Verbal mediation of number knowledge: Evidence from semantic dementia and corticobasal degeneration

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Abstract

Patients with corticobasal degeneration (CBD) appear to have impaired number knowledge. We examined the nature of their number deficit while we tested the hypothesis that comprehension of larger numbers depends in part on verbal mediation. We evaluated magnitude judgments and performance on number conservation measures rooted in Piagetian theory in nonaphasic patients with CBD (n = 13) and patients with a fluent form of progressive aphasia known as semantic dementia (SD; n = 15). We manipulated the numbers of the arrays and the visual–spatial properties of the stimuli being compared during magnitude judgments and Piagetian conservation measures. CBD patients were consistently impaired judging the magnitudes of larger numbers (4–9), while they had minimal difficulty with smaller numbers (magnitudes ≤3). By comparison, SD patients performed all measures of number knowledge at a ceiling level regardless of number magnitude. Neither patient group was significantly impacted by manipulations of the spatial properties of the stimuli. CBD patients’ impairment with larger numbers despite minimal aphasia, and SD patients’ intact performance despite an aphasia, challenge the proposal that understanding larger numbers depends on verbal mediation.

Keywords: Corticobasal; Frontotemporal; Semantic dementia; Number; Calculation

1. Introduction

Number knowledge is a semantic domain that supports understanding quantities such as “five” independent of the objects (e.g., dots, dolls, books, etc.) or symbols (e.g., 5, V) that happen to be used to illustrate and represent “fiveness.” One hypothesis holds that adult humans possess an analogue representation for number magnitude in the form of a mental number line or “ruler” (Dehaene, 1997). This “mental ruler” tends to compress larger numbers into a progressively smaller, less differentiated space like a logarithmic scale where equal space is allocated to the interval between the numbers 1 and 2, between 2 and 4, between 4 and 8, etc. This property results in decreasing accuracy and speed during magnitude estimations and calculations across the entire set of numbers as the target number increases in magnitude (Dehaene et al., 1998). In order to use larger numbers in a precise way, these investigators argue that they must be mediated by a verbal system. Accordingly, without intact language functioning, we are forced to approximate magnitude based on the perceptual–spatial properties of an array. Occasional observations, however, appear to be inconsistent with this approach.
Some aphasic patients have been reported with preserved number knowledge and calculation skills despite their language impairment (Thioux et al., 1998; Warrington, 1982). Moreover, a small number of patients has been described who suffer from a profound loss of number knowledge and acalculia without aphasia (Delatolas, Deloche, Basso, & Claros-Salinas, 2001; Lucchelli & De Renzi, 1993; Rossor, Warrington, & Cipolotti, 1995; Whetstone, 1998).

In the present study, we attempted to determine more directly whether number knowledge depends in part on verbal mediation by investigating whether patients with corticobasal degeneration (CBD) are relatively more impaired on tasks requiring knowledge of larger numbers (4–9) compared to smaller numbers (1–3). We reported previously that patients with CBD are significantly impaired in their calculation performance relative to patients with Alzheimer’s disease and frontotemporal dementia (Halpern et al., 2003; Halpern et al., 2004). Importantly, aphasia is said to be relatively modest in CBD (Rinne, Lee, Thompson, & Marsden, 1994; Snowden, 1994; Stover & Watts, 2001), involving primarily some naming difficulty (Grossman et al., 2004). Dehaene (1997) has argued that the comprehension of larger numbers is verbally mediated. To the extent that CBD patients are impaired with larger numbers despite the absence of aphasia, it becomes difficult to argue that comprehension of larger numbers is verbally mediated.

To assess the role of language in number knowledge from another perspective, we also examined the performance of patients with semantic dementia (SD). SD is a rare neurodegenerative condition where naming and comprehension of familiar words and objects is compromised (Hodges, Patterson, Oxbury, & Funnell, 1992; Snowden, Goulding, & Neary, 1989). Despite difficulty with word and object knowledge, two case studies (Cappelletti, Butterworth, & Kopelman, 2001; Diesfeldt, 1993) and one group study (Halpern et al., 2004) revealed preserved number knowledge in SD. Preserved knowledge of larger numbers in SD would provide additional evidence against the claim that knowledge of larger numbers depends on verbal mediation.

