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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

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[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).

a wide range of early mathematics skills, including verbal abilities and the mental number line, in persons with WS relative to mental-age matched individuals without WS. We used the Test of Mathematical Abilities, second edition (TEMA-2; Ginsburg & Baroody, 1990). The TEMA-2 is an excellent tool for examining the level of mathematical abilities in WS because it is a standardized for use with children from 2- to 8-years of age, consistent with the mental-age of participants with WS; moreover, it tests a broad set of basic mathematical abilities. The TEMA-2 was first introduced in 1983 to identify children at risk for learning problems in mathematics, and has been used effectively to examine math deficits in people with developmental disorders and mathematics learning disability (Mazzocco, 2001; Murphy et al., 2006). Our first goal was to determine whether math abilities were particularly impaired in people with WS. If so, individuals with WS should have *lower* overall test scores on the TEMA-2 than typically developing children matched for mental-age. This level of performance would be similar to that found in WS people on visuospatial tasks such as block construction. Our second goal was to examine whether the WS pattern of performance on particular questions reflects weaker performance on tasks requiring representation of magnitude, or the mental number line, and stronger performance on tasks engaging verbal abilities, as has been suggested previously albeit with mixed results (Ansari et al., 2003; Paterson et al., 2006).

2. Methods

2.1. Participants

Fourteen people with Williams syndrome (7 males, 7 females) with a mean age of 17 years, 9 months ($SD = 7$ years, 3 months), positively diagnosed by a geneticist and the FISH test (Ewart et al., 1993), participated in the study. Typically developing children matched for mental-age (6 males, 8 females) were, on average, 6 years, 2 months old ($SD = 1$ year, 1 month). People with WS were recruited through the Williams syndrome Association, and typically developing children were recruited through local preschools or other studies. They were individually matched to the WS participants on mental-age (MA-matches), using the raw score from the Matrices subtest on the Kaufman Brief Intelligence Test (KBIT-1; Kaufman & Kaufman, 1990).[4] All but one participant was also matched on gender. The individuals with WS also did the block construction subtest from the Differential Abilities Scales (Elliott, 1990) as part of a standard battery administered to participants with WS.

The Matrices subtest on the KBIT-1 requires picture-based category matching and has relatively few spatial items; therefore it does not overly penalize WS people for their spatial deficit (*M*s: $\text{WS} = 21.1 \pm 4.7$; $\text{controls} = 20.9 \pm 3.3$; $t(26) = .09$, $p = .93$). The Verbal subtest requires picture naming, a relative strength in WS, and the WS group performed better than controls, but not significantly so (*M*s: $\text{WS} = 40.1 \pm 7.5$; $\text{controls} = 37.4 \pm 5.3$; $t(26) = 1.10$, $p = .28$). The WS profile emerging from these participants was consistent with that reported in other studies of WS, with a mean IQ of 65 ± 16 (KBIT-1), and most participants performing at or below the first percentile for age on block construction (Elliott, 1990; See Table 1). This suggests that the participants in the present study are representative of the broader population of individuals with WS. Informed consent was properly obtained from all participants or their legal guardians.

2.2. Design, stimuli, and procedure

The TEMA-2 was administered in a quiet testing room by a trained experimenter who followed the standardized instructions (see Ginsburg & Baroody, 1990). The first questions are simple and then the questions progressively become more difficult. MA-matches started at the item suggested in the standardized instructions, on the basis of their age. Most of the WS participants started at the level suggested for age 4, though a few started earlier or later depending on their ability. If participants had difficulty with the first few items, the experimenter reversed course and administered earlier items. All participants were given encouragement, but no feedback on their performance. Testing continued for all participants until five consecutive items were missed, consistent with the standardized instructions for achieving a test ceiling.

