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Arithmetic knowledge in semantic dementia: Is it invariably preserved?

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A B S T R A C T

There is accumulating evidence of preserved arithmetic knowledge in semantic dementia (SD), contrasting with patients' striking impairment in other domains of semantic memory. This important finding exemplifies domain specificity in the breakdown of semantic memory and supports notions of the functional independence of semantic number knowledge. Nevertheless, evidence for preserved arithmetic knowledge in SD comes largely from single case studies. It is not known whether such preservation is a universal finding, or whether it persists irrespective of disease severity. The present study examined performance of 14 SD patients, varying in the severity of their semantic impairment, on tasks assessing knowledge of arithmetic signs, and on single-digit and multi-digit calculation problems, permitting evaluation of fact retrieval and use of procedures. SD patients performed generally well compared to 10 healthy controls on tests of addition and subtraction. However, abnormalities were elicited, which were not explained by education or hemispheric side of atrophy, but increased as a function of semantic severity. Patients had difficulty identifying arithmetic signs. They used increasingly basic, inflexible strategies to retrieve multiplication table 'facts', and in multi-digit calculations they made procedural errors that pointed to a failure to understand the differential weighting of left and right hand columns. The pattern of responses and error types mirrors in reverse that found in children as they acquire arithmetic competence, and suggests a progressive degradation in conceptual understanding of arithmetic. Longitudinal study of two SD patients demonstrated an association between semantic decline and impaired arithmetic performance. The findings challenge the notion of arithmetic knowledge as a totally separate semantic domain and suggest that the temporal lobes play an important role in arithmetic understanding.

1. Introduction

There is widespread consensus that calculation draws on different forms of arithmetic competence. Influential contemporary models in the adult neuropsychological literature (McCloskey, Caramazza, & Basili, 1985; Dehaene, 1992; Dehaene & Cohen, 1995, 1997) distinguish between comprehension of operation symbols (e.g. +), knowledge of arithmetic facts (e.g. that $2 \times 3 = 6$) and application of procedures (i.e. the sequence of steps necessary for performing a calculation).

An important theoretical issue is how these components of arithmetic competence relate to meaningful knowledge. An assumption of the neuropsychological models outlined above is that facts and procedures are supported by dedicated and autonomous networks in associative memory, and can be retrieved automatically without recourse to meaning (Ashcraft, 1982; Campbell & Graham, 1985; Girelli & Delazer, 1996; McCloskey et al., 1985). Meaningful understanding, often termed "conceptual knowledge of arithmetic" is thought to comprise a functionally independent component of the calculation system. It is defined as knowledge of stable, overarching principles and laws relating to arithmetic operations (Hittmair-Delazer, Semenza, & Denes, 1994; Sokol, McCloskey, & Cohen, 1989).

In the child development literature, the 'semantic' model of calculation offers a very different perspective (Baroody, Wilkins, & Tiilikainen, 2003; Baroody & Ginsburg, 1986; Brownell, 1935; Carpenter, 1986; Resnick, 1992). Arithmetic meaning is not thought to develop independently of other arithmetic processes. Rather, successful production of arithmetic facts and procedures depends on semantic understanding of arithmetic relationships. Young children's understanding is initially very limited so that it can only support very basic and inflexible counting strategies to retrieve single-digit combinations (e.g. $5 + 3 \rightarrow$ "5-6-7-**8**") (Carpenter & Moser, 1982; Siegler, 1988) and leads to systematic procedural errors when performing multi-digit calculations (e.g. $75 - 38 = 43$) (Brown & Burton, 1978; Resnick, 1982; Young & O'Shea, 1981). As understanding of arithmetic relations and operations grows, strategies become more interconnected, complex and flexible, thus increasing efficiency (Butterworth, Marchesini, & Girelli, 2003; Lemaire & Siegler,

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1995). The implication is that knowledge of arithmetic facts and procedures in adults is supported not only by associative memory but also by rich and interconnected conceptual knowledge of arithmetic relations (Baroody, 1985; Baroody & Ginsburg, 1986; Resnick, 1992; Sophian, 1996).

According to these developmental models, meaningful knowledge does not necessarily imply stable knowledge of overarching principles. Conceptual knowledge ranges from very basic understanding of numerical manipulations to a more elaborate and integrated understanding of wider principles. Crucially, there is not a fundamental distinction between low level and high level knowledge, but rather gradations in conceptual understanding (Baroody & Ginsburg, 1986; Baroody et al., 2003; Carpenter, 1986; Resnick, 1992).

Arithmetic knowledge is of immense theoretical interest. There is converging evidence that it forms a biologically determined and functionally distinct domain of semantic knowledge. Lesion studies (Cipolotti, Butterworth, & Denes, 1991; Polk, Reed, Keenan, Hogarth, & Anderson, 2001; Thioux et al., 1998) and brain-imaging studies (Pesenti, Thioux, Seron, & De Volder, 2000; Thioux, Pesenti, Costes, De Volder, & Seron, 2005) have identified dedicated neural substrates for number knowledge and calculation in the inferior parietal lobes, whereas conceptual understanding of words and objects implicate the temporal lobes (Kapur et al., 1994; Lauro-Grotto, Ciaramelli, Piccini, & Treves, 2007; Noppeney et al., 2007). Perhaps the most compelling evidence comes from neuropsychological studies on patients with semantic dementia (SD), a selective disorder of semantic memory arising from focal degeneration of the temporal lobes (Hodges, Patterson, Oxbury, & Funnell, 1992; Snowden, Goulding, & Neary, 1989). Patients show loss of word meaning, exemplified by poor comprehension, profound anomia and the presence of semantic errors (e.g. horse → "dog"). The semantic disorder is multi-modal, involving difficulty in recognising objects, faces, smells and environmental sounds (Bozeat, Lambon Ralph, Patterson, Garrard, & Hodges, 2000; Snowden, Neary, & Mann, 1996). In contrast, non-semantic cognitive skills, including spatial abilities and autobiographical memory, remain well preserved. The highly selective but profound nature of the conceptual breakdown in this disorder offers a unique opportunity to study the cerebral organisation of semantic knowledge.

