LDS showed a very similar performance, being able to report only a few words displayed in the RVF likely because of their homonymous hemianopia,

	LVF					RVF				
	Performance	%	3	4	5	Performance	%	3	4	5
AT	46/60	77	15	17	14	2/59	3			2
C1	46/60	77				56/60	93			
C2	47/60	78				55/60	92			
FC	37/120	31	14	31	10	21/120	18	9	7	5
LDS	37/118	31	14	14	9	14/120	12	4	4	6

Table 6.1: Word reading in the LVF and RVF. Proportion, percentage and number of words reported in each hemifield by AT, by his matched controls C1 and C2, and by the two pure alexic patients FC and LDS.

and a low percentage of words displayed in the LVF. This result confirms that they are pure alexic patients.

Letter naming

As for FC and LDS, AT was presented with the letter naming task, run with the same procedures used for the 2 LBL readers. AT performed the task twice.

As shown in Fig. 6.4, AT's performance is the same as FC's one, therefore in the normal range compared to his matched controls.

6.9.2 Visual span

As with FC and LDS, AT was presented with a visual span task. The procedures are the same as those used before.

Results are reported in Table 6.2. AT's performance was significantly worse than C1 and C2 with letters (Wilcoxon test, z = -3.01, p $<$.003; Wilcoxon test, z = 3.02, p $<$.002, respectively), but it was not significantly different from one of his matched controls with digits (Wilcoxon test, z = -1.6, p $>$.109). His performance was comparable to FC's with letters (Wilcoxon test, z = -.539, p $>$.59) as well as with digits (Wilcoxon test, z = -.775, p $>$.439) whereas it was significantly

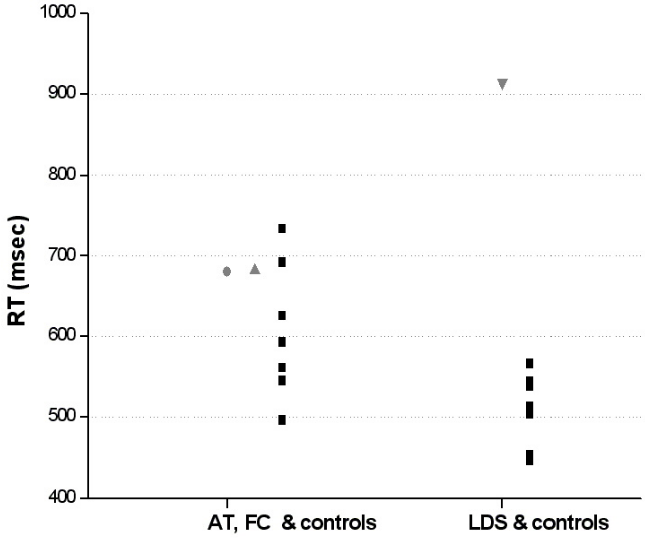


Figure 6.4: Letter naming task with AT. RTs for AT, FC and his controls and for LDS and her controls.

	Performance								
		Horizontally				Vertically			
		100	150	250	Avarege	100	150	250	Average
AT	Letter	75	71	79	75	87	96	92	92
	Digit	79	87	87	85	92	92	100	94
C1	Letter	100	100	100	100	87	96	87	90
	Digit	100	100	100	100	96	96	100	97
C2	Letter	100	100	96	99	87	96	87	90
	Digit	96	100	100	99	83	83	100	89
FC	Letter	83	83	92	86	75	75	79	76
	Digit	83	83	96	87	75	87	100	87
LDS	Letter	62	46	62	57	58	50	71	60
	Digit	71	62	71	68	58	71	67	65

Table 6.2: Visual span with AT. Percentage of letters and digits reported by AT, his matched controls, by FC and LDS at each exposure duration (100ms, 150ms and 250ms). The three letters and digits were presented at the left of fixation (horizontally) and just below the fixation (vertically).

better than LDS' with all the stimuli. AT showed an improvement when items were presented vertically (F=17.33, p $<.0001$) and his performance was better with digits than with letters (F=3.9, p=.052), as LDS and FC.

6.9.3 The ability to orient attention

As with the Italian pure alexic patients, AT was presented with an attentional task aimed to assess whether he is able to orient and to move his attention in the space adequately. This is a non-reading task where the typical visual search paradigm developed by Triesman and Gelade (1980) has been used.

Materials and procedures are the same as those used for the Italian pure alexic patients (see chapter 4). AT did the experiment 3 times, FC and LDS twice.

Results

As for FC and LDS, only the target present trials were analysed; AT made 12 mistakes (7/432).