The TEMA-2 includes a wide range of items, with multiple items of the same type, progressing from easy to more difficult. These items include: counting (including backwards and skip counting); perception and conception of more; cardinality; number constancy; reading and writing numerals; mental number line; speeded subtraction, addition and multiplication; written addition and subtraction; and simple word problems. Each item is also categorized by Ginsburg and Baroody (1990) as requiring formal or informal mathematical knowledge. Informal knowledge is generally introduced before starting school, and is often related to counting routines; formal knowledge includes understanding conventions (e.g., 2 means two), number facts (e.g., memorized addition tables), calculation (e.g., column addition), and other concepts learned in school.

All items were scored as prescribed in the manual, with participants earning either a 1 (correct) or a 0 (incorrect) on each item. It was assumed that participants would have answered the questions prior to their start point correctly; therefore, these questions were coded as 1 or correct. The TEMA-2 raw score, as described in the manual, included as correct all the items prior to the basal, whether correct

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[4] In many of our studies, we match WS people to controls on both the matrices and verbal subtest raw scores. In the present case, we matched only on the matrices score, because several of the adults with WS had very high verbal scores, making it impossible to match their profile on both verbal and matrices subtest to typically developing children.

Table 1
Demographic information and KBIT-1, TEMA-2, and DAS performance

Demographic			KBIT-1			TEMA-2		DAS block
Subject #	Age	Gender	IQ	Verbal age-equivalent[a]	Matrices age-equivalent[a]	Raw score	Age-equivalent[a]	Age-equivalent
Williams syndrome								
106	10;5	M	69	5;10	6;4	30	6;1	3;7
115	10;9	F	90	10;4	8;0	42	7;9	5;10
103	12;7	F	86	9;0	9;6	50	8;6	5;10
102	13;0	M	88	9;0	13;1	59	>8;11[b]	9;3
107	13;2	F	74	8;9	7;1	31	6;2	3;1
101	14;1	F	46	6;4	6;4	24	5;6	4;4
111	15;9	M	62	10;6	7;1	39	7;5	5;1
113	17;3	F	51	9;10	6;0	34	6;10	4;7
114	18;11	F	59	10;1	7;6	39	7;5	4;7
110	19;4	M	40	7;10	6;0	35	7;0	5;10
105	20;4	M	51	7;10	8;4	35	7;0	4;7
104	21;0	M	78	12;1	11;4	43	7;10	
112	22;9	M	57	8;3	8;0	31	6;2	5;10
109	38;10	F	53	8;3	4;7	34	6;10	3;10
MA-matched controls								
249	4;7	F	128	6;10	5;7	27	5;9	[c]
214	4;9	F	134	8;1	6;4	31	6;2	
206	4;10	M	140	6;4	7;1	31	6;2	
221	5;0	F	124	7;10	5;7	20	5;0	
210	5;5	F	119	7;1	7;1	22	5;2	
208	6;4	M	122	8;1	7;4	35	7;0	
273	6;5	F	121	8;3	7;1	39	7;5	
272	6;8	F	137	9;4	10;1	41	7;8	
202	6;8	M	127	9;4	7;1	38	7;3	
216	6;8	M	131	8;3	8;0	51	8;8	
203	6;8	M	130	9;7	8;0	39	7;5	
213	6;11	M	127	8;0	7;6	50	8;6	
211	7;0	F	131	8;6	8;4	46	8;2	
204	8;5	F	131	9;7	11;4	63	>8;11[b]	

[a] These age-equivalents were estimated by finding the approximate age at which the raw score fell at the 50th percentile or at an IQ of 100.

[b] The TEMA-2 was not normed at this raw score.

[c] DAS block construction scores were not carried out on control children. However, on the basis of previous studies from our lab, we would expect them to perform at about the level expected on the basis of their chronological age (e.g., Hoffman et al., 2003).

or incorrect. Between the basal and ceiling, only correct items were scored as a 1. In addition, we computed a secondary score—TEMA-2 raw, excluding mistakes—that scored mistakes before the basal as missed items, in contrast to the official method of scoring. We did this in case participants with WS exhibited less consistent performance than MA-matches (i.e., missed more early items). We examined differences between groups in these overall scores, and in several composite scores consisting of performance on a subset of questions, using *t*-tests. We also did an analysis on individual items using chi-squares to examine the number of participants in each group getting a problem correct.

