Acquired disorders of reading and writing: Cross-script comparisons

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Acquired Disorders of Reading

Matthew A. Lambon Ralph and Karalyn Patterson

Introduction

> "The problem is, I don't know what some of these words mean"
> Patient EK with semantic dementia

One of the many fascinations of working with neurological patients is their uncanny ability to highlight the key deficit that underpins their poor performance on a range of specific mental activities. Semantic dementia is a neurodegenerative condition of the temporal lobes bilaterally that results in a selective yet progressive loss of conceptual knowledge (Hodges, Patterson, Oxbury, & Funnell, 1992; Snowden, Goulding, & Neary, 1989). It is not uncommon for such patients to produce statements very similar to that noted for patient EK above. Although not unexpected in the context of defining a word's meaning, these frustrations are often expressed, as they were in this instance, when patients are simply asked to transform or maintain the surface form of the items; for example, in reading aloud, delayed repetition, or immediate serial recall tasks. As we will argue in this chapter, patient EK quite rightly diagnosed that her difficulties with a range of simple language activities were all due to a common underlying cause. More specifically, there is a growing body of evidence that the major types of acquired dyslexia (and other aphasic deficits) can all be described in relation to impairments of a limited set of interacting, primary brain systems (Patterson & Lambon Ralph, 1999).

An important aspect of this approach is that acquired dyslexias are considered in the context of the patients' other language and cognitive deficits with the assumption that there are important relationships between them. This represents a recent development in the methodology applied to acquired dyslexia and cognitive neuropsychology more generally. We begin with a short overview of the classic cognitive neuropsychological approach to acquired disorders of reading and proceed to discuss this more

contemporary approach in which data from such patients have been reinterpreted within a highly interactive, computationally explicit model of reading.

The classic cognitive neuropsychological approach to acquired dyslexias

The study of acquired reading disorders has played a central role in the development of cognitive neuropsychology ever since the seminal study of Marshall and Newcombe (1973). These early, insightful studies not only gave descriptions of the major acquired dyslexias but also grounded these in the context of models of normal reading. Normal and disordered processing were encapsulated in box-and-arrow type diagrams (Coltheart, Patterson, & Marshall, 1980; Ellis & Young, 1988). With the accumulation of patient data, particularly in the form of behavioral dissociations, increasingly elaborate theories have been produced. The process of reading starts in these models with an orthographic recognition system that specifies the identity and position of the letters within each word. This orthographic information is converted into the phonological form of the word via three routes or processes. A direct pathway translates letters into sounds using grapheme–phoneme conversion (GPC) rules. Although it is a serial and thus somewhat slow process, the GPC route is able to provide the correct pronunciation for familiar words that have a regular or predictable relationship between spelling and sound, and a correct pronunciation of novel orthographic strings (nonwords). For real words two other lexical pathways are available. Both are initiated by recognizing the word, which means matching the target letter string to a familiar word form in a visual input lexicon. This module passes information onto the semantic system allowing the meaning of the word to be retrieved. The semantic representation can, in turn, activate the corresponding representation in the speech output lexicon and thereby release its phonological form. The original conception of the dual-route model contained only this semantic-based lexical route in addition to the nonlexical GPC procedure (Marshall & Newcombe, 1973). Some single-case studies, however, led to the proposition that a direct pathway existed between the visual input lexicon and the speech output lexicon, allowing lexically based reading without involvement of the semantic system (see below for further discussion of patients who read despite significant semantic impairments).

Acquired dyslexias are assumed to arise from damage to one or more of the elements or pathways within this model. Peripheral dyslexias such as pure alexia, visual, attentional, and neglect dyslexia are usually explained in terms of insufficient or inaccurate visual information arriving at the orthographic recognition system. Pure alexia, for example, refers to a reading disorder in which there is a specific difficulty in recognizing printed words, without accompanying aphasia or agraphia. This combination of alexia without agraphia leads to the striking clinical observation of patients who are unable to read what they, themselves, have just written. Many of these patients try to aid their visual recognition in reading by using a letter-by-letter strategy either overtly or covertly (as indeed normal readers tend to do when reading with poor lighting or in other unfavorable conditions). This strategy produces the cardinal feature of letter-by-letter (L-by-L) reading – significant length effects on reading times that are typically absent in normal readers, at least until words get longer than about seven letters.

Surface dyslexia refers to an abnormal form of reading signaled primarily by a much exaggerated sensitivity to the joint impact of word frequency and spelling-sound consistency (Bub, Cancelliere, & Kertesz, 1985). Surface dyslexic patients are particularly prone to error in reading aloud low-frequency, inconsistent words. Their errors predominantly correspond to the more typical pronunciation for the orthographic elements within the word (e.g., reading PINT as though it rhymed with "mint": Marshall & Newcombe, 1973). In contrast, reading of regular words and nonwords is significantly better and, in the purest cases, is close to or within normal limits. In traditional dual-route accounts of reading, surface dyslexia is assumed to reflect an impairment of lexical reading; the most typical interpretation is that it results from damage to the orthographic input lexicon itself. Regular words and nonwords can be pronounced without error through the GPC route, which does not require word recognition. High-frequency irregular words are less affected than their low-frequency counterparts because, even in its normal state but particularly after damage, the efficiency of the lexical route is modulated by word frequency (Coltheart, Rastle, Perry, Langdon, & Ziegler, 2001).

Phonological dyslexia refers to a pattern in which oral reading accuracy exhibits a significant and abnormal lexicality effect; that is, very poor performance (including lexicalization errors) in response to nonwords. In traditional cognitive neuropsychological accounts, phonological dyslexia reflects damage to the GPC route. Real words, regular and irregular, are unhindered, as these can be read efficiently by the lexical routes. In contrast, nonwords cannot be read correctly, as these orthographic strings can only be transformed into phonological representations via the application of GPC rules. The fact that phonological dyslexic patients sometimes produce lexicalization errors (e.g., BEM → "Ben") is assumed to reflect the patients' attempt to read nonwords via the lexical reading routes.

