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The benefits and protective effects of behavioural treatment for dysgraphia in a case of primary progressive aphasia

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Abstract

Background: Spoken and written language difficulties are the predominant symptoms in the progressive neurodegenerative disease referred to as primary progressive aphasia (PPA). There has been very little research on the effectiveness of intervention on spoken language impairments in this context and none directed specifically at progressive written language impairment.

Aims: To examine the effectiveness of behavioural intervention for dysgraphia in a case of primary progressive aphasia.

Methods & Procedures: We carried out a longitudinal single-case study that allowed us to examine the effectiveness of a non-intensive spell-study-spell intervention procedure. We did so by comparing performance on four sets of words: trained, repeated, homework, and control words at five evaluations: baseline, during intervention, after the intervention, and at 6- and 12-month follow-up.

Outcomes & Results: We find that: (1) at the end of the intervention, Trained words show a small but statistically significant improvement relative to baseline and an advantage in accuracy over Control, Homework, and Repeated word sets. (2) All word sets exhibited a decline in accuracy from the end of treatment to the 6-month follow-up evaluation, consistent with the degenerative nature of the illness. Nonetheless, accuracy on Trained words continued to be superior to that of Control words and not statistically different from pre-intervention baseline levels. (3) Repeated testing and practice at home yielded modest numerical advantages relative to Control words; but these differences were, for many comparisons, not statistically significant. (4) At 12 months post-intervention, all words sets had significantly declined relative to pre-intervention baselines and performance on the four sets was comparable.

Conclusions: This investigation documents—for the first time—that behavioural intervention can provide both immediate and short-term benefits for dysgraphia in the context of primary progressive aphasia.

Language difficulties may be the predominant symptom in progressive, neurodegenerative disease (Mesulam, 1982). In many of those cases, written language difficulties may accompany spoken language impairments, or may even be the primary presenting language deficit (Westbury & Bub, 1997). There has been very little research on the effectiveness of intervention (behavioural or pharmacological) on spoken language impairments in progressive neurological disease and, to our knowledge, none directed specifically at progressive written language impairment. Here we report on a study of the effectiveness of behavioural intervention for dysgraphia in a case of primary progressive aphasia. We use a spell-study-spell training procedure that has been used with some success in a number of

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cases of acquired dysgraphia (Beeson, 1999; Rapp & Kane, 2002). The findings of this investigation indicate that the behavioural intervention yielded significant immediate gains as well as medium-term benefits for trained words relative to untrained control words.

PROGRESSIVE APHASIA AND DYSGRAPHIA

A decline in spoken language abilities is a common feature of a wide range of different neurodegenerative diseases and, in some cases, may be the predominant symptom in the initial stages or even throughout the course of the disease. Primary progressive aphasia (PPA) refers to specifically to those degenerative conditions in which spoken language represents the only, or salient, domain of cognitive decline for at least the first 2 years of the disease (Mesulam, 1982; Mesulam, Grossman, Hillis, Kertesz, & Weintraub, 2003; Weintraub, Rubin, & Mesulam, 1990). This investigation will focus on written language production which may be disrupted in a wide range of neurodegenerative diseases (see Graham, 2000, for a review) including Alzheimer's disease, frontal and/or temporal lobe atrophy, posterior cortical atrophy, and cortico-basal degeneration. Furthermore, Westbury and Bub (1997), in their review of published reports of 112 individuals diagnosed specifically with progressive aphasia, indicated that although written language abilities were not routinely evaluated in the context of PPA, in 8% of the cases in their total sample, written language difficulties (dysgraphia: 5%, dyslexia: 3%) were actually the presenting symptoms. There have also been a few reports of individuals in whom dysgraphia was not only the presenting, but also the predominant, symptom for an extended period of time (Graham, Patterson, & Hodges, 1997; Luzzi & Piccirilli, 2003), a condition referred to by Graham (2000) as "primary progressive dysgraphia". In contrast, it has also been reported that written language abilities may be relatively preserved throughout the course of the progressive spoken language deterioration or that written language abilities may be affected only in later stages (see Luzzatti, Iaiacona, & Agazzi, 2002, for a review). In fact, some researchers have commented that for a number of individuals with progressive aphasia, written language remains a relative strength and they sometimes rely on written language for their communication (Cress & King, 1999; Holland, McBurney, Moossy, & Reinmuth, 1985; Kertesz, Hudson, Mackenzie, & Munoz, 1994; Mesulam & Weintraub, 1992; Murray, 1998;Weintraub et al., 1990). Despite the relative paucity of data on written language in neurodegenerative disease, the picture that emerges is one in which written language impairment is a common symptom, often occurring alongside spoken language deterioration; however, written language may be either selectively affected or preserved for considerable periods of time during the course of the disease.

The specific dysgraphias that have been described in the context of neurodegenerative diseases are apparently similar, in terms of symptomatology and underlying functional deficits, to those observed in patients with non-progressive brain lesions that result from stroke, head injury, or surgery (Graham, 2000). For example, in neurodegenerative cases there are very frequent reports that include spelling impairments that primarily affect the retrieval of the spellings of familiar words that are stored in long-term memory (Ardila, Matute, & Inozemtseva, 2003; Croisile, Carmoi, Adeleine, & Trillet, 1995; Hillis et al., 2006; Platel, Lambert, Eustache, & Cadet, 1993; Rapcsak, Arthur, Bliklen, & Rubens, 1989). This memory system is often referred to as the *orthographic lexicon* (see Figure 1), and deficits to this system are sensitive to the frequency of words and typically result in the production of phonologically plausible spelling errors (e.g., spelling "yacht" as YOT) if the sublexical phonology-to-orthography conversion process remains relatively intact. The deficit pattern that emerges from damage to the orthographic lexicon (or the lexical system more generally) is referred to clinically as "surface dysgraphia". There are also reports of cases of neurogenerative disease in which the written language deficits especially affect the processes involved in using the knowledge of the sublexical, regular relationships between sounds and letters that are necessary for the spelling of unfamiliar words or pseudowords—the *phonology-to-orthography conversion system* (Luzzatti, Iaiacona, & Agazzi, 2003). This deficit is characterised by difficulties in spelling pseudowords and is often referred to as "phonological dysgraphia".

In addition, deficits specifically affecting the ability to hold letters in the *graphemic buffer* (i.e., orthographic working memory) during the serial process of producing a written or oral spelling have also been described in cases of progressive disease (O'Dowd & Zubicaray, 2003), as have been a range of peripheral impairments. The peripheral impairments arise at different stages of letter shape production, and include not only fairly commonly reported deficits of motor execution that result in poorly formed letters, but also deficits that are apparently due to such things as a failure to remember the shapes of letters (Graham, Zeman, Young, Patterson, & Hodges, 1999), difficulties with a specific case or font (e.g., Graham et al., 1997), difficulties with spacing of letters, and other spatial aspects of writing, and so forth. In many cases, and particularly with disease progression, multiple processing mechanisms within the spelling system are affected. It is a matter of debate whether or not there is a specific order in which deficits to the spelling system arise with disease progression. Some researchers have suggested that the norm is a progression from central (lexical) impairments to more peripheral deficits (Graham, 2000; Hughes, Graham, Patterson, & Hodges, 1997), while others have claimed that there is no fixed order and that the progression depends, for each individual, on the specific neuro-geography of the disease progression (e.g., Luzzatti et al., 2003).

INTERVENTION IN CASES OF PROGRESSIVE APHASIA

There have been only a few papers reporting the results of specific interventions in cases of PPA (Cress & King, 1999; McNeil, Small, Masterson, & Fossett, 1995; Murray, 1998; Pattee, von Berg, & Ghezzi, 2006; Schneider, Thompson, & Luring, 1996). These papers vary in the extent to which they include control conditions and/ or assessment measures that allow for a statistical evaluation of the effectiveness of the treatment approaches. Nonetheless, the existing literature consistently indicates promising outcomes for a range of treatment approaches.

Cress and King (1999), Pattee et al. (2006), and Murray (1998) all report on the use of augmentative and alternative communication (AAC) strategies that were developed to supplement the communication strategies and abilities of individuals with PPA. For example, Cress and King (1999) developed and provided training for devices such as communication books and boards, as well as the coaching of communication partners. Their evaluation of one individual with whom they carried out an intensive 1-month intervention indicated, among other things, that he was able to learn the majority of the symbols on the boards, initiated more communicative interactions with AAC support than without AAC support and that many of these activities continued even 1 year after the intervention. Similarly, Pattee et al. (2006) documented that an individual with PPA improved her use of a text-to-speech device as well as her ability to make use of and expand upon previously acquired American Sign Language signs and fingerspelling. Furthermore, the individual showed greater benefits and a clear preference for ASL over the text- to-speech device. Murray (1998) also showed benefits for a number of different treatment approaches instituted over a 2½-year period in order to meet the changing communicative needs of an individual with PPA. The interventions included: Dyna Vox (Sentient Systems), dyad-oriented therapy with her spouse, and the use of drawing to supplement expressive communication. Using these approaches, Murray (1998) reported benefits in the face of decline in language functions (such as repetition) that were not trained.