3. Results

3.1. Overall test performance

As is evident from Fig. 1, there were no differences between groups on the TEMA-2 raw score ($M_{\mathrm{WS}} = 39.3 \pm 8.7$, $M_{\mathrm{MA\text{-}matches}} = 38.9 \pm 12.3$; $t(26) = .09$, $p = .93$) or the TEMA-2 raw score, excluding mistakes (an alternate scoring that excluded mistakes prior to the basal: $M_{\mathrm{WS}} = 37.6 \pm 8.9$, $M_{\mathrm{MA\text{-}matches}} = 38.1 \pm 11.8$; $t(26) = -.13$, $p = .90$). This suggests that people with WS perform as well as children matched for MA on a developmentally appropriate standardized test of mathematical ability (See also Table 1). This is in contrast to many other tasks, in particular visuospatial tasks, in which people with WS perform more poorly than do MA-matches (e.g., Farran, 2005; Bertrand et al., 1997; Hoffman et al., 2003; O'Hearn et al., 2005). In both the WS and the comparison groups, overall performance on the TEMA-2 was significantly correlated with raw score on both the verbal (WS group: $r = .54$, $p = .05$: MA-matches $r = .71$, $p = .005$) and the matrices subtests of the KBIT-1 (WS group: $r = .77$, $p = .001$: MA-matches $r = .80$, $p = .001$).

Although the total score was similar between groups, secondary item analyses did reveal some differences between groups. Chi-squares were used to analyze the

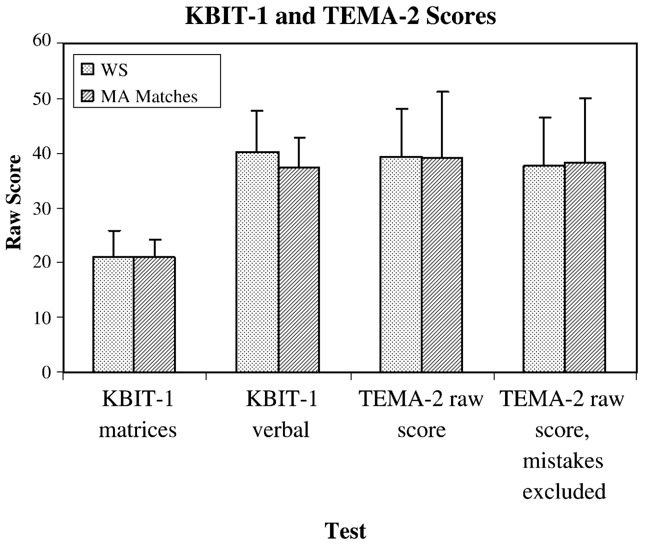

Fig. 1. Bar graphs showing raw scores on the KBIT-1 verbal and matrices subtest and the TEMA-2 scores (both the official Tema-2 score and the alternate scoring excluding mistakes) in participants with WS and MA-matched controls. Error bars are the standard error of the mean.

number of participants in each group answering each item correctly. We also created several composite scores to further examine performance differences between groups using *t*-tests.

3.2. Performance on individual items

In many ways, the performance by the two groups was similar. On the easiest questions that appeared near the beginning of the test (1–13), almost all participants from both groups performed accurately, regardless of question type. This included counting up to 10 objects (fingers, pictures, pennies), cardinality (enumerating the set correctly without counting the individuals), number constancy in spite of shape changes, reading and writing single digit numbers, and counting to 21.