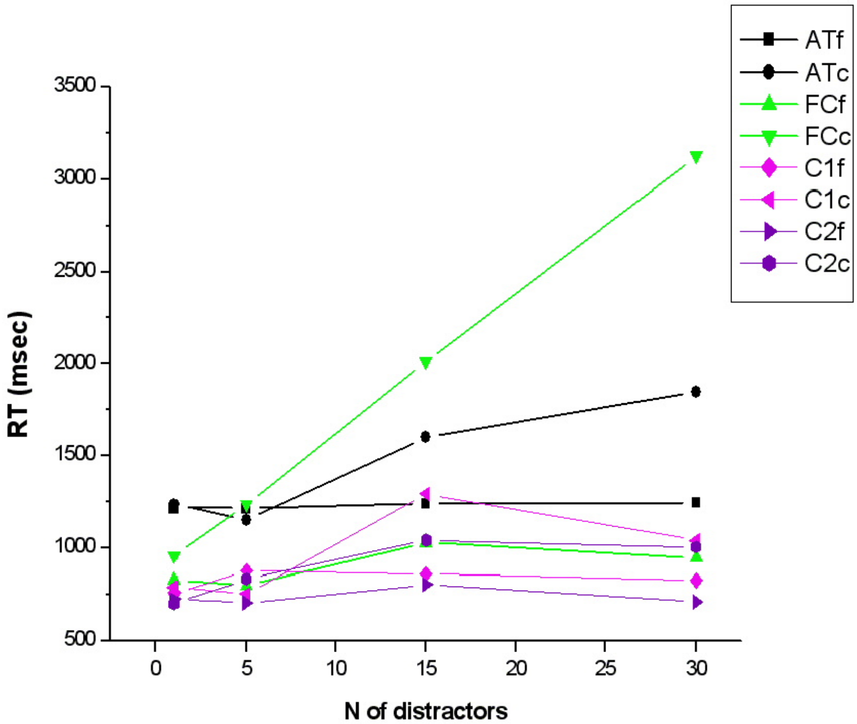


Figure 6.5: Treisman attentional task with AT, FC and their matched control subjects C1 and C2 on visual search as a function of display size (N of distractors) for the feature (f) and the conjunction (c) tasks.

As shown in Fig. 6.5, the analysis of variance of AT's RTs (with feature/conjunction and number of distractors as factors) indicated a display size effect in the conjunction task ($F(1, 99) = 23.3$, $p < .0001$) but not in the feature task ($F(1, 101) = .078$, $p > .78$). This is in line with results of the experiment by Treisman and Gelade (1980).

These results are in agreement with those reported for FC and LDS: there is no effect of display size in the feature task even with 30 distractors. This suggests that AT does not have difficulties in distributing his attention in tasks where letter processing is not requested.

6.9.4 Study of AT's elongations

By studying AT's performance on different reading tasks, we noticed that he tended to perceive words longer than their objective length when they were displayed briefly (on average 350ms). In particular, AT has been presented with more than 1000 words, in different conditions such as in tasks of semantic categorization, of lexical decision, or in the cumulative and successive presentations. The results of all these tasks are not relevant for the study of his hemianopic alexia; however the elongations that AT made were very interesting and have been studied carefully.

AT was presented with 1046 words: 527 (50%) have been read, 275 (26%) resulted in the typical ”I don't know” answers and the last 244 (23%) resulted in incorrect answers. We studied the last category of responses. Words had different lengths (from 4 to 8 letters) and most of the times were presented at the left of the fixation point, although the relatively long duration allowed shifting of the eyes.

AT's errors had neglect aspects: they shared the same initial letters but diverged towards the end. Examples are: parola (word) >paragrafo (paragraph); cenere (ash) >centimetro (centimeter); estate (summer) >estrazione (extraction); stanza (room) >stangata (blow); anna >ammalato (ill). The frequency of AT's incorrect answers was not higher than the target's. Moreover with respect to neglect dyslexia there is no maintenance of the target word length in the answer.

To study AT's elongation, all the words have been divided in 4 parts (A, B, C, D) that are 4 spatial quartiles. For instance with a four-letter word each quartile had only 1 letter, while with an eight-letter word each quartile had 2 letters.

In the first analysis we studied the number of omitted letters per error. As shown in the Fig. 6.6, the longer the word, the more last letters are omitted (e.g. avvocato (lawyer) >avvolgere (to wrap); cammello (camel) >cammino (path)).