A related issue concerns the visual perceptual–spatial properties of an object array. Without verbal mediation, Dehaene (1997) suggests that number representations may be influenced by the perceptual–spatial properties of the array. Under these circumstances, faulty verbal representations of numbers may result in reliance on the spatial extent of the materials rather than the numerosity of an array of stimuli. To assess whether numerosity is inappropriately influenced by the spatial magnitude of the stimuli, we manipulated the spatial dimensions of the stimuli. We thus sought to determine whether patients with impaired number knowledge attend to the area of stimuli during an assessment of the relative magnitude of two arrays rather than the number of the stimuli.

2. Methods

2.1. Subjects

We examined 13 patients who were diagnosed with corticobasal degeneration (CBD) and 15 patients diagnosed with semantic dementia (SD). These patients also participated in our previous study (Halpern et al., 2004). Demographic features of these patients are summarized in Table 1. All patients were identified in the out-patient clinic of the Department of Neurology at the University of Pennsylvania Medical Center. The clinical diagnosis was made by a board-certified neurologist with expertise in the diagnosis of dementing conditions. We are unaware of any published consensus criteria for the clinical diagnosis of CBD, although experts have suggested specific clinical features that may be important in diagnosing CBD (Riley & Lang, 2000). The criteria we developed, based on a review of literature concerned with clinical–pathological diagnosis of CBD, include the insidious onset and gradual progression of: cortical sensory deficit, apraxia, gait instability and axial rigidity, unilateral alien hand, and/or asymmetric extrapyramidal features such as myoclonus, dystonia, and rigidity, but little resting tremor. All CBD patients presented with extrapyramidal motor features that were somewhat bilateral, including 9 of 13 CBD patients with extrapyramidal motor features primarily on the left, 3 of 13 patients with extrapyramidal features more prominently on the right, and 1 CBD patient with fairly symmetric involvement. We showed previously that these patients have mild naming difficulty (Halpern et al., 2004). There was no evidence for an ocular motility disorder, and these patients did not show evidence of neglect. However, these patients were more impaired in their visual–spatial judgments of line orientation than SD patients [U = 17.5; p < .005] (Benton, 1978).

The clinical diagnosis of SD was based on the identification of patients with Frontotemporal Dementia (FTD) using published criteria (The Lund & Manchester Groups, 1994; McKhann et al., 2001). Based on the con-

Table 1
Means and standard deviations for age, years of education, and MMSE in semantic dementia patients and corticobasal degeneration patients

<table>
<thead>
<tr>
<th></th>
<th>Semantic dementia</th>
<th>Corticobasal degeneration</th>
</tr>
</thead>
<tbody>
<tr>
<td>N</td>
<td>15</td>
<td>13</td>
</tr>
<tr>
<td>Age (year)a</td>
<td>67.79 ± 9.1</td>
<td>66.46 ± 10.2</td>
</tr>
<tr>
<td>Gender</td>
<td>8 M; 6 F</td>
<td>7 M; 6 F</td>
</tr>
<tr>
<td>Education (year)b</td>
<td>16.46 ± 3.4</td>
<td>15.15 ± 4.6</td>
</tr>
<tr>
<td>MMSEb score (max = 30)</td>
<td>22.64 ± 3.8</td>
<td>19.62 ± 5.1</td>
</tr>
</tbody>
</table>

a T tests showed that patient groups did not differ for age [t(26) = 0.72; ns] or education [t(26) = 0.83; ns].
b T tests showed that patient groups did not differ for MMSE (Mini Mental Status Examination) [t(26) = 1.95; ns].
sensus evaluations of a semi-structured interview, detailed mental status evaluation, and clinical neurological assessment, we divided the group of FTD patients into clinical subgroups guided by specific criteria (Davis, Price, Moore, Campea, & Grossman, 1998; Grossman & Ash, 2004; Neary et al., 1998; Price, Davis, Moore, Campea, & Grossman, 2001). SD, one subgroup of FTD, demonstrates fluent, circumlocutory speech with frequent word-finding pauses, paraphasias, and naming and comprehension difficulty for single words.

Other causes of dementia were excluded by history, physical exam, serum studies, and structural brain imaging. Structural and functional neuroimaging (when available) were consistent with the clinical diagnosis in all patients. We assessed overall dementia severity with the Mini Mental State Examination (MMSE) (Folstein, Folstein, & McHugh, 1975), a general overview of cognitive functioning that evaluates orientation, anterograde memory, language, executive functioning, and visual construction on a 30-point scale. Although most of the patients were taking cholinergic-supplementing agents (e.g., donepezil and rivastigmine) and some were taking modest dosages of serotonin-specific re-uptake inhibitor anti-depressants (e.g., sertraline and paroxetine), the patients were not taking sedating medication at the time of assessment. This study was approved by the IRB of the University of Pennsylvania, and all patients and caregivers participated in the Informed Consent procedure.