Among items in the middle range, both groups began to miss some items and, in this section, there were some differences on which items they missed. There were no differences between groups ($ps > .1$) evident on questions examining counting dots (up to 16), counting out loud, counting by 10s, adding objects, conception of more, or continuing the count sequence (i.e., "34, 35, and then...). People with WS tended to be more accurate at counting out 19 manipulatives, reading 2-digit numerals in both the teens and above, and writing 2-digit numerals (all $ps = .07$). The latter two reflect 'formal' knowledge and verbal skills trained in school. In contrast, people with WS performed more poorly than controls on measures of the mental number line. On these items, 3 numbers are presented in a triangle (e.g., 6 at the top; 5 and 9 below). Participants were asked to choose the number that was quantitatively closer to the target number at the top (e.g., "is the 5 or the 9 closer to the 6": 1 digit, $p = .01$; 2 digit, $p = .058$). This type of 'informal' mathematical knowledge is thought to require the use of a spatial representation subserved by the parietal lobe (Dehaene et al., 1999).[5]

After item 33, the number of participants answering each question gradually decreased because individuals from both groups began to reach their performance ceiling. These more difficult questions often required 'formal' knowledge to which participants may not yet have been exposed. Nevertheless, there were group differences in which items were missed within this ceiling range. People with WS were more accurate than the MA comparison group at reading 3-digit numerals ($p = .007$), consistent with their strong verbal skills. In contrast, the MA-matches were better at speeded addition facts (sums of 10, $p = .06$: large doubles [e.g., $7 + 7$] $p = .07$) and speeded subtraction facts ($p = .06$). These weaknesses in WS might reflect their different mathematics training, memory impairments, or reaction time differences.

Although most participants across groups could not complete the more complex problems at the very end of the test (e.g., speeded multiplication, 2-digit subtraction, word problems), two participants from each group completed the entire battery (i.e., never reached a ceiling). The two participants with WS who had the highest TEMA-2 scores accurately completed the mental number line problems, some word problems, and skip counting (by 4s); the participant with WS scoring the highest could also do 3-digit addition and speeded multiplication facts.

3.3. Composite scores

In order to further examine differences between groups, we made several composite scores. One score was mean performance for the three items examining the mental number line (#s 23, 31, 51), and another was mean performance for the five items that tested reading numbers (#s 12, 25, 29, 38, 53). MA-matches performed better on the mental number line composite score than participants with WS [$t(26) = -2.79$, $p = .01$], while participants with WS performed better on the reading composite score than MA-matches [$t(26) = 3.41$, $p = .002$]. We also divided the items from #14 to #32 into questions testing non-verbal (or less verbal) math knowledge versus those testing more verbal math knowledge. We included only these items because, prior to item 14, almost all participants answered correctly and, after question 32, participants started to reach ceiling, so the entire sample did not complete the items. The 'non-verbal' composite score included items testing the mental number line, adding objects, and conception of more (#s 14, 16, 19, 22, 23, 31); the 'verbal' composite score included counting—objects, dots, backwards, by tens, and count after me problems—and reading and writ-

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[5] While the formal–informal distinction may also reflect the verbal–spatial components reviewed in the introduction, we feel the latter division is clearer with regards to the predictions in Williams syndrome.

ing numerals (#s 15, 17, 18, 20, 21, 24, 25, 26, 27, 28, 29, 30, 32). The MA-matched comparison children performed better than participants with WS on the non-verbal composite score [$t(26) = -3.15$, $p = .004$]; the difference between the groups on the verbal composite score did not reach significance [$t(26) = 1.37$, $p = .18$].

