Agraphia and Alexia

D Fiset, Université du Québec en Outaouais, Gatineau, QC, Canada
D Bub, University of Victoria, Victoria, BC, Canada

Glossary

Cognitive architecture The component processes and their interconnections that make up a more complex mechanism involved in a task like reading single words aloud or writing words to dictation.

Grapheme The smallest combination of letters associated with an elementary sound unit. Graphemes can be as small as a single letter. For example, the letter P corresponds to the pronunciation 'puh' while the letter pair PH (a bigram) corresponds to 'fuh.'

Hemifield One half of the field of view defined according to retinal coordinates.

Hemispatial neglect A neuropsychological condition that affects attention, exploration, and awareness of the hemispace opposite the damaged hemisphere. Clinical manifestations of neglect include bumping into objects and walls, ignoring objects, persons, and sounds coming from the affected side, forgetting to shave (or applying make-up) to one part of the face, etc.

Modular Refers to the idea of separable cognitive components working together to carry out a task. For example, the orthographic lexicon is a component that is modularly distinct from the phonological lexicon. Modularly distinct components can be independently affected by neurological damage.

Orthographic lexicon The stored representation of the spelled form of words in the reader's vocabulary. The orthographic lexicon maintains each word as a sequence of abstract letter identities.

Phonological lexicon The stored representation of the pronunciation of the words in a speaker's vocabulary.

Retinocentric A spatial coordinate system centered on the retina.

Syndrome In cognitive neuropsychology, the term refers to a cluster of impairments on a number of different tasks, and the co-occurrence of symptoms reflects a theoretically important principle. The term is not applied to impairments that co-occur simply because neurological damage has fortuitously affected a number of unrelated processes.

The advent of the visual word as a means of communicating ideas and preserving knowledge is of fundamental importance to human development. Because of the crucial role played by written language in so many domains it is no surprise that learning how to fluently read and spell occupies much of our initial efforts at school. As a general rule, good readers are also good spellers, while bad readers have a harder time producing the correct form of written words. This correlation suggests that these two tasks share common substrates. But given the demands specific to each task, reading and writing cannot depend on exactly the same underlying mental processes. For example, the rapid identification of a letter string depends on certain mechanisms unique to vision, whereas the retrieval and execution of letter shapes require motor processes specific to handwriting.

The main goal of this chapter is to describe and understand how certain neurological lesions can induce deficits in reading and writing (i.e., alexias and agraphias). What these different pathologies can tell us about the organization of the mental processes responsible for reading and writing in normal individuals will also be addressed. Neurological damage can selectively affect the peripheral components of word processing (e.g., the perception or production of letters) or its central components (e.g., the relationship between the orthographic form of a word and its sound or meaning). Generally speaking, the more peripheral the damage, the more likely the impairment will be specific to either reading or writing. Throughout this article, the reading and writing system will be deconstructed into different components, and the disorders associated with their impairment will be described. First, the neuropsychological disorders related to the uptake of visual information will be presented. Next, deficits resulting from damage to more central mechanisms will be explained. These central mechanisms relate to orthographic, phonological and semantic knowledge, and often affect both reading and writing simultaneously. Lastly, the deficits that follow damage to orthographic output or, more specifically, to letter production, will be described.

Peripheral Dyslexias

Skilled reading demands accurate and rapid visual access to the symbols displayed on a page or computer screen. The quality of the visual information perceived is in part determined by the capacity to efficiently direct one's gaze and/or attention toward the text on the page (e.g., find the first paragraph). At the same time, in order for reading to be rapid and efficient, one must quickly recognize the letters in each word while maintaining their relative positions (e.g., LISTEN is not the same word as SILENT). In brain-lesioned patients who suffer from reading disorders as a result of damage to peripheral systems, the visual information perceived is inadequate to support accurate or fluent reading. Although these lesions will frequently have major repercussions on reading abilities, they will not affect spelling. Three types of peripheral alexias will be described in this section: pure alexia, attentional dyslexia, and neglect dyslexia. Note that we will use the term 'alexia' for some varieties of reading disorder and 'dyslexia' for others. 'Alexia' has

generally been the term that denotes reading disorders acquired as a result of neurological injury, while 'dyslexia' has more commonly been used to refer to developmental disorders of reading. However, the term 'dyslexia' has also been applied to certain forms of acquired reading disorder (e.g., deep dyslexia, attentional dyslexia, and neglect dyslexia) as has the term 'dysgraphia' to certain acquired writing/spelling disorders. We have varied our labeling, therefore, so as to be consistent with the published nomenclature.