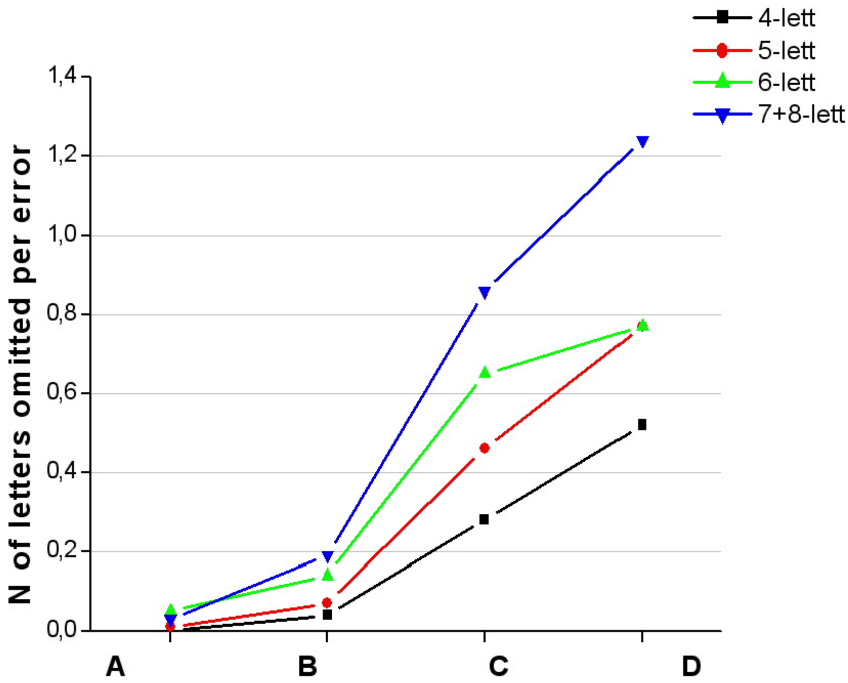


Figure 6.6: Study of AT's elongations. Number of letters omitted per error. A, B, C and D represent spatial quartiles in which words were divided; the figure shows that the longer the word, the more last letters are omitted.

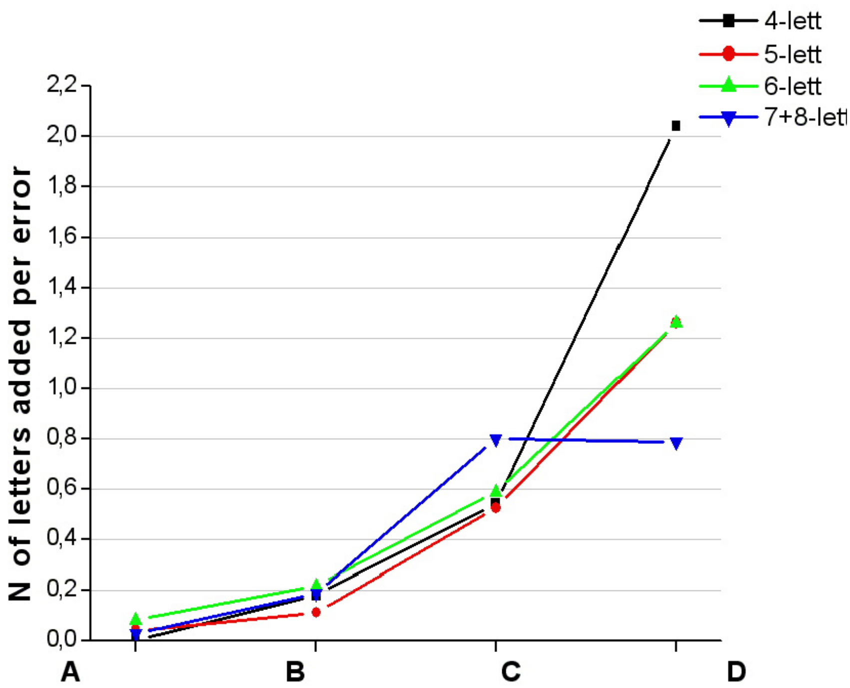


Figure 6.7: Study of AT's elongations. Number of letters added per error. A, B, C and D represent spatial quartiles in which words were divided; the figure shows that the shorter the word, the more letters are added.

The difference between 4- and 7+8- letter words in terms of number of letters omitted per error is significant in position C ($p < .0001$) and in position D ($p < .0001$) when analyzed with the t-test.

In the second analysis, we studied the number of letters added per error (Fig. 6.7). In this case the shorter the word, the more letters were added (dito (finger) >dittatura (dictatorship); vino (wine) >vincere (to win); bici (bike) >bistecca (steak); cocco (coconut) >cocomero (cucumber); tenda (curtains) >tendaggio (drapery)). The difference between 4- and 7+8- letter words in terms of number of letters added per error is significant in position D ($p < .0001$).

Word reading at different durations in the LVF

To investigate where these elongations arise from, AT was presented with a set of words presented in the LVF at different durations.

A total of 180 words 60 each of 4-, 5- and 6- letter in length were collected and matched for frequency; they were displayed at the left of the fixation point (a cross) for 3 durations (170, 350 and 600ms) in uppercase (Arial, font 25) in a randomized order. At each trial AT was asked to fixate the central cross before the word appeared.