4. Discussion and conclusion

The present paper demonstrates that, on a standardized instrument testing a range of math skills, math ability overall in WS is at the level expected on the basis of mental-age; all our participants with WS did well at the basic mathematics questions at the beginning of the TEMA-2. This is unexpected in light of speculation that people with WS are severely impaired at mathematics (Paterson et al., 2006). When we examined items individually, however, there was evidence of peaks and valleys in mathematical abilities, with the WS group exhibiting some abilities above mental-age (reading numbers) and some below mental-age (mental number line). Such an uneven profile has been observed within the spatial domain (Landau & Hoffman, 2007), and conflicting reports of strength and weakness in language suggest that the same may be true of language (Clahsen & Almazan, 1998; Karmiloff-Smith et al., 1997; Musolino et al., 2006). While other investigators have suggested this possibility for the domain of number (Paterson et al., 2006), this is the first evidence of this profile.

While the WS group displayed a distinct pattern of performance which differed from MA-matches, we were surprised by the skill of the people with WS on the TEMA-2. While visuospatial abilities (e.g., block construction) in WS are at the level of typically developing children 4 years of age, mathematical skills are at the level of children 6–8 years of age. All individuals with WS performed at a higher age-equivalent on the TEMA-2 than on block design, the hallmark deficit in WS (See Table 1). Thus, their overall mathematical skills are better than their block construction ability (Bellugi et al., 2000; Hoffman et al., 2003), as well as other visuospatial tasks impaired to a similar degree (e.g., drawing, Bertrand et al., 1997; multiple object tracking, O'Hearn et al., 2005; and rapid enumeration or subitizing, O'Hearn, Landau, &Hoffman[6]). Several of our participants reached a high level of math ability, at or better than the level of a typical 8-year-old, including accurate performance on the mental number line questions.

This similarity in overall score between groups occurred in spite of potential differences in mathematics experience between groups.[7] For instance, the WS group may have had much more practice on rote counting, as they are older than MA-matches, and this might account for the greater skill in WS on some items (though additional training did not help them on other basic items). On the other hand, some more advanced concepts, such as multiplication, may not have been introduced to them because of their level of intellectual functioning. In spite of these issues, participants with WS performed at the level expected by mental-age, suggesting that their mathematical instruction was most likely comparable to children at similar levels of competence. Perhaps more importantly, it suggests that mathematical understanding proceeds relatively normally given a reasonable introduction to number orthography and counting routines.[8]

Our data do suggest that people with WS have particular difficulty on tasks requiring a well-articulated number line, a weakness that is likely to reflect the abnormalities in parietal areas found in WS. The mental number line has been linked to activity in parietal areas thought to support spatial representation, most explicitly areas in the intraparietal sulcus (IPS; Dehaene et al., 2003). Thus, poor performance on the mental number line question in the present study is consistent with previous reports of atypical parietal lobe structure and function in WS (Meyer-Lindenberg et al., 2004; Reiss et al., 2004). The structural abnormalities in the dorsal stream of people with WS include decreased volume in parietal areas (IPS; Meyer-Lindenberg et al., 2004; Reiss et al., 2004; also superior parietal lobe, Eckert et al., 2005) and, in some reports, the occipital lobe (Reiss et al., 2000). In addition to structural differences, functional activation in these dorsal regions is atypical. Meyer-Lindenberg and colleagues (2004) report decreased activation in parietal regions (closely located to the structural differences in IPS) during both a location memory task and a visuospatial construction task, but no differences in activation level in temporal areas for an object memory task.

Many of the abilities that are particularly impaired in WS may be related to parietal abnormalities. Studies of typical adults indicate that IPS and surrounding regions are utilized in both number and spatial tasks (e.g., Culham, Cavanagh, & Kanwisher, 2001; Dehaene et al., 2003). Therefore, deficits on the mental number line question may be related to not only impairments in estimating number (Ansari et al., in press; O'Hearn et al., under revision) and judging the distance between numbers (Paterson et al.,

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[6] This last task is linked to mathematical abilities, both because the task requires accurate counting and because skill at subitizing may underlie counting early in development (Benoit, Lehalle, & Jouen, 2004). Our evidence suggests that counting itself is not impaired in WS, relative to mental-age matches. This in turn suggests that subitizing does not play a strong causal role in WS counting, at least at the time of our test; any early delay in counting that is caused by impaired subitizing may be later overcome or compensated for in WS.