Pure Alexia (Alexia Without Agraphia)

A dramatic example of a reading disorder that occurs without corresponding difficulties in spelling or writing is pure alexia (or alexia without agraphia). Jules Dejerine first described this syndrome over a hundred years ago at the Biological Society in Paris. In this seminal work, Dejerine documented the remarkable case of 'Monsieur C' both behaviorally and anatomically. Monsieur C, a highly educated businessman who had suffered a left hemisphere stroke, showed preserved writing and spoken language skills. He wrote fluently both spontaneously and to dictation but showed severe word blindness; that is, Monsieur C could not even read sentences he himself had written once the memory of the text had faded.

From neuroanatomical and behavioral observations, Dejerine concluded that pure alexia is a deficit caused by a disconnection between vision and language-based areas of the brain, a theory that Geschwind also resurrected in the mid-twentieth century. The disconnection theory Geschwind proposed entails that at least two lesions are necessary to isolate words from visual input, resulting in pure alexia: one in the left occipital lobe and the other in the splenium of the corpus callosum. Although it is a possible explanation for some of the cases reported over the years, current research suggests a more parsimonious neuroanatomical explanation. Most patients with pure alexia show a lesion of a small section of the cortex in the left mid-fusiform gyrus. This region is the only visible damage in at least one documented case of pure alexia. However, the specific role of this brain region and its importance for efficient reading remains a matter of intense debate.

The majority of cases showing alexia without agraphia are not as severely impaired as Monsieur C, who was unable to identify even single letters, treating them as unfamiliar shapes (e.g., he indicated that the letter A looked something like an artist's easel and S like a snake). The majority of pure alexics are able to identify individual letters but are forced to adopt a laborious approach to decipher even short familiar words. As a result, pure alexic readers (or letter-by-letter dyslexics as the syndrome is now often called) show very slow word identification (e.g., more than 2.5 s on average for even short, familiar words). A defining feature of letter-by-letter reading is the so-called word-length effect; that is, a linear relationship between the number of constituent letters and the time taken to read a word. The abnormally large word-length effect in this reading disorder is in striking contrast with the reading profile of normal readers, who show no word-length effect in naming latencies for words of less than seven letters.

The effect of length on performance suggests that patients with this form of reading disorder are limited to deciphering words by analyzing each letter in sequence, so that the longer the word, the slower the reading speed (hence the term letter-by-letter dyslexia). Interestingly, though, this surface feature of the disorder is misleading. Good evidence indicates that it is generally not the number of constituent letters that impacts the reading of many letter-by-letter dyslexics but the extent to which the word contains letters that are perceptually confusable with other letters of the alphabet. This confusability index tends to be greater for longer words, simply because the longer the word, the more letters there are to be identified that are potentially confusable with other letters. It is possible to vary length while holding constant the summed confusability scores of the letters making up the word. When word-confusability scores are controlled in this way, letter-by-letter readers continue to read slowly, but their performance is generally not slower for longer than for shorter words. Thus, reading in this syndrome, despite the modern label for the disorder, does not seem to be invariably confined to a sequential analysis of letter identities. Rather, simultaneous analysis of multiple letters generates perceptual noise that delays recognition, and the degree of perceptual noise is determined by the overall confusability of the letters in a word.

Pure alexia offers a rich window into the visual mechanisms dedicated to word recognition. How specific is the disorder to reading? There are two alternative views on this question. The first attributes the reading disorder to damage of a specialized system for the visual analysis of alphabetic stimuli (i.e., words and letters). This account is based on the hypothesis that visual experience and expertise with words is sufficient to create area(s) of visual cortex responsive uniquely to visual words. Theories postulating that only alphabetic stimuli are impaired in pure alexia are examples of this specific account. The second account proposes that no brain part is uniquely devoted to visual word recognition per se but that some neural mechanisms that determine efficient perception of words are less crucial for other kinds of objects (e.g., faces, animals, man-made three-dimensional objects, etc.). This last account predicts that pure alexia is not a deficit specific to alphabetic material but rather includes an impairment in any task where the necessary visual resources coincide with those needed for reading. For example, one account proposes that visual word recognition requires simultaneous integration of multiple letters and that this integration mechanism is not specific to reading.