Results indicated a good performance: AT scored 160/180 (89%). He gave 7 answers that were longer than the target length (e.g. naso (nose) >nasco (I am born); sugo (sauce) >sughero (cork)), 2 that were shorter (e.g. roccia (rocks) >rocca (fortress)) and 4 which were equal (e.g. pietra (stone) >pietro (Peter); moda (fashion) >modo (manner)). The elongations AT made were 1 at 170ms, 4 at 350ms and 2 at 600ms.

As a result, the low number of words ”hallucinated” suggests that AT can read words correctly when displayed in the LVF, producing a few elongations.

Word reading at different duration centrally

The same task was administered in central vision. The same set of words was used and displayed centrally so that the fixation point (a cross) coincided exactly with the central point of each word. Words were displayed for 170, 450 and 650ms, slightly longer than before because of his visual field defect. AT was encouraged to fixate the cross at each trial.

Results are shown in Fig. 6.8; a trial was removed because AT was distracted. AT's performance was less accurate than before (134/179, 75%) and with a higher number of elongations: (24/179, 13%). As shown in Fig. 6.8, the percentage of AT's elongations was significantly higher than his answers with a shorter length (Chi2 = 30, $p<.0001$).

Results show that AT starts to elongate when words are presented centrally

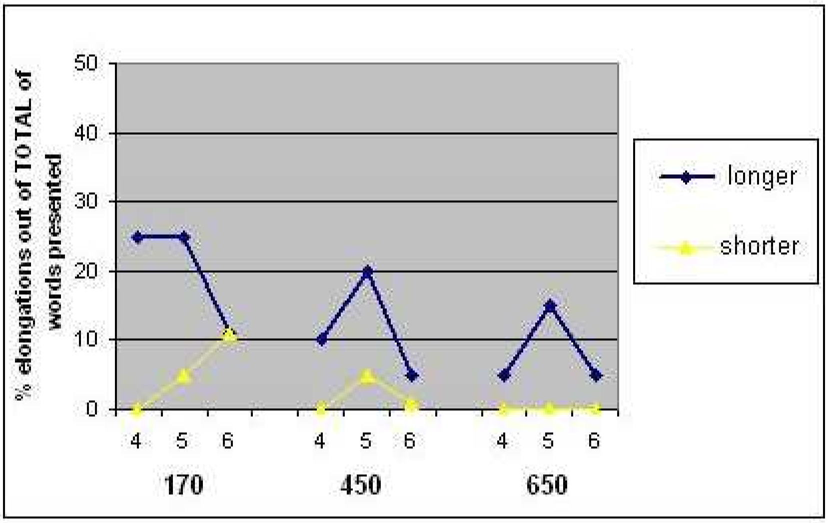


Figure 6.8: Reading of words presented centrally at different durations. The figure shows the percentage of words elongated and shortened out of the total of words presented.

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Quantification: how many items do you see?

In order to understand whether his elongations are specific to the reading process, AT was presented with a task which required him to judge the length of the items displayed.

A total of 840 consonant strings containing 210 items each of 3-, 4-, 5- and 6- letter strings were displayed for 350ms in 5 different positions. The letters used were c, n, h, p, d, g, s and q because they had the same length (as opposed to i or m). In position 1 strings finished at the left of the fixation point, in position 2 exactly on the fixation point, in position 3 a bit more on the right and so on (see Fig.6.9). First a fixation point appeared centrally and AT was asked to fixate it; then the letter string appeared for 350ms. He was asked to say how many items the string had.

							+			
pos						**1**	**2**	**3**	**4**	**5**
length							+			
3				c	s	d				
3					g	h	p			
3						n	g	q		
3							s	d	n	
3								c	h	d
							+			
4			s	p	g	h				
4				g	q	n	c			
4					d	h	n	s		
4						s	g	p	d	
4							n	d	g	p
							+			
5		g	p	s	h	d				
5			s	h	q	g	n			
5				n	d	h	s	q		
5					n	c	d	h	q	
5						q	s	d	g	d
							+			
6	p	s	g	h	d	n				
6		n	h	g	s	p	q			
6			q	n	g	h	d	s		
6				c	p	h	n	q	g	
6					d	s	p	d	q	n

Figure 6.9: Quantification task with consonant strings. The figure shows the different positions that the letter-strings had with respect the fixation point.