In contrast to work with augmentative and alternative communication strategies, McNeil et al. (1995) and Schneider et al. (1996) applied interventions directed at strengthening specific areas of language weakness. In a single-case study McNeil et al. (1995) targeted spoken word production in a 5-month, multiple-baseline, multiple-probe study using a cueing hierarchy to elicit synonyms or antonyms for a set of adjectives. These researchers found improved spoken word production as well as evidence of generalisation to words in untrained sets. These gains were observed in the context of decline in other language abilities. A pharmacological intervention with dextroamphetamine was included along with the behavioural intervention, but the results did not show any added benefit of the pharmacological agent. Schneider et al. (1996) targeted the production of verb tense with a treatment that involved oral and oral + gestural training. They found improved sentence production for trained verbs as well as generalisation to untrained verbs of the same tense, but not to other tenses. In this study (as in a number of the other studies) the evidence of the benefits of the interventions comes from observation of improvements in the specifically trained behaviours in the face of decline in non-trained language functions.

In sum the research to date, although extremely limited, is promising in that it indicates that individuals with progressive language deficits, despite suffering from neurodegenerative disease, are nonetheless able to strengthen specific areas of language decline, as well as learn to use strategies and devices to supplement their communicative abilities. The research raises a vast number of questions regarding the nature of the interventions, their timing, patient characteristics, etc. It is clear that what is needed is extensive research designed to document the conditions under which intervention benefits can be maximised in order to extend language use and effective communication skills for as long as possible.

TREATMENT OF ACQUIRED, NON-PROGRESSIVE DYSGRAPHIAS

The investigation described in this paper involves a single case study of an individual with progressive aphasia and dysgraphia in which intervention is targeted at strengthening the representations of words spellings (the orthographic lexicon). To our knowledge there has been no published research involving treatment of progressive dysgraphia, and even the literature regarding behavioural intervention in acquired dysgraphia is relatively small. The training procedures implemented in this study are similar to those that have been applied in various cases of acquired, non-progressive dysgraphias that resulted from stroke, infection, tumour resection, head injury, and other aetiologies. We briefly review these here.

A number of treatment studies have targeted specific components of the spelling process (see Figure 1), such as the semantic component (Hillis, 1991, 1992), the sublexical phonology-to-orthography conversion system (Carlomagno, Iavarone, & Colombo, 1994; de Partz, Seron, & Van der Linden, 1992; Hatfield, 1983; Luzzatti, Colombo, Frustaci, & Vitolo, 2000), the graphemic buffer (Hillis, 1989), or the orthographic lexicon (Aliminosa, McCloskey, Goodman-Schulman, & Sokol, 1993; Beeson, 1999; Beeson & Hirsch, 1998; Beeson, Hirsch, & Rewega, 2002; Behrmann, 1987; Behrmann & Byng, 1992; Carlomagno et al., 1994; Clausen & Beeson, 2003; de Partz et al., 1992; Hatfield & Weddel, 1976; Hillis & Caramazza, 1987; Rapp, 2005; Rapp & Kane, 2002; Raymer, Cudworth, & Haley, 2003; Schmalzl & Nickels, 2006; Seron, Deloche, Moulard, & Rousselle, 1980; Weekes & Coltheart, 1996). In addition, there have been some studies that have targeted the interaction between lexical and sublexical processes (Beeson, Rewega, Vail, & Rapcsak, 2000) and others that have focused on components that are specifically required for written as compared to oral spelling (Mortley, Enderby, & Petheram, 2001; Pound, 1996).

The majority of these treatment studies have been directed at strengthening the representations of word spellings stored in the orthographic lexicon. These studies have generally employed techniques in which the correct spellings of words are repeatedly presented for study and spelling and/or delayed copy. This general method is based on the assumption that neural injury has weakened/damaged the representations of words in long-term memory and that the repeated presentation of words will strengthen or facilitate retrieval of these representations. Various other techniques have also been used (for a review see Beeson & Rapcsak, 2002) for improving different aspects of the spelling process. For example, Hillis and Caramazza (1987) taught error detection and correction strategies to an individual with a deficit affecting graphemic buffering, and Hillis (1989) used a cueing hierarchy in another case. A number of the studies that have been directed at remediating deficits affecting the sublexical processes of the phonology-to-orthography conversion system have focused on re-teaching phoneme–grapheme relationships, often by pairing each phoneme with a “key word” that the individual can spell (Carlomagno & Parlato, 1989; de Partz et al., 1992; Hatfield, 1983; Hillis Trupe, 1986; Luzzatti et al., 2000). There are also reports of studies that use computers and word processors to facilitate repetitive practice or to complement an intervention that targets specific cognitive processes (e.g., Mortley et al., 2001).

The treatment outcomes in these studies are generally positive with regard to improvement on treated items, although there is considerable variability with regard to generalisation and maintenance. Despite the relatively small number of controlled studies of treatment outcomes in non-progressive dysgraphia, it is clear that spelling deficits arising from impairments to different functional components are amenable to improvement even many years after the onset of the dysgraphia. Furthermore, in at least some cases, this improvement has been shown to have a functional impact on daily life (see Beeson & Rapscak, 2002, for discussion). Also important is the observation that in at least some individuals with both aphasia and dysgraphia, the dysgraphia has been shown to be more amenable to intervention than the aphasia (Beeson, 1999; Beeson et al., 2002; Beeson, Rising, & Volk, 2003; Robson, Marshall, Chiat, & Pring, 2001). That is, the effectiveness of dysgraphia treatment is not always dependent on the integrity of the spoken language system. This is consistent with other evidence of the relative independence of at least some of the cognitive components of written and spoken language processing (for a review see Rapp, Benzing, & Caramazza, 1997; Rapp & Caramazza, 2002). The independence of certain aspects of the written and spoken language systems may have implications for the role of dysgraphia treatment in the context of progressive spoken language impairment.

INTERVENTION IN A CASE OF PROGRESSIVE DYSGRAPHIA

It is important to determine whether or not treatment approaches that have been successful in cases of non-progressive dysgraphia can be applied in cases of neurodegenerative disease to either improve written language abilities or prolong skill levels in the face of the degenerative disease process: This is the aim of this paper. We report on a single case study of a woman diagnosed with primary progressive aphasia with dysgraphia, for whom dysgraphia was the initial symptom. The two behavioural interventions used in this study were a spell-study-spell type procedure implemented in the laboratory as well as a modified version of the same procedure that was carried out independently in the home. Two control conditions were included to determine treatment effectiveness and generalisation to untrained words. Finally, the maintenance of gains was assessed in follow-up evaluations at 6 and 12 months after the end of the intervention. In the General Discussion we consider, among other issues, the role of dysgraphia intervention in the context of spoken language deterioration and the similarities and differences between interventions in non-progressive versus progressive dysgraphia.

CASE STUDY

CB was a right-handed, college-educated, professional artist and illustrator of children's books, who was born in 1941. She was an avid reader and excellent speller, and had planned to begin writing professionally before her neurodegenerative illness made this impossible. She reports that a grandfather and an aunt suffered from dementia. CB recalled experiencing the earliest symptoms of language difficulties in 1996 at the age of 55. At that time she found that she was occasionally unable to spell common, irregular words (such as BELIEVE) and that she occasionally found reading for meaning to be more difficult and slower than it had been previously. She reported that in the subsequent 5 years she began to experience word-finding difficulties, and difficulties with calculation as well as with her artwork. In 2001 she sought evaluation, and clinical neuropsychological testing was carried out at that time and as well as in 2003 and 2004[1] (see Table 1 for a time-line). She was able to live alone and fairly independently with only some outside assistance until 2006, at which time family and friends began to make arrangements for a living situation in which she would receive more supervision.

Neuropsychological testing

A diagnosis of PPA was made on the basis of CB's performance on the 2001 and 2003 neuropsychological evaluations (Table 2). In 2001, CB was diagnosed as having moderately severe neurocognitive impairment of an undetermined aetiology. Her symptoms included mild dysnomia, marked dyscalculia, dysgraphia, mild-to-moderate deficits of attention and new learning, psychomotor slowing, and mild constructional difficulties. In contrast, delayed recall and verbal recognition memory and frontal lobe-type functions were within normal limits. MRI scanning at the time revealed asymmetric temporal parietal atrophy (see Figure 2). The 2003 evaluation revealed a continued anomia, with the written modality more impaired than the spoken, and a deterioration in language functions in the face of fairly stable performance in memory and non-language tasks. On this basis CB was diagnosed with primary progressive aphasia of a fluent-type, most likely due to frontotemporal lobar degeneration or Pick complex.