All thoroughly tested patients with pure alexia show some form of visual processing impairment not specific to alphabetic stimuli. Hence, the hypothesis of a general visual deficit is actually plausible. But the nature of the perceptual mechanisms, so important for reading yet of much less significance in other visual tasks, remains to be determined.

Attentional Dyslexia

Attentional dyslexia is a rare form of reading disorder that occurs after a brain lesion to the left parietal cortex. The main behavioral symptom is a failure to correctly perceive one item of a given category (e.g., the letter A) when this item is presented simultaneously with other items of the same category (e.g., BDGAH). A typical patient is able to read isolated words or letters with relative ease but his performance declines when

he is asked to identify those same items in a sentence or a letter string – because many items are visible at the same time. These patients also make many errors when they are asked to identify a single digit flanked by other digits but the error rate decreases significantly when the target is a set of dots to be counted. This behavioral observation implies that naming (in this instance, of numbers) is not accountable for the deficit. The main deficit in attentional dyslexia is a lack of control of a filtering mechanism that suppresses visual processing of unattended items in the visual field. Thus, these patients are unable to direct their attentional spotlight to a specific region of interest. As a result, more input than expected falls within the attentional spotlight and the wrong features are integrated together leading to perceptual errors. Within this framework, error rates are larger when the surrounding stimuli are from the same category.

Neglect Dyslexia

Although it is frequently associated with hemispatial neglect and right hemisphere damage, neglect dyslexia can also be observed in isolation. In fact, at least one patient has been known to have a right-side hemispatial neglect following a left hemisphere lesion in conjunction with left-side neglect dyslexia following right hemisphere damage. This exceptional patient undoubtedly proved the existence of a double dissociation between nonverbal neglect and neglect dyslexia. In this form of peripheral dyslexia, reading is impaired because the patients tend to omit or misread the left side of a page or the first letters of letter strings or words of a sentence. At the word level, visual errors made by left neglect dyslexics can be additions of letters at the beginning of words ('love' read as 'glove'), omissions ('cowboy' read as 'boy') or substitutions ('mother' read as 'bother'). In right neglect dyslexia, these errors occur at the end of the word. By itself, the existence of neglect dyslexia suggests that spatial attention plays an important role in normal reading. However, neglect dyslexia is quite variable between patients and different attentional mechanisms have been proposed to explain this variability.

It has been argued that at least three spatial frames of reference are necessary for understanding visual attention in words. The first frame is retinocentric; spatial information is represented by means of a coordinate system centered on the retina. When this form of spatial attention is impaired in neglect dyslexia, omissions and errors tend to be on the letter strings that appear in one hemifield, generally the left in patients with right hemisphere damage. Indeed, a word written on the left side of a sheet will be ignored or misread while it will be read correctly if it is presented on the right, no matter the word's orientation (right to left, left to right, vertically, etc.).

The next frame of reference is a stimulus-based one. When this frame is impaired for reading, words' spatial representations are altered. The left side of the word as a string of letters is misread, regardless of whether the orientation demands a left-to-right or right-to-left perusal. A word such as 'marble' might be read 'cable' if presented from left to right but as 'march' if demanding a right-to-left analysis of letters (i.e., 'elbram'). In contrast to retinocentric impairment, damage to the second frame of reference leads to the same error rates no matter where the letters string is presented in the visual field.

The third and last frame of reference implicates cognitive, higher-level representations of known words, and the processing is no longer at the visual extraction level. When this reference frame is impaired for reading, a word such as 'bible' could be read as 'cable' even if presented from left-to-right or right-to-left. Interestingly, this kind of patient tends to commit similar errors in writing or spelling, thus suggesting that at this level, attention is applied to more central orthographic representations, akin to the reading and spelling errors in central alexias and agraphias.