2.2. Materials

A pencil and paper formatted series of tests was presented to all patients to evaluate number knowledge.

2.2.1. Dot array magnitude comparison

Patients saw 36 paired sets of filled, black, 1 cm circles or “dots” in boxes aligned vertically and presented in a fixed, random order. The pairs were divided into six pairs of small numbers (1–3), six pairs of medium-sized numbers (4–6), and six pairs of large numbers (7–9). In addition, we presented patients with mixed conditions, including six pairs of small and medium-sized numbers, six pairs of small and large numbers, and six pairs of medium-sized and large numbers. To assess the performance of CBD patients and SD patients on all tasks of number knowledge in a manner sensitive to magnitude, we split the problem set in half by categorizing all conditions involving small numbers as “smaller,” including mixed cases of small and medium numbers and small and large numbers. The remaining conditions, involving medium-sized and large numbers were categorized as “larger.” The larger number of each pair was presented equally often in the top or bottom box.

We also presented patients with 36 pairs of boxes aligned vertically and filled with different numbers of dots of unequal size. One box contained an array of smaller dots (1 cm diameter) and the other box contained an array of dots that were physically larger in size (2 cm diameter). Half of these paired arrays, which we name the “divergent” stimuli, consisted of smaller numbers of larger dots and larger numbers of smaller dots. The remainder of these array pairs, which we name the “congruent” stimuli, consisted of larger numbers of larger dots and smaller numbers of smaller dots. Patients were asked to determine which box contained the greater number of dots.

2.2.2. Number conservation

We administered a modified version of a Piagetian measure of number conservation (Piaget, 1964). Patients were presented with two sets of equally numbered poker chips (3 cm diameter) that were arrayed linearly and parallel to each other with 4 cm separating each chip. We presented patients with sets containing a smaller number of chips (3), a medium-sized number of chips (6), and a larger number of chips (9). An initial equivalence between the two equally numbered sets was established by pairing the items in each set when arrayed linearly opposite each other. Then, we added (2 trials for each number) or subtracted (2 trials for each number) one poker chip to or from one of the sets while maintaining an equal spacing between chips within each set, and the equality of the overall length across the two sets. We asked “Do these two sets still contain an equal number of chips?” As a control condition, we lengthened (2 trials) or shortened (2 trials) one of the sets of poker chips by 4 cm and asked “Do these two sets still contain an equal number of chips.” These judgments were presented in a fixed, random order.

2.2.3. Volume conservation

We administered a modified version of a Piagetian volume conservation task (Piaget, 1964). We first established an equivalence between two equal volumes (water height = 9 cm, diameter = 7.75 cm, volume = 424 cm³) of water in transparent glass containers (container height = 12 cm, diameter = 7.75 cm, and total possible volume = 566 cm³). The examiner then emptied one container into a shorter (container height = 5 cm) but much wider (diameter = 17.5 cm) glass container (total possible volume = 1202 cm³). The water height in the wider container was 4 cm. Patients were asked: “Do the two containers still contain the same volume of water?” We also presented patients with a task in which we first established initial equivalence between two equal volumes of water (water height = 4.5 cm, diameter = 7.75 cm, volume = 212 cm³) in two equally filled, transparent containers (container height = 12 cm, diameter = 7.75 cm, total possible volume = 566 cm³) of water, and then we poured the water of one container equally divided into two smaller and thinner (container
height = 7.0 cm, diameter = 5 cm, total volume = 137 cm$^3$) containers. Because of the shape of the smaller containers, the height of the water was higher above the table surface (6.5 cm) than it was in the original, larger container. We then asked: “Do these two containers together contain the same amount of water as the original container?” We administered another version of this task by pouring a glass container (total possible volume = 522 cm$^3$, height of water = 8 cm, diameter = 7.75 cm, volume = 401 cm$^3$) into four smaller glass containers (container height = 7.0 cm, diameter = 5 cm, total possible volume = 137 cm$^3$). We asked: “Do these four containers contain the same amount of water as the original container?” These trials were presented in a fixed, random order. We combined performance on these three tasks and report patient overall accuracy.