The cardinal feature of deep dyslexia is the production of semantic paralexic errors (e.g., MERRY → "happy," CARNATION → "narcissus"), which makes this, perhaps, the most striking of the acquired dyslexias. In addition to the production of semantic errors, patients with deep dyslexia tend to have a range of other reading "symptoms" – that is to say, a set of co-occurring reading deficits and characteristics. These include poor phonological activation directly from print (i.e., poor or abolished nonword reading); relatively better reading of concrete than abstract words; a graded difficulty with words from different parts of speech (nouns, adjectives, verbs, function words, in descending order of accuracy – which may in fact be just another manifestation of the marked advantage for concrete over abstract words); and the production of morphologically related (e.g., WASH → "washing"; LOVELY → "love") and visually related paralexic errors (e.g., SIGNAL → "single," MOMENT → "money"). In traditional cognitive neuropsychological accounts, deep dyslexia is assumed to reflect major, widespread damage to the reading architecture. As in phonological dyslexia, poor nonword reading results from abolished grapheme–phoneme conversion. Nonsemantic lexical reading (along the direct connection between the visual input and speech output lexicons) is also assumed to be defunct, requiring patients to read solely by an impaired lexical-semantic route. The remaining efficiency of this pathway is graded by the strength or richness of the semantic representations – leading to the imageability effect on reading accuracy. Semantically related errors reflect inaccurate translation via word meaning or corruption to the semantic representations themselves.

These traditional models typically represent the cognitive architecture thought to underlie reading specifically and say little or nothing about how these modules link with the rest of linguistic and visual processing. At least two factors may have influenced the shift to a different way of considering various types of acquired dyslexia and other neuropsychological deficits. While considerable progress has been made through focused study of each individual patient's reading disorder, this approach has tended to avoid consideration of the potential relevance of the patient's other impairments. As we shall describe in some detail below, when researchers began to look across cases with certain types of acquired dyslexia, interesting associations with more general language and visual impairments were noted. Serendipitously, these evolving explanations for acquired dyslexia provide increasing synergies with the developmental dyslexia literature. The potential for linking accounts of the development and dissolution of reading is exciting and may be regarded as further motivation for pursuing this theoretical approach. A second factor relates to the rise of computational models to simulate normal and disordered performance. Rather than elaborating the cognitive architecture of the reading system as has tended to occur with box-and-arrow diagrams, computational models have typically treated reading as part of a larger language system, impairments to which would have consequences for other language tasks.

The triangle model: a theoretical framework

The "triangle" model of reading (Harm & Seidenberg, 2004; Plaut, McClelland, Seidenberg, & Patterson, 1996; Seidenberg & McClelland, 1989; the descriptive term "triangle model" was first used by Patterson & Behrmann, 1997) contains three principal components: (1) a visual component that, with respect to reading, handles orthographic processing, (2) phonology, and (3) semantics (see figure 22.1). None of these components is specific to reading; all operate in and indeed evolved for use in other language and cognitive activities (Patterson & Lambon Ralph, 1999; Savin, 1972). In effect, reading is taken to be parasitic on ontogenetically older brain systems and represents the interaction between fine visual processing and language mechanisms. The semantic system deals with all forms of verbal and nonverbal comprehension as well as any production task that requires a semantic source of activation, including speech production and drawing objects in response to their names (Bozeat, Lambon Ralph, Patterson, Garrard, & Hodges, 2000; Caramazza, Hillis, Rapp, & Romani, 1990; Lambon Ralph & Howard, 2000). Likewise the phonological representations involved in reading aloud are activated for any task requiring spoken production such as naming, repetition, immediate serial recall in addition to reading aloud (Lambon Ralph, Cipolotti, & Patterson, 1999; Lambon Ralph, Moriarty, & Sage, 2002). Again, it is interesting to note that similar arguments have been made in the developmental literature (e.g., Wagner & Torgesen, 1987; Hulme & Snowling, 1991). Finally, written word processing is accomplished by the generic visual system rather than a dedicated, reading-specific process (Behrmann, Nelson, & Sekuler, 1998a; Farah & Wallace, 1991). Some researchers have argued that a section of the left inferior occipito-temporal region – a part of the visual object recognition system – becomes specialized to orthographic recognition when people learn to read (the visual

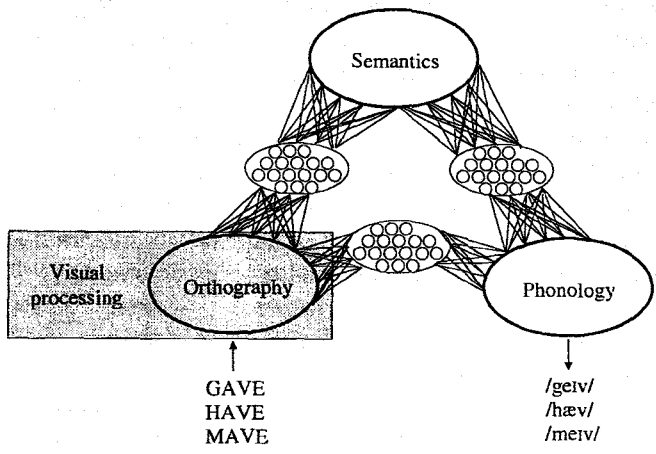

Figure 22.1 The triangle model of reading.

word form area: McCandliss, Cohen & Dehaene, 2003). This proposal is a topic of current debate, with other researchers questioning the specificity of this region to orthographic recognition (i.e., the same region is differentially activated by other nonvisual and nonreading tasks: Price & Devlin, 2003). In any event, the points we wish to emphasize are (1) that there is no section of the brain set aside for reading before we learn to read; and, (2) that given a nonreading task that necessitated the same kind of visual processing required for reading (i.e., rapid discrimination and identification of confusable symbols at least partly in parallel), we firmly predict that this task would recruit the same neural substrates as orthographic processing.