Central Dyslexias and Dysgraphias

Central dyslexia and dysgraphia are the result of a disruption to the linguistic mechanisms (i.e., orthographic, semantic, or phonological) that mediate reading and spelling, respectively. Neuropsychological studies of brain-damaged patients have contributed substantially to the development of a model specifying the cognitive processes and their interconnections that determine reading and spelling (see Figure 1). The structure of the model – its cognitive architecture – is founded on the assumption that separate routines from print to sound (or in the case of writing, from sound to print) are recruited for familiar and novel words. A familiar word contacts its stored visual form and meaning, and these representations are then available for reading aloud and writing. If a novel word is encountered (or in a task devised for experimental purposes, a pseudoword), no stored form or meaning is available. Reading aloud or writing to dictation is then based on subword graphemic units assigned to corresponding phonemic segments; for example, the written sequence FENT is pronounced

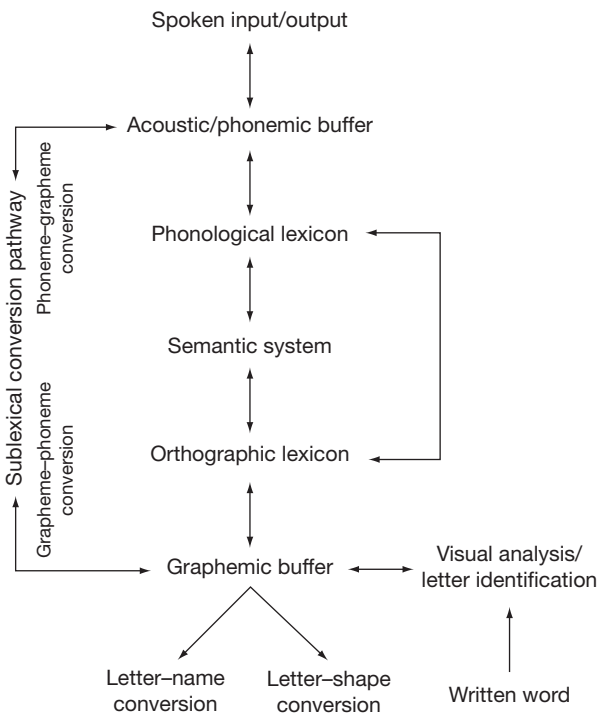

Figure 1 Modular architecture of reading and writing.

by knowing the sound associated with the letter F, and the typical rendering of the segment ENT. The framework offers an account of certain aspects of normal as well as pathological performance but is not without controversy. An alternative connectionist architecture rejects the view of two independent procedures for transcoding familiar and novel words. This rival approach has three sets of simple processing units, and so is referred to as the 'triangle model': grapheme units, representing orthography, phoneme units representing sound, and an array of semantic units for meaning. A spelling pattern generates recurrent activity over the grapheme units which then propagates through the network, resulting in a pronunciation for reading aloud (writing would entail a similar propagation of activity but from phoneme-to-grapheme units).

The triangle model, like its rival, includes two pathways between print and sound; a direct mapping between graphemes and their corresponding pronunciation, and a second pathway from print to sound via semantic units. Nevertheless, there are important differences between connectionist approaches and the more traditional dual-route model depicted in Figure 1, the most basic of which is that the same processing mechanism is applied to the way familiar and unfamiliar words are mapped between spelling and sound. For expository purposes, we will follow the logic of the traditional model to describe central reading and spelling disorders. We will therefore begin our discussion on the central deficits of reading and writing with a short portrayal of the main components of this model.

In order to read, children must learn to assign a pronunciation to spelling units (e.g., the pronunciation of a sequence like INE). For writing, a similar mechanism is required but operating in reverse, to convert a pronunciation into a valid graphemic pattern. Note that the cognitive constraints on spelling-to-sound and sound-to-spelling conversion procedures are rather different. For example, the letter string HAIR has only one possible correct pronunciation but the spoken sequence/hɛər/has at least two corresponding orthographic forms (e.g., HARE, HAIR).

As reading and spelling develop, children learn to access or retrieve familiar words by directly contacting their stored visual forms. The ability to read or spell by using whole words rather than subword units implies three processing components for a word, one for its orthography, the second for meaning, and the last for pronunciation. This lexical–semantic route has some similarity with a dictionary: the orthographic lexicon contains the word's spelling (e.g., A-P-P-L-E), the semantic system containing its definition (e.g., the fleshy usually rounded red, yellow, or green edible fruit) and the phonological lexicon containing its pronunciation (e.g., /ˈa-pəl/). Theoretically speaking, the orthographic lexicon contains representations of all the words that a normal subject has learned to recognize in a single glance or to spell with high efficiency. The semantic system is responsible for storing the meanings of words. It is shared by other language mechanisms such as oral or sign language. The phonological lexicon is a system involved in the retrieval of spoken words in speech production. It plays an essential role in the spoken component of reading.

We will see that each of these processing routes and subsystems can be damaged independently and cause different patterns of dyslexia and dysgraphia.