2.3. Statistical analysis

Nonparametric statistical tests were used to test the null hypothesis that differences in number knowledge do not exist between SD patients and CBD patients. We used non-parametric statistics because SD patients were at or near ceiling for all measures of numerosity and quantity knowledge. We also performed analyses of individual patient performance profiles by converting each patient’s accuracy for each task to a $z$ score based on the performance of the entire patient sample.

3. Results

3.1. Dot array magnitude comparisons

3.1.1. Equally sized dots

A Friedman Analysis of Variance (ANOVA) by ranks with a group (2-SD, CBD) × dot array size (2-smaller, larger) design revealed that SD patients and CBD patients differ in their performance depending on the number of dots in the array [$\chi^2(1) = 9.31; p < .002$]. Fig. 1 summarizes these findings. Between-group comparisons showed that CBD patients are more impaired for conditions involving larger arrays of dots relative to SD patients [$U = 28.50; p < .001$], although these patient groups did not differ for smaller arrays of dots [$U = 88.00; ns$]. Within-group comparisons, using a Wilcoxon Signed Ranks test, revealed an impairment in CBD patients for conditions involving larger numbers relative to conditions involving smaller numbers [$Z = 2.81; p < .005$]. SD patients did not show this difference. Visual–spatial judgments did not correlate with magnitude judgments in CBD. Moreover, between-group comparisons using Mann-Whitney $U$ tests showed that CBD patients with more prominent right-sided clinical features (greater left hemisphere disease) were equally impaired on measures of number knowledge compared to CBD patients with more prominent left-sided clinical features (greater right hemisphere disease).

Analyses of individual patient performance confirmed these group findings. For larger numbers, we found that 10 (77%) of 13 individual CBD patients encountered clear difficulty (i.e., multiple errors), while 13 of 15 SD patients were perfect in their performance and only 2 (13%) of 15 SD patients made a single error. Using the $z$ scores of patient performance accuracy to identify abnormal performance (at the $p < .05$ level) relative to the entire patient sample, we found a significant difference in the proportions of impaired CBD patients compared to SD patients for larger numbers [$\chi^2(1) = 11.37; p < .001$]. However, only 2 (15%) of 13 individual CBD patients and only 1 (7%) of 15 SD patients made a single error when judging smaller numbers on the dot array magnitude comparison task. These proportions did not differ, according to $z$ scores of patient performance accuracy. These findings suggest that CBD patients have more difficulty judging larger arrays than smaller arrays, although SD patients with an aphasia were quite unimpaired in this task.

3.1.2. Unequally sized dots

A Friedman Analysis of Variance (ANOVA) by ranks of the divergent arrays of unequally sized dots with a group (2-SD, CBD) × dot array size (2-smaller, larger) design revealed that SD patients and CBD patients differ in their performance depending on the numerosity of the arrays [$\chi^2(1) = 5.44; p < .02$]. Fig. 2 summarizes these findings. Between-group comparisons revealed a difference for the larger arrays but not the smaller arrays.
Within-group comparisons revealed an impairment in CBD patients for larger arrays relative to smaller arrays of dots ($Z = 1.90; p < .058$), but SD patients did not show this difference. Thus, spatial manipulations of dot arrays did not impact patient performance.

Individual analyses revealed that 9 (69%) of 13 CBD patients were clearly impaired (i.e., multiple errors) for larger-numbered divergent stimuli, whereas 13 of 15 SD patients were perfect and only 2 (13%) of the SD patients made a single error. Using the $z$ scores of patient performance accuracy to identify abnormal performance (at the $p < .05$ level), we found a significant difference in the proportions of impaired CBD patients compared to SD patients on this task [$\chi^2(1) = 6.48; p < .01$]. However, we found that only 4 (31%) of 13 individual CBD patients made a single error and not one SD patient made a single error when judging smaller-numbered stimuli, and these proportions did not differ statistically.

A Friedman Analysis of Variance (ANOVA) by ranks of the congruent arrays of unequally sized dots with a group (2-SD, CBD) x dot array size (2-smaller, larger) design demonstrated that SD patients and CBD patients differ in their performance depending on the numerosity of the arrays [$\chi^2(1) = 4.46; p < .04$]. Fig. 2 summarizes these findings. Between-group comparisons revealed a difference for the larger arrays but not the smaller arrays. Within group comparisons revealed an impairment in CBD patients for larger arrays of dots relative to smaller arrays of dots [$Z = 2.10; p < .04$], but SD patients did not show this difference.