A full description of the triangle model can be found elsewhere (Plaut, this volume); however, some of its key characteristics are reviewed here prior to a consideration of how damage to one or more of its primary systems leads to the acquired dyslexias described above. Reading aloud is accomplished in this model by the interaction of the three primary systems via sets of connections that allow activation to pass and reverberate between the three groups of units. This means that the final form of representation computed on the phonological units, for example, is influenced by both the semantic and orthographic representations. The strengths or weights of the connections between units in each set of representations change gradually as the model is trained. For learning to read English words, the direct mapping between orthography and phonology (hereinafter O→P) comes to play the dominant role (Plaut et al., 1996). This is because in alphabetic languages, such as English, graphemes and phonemes are systematically related and this tight relationship only varies a little across words. Even in a frankly irregular word like YACHT, there is a transparent pronunciation of the *Y* and the *T*, and *A* receives one of its common pronunciations (cf. FATHER). The O→P computation is supplemented by the

interaction with word meaning, particularly through the interplay between semantics and phonology. Indeed, the S→P mapping is highly developed for speech comprehension and production considerably before children learn to read. In contrast, the efficiency of O→P develops as children learn to read and is influenced by word frequency and spelling-sound consistency. The direct mapping is least efficient for low-frequency, inconsistent words because the model has few opportunities to learn their particular, atypical mappings. Although the efficiency of S→P has to be high for all words (to support comprehension and speech production), the additional activation of phonology by word meaning is considered particularly important for accurate reading of these low-frequency inconsistent words.

Although this chapter is concerned with disordered reading, it is worth noting in this context that there is evidence that *normal* reading of low-frequency, inconsistent words is more influenced by a semantic variable – imageability – than higher-frequency or consistent words (Shibahara, Zorzi, Hill, Wydell, & Butterworth, 2003; Strain, Patterson, & Seidenberg, 1995). In addition, Rosson (1985) found that pronunciations given to nonwords such as LOUCH were influenced by a preceding word (e.g., *feel*) that semantically primed one sound-spelling pattern (*touch*) rather than another (*couch*).

In the remainder of this chapter, we will review data suggesting that each of the major acquired disorders of reading (surface dyslexia, phonological-deep dyslexia, pure alexia) is produced by impairment of a primary system (semantics, phonology, and fine visual processing, respectively).

Semantic Memory and Surface Dyslexia

The primary systems hypothesis relates surface dyslexia to a semantic impairment or, more precisely, to a dramatic reduction in the activation of phonology by word meaning (S→P). The literature on this type of reading disorder is dominated by patients with semantic impairment and profound anomia, most commonly in the context of semantic dementia (e.g., Patterson & Hodges, 1992; Shallice, Warrington, & McCarthy, 1983; Warrington, 1975) but sometimes after acute brain damage (Bub et al., 1985). The clinically derived link between semantic impairment and surface dyslexia can be traced back at least to 1980, when Shallice and Warrington in fact referred to this reading disorder as semantic dyslexia (Shallice & Warrington, 1980).

A more formal, computational description for the link to semantic impairment is given by the triangle model (simulation 4: Plaut et al., 1996). As noted above, the direct O→P computation in the triangle model is relatively inefficient for low-frequency, inconsistent words and this is compensated for by the contribution from word meaning (S→P). Without this additional semantic input, the pronunciation of words simply corresponds to the computation along O→P. The weights learned by O→P primarily reflect the mappings in consistent words, plus word-specific mappings for frequently encountered words; accordingly, when additional input from S→P is reduced, the model then gives a more typical, consistent pronunciation for low-frequency, inconsistent words – just like patients with surface dyslexia (e.g., YACHT → "yatched," SEW → "sue").

The last decade has witnessed increasing evidence in favor of the link between semantic impairment and surface dyslexia. First, the vast majority of patients with semantic dementia (SD) have surface dyslexic reading at presentation or, if they are very mild, after there has been sufficient decline in their semantic memory (Caine, Breen, & Patterson, 2002). Only three semantic dementia patients in the literature run counter to this prediction; these are discussed in more detail below. In addition, the onset of semantic impairment in Alzheimer's disease is associated with the emergence of surface dyslexia (Strain, Patterson, Graham, & Hodges, 1998), though again there are a few exceptions to this pattern (Lambon Ralph, Ellis, & Franklin, 1995; Raymer & Berndt, 1994, 1996).

In addition to the link between semantic impairment and surface dyslexia at the level of associated deficits, an item-specific relationship has also been demonstrated. Irregular words that individual semantic dementia patients still understand relatively well are read more accurately than words that have become semantically degraded (Funnell, 1996; Graham, Hodges, & Patterson, 1994). In a similar vein, there is now evidence for category specificity in surface dyslexia: semantic dementia patients comprehend number words relatively well, and their reading of number words with atypical spelling-sound correspondences (e.g., ONE, TWO, FOUR) is more accurate than for comparable non-number words (Butterworth, Cappelletti, & Kopelman, 2001).

Evidence for the primary systems hypothesis from behaviors beyond reading

A key aspect of the primary systems hypothesis is that the three principal components are assumed to underpin a variety of mental activities. Patients with semantic impairment demonstrate consistency by frequency interactions not only in reading but also in a whole range of verbal and nonverbal activities. When given the spoken stem of a verb and asked to produce its past tense form, for example, semantic dementia patients are particularly poor with low-frequency, irregular items (e.g., grind → ground) for which they produce regularization errors, "grinded" (Patterson, Lambon Ralph, Hodges, & McClelland, 2001). This phenomenon can also be observed in receptive as well as expressive tasks. Rogers, Lambon Ralph, Hodges, and Patterson (2004) asked semantic dementia patients to make two-alternative forced-choice lexical decisions to written words, each paired with a nonword that was either more or less consistent with typical English orthography (as measured by bigram and trigram frequencies). With greater degrees of semantic impairment, patients' lexical decisions became increasingly guided by a preference for typical orthographic patterns, whether or not these actually corresponded to real words. As with reading aloud and generating past tense verbs, the semantic dementia patients were particularly inaccurate for low-frequency words with atypical letter patterns and demonstrated a strong preference for the accompanying nonword with a more typical letter sequence (e.g., preferring GOAST to GHOST).