The dysgraphia intervention study reported in this paper began in 4/2004 shortly after the second cognitive neuropsychological evaluation (in 12/2003) and ended 4 months later in 8/2004, with follow-up evaluations 6 and 12 months after the end of the intervention. CB underwent a third cognitive neuropsychology evaluation in 9/2004, shortly after the intervention phase of the study. As indicated in Table 2, this third evaluation revealed an accelerating pace of language deterioration—e.g., Controlled Oral Word Association Test (Benton & Hamsher, 1976), a written noun/verb picture naming task (unpublished)) as well as impairment of verbal memory (Rey Auditory Verbal Learning Test; Schmidt, 1996), and general cognitive functions (Mini Mental Status Examination; Folstein, Folstein, & McHugh, 1975); Wechsler Memory Scale (Wechsler & Stone, 1973). However, visual perception remained relatively stable (Rey-Osterreith Complex Figure Test; Osterreith, 1944; Rey, 1942). Further information regarding patterns of spared and impaired abilities during the period of 2003–2005 will be reported below in the context of the intervention study.

DYSGRAPHIA INTERVENTION STUDY

In addition to the clinical neuropsychological evaluations, a number of tasks were administered as part of the dysgraphia intervention study itself with the following two

[1]We are grateful to Drs Hillis and Selnes at Johns Hopkins Hospital for providing us with the results of the neuropsychological diagnostic testing that they carried out with CB.

objectives: (a) to assess a range of cognitive and language abilities at three time points: prior to the onset of training, after the end of training and during the follow-up period, and (b) to determine the specific deficits affecting CB's spelling system.

General assessment

It was important to evaluate a range of skills in order to characterise the backdrop against which intervention-related changes might take place. That is, in order to attribute any significant changes specifically to the behavioural intervention it was important to evaluate if other skill domains were stable or deteriorating during the period of the intervention study.

As indicated in Table 3, at the onset of the investigation CB showed good visuo-spatial skills, as well as language comprehension. In language comprehension she showed fairly intact performance in both written and spoken domains. With regard to spoken language, her comprehension was adequate at both the single word and sentence level, as indicated in the tasks in Table 3 as well as in informal interactions. As concerns spoken language production, CB showed excellent abilities in word and nonword repetition as well as in sentence completion (completing sentence onsets such as: "John wanted to leave, but…"). These areas of fairly intact performance contrasted with her moderately impaired spoken picture (confrontation) naming. Errors in picture naming were semantic (ant → "mosquito"; 29%), phonological, including phonologically similar words (camel → "cannibal"; 14%) and nonwords (penguin → "kerwin";14%), circumlocutions and definitions (lettuce → "salad stuff, greens"; 43%). Anomia was also observed in her spontaneous speech, which was grammatical and fluent although slowed by occasional word-finding difficulties and circumlocutions. Her writing to dictation of both words and nonwords was severely impaired and will be discussed in greater detail below.

Although not all tasks were administered at all subsequent time points, the results (Table 3) indicate that throughout the entire period of this investigation CB's performance was stable for a range of visual abilities as measured by certain subtests of the BORB and the Benton Visual Retention Test. Spoken language was fairly stable from pre-training to post-training and through at least the first follow-up assessment in the following areas: single word auditory comprehension, single word repetition, and sentence completion. In contrast to these areas of stability, the testing documents significant deterioration during this same time period in spoken confrontation naming, $\chi^2(1)=5.9$, $p<.01$, and in the spelling to dictation of nonwords, $\chi^2(1)=4.9$, $p<.05$.

In sum, with respect to language, comprehension was better preserved than production, both in the written and spoken modalities, and written production was more affected than spoken production In contrast, visuo-spatial skills remained quite stable throughout the entire 18-month period of intervention and follow-up.

Dysgraphia assessment

The primary purpose of the dysgraphia assessment was to identify the components of the spelling system that were affected, focusing especially on those involved in writing words to dictation, as this was the task that was used in the intervention study. All spelling accuracies are reported as number of letters correct/letters in the stimuli. Each target letter was assigned a score of 0, 0.5, or 1; with a score of 1 assigned if the target letter was present and in the correct position, 0.5 if it was present but in the incorrect position, and 0 if the target letter was absent. Using this scoring method, for any given word set, percent correct equals the total score/total number of letters in all of the target words in the set. Although not reported, responses were also scored according to whether each word was correct or incorrect. The results obtained by considering letter or word accuracy revealed essentially the same pattern; however, we prefer letter accuracy as it is a far more sensitive measure.

According to Figure 1, writing to dictation of familiar words first requires understanding the dictated word. This is accomplished by identifying the word among the forms of familiar words stored in phonological long-term memory (the phonological lexicon) and then using this as a basis for accessing its stored meaning in the semantic system. CB's good performance on single word comprehension as measured by the PPVT (Table 3) through the first follow-up evaluation, allows us to be fairly certain that her spelling difficulties did not originate in her failure to understand the words dictated to her. Furthermore, for all spelling-to-dictation tasks, CB was asked to repeat every word before spelling it and on the rare occasion that she failed to repeat the stimulus correctly, it was repeated to her until she was able to produce it correctly.

Subsequent to auditory comprehension, the spelling-specific components involved in spelling to dictation include the orthographic lexicon, the graphemic buffer, letter shape selection, and motor execution. The orthographic lexicon is the long-term memory store of the spellings of familiar words. Once a representation has been accessed in the orthographic lexicon it is maintained active by the graphemic working memory process (graphemic buffer) while the form of each letter is selected by the letter-shape conversion process to be produced in serial manner by the motor system. The dysgraphia assessment evaluated the integrity of each of these spelling-related components.

We can infer that significant damage to post-buffering letter-shape selection and motor execution processes are unlikely to have been important contributors to CB's spelling difficulties on the basis of two sets of observations. First, there were no significant differences between CB's written and oral spelling accuracies on the same list of 27 words (approximately half low and half high frequency, half four-letter and half eight-letter words) letter accuracy: 62% (98/160) vs 59% (94/160); $\chi^2(1)=.21$, *ns*. This similarity indicates that there is no special difficulty in producing written spellings, and thus indicates that the spelling errors probably originated at a point in processing before written and oral spelling diverge. Consideration of Figure 1 indicates that either the orthographic lexicon and/or the graphemic buffer are likely deficit sites. Second, although, as indicated in Table 4, CB's performance in writing single letters to dictation (92% and 90% correct) and in transcribing from one case to another (89% to 92% correct) are not perfect, and indicate that some errors might have arisen in the course of letter-shape selection, the magnitude of such a deficit is not commensurate with the magnitude of her spelling difficulties.

There are specific findings indicating that both the orthographic lexicon and the graphemic buffer have been affected, and it is likely that both deficits contributed to CB's spelling difficulties. Damage to the orthographic lexicon is indicated by a significant difference in her combined oral and written spelling accuracy for high vs low frequency words: 73% (166.5/229) vs 57% (138/241); $\chi^2(1)=12.3$, $p<.001$. Damage to the graphemic buffering process is indicated by a significant effect of word length. CB was able to produce the letters of four-letter words significantly more accurately than those of eight-letter words: 71% (79/112) vs 54% (108.5/200); $\chi^2(1)=7.9$, $p<.01$.

Although CB's nonword spelling abilities were not extensively examined, the data in Table 3 indicate significantly impaired performance in nonword spelling with errors such as "kittle" → CIKOL; "reash" → CHEACH; "bruth" → BURTH). Nonetheless, it is noteworthy that accuracy with nonwords was superior to her performance with words. Difficulties in nonword spelling are expected when there are graphemic buffering difficulties as the buffering process is required for both words and nonwords. For this reason it is difficult to know if CB's difficulties with nonwords can be accounted for solely by the buffering deficit or if there is an additional deficit to the sublexical system. Relevant to this question is the fact that in word spelling CB sometimes produced phonologically plausible errors ("candle" → CANDIL), an indication that, at least on some occasions, when retrieval from the orthographic lexicon failed, the sublexical conversion system successfully generated a plausible spelling.