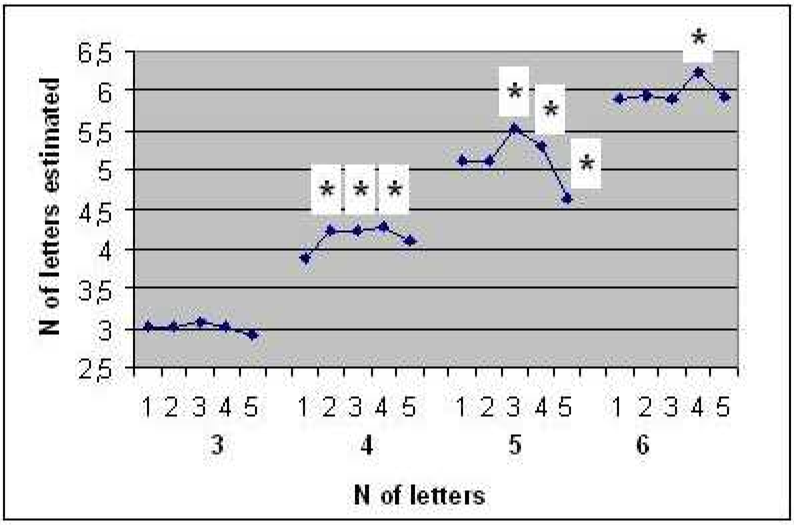

Figure 6.10: Task of length judgment. Number of letters estimated of consonant strings as a function of the consonant string length (3-, 4-, 5- and 6- letter long) and the position of the string with respect to the fixation point (1=the string finishes at the left of the fixation point; 2=it finishes on the fixation point; 3=at the right of it etc.).

Results

The results (Fig. 6.10) showed that AT was able to judge the length of the string correctly when it comprised 3 letters. However when it had 4 consonants AT started to elongate as soon the string was shifted to the right. Four-letter strings were estimated significantly longer in position 4, 5 and 6 (t-test = 3.16, $p < .005$; t-test = 3.55, $p < .005$; t-test = 2.74, $p < .009$, respectively). Again, 5-letter strings were perceived longer in position 3 and 4 (t-test = 7.2, $p < .0001$; t-test = 3.66, $p < .001$). By contrast, in position 5, the last one, AT perceived the 5-letter strings significantly shorter (t-test = 3.52, $p < .001$). With 6-letter strings, AT estimated the length as longer only in position 4 (t-test = 3.2, $p < .002$).

The results suggest that AT can estimate the length of 3-letter strings correctly. However he starts to elongate when letter strings comprise more than

3 letters and when they are displayed centrally. If most of the letter string is displayed on the damaged right visual field (i.e. about 3 letters), AT judges the length either correctly or shorter. It appears that the visual field defect has a crucial role in these elongations.

6.9.5 Discussion

In the present chapter a new case of acquired dyslexia has been investigated. AT is a patient with hemianopic alexia after damage in the occipito-temporal region of the left hemisphere (which interestingly included the VWFA). AT can speak perfectly, comprehend and write; however as a result of his visual field defect, he is slow at reading words presented centrally, but he can read words presented briefly in his intact visual field perfectly. AT is an unusual case because when words are presented briefly (about 350ms) he tends to perceive them as longer.

In the introduction it has been claimed that AT does not have right neglect dyslexia, nor position dyslexia. Although his errors tend to involve only the last letters, the pattern appears somewhat different from neglect since there is not maintenance of the target word length in his response errors (pollo (chicken) >polonia (Poland); zucca (pumpkin) >zucchero (sugar); elena (Helen) >elemosina (alms); strada (street) >stradina (little street)), nor does he shorten words making ”missing left errors” (Ladavas et al., 1997). Moreover when words appear without time constraints AT can read them correctly.

AT does not show typically positional dyslexia, because he can perceive the last letters within the words in their correct positions (e.g. tram >tramite (intermediary); braccia (arm) >bracciale (bracelet); fede (faith) >federa (pillowcase)). As shown in Fig. 6.6 the longer the word, the more last letters were omitted, but this did not occur with short words, as it is evident also in the examples here reported, so it was not a deficit in locating final letters within the word. Moreover, when AT has time, he is able to read accurately. Finally, the patient TB described by Patterson and Wilson (1990) and JM by Katz and Sevush (1989) showed left posterior lesion with right homonimous hemianopia, but the letters more vulnerable to reading were the initial ones. Therefore the visual field seems not to play any role in the reading deficit.

The present case appears similar to those described by Chatterjee in 1995: AT tends to elongate words, especially when these are short. However the cases reported by Chatterjee (1995) extended very short words (2-, 3- letter strings) and tended to add less than 1 letter on average. As in Chatterjee's (1995) cases, such a pathological behavior of word elongation is less associated to completion phenomena and fits better with an account of confabulation of letters. The explanation suggested is very interesting and it can explain our case as well.

Normally, every region of the cortex has a fine balance between synaptic excitation and inhibition. For instance in the visual cortex there are neurons with well-defined areas of excitation and inhibition, like simple cells in V1 which respond to stimuli with a certain orientation, in a specific position. Brain damage disrupts this balance, likely in favor of excitation, and this causes a disinhibition. Neurons are hyperexcitable and might show a spontaneous activity so high as to produce confabulations, perception of things that are physically absent. This is the basic account. Chatterjee (1995) suggests that these patients show a failure of normal inhibitory processes, with disinhibition at the left edge of the attentional window.