Analogous behavior has been observed in receptive and expressive nonverbal tasks once measures of consistency (i.e., the reliability of how elements in the input map onto their corresponding output representations) have been defined for those domains. In a parallel study, Rogers, Lambon Ralph, Hodges, and Patterson (2003) found an increasing influence of consistency in object decisions as a function of degree of semantic deficit.

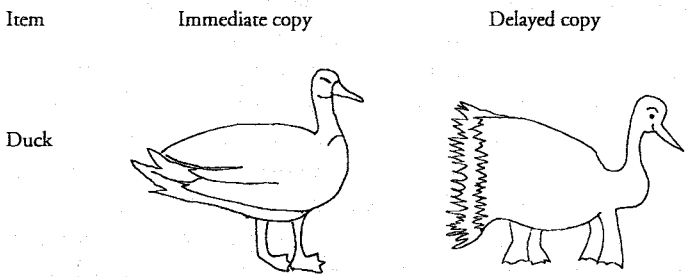

Figure 22.2 An example of drawing overregularization from a patient with semantic dementia.

SD patients were more likely to indicate that line drawings of animals or objects were real if their component elements were typical/consistent of those found in that domain (e.g., for animals – four legs, ears, etc.) than if the depictions contained atypical/inconsistent features (e.g., humps, stripes, antlers). Again the rate of errors was highest when the correct real object not only had atypical elements but was also of lower familiarity. This pattern can also be observed in drawing to name and delayed copying of object drawings (expressive measures of visually based knowledge). Patients are increasingly likely to omit features in their drawings that are unusual (inconsistent) for the relevant semantic domain and become more likely to include erroneous features that are typical/consistent (Bozeat et al. 2003). With sufficient semantic impairment, patients can produce striking and memorable errors such as drawing ducks with four legs (see figure 22.2 and Lambon Ralph & Howard, 2000).

A final example of semantic impairment leading to frequency by consistency effects in nonverbal tasks is that identified in object use. By using a feature-type database, Bozeat et al. (2003) were able to define not only the elements involved in object use (e.g., features of hold, manipulation, and object structure) but also co-occurring pairs of structure-hold and structure-manipulation relationships (i.e., affordances: for example, if an object has a handle it is very likely that the correct manipulation includes gripping the object in question; Bozeat, Lambon Ralph, Patterson, & Hodges, 2002). One can think of these as being analogous to typical spelling-sound correspondences in reading and thus make the prediction that afforded elements, like regular words, would be relatively impervious to the familiarity of the object or the degree of semantic impairment each patient has. Consistent with this hypothesis, Bozeat et al. (2002) found that semantic dementia patients exhibited a consistency (afforded vs. nonafforded manipulations) by familiarity interaction in their object use accuracy.

Challenges to the primary systems hypothesis

Given the apparent wealth of evidence for the role of conceptual knowledge in reading (Kintsch, this volume) as well as in a range of other verbal and nonverbal tasks, we would argue that the semantic corner of the triangle model is firmly established. The small

number of patients described in the literature who have significant semantic impairment without the predicted surface dyslexia, however, provide a clear challenge to this approach. These include some cases with Alzheimer's disease (Lambon Ralph et al., 1995; Raymer & Berndt, 1996) as well as three with semantic dementia (Blazely, Coltheart, & Casey, in press; Cipolotti & Warrington, 1995; Schwartz, Saffran, & Marin, 1980). It can be very difficult to establish the degree of semantic impairment in Alzheimer's disease because this disorder leads to a variety of cognitive and perceptual impairments, many of which can compromise performance on tests designed to tap conceptual representations (Nebes, 1989). This is made all the more likely in Alzheimer's disease in that patients are often moderately to severely demented before clear semantic impairments emerge (Strain et al., 1998). In such circumstances, it can be easy to overestimate the true degree of semantic impairment and conclude that such patients are reading without meaning. Because of the selective nature of semantic impairment in semantic dementia, these cases provide a clearer test of the putative role of word meaning in reading aloud.

The first and, perhaps, still best known counterexample to surface dyslexia in semantic dementia is case WLP (Schwartz et al., 1980b). At one point during her decline, WLP was able to read irregular words that she failed to understand, for example correctly reading aloud the irregular word HYENA and then commenting: "hyena . . . hyena . . . what in the heck is that?" At the time, this dissociation was deemed sufficient to motivate the addition of a nonsemantic, lexical pathway to dual-route models of reading, turning them effectively into triple-route models. This conclusion is tempered though by observation that, a little later in her progression, WLP developed a surface dyslexic pattern of reading just as one would predict from the triangle model. Indeed, more recent longitudinal studies indicate that if semantic dementia patients are very mild when they first present, then the semantic impairment can be insufficient to produce surface dyslexia (Caine et al., 2002). Careful study of one relatively full computational instantiation of the triangle model (simulation 4: Plaut et al., 1996) demonstrates that the relationship between semantic impairment and reading accuracy is nonlinear. Because O→P takes the dominant role in reading, the influence of semantic representations is subtle and surface dyslexia only begins to emerge once there is considerable semantic impairment. The model is consistent, therefore, with the pattern described by Caine et al. (2002) and by Schwartz et al. (1980b) – relatively normal reading in the earliest phase of semantic dementia but emergent surface dyslexia as the inevitable semantic deterioration takes hold.

Two more recently described cases, however, do not fit with this predicted relationship. Patients DRN (Cipolotti & Warrington, 1995) and EM (Blazely et al., in press) both had semantic impairment of a degree sufficient to produce surface dyslexia in numerous other semantic dementia patients but not in these two cases. One obvious, albeit dull, explanation for the difference, that apparent variations between patients reflect differences in the test materials employed, can be immediately ruled out in these cases because DRN and EM were tested on some of the same assessments that other studies have used to demonstrate associations between semantic impairment and surface dyslexia (Graham et al., 1994; Patterson & Hodges, 1992).

