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Mathematical skill in individuals with Williams syndrome: Evidence from a standardized mathematics battery

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Abstract

Williams syndrome (WS) is a developmental disorder associated with relatively spared verbal skills and severe visuospatial deficits. It has also been reported that individuals with WS are impaired at mathematics. We examined mathematical skills in persons with WS using the second edition of the Test of Early Mathematical Ability (TEMA-2), which measures a wide range of skills. We administered the TEMA-2 to 14 individuals with WS and 14 children matched individually for mental-age on the matrices subtest of the Kaufman Brief Intelligence Test. There were no differences between groups on the overall scores on the TEMA-2. However, an item-by-item analysis revealed group differences. Participants with WS performed more poorly than controls when reporting which of two numbers was closest to a target number, a task thought to utilize a mental number line subserved by the parietal lobe, consistent with previous evidence showing parietal abnormalities in people with WS. In contrast, people with WS performed better than the control group at reading numbers, suggesting that verbal math skills may be comparatively strong in WS. These findings add to evidence that components of mathematical knowledge may be differentially damaged in developmental disorders.

Keywords: Williams syndrome; Mathematics; Number; Magnitude representation; Spatial representation; Developmental disorder

1. Mathematical abilities in Williams syndrome

Williams syndrome (WS)[1] is a genetic disorder (1:7500) that generally causes mild to moderate retardation, distinctive facial morphology, small stature and other physical anomalies (e.g., heart defects: Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Ewart et al., 1993; Mervis, Bertrand, Morris, Klein-Tasman, & Armstrong, 2000; Meyer-Lindenberg, Mervis, & Berman, 2006). People with WS also exhibit a strikingly uneven cognitive profile which includes relatively spared language together with severely impaired visuospatial abilities (Bellugi et al., 2000; Mervis et al., 2000). Language, especially vocabulary, is a strength for these individuals, and some aspects of syntax and semantics are also quite strong (Musolino, Landau, & Chunyo, 2006; Zukowski, 2005). In contrast, visuospatial abilities such as block construction and drawing are severely impaired, with performance at the level of 3- or 4-year-old normally developing children (Bertrand, Mervis, & Eisenberg, 1997; Farran, Jarrold, & Gathercole, 2003; Georgopoulos, Georgopoulos, Kurz, & Landau, 2004; Hoffman, Landau, & Pagani, 2003).

The uneven profile in WS is evident even within the realm of visual processing. Visuoconstructive impairments have been linked to abnormalities in parietal areas of the WS brain, part of the dorsal stream of visual processing (the "where" or "how" stream; Goodale & Milner, 1992; Meyer-Lindenberg et al., 2004; see also Atkinson et al., 1997). Consistent with damage to parietal areas, people with WS are also particularly impaired in tasks such as posting a letter or visual object tracking, which engage these areas (Atkinson et al., 1997; O'Hearn, Landau, &

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[1] *Abbreviations*: WS, Williams syndrome; TEMA-2, Test of Early Mathematical Ability, version 2; MA, mental-age; KBIT-1, Kaufman Brief Intelligence Test, version 1; MLD, math learning disabled.

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Hoffman, 2005). In contrast, other visual abilities such as perception of biological motion, motion coherence, and object recognition are at or above the level expected on the basis of mental-age (Jordan, Reiss, Hoffman, & Landau, 2002; Landau, Hoffman, & Kurz, 2006; Reiss, Hoffman, & Landau, 2005). Biological motion is supported by activity in the superior temporal sulcus (Puce & Perrett, 2003) and object recognition is supported by areas of the inferotemporal lobe, part of the ventral stream. In general, functions that utilize ventral visual areas in the temporal lobe (the "what" stream) are strong (Meyer-Lindenberg et al., 2004). As one striking example, people with WS are relatively skilled with face perception and recognition (Paul, Stiles, Passarotti, Bavar, & Bellugi, 2002; Tager-Flusberg, Plesa-Skwerer, Faja, & Joseph, 2003). This strength may be linked to a preference for faces, which accords well with the outgoing and friendly personality often found in WS (Meyer-Lindenberg et al., 2006).