Similarly, Toraldo and Reverberi (2004) hypothesize that generally in neglect there is a misprojection of landmarks onto the internal spatial representation, due to a lack of inhibition on the right of this spatial representation.

As regards AT, we claim that his errors are caused by the brain damage which resulted in visual field defect. The upper quadrant hemianopsia may induce an input loss in the orthographic representation, which causes a lack of inhibition on the right, producing extra-letters at the end of words. As has been shown in the experiments the addition of letters interacts with the position in the visual field. This confabulation resolves fairly quickly, after a few hundred milliseconds. As shown, the addition of letters seems not to be specific to reading, since it also occurs in a task of length judgment. It is noteworthy that since we still used orthographic material (consonant strings), we cannot rule out the possibility that with non-orthographic material this does not happen.

This phenomenon is consistent with other accounts of neglect (Chatterjee, 1995; Toraldo and Reverberi, 2004). However, since AT does not present neglect, this phenomenon may dissociate from it and therefore it can be considered as a more simple, peripheral explanation.

Chapter 7

Conclusions

Reading is one of the most important human cultural inventions. It is central to visual and language processes and it rests on the ability to identify several letters rapidly and in parallel. Reading is a complex orchestrated activity which involves different components. This skill can be disrupted by brain damage in relative isolation from other deficits, a disorder called acquired dyslexia. The present work looks at the visual recognition of words in patients with peripheral dyslexia. By studying the effects that lesions have on the performance of patients we can make inferences on how this ability is organized and thus built models of the normal reading function. Patients with pure alexia and hemianopic alexia have been studied to investigate the early visual processing of words.

7.1 The qualitatively different forms of pure alexia

As described in Chapter 2, pure alexia is a reading disorder that occurs in previously competent readers following brain damage. It is associated with left occipito-temporal lesions and it characterized by a slow letter-by-letter reading procedure. Patients usually take an abnormally long time to read even single words, although the severity of the deficit can vary. Different accounts have been proposed so far in the attempt to explain the specific functional damage. As discussed in Chapter 3, some theories have claimed that pure alexia is caused by a single type of deficit (e.g. Behrmann, Plaut and Nelson, 1998) others have hypothesized that the deficit can vary, although the results are not so clear-cut (Price and Humphreys, 1992; Patterson and Kay, 1982; Hanley and Kay, 1996).

In our investigation with the pure alexic patients we addressed an issue that is central to understanding the disorder, whether there is one source of deficit or multiple sources of deficit from a neuropsychological point of view. In particular we were interested in investigating the early stage of word coding that comprises the processing of single letters as well as their integration in subparts like syllables and then words. As discussed in Chapter 2 this intermediate level has been ignored by most of the computational models of reading (e.g. Coltheart et al., 2001; McClelland and Rumelhart, 1981); only recently has it attracted more attention (Grainger and Whitney, 2004; Dehaene et al., 2005). In languages such as Italian or Spanish with clear syllabic boundaries an intermediate syllable level can be easily investigated (Carreiras, Vergara and Barber, in press).

The Italian patients have been presented with several tasks, testing the ability to process single letters and to integrating them together, implicit reading with semantic categorization tasks, a visual imagery of words and the use of the LBL reading strategy. The results have shown a clear double dissociation: the patient FC was able to process single letters as rapidly and accurately, as his matched controls, but he was unable to group together the letters he had correctly identified. By contrast, the patient LDS was slower and more impaired at letter processing (also at letter recognition), but she was able to integrate letters in syllables and words.

We have hypothesized that FC has a classical disconnection syndrome, as

Déjerine reported (1892): the word-form system visual input is intact but disconnected from the visual input. This hypothesis fits with the anatomy, since FC has a lesion in the left occipital lobe and in the paraventricular white matter and also fits with the results obtained from a visual imagery task of words, where FC could easily access to the visual word-form. The results suggest that the visual word-form system cannot be accessed from the visual modality.

By contrast LDS has a partial impairment of the visual word-form system, starting at the letter-form level. In fact she showed a deficit in letter processing but she was able to integrate letters together; this ability was significantly better than FC's but worse than her matched controls. The hypothesis of partial damage of the word-form system fits with the results from a visual imagery task of words where she was able to carry out the task, albeit more slowly and with more mistakes than FC and her controls. It also fits with LDS's lesion that was in the occipito-temporal lobe affecting the visual word form area (VWFA). It can be hypothesized that the VWFA is partially spared although closely surrounded by lesioned cerebral tissue, as has been shown in patient F described by Cohen et al. (2003). Interestingly, patient F had an occipito-temporal lesion, as had LDS and his pattern of reading was very similar to LDS's. Therefore it is possible that that area might have contributed, at least in the case of LDS, to a more efficient integration of letters.

