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What makes counting count? Verbal and visuo-spatial contributions to typical and atypical number development

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Abstract

Williams Syndrome (WS) is marked by a relative strength in verbal cognition coupled with a serious impairment in non-verbal cognition. A strong deficit in numerical cognition has been anecdotally reported in this disorder; however, its nature has not been systematically investigated. Here, we tested 14 children with WS (mean age = 7 years 2 months), 14 typically developing controls individually matched on visuo-spatial ability (mean age = 3 years 5 months) as well as a larger group of typically developing controls (mean age = 3 years 4 months) on two tasks to assess their understanding that counting determines the exact quantity of sets (cardinality principle). The understanding of the cardinality principle in children with WS is extremely delayed and only at the level predicted by their visuo-spatial MA. In this clinical group, only language accounted for a significant amount of the variance in cardinality understanding, whereas in the normal comparison group only visuo-spatial competence predicted the variance. The present findings suggest that visuo-spatial ability plays a greater role than language ability in the actual development of cardinality understanding in typically developing children, whereas the opposite obtains for the clinical group.

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Williams syndrome (WS) is a rare genetic disorder occurring in 1 in 20,000 live births (Beuren, 1972; Greenberg, 1990). WS is caused by a hemizygous sub-microscopic deletion of some 20 genes on chromosome 7q11.23 (Ewart et al., 1993; Tassabehji et al., 1996). In recent years WS has attracted a great deal of attention from cognitive neuroscientists due to the cognitive profile that it presents: whereas aspects of language, social cognition and face processing show relative behavioural proficiency in WS, visuo-spatial cognition, planning and problem solving are seriously impaired (Arnold, Yule, & Martin, 1985; Donnai & Karmiloff-Smith, 2000; Mervis et al., 2000).

There is already some indication in the literature that people with WS also have problems with numerical tasks (Bellugi, Marks, Bihrle, & Sabo, 1988; Paterson, Girelli, Butterworth, & Karmiloff-Smith, 2002; Udwin, Davies, & Howlin, 1996). However, there exists no systematic investigation of the development of basic number skills in WS and whether these follow a typical or atypical developmental trajectory. Added motivation for the investigation of number abilities in WS derives from the current general interest in verbal and non-verbal aspects of numerical cognition (Dehaene, 2001; Dehaene, Spelke, Pinel, Stanescu, & Tsivkin, 1999) and their potential contribution to impairments of numerical cognition (Ansari & Karmiloff-Smith, 2002). Given that individuals with WS present with a relative strength in language coupled with a strong weakness in non-verbal cognitive processes, differential degrees of impact from verbal and non-verbal processes on number may exist in this developmental disorder.

Children and adults with learning disabilities cannot be tested on tasks employed with normal adults. Simpler techniques must be selected to trace cognitive impairments back to their developmental origins. One such task is counting and the understanding of cardinality. The ability to count and to use counting to determine exact quantities is considered to be a fundamental numerical ability (Briars & Siegler, 1984; Fuson, 1988; Gelman & Gallistel, 1978; Wynn, 1992). Only when children abide by the cardinality principle (i.e., the last tag in a count sequence represents the total number of items in a counted set) can they be said to have a concept of the meaning of counting, i.e., to determine the total numerosity of a set. It is one thing to know *how* to count and quite another to know *why* one counts. Studying how children with Williams syndrome develop an understanding of the cardinality principle is a good starting point to explore whether their relative strength in language affords them a proficient development of exact number representation and whether this follows a normal developmental trajectory.

How is the understanding of the cardinality principle best assessed? The most common way has been to have children count a number of objects and subsequently ask them “how many” objects they counted (Gelman & Gallistel, 1978; Wynn, 1990). This task has been criticised for being potentially misleading, encouraging the child to infer that the experimenter is asking him to recount or that his first count was erroneous. The "give-a-number" task designed by Wynn (1990, 1992) represents an alternative to the "how many" task. Here, the child is asked to give a puppet different numbers of objects. To be successful, the child needs to understand the cardinal value of the number of objects he is being asked to give to the puppet. Wynn (1990) found that very young children generally just grabbed a large number of objects; they were labelled "grabbers." Those children who consistently stopped at the correct number were called "counters." Children younger than $3\frac{1}{2}$ years were able to give correctly 1, 2, and 3 objects, but they consistently grabbed when required to give a number of objects greater than 3. By contrast, children older than $3\frac{1}{2}$ years succeeded on numbers greater than 3. Wynn argues that at around 3 years of age a conceptual shift occurs, in which children connect the concept that counting determines numerosity, on the one hand, to all numbers within their counting range, on the other.