This state of affairs is a challenge to the triangle model, which needs to address the atypical cases, and also to dual-route theories. The latter framework (see Coltheart, this volume) needs to explain the nearly, though not quite perfectly, predictable impact of

semantic impairment on reading as well as on other verbal and nonverbal tasks. The only explanation that has been offered from this perspective is that these are accidentally associated deficits based on anatomical proximity of regions responsible for conceptual knowledge and reading aloud. On the basis of a steady increase in both the number of cases demonstrating the association, and the number of quite different tasks (e.g., reading, spelling, verb inflection, lexical decision, object decision, drawing, object use) demonstrating a frequency-by-typicality modulated impairment in conjunction with semantic deficits, we suggest that this "accidental association" becomes a less plausible account.

To explain such dissociations when they do (rarely) occur, Plaut (1997) demonstrated that two forms of individual difference could modulate the relationship between semantic impairment and reading accuracy sufficiently to account for all the patients reported in the literature, at least to that date. The efficiency of O→P can be modulated either by restricting the influence of semantic representations during learning to read or by increasing the computational resources of the direct O→P mapping. These individual differences continua mean that the degree of surface dyslexia produced by the same semantic impairment varies and, at one end point of these continua, reading accuracy can remain intact. Demonstration that this possible explanation actually works at the computational level is a key step in producing a complete theory. Future research will have to extend this to behavioral studies of patients with semantic dementia and will need to provide methods for measuring such individual differences across patients.

Phonology and Phonological-Deep Dyslexia

When the first case of phonological dyslexia was described, Beauvois and Dérouesné (1979) suggested that this type of disorder might represent a qualitatively different reading impairment to deep dyslexia. Although both phonological and deep dyslexic patients have problems reading nonwords, the patient they described (RG) did not make semantic errors and had little in the way of other aphasic impairments. The alternative possibility, that phonological and deep dyslexia might be more intimately related, was also noted in these early descriptions: Beauvois and Dérouesné suggested that patient RG might just have been a very pure, mild example of a deep-dyslexic-like reading disorder. Likewise in the second report of phonological dyslexia, Patterson (1982) noted that she had originally assumed patient AM to be a mild deep dyslexic rather than suffering from a different form of reading disorder.

More recently, the link between phonological and deep dyslexia has been championed by Friedman and colleagues (Friedman, 1996; Glosser & Friedman, 1990). In a review of the literature, Friedman (1996) noted a number of patients originally presenting with deep dyslexia whose reading pattern had evolved into phonological dyslexia over time. More specifically, Friedman argued that there was a succession of symptoms along a severity-based continuum of phonological-to-deep dyslexia: in the mildest patients only nonword reading would be affected; then, as a function of increasing severity, morphological and visual errors would appear, followed by part-of-speech effects, then imageability effects and finally the production of semantic errors.

The appraisal of this continuum hypothesis is hindered by the fact that previous case studies have not used comparable test materials. To circumvent this difficulty, Crisp and Lambon Ralph (submitted) investigated 12 patients with phonological or deep dyslexia using the same test battery in order to facilitate direct comparisons between cases. With the exception of semantic paralexic errors, all patients including those with mild reading difficulties demonstrated most of the symptoms of deep dyslexia, including significant effects of imageability (taken by Friedman to be the penultimate symptom before semantic reading errors). Thus, the findings of this study strongly reinforced the notion of a severity-based continuum between deep and phonological dyslexia, although there was little evidence in favor of the predictable succession of symptoms suggested by Friedman (see above). In short, the phonological-deep dyslexia continuum involves a common co-occurring set of symptoms, the severity of which varies in line with the overall degree of reading impairment.

Given this fairly clear evidence favoring a phonological-deep dyslexia continuum, the most obvious next question concerns the cognitive basis of the reading disorder. The triangle model suggests that it is a general phonological impairment in which the phonological component of the language system is pathologically weak or underactivated (Patterson, Suzuki, & Wydell, 1996). Note that a significant relationship between poor phonology and impaired reading is a standard finding in the developmental literature (Harm & Seidenberg, 1999; Snowling, 2000; Vellutino & Fletcher, this volume). A generalized deficit of phonology should have an impact on reading aloud of all types of orthographic string, but there are at least two reasons why it might be expected to have disproportionate consequences for unfamiliar strings (i.e., nonwords). First, setting aside the computation from O→P, the phonological system itself develops on the basis of experience with words in the speaker's language, and it will always operate more efficiently with the familiar phonological sequences of the words that it knows than with the nonwords that it does not; this will be true even in its normal state, but the familiarity effect should be magnified by damage. The second important distinction comes from the fact that words, unlike nonwords, have meaning. As noted in the introduction at the beginning of this chapter, communication between S and P (in both directions) has to be highly efficient because it supports all forms of speech comprehension and production. Studies of phonological short-term memory in semantic dementia patients and related studies with neurologically intact participants have established that semantic representations play a role in constraining and maintaining the phonological coherence of words (e.g., Hulme, Maughan, & Brown,1991; Walker & Hulme, 1999; Jefferies, Jones, Bateman, & Lambon Ralph, in press; Patterson, Graham, & Hodges, 1994). One could easily imagine, therefore, that this interplay between semantics and phonology may assume exaggerated importance when phonology itself is compromised. Weakly activated phonological patterns can be enhanced by the interaction with intact semantic representations for words that are obviously absent for nonwords.