Table 5 reports the distribution of error types that CB produced in spelling a number of words lists. Her overall letter accuracy was 68% on these lists and, with respect to error types, it is noteworthy that she did not produce semantic or morphological errors in spelling to dictation. Her errors included not only phonologically plausible errors ("mercy" → MIRCEY; "method" → METHED) but also visually similar word errors in which the word response contained at least 50% of the target's letters ("crown" → COW; "hook" → HONK), visually similar nonword errors in which the response contained at least 50% of the target letters ("shrug" → SHUG; "prison" → PRISTON), as well as nonword responses that differed from the target by more than 50% of letters ("schedule" → SHEN; "province" → PARVERIS; note that we did not distinguish between orthographically vs phonologically similar responses). These types of errors are certainly consistent with a system in which there are deficits to both the orthographic lexicon and graphemic buffering while the remaining components remain relatively intact.

In sum, at the onset of the investigation both auditory comprehension and the peripheral post-buffering spelling processes seemed to be quite intact and were almost certainly not the major cause of the spelling difficulties. Instead, these difficulties were apparently the consequence of deficits affecting the orthographic lexicon, graphemic buffer and, possibly, sublexical phonology-to-orthography conversion.

Training protocol

There were four phases: pre-training, training, post-training, and follow-up. All four stimulus sets were presented for spelling to dictation twice during pre- and post-training evaluations and at the first follow-up, and only once at the second follow-up. Subsequent to the two pre-training (baseline) evaluations, training was carried out typically in biweekly sessions of approximately 3 hours, for a total of 11 complete administrations of the Trained and Repeated word sets. At each training session, the Trained words and the Repeated words were administered in a blocked fashion, with the order of blocks alternating from session to session. Each word set was divided into halves, as it typically required two sessions to administer both sets of words. After baseline testing, the duration of the intervention was 15 weeks (there were some interruptions in the training due to CB's travel or illness). Therefore, each word of the Trained and Repeated word sets was presented for a total of 11 times over a 15-week period. The follow-up evaluations were administered at approximately 6 and 12 months following the end of the training phase.

Stimulus categories

Stimuli consisted of four word sets (*n*=20 for each set) that were matched for lexical frequency, letter length, regularity, and concreteness: Trained, Repeated, Homework, and Control words. Table 6 reports on the mean values for these variables for each list. For the second follow-up evaluation, reduced word sets for all word categories were used in order to limit the amount of time needed for the evaluation. This was done to accommodate CB's deteriorating and increasingly slow spelling performance. Each reduced set consisted of a subset of 10 of the items from each original 20 word sets, selected so that: (a) the four lists were matched with one another (as well as to the original 20-word lists) with respect to frequency, length and concreteness and (b) each subset of 10 items was matched in accuracy with the accuracy of the list it was derived from at Follow-up 1.

Training procedures

A spell-study-spell procedure, similar to that used in a number of the studies described in the Introduction, was applied to the Trained words, such that, at every training session, CB was asked: (1) to spell each word to dictation, (2) to study the word presented on a written notecard while the experimenter repeated the word and orally named each of its letters and then, if the word had been spelled incorrectly, (3) to attempt to spell it again. This procedure was repeated until the word was correctly spelled (or for three repetitions). To be clear, when a word was spelled correctly on the first attempt, the word was still presented for CB to study and only then did the experimenter continue to the next word.

Repeated words were simply presented for spelling to dictation at each training session without feedback. Homework words were repeatedly copied and self-tested at home. Specifically, CB was given sheets of paper each with one of the target words written at the top. She was instructed to copy the word 10 times, turn the page, and then try to spell it from memory. She then was to turn back the page to check to see if it was correctly spelled and, finally, she was supposed to study the correctly spelled word. It is important to note that CB did not carry out homework with consistency and regularity and so it is not possible to consider this study to be an adequate test of the real efficacy of the homework condition. Nonetheless, we present the results for the Homework condition as it may reflect “real-world” limitations on the application of this type of treatment (especially with clients who live alone and don’t have someone to help supervise their homework). Control words were presented for spelling to dictation only at the time of the pre and post-treatment evaluations and at the two follow-ups.

The protocol structure allowed us to evaluate the following four effect types: word-specific treatment effects, word-specific homework effects, generalised learning effects, word-specific repetition effects. The evaluation of these effects is influenced by the fact that we are considering a neurodegenerative disorder. In the case of relatively stable diseases and conditions, we assume that during the intervention period, performance will remain unchanged in the absence of treatment. In the case of degenerative disease, while stability is one possibility, another is that in the absence of treatment, performance will deteriorate. These two possibilities and their impact on data interpretation will be considered in the following discussion.

Word-specific treatment effects refer to treatment benefits that affect the Trained words themselves and cannot be attributed simply to repeated attempts to spell the word or to natural progression (improvement or deterioration). In cases of *non-progressive* deficits, a treatment effect is indicated if there is better spelling performance on Trained words at the end of treatment than at the beginning, accompanied by better spelling performance on Trained versus Repeated words at the end of treatment. The latter is necessary to show that it was not merely the repeated testing that was efficacious, but that the treatment procedure itself was beneficial. However, in the context of a neurodegenerative disease, to show the effectiveness of treatment it is not required that the treatment actually improves performance relative to pre-treatment baseline. Instead, it must be shown that the treatment provided some benefit relative to the possible deterioration that might otherwise have occurred during the same time period. Performance on the Repeated words provides both an index of degeneration and takes into account any possible benefits of repeated testing. Therefore, in degenerative cases all that is required to show word-specific treatment effects is a significant difference between Trained words and Repeated words at the end of treatment.

Word-specific homework benefits are also treatment effects and therefore are subject to the same logic as word-specific treatment effects. That is, to demonstrate the specific benefits of Homework, these words should either (a) be spelled better at post-treatment than at baseline, and also better than Repeated words at post-treatment; or (b) simply be spelled better than Repeated words at post-treatment.

Generalised learning effects, in the case of non-progressive deficits, would be manifested by improvement in accuracy levels for the Control words from the beginning to the end of treatment. This is because we assume that, in the absence of treatment, control words will be stable from pre to post treatment. In the case of neurodegenerative disease, if we were to find such an improvement for the Control words it would, of course, indicate generalisation. However, if we were to find that performance on Control words stayed the same, or deteriorated between pre and post intervention evaluations, we could not reject the possibility that there might have been generalisation. This is because we have no way of knowing the extent of deterioration in the absence of treatment, and so we cannot be sure that the observed deterioration is equal to or less than would have occurred without treatment. There doesn't seem to be any good way to resolve this ambiguity; therefore even in the absence of improvement for control words, generalisation cannot be ruled out.

Word specific repetition effects are manifested, in the case of non-progressive deficits, by better spelling performance on the Repeated words at the end of treatment compared to the beginning accompanied by either (a) no generalised learning effect, or (b) an effect of repetition beyond that of generalised learning (spelling performance with the Repeated words better than that of the Control words at the end of treatment). This is because a repetition effect can only be distinguished from a generalised learning effect if the benefit to repeated words is greater than any benefits that apply to all words. In degenerative cases either of these two patterns would, of course, also constitute evidence of word specific repetition effects. However, in the case of a degenerative disorder, if there are no clear generalisation effects, word-specific repetition effects would be indicated (whether or not performance on Repeated words improved from pre to post intervention) if there were better performance on Repeated versus Control words at the end of treatment (i.e., control words show greater deterioration in performance).

RESULTS

Pre to post treatment effects

Two sets of analyses were used to evaluate the effect types described above. The first set of analyses involves comparisons of the spelling accuracies of the four word sets at pre versus post treatment evaluations. For comparisons of the *same* word sets at different time points we used the Wilcoxon matched pairs (signed rank) test, and for comparisons of *different* word sets at the same time point we used the chi-square test. Two-tailed tests are reported unless otherwise noted. The second analysis involves a regression analysis comparing the trajectory of change for the Trained versus Repeated conditions from pre to post-treatment. This regression analysis considers not only performance at pre and post-treatment "endpoints" but also includes all of the intermediate accuracy scores. For Repeated words these intermediate scores correspond to the results of administering the Repeated words for spelling to dictation at each treatment session; for Trained words these scores correspond to CB's first response to each of the words during the training trials (before she went on to carry out the study-spell procedure). This regression analysis was only possible for Trained versus Repeated words as none of the other stimulus conditions involved acquiring accuracy data at points other than at pre and post-treatment (and follow-up).[2]

Analysis 1.—Pre and post treatment letter accuracies for each list were combined (added) across the two evaluations carried out at each of these time points. This was done in order to reduce variability and increase power. A comparison of the accuracy levels between stimulus conditions at Pre intervention reveals no significant differences between Trained, Repeated, Homework, or Control words (chi square values for these comparisons ranged from .01 to .80), indicating that all word sets were starting from a comparable level.