The aim of the present work is to explore the nature and extent of cardinality understanding in children with WS and to ascertain whether they develop an understanding of the cardinality principle via a similar conceptual shift to that postulated by Wynn (1990). It is hypothesised that children with WS will be proficient at reciting the count sequence. Furthermore, it is predicted that children with WS may rely on their relative strength in language to scaffold their understanding of the meaning of counting, the mechanisms of which remain to be elucidated.

Two tasks were used to assess the cardinality principle. First, children counted 2–6 objects and were then asked how many objects they had counted ("how many" task). Subsequently, they were requested to give 1–6 marbles to a puppet ("give-a-number" task). In both tasks, two comparison groups were used. Children with WS were compared to an equal number of typically developing children, individually matched on visuo-spatial ability. This very much younger control group served to explore the level of cardinality understanding in children with WS relative to their visuo-spatial mental age matched controls. A second, larger comparison group was used to assess the extent to which visuo-spatial or language scores account for the variance in cardinality understanding in the control and clinical populations.

Method

Participants

A sample of 15 children with Williams syndrome was recruited through the Williams Syndrome Foundation, a UK-based charity that supports parents and affected individuals. All of these children had been diagnosed clinically as well as by means of the fluorescence in situ hybridisation (FISH) genetic test for deletion of the elastin gene. The mean chronological age (CA) of the children with WS was 7 years, 2 months (range: 6;0 years to 11;5 years). This age range was chosen because we assumed, on the basis of previous work, that children with WS could be expected to be functioning at a level roughly half their chronological age. It is at this level of

MA where, on the basis of previous research (Gelman & Gallistel, 1978; Wynn, 1990) one would expect the understanding of counting to be in the process of developing in healthy controls.

One child with WS failed to score above zero on the test of visuo-spatial cognition, and the data from this child were therefore excluded from further analysis. Fourteen control children were also tested. These children were individually matched to the children with WS on both gender and visuo-spatial mental age, using age equivalent scores of the Pattern Construction Subscale of the British Abilities Scales II (Elliot, Smith, & McCulloch, 1996). Our decision to match on visuo-spatial mental age, rather than language, was due to the fact that the young controls could be expected to be at an age where they would still show a range of abilities on the counting task. If the children with WS had been individually matched on verbal mental age to a group of typically developing children, the typically developing children would have been at ceiling on the counting task and would therefore not have represented a suitable comparison group. Both groups comprised 5 girls and 9 boys. The mean CA of the individually matched control children was 3 years, 5 months (range: 3;0 years to 4;7 years). Thus the individually matched control children were much younger than the children with WS.

One of the aims of this paper was to establish the extent to which language ability and visuo-spatial competence account for the variability in cardinality understanding among children with WS and how this may differ from typically developing children. Because the individually matched control children were much younger than the children with WS, they obviously did not exhibit the same range and variability in verbal mental age as the children with Williams syndrome. A comparable variance and range is, however, necessary when evaluating differences in the predictors of performance between groups. Therefore, to ensure that both groups had similar variability in verbal mental ages and visuo-spatial scores, an additional 14 typically developing children were tested. This group of children had a mean CA of 3 years, 4 months (range: 2;6 years to 5;3 years). Together with the children in the matched control group ($N = 14$), the new comparison group consisted of a total of 28 children, with a mean CA of 3 years, 4 months (range: 2;5 years to 5;3 years). All but 2 of the children in this large comparison group were also tested for both visuo-spatial ability and verbal mental age.

Background measures

All participants with WS were tested on the Early Years Version of the British Abilities Scales, BAS (Elliot et al., 1996) as well as on the British Picture Vocabulary Scales, BPVS (Dunn et al., 1992). The mean General Cognitive Ability (GCA) of the children with WS on the BAS was 57 (range: 42–81). The GCA is roughly an IQ equivalent with a mean of 100 and a standard deviation of 15. The mean verbal mental age (VMA) of the WS group on the BPVS was 4 months, 8 years (range: 2;11 years to 7;4 years). The means and standard deviations of the chronological ages, verbal and spatial mental ages as well as the raw scores on the pattern construction test for all groups tested can be found in Table 1.