Individual analyses revealed that, for larger numbers, 9 (69%) of 13 CBD patients were clearly impaired (i.e., multiple errors) for the congruent stimuli, whereas 14 of 15 SD patients were perfect and only 1 (7%) of the SD patients made a single error. Using the $z$ scores of patient performance accuracy to identify abnormal performance (at the $p < .05$ level), we found a significant difference in the proportions of impaired CBD patients compared to SD patients on this task [$\chi^2(1) = 9.67; p < .01$]. However, we found that only 4 (31%) of 13 individual CBD patients made a single error and not one SD patient made a single error when judging smaller-numbered stimuli, proportions that did not differ statistically.

Within-group comparisons across the three magnitude judgment tasks, including equally sized dots, divergent sets of unequally sized dots, and congruent sets of unequally sized dots, revealed that the performances of SD patients and of CBD patients did not differ, according to Wilcoxon Signed Ranks tests. CBD patients were equally accurate with smaller arrays under all conditions and were equally impaired with larger arrays under all conditions. SD patients were equally accurate across all conditions regardless of array size. This provides additional evidence suggesting that CBD patients were uninfluenced by the spatial properties of the stimuli, despite their number deficit.

### 3.2. Piagetian measures of number conservation

We used a Friedman ANOVA with a group (2-SD, CBD) by set size (3-smaller, medium, larger) design to compare patient performance when presented with two sets of equal length containing different numbers of poker chips. This revealed a significant difference between CBD patients and SD patients depending on the number of the array [$\chi^2(2) = 10.97; p < .004$]. Mann–Whitney $U$ tests showed that CBD patients are more impaired than SD patients for mid-sized sets [CBD = $83.33 \pm 19.1\%$ correct; SD = 100% correct; $U = 30.0; p < .01$] and larger-numbered sets [CBD = $69.05 \pm 24.9\%$ correct; SD = $98.81 \pm 4.1\%$ correct; $U = 26.0; p < .01$], although CBD patients (88.10 ± 19.1% correct) did not differ from SD patients (100% correct) for smaller-numbered sets of poker chips [$U = 42.0; ns$]. Within-group comparisons, using Wilcoxon Signed Ranks tests, revealed that CBD patients are more impaired for larger sets relative to smaller sets [$Z = 2.40; p < .02$] and mid-sized sets [$Z = 2.21; p < .03$]. Individual patient analyses revealed that 9 CBD patients make many errors, while 14 of 15 SD patients made no errors and only 1 SD patient made a single error. Using the $z$ scores of patient performance accuracy on this task, we found a significant difference in the proportions of impaired CBD patients (75% impaired) and SD patients (8.33% impaired) [$\chi^2(1) = 7.81; p < .01$].

A Friedman ANOVA with a group (2-SD, CBD) by set size (3-smaller, medium, larger) design was used to compare patient performance when presented with two equally numbered sets of poker chips that are of unequal lengths. SD patients (100% correct for all cases) and CBD patients (smaller = 91.67% correct;
patients differ only marginally from SD patients on the 3.3. Piagetian measures of volume conservation

Between-group comparisons revealed that CBD patients differ only marginally from SD patients on the three volume tasks \( U = 41.5; p < .08 \). While a majority of individual CBD patients were impaired on all tasks involving enumeration, we found that only 6 of 13 CBD patients make errors on these tasks involving volume judgments. Not one SD patient made a single error. A Chi-Square test was used to assess the relative impairment of individual CBD and SD patients. Using the \( z \) scores of overall accuracy for all of these tasks combined, we found a significant difference in the proportions of impaired CBD patients and SD patients \( \chi^2(1) = 5.10; p < .05 \).

4. Discussion

Scattered reports of single patients with CBD have described difficulty on mental calculation tasks that require number knowledge (Caselli & Jack, 1992; Rebeiz, Kolodny, & Richardson, 1968). Two previous studies quantified this impairment, showing a profound deficit on simple oral calculations, dot array magnitude estimations, and magnitude comparisons of both pairs of numerals and pairs of dot arrays (Halpern et al., 2003, 2004). In the present study, we aimed to test hypotheses about the basis for this number impairment in a mildly demented cohort of CBD patients. Group findings for the dot array magnitude comparison task revealed that CBD patients are impaired only for larger-numbered arrays of dots. The great majority of CBD patients made several errors with the larger numbers, while only a small minority of these patients made a single error on this task using smaller numbers. Their performance was comparably impaired on measures of magnitude comparison involving larger numbers, despite visual–spatial manipulations. Group comparisons for the Piagetian measure of number conservation revealed a similar pattern of impairment only for sets containing more than a smaller number of poker chips. By comparison, the majority of SD patients with progressive aphasia performed all of the numerical tasks in this study at a ceiling level. Their performance for smaller and larger stimuli did not differ, and visual–spatial manipulations did not have any impact on their performance accuracy.