Phonological Dyslexia and Dysgraphia

In phonological dyslexia and dysgraphia, patients have difficulty in reading and spelling unknown words and pseudowords. Their performance, though, is clearly better or even in the normal range for known words, particularly for high-frequency words. Interestingly, most errors made by these patients are real words that are visually similar to the target word (this kind of error is termed a lexicalization; e.g., the nonsense word SIFE read as 'sift').

The cognitive architecture implies a division between the mechanisms responsible for known words and those responsible for unknown words and pseudowords. The fact that a cerebral lesion can affect reading and writing of pseudowords without affecting known words is consistent with the claim that distinct mechanisms are implied for processing these stimuli. In phonological dyslexia and dysgraphia, the lexical route is preserved, enabling the reading and spelling of familiar words, while the nonlexical route is impaired, affecting performance on spelling patterns that have no representation as real words.

Surface Dyslexia and Dysgraphia

At this point in our description of central deficits, the assumption that a distinction exists between a lexical and a sublexical pathway is not the only plausible interpretation of phonological dyslexia and dysgraphia. It is possible that pseudowords are simply harder to read and write than known words and that neurological damage only exacerbates this difference. The fact that normal subjects are slower and less accurate when reading and writing pseudowords versus real words is consistent with this possibility.

The finding of patients with the opposite behavioral profile – that is a deficit for words but no deficit for pseudowords – would eliminate this alternative possibility, however. Marshall and Newcomb were the first to describe such a patient in 1973. They documented what we now call surface dyslexia, that is, an acquired reading deficit in which the reader has difficulty reading 'exception' words (i.e., words with an irregular spelling-to-sound mapping; e.g., colonel) but makes fewer errors when asked to read regular words (e.g., fairy) and pseudowords (e.g., peratine). The initial cases described were less clear than expected given the assumption of a categorical distinction between whole-word and subword procedures for translating spelling to sound. Surface dyslexics often produced errors that indicated impairment of both procedures. This lead to some doubt about the validity of the distinction, until a number of unambiguous cases were documented. From the perspective of the model, the reading impairment in surface dyslexia results from damage to the lexical route. Since this processing route is unavailable, surface dyslexic patients attempt to read and spell by the sublexical (i.e., grapheme-to-phoneme and phoneme-to-grapheme conversion) processing systems. Put differently, surface dyslexic patients often treat irregular words as if they were regular words or pseudowords. For example, they will pronounce COLONEL as /ˈkoʊləneɪl/ instead of /ˈkɜrnl/. Such errors are called regularizations and constitute the diagnostic pattern of surface alexia. Importantly,

these errors are more likely to occur for less common exception words (e.g., MOTHER might be read and spelled correctly but not LEOPARD).

Similar symptoms have been described for writing and spelling. In this case, patients make a large quantity of spelling errors for irregular words all the while correctly spelling regular words and pseudowords. The error rate in general is actually higher for this form of dysgraphia than for the corresponding surface dyslexia. In writing, the number of words with unpredictable phoneme-to-grapheme correspondences is quite high (e.g., the pronunciation 'boat' can be spelled in at least two ways, if we simply apply general principles of spelling-to-sound translation, including BOTE). Of course, some words, for example, DOG or CAT, have highly predictable sound-to-spelling correspondences. These words can be correctly spelled using the nonlexical conversion module. But many words have an unpredictable orthography, and the lexical route is needed for these to be correctly spelled.

It seems reasonable that surface dysgraphia should be seen together with surface dyslexia; both reading and writing errors can be attributed to a failure to contact a central representation of the word's orthographic form, and responses are based on more generic procedures that yield regularization errors. In very rare cases, though, surface dysgraphia occurs selectively (i.e., reading via word-specific procedures is demonstrably intact) or in association with another form of central dyslexia (e.g., phonological dyslexia). What do these unusual dissociations imply about the orthographic representations accessed for reading and spelling? One possibility is that different word-specific representations are available for input (reading) and output (spelling). The alternative is that the representations are indeed unitary but that distinct access and retrieval procedures exist that can be separately damaged to yield the dissociations seen in rare cases. The fact that such dissociations are so infrequent, though, indicates that the neuroanatomical substrates underlying any functional distinction between word-specific representations involved in reading and spelling must be so organized that they are mostly affected together.