The interaction with meaning might also provide an explanation for some of the other symptoms found in phonological-deep dyslexia. The quality of the interplay between semantics and phonology is likely to be graded according to characteristics of the words themselves. In particular, imageability effects might reflect the relative strength of the semantic support, given that high-imageability words are thought to have relatively better

specified, richer, and more context-independent semantic representations than their abstract counterparts (Jones, 1985; Plaut & Shallice, 1993). This difference might also be germane to an understanding of semantic paralexias. For patients with a very severely impaired phonological system, activation of word meaning will be computed only on the basis of the O→S pathway, the weakest link in the triangle (Harm & Seidenberg, 2004). If this results in only partial semantic activation, then the S→P computation may produce several semantically related candidates in the phonological system – those that share features with the underspecified representation activated in semantic memory (e.g., for the target HARE the group of semantically related targets might include *rabbit, squirrel, guinea pig*, etc.). For the most part, the combination of any residual O→P activation together with partial reinforcement from word meaning will favor the target word form. In very impaired cases, however, it is possible that S→P will override the target form with phonological activation of a semantically related neighbour (e.g., HARE → "rabbit"). This is particularly likely if that item is more readily activated in normal circumstances; for example, if the incorrect word has a richer semantic representation or benefits from other factors that influence the efficiency of speech production, such as imageability, age-of-acquisition, and frequency (Gerhand & Barry, 2000). It also fits with the observation that semantic errors tend to be more imageable/concrete than the target words to which they are incorrect responses (Newton & Barry, 1997; Shallice & Warrington, 1975).

Where appropriate testing has been carried out, most if not all patients with phonological and deep dyslexia have been shown to have deficits of phonology outside the domain of reading. For example, all 17 patients with phonological dyslexia reported in a special issue of *Cognitive Neuropsychology* (no. 6, 1996) performed below the normal range on nonreading phonological tasks. Impaired phonology can be revealed by poor immediate and delayed repetition of words or nonwords, though stringent assessments, such as rhyme judgment, phonological segmentation and blending tasks, or measures of verbal working memory, are more likely to highlight the phonological impairment in these patients.

As noted above, the phonological-deep dyslexic literature is dominated by single-case studies. This means that the association with poor phonology across these different cases has been the key evidence for this explanation of phonological-deep dyslexia. There are three studies, however, that greatly strengthen the case for a causal link. The first two demonstrate a tighter relationship between phonological impairment and phonological-deep dyslexia by highlighting overlapping characteristics of patients' performance in reading and nonreading tasks. If this reading deficit, characterized by such a strong lexicality effect, is underpinned by a general phonological impairment, then one might expect the lexical status of the target form to influence success in any task requiring a phonological response, whether the stimulus is orthographic or not. Patterson and Marcel (1992) found exactly this. In their study, patients were more accurate when attempting to perform phonological manipulations (segmentation or blending on the basis of auditory input) when the correct manipulation resulted in a word response ("Take the first sound off the word 'mother' and say what remains") than a nonword response ("Take the first sound off the word 'father' and say what remains").

This logic can also be extended to deep dyslexia. Beland and Mimouni's (2001) study of an Arabic–French bilingual deep dyslexic patient ZT was primarily aimed at explor-

ing the impact of different writing systems on the symptoms of this reading disorder. In addition the authors completed a thorough investigation of the patient's nonreading language skills. Like many other patients with deep dyslexia, ZT was able to repeat single real words with very few errors. When a 5- or 10-second delay was inserted between the spoken stimulus word and ZT's repetition response, however, his repetition accuracy dropped by 30% to 40% in both languages. More interestingly, once his phonological system had been stressed in this way, the distribution of the patient's repetition errors mimicked those found in reading, including phonological, morphological, and omission errors, and perhaps most striking, semantic paraphasias too.

The case for a causal link between semantic impairment and surface dyslexia has been strengthened significantly by the demonstration that, as the degree of semantic impairment varies, so does the resultant reading accuracy (either across patients or in studies that focus on a by-items analysis). Until very recently, it has been difficult to compile parallel data for the link between phonological impairment and phonological-deep dyslexia because of the lack of comparable data across individually studied patients. This goal was tackled, however, by Crisp and Lambon Ralph's (submitted) case-series study noted above. As predicted, the patients' nonword reading accuracy was significantly correlated with their varying degrees of success on phonological manipulation tasks. In addition, the size of the patients' lexicality effect was strongly correlated with their ability to derive meaning not only from written but also spoken words (as measured by various synonym judgment tasks). This suggests that phonological-deep dyslexia reflects the interplay between the patients' general phonological impairment and the status of their semantic memory (or the efficiency of its interaction with surface word forms).

Extension to nonreading activities

A critical aspect of the triangle framework is that any acquired reading disorder should be accompanied by a predictable range of other deficits because it arises from impairment to one of the primary systems. As noted above, semantic impairment apparently leads to consistency-by-frequency interactions not only in oral reading but in a whole set of other verbal and nonverbal tasks. There is some evidence that the same cross-task associations are true for phonological impairments. For example, in the previous section on the impact of semantic impairment, we noted that when semantic dementia patients are given the spoken stem of a verb, they demonstrate relatively poor performance for low-frequency, irregular verbs and produce regularization errors (Patterson et al., 2001). In contrast, patients with Broca-type aphasia are impaired at generating past tense forms of verbs, and particularly so for novel verb forms (i.e., they exhibit a lexicality effect). Some of these cases also demonstrate relatively lower accuracy on the past tense of regular than irregular real verbs (Ullman et al., 1997). Bird, Lambon Ralph, Seidenberg, McClelland, and Patterson (2003) argued that the Broca patients' poor verb abilities were due to their phonological deficit and, more specifically, that the difference between regular and irregular verbs reflected the variation in phonological complexity of the items (the phonological complexity of the offset or the syllable length is automatically increased when regular verbs are inflected for the past tense). Interestingly, this case-series of ten Broca aphasic

patients was also impaired on a range of phonological awareness tasks and, as predicted, all had phonological-deep dyslexia.