Table 1
Participant background data

	Group							
	WS		Individually matched controls (a)		Additional 14 children (b)		Larger comparison group (a + b)	
	Mean	*SD*	Mean	*SD*	Mean	*SD*	Mean	*SD*
Chronological age	7.2	1.5	3.5	.51	3.4	.92	3.4	.73
BPVS age equivalent (VMA)	4.8	1.6	3.0	.73	3.8	1.6	3.5	1.3
Pattern construction age equivalent	3.0	1.0	3.0	1.0	3.9	1.1	3.4	1.2
Pattern construction raw score	4.1	3.2	3.5	2.5	7.0	4.6	5.2	4.0

Note. Two children in the individually matched control group were only tested on the pattern construction subscale of the BAS and not the BPVS. Therefore the mean Verbal mental age (VMA) for the individually matched controls in Table 1 is representative of 12/14 participants. The larger comparison group consists of 28 children, consisting of the 14 individually matched controls and 14 additional children.

Because 7 children with WS were found to have raw scores within the range for the floor age equivalent score on the pattern construction task, we ensured that children with WS did not differ significantly in their raw scores from the individually matched controls. No significant difference in raw scores was found between WS and their controls, $t(26) = 0.5, p = .60$. Also, due to the fact that 7 children had floor age equivalent scores on the pattern construction tasks, raw scores were used in all the statistical analyses, since these scores better discriminate between individuals' performance.

Procedure

All the children with WS were tested at home on two separate occasions that were no more than 1 month apart. On the first occasion they were tested on the background measures. During a second session they were tested on the two counting tasks. Children in both control groups were also tested on the background measures and the counting tasks on two separate occasions, no more than 1 month apart, in their nursery and primary schools.

"How many task"

Children were introduced to a hand-held puppet called "Marvin the Mole." They were told that the puppet had forgotten how to count, and that they were to teach him. The children were then shown different numbers of plastic animals glued to cardboard, with a 7 cm space between each animal. Participants were presented with displays of 2, 3, 4, 5, and 6 animals. Within these sets, only one type of animal was used per set (homogeneous sets). In addition, children were tested with two other sets, consisting of 3 and 4 animals, where the type of animal was varied randomly within each set (heterogeneous sets). Participants were randomly presented with these 7 sets of animals and asked to count them for the puppet, for a total of 7 trials. When they had finished counting each set, they were asked, “How many animals were there?” The experimenter recorded both whether or not the child had counted the sets of animals correctly, without skipping or double counting, as well as the answer given to the “how many” question, and whether they recounted the set. Throughout the 7 trials the experimenter repeatedly emphasised that the puppet had forgotten how to count and needed the help of the child.

“Give a number” task

The same children were then tested on the “give a number” task, which was always presented after the “how many” task, to ensure first that children were able to recite the count sequence correctly. Children were presented with two ceramic bowls. Into one bowl the experimenter emptied 93 marbles, all of which had the same colour. The bowl full of marbles was placed in front of the child and an empty bowl was put in front of the experimenter holding the puppet. Children were again told that the puppet had forgotten how to count and were asked, in random order, to put a given number of marbles into the bowl (1, 2, 3, 4, 5, or 6 marbles). Each number was repeated three times, leading to a total of 18 responses.

Results

“How many” task

Children with WS were proficient at reciting the count sequence when counting 2–6 objects. Only 14% of children with WS (2/14) did not count all 7 objects correctly. Of the 2 children who failed to obtain a ceiling score on this task, one made a single mistake and the other failed to count 3 of the 7 sets correctly. In the group of individually matched controls, 28% of the children (4/14) did not count all 7 objects correctly, but only 1 of these children made more than 3 mistakes. There was no significant difference between the young controls and the group of children with WS in their ability to attach the correct number words onto the objects they counted: $t(26) = 1.10$, $p = .27$.

On average, individuals with WS gave the correct cardinality response (stating the correct total number of items counted rather than recounting the set) only 43% of the time. Individually matched controls gave the correct cardinality response 52% of the time. There was no significant difference in the number of correct cardinal responses between children with WS and their much younger, individually MA-matched controls: $t(26) = .56$, $p = .57$.

“Give a number” task

In order to compare the performance on how many times individuals gave the correct number of small (1–3) and large numbers (4–6) within and between groups,

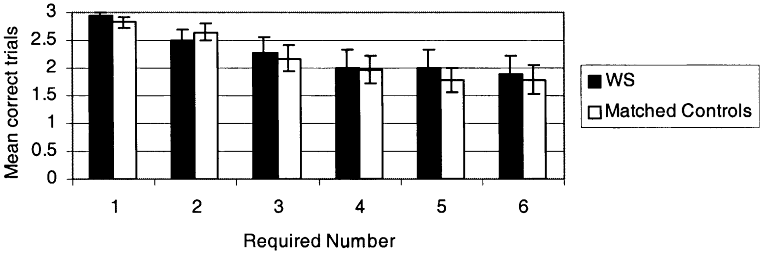

Fig. 1. Comparison of mean number of times correct number of marbles given (out of 3 trials) for numbers 1–6 for Williams syndrome and individually spatially matched control group in "give a number" task. Error bars represent the standard error of the mean.

a 2 (large vs. small) × 2 (WS vs. individually matched control group) mixed ANOVA was computed. As can be seen from Fig. 1, both WS and individually matched controls gave the wrong number of marbles significantly more often for large numbers in comparison with small numbers, $F(1,26) = 20.4$, $p < .0001$. There was no effect of group, $F(1,26) = .53$, $p = .47$, and the interaction between group and size was also found to be non-significant, $F(1,26) = .56$, $p = .48$.