Surface dyslexic and dysgraphic errors are the result of a failure to retrieve a word's pronunciation based on its stored orthographic form, and an overreliance on more general correspondences between spelling and sound units. We turn now to a very different phenomenon, involving reading or spelling impairment that ensues when the meaning of the word remains the only point of translation between its visual form and pronunciation, because all other routes have been destroyed.

Deep Dyslexia and Dysgraphia

In any cognitive neuropsychological task, the occurrence of semantic errors is probably one of the most spectacular kinds of error. In word reading, semantic errors are the main diagnostic behavior of deep dyslexic patients. For example, the typical patient could produce 'freedom' when confronted with the word 'LIBERTY' or 'knight' in response to 'CASTLE.' The first case of deep dyslexia was described in 1931 but the burgeoning interest in this reading disorder began in the late 1960s with the report of Marshall and Newcombe. In writing, the first case was described by one of us (D.B.) in the early 1980s. The limits on the performance of patients with deep dyslexia or dysgraphia are rather similar to the constraints suffered by patients with a phonological alexia or agraphia. In both types of disorder, performance for nonsense or unfamiliar words is affected while words yield better accuracy. However, deep dyslexic or dysgraphic patients are unable to cope with even a short nonsense word made up of a single syllable, thus suggesting a total destruction of the sublexical pathway. Of course, the syndrome is also associated with semantic errors produced by familiar words.

A comparatively large amount of research has been conducted on deep dyslexia. The reading pattern is probably the most complex and dramatic of all acquired reading impairments. Although semantic errors are the most frequent and diagnostic of errors in deep dyslexia, this kind of mistake is only one of the many characteristics of the syndrome. The most prominent features are (1) semantic errors, (2) visual errors (e.g., SYMPHONY read as 'sympathy'), (3) function-word substitutions (e.g., BUT read as 'and'), (4) derivational errors (e.g., MARRIAGE is read 'married'), (5) a complete failure to read pseudowords, (6) some classes of words found to be harder than others in reading aloud (function words > (i.e., harder than) verbs > adjectives > nouns), (7) strong concreteness (i.e., imageability) effect (TABLE easier than FATE).

To explain deep dyslexia, the dual-route model needs to postulate a number of deficits caused by independent damage to different functional components. Damage to the sublexical print-to-sound translation pathway is needed to explain the striking impairment in pseudoword reading. The presence of semantic errors as well as the concreteness effect suggests impairment to the semantic system itself. Destruction of the direct pathway between the stored visual form of the word and its pronunciation is also necessary since this procedure is clearly not available in deep dyslexia. If it was, the semantic errors would not occur because the pronunciation of the word could be retrieved without requiring that its meaning be contacted. Finally, visual errors suggest damage to the structural analysis of words; a possible source of these errors would be faulty processing within the orthographic lexicon. This unlikely coincidence of deficits observed in most deep dyslexic has led some researchers to argue that deep dyslexia is of no relevance for comprehension of normal reading mechanisms. Indeed, these researchers propose that deep dyslexics' left hemisphere lesion is so extensive that it is no longer available for reading. In this extreme case, reading would be mediated by a weakly literate right hemisphere. The attractiveness of this hypothesis comes from the reading similarities observed between deep dyslexics and patients with an isolated right hemisphere. However, the idea seems less plausible given evidence from a single case demonstrating many deep dyslexic characteristics, but whose reading was abolished after a second left hemisphere stroke.

The question remains, then. Why does the complex of symptoms characteristic of deep dyslexia occur, assuming that the pattern reflects the performance of a partially damaged left hemisphere reading system and not the normal reading capabilities of a linguistically restricted right hemisphere. Some progress on the issue has been made by researchers

constructing and analyzing connectionist models of reading that map the meaning of a visual word to its pronunciation. The relevant architecture consists of a primary layer of grapheme units representing the visual form of words, connected via intermediate units to 'sememe' units. The only function of intermediate units is to learn the pattern of associations that map specific letter patterns to a particular meaning, while sememe units represent the meaning of a word in terms of a limited set of semantic features like 'can be eaten,' or 'made of wood.' Sememe units connect to phoneme units via an additional set of intermediate mapping units.