Language deficits have been hypothesized to explain impairment on tasks requiring knowledge of larger numbers. From this perspective, language must be used to differentiate between larger numbers (Dehaene, 1997; Dehaene & Cohen, 1997). However, we believe that the deficit seen in number knowledge in CBD, specifically for numbers larger than 3, is independent of language functioning. Naming difficulty has been reported in clinical series (Grossman et al., 2004; Kompoliti et al., 1998; Riley et al., 1990) and autopsy-proven cases (Gibb, Luthert, & Marsden, 1989; Kertesz, Hudson, Mackenzie, & Munoz, 1994; Wenning, Litvan, & Jankovic, 1998) of CBD, but comprehension of single words seems to be less severe than their naming difficulty (Ikedo, Akiyama, & Iritani, 1996; Kertesz et al., 1994; Sakurai, Hashida, & Uesugi, 1996), and aphasia is considered to be uncommon (Black, 2000; Kompoliti et al., 1998; Riley et al., 1990). In a previous study, we demonstrated that this group of CBD patients has far more difficulty with representations of numbers than with representations of object names (Halpern et al., 2004). Thus, significantly degraded number knowledge, despite little aphasia, is inconsistent with the proposal that the use and comprehension of numbers is verbally mediated.

Additional evidence against verbally mediated processing of larger numbers comes from observations of the SD patients assessed in this study. Despite language deficits as seen in their performance on an object naming task (Halpern et al., 2004), SD patients performed calculations and magnitude judgments of larger numbers at a ceiling level, and this did not differ from their highly accurate performance with smaller numbers. Single case descriptions also have shown that number meaning is preserved in SD patients (Cappelletti et al., 2001; Diesfeldt, 1993). Thus, despite the presence of an aphasia, a deficit for larger numbers has not been observed in SD. This provides further evidence against the hypotheses that larger numbers depend on verbal mediation.

Visual–perceptual manipulations of stimuli may influence the performance of aphasics with larger numbers. According to this argument, aphasics depend on the spatial extent of an array to make magnitude judgments, especially for larger numbers, since they cannot rely on intact verbal mediation. However, we found that SD patients performed all measures at a ceiling level, despite manipulating the spatial properties of stimuli. Their performance with the divergent stimuli on the dot array magnitude comparison task thus was not influenced by manipulations of dot size. SD patients also performed the number conservation task flawlessly even for larger sets of poker chips. Despite aphasia, SD patients seem to have intact knowledge of quantity regardless of the magnitude or visual–spatial manipulations of the stimuli.

Visual perceptual–spatial impairments have been documented in CBD (Frisoni et al., 1995; Gibb et al., 1989; Soliveri, Monza, & Paridi, 1999), and a study of 14 patients with autopsy-confirmed CBD reported vi-
sual or sensory neglect (Wenning et al., 1998). We, too, found that CBD patients were relatively impaired on a visual–spatial task. However, their performance did not correlate with their magnitude judgments of equally sized dots regardless of the size of the arrays. We minimized the impact of potential lateralized neglect in CBD by arraying stimulus materials vertically. A visual perceptual–spatial deficit presumably would compromise performance with both smaller and larger-numbered dot arrays. Instead, the CBD patients that we assessed were selectively impaired for larger-numbered arrays. We also found that the level of impairment for larger numbers did not worsen in CBD, despite visual–perceptual manipulations. Their relative impairment with the number conservation tasks was consistent with their impairment for other measures of number as well. Thus, visual–spatial deficits cannot fully explain the observed impairments in CBD. These observations provide further evidence that knowledge of larger numbers in CBD is degraded despite an absence of aphasia and little relationship to visual–perceptual impairments.