First, visual inspection of changes in pre to post training accuracies (Figure 3) indicates a fairly clear overall pattern of results such that: Trained words increase in accuracy, Control words decrease in accuracy, and Homework and Repeated words remain fairly stable at an intermediate level of accuracy. Statistical evaluation reveals the following:

1. A *word-specific treatment effect* was indicated by a significant improvement in letter accuracy for Trained words from pre to post training evaluations, T=1596 (N=91) p<.05. This effect was not due simply to repeated evaluations, as the accuracy of Trained words was greater than that of Repeated words at post-treatment, $\chi^2(1)=4.94$, p<.05. In order to provide a more concrete sense of the types of changes observed for the Trained words, Table 7 reports CB's responses for the first pre-training evaluation and the final post-training evaluation.

2. There was no significant improvement for Homework words from Pre to Post Treatment, T=1411 (N=77) p<.65, nonetheless, there was evidence of a *word-specific homework effect* given CB's superior performance with Homework vs Control words at post-treatment that is statistically significant with one-tailed testing, $\chi^2(1)=2.91$, p<.01, 1-tailed. However, this benefit for Homework words cannot be distinguished from a benefit for repeated testing, as Homework words were not produced significantly more accurately at Post-treatment than Repeated words, $\chi^2(1)=.04$, *ns*.

3. We could not discern any *generalisation effects* as performance with Control words declined from Pre to Post training, although this decline only trended towards being significant, T=1914 (N=97) p<.1. As indicated above, it is possible that performance with Control words declined less than it would have without treatment, but there is no experimental methodology that allows us to determine whether that was the case in the context of the neurodegenerative disease.

4. With regard to a *repetition effect*, we first note that accuracy on Repeated words did not significantly improve from Pre to Post treatment, T=1675 (N=86) p>.4. However, there was a suggestion of a benefit of repetition as Repeated words were spelled more accurately than Control words at post-treatment, although this difference was not significant, $\chi^2(1)=1.98$, *ns*.

Analysis 2.—The strongest finding from Analysis 1 is that of a treatment benefit for Trained words. The finding of a treatment benefit is indicated by a significant accuracy difference between Trained words at Pre and Post-treatment as well as by a significant difference between Trained and Repeated words at Post-treatment. The difference between Trained and Repeated words is critical for establishing the benefits of treatment and this second analysis provides a more stringent test of this difference between conditions. It does so by comparing the Trained and Repeated not only at the endpoints of the intervention period, but by including all of the intermediate evaluations as well.

[2]We had hoped to acquire similar data for the Homework words but because CB did not do the homework consistently and because she had no one at home to supervise her work and help organise the materials, we couldn't be certain of the accuracy of the homework data, nor of the conditions under which they were obtained.

The question addressed in this analysis is whether or not the differences between the Trained and Repeated words increase systematically with training. This is evaluated by means of a regression analysis that essentially determines the degree to which the accuracy differences between Trained and Repeated words are related to the evaluation number. Figure 4 reports the difference scores (difference in proportion correct) for Trained and Repeated word sets for each of the individual 13 evaluations (including two Pre-treatment evaluations). Also included in the figure is the best linear fit to these difference scores (R^2=.34). The positive slope of this line indicates that the differences in accuracy between Trained and Repeated words are increasing with training session—indicating a training benefit. The critical statistical question is whether or not the positive slope of this line is significantly greater than 0. In fact, the regression analysis indicates a clearly significant positive slope (F=5.57, p<.04), confirming the significant treatment effect established in Analysis 1. A number of variations on this analysis were carried out as well as evaluations of various characteristics of the data. These findings were incorporated into a more complex regression analysis (described in footnote 3). All additional analyses revealed significant findings, comparable to those reported just above.[3]

Maintenance of benefits and medium- to long-term treatment effects

Figure 5 reports the accuracy for each of the four stimulus conditions at Pre and Post-treatment as well as Follow-up 1 (6 months post treatment) and Follow-up 2 (12 months post treatment). It provides an overview of the entire intervention and the trajectories of all stimulus conditions.

Maintenance: From post-treatment to follow-ups.—The degree to which treatment benefits were *maintained* over the 12-month period following the end of treatment can be evaluated by comparing post-treatment accuracy levels with those at Follow-ups 1 and 2.

Comparisons of Post-treatment accuracy levels with those at Follow-up 1 reveal a numerical decline in performance for all stimulus categories. These changes were statistically significant for all categories except for Homework words—Homework: T=1365 (N=82) p<. 12, Trained: T=1287 (N=94) p<.0001, Control words: T=2281 (N=108) p<.04, Repeated words: T=1560 (N=93) p<.02. Despite this decline, 6 months after the end of treatment at Follow-up 1, Trained words still remained significantly more accurate than Control words, $\chi^2(1)$=6.18, p<.05, although they were no longer statistically more accurate than either Repeated, $\chi^2(1)$=0.97, *ns*, or Homework words, $\chi^2(1)$ =0 .01, *ns*. Furthermore, at Follow-up 1 both Homework and Repeated words continued to be more accurate than Control Words, although this difference was statistically significant only for the Homework set—

[3]The regression analysis can be carried out on the raw proportion differences depicted in Figure 5 or on statistically appropriate transformations of the data. For example, we also applied a logit transformation to the proportion scores obtained for each of the two kinds of word sets in each session. The logit transformation is the log-odds or logarithm of the odds ratio of the proportion of letters correctly identified. In this case, this provides us with transformed scores that are on the real line and, consequently, greater comfort with the assumptions in the linear model. We modeled the differences in the logit of the scores in terms of a simple linear regression set-up with time as the explanatory variable. The differences in log-odds ratios of the trained and repeated sets varied significantly with time (coefficient=0.0567, p=.0403). (In the context of the original untransformed data, this implies that the relative proportions of the odds ratios of the scores for the repeated and trained word sets increases exponentially by 0.0567 for every unit change in time.) The model reported an R-squared of 0.3293 and an adjusted R-squared of 0.2684. Detailed diagnostics of the residuals indicates that the set of scores for the second observation was both outlying as well as influential. Otherwise, the homoscedasticity and normal assumption for the errors in the differences of logit-transformed scores was reasonable. Given this, the analysis was redone with the second set of observations omitted. The main finding of a significant relationship over time (slope=0.0184, p=.0003) was strongly supported. The residuals have an autocorrelation coefficient of −0.329, so a model incorporating a first-order autocorrelation structure was also fitted, yielding significant results comparable to those of all previous analyses (slope=0.08903; p<.0001). Finally, a chi-squared test statistic evaluating the goodness of fit of the model incorporating the autocorrelation (χ^2=2.49) indicates that the first-order autoregressive structure for the model is not significant (p-value=.115). In other words, the previous model that did not include the autocorrelation was adequate.

Homework: $\chi^2(1)=5.60$, $p<.05$—with only a trend towards significance for the Repeated set —Repeated: $\chi^2(1)=2.27$, $p<.1$.

From 6 to 12 months after the end of treatment there continued to be a decline for all stimulus categories. All of these decreases were statistically significant except for the Control words—Control words: T=296.5 (N=36) $p<.6$, Trained: T=118.5 (N=35) $p<.002$, Repeated words: T=177 (N=38) $p<.006$, and Homework: T=841.5 (N=70) $p<.02$. Finally, by 1 year after the end of treatment at Follow-up 2, there were no statistically significant differences among the four word categories. In sum, there was clear evidence of the benefit of treatment, although the magnitude of the post-treatment benefit to Trained words had reduced by 6 months after treatment. This is indicated by the fact that at 6 months after the end of treatment, Trained words were still spelled significantly more accurately than Control words. Furthermore, both Homework and Repeated words maintained their advantage over Control words from post-treatment to Follow-up 1, although this difference was only significant for Homework words. Finally, no benefits of treatment were discernible for any of the word sets 12 months after the end of the treatment.

Medium- and long-term treatment benefits: From pre-treatment to follow-ups.—An evaluation of the medium- and long-term effectiveness of treatment was carried out by comparing *Pre*-treatment accuracy levels with levels at Follow-ups 1 and 2.

Compared to the pre-treatment accuracy levels, at 6 months after the end of treatment there were numerical declines in performance for all stimulus categories. These declines were statistically significant only for Control words, T=1849 (N=110), $p<.0005$, while accuracy levels for Homework words, T=1708.5 (N=93), $p<.07$, Repeated words, T=2248.5 (N=103) $p<.2$, and Trained words, T=1883 (N=94) $p<.2$, were statistically unchanged. This is evidence that, in the face of clear deterioration of untrained items, the training protocol helped to *maintain* the integrity of Trained, Homework, and Repeated items through the treatment period and out to 6 months beyond the end of treatment.

Compared to pre-treatment, at 12 months after the end of treatment there were significant declines in all four categories (all ps<.01). Therefore, despite the immediate (post-treatment) improvement for Trained words and medium-term maintenance of pre-treatment accuracy levels for Trained, Homework, and (to a lesser extent) Repeated words, there was no evidence of any treatment benefits by 1 year after the end of treatment.