This type of architecture has trouble learning to associate visually similar words with distinct semantic representations; however, the inclusion of a layer of 'clean-up' units that interacts with the sememe layer dramatically improves the mapping between the orthographic form of words and its meaning, by minimizing the confusion between visually similar words. Of great interest is the fact that damage to a percentage of clean-up units (by randomly altering a subset of weights) yields a number of the symptoms of deep dyslexia: visual errors occur as well as semantic errors, because the clean-up units no longer can differentiate between the patterns of activation generated by visually or semantically similar words. In addition, both deep dyslexic cases and the damaged connectionist model produce more complex errors, involving a semantic error driven by a visual substitution (SYMPATHY read as 'orchestra,' via the visual confusion 'symphony'). A concrete word like BOOK resists damage to the network more than an abstract word like FATE, because the former has many more semantic features than the latter. The clean-up units have greater redundancy to work with and so converge on the right meaning even after significant damage. Finally, because correct recognition of concrete words depends heavily on clean-up units, severe enough damage to this layer reproduces a striking reverse-concreteness effect also observed in one rare case of deep dyslexia; abstract words are read better than concrete words. Despite the undeniable value of these computational accounts, however, this approach has not yet produced a unified, coherent interpretation of the full symptom complex.

Deep dysgraphia is of considerable theoretical interest because the syndrome may sometimes occur in the absence of deep dyslexia, though the writing disturbance is invariably seen in association with some form of reading and language disorder. The dissociation between deep dyslexia and dysgraphia is evidence that procedures affording access to meaning from print may be partially distinct from procedures that operate in reverse to retrieve the spelled from of a word from its meaning. In addition, cases of deep dysgraphia show errors in their written performance indicating a failure to maintain letter identities in an output buffer holding active the graphemic representation of a word for sequential output. We discuss the reason for this surprising association of seemingly disparate effects when we consider writing disorders linked to impairment of the graphemic buffer.

Semantic Dyslexia and Dysgraphia

One possible outcome of severe damage to semantic representations is surface dyslexia, that is, a deficit in reading irregularly spelled words. It seems reasonable to expect that words will be treated as nonsense words given severe enough damage to semantic representations. If the sublexical conversion pathway remains intact, orthographically irregular words will be regularized. Surprisingly, some patients – despite a severe semantic impairment – retain an ability to read and even spell both regular and irregular words. Semantic dyslexic cases suffer from Alzheimer's dementia or semantic dementia. They are able to read any kind of letter string (i.e., regular and irregular words as well as pseudowords), but they show no understanding of the words they can produce correctly. This kind of patient demonstrates that three processing routines are available to access pronunciation from print; the spelling-to-sound conversion procedure, the lexical–semantic pathway and a lexical nonsemantic pathway directly mapping word-specific orthographic forms to a pronunciation. It is this route that is used in semantic dyslexia to read irregular words correctly. Notably though, semantic impairment often does lead to surface dyslexia rather than semantic dyslexia. Why the different outcome in different cases? An interesting answer is that there are substantial individual differences in the utilization of semantic representations for reading. Some readers may need to rely more on semantic access to read irregular words whereas other readers are less dependent on word meaning.

Graphemic Buffer Impairment

As depicted on Figure 1, the spelled form of words is produced by accessing a graphemic buffer, a component that receives information from both the orthographic lexicon and the phoneme–grapheme conversion mechanism. This component maintains abstract letter information (i.e., case invariant information) while peripheral conversion mechanisms operate sequentially on each grapheme to produce the spelled form (e.g., letter–name conversion for oral spelling, letter–shape conversion for writing, letter–motor scheme for typing).

How should performance be affected if the graphemic buffer were impaired, assuming that the damage is not severe enough to preclude any form of output? The buffer holds the final spelling of both familiar words as well as graphemic sequences generated by spelling-to-sound conversion. Performance should therefore be unaffected by word-specific variables like concreteness, familiarity, and grammatical class. If the capacity of the buffer to maintain letter identities has been reduced, longer words should yield more impairment than shorter words. In addition, if activation in the buffer has been compromised so as to generate interference between letter identities, spelling errors should be more frequent in the middle than the end of words, because terminal letters have fewer adjacent letters to compete with their activation. This profile has been documented in a substantial number of dysgraphic cases. Their errors include doubling applied to the wrong letters (e.g., STREET spelled as STRETT; consistent with the idea that the doubled status of the letters is represented separately from the letters themselves), ordering errors (transpositions), substitutions, and omissions. A successful computational account of this dysgraphic subtype has been implemented (a 'competitive queuing model') based on principles derived from a class of models that deal with the

control of sequential output. The model includes the idea that response elements organized for sequential output are selected by means of an activation gradient; the more active an element the sooner it is produced. In addition, competition occurs between response elements. Random noise added to the sequence generation process yields the key features of a graphemic buffer dysgraphia.

A surprising variant of the syndrome shows many of the cardinal signs indicative of a damaged buffer but in addition, elements of deep dysgraphia, including word class effects (like concreteness) and semantic errors. Nonsense words are spelled very poorly. Like the more typical instances of the syndrome, letter deletions, substitutions, and transpositions occur and spelling is worse for longer than shorter words. However, errors increase monotonically from the beginning to the end of the word (whereas in the more typical case, as we have seen, lower error rates occur at the start and end of words). This variant can be understood if the output of the semantic pathway yields a degraded representation of letters in the graphemic buffer. Subthreshold activation of letter identities results in numerous errors, including premature termination of the word if the very low signal from the last few letters is taken to indicate that the end of the word has been reached.

It was originally assumed that the graphemic buffer's role was restricted to the writing domain. However, the buffer may also be involved in reading, maintaining the level of activation generated by letters in the word. Accordingly, patients with a lesion to the graphemic buffer frequently have difficulty reading items comprising many letters, especially when pseudowords must be read and the orthographic lexicon cannot contribute to the response. Normal readers supply converging evidence for a short-term visual memory needed to derive the pronunciation of unfamiliar words. In this framework, the only strictly peripheral agraphias are those coming from lesions to the letter–shape and letter–name conversion mechanisms.

More Peripheral Agraphias

Impaired writing can occur even though the spelled form of the word has been correctly retrieved. In this type of agraphia, the word TABLE would be spelled aloud as 'tee, ay, bee, ell, eeh,' yet errors occur when the letters are produced in written form. The spelling of the word is clearly available, therefore, at least in a form suitable for naming (but also for other tasks; e.g., constructing the spelled word using block letters), but their shapes are not correctly translated into movements. If the writing impairment is due to a failure in motor processes that govern neuromuscular execution and control, written letters are distorted often to the point of being unrecognizable, and errors include misplaced as well as incorrectly formed strokes of the pen.

More surprisingly, it is possible to observe instances of peripheral agraphia where the dominant error consists of well-formed letter shapes; TABLE would be written as F-A-P-L-E. The question of interest is whether such errors are visually related to the target or whether some other relationship exists, based on the sequence and placement of the movements needed to construct the letter. A question of additional import is whether the action representation at this level of organization in the writing mechanism concerns movements of the hand or more generally, to any action that is intended to construct letter forms. The answer to the second question is straightforward; the representation affected is sufficiently abstract that impairment is seen even when letters are produced by tracing the shape with a foot. The first question is more difficult. Letters that require similar strokes for their production tend also to be visually similar; how then can we distinguish visually based from action-based errors? A reasonable way of proceeding is to invoke two empirically driven methods to arrive at separate metrics of visual and motor similarity between letters. For example, visual confusability can be assessed by examining the errors of normal readers under restricted viewing conditions; the higher the probability a letter is confused with another, the more they are perceptually similar. Motor similarity can be operationally defined by constructing a reasonable taxonomy involving, say, number of strokes, shape (line vs. curve), direction (up, down, clockwise, counterclockwise), and so on. In this way, it is possible to isolate letter pairs that are more confusable with respect to an underlying metric of motor than visual similarity. An analysis of errors using this methodological approach has disclosed that in some cases of peripheral agraphia, errors are based on stroke similarity between letters rather than visual similarity.

Peripheral agraphias offer the possibility of valuable insights into the processes that govern the production of letters, as the previous example illustrates. Additional evidence from individual cases reveals a degree of categorical organization imposed on the processes that translate letter identities into movement. Impairment can affect the formation of letters while numbers are unaffected; the letter O may be incorrectly written, in other words, but not the number zero! Other forms of peripheral dysgraphia disclose mechanisms that concern the distinction between upper and lower case forms of the same letter (allographs); patients may have severe difficulty in producing uppercase printed letters for example, but not lowercase cursive script. Finally, dysgraphia may involve a disconnection between hemispheres, so that one hand no longer has access to the representations that govern the production of letter shapes.

See also: Dyslexia; Motor Control; Neuroimaging of Dementia; Psychology of Reading; Reading and Phonological Processing; Semantic Memory; Spatial Perception.

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