CBD patients were not as impaired in their performance on the volume conservation tasks as the number conservation tasks. Both kinds of tasks challenged patients to ignore superficial, misleading visual–spatial attributes of the stimuli in order to attend to the numerosity or volume of the stimuli, and we discussed above some of the reasons why a visual–spatial impairment is unlikely to explain the pattern of performance on the measures of volume conservation in CBD. Their modest difficulty may be due in part to the fact that the volume task is easier in some overall manner than the number task. Developmental work suggests that volume conservation is acquired earlier than number conservation (Piaget, 1964). An alternative possibility is that tasks specifically requiring enumeration are more difficult for CBD patients. Additional work is needed to resolve this issue.

If verbal mediation cannot explain number difficulty in CBD, and if this deficit cannot be attributed to a visual–spatial deficit, what could explain CBD patients’ number deficit? One approach proposes two distinct categories of numbers, and suggests that each category is preferentially processed by a distinct mechanism. One process, known as subitizing, involves the ability to quantify a smaller number of objects—usually no more than 3 or 4—rapidly and accurately, and is thought to support comprehension of very small numbers (Kaufman, Lord, Reese, & Volkmann, 1949). The second process, known as counting, involves the slow and error-prone process of serially incrementing a number series—typically by one unit. This process is invoked to support the enumeration of numbers larger than 3. Our findings are not inconsistent with the hypothesis that number knowledge is structured to distinguish between subitizing smaller numbers and counting larger numbers. While we have no direct evidence for this, smaller numbers seem to have a special status that is not compromised in CBD. Evidence consistent with this claim comes from animal studies (Boysen & Capaldi, 1993; Davis & Peruss, 1998; Gallistel, 1989) and infant studies (Starkey & Cooper, 1980; Wynn, 1992) showing the ability to discriminate numbers up to about 3. The distinct nature of these processes receives support from single case studies suggesting a double dissociation between subitizing and counting in brain-damaged patients. Acalculia patients suffering from brain damage have been reported to have selective preservation of elementary numerical abilities associated with subitizing (Dehaene & Cohen, 1991, 1997; Takayama, Sugishita, Akiyoshi, & Kimura, 1997; Warrington, 1982). Insult to the right parietal cortex showed selectively impaired counting but intact subitizing (Dehaene & Cohen, 1994). Conversely, two patients with developmental dyscalculia demonstrated impaired subitizing but intact counting (Butterworth, 1999; Cipolotti, Butterworth, & Denes, 1991). Our findings suggest the selective preservation of a primitive number processing system associated with subitization in CBD.

Clinical observations suggest that the cortical component of the neurodegenerative disease in CBD appears to be centered in the parietal lobe, based on clinical features such as apraxia and cortical sensory loss (Pillon et al., 1995; Riley et al., 1990). Neuroimaging studies (Blin et al., 1992; Brooks, 2000; Caselli & Jackson, 1992) and neuropathological reports that include an analysis of the neuroanatomic distribution of disease (Forman et al., 2002; Kertesz et al., 1994; Solviero et al., 1999) consistently report parietal disease in CBD. Using voxel-based morphometric analyses of structural fMRI with high spatial resolution, CBD patients appear to have cortical atrophy centered in the parietal lobe (Grossman et al., 2004; Halpern et al., 2004). These deficits for number knowledge in CBD appear to be associated with gray matter atrophy in parietal cortex (Halpern et al., 2004). Reports of parietal lobe lesions mostly due to stroke demonstrate impaired calculation and magnitude estimation (Dellatolas et al., 2001; Grafman, Passafiume, Faglioni, & Boller, 1982; Roselli & Ardila, 1989). Case studies with neuroanatomic correlation (Dehaene & Cohen, 1997; Gruber, Indefrey, Steinmetz, & Kleinschmidt, 2001) and functional neuroimaging studies of healthy adults implicate parietal cortex in tasks requiring number knowledge (Dehaene et al., 1998; Eger, Sterzer, Russ, Giraud, & Kleinschmidt, 2003; Posenti, Thioux, Sera, & de Volder, 2000; Rickard et al., 2000; Simon, Mangin, Cohen, Le Bihan, & Dehaene, 2002; Stanescu-Cosson et al., 2000). Lesion studies and fMRI data in brain-damaged patients have demonstrated that bilateral parietal lobe defects compromise number knowledge (Chochon, Cohen, van de Moortele, & Dehaene, 1999; Cohen, Dehaene, Chochon, Lehericy,
& Naccache, 2001). Further observations are needed to establish the precise role of parietal cortex in number knowledge.

References