Summary of results

The results can be summarised as follows:

1. The spell-study-spell treatment provided a modest (10% of letters), but statistically significant, advantage to words that are trained compared to words that were either untrained, practised in homework, or simply repeatedly tested.
2. The benefit for Trained words was fragile, such that by 6 months after the end of the study Trained words could not be statistically distinguished from Homework and Repeated items.
3. However, the intermediate range benefits of treatment manifested themselves at 6 months after the end of treatment in the maintenance of pre-treatment levels of accuracy for Trained words. This occurred in the face of clearly declining spelling abilities as indexed by deteriorating performance with Control words.
4. More modest benefits of homework practice or repeated testing were indicated by the greater stability in the accuracy levels of Homework and Repeated items

relative to control items in comparisons of pre-treatment with post-treatment and with the 6-month follow-up.

5. No generalisation effects could be documented.
6. No benefits of treatment could be discerned by one year after the end of treatment.

GENERAL DISCUSSION

We find both immediate and short-term benefits of behavioural intervention for dysgraphia in a case of primary progressive aphasia. Immediately following treatment, accuracy on Trained words increased relative to pre-treatment baseline and relative to all other control word types. Six months after the end of treatment, Trained words were protected from the significant deterioration experienced by untrained Control words. Repeated testing or practice at home yielded modest benefits, resulting in accuracies intermediate to those of the Trained and untrained Control words. These findings are among the first to provide empirical support for the potential of behavioural therapy to prolong language abilities in primary progressive aphasia. This occurred in a context of non-intensive training (typically 2 sessions/week) with overall relatively modest input (11 sessions).

The relationship between treatment in progressive and non-progressive disorders

A fundamental question in the study of PPA is the extent to which the response to treatment is similar or different in progressive compared to non-progressive disorders resulting from stroke, surgery, or head injury. Of course, in non-progressive disorders there may be changes in symptomatology subsequent to the neurological insult, but given that the aetiology is not degenerative the changes tend to be quantitative rather than qualitative. In previous work, Rapp and Kane (2002) and Rapp (2005) reported three individuals with acquired dysgraphia subsequent to stroke who were tested with essentially the same treatment protocol (without the Homework condition) as was used in the case of CB. Although none of the individuals had both the orthographic lexicon and graphemic buffering deficits that CB demonstrated, one (MMD) had a clear lexicon deficit and the other two (JRE, RSB) suffered from graphemic buffer deficits. These differences notwithstanding, it may be informative to consider the similarities and differences between CB's response to treatment and the responses of these individuals with non-progressive deficits. Figure 6 presents the results from MMD, JRE,[4] and CB. For MMD and JRE we include results from follow-ups at 5 and 10 months post treatment as these are the closest to CB's 6 and 12 month follow-ups.

First of all, Rapp and Kane (2002) and Rapp (2005) found that all individuals (regardless of deficit type) exhibited a benefit for Trained words. That is, all three significantly improved their performance for Trained words pre to post treatment, and Trained words were spelled more accurately than Repeated words at the end of training. Furthermore, this treatment benefit persisted until 10 months after the end of treatment, such that although performance decreased on the Trained words during this time period, accuracy on these words was still significantly better at 10-month follow-up than it had been prior to the onset of treatment. In this regard, CB's response to treatment was very similar to the individuals with non-progressive deficits. She too showed significant improvement for Trained words and the effects of this treatment were still evident at 6 months after the end of treatment. However, for CB, the benefits at the 6-month follow-up were manifested not in continued superior performance relative to pre-treatment but in a failure to exhibit a significant decline from

[4]Both JRE and RSB responded very similarly to treatment except that JRE's maintenance of benefits was more short-lived than RSB's; for the sake of brevity, we report only the results for JRE.

pre-treatment. In addition, for CB, the advantage of Trained over Control words at the 6-month follow-up occurred in the context of significantly deteriorating performance for Control words. For the individuals with non-progressive deficits, Control words either remained unchanged from pre-treatment (MMD) or they continued to show generalisation benefits at 5 months post-treatment (both JRE and RSB).

Second, the individuals with non-progressive deficits showed different generalisation effects that we attribute to their different underlying deficits. Specifically, both individuals with a graphemic buffer deficit (see JRE in Figure 6) showed generalisation effects manifested by improved performance for Control words from pre to post treatment. This generalisation benefit remained significant for JRE until 12 weeks after treatment while for RSB it was still apparent 2 years after the end of treatment. In striking contrast, MMD (with a lexicon deficit) showed no generalisation effect, as accuracy with control words remained virtually unchanged from pre to post treatment. Instead, she showed a significant benefit for the Repeated words such that these not only exhibited improvement relative to pre-treatment, but they were superior to Control words at post-treatment, and the benefits of repetition persisted until the 10-month follow-up. In the graphemic buffer cases, while there were significant improvements for Repeated words, these could not be distinguished from the generalisation effects, as Repeated words were not significantly better than Control words. In this regard, CB patterned more closely with MMD, showing no clear evidence of generalisation. In CB's case there was a significant decline in performance with Control words, while for MMD accuracy with the Control words remained stable across the course of the investigation. In addition, with respect to the effects of repetition, CB was similar to MMD in showing a trend for a benefit for Repeated words such that these were spelled consistently better than Control words at post-treatment and at the first follow-up. However, these similarities between CB and MMD are only suggestive given that: (a) a failure to generalise cannot be established in CB's case, and (b) the effects for Repeated words were not significant for CB and, therefore, clear benefits of repetition cannot be inferred.

In sum, overall CB responded quite similarly to the non-progressive cases, showing statistically significant and persistent benefits of training, as well as some evidence of benefits for repetition (and homework). These effects took place against a backdrop of generally declining performance, while for the non-progressive cases the backdrop was one of stability or even generalisation. In terms of absolute benefits, CB's 10% improvement for Trained words by post-treatment appears more modest than the 18–25% improvement of the non-progressive cases. However, if benefit is evaluated as the difference between Trained and Control words at post-treatment then CB shows a 16% benefit, which is quite close to that exhibited by the non-progressive participants. This may be the more appropriate measure as it takes into account the fact that the intervention takes place against the backdrop of deteriorating performance. In fact, the differences between Trained and Control words from post-treatment, through Follow-ups 1 and 2 for CB are 16%, 12%, and 0% respectively, and for MMD these are quite comparable at 17%, 13%, and 6%.

There are also certain differences between CB and the non-progressive cases that are worth discussing. The most obvious difference is the deteriorating performance of Control words across the entire intervention period. In addition there is the deterioration for all word classes from pre-treatment to 12 months post-treatment. More subtle differences include the fact that during the treatment sessions, CB required a greater number of repetitions of each item. She averaged 1.8 repetitions, while the individuals with non-progressive deficits averaged only .22 to .45 repetitions. Furthermore, non-progressive participants received training until reaching the criterion of 95% accuracy or better for two consecutive sessions. CB was never able to achieve these accuracy levels; instead training was discontinued when accuracy for Trained words stabilised between 70% and 75% of letters correct. Nonetheless, for both groups, stable effects were achieved with roughly comparable numbers of training sessions (11 for CBD, 11–20 for the non-progressive cases). Overall, therefore, CB's ability to relearn the spellings of words or recover some of her ability to access these words seemed roughly comparable to that of individuals who did not suffer from neurodegenerative disease, yet there is clear evidence from the number of repetitions required, the level of accuracy at which asymptotic performance is reached, and the fragility of the training benefits, that CB's relearning was hampered by the neurodegenerative disease.

Costs, benefits, and other considerations

Therapy is costly in terms of time and financial resources, and therefore it is important to weigh the costs and benefits of any therapy approach. Unfortunately, it is of course difficult to evaluate cost/benefit ratios in the best of circumstances, and clearly not possible with the limited data from this study. Nonetheless, it may be worthwhile to consider some of the relevant issues.

This study serves as a type of "proof of concept" study demonstrating that it is possible to get improvement and "protection" from degeneration for specific words that are trained. It is clear that the degree of improvement or protection conferred on the Trained words is not sufficient to make an impact on CB's quality of life. This is not only because the benefits were quantitatively modest, but also because the words that were selected for treatment were not words that were important to CB's everyday life (words such as CANDLE, TOUGH, BACTERIA). However, the study does show that, even under these less than optimal functional circumstances, measurable and significant benefits can be obtained. Certainly it will be important to evaluate the degree to which effectiveness of training is affected by the personal relevance of the items, as well as by whether or not the training itself explicitly works to integrate the words into activities of daily living.