Relation between language, visuo-spatial cognition and understanding of cardinality

A high degree of variability existed in the Verbal Mental Ages (Mean = 4 years, 8 months; $SD = 1$ year, 6 months) of the participants with WS. To explore whether this variability was related to children's performance on the "give a number" task, the group of children with WS was split according to VMA. We took the mean VMA in the WS group (4 years, 8 months) as a basis for our subgroup split. Using this split we compared a group of children ($N = 5$) with a VMA equal to or below 4 years and 8 months (low language ability group) with a group of children ($N = 9$) (high language ability group) who obtained a VMA higher than 4 years and 8 months.

An independent t test revealed a significant difference in VMA between the subgroups, $t(12) = 5.6$, $p < .0001$. A further independent t test revealed that the two subgroups did not differ significantly in terms of visuo-spatial ability, $t(12) = 1.3$, $p < .199$. To establish whether these groups differed in their understanding of cardinality, performance on the "give a number" task was compared between groups. This was achieved by computing a 2 × 2 mixed ANOVA with number of marbles (small vs. large) as the within-subjects variable and group (high vs. low language WS) as the between-subjects variable.

There was a significant main effect of size, $F(1,12) = 45.8$, $p < .0001$. Furthermore a significant interaction between size (large vs small) and group was found, $F(1,12) = 22.5$, $p < .0001$. As can be seen in Fig. 2, for large (4–6) numbers, children with WS in the high language group were at or near ceiling, whereas children with WS in the low language group were at or near chance. Visual inspection of Fig. 2 suggests that the interaction between size and group is not only due to the difference in the amount of large numbers correctly given between high and low language WS

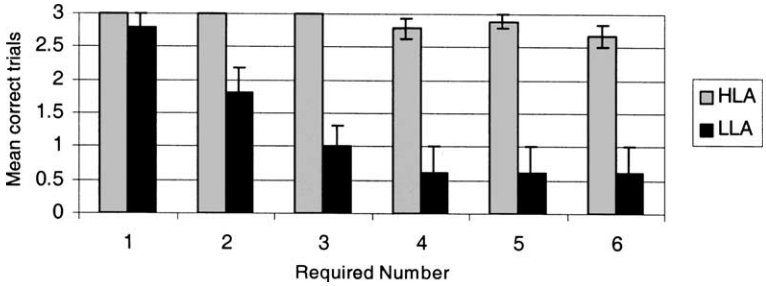

Fig. 2. Comparison of mean correct number of marbles given (out of 3 trials) for numbers 1–6 for individuals with WS in the high language ability group (HLA) and individuals with WS in the low language ability (LLA). Error bars represent the standard error of the mean.

groups. This can be explained by the finding that children in the low language WS group were also poorer than the high language group at giving small (1–3) numbers. Post-hoc *t* test confirmed that the two WS groups differed both in terms of small number of marbles (1–3), $t(12) = 7.8$, $p < .0001$, as well as large number of marbles (4–6), $t(12) = 7.3$, $p < .0001$.

These findings raise the possibility that language ability rather than visuo-spatial ability is correlated with and accounts for a greater amount of the variance in cardinality understanding in WS. To explore whether there is a difference between the extent to which language accounts for this variance and whether this differs from typically developing controls, a set of bivariate and partial correlations in addition to hierarchical regressions were run. Given the small number of trials (7) in the "how many" task and therefore the relatively low statistical power, the performance on the "give a number" task was used as dependent variable.

Due to the small variance in VMA in the individually matched control group ($SD = .73$), the correlational and regression analyses with VMA and pattern construction raw score were run with the larger comparison group ($N = 28$). This group consisted of the individually matched controls ($N = 14$) as well as the additional 14 unmatched typically developing children. There was a larger amount of variance in the VMA's of this larger control group ($SD = 1.3$), making them more similar to the variability in VMA's observed in the group of children with WS ($SD = 1.6$).