Despite the pervasive semantic impairment, conceptual understanding of number and calculation in SD appears to be strikingly spared (Cappelletti, Butterworth, & Kopelman, 2001; Cappelletti, Kopelman, Morton, & Butterworth, 2005; Crutch & Warrington, 2002; Diesfeldt, 1993; Jefferies, Bateman, & Lambon Ralph, 2005; Lemer, Dehaene, Spelke, & Cohen, 2003; Zamarian, Karner, Benke, Donnemiller, & Delazer, 2006). There are anatomical reasons why this should be so. SD is associated with circumscribed atrophy of the anterior temporal lobes, particularly the inferior and middle temporal gyri (Chan et al., 2001; Mummery et al., 2000), and the focal distribution of atrophy is maintained even at end stage disease (Snowden et al., 1996). The parietal lobes, which are thought to underlie numerical concepts and calculation skills, are notably spared. The apparent dissociation between number knowledge and other domains of semantic knowledge thus provides support for the functional and anatomical independence of conceptual number knowledge.

Nevertheless, most of the evidence for preserved number knowledge in SD comes from single case studies. It is not known whether such patients are representative of the SD population. Indeed, one recently reported patient was described as highly educated, with substantial pre-morbid experience of numbers (Cappelletti et al., 2005). It would be important to establish whether preserved calculation is demonstrable across a relatively large, unselected series of pure SD patients, regardless of their educational level. Furthermore, most case studies are cross-sectional. It is therefore not clear whether preservation is maintained regardless of the severity of patients' semantic disorder. This is a pertinent question: loss of understanding in SD does not occur in an all-or-none fashion. Rather, degradation of concepts is gradual and knowledge may be partial. Is arithmetic knowledge totally immune from this gradual breakdown (as predicted by the functional independence hypothesis) or does semantic impairment impact also to some extent on calculation skills?

Case studies have, understandably, emphasised the marked disparity between SD patients' performance on number and other semantic tasks. However, there is some evidence that calculation performance is not entirely normal. Patients may have difficulty recognising arithmetic signs (Cappelletti et al., 2001; Crutch & Warrington, 2002; Diesfeldt, 1993), retrieving arithmetic facts (Cappelletti et al., 2005; Diesfeldt, 1993; Jefferies et al., 2005) and even applying arithmetic procedures (Cappelletti et al., 2005). These deficits have been attributed to the verbal demands of the task, arising secondary to patients' declining language abilities (Cappelletti et al., 2005; Jefferies et al., 2005; Lemer et al., 2003). By contrast, meaningful understanding of arithmetic operations and relationships is assumed to remain preserved (Cappelletti et al., 2005; Lemer et al., 2003).

There has, however, been little systematic exploration of qualitative performance characteristics so that the basis for failures is not always easy to determine. If breakdown is secondary to verbal deficits, then one might predict particular impairment in rapid access to verbal number 'facts' (e.g. 5×4), but preserved use of back-up strategies. Poorer performance might also be expected in patients with greater left than right temporal atrophy. Furthermore, if conceptual understanding of arithmetic is preserved then there is no reason to predict a correlation between calculation performance and severity of semantic impairment. One might expect too that errors should be plausible, reflecting an implicit understanding of numerical relationships.

By contrast, if semantic breakdown does impact on arithmetic knowledge, there should be a direct relationship between calculation performance and severity of semantic impairment. The left versus right predominance of temporal atrophy ought not to influence calculation performance. Furthermore, one might expect a reversal of the developmental processes seen in children's acquisition of arithmetic, affecting rapid, automatic retrieval of number facts *and* correct application of arithmetic procedures. With increasing semantic impairment, patients would be expected to rely increasingly on compensatory strategies, showing a gradual transition from use of flexible, principled strategies to the more basic strategies observed in young children.

We had the opportunity to investigate calculation performance in a large group of 14 SD patients, differing in level of education, left/right topography of temporal lobe atrophy and severity of semantic impairment. Analysis of quantitative and qualitative performance characteristics permitted exploration of arithmetic sign knowledge, 'fact' retrieval and procedural application as well as their relationship to formal education, distribution of temporal atrophy and semantic impairment. A longitudinal analysis of two patients allowed us to explore further the effect of semantic severity on arithmetic performance.

2. Methods

2.1. Participants

2.1.1. Semantic dementia

The SD group comprised 14 patients (9 men, 5 women), currently attending a specialist dementia clinic (Table 1). All patients had presented with problems in naming and comprehension, in the context of well preserved day-to-day mem-