[7] Both our WS group and our MA-matches had a diverse set of experiences, including home schooling.

[8] An anonymous reviewer asked whether the two WS participants who completed the TEMA had unique training, which might suggest particular kinds of instruction that could be particularly helpful to people with WS. These two participants had been mainstreamed in public schools. One participant had not had additional training in mathematics, while the other participant had had tutoring in reading, writing, and mathematics. Both sets of parents reported that mathematics was not particularly difficult for their children.

2006; the present evidence), but also other spatial deficits (see Landau & Hoffman, 2007 for review on spatial impairment). Other areas of particular weakness in WS that may be related to parietal activation include such diverse tasks as mental rotation (Landau, personal communication), multiple object tracking (O'Hearn et al., 2005), and accurately posting a letter (Atkinson et al., 1997; Dilks, Landau, Hoffman, & Siegfried, 2001; Dilks, Landau, & Hoffman, 2007).

Our study also suggests that verbal aspects of mathematical skills, particularly reading numbers, are relative strengths for people with WS. Despite relatively similar scores on the verbal subtest of the KBIT, the WS group performed better than controls at reading numbers. This suggests that parietal involvement may not be needed for reading numbers; if it was, one might expect the WS group to perform more poorly than controls, which is clearly not the case.[9] One caveat is that the WS group did perform slightly better (non-significantly) than the MA-matches on the verbal subtest. To make the verbal scores even more similar between groups, we removed the WS participant-MA-match pair with the greatest discrepancy in scores on the verbal subtest and then re-examined our results. This change did not affect the pattern of results, suggesting that the significant difference between groups at reading numbers did not simply reflect the slightly better score on the verbal subtest of the KBIT.

The WS profile for number is consistent with the overall pattern of the strengths and weaknesses found in this disorder, with relatively spared verbal abilities and severe impairment on spatial tasks (Bellugi et al., 2000; Mervis et al., 2000). Is the pattern unique to WS? Although we do not know, several facts suggest that the profile is not observed in other math-impaired populations. First, the pattern of performance in WS does not seem to be simply a reflection of low math abilities. Mazzocco and Thompson (2005) found that 5-year-olds later identified as math learning disabled (MLD), while impaired on the mental number line question on TEMA-2, were also impaired on a host of other questions, including reading numerals, items on which participants with WS did particularly well. Thus, the performance profile on the TEMA-2 in WS differs from that found in children with poor math skills (i.e., MLD). Second, the profile of individuals with WS also differs from profiles of TEMA-2 scores reported for individuals with fragile X or Turner syndrome (Murphy et al., 2006). Whereas 6-year-old girls with fragile X syndrome showed the same difficulty on mental number line problems as observed in persons with WS, these girls also had difficulty with the counting principles, including cardinality—difficulties not evident in the WS group we studied. In this study, girls with Turner syndrome showed no item-specific performance profile on the TEMA-2. Thus, it is not the case that children with developmental disability, in general, show a particular profile on the TEMA-2.

It is possible that other developmental disorders that affect parietal lobe, for example Turner syndrome (Brown et al., 2004), may lead to distinct mathematical impairments that nonetheless share some elements with the deficit found in WS. Although the study by Murphy et al. (2006) found that girls with Turner syndrome had no item-specific performance profile on the TEMA-2, a study by Bruandet and colleagues (2004) suggests that women with Turner syndrome have a relatively specific impairment in magnitude representation that is linked to parietal lobe dysfunction. This pattern would be similar to that found in WS. It is unclear what caused the difference in the pattern of results in the two studies of Turner's syndrome, but one possibility is this deficit has several subclasses, one of which is more similar to WS. Another possibility is that the deficit in Turner syndrome is more subtle than that found in WS, thus not identifiable on the TEMA-2 but evident on other tests (relatedly, the deficit may emerge later in development than the ages studied in Murphy et al., 2006). In any case, systematic comparisons of the patterns of mathematical abilities across different developmental disorders should ultimately lead to models of distinct neurodevelopmental pathways to difficulties with mathematics (Murphy et al., 2006).