It has been suggested that people with WS have particular problems with mathematics in addition to, and possibly related to, their visuospatial deficits and parietal lobe abnormalities (Ansari & Karmiloff-Smith, 2002; Paterson, Girelli, Butterworth, & Karmiloff-Smith, 2006). However, only a few studies have directly assessed mathematical knowledge in WS across a range of tasks (Paterson et al., 2006; Udwin, Davies, & Howlin, 1996), and results from these studies are difficult to interpret. For example, Udwin and colleagues (1996) found that scores on standardized arithmetic tests did not improve between adolescence and adulthood, in contrast to general IQ scores, which did improve. However, Udwin and colleagues urge caution in interpreting the results, as different assessments were used at different timepoints, and many of the arithmetic problems were beyond the skill level in WS.

One possibility is that some but not all components of mathematical reasoning are impaired in individuals with WS. This possibility reflects evidence that different components of mathematics are functionally distinct in adults. Distinct psychological and neural representations appear to be engaged for verbal versus magnitude/quantity components of numerical reasoning (Cipolotti, Butterworth, & Denes, 1991; Dehaene, Spelke, Pinel, Stanescu, & Tsivkin, 1999; Dehaene, Piazza, Pinel, & Cohen, 2003; Lemer, Dehaene, Spelke, & Cohen, 2003). Linguistic or verbal knowledge uses number words as symbols to refer to exact quantities, is sensitive to the language in which it was encoded, may support species-specific reasoning about number, and may be represented near language association areas on the left side of the brain (Dehaene et al., 1999, 2003). In contrast, a non-symbolic magnitude representation that is approximate may underlie reasoning about quantity (i.e., a mental number line). This representation does not seem sensitive to the language in which it was learned, is available to infants and non-human animals, and may utilize bilateral dorsal areas in the parietal lobe (See Dehaene, 1997; Dehaene et al., 1999, 2003). Given other evidence on the parietal lobe, this type of number representation may be closely linked to spatial representations. This division of labor—between aspects of mathematical reasoning that are supported primarily by verbal knowledge and those supported by a magnitude representation—may also be evident in some developmental disorders (e.g., Turner syndrome; Bruandet, Molko, Cohen, & Dehaene, 2004; but see Murphy, Mazzocco, Gerner, & Henry, 2006).[2]

Several investigators have proposed that mathematics might be selectively damaged in WS in accord with this division of function—with weak spatial/magnitude abilities and strong verbal skills (Ansari, Donlan, & Karmiloff-Smith, in press; Paterson et al., 2006). If so, mathematical tasks that rely on the representation of numerical magnitude could be more impaired than tasks that rely on verbally encoded number. Consistent with this, recent work on estimating number (Ansari et al., in press) and the symbolic distance effect (Paterson et al., 2006)[3] suggest that representing magnitudes in mathematical tasks may be particularly impaired in WS. For instance, Ansari et al. (in press) found that the ability of children with WS (mean age 9.7 years) to estimate a small number of dots (up to 12) displayed briefly was comparable to 4-year-old typically developing children, while adults with WS performed more like 6-year-olds. Evidence is mixed on whether the verbal skill in individuals with WS leads to better performance on those mathematical tasks having a strong verbal component. Ansari and colleagues (Ansari et al., 2003) reported that an understanding of cardinality—that the last number counted equals the total number of items in a set—in a group of 6- to 11-year-olds with WS was on par with mental-age (MA) matches (mostly 3- and 4-year-olds). Moreover, they found that verbal mental-age, but not block construction scores, accounted for the variability in cardinality judgments in children with WS whereas the opposite pattern held in typically developing children. These findings are consistent with the idea that verbal strategies facilitate performance of individuals with WS on numerical tasks. In contrast, Paterson et al. (2006) found that the strong verbal skills in WS did not facilitate performance on the verbal components of a test of mathematics, compared to typically developing individuals and people with Down's syndrome matched for non-verbal ability.

To better understand whether there is particular impairment in mathematical reasoning, the present study assessed

[2] Some have suggested that impairment in a fundamental ability supporting mathematical reasoning, such as magnitude representation, might cascade over development, leading to mathematical deficits; this might occur for WS and other developmental disorders (Ansari & Karmiloff-Smith, 2002).

[3] A magnitude representation is thought to be responsible for the symbolic distance effect, in which participants are faster to discriminate numbers that are farther apart than those that are close together, presumably due to greater overlap in the representations of numbers that are close together (Moyer & Landauer, 1967; Sekuler & Mierkiewicz, 1977).