A similar association across tasks was noted in a study designed to investigate the link between semantic/phonological impairments and word-finding difficulties in patients with aphasia following cerebral vascular accidents (stroke: Lambon Ralph et al., 2002). As noted throughout this chapter, activation from S→P is required for a variety of language activities including speech production. This predicts that the degree of word-finding difficulties found in aphasic patients should be related to the degree of their phonological and semantic deficits. Correlational analyses across the case-series of patients yielded strong support for this proposal – in fact, measures of semantic and phonological ability were as good at predicting naming scores as a separate naming test (i.e., as good a predictor as the test validity). Furthermore, phonological impairments dominated the neuropsychological profiles of this series of CVA patients, and, just as the triangle model would predict, the patients all had phonological dyslexia.

Visual Processing and Pure Alexia (Letter-by-Letter Reading)

Although there is considerable debate about the locus of impairment that gives rise to pure alexia (Coltheart, 1998), virtually all current explanations share the notion that this reading disorder reflects a relatively early impairment in the visual-orthographic system, leading to poor whole-word recognition. Early investigations of pure alexia in the cognitive neuropsychology literature, like other studies of acquired dyslexia, used reading-specific models to interpret the patients' data. These studies suggested that pure alexic patients either had impaired letter recognition or a breakdown between letter and word representations (e.g., Patterson & Kay, 1982). As noted in the introduction above, the primary systems hypothesis views the reading process first and foremost as an interaction between visual and language processes. We have argued that surface, phonological, and deep dyslexia can all be linked to nonreading impairments within the language system. In the same way, it is also possible to consider pure alexia in terms of a more generalized visual impairment (Behrmann et al., 1998a; Farah & Wallace, 1991; Mycroft, Behrmann, & Kay, in press).

Word recognition requires the reader to discriminate amongst and identify visual symbols rapidly and, at least to some degree, in parallel. It is not hard to imagine that this places considerable demands on the visual system and, therefore, that reading might be especially vulnerable to even a mild deficit of visual processing. In support of a link between pure alexia and visual perceptual deficits, Behrmann et al. (1998b) reviewed 57 published cases of L-by-L reading and found that 50/57 showed frank deficits in single-letter identification, at least in speed/efficiency if not in accuracy; for the remaining 7, there was insufficient evidence to rule out an impairment in this rather peripheral aspect of the reading process. This rate of associated deficits is remarkably high given that most of the patients had not been assessed on tasks with nonorthographic stimuli that might tax visual discrimination/identification as heavily as word reading does. A small number of studies have carefully constructed the visual stimuli so that these nonreading, visual tasks possess some of the same processing demands as reading. When this kind of careful

empirical assessment has been done, the patients were impaired on these tasks as well (Behrmann et al., 1998a; Mycroft et al., in press).

To date, there have been no case-series studies of L-by-L readers that have attempted to relate the degree of difficulty in visually demanding nonreading tasks to the severity of the patients' reading impairment (measured, perhaps, by the gradient that relates reading times to word length) as has been done for semantic impairment – surface dyslexia, and phonological impairment–phonological dyslexia. Such a study would first have to specify the nature of the critical, underlying visual deficits more precisely – a non-trivial matter. Evidence for a causal link between visual impairment and pure alexia has been provided, however, by a recent study that investigated the characteristics of reading and nonreading processing across the same set of seven patients (Mycroft et al., in press). As argued for the generalized phonological impairment underpinning phonological-deep dyslexia, the proposal for a generalized visual impairment causing L-by-L reading would be strengthened if some of the classic traits of pure alexia could be observed in nonreading tasks. Using carefully constructed visual search tasks (either for letters or symbols within a string of the same type of stimuli), Mycroft et al. demonstrated that patients' decision times were affected by the left-to-right location of the target stimulus as well as the length of the strings in the same way for both the reading and nonreading versions of the tasks. In stark contrast, normal readers showed no evidence for the influence of these factors on their decision times in either version of the task.

When considering the effects of semantic or phonological impairments, it is possible to find evidence for the impact of such deficits on a predictable set of other tasks. The same should also be true for the ramifications of a generalized visual deficit. More specifically, if pure alexia reflects the inability of the damaged visual system to support the high processing demands of word recognition, then it should also impair recognition of other classes of stimuli that share the same visual characteristics. The evidence that exists in the literature, in this regard, is mixed. At least some well-documented L-by-L readers are also significantly impaired in identifying numbers (Miozzo & Caramazza, 1998), musical notation (Horikoshi et al., 1997), and/or faces (De Renzi & di Pellegrino, 1998). There are, however, a small number of patients for whom number recognition seems significantly better than letter identification (Farah, 1999). This apparent dissociation between number and letter recognition requires further investigation and will necessitate a careful analysis of the characteristics of both types of stimuli. More generally, a major challenge for future studies of pure alexia will be to use the growing knowledge about the neural bases of visual processing to specify the elements that make up the visual corner of the triangle model. This might facilitate identification of the visual processing deficits that produce L-by-L reading behavior (which might actually vary in type across cases), and also of the basic impairments responsible for other forms of peripheral dyslexia such as attentional, neglect, and visual dyslexias (Ellis, Flude, & Young, 1987; Lambon Ralph & Ellis, 1997; Warrington, Cipolotti, & McNeil, 1993).

To finish this section on pure alexia, we will consider a further challenge to the visual impairment hypothesis – namely, the fact that some patients with severe L-by-L reading exhibit the "Saffran effect" (Lambon Ralph, Hesketh, & Sage, 2004). Although reports of implicit word recognition in pure and global alexics first appeared in the literature in the early 1960s (Kreindler & Ionasescu, 1961), the most comprehensive investigations of

such patients were reported by Saffran and her colleagues (Coslett & Saffran, 1989; Coslett, Saffran, Greenbaum, & Schwartz, 1993; Saffran & Coslett, 1998). The clinical manifestation of this phenomenon is that a patient succeeds, at a rate well above chance, in categorizing the meaning of a written word (e.g., is it the name of a living or a nonliving object?), or the lexical status of a letter string (is it a word or nonword?), despite being unable to identify the words themselves (i.e., to read them aloud). In the first detailed investigation of the Saffran effect (Shallice & Saffran, 1986), ML, a slow L-by-L reader, performed above chance on a series of forced-choice lexical decision and categorization tasks, even when the words were presented too briefly for him to report the identity of the word or its constituent letters. Subsequent descriptions of other patients found a similar disparity between above-chance (though never perfect) performance in the context of brief presentation and little or no explicit word recognition (for an overview, see Saffran & Coslett, 1998).