A limitation of the present study is that our sample of MA-matches have high IQs on the KBIT-1; their verbal scores (i.e., from object naming) were particularly strong, but their matrices scores were also higher than average. This problem, common with control groups, brings up two possibilities which we cannot distinguish: (1) control children with average IQs would have been slightly older than our present controls but would have performed similarly, and (2) older children with average IQs would actually show a different pattern of performance than our present controls on the TEMA-2, and thus some of our differences between groups might reflect characteristics of our control group (See Mervis & Klein-Tasman, 2004). In spite of this limitation, our evidence is informative, as our two groups were matched on the performance on the matrices subtest, and distinct patterns of performance emerged in the two groups. In addition, use of a standardized measure such as the TEMA-2 is helpful in interpreting the data—overall, the control group's performance was advanced but appeared relatively normal. Both control and WS group performance progressed as expected through the TEMA-2 (i.e., all participants got the first problems correct, some of the middle section, and missed many questions at the end) and, overall, age-equivalents on the TEMA-2 were closely linked to age-equivalents on the KBIT-1 matrices. Regardless of how to interpret the data from the control group, the relatively skilled performance of the WS group is evident on the basis of age-equivalents on the TEMA-2.

Our results on the number line questions are consistent with reported deficits in the symbolic distance effect

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[9] Thanks to an anonymous reviewer for suggesting this possibility.

(Paterson et al., 2006). However, our results appear to differ from Paterson and colleagues on their mathematical battery. They found that, in spite of an early intact ability to discriminate small numbers, people with WS appeared more impaired on a mathematical battery than were people with Down's syndrome or MA-matches, and did not show the expected strengths at reading numbers. Significant differences between the WS group and the Down's syndrome group were evident on two tasks: seriation tasks (arranging numbers and dots in sequential order; MA-matches also did poorly on the numbers) and the 'what comes before/after' question.[10] While it provides an interesting comparison, differences between WS and another special population such as Down's syndrome are difficult to interpret because—like other developmental disorders (Murphy et al., 2006)—their abilities may not be evenly impaired and their pattern of impairment is likely to differ from that of people with WS. Other differences between Paterson's and our studies include a different battery (theirs was not standardized), differences in numbers of subjects tested[11] and their use of slightly older MA-matches.

Many of our participants with WS reported that they "weren't good with numbers". They were pleased when it became clear that they could do many of the problems on the TEMA-2. Our evidence indicates that, at least on a standardized mathematics battery, people with WS can learn mathematics appropriate to their overall level of mental functioning. However, our evidence is also consistent with prior research indicating they may have difficulty with the spatial aspects of some mathematical problems, in this case questions involving a mental number line. This deficit is supported by previous work on WS in number estimation tasks (Ansari et al., in press) and tests of the symbolic distance effect (Paterson et al., 2006), supporting the possibility that this TEMA-2 question does indeed require a magnitude representation. Since magnitude representation has been linked to the parietal lobe (Dehaene, 1997; Dehaene et al., 2003), the behavioral deficit in WS is also consistent with neurological data showing that individuals with WS have decreased gray matter and hypoactivation in posterior parietal areas (Meyer-Lindenberg et al., 2004). Mathematical interventions for people with WS may be more effective if they provide different strategies, including verbal ones, which help support understanding of the number line.

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[10] The 'what comes before/after' question would appear to require a strong verbal component; it is unclear what type of knowledge the seriation task might use (with numbers, it might reflect the verbal component; with dots, possibly use of the mental number line).

[11] Paterson et al. (2006) included eight individuals with WS and nine with Down's syndrome but two with Down's syndrome did not do the math battery and some others could not complete all items on the battery.

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