In the context of evaluating the benefits of this treatment, one might also question the wisdom of expending resources on written rather than spoken language rehabilitation. One thing that is relevant in this regard is that, in the context of non-progressive deficits, it has been sometimes reported that for certain individuals with both spoken and written language impairments, remediation of the written language deficits is sometimes more successful than remediation of the spoken deficits (Beeson, 1999; Robson et al., 2001). This is not entirely surprising given the fairly extensive evidence showing the independence of a number of the cognitive mechanisms involved in written and spoken language production. That is, there have been a number of cases of individuals who can write words they cannot say, and vice versa (for a review see Rapp et al., 1997). In the context of progressive aphasia we noted in the Introduction that it has been reported that, for some individuals, written language remains a relative strength and they therefore rely on this for communication (Cress & King, 1999; Holland et al., 1985; Kertesz et al., 1994; Mesulam & Weintraub, 1992; Murray, 1998; Weintraub et al., 1990). Cress and King (1998) specifically remarked on relative preservation of reading in the course of PPA, at least in with regard to overlearned written materials. They state (p. 257):

> It is important to determine whether people with PPA can successfully overlearn critical written information during periods of better reading and language skills or whether this skill is restricted to highly familiar environmental writing…. if deliberate practice of specific written vocabulary can keep those vocabulary items in active use, then overlearning of essential information may be an effective strategy in maintaining functional reading and recognition skills for persons with PPA [italics added].

Given the findings we have reported here indicating that learning of written material is possible in cases of PPA, it will be important for future research to examine the role that the timing of the intervention plays in the development and strengthening of skills and representations that will be durable in the face of the degenerative process.

Conclusions

In sum, we have reported evidence that the relearning, strengthening, or improved accessibility of word spellings are possible in a case of primary progressive aphasia. This recovery, at least superficially, resembles what is observed in cases of non-progressive dysgraphia. Certainly, however, the underlying nature and mechanisms of the improvement are not understood in any of these cases and this represents yet another area that will require future investigation. However, what is most encouraging is that the benefits provided by the training allow the targeted words to remain available for use for a longer period of time. The study raises a number of questions regarding the best means of harnessing this learning capacity so as to best serve the needs of individuals with neurodegenerative language disorders.

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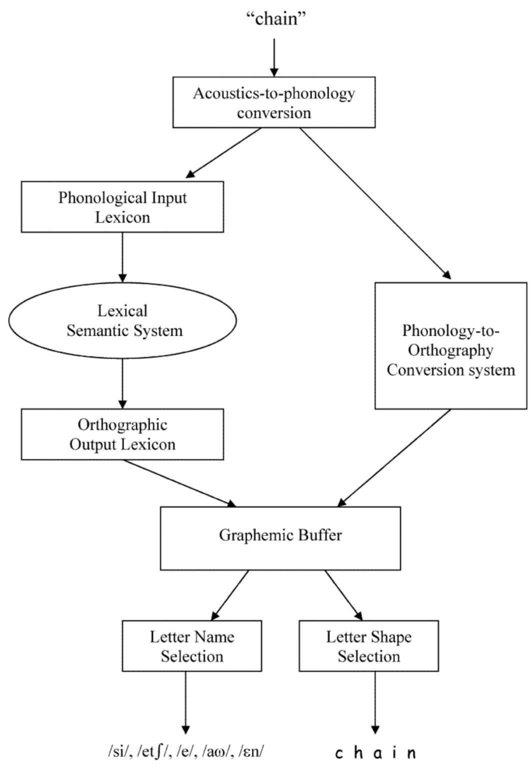

Figure 1.
The cognitive mechanisms involved in written (or oral) spelling to dictation. Adapted from Buchwald and Rapp (2006).

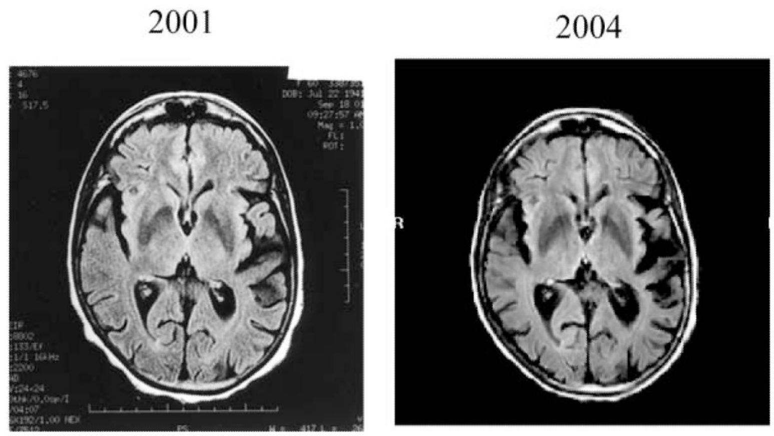

Figure 2.
MRI scans taken of CB in 2001 and 2004, showing asymmetric temporal parietal atrophy.

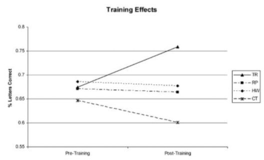

Figure 3.
Percent letters correct at pre and post training (combined over two administrations of each list at each time point). TR=trained list, RP=repeated list, HW=homework list, and CT=control list (*n*=228 letters for each word list at each time point).

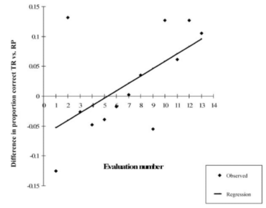

Figure 4.

Differences in proportions correct on Trained (TR) minus Repeated (RP) word sets at each of 13 evaluations (from baseline to post-training) are represented with diamonds. Also included is the best linear fit to the data (slope=.012, F=5.57; p<.04).

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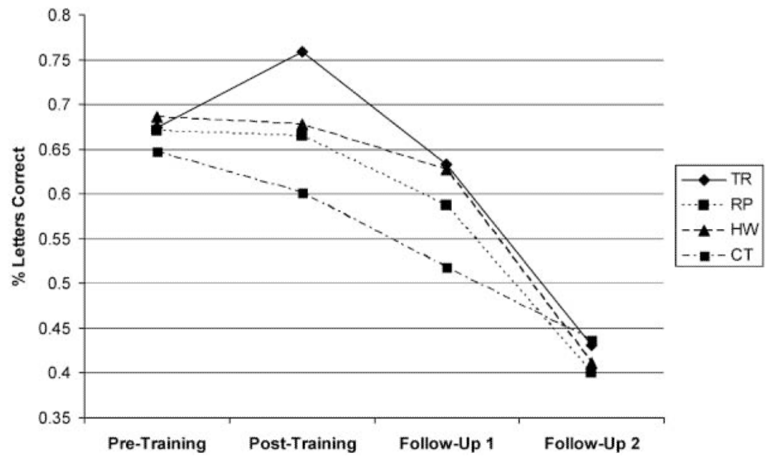

Figure 5.
Accuracy for all word sets (TR=trained, RP=repeatedly tested, HW=homework, and CT=control (*n*=228 letters for all sets at all time points, except Follow-up 2 where *n*=58 letters for TR and RP and *n*=56 for HW and CT).

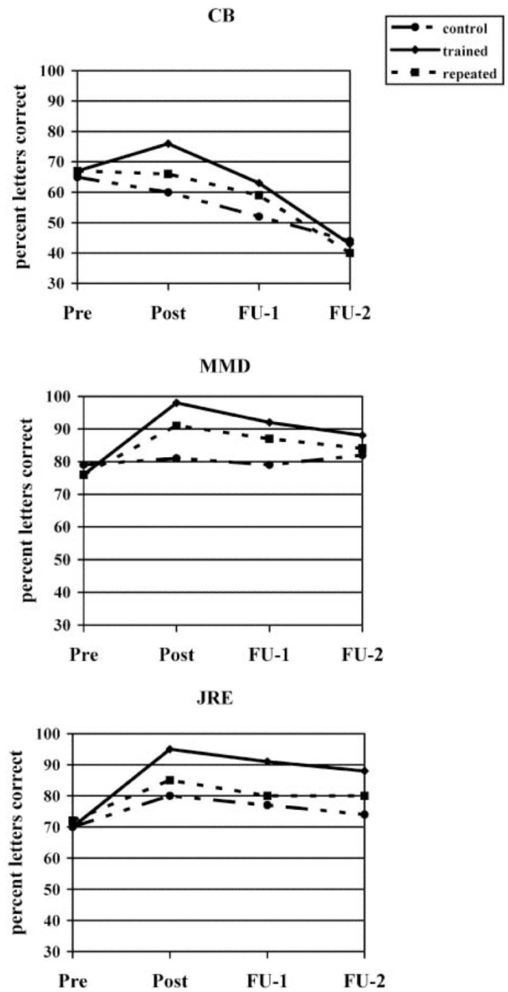

Figure 6.