It is worth noting one limiting factor on the potential significance of this contrast between word identification and word categorization: the two tasks have very unequal demands. The task at which pure alexic patients are so impaired – identification – essentially asks readers to decide what word they are looking at when the correct answer could be (almost) any word in the language; the task at which they are significantly better – categorization – asks them to assign the target word to one of only two classes. A fairer contrast would be to make the identification task two-choice as well: given a brief presentation of the word HARE, for example, the patients could be asked in the classification task if it was a living or a nonliving thing, and in the identification task if it was the word "hare" or "hate." If they were still much more successful at the former judgment than the latter, the contrast would be more impressive. Exactly this comparison has been made in a recent study of the Saffran effect in a L-by-L reader (Lambon Ralph et al., 2004). This investigation found that the apparent difference between identification and classification was removed when a two-forced choice method was used in both tasks.

The ability to categorize words without overtly recognizing them is, perhaps, the most striking form of the Saffran effect, but a range of other results also suggests activation of higher-level, lexical-semantic representations in pure alexic patients. For instance, some L-by-L readers have exhibited a significant word superiority effect (better letter recognition under masked presentation if the stimuli correspond to words, or wordlike nonwords, than letter strings: Reuter-Lorenz & Brunn, 1990). In addition, pure alexics show influences of variables associated with lexical-semantic processes (frequency and imageability: Behrmann et al., 1998b) and can also demonstrate standard Stroop interference (McKeeff & Behrmann, 2004).

At face value, explanations of pure alexia based on early visual impairment seem at odds with the Saffran effect: the early visual deficit is supposed to prevent activation of whole-word representations, thus necessitating the L-by-L strategy for word recognition; but the Saffran effect is evidence for activation of high-level representations, including word meaning. Indeed, the explanation championed by Saffran, Coslett, and their colleagues (1998) separated the key behavioral elements observed in these patients into two groups and posited a separate, reading-specific system to account for each. They argued that a left hemisphere system supports explicit letter and word recognition and, when damaged, resorts to L-by-L reading as a compensatory strategy, whereas a second reading

system in the right hemisphere reading system underpins implicit letter and word recognition. Under this proposal, when the left hemisphere system is damaged, as it is in pure alexic patients, their L-by-L behavior derives from the abnormal processing in the left hemisphere while the Saffran effect is generated by the right hemisphere.

The Saffran effect can, in fact, be explained by the generalized visual impairment hypothesis. This explanation assumes that inadequate visual input (whether in pure alexics or in normal readers operating under visually difficult conditions) drives two processes: a *single* word-processing system (i.e., the triangle model), and a separate, compensatory L-by-L strategy. Under this proposal, the Saffran effect simply reflects the partial, remaining activation of the usual (but now inadequate) word recognition system (Behrmann et al., 1998b; Feinberg, Dyches-Berke, Miner, & Roane, 1994; Lambon Ralph et al., 2004; Shallice & Saffran, 1986). This explains why performance on lexical decision and categorization tasks is significantly above chance but never perfect (indeed sometimes a long way from perfect) in these cases. The combination of early visual-orthographic impairments and partial semantic access would only be surprising if the reading system were thought to consist of discrete processes. In a cascading or interactive activation system, degraded input will still produce partial activation in subsequent parts of the processing system (Lupker, this volume; Morton & Patterson, 1980; Shallice & Saffran, 1986). In McClelland and Rumelhart's (1981) interactive activation word recognition model, reduced input to letter features still produced partial letter and word level activation. This was, of course, the basis for their explanation of the word superiority effect in normal readers under masked, brief presentation. Behrmann et al. (1998b) argued that it also explains why pure alexics can show the word superiority effect (feedback from lexical-semantic knowledge), effects of frequency and imageability (frequent and concrete words are more readily activated even under reduced visual input), and above chance performance on lexical decision and categorization tasks (partial activation of lexical and semantic representations).

Future research directions

Up to now, the literature on acquired disorders of reading, like much of the rest of cognitive neuropsychology, has been based on the study of patients who have reached a chronic stable phase, normally several months or years after the onset of their brain injury. In contrast, studies of the decline in patients with neurodegenerative disease have provided important insights about the link between reading impairments and the underlying primary brain systems involved. This suggests that the changing brain provides useful information about the normal underlying processes that support reading as well as other aspects of cognitive performance. An area that is overlooked by most studies is the period of recovery following acute brain injury. A few notable exceptions to this "neglect" can be found in the acquired dyslexia literature and have led to important insights about the nature of the underlying systems (e.g., the recovery of deep into phonological dyslexia: Friedman, 1996). Indeed, in a recent exploration of plasticity in the context of a computational model of O→P, no clear form of acquired dyslexia emerged immediately following damage to the network (as described in earlier work: Patterson, Seidenberg, &

McClelland, 1989) but the pattern of surface dyslexia did crystallize out of the model after a period of recovery (Welbourne & Lambon Ralph, in press).

In our overview of the acquired dyslexia literature, we have attempted to summarize a growing body of evidence in favor of the idea that reading is parasitic on the pre-existing brain systems that support visual processing and language (Savin, 1972). Although there are now a considerable number of studies supporting each element of the primary systems hypothesis (Patterson & Lambon Ralph, 1999), this approach still faces a number of important challenges. Future studies of acquired disorders of reading should be able to address many of these challenges through additional neuropsychological studies and computational modeling. In addition, information from neuroimaging and transcranial magnetic stimulation studies of intact and brain damaged individuals will bring new perspectives on acquired dyslexia (Leff et al., 2001a; Leff, Scott, Rothwell, & Wise, 2001b; Price, 2000; Price & McCrory, this volume).