We have also discussed the logic of the findings of complementary classical and strong dissociations, as it supports the existence of a qualitative difference between the patients. Performance on the letter processing tasks corresponds to a classical dissociation in the sense of Shallice (1988): FC's performance not only is better than LDS's, but is well within the normal range. By contrast, LDS is worse at letter level processing, but her performance on letter integration is superior to FC's. However, she still performs worse than her matched controls, thus showing a strong dissociation. In reading, letter identification occurs before letter integration: this implies that impaired letter processing will result in a less efficient letter integration process, even if this process is not itself damaged. With this organization of the system, one could never observe two classical dissociations. Complementary classical and strong dissociations are the most powerful combinations that could be observed.

In conclusion, data, showing a double dissociation, suggested that the nature of the deficit in pure alexia may differ qualitatively across patients. The results are consistent with the theories claiming the existence of different functional deficit in pure alexia (Price and Humphreys, 1992; Patterson and Kay, 1982; Hanley and Kay, 1996), differently from those arguing that a single type of general perceptual deficit is common to all the patients (Behrmann at al., 1998). Moreover the ability of the patients on object naming was not different from their matched controls, suggesting that not only the deficit can vary in pure alexia, but it seems also fairly pure, at least for our cases. The dissociation of FC and LDS is not compatible with any of the existing computational models, because most of them did not focus on the early stage of the visual recognition of words and more elaborations would be needed.

We claim that pure alexia can result from deficit at different levels of the visual word processing, from a more general perceptual deficit, as probably in some cases reported by Behrmann and colleagues (Sekuler and Behrmann, 1996; Behrmann et al., 1998), from a deficit at letter level, as in our case LDS and even from the integration of letters, as our case FC showed.

As secondary aspects, the results on implicit reading studied with semantic categorization tasks have shown that both the patients were unable to access the semantic information from briefly presented words, as has been reported by other authors (Chialant and Caramazza, 1998; Patterson and Kay, 1982). Finally we have studied the LBL reading strategy: the results have shown that, at least in Italian where spelling is not commonly used, reading can be carried out without using a spelling procedure. A strategy more visual in nature is likely to be used.

As regards the English pure alexic patients, the results have shown that all of them have letter identification problems. Moreover they have weak integration abilities, although these are differently distributed: AT2 is very impaired, TS shows some facilitation in conjoining letters together and SC has a performance in between. Finally the results have confirmed that the LBL reading strategy is not necessarily dependent on the spelling system.

We argue that it is more difficult to find qualitative distinctions beyond the letter level in English than in Italian mainly due to cross-linguistic differences in the orthography. In English letter integration is more difficult because of the inconsistency of the language; moreover, normal readers are likely to place a greater emphasis on the lexical procedures than the sublexical one (Paulesu et al., 2000) and therefore a LBL reading strategy might be more disrupting in English than in Italian. However, the results are compatible with our hypothesis of the different types of functional deficit in pure alexia, although they are less strong than those found with the Italian patients.

7.2 The effects of the damaged visual field in hemianopic alexia

Finally a case of hemianopic alexia has been studied. AT is an interesting case because under normal conditions he can read words accurately albeit more slowly, as a result of his visual field defect. However, with briefly presented words AT perceives words longer, adding extra-letters at the end of the words. In a series of different tasks, a total of more then 1000 words has been presented to AT: 23% of the words resulted in incorrect answers and these have been carefully studied in an analysis which has divided words into spatial quartiles. The results showed that the longer the word, the more last letters were omitted (e.g. avvocato (lawyer) >avvolgere (to wrap); cammello (camel) >cammino (path)), but the shorter the word, the more letters were added (dito (finger) >dittatura (dictatorship); vino (wine) >vincere (to win); bici (bike) >bistecca (steak); cocco (coconut) >cocomero (cucumber)). Moreover, in tasks where the position of the words was more controlled, the results indicated that the elongations increased when words were presented in the damaged visual field.

The results suggest that his parafoveal defect is responsible for the abnormal elongations and that the latter interact with the letter string position. His deficit might be not specific for reading, as it was evident also in a length judgment task.

AT does not show either a proper form of neglect dyslexia or a positional dyslexia since his errors tend to be longer than the target word, the elongations resolve fairly quickly and although they occur at the end of the words he knows the position of the letters. AT's damage is not parietal and is evident with words, therefore it is unlikely that his errors are the result of completion phenomena: completion usually refers to the ’completion’ of simple geometric visual patterns over a blind region. Interestingly, his brain damage is exactly in the occipital-temporal region, where the VWFA is supposed to be; according to the theory of Cohen and Dehaene AT should not be able to read. It would be interesting to see with an imaging study which brain areas allow him to read efficiently. It can be hypothesized that either that area is still active in word reading tasks and contributes to word recognition or that it is functionally damaged and therefore some other areas have taken over or that another area is involved, differently from Cohen and Dehaene's claim.