Simple and partial correlations (controlling for chronological age) between verbal mental age, pattern construction raw score and the total number of correctly given marbles were run. As can be seen from the correlation coefficients presented above the diagonal in Table 2, VMA correlated significantly with the total number correctly given in both the group of children with WS as well as in the larger comparison group. However, visuo-spatial ability was correlated significantly in the comparison group but not in WS group. Results from the partial correlations controlling for the effects of chronological age (CA), presented below the diagonal in Table 2, show that VMA was still significantly correlated with performance in the group of children with WS after controlling for the effects of CA. However, this was not true of the comparison group. By contrast, visuo-spatial competence still correlated significantly in the comparison group after controlling for CA.

Table 2
Simple and partial (controlling for chronological age) Pearsons correlations between chronological age (CA), verbal mental age (VMA), pattern construction raw score and total number of marbles correctly given in the "give a number" task

	Total number correctly given		CA		VMA		Pattern construction raw score	
	WS	Controls	WS	Controls	WS	Controls	WS	Controls
Total number correctly given	—	—	.202	.739**	.766**	.504*	.440	.761**
CA	—	—	—	—	−.077	.659**	.139	.542**
VMA	.800**	.049	—	—	—	—	.455	.530**
Pattern construction raw score	.424	.635**	—	—	.471	.265	—	—

Note. Simple correlations appear above the diagonal; partial correlations, below the diagonal. $*p < .01$, $**p < .001$.

From these correlational analyses it cannot be established whether VMA accounts for a significant amount of variance in performance on the "give a number" task over and above pattern construction raw score and CA. Therefore, hierarchical regression models were run to identify the best predictor of understanding of the cardinality principle.

Two hierarchical regression models were run in which CA was always entered in the first block. In Model 1 VMA was entered in step 2 and visuo-spatial raw score was entered in step 3. In Model 2 the order of steps 2 and 3 was reversed. As can be seen from the results presented in Table 3, in the WS group, CA did not account for a significant amount of variance in the dependent variable (total number cor-

Table 3
Hierarchical regression models with total number of marbles correctly given as the dependent variable

Step	Predictor	WS			Controls		
		β	R^2	ΔR^2	β	R^2	ΔR^2
Model 1							
1	CA	.202	.041	.041	.728**	.530	.530**
2	VMA	.758**	.656	.615**	.047	.531	.001
3	Pattern construction	.060	.658	.003	.542**	.727	.196**
Model 2							
1	CA	.202	.041	.041	.728**	.530	.530**
2	Pattern construction	.420	.214	.173	.515**	.719	.190**
3	VMA	.758*	.658	.455*	−.121	.727	.007

Note. Step denotes the order of entry of variables in the model. Chronological age (CA) is always entered in Block 1. In Model 1 Verbal Mental Age (VMA) is entered in Block 2 and Pattern construction raw score in Block 3. The order of entry for VMA and Pattern construction raw score is reversed for Model 2. β is the standardised regression coefficient, R^2 is the value of R Square, and ΔR^2 refers to the value for R Square Change. Significance level is denoted by $* = p < .005$ and $** = p < .001$.

rectly given in the “give a number” task). VMA accounted for a significant amount of the variance in the dependent variable while pattern construction score did not. This was true for the clinical group, regardless of the order in which these variables were entered. The findings from the larger comparison group demonstrate that CA accounts for a significant amount of the variance in task performance in typically developing children. Moreover, pattern construction raw score, but not VMA, significantly accounts for variance in cardinality understanding after allowing for CA, regardless of the order of the variables entered. This once again replicates the results of previous analyses.

Discussion

The results reported in this paper represent a first step towards a systematic investigation of numerical cognition in young children with Williams syndrome. Our findings show that children with WS can recite the count sequence for small numbers and make almost no errors. This is in line with results from a study by Thomas et al. (2002) using speeded naming, in which individuals with WS were accurate and fast at naming numerals. However, the present findings highlight the fact that counting of the number sequence does not predict the actual understanding of counting in this clinical group. Children with WS responded correctly to the “how many” question less than 50% of the time and, despite a mean CA above 7, they only reached the level of 3 year olds, i.e., their much younger individually matched MA controls, indicating that their understanding is merely at a level expected for their visuo-spatial mental age.

The data from the “give a number” task paint a similar picture. Overall children with Williams syndrome (WS) are significantly better at giving small (1–3) compared to large (4–6) numbers and do not differ significantly from the much younger controls. As pointed out by Wynn (1990, 1992), the ability to give numbers 1–3 correctly does not necessarily reflect an understanding of the cardinality principle, but rather a reliance on perceptual, