REVIEW

Dysgraphia in dementia

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Abstract

This paper reviews the spelling and writing deficits associated with the major forms of dementia. In dementia of the Alzheimer's type (DAT), the finding of surface dysgraphia is typical, although not universal, and with disease progression non-phonologically plausible errors often increase; additional difficulties with handwriting are common. Surface dysgraphia is a predictable feature in semantic dementia, but in contrast to DAT, the mechanics of handwriting are usually preserved. In patients with posterior cortical atrophy [including some (atypical) DAT patients], spatial dysgraphia is an early symptom. Spelling and writing disorders have been documented, although not thoroughly studied, in other forms of dementia (e.g. vascular, frontal, dementia with Lewy bodies, etc.). Research on dysgraphia in dementias associated with movement disorders (e.g. cortico-basal degeneration, progressive supranuclear palsy, Huntington's disease, etc.) has focused mainly on problems with writing, which is often poorly formed, but spelling impairments have also been observed. The precise characteristics and prevalence of spelling disorders in these diseases are, however, unknown as there has been little systematic study. Additional investigations, which assess both central (linguistic) and peripheral aspects of writing, and which include a longitudinal component, will help to elucidate the nature and progression of dysgraphia in different types of dementia.

Introduction

Dysgraphia is a disorder in writing or spelling, and is a common symptom in a variety of dementias. The first description of a dysgraphic impairment in a dementia patient was provided by Alois Alzheimer (1907; translated in 1977), who observed omissions and duplications of written syllables. Since that time, the neuropsychological impairments in the dementias have been extensively investigated, but the dysgraphic deficits have received relatively little attention.

Historical overview of research on dysgraphia

Until the 1980s, research on disorders of spelling and writing almost exclusively involved patients with stable brain lesions, rather than the progressive atrophy seen in dementia. Studies were mainly focused on describing syndromes and attempting to localize them anatomically. The earliest report of writing impairment caused by cortical lesions was published in 1856 by Marcé [cited in Hécaen *et al*. (1963)]. Jackson (1864, 1866) also provided early descriptions of dysgraphic patients and noted that writing and speech can be damaged separately. The term 'agraphia' was coined apparently independently by both Benedikt [1865; cited in Leischner (1969)] and Ogle (1867) to refer to acquired disorders of writing, and has

subsequently been used interchangeably with the term 'dysgraphia' (McCarthy and Warrington, 1990). Ogle distinguished two types of dysgraphia, which affect either linguistic or motor aspects of writing, and which arise from cortical lesions. In 'amnemonic agraphia', letters are well formed, but incorrect letters or words may be produced, while in 'atactic agraphia' letters are poorly formed and often illegible. This general distinction between linguistic and motor factors in writing is now universally accepted. Using more recent terminology [coined by Ellis (1982)], central dysgraphias arise from a linguistic problem affecting the spelling system, while peripheral dysgraphias reflect a modality-specific disorder affecting writing, oral spelling or typing.

The rise of the information-processing approach in the 1970s led to a change in the study of dysgraphia. Prior to that time, classifications of dysgraphia revolved around neuroanatomical locations or aphasic categories. Subsequently, researchers following the information-processing approach sought to specify the underlying processes involved in writing and spelling, and to identify those that may be disturbed in dysgraphic patients. Classifications of dysgraphia came to be based upon hypothesized functional loci of damage in models of spelling and writing. These models will now be described.

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Models of spelling

The dual-route model

For nearly two decades, the dual-route model has dominated the field of spelling research, and has motivated much progress. The model was based upon a pre-existing model of reading, originally developed by Marshall and Newcombe (1973), and adapted some years later to explain processes in spelling (Morton, 1980; Newcombe and Marshall, 1980; Ellis, 1982). Since then, the model has been elaborated somewhat, but in essence has changed little. There are thought to be two basic procedures involved in spelling (Ellis, 1982; Margolin, 1984; Goodman and Caramazza, 1986; Patterson, 1986, 1988; Baxter and Warrington, 1987; Ellis and Young, 1988; Shallice, 1988; McCarthy and Warrington, 1990; Hillis and Caramazza, 1991; Barry, 1994; Zesiger and de Partz, 1997): (i) accessing stored orthographic representations of specific known words, generally known as the lexical or lexical–semantic procedure; and (ii) deriving the spelling of any word (particularly unfamiliar or nonsense words) on the basis of sound-to-spelling correspondence rules, referred to as the non-lexical, assembled or phonological procedure. There are different variants of the dual-route model, and a third route is often included; this involves accessing orthographic representations for output directly from phonological input representations, without involving the semantic system, and is referred to as the lexical non-semantic, or lexical phonological, route.

Once the spelling of a word has been accessed or derived, the relevant graphemes (i.e. abstract representations for letters) are held in the graphemic buffer (also known as the orthographic buffer), while more peripheral output processes are executed. This stage is common to all three spelling routes, and damage here would lead to a similar spelling impairment in all output modalities.

Spelling by analogy

Campbell (1983) noted that the two routes in the dual-route model are not clearly distinguishable, and suggested that lexical and assembled spelling are accomplished by the same mechanism. She reasoned that spelling of non-words may be accomplished by utilizing the spelling of similar sounding words in an 'analogical lexically based parsing system'. This theory, which has subsequently become known as 'spelling by analogy', is notable because it was, until recently, the only significant alternative to the dual-route model, but it has been criticized on a number of grounds. For example, Shallice (1988) noted (as did Campbell) that if non-word spelling is accomplished by lexical analogy, then damage to the orthographic lexicon should lead to a non-word spelling impairment because the number of words on which analogies could be based would be reduced; but in contrast with this prediction, lexical and non-word spelling impairments are not always associated. It seems, however, that Campbell's general idea that non-word spelling is accomplished on the basis of knowledge about the spelling of words is again gaining favour because of the recent success of implemented models of spelling, in which both lexical and non-word spelling are accomplished by one mechanism (see below).

Connectionist models of spelling

Connectionist models are a relatively recent alternative to symbolic information-processing models, such as the dual-route model. They are composed of neuron-like processing units with weighted connections between the units; knowledge is represented as graded patterns of activation across the units. Connectionist models are often implemented as computer simulations, and can learn to associate input and output patterns without being given explicit rules; 'learning' entails repeated exposure to an association between patterns of activation across different sets of units, which leads the model gradually to change the weights on the connections (in accordance with the particular learning algorithm built into the model). In this way, multiple associations can be learned by the same set of connections. Models can be damaged by removing units or connections, or by adding random noise to the weights, thereby allowing researchers to simulate the behaviour of patients with brain damage.

A small number of modellers have attempted to simulate the generation of the spelling of words (Brown *et al.*, 1991; Loosemore *et al.*, 1991; Brown and Loosemore, 1994; Bullinaria, 1994, 1997; Olson and Caramazza, 1994). Perhaps the most significant result from these simulations is that the networks were able to learn to spell both the regular and exception words in the training set, and also to generalize to the spelling of novel words outside the training corpus, using only one mechanism. This was done without either a lexicon or explicit phoneme–grapheme conversion rules, and is in contradiction with the dual-route model, which suggests that regular and exception words are spelled via separate mechanisms. A version of the dual-route model has also recently been implemented (Houghton and Zorzi, 1998).

Models of peripheral output processes in writing and oral spelling

As noted above, once the spelling of a word has been accessed or derived, abstract representations of the letters in the word are temporarily stored in the graphemic buffer while peripheral output processes are executed. For writing, the first stage beyond the graphemic buffer is the allograph level, where information about physical letter shapes (including upper and lower case, script and print, etc.) is specified (Ellis, 1982); allographs are sometimes referred to as physical letter codes (Margolin, 1984; Margolin and Goodman-Schulman, 1992). Patients with impairment at the allographic level have difficulty with recalling letter shapes, but once a letter has been recalled or made available to copy, they fluently produce well-formed letters. The next stage in the production of written output involves accessing graphic motor

patterns that specify the strokes required to form each letter (Ellis, 1982; Margolin, 1984; Margolin and Goodman-Schulman, 1992). Production of poorly formed letters (in the absence of general motor or praxic deficits) is usually attributed to impairment in selection or execution of graphic motor patterns. For oral spelling, the first stage beyond the graphemic buffer involves accessing the names of letters, and the next involves execution of articulatory motor programmes (Margolin and Goodman-Schulman, 1992).

Dysgraphia in dementia

Dementia can occur in a variety of disease processes which impair intellectual functioning, and until recently was characterized as a global or generalized intellectual impairment. Advances in diagnosis have demonstrated, however, that in the early stages the diseases that cause dementia tend to produce distinct and often focal neuropsychological deficits which reflect the disease topography (Hodges, 2000). A more current definition of dementia describes it as an acquired, often progressive decline in memory, plus one or more additional aspects of cognitive functioning, including language, visuospatial or perceptual skills, praxis, abstract thinking and judgement, and personality and social behaviour (American Psychiatric Association, 1994; Hart and Semple, 1994; Hodges, 1994).

The dysgraphic deficits associated with different types of dementia will now be outlined. The interpretation of these deficits in current models of spelling and writing will also be explicated.

Alzheimer's disease (AD)

Although Alzheimer's initial report of a patient with this disease was published in 1907, the disease did not become a focus of extensive study until the 1980s, and work since then has been prolific. The first studies which examined the dysgraphic deficits in dementia of the Alzheimer's type (DAT) did so as a small part of test batteries which assessed multiple cognitive functions, or addressed clinical issues, and the assessments of spelling or writing were, therefore, somewhat superficial. These early studies established that dysgraphia is a common symptom in DAT (Folstein and Breitner, 1981; Appell et al., 1982; Seltzer and Sherwin, 1983; Breitner and Folstein, 1984; Cummings et al., 1985, 1988; Kertesz et al., 1986; Whitworth and Larson, 1989), and that this impairment is often more severe than the spoken language impairments (Appell et al., 1982; Kertesz et al., 1986).

Perhaps the earliest paper that focused specifically on written output in DAT (Behrendt, 1984) aimed to provide information for document examiners, who deal with wills, codicils, etc. Although the data set presented was rather meagre, results demonstrated that handwriting may progressively deteriorate, and that spelling errors may be observed.

Subsequent work has demonstrated that DAT is associated with breakdown at multiple levels in the writing process. These will be outlined in turn.

Narrative writing in DAT. Assessments of narrative writing, in which patients are asked to write a sentence (Folstein and Breitner, 1981; Kumar and Giacobini, 1990; LaBarge et al., 1992; Kemper et al., 1993) or a description of a picture (Horner et al., 1988; Neils et al., 1989; Henderson et al., 1992; Croisile et al., 1995, 1996; Carey et al., 1999), have shown a range of impairments. The written output produced by DAT patients was found to be shorter than that of controls (Neils et al., 1989; Henderson et al., 1992; Kemper et al., 1993; Croisile et al., 1995, 1996), and to contain less information (Henderson et al., 1992; Kemper et al., 1993; Croisile et al., 1995, 1996). In addition, errors in vocabulary (e.g. semantic substitutions or neologisms) (Horner et al., 1988; Neils et al., 1989; Henderson et al., 1992; LaBarge et al., 1992; Croisile et al., 1996) and syntax (Horner et al., 1988; Croisile et al., 1996) may be prominent, even though DAT patients tend to use simpler grammatical constructions than controls (e.g. fewer subordinate or embedded clauses) (Kemper et al., 1993; Croisile et al., 1996). The vocabulary errors and reduced information content are hypothesized to be related to the semantic impairment that is typical in DAT (Neils et al., 1989; Kemper et al., 1993). Intrusion of incorrect or irrelevant information marred the output of some DAT patients (Horner et al., 1988; Croisile et al., 1996), as did perseveration on previously used words, phrases or ideas (Horner et al., 1988). Some studies reported that a minority of patients produced poorly formed letters, and/or spatially disordered output (Horner et al., 1988; LaBarge et al., 1992; Croisile et al., 1996). Taken together, these results indicate that the narrative writing of DAT patients shows many types of deficit. This is not surprising, given that the task requires the complex integration of multiple cognitive functions.

Central or linguistic dysgraphia in DAT. Many of the studies involving narrative writing demonstrated that DAT patients produce more spelling errors than controls (Horner et al., 1988; Neils et al., 1989; Henderson et al., 1992; LaBarge et al., 1992), but greater progress in understanding the nature of the spelling impairment has been achieved using tasks in which patients are asked to write or spell orally single words to dictation. This type of task enables researchers to manipulate linguistic variables that are pertinent to spelling. As will be outlined below, there is some inconsistency across studies, but the finding of surface dysgraphia is the most common. This type of spelling impairment (also known as lexical agraphia) is characterized by impaired spelling of words with exceptional or unpredictable sound-to-spelling correspondences (e.g. cough, yacht), with a tendency to produce phonologically plausible errors (e.g. tomb → TOOM, crane → CRAIN). Spelling of regular words or non-words is better preserved, as phoneme-to-grapheme conversion skills are typically unaffected. This pattern of performance finds a ready explanation in the dual-route model of spelling: damage to the lexical spelling route leads to reliance upon assembled spelling. Most studies of patients with surface dysgraphia

have focused on patients with stable brain lesions resulting from stroke or head injury, but this syndrome was first documented in DAT patients by Rapcsak *et al.* (1989). Results showed normal spelling of both regular and non-words, but the patients were impaired at spelling exception words, and tended to produce phonologically plausible errors. The researchers noted that the surface dysgraphia observed in the DAT patients was 'clinically indistinguishable' from that observed in patients with focal lesions.

Further studies (Platel *et al.*, 1993; Croisile *et al.*, 1995; Hillis *et al.*, 1996) have confirmed that patients with mild dementia tend to produce phonologically plausible spelling errors, and have also shown that as the disease progresses patients make increasing numbers of non-phonologically plausible errors. Thus, patients are initially better at spelling regular than exception words (i.e. they show a regularity effect), but as the non-phonologically plausible errors increase, spelling of regular words and non-words becomes affected. Similar findings were documented in a longitudinal study of two patients with progressive left temporal atrophy in association with presumed Pick's disease (Graham *et al.*, 1997). On the dual-route model of spelling (described above), these results would be interpreted as an initial impairment in the lexical spelling route, followed by an additional problem in the assembled route. Interestingly, this pattern of performance was observed in an implemented model of spelling (Olson and Caramazza, 1994) in which familiar and novel words are spelled by the same procedure (unlike in the dual-route model); mild damage to the network led to phonologically plausible errors, while more severe damage led to a greater number of errors, many of which were non-phonologically plausible.

The correlates of the non-phonologically plausible spelling errors have been examined. Béland *et al.*'s (1999) detailed single-case study of a DAT patient showed that greater phonological complexity in the stimuli led to more non-phonologically plausible errors, suggesting that these errors arise from a phonological impairment; for the purpose of the study, stimuli deemed to be phonologically complex incorporated a syllabic context that is forbidden in at least one world language, while phonologically simple stimuli comprised onset-rime syllables. In contrast with Béland *et al.*, Neils *et al.* (1995a) found that the production of non-phonologically plausible errors was correlated with impairment on attentional tasks such as letter cancellation and digit span, and suggested that impairment in attentional processing has a detrimental effect on the functioning of the graphemic buffer. Their finding that spelling was adversely affected by increased word length is consistent with this hypothesis, since length effects are expected in graphemic buffer impairment (Caramazza *et al.*, 1987).

Some studies have tried but failed to find effects of regularity or lexicality. The DAT patients studied by Neils and Roeltgen (1994) and Aarsland *et al.* (1996) were equally impaired on spelling regular and exception words and non-words. The discrepancy with the results outlined above may

be attributable to individual differences in patients, in stage of disease (in both studies patients were in the early stages of their illness), or in the sensitivity of the spelling tests used.

Glosser *et al.* (1999a,b) found that although the spelling scores of DAT patients were depressed relative to controls, the overall pattern of performance in the two groups was similar. The exception to this was that the patients showed a 'slightly', although significantly, larger effect of regularity. The similarity in the spelling performance of the two groups led the authors to conclude that the patients' impairment was not caused by a deficit specific to orthographic processing. This claim was supported by noting that in the literature, deficits outside the orthographic system have been found to correlate with spelling performance, including attentional, visuospatial, graphomotor and apraxic problems. One difficulty with this interpretation is that although these deficits can disrupt spelling and/or writing, they would not be expected to lead to a larger effect of regularity in patients than controls.

One final study of spelling in DAT warrants mention. Penniello *et al.* (1995) combined a behavioural study of writing regular words, exception words and non-words to dictation with PET measurements of glucose metabolism. In line with the cognitive findings described above, amongst the DAT patients whose spelling was impaired, the most common pattern was one of disrupted spelling of exception words, suggesting surface dysgraphia. The neuroanatomical results indicated selective involvement of two left-hemisphere regions, the supramarginal gyrus and the angular gyrus, in phonological and lexical processes of spelling, respectively.

It is clear from this brief review that there is no single predictable pattern of spelling ability or impairment associated with DAT. It does seem, however, that once the disease has progressed beyond the early stages, spelling impairment is inevitable. The most often reported pattern is one of surface dysgraphia in the mild stages of disease, followed by an increase in non-phonologically plausible errors as the disease progresses.

Peripheral dysgraphia in DAT. Although most research on dysgraphia in DAT has focused on the central spelling deficits, there is also evidence of peripheral dysgraphia. For example, production of poorly formed or illegible letters has been reported by several researchers (Behrendt, 1984; Horner *et al.*, 1988; LaBarge *et al.*, 1992; Platel *et al.*, 1993; Neils *et al.*, 1995b; Piras *et al.*, 1998). Impaired letter production is usually a relatively late feature, emerging only when the disease has reached a moderate stage (Platel *et al.*, 1993), and eventually rendering some patients unable to write (Rapcsak *et al.*, 1989; Platel *et al.*, 1993).

Neils *et al.* (1995b) did not, however, find a systematic relationship between dementia severity and 'graphomotor impairment'. Moreover, the qualitative description provided by Piras *et al.* (1998) suggested that problems with handwriting appear early in the course of DAT. Similarly, Neils-Strunjas *et al.* (1998) reported a case study of a DAT patient whose peripheral dysgraphia was apparent even when his

dementia was mild; he produced frequent letter formation errors, and in addition tended to perseverate on strokes and letters, a finding which is not commonly reported in DAT. Although no imaging or pathological data were reported, the authors speculated (on the basis of relevant literature) that the writing deficit resulted from right parietal dysfunction. This seems plausible given that the pattern of the dysgraphia in this patient is consistent with that seen in posterior cortical atrophy (see below); this type of atrophy can be caused by AD, although the associated dementia (and dysgraphia) differs somewhat from that in typical DAT (Kiyosawa *et al.*, 1989; Berthier *et al.*, 1991; Ross *et al.*, 1996).

A peripheral writing deficit involving poorly formed letters, such as that observed in typical DAT, is likely to arise from functional impairment at the level of graphic motor patterns. Hughes *et al.*'s (1997) results are consistent with this, and indicate that DAT patients may typically have an additional deficit at the allographic level. These authors tested DAT patients with either minimal or mild dementia on copying (e.g. b → b, F → F) and cross-case transcription (e.g. b → B, F → f) of single letters. The minimally impaired subgroup showed normal performance on these tasks. In contrast, the more impaired subgroup showed a deficit on cross-case transcription, particularly when responding in lower case, as well as a milder impairment on copying of lower (but not upper) case letters. The selective impairment in producing lower case letters is difficult to interpret; it could arise from differential difficulty (perhaps lower case letters require more complex motor planning or are less visually distinctive than upper case letters), but this seems unlikely to be the (entire) explanation because, across patients, case doubly dissociates [see, for example, Patterson and Wing, 1989; Kartsounis, 1992; Graham *et al.*, 1997). The disproportionate deficit on transcription would result from a breakdown at the allographic level, where information about the shapes of letters is accessed (Ellis, 1982; Margolin, 1984; Margolin and Goodman-Schulman, 1992). An additional deficit, at the level of graphic motor patterns, must be hypothesized to explain the impairment on letter copying.

Frontotemporal dementia

Patients with progressive frontal and/or temporal lobe atrophy were first reported a century ago by Arnold Pick [1892; cited in Hodges (1993)], but the term 'frontotemporal dementia' was adopted only recently (Lund and Manchester Groups, 1994). There are three prototypic variants of frontotemporal dementia, which produce distinct neurobehavioural syndromes (Neary *et al.*, 1998): semantic dementia (SD), non-fluent progressive aphasia (NFPA), and the frontal variant (dementia of the frontal type, DFT). A further variant, in which relatively pure progressive dysgraphia was associated with left temporal lobe atrophy, has also been reported. The dysgraphia associated with these variants will be discussed in turn.

Semantic dementia. Spelling impairment is a prevalent feature in SD. It has often been noted that these patients tend to produce phonologically plausible spelling errors on words with unpredictable or exceptional spellings (Baxter and Warrington, 1987; Snowden *et al.*, 1989, 1992, 1994, 1996a,b; Diesfeldt, 1992, 1993; Patterson and Hodges, 1992; Parkin, 1993; Hodges *et al.*, 1995, 1998; De Bleser *et al.*, 1996; Hodges and Patterson, 1996; Kertesz and Munoz, 1997; Kertesz *et al.*, 1998; Schwarz *et al.*, 1998). Preserved spelling has also been reported, but rarely (Lauro-Grotto *et al.*, 1997; Schwarz *et al.*, 1998). Handwriting is generally thought to be unimpaired (Warrington, 1975; Snowden *et al.*, 1989, 1996a,b; Schwartz and Chawluk, 1990; Diesfeldt, 1993; Scholten *et al.*, 1995; Schwarz *et al.*, 1998), presumably because the abilities upon which it depends are typically preserved in SD (e.g. praxis, visuospatial and constructional skills).

Three single-case studies and one group study have examined the spelling skills of SD patients in detail. Although none of the case studies stated that the patients involved had SD, each of the three patients fulfilled diagnostic criteria, and has subsequently been assigned this label (KT and TOB: Patterson and Hodges, 1992; Sasanuma and Patterson, 1995; WLP: Hodges *et al.*, 1998). The data reported in the case studies confirmed the qualitative descriptions (outlined above), in showing that the patients had a spelling impairment characterized by a tendency to produce phonologically plausible spelling errors (Schwartz *et al.*, 1979; Baxter and Warrington, 1987; Parkin, 1993).

Graham *et al.* (2000) provided a detailed study of spelling skills in a group of SD patients. Results indicated that impairment in spelling is a predictable feature in the syndrome, except in the earliest stages of disease. All 14 of the patients studied exhibited spelling deficits, and longitudinal follow-up of seven patients revealed further deterioration in spelling. Performance on words with unpredictable or exceptional sound-to-spelling correspondences was most affected, and the majority of errors were phonologically plausible. Non-word spelling was preserved in most patients, and there was little difference between written and oral spelling. The spelling impairment was correlated with, and was attributed to, the semantic deficit. This result was predicted on the basis of the connectionist model of lexical processing developed by Seidenberg and McClelland and colleagues (Seidenberg and McClelland, 1989; Plaut *et al.*, 1996), in which the computation of orthography from phonology is partly mediated by semantics.

Non-fluent progressive aphasia. Investigations of the spoken language deficit in NFPA have been numerous, particularly in the last decade, but as is often the case, written language skills have received little attention. Croot (1997) reviewed all cases of NFPA described in the (English language) literature between 1982 (when a paper by Mesulam stimulated interest in the syndrome) and 1997. Writing skills were described (mostly qualitatively) in 41 of 63 cases, and were considered to be impaired in 34 (82.9%). Thus, the majority

of NFPA cases showed a writing deficit. This deficit is often said to 'mirror' the spoken language impairment, in that writing may be telegraphic and contain morphological errors (Holland *et al.*, 1985; Mesulam and Weintraub, 1992b; Snowden *et al.*, 1992, 1996b; Snowden and Neary, 1993; Grossman *et al.*, 1996), and become progressively more telegraphic on follow-up (Holland *et al.*, 1985; Weintraub *et al.*, 1990; Mesulam and Weintraub, 1992b; Grossman *et al.*, 1996). Although spoken and written language may decline in parallel (Weintraub *et al.*, 1990; Mesulam and Weintraub, 1992a), it is often reported that patients with NFPA use writing to aid communication (Holland *et al.*, 1985; Weintraub *et al.*, 1990; Mesulam and Weintraub, 1992a; Kertesz *et al.*, 1994). Some patients, usually in the early stages of their illness, have normal spelling, but impairment in this domain is a common finding (Weintraub *et al.*, 1990; Caselli and Jack, 1992; Mesulam and Weintraub, 1992a; Snowden *et al.*, 1992, 1996b; Snowden and Neary, 1993; Kertesz *et al.*, 1994; Greene *et al.*, 1996; Hodges and Patterson, 1996; Watt *et al.*, 1997). The types of errors have only rarely been described, but seem to be generally non-phonologically plausible, and include omissions or transpositions of letters (Kartsounis *et al.*, 1991; Snowden *et al.*, 1996b). It has also been noted that the motoric aspects of writing may be executed somewhat slowly, although letters are well formed (Kartsounis *et al.*, 1991; Snowden *et al.*, 1996b).

Frontal variant of frontotemporal dementia. Despite growing interest in DFT and the associated language problems, spelling and writing skills in this syndrome have received little study, the few descriptions in the literature being mainly qualitative. For example, Snowden *et al.* (1996b) noted that written output may be reduced, and irrelevant in content, possibly because of general problems in attention, motivation and monitoring. Perseveration in writing, at the level of the sentence, has also been observed (Snowden and Neary, 1993). Spelling skills are preserved in some patients, although others may show (unspecified) spelling impairment (Snowden *et al.*, 1996b). Poor spelling has also been reported in the comparatively rarer syndrome which combines DFT and motor neuron disease (Ferrer *et al.*, 1991).

Primary progressive dysgraphia. This novel dementia syndrome was described by Graham *et al.* (1997), who reported a patient whose presenting and predominant symptom was dysgraphia. The dysgraphia was associated with left temporal lobe atrophy, but was not accompanied by the selective semantic deficit seen in semantic dementia. Ultimately, the patient developed semantic impairment, but this occurred in the context of a fairly generalized dementia. A second patient exhibited a similar dysgraphic syndrome, but this was accompanied by a striking anomia, even at presentation. Both patients showed initial surface dysgraphia and, over time, showed an increasing tendency to produce non-phonologically plausible spelling errors; these eventually dominated performance. An additional peripheral dysgraphia in both cases consisted of difficulty with producing letters, particularly in lower case, without a model to copy.

Posterior cortical atrophy (PCA)

PCA was initially described by Benson *et al.* in 1988. The dementia is dominated by problems with visual function, including visual agnosia, dyslexia, and features of Balint's syndrome (visual disorientation, optic apraxia and simultanagnosia). Other aspects of cognitive functioning are, at least initially, relatively preserved. Atrophy is in the occipito-temporal or occipitoparietal areas, and may be caused by different pathologies, including AD, subcortical gliosis and Creutzfeldt–Jakob disease (Berthier *et al.*, 1991; Victoroff *et al.*, 1994).

Dysgraphia is a one of the main features of the dementia associated with PCA (Benson *et al.*, 1988). Problems with writing are often apparent at presentation, and increase in severity as the illness progresses, frequently to a point of complete inability to write (Kiyosawa *et al.*, 1989; Graff-Radford *et al.*, 1993; Levine *et al.*, 1993; Ross *et al.*, 1996). Most descriptions in the literature of the dysgraphia in PCA are clinical (Benson *et al.*, 1988; Kiyosawa *et al.*, 1989; Berthier *et al.*, 1991; Graff-Radford *et al.*, 1993; Victoroff *et al.*, 1994; Perez *et al.*, 1996; Rogelet *et al.*, 1996), but a small number of studies have provided relevant data (Freedman *et al.*, 1991; Levine *et al.*, 1993; Ross *et al.*, 1996; Ardila *et al.*, 1997). The dysgraphia is mainly peripheral, often characterized by severe problems with legibility, placement of letters on a page, appropriate spacing between letters or words, and writing on a line; in addition, letters and strokes may be omitted or added (Levine *et al.*, 1993; Rogelet *et al.*, 1996; Ross *et al.*, 1996; Ardila *et al.*, 1997). This writing disorder is referred to as 'spatial dysgraphia' (Hécaen and Marcie, 1974; Ardila and Rosselli, 1993) or 'afferent dysgraphia' (Ellis *et al.*, 1987). Some studies have also reported a central spelling impairment in patients with PCA (Benson *et al.*, 1988; Freedman *et al.*, 1991; Graff-Radford *et al.*, 1993; Ross *et al.*, 1996; Ardila *et al.*, 1997). The nature of the errors has only rarely been characterized, but Ardila *et al.*'s results showed that their patient was surface dysgraphic. In contrast, one of the patients reported by Ross *et al.* (Case 1, the only one in whom the errors were described) produced non-phonologically plausible errors consisting of letter omissions or substitutions.

Taken together, these studies illustrate that the dysgraphia in PCA is most likely to be peripheral and to show features of spatial dysgraphia; this is presumably due to the severe visual problems associated with PCA. When there is a spelling disorder, the nature may vary, presumably with the precise neuroanatomical location of the pathology, and/or disease severity. Further studies will be required to learn whether spelling impairment is ubiquitous in PCA, and if there is a typical pattern of progression.

Dementia with Lewy bodies (cortical Lewy body disease)

Dementia with Lewy bodies was recognized as a separate clinico-pathologic entity, and diagnostic criteria defined (McKeith *et al*., 1996) only relatively recently. In addition to the pure syndrome, caused by (mainly) cortical Lewy bodies, some authors have reported the existence of an overlap syndrome in which both Lewy bodies and plaques (a feature of AD) are observed on neuropathological examination of the brain (Hansen *et al*., 1990; Cercy and Bylsma, 1997; Connor *et al*., 1998; McKeith, 1998). The neuropsychological impairments seen in dementia with Lewy bodies have really only been studied over the last 10 years, and little attention has been paid to the skills of spelling and writing. I know of no study that has examined these skills in patients with purely Lewy body pathology, but two included relevant tasks as a minor component of studies comparing the Lewy body variant (LBV) of AD with pure AD (Hansen *et al*., 1990; Connor *et al*., 1998). Hansen *et al*.'s (1990) results indicated that patients with LBV were poorer at writing words to dictation than AD patients with a similar severity of dementia, but neither the stimuli nor the errors were described, so the nature of the dysgraphia in LBV was not illuminated. Connor *et al.* (1998) found that LBV patients did not perform as well as those with AD on writing a sentence, but again the errors were not described. Severe visuospatial impairment is typical in dementia with Lewy bodies (McKeith *et al*., 1996), and one of the studies comparing LBV and AD found that these skills were more affected in the LBV group (Hansen *et al*., 1990). Thus, the impairments in writing sentences, and words to dictation, may be due to a peripheral writing disorder caused by the visuospatial deficit. A central spelling disorder cannot, however, be ruled out. Further studies will be required to elucidate the nature and frequency of dysgraphia in Lewy body dementia.

Vascular dementia

Little is known about the nature or even the prevalence of the dysgraphic deficits in vascular dementia because few studies have included relevant assessments. A small number of investigations comparing neuropsychological deficits in vascular dementia versus DAT have included assessment of writing, and all found evidence of greater impairment in vascular patients (Erkinjuntti *et al*., 1986; Powell *et al*., 1988; Kertesz and Clydesdale, 1994; Carey *et al*., 1999). Two of these studies evaluated performance on the writing subtest from the Western Aphasia Battery (WAB). In the earlier one, Powell *et al.* (1988) found that vascular and DAT patients with equivalent severity of dementia were equally impaired on narrative writing and writing to dictation, but the vascular patients showed more problems with the mechanics of writing. The other study (Kertesz and Clydesdale, 1994) found that vascular patients were more impaired overall on the WAB writing subtest than DAT patients with equivalent severity and duration of dementia. Moreover, in contradiction with Powell *et al*.'s (1988) findings, the writing subtest was one of the most useful tests for discriminating between the patient groups. Kertesz and Clydesdale also observed that vascular patients had particular difficulty (relative to DAT patients) with writing letters to dictation and copying of a sentence, suggesting greater impairment on the peripheral aspects of writing. This deficit could have arisen at the allographic or graphic motor pattern levels, but further investigation would be needed to confirm and specify this hypothesis, and to determine whether this is a typical pattern in vascular dementia. Carey *et al.* (1999) compared vascular and DAT patients on a narrative writing task, and (in contrast with Powell *et al.*, who observed no difference between groups on a similar task, see above) found that the vascular dementia patients were more impaired: they produced more spelling errors and grammatically simpler sentences, and had greater difficulty with writing in straight horizontal lines. This indicates that the vascular dementia patients were impaired on both central and peripheral aspects of writing.

One study provided a detailed investigation of the spelling impairment in a patient with vascular dementia (Lesser, 1990). Patient TF showed an unusual deficit: he could not write words to dictation unless he spelled them orally first. Attempts to write dictated words (or letters) without prior oral spelling resulted in illegible strokes. TF also had a spelling deficit (observed on oral spelling) with features of surface dysgraphia, implying difficulty in accessing the orthography of specific words. To explain the discrepancy between oral and written spelling, Lesser suggested that responses in the two modalities are derived from the spelling system independently. This type of disorder could, however, have arisen from problems in peripheral aspects of the writing process.

Taken together, these studies suggest that patients with vascular dementia may show impairment in both central and peripheral aspects of the writing process, but this conclusion is based on small numbers of data and subjects, and therefore must be taken with caution. The specific nature of the spelling and writing deficits in vascular dementia has not yet received systematic study.

Cortico-basal degeneration (CBD)

Although problems with writing are a common presenting complaint in CBD (Rebeiz *et al*., 1968; Riley *et al*., 1990; Moreaud *et al*., 1996; Mimura *et al*., 1997; Blasi *et al*., 1999; Graham *et al*., 1999), the dysgraphia in this clinico-pathologic entity has received little study. The difficulties with writing presumably arise from the dyspraxia, which is a diagnostic feature in CBD (Lang *et al*., 1994; Rinne *et al*., 1994), and which may ultimately render sufferers unable to write. Most studies which examined the writing impairment in this illness have provided only a qualitative description. For example, the patient studied by Moreaud *et al.* (1996) wrote slowly

and hesitantly, and had difficulty forming individual letters. Graham et al.'s (1999) patient was able to execute the motor act of writing quite well, despite his dyspraxia, but he had difficulty with cross-case transcription of single letters (e.g. a → A, D → d), seemingly because he had difficulty in remembering the appearance of the letters; this suggests a functional impairment at the allographic level in models of writing.

Impairment in spelling has also been reported in CBD. Graham et al. (1999) investigated this skill in their patient because it was a presenting complaint. Results showed that early in the course of his illness the patient was surface dysgraphic, but on follow-up, non-phonologically plausible errors became the predominant error type. This pattern of progression in spelling deficit has also been reported in patients with DAT (see above). Another case study involving a CBD patient reported an (unspecified) spelling impairment (Beatty et al., 1995), but spelling has not been extensively studied in this disorder, and one therefore cannot predict whether most sufferers should be expected to have impaired spelling.

Huntington's disease (HD)

A thorough description of language functions in HD was provided by Podoll et al. (1988), who administered the Aachen Aphasia Test (AAT) to 45 (German-speaking) patients. Writing and spelling were assessed by having subjects write seven words and three sentences to dictation. Not surprisingly, the involuntary choreiform movements that are a symptom of HD interfered with the execution of writing, and in severe cases led to inaccurate placement of letters, inconsistency in the slant of letters within words, and large variations in the pressure exerted on the page. Two patients were prevented from writing by their chorea. A spelling deficit characterized by omission, addition, substitution and perseveration of letters was observed in the middle to later stages of the disease. Although the authors attributed these spelling errors to constructional impairment, this is not the only possible interpretation; the types of errors documented by Podoll et al. have also been reported in association with hypothesized damage to the graphemic buffer—the temporary store where abstract graphemic representations are held while output processes such as writing are executed (Ellis, 1982; Caramazza et al., 1987). Further investigations of spelling, including oral spelling (which would not be affected by constructional difficulties) and a larger number of target words of varying lengths (length effects are expected in graphemic buffer impairment), will be required to determine the functional locus of the spelling impairment in HD.

Progressive supranuclear palsy (PSP)

Studies that have examined dysgraphia in PSP have focused mainly on disturbances in handwriting, rather than spelling. Qualitative descriptions indicate that handwriting becomes cramped, and ultimately illegible (Steele et al., 1964). Micrographia has also been reported (Dix et al., 1971). Podoll et al. (1991) evaluated spelling as well as writing in their study of six (German-speaking) PSP patients, who were asked to write seven words and three sentences to dictation (from the AAT). The legibility of writing was detrimentally affected in each patient, and this was attributed to loss of dexterity in finger movements. The majority of spelling errors (79%) involved omissions of letters, and omission of words within sentences was also observed. This is an unusual pattern of spelling disorder, and Podoll et al. speculated that it may be due to problems with visual monitoring of writing, presumably resulting from the vertical gaze palsy that is typical in PSP. This seems unlikely to be the entire explanation, however, as lack of visual feedback during writing led neurologically intact subjects to produce errors involving duplications of strokes and letters, as well as omissions (Lebrun, 1976).

Parkinson's disease (PD)

Difficulty with writing is an early and common symptom in PD (Selby, 1990). Writing may be untidy and is often slow (Margolin and Wing, 1983; Selby, 1990). In keeping with this, Cummings et al. (1988) found that PD patients were more impaired than those with DAT on 'writing mechanics'. Micrographia affects a minority of PD sufferers (McLennan et al., 1972): the size of writing is small, and often diminishes from the beginning to the end of a line. Studies have shown that patients can voluntarily increase the amplitude of their writing (at least temporarily) if they are given visual cues (marks or lines on a page indicating the required size) or auditory reminders (McLennan et al., 1972; Oliveira et al., 1997).

Investigations of the cause(s) of diminished writing size have focused on the allocation of time and force to strokes in writing. Margolin and Wing (1983) found that a decrease in letter size was associated with an increase in movement time, suggesting that the micrographia was not due to slowness, but to inadequate force. When movements take longer, one would expect letters to get larger, rather than smaller, if the force were unchanged. Van Gemmert et al. (1999) also suggested that PD patients have difficulty in maintaining the appropriate level of force needed in writing to produce the appropriate stroke size.

Aside from the problems with the size of their writing, the PD patients studied by Margolin and Wing (1983) produced well-formed written output, which included the appropriate strokes. This indicates that the writing impairment arises at a peripheral level, after information about the shapes of letters has been accessed, and suggests problems with the execution of graphic motor programmes. The problem with maintenance of adequate force is not, however, specific to writing, as this mechanism has also been proposed as an explanation for hypometric movements in walking, arm

movements, etc., in PD patients (see Van Gemmert *et al.*, 1999).

Conclusions

Dysgraphia is a common symptom in dementia, and may take varying forms. Although similar dysgraphic impairments are sometimes associated with different dementia syndromes or disease processes, assessments of spelling and writing may have some relevance in differential diagnosis. For example, although surface dysgraphia has been documented in both DAT and SD, problems with execution of graphic motor patterns are more likely to occur in AD. Similarly, symptoms of spatial dysgraphia suggest posterior cortical atrophy, while observation of mainly non-phonologically plausible spelling errors (arising from the central spelling system, rather than problems with writing) in the early stages of dementia may indicate non-fluent progressive aphasia. The latter suggestion is somewhat tenuous, however, as it is based mainly on qualitative descriptions in the literature.

There is a paucity of investigations of the dysgraphic deficits associated with many (often rare) forms of dementia, including NFPA, DFT, dementia with Lewy bodies, and vascular dementia. The spelling and writing deficits in these syndromes are, therefore, not well understood. Similarly, the dysgraphic deficits in dementias associated with movement disorders (e.g. CBD, HD, PSP and PD) are poorly documented. Because some of the symptoms in these dementias can affect writing adversely (e.g. oculomotor difficulties, dyspraxia, hyper- or hypokinetic movements, etc.), most studies have focused on this aspect, and the spelling deficits (if any) have received little investigation.

The spelling deficits observed in dementia are generally parallel to those documented in patients with stable brain lesions resulting from stroke or head injury, but are often less pure: although similar types of errors are observed in progressive and stable lesions, the distribution of these errors seems to differ. There are numerous reports of patients with stable lesions who produce mainly phonologically plausible (e.g. Beauvois and Derouesné, 1981; Goodman-Schulman and Caramazza, 1987; Rothi *et al.*, 1987; Behrmann and Bub, 1992; de Partz *et al.*, 1992) or non-phonologically plausible (Caramazza *et al.*, 1987; de Partz, 1995; Annoni *et al.*, 1998) spelling errors. In contrast, many of the studies reviewed here reported that dementia patients made a mixture of error types (i.e. both phonologically plausible and non-phonologically plausible), although often predominantly one type or the other (e.g. Croisile *et al.*, 1995; Neils *et al.*, 1995a; Graham *et al.*, 1997, 2000; Béland *et al.*, 1999; Glosser *et al.*, 1999b).

There are at least two possible interpretations of the different findings in dementia versus stroke patients. First, the spelling impairments in the two types of aetiologies may be distinct. On the widely accepted dual-route model of spelling, the mixture of error types reported in dementia implies more than one locus of impairment (see above). In

keeping with this, it would not be surprising that patients with dementia are more likely than those with stable lesions to show more than one spelling impairment. A second possible interpretation, however, is that the spelling impairments in the two types of aetiologies are more similar than the literature leads us to believe. The numerous reports of strong dissociations between error types in patients with stable lesions may give a misleading impression regarding the frequency of occurrence of these dissociations. This is because cases with stable lesions who make only or mainly one type of error may be considered to be of greater interest, and may therefore be more likely to be reported. On this interpretation, clear dissociations may be the exception rather than the rule. Indeed, it has been noted that within the field of cognitive neuropsychology, strong dissociations are rare and may represent anomalies (Goldberg, 1995; Appelbaum and Bates, 1999). The results of a multiple single-case study ($n = 53$) of spelling in aphasic patients with stable lesions (Luzzatti *et al.*, 1998) are consistent with this view: although many patients showed dysgraphia, suggesting impairment primarily in either the lexical or assembled routes, this often occurred in conjunction with a milder impairment in the alternate route, suggesting that having more than one locus of impairment is not uncommon in patients with stable lesions. Additional studies using unselected series of patients and stringent criteria for dissociations would allow us to distinguish whether clear dissociations between error types are indeed more likely to occur in patients with stable lesions, as compared to those with dementia.

In conclusion, further research which is guided by information-processing models, and which assesses both central and peripheral mechanisms in writing, will help us to understand the dysgraphic disorders in the different dementias. In addition, although relevant longitudinal studies have been carried out (e.g. Platel *et al.*, 1993; Neils and Roeltgen, 1994), they were done on a relatively short time scale (of 1 year or less), and the change over such a short time may be minimal. Longer-term studies would enable us to document dysgraphic deficits at different stages, and to look at the evolution of the impairment(s).

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