Letter accuracy at Pre-intervention, Post-intervention, Follow-up 1, and Follow-up 2 for CB, MMD, and JRE (details regarding MMD and JRE’s data are available in Rapp, 2005, and Rapp & Kane, 2002).

TABLE 1

Timeline of the major events referred to in this investigation

Date	*Evaluation/Observations*
1996	Earliest symptoms of language difficulties
Dec 2001	*Neuropsychological Evaluation 1:* Mild cognitive impairment
Dec 2003	*Neuropsychological Evaluation 2:* Diagnosis of PPA
Jan–April 2004	*Pre-intervention Evaluation*
May–Aug. 2004	*INTERVENTION STUDY*
Sept 2004	*Neuropsychological Evaluation 3:* Further cognitive deterioration
Dec–Feb 2005	*Post-Intervention Evaluation and Follow-up 1*
July 2005	*Post Intervention Follow-up 2*

TABLE 2

CB's performance on a set of neuropsychological tests at three time points

	12/2001	*12/2003*	*9/2004*
MMSE (Mini Mental Status Examination)			
[29 (1.3)]	26/30 (Z=−2.31)	26/30 (Z=−2.31)	20/30 (Z=−6.92)
WMS (Wechsler Memory Scale)			
Information and Orientation [13.5 (0.6)]	13/14 (Z=−0.83)	14/14 (Z=.83)	12/14 (Z=−2.5)
Grooved Pegboard Test			
Dominant	13%ile	<1%ile	<1%ile
Nondominant	12%ile	<1%ile	<1%ile
Trail making			
A [35.8 (11.9)]	53 sec	98 sec	93 sec
B [81.2 (38.5)]	166 sec	discont @ 300s	discont@ 200s
Rey Auditory Verbal Learning Test (RAVLT)			
Total [49.4 (7.5)]	28/75 (Z=−2.85)	27/75 (Z=−2.98)	12/75 (Z=−4.98)
Delayed recall [10.2 (2.5)]	8/15 (Z=−0.88)	8/15 (Z=−0.88)	3/15 (Z=−2.88)
WMS (Wechsler Memory Scale)			
Digits forward span [8.4 (1.9)]	7	7	5
Backward span [6.5 (2.0)]	4	3	2
Warrington Recognition Memory Test	68th %ile		
Words	25th %ile	–	–
Faces			
Rey-Osterreith Complex Figure			
Direct [30 (4.21)]	33/36 (Z=.71)	27/36 (Z=−.71)	26/36 (Z=−0.95)
Immediate [16.57 (7.53)]	11.5/36 (Z=−.67)	12.5/ 36(Z=−0.54)	14/36 (Z=−0.34)
Delayed [14.21 (7.5)]	–	12/36 (Z=−0.29)	15/36 (Z=0.11)
Stroop			
Color	Z=−4.39	Z=−104	–
Color word	Z=−4.61	Z=−4.34	–
Boston Naming Test [55.6 (3.5)]	25/30	48 /60 (−2.17)	–
Controlled Oral Word Association Test (F-A-S)	37 words	22 words	5 words
[42 (12.1)]	(Z=−0.41)	(Z=−1.67)	(Z=−3.05)
Noun/verb picture naming			
spoken: N/V	–	87% / 90%	87% / 83%
written: N/V		53% / 47%	27% / 10%

Wherever possible scores are reported as Z scores or percentiles. Numbers in [ ] correspond to the mean score of normal subjects and their (SD). MMSE (Folstein et al., 1975); WMS (Wechsler & Stone, 1973); Grooved Pegboard (Trites, 1984); Trail making (Reitan, 1992); RAVLT (Schmidt, 1996); WMS (Wechsler et al., 1973); Warrington Recognition Memory Test (Warrington, 1984); Boston Naming Test (Kaplan et al., 1983); Rey-Osterreith Complex Figure (Osterreith, 1942; Rey, 1942); Controlled Oral Word Association Test (Benton & Hamsher, 1976); Stroop (adapted from Golden & Freshwater, 2002); Noun/verb picture naming (unpublished).

TABLE 3

Results of additional assessment of language and other cognitive functions

	Pre-training (12/03-4/04)	*Post-training (9/04-10/04)*	*6 month Follow-up 1 (12/04-2/05)*	*12 month Follow-up 2 (7/05-9/05)*
Language comprehension				
Spoken				
-Word (PPVT) (*n*=175)	68%ile	63%ile	68%ile	19%ile
-Sentence (JHU Screener) (*n*=26)	96%	92%	-	77%
Written				
-Visual Lexical Decision (PALPA 25) (*n*=120)	96%	85%	-	80%
Language Production (JHU Language Screeners)				
Spoken				
-Picture naming (*n*=41)	83%	59%	-	-
-Oral reading (*n*=35)	83%	71%	-	-
-Single word repetition (*n*=15)	100%	93%	-	93%
-Sentence completion (*n*=10)	100%	90%	-	90%
Written *				
-Spelling to dictation				
-words (*n*=79)	15%	13%	-	-
-nonwords (*n*=33)	39%	15%	-	-
Visuo-spatial				
-BORB-length match (*n*=30)	90%	93%	-	-
-BORB-object recognition (*n*=62)	98%	100%	-	-
-BVRT	-	100% / 20%	-	97% / 23%
(direct copy/15 sec delay) (*n*=20)				

PPVT=Peabody Picture Vocabulary Test (Dunn & Dunn, 1981); BORB=Birmingham Object Recognition Battery (Riddoch & Humphreys 1993). JHU=Johns Hopkins University Language Screeners (unpublished-different items are used in different modalities; lists include a range of word frequencies and lengths); PALPA=Psycholinguistic Assessment of Processing in Aphasia (Kay et al., 1992); BVRT=Benton Visual Retention Test (Benton Sivan, 1991).

* Spelling accuracy is reported as: # of correct strings/total strings attempted (rather than as letters correct) so as to be more comparable to other word tasks in this table.

TABLE 4

CB's performance across a range of single-letter tasks both pre and post intervention

Task	*Upper/Lower Case*	*Pre-Training*	*Post-Training*
Direct Copy	LC	100% (26/26)	100% (26/26)
	UC	100% (26/26)	100% (26/26)
Naming	LC	96% (25/26)	92% (48/52)
	UC	100% (26/26)	94% (49/52)
Direct Copy Transcoding	UC to LC	92% (48/52)	89% (23/26)
	LC to UC	90% (47/52)	89% (23/26)
Writing to Dictation	LC	92% (48/52)	90% (47/52)
	UC	98% (51/52)	98% (51/52)

UC=upper case; LC=lower case; *n*=26 includes the 26 letters of the alphabet evaluated once; *n*=52 includes the 26 letters evaluated twice.

TABLE 5

Distribution of CB's error types in word lists administered for spelling to dictation during pre-training evaluations

	Pre-Training
Phonologically plausible	11% (5/44)
Visually Similar Word	14% (6/44)
Visually Similar Nonword	55% (24/44)
Semantic	0%
Morphological	0%
Other	21% (9/44)

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TABLE 6

Average (Avg) and standard deviation (in parentheses) for length in letters, concreteness and frequency for words in the four experimental lists

	Avg Length	*Avg Concreteness*	*Avg. PG probability*	*Avg Frequency*
Trained	5.7 (1.34)	496.85 (112.3)	.69 (.18)	43.15 (60.7)
Repeated	5.7 (1.34)	509 (104.0)	.73 (.14)	42.9 (60.6)
Homework	5.7 (1.34)	491.1 (121.3)	.71 (.17)	44.85 (68.7)
Control	5.7 (1.34)	510.1 (107.4)	.70 (.13)	41.5 (53.9)

Avg. PG probability corresponds to average phoneme–grapheme probability computed as an average of every PG mapping in each list based on the values in Hanna et al. (1996).

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TABLE 7

CBD's written responses to Trained words at the first pre-treatment evaluation and at the final post-treatment evaluation

Target	*PRE*	*POST*
DIME	DINE	√
MAZE	MASE	MAC
BONE	√	√
BOMB	DOOM	BOME
PAPER	PAPAR	√
TWIST	TIST	√
GHOST	SO	GOST
FLAME	FASME	FALM
TOUGH	THOUT	TUS
PIANO	PION	PIPO
CANDLE	CANDLED	DANLL
SHIELD	SHERD	SHEID
PHRASE	CA	PRAC
TALENT	TALLANT	TALINT
SKETCH	SKECK	SKECK
JOURNAL	GO	JORNE
ANTIQUE	ANTIC	ANTQUE
QUESTION	√	√
BACTERIA	BATINC	BTERI
ORDINARY	ONR	√