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Research report

Dissociations in mathematical knowledge: Case studies in Down's syndrome and Williams syndrome

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ABSTRACT

A study is reported of mathematical vocabulary and factual mathematical knowledge in PQ, a 22 year old with Down's syndrome (DS) who has a verbal mental age (MA) of 9 years 2 months and ST, a 15 year old with Williams syndrome (WS) who has a verbal MA of 9 years 6 months, matched to typically developing controls. The number of mathematical words contained within PQ's lexical stores was significantly reduced as reflected by performance on lexical decision. PQ was also impaired at both naming from descriptions and describing mathematical words. These results contrast with normal lexical decision and item descriptions for concrete words reported recently for PQ (Robinson and Temple, 2010). PQ's recall of mathematical facts was also impaired, whilst his recall of general knowledge facts was normal. This performance in DS indicates a deficit in both lexical representation and semantic knowledge for mathematical words and mathematical facts.

In contrast, ST, the teenager with WS had good accuracy on lexical decision, naming and generating definitions for mathematical words. This contrasted with the atypical performance with concrete words recently reported for ST (Robinson and Temple, 2009). Knowledge of addition facts and general knowledge facts was also unimpaired for ST, though knowledge of multiplication facts was weak.

Together the cases form a double dissociation and provide support for the distinct representation of mathematical and concrete items within the lexical–semantic system during development. The dissociations between mathematical and general factual knowledge also indicate that different types of factual knowledge may be selectively impaired during development. There is further support for a modular structure within which mathematical vocabulary and mathematical knowledge have distinct representations. This supports the case for the independent representation of factual and language-based knowledge within the semantic system during development.

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1. Introduction

Children with learning difficulty provide models for the exploration of lexical and semantic representation of vocabulary and knowledge. This paper focuses upon mathematical vocabulary and knowledge in two contrasting genetic disorders, reporting a case study of Down's syndrome (DS) and a case study of Williams syndrome (WS).

DS is a pervasive developmental disorder most commonly caused by triplication of chromosome 21 (LeJeune et al., 1959) but also arising in 3% of cases from translocation of part or all of chromosome 21 with chromosome 14. The latter form has no relationship to maternal age and the translocation may be of maternal or paternal origin. Mental retardation is a consistent feature of DS, with IQ typically situated in the moderate to severely retarded range (Carr, 2003). Language development is reported as delayed relative to general cognitive ability, with a more pronounced deficit emerging in verbal production relative to verbal comprehension around 18 months of age (Fowler, 1990). On standard receptive vocabulary tasks, performance has been consistently documented at mental age (MA), with MA appropriate semantic verbal fluency and word frequency effects also reported (Pennington et al., 2003; Robinson and Temple, 2010; Vicari et al., 2004). In contrast, productive vocabulary has been consistently reported as below MA, with reduced mean length of utterance despite comparable lexical store size (Pennington et al., 2003; Robinson and Temple, 2010; Vicari et al., 2000; Ypsilanti et al., 2005).

WS is a rare contiguous gene disorder caused by a microdeletion of the long arm of chromosome 7, at band 7q11.23 (De Silva et al., 2002; Ewart et al., 1993). Mental retardation is also a consistent feature of WS, with IQ typically situated in the mild to moderately retarded range (Bennett et al., 1978; Morris et al., 1990; Udwin et al., 1987). Poor visuo-spatial processing skills are reported, with atypical global processing and difficulties with perceptual grouping and spatial relations (Bihrle et al., 1989; Bertrand et al., 1997; Farran and Jarrold, 2003; Farran et al., 2003). Language development is also atypical, with selective impairments and dissociations in cognitive skills typically associated with early lexical acquisition, such as, referential pointing, object categorization and the use of whole object constraints (Stevens and Karmiloff-Smith, 1997; Mervis and Bertrand, 1997; Nazzi et al., 2005). On receptive vocabulary tasks, when broad semantic knowledge is required performance has been reported as intact or elevated relative to verbal MA, however, when fine-grained semantic knowledge is required performance is reported as impaired (Clahsen et al., 2004; Bellugi et al., 1990; Temple et al., 2002; Volterra et al., 1996). Consistent with this, reduced specification of 'sensory' information within semantic representations has been documented (Robinson and Temple, 2009). Atypical word frequency effects have also been reported on tasks of semantic verbal fluency, despite an increase in the number of items generated (Bellugi et al., 1990; Temple et al., 2002; Volterra et al., 1996). Productive vocabulary has also been reported as impaired by some (Clahsen et al., 2004; Clahsen and Temple, 2003; Temple et al., 2002; Volterra et al., 1996), though not others (Bello et al., 2004; Lukacs et al., 2001), with recent evidence of category-specific deficits (Robinson and Temple, 2009). These studies suggest that relative to verbal MA, lexical stores are elevated in size but semantic knowledge representations are inadequately specified or poorly activated, which may affect certain categories of knowledge to a greater extent than others.

1.1. *Mathematical skills*

Mathematical difficulties have been consistently reported in DS. Impaired performance has been documented on the *arithmetic* subtest of the Wechsler scales (Devenny et al., 2000; Kitler et al., 2004). Children and infants with DS have also been reported to have difficulties discriminating between familiar and unfamiliar numerosities and solving novel counting problems, compared to MA matched typically developing (TD) controls (Gelman and Cohen, 1988; Patterson, 2003). In a longitudinal study of 79–102 individuals with DS aged between 8 and 21 years, Turner and Alborz (2003) reported impaired mathematical skills, with a plateau in performance around 16–18 years of age. In relation to the UK National Curriculum targets, 75% achieved Foundation level skills, 50% achieved Key Stage 1 level skills (using and applying mathematics) and 25% demonstrated Key Stage 2 level skills (number and algebra) with some Key Stage 3 level skills (shape, space and measures). Similarly, in a study of 41 adults with DS, Carr (1988) reported that around 53% could recognize numbers and count, 19% could add two figures between zero and nine and 12% could subtract figures between zero and nine. Shepperdson (1994) also found that, in a study of 49 individuals with DS, 18% had no numerical abilities as teenagers; however, by early adulthood 31% had no numerical abilities. This indicated a decline of mathematical skills in post-adolescents with DS. It is unclear whether this is simply part of the general decline of cognitive skills in adulthood in DS associated with the early onset dementia in this syndrome (Devenny et al., 2000).

Mathematical knowledge in individuals with DS has however been shown to be influenced by the school attended, with those attending mainstream schools achieving higher levels of numerical skills than those in special education (Casey et al., 1988; Sloper et al., 1990). Mathematical skills in DS have also been shown to correlate with MA, receptive vocabulary and grammatical comprehension (Caycho et al., 1991; Nye et al., 1995; Sloper et al., 1990), suggesting that underlying language skills may be related to performance. Children with DS in mainstream schools may be more likely to have higher MA and language skills to begin with, which may account for the effect of school attendance upon mathematical knowledge. Alternatively, experience in a mainstream school may expose the child with DS to more or differently constructed mathematical teaching than the child with DS in special education.

In WS, there are reports of mathematical difficulties in everyday life, such as, managing change and cooking from recipes (Bellugi et al., 2000). Poor performance is documented for the *arithmetic* subtest of the Wechsler scales (Howlin et al., 1998; Temple et al., 2002; Udwin et al., 1987). Delayed understanding of the cardinality principle (knowing that the last tag