Most importantly, AT does not show neglect and his symptoms can be better described as confabulations following his brain damage which resulted in a visual field defect. This phenomena might be common to other syndromes like neglect without postulating more complex accounts.

7.3 Prospectives

Several questions for future research can be formulated to further investigate the visual recognition of words. As regards our patients, it would be interesting to investigate how the integration of letters occurs in sublexical parts. There is a growing body of evidence supporting the role of the syllable as a relevant sublexical unit in reading words, at least in languages such as Spanish and Italian which have clear syllabic boundaries (Carreiras and Perea, 2002; Alvarez, Carreiras and Taft, 2001). However, it is not clear whether syllabic effects arise from a sublexical phonological level or from a sublexical orthographic level, since there is evidence for both positions. It would be interesting to disentangle syllabic overlap from orthographic overlap in tasks of word reading (possibly with masked priming techniques) with our Italian patients, so that we can gain evidence whether syllables are phonological or orthographic in nature.

Moreover, can letter integration be considered similar to the integration process that we carry out when we see an object? Namely, can we assume the existence of a spatial integration system common to both word and object processing which comes into play when the spatial relations between distinctive parts within an object are critical for its identification? Then, for which reasons can the integrative process be impaired? Can it be accounted for by the problem of letter similarity, as suggested by some authors (e.g. Arguin and Bub, 2005)?

It could be interesting to study the temporal dynamics of letter and integration processes, for instance with MEG, with the patients who have different types of functional deficit, to see how the processing stream develops in different loci in time and space.

Then, we could study the relation between word and object processing. Does the letter-level deficit in patients like LDS or the English AT2 result from a more general perceptual deficit? Even if this was the case, the deficit impairs reading more than other forms of visual processing: so what makes words a special class of objects?

Finally, for the study of the hemianopic alexia shown by AT, it would be interesting to study whether the same elongations also occur in tasks with non-orthographic stimuli, like pairs of bisected lines presented briefly.

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Appendix A

List of orthographic syllable and nonsyllable used in the Syllable task (Chapter 4, Section 4.12.2)

N	orthographic syllable	N	orthographic syllable
1	bal	31	por
2	ber	32	pre
3	bor	33	pri
4	bra	34	psi
5	bre	35	pun
6	bri	36	rin
7	bru	37	riu
8	cir	38	san
9	cli	39	sba
10	cor	40	sca
11	cre	41	sco
12	cru	42	scr[a]
13	den	43	ser
14	dol	44	sfu
15	dre	45	sgu
16	fio	46	sin
17	fiu	47	sma
18	for	48	spa
19	fra	49	spe
20	fun	50	spi
21	gra	51	ste
22	gri	52	sti
23	gua	53	stu
24	gui	54	sve
25	lam	55	tan
26	len	56	tar
27	mal	57	ter
28	man	58	tom
29	par	59	tro
30	pen	60	tru

Table A.1: Set of orthographic syllables used in the Syllable task (Chapter 4). The set is consistent with the database of the orthographic syllables in written Italian (Stella and Job, 2001).

[a]This item has been removed because it is not an orthographic syllable according to the database of the orthographic syllables in written Italian (Stella and Job, 2001)

N	Orthographic nonsyllable (O-NSYLL)	Might the (O-NSYLL) be a phonological syllable in Italian?
1	bqo	no
2	etm	no
3	gme	no
4	igf	no
5	ipl	no
6	nds	no
7	plc	no
8	qco	no
9	stc	no
10	svr	no
11	tpa	no
12	uzt	no
13	vni	no
14	dsi	only if pronounced as /çi/ (but it is written *zi*)
15	mhi	only if pronounced as /mi/ (but it is written *mi*)
16	och	only if pronounced as /ok/ (but it is written *oc*)
17	qem	only if pronounced as /kem/ or /quem/ (but it is written *chem* or *quem*)
18	qer	only if pronounced as /ker/ or /quer/ (but it is written *cher*) or *quer*
19	tsa	only if pronounced as /ça/ (but it is written *za*)
20	vho	only if pronounced as /vo/ (but it is written *vo*)

Table A.2: Set of orthographic nonsyllables used in the Syllable task (Chapter 4). None of them is present in the database of the orthographic syllables in written Italian (Stella and Job, 2001). However some of them *might* be phonological syllables (although the assimilation, when possible, is not even automatic), but in that case they would have a different orthographic representation. In any case the ability of the patients to report the 3 letters of the orthographic nonsyllable is not influenced by the possibility of being a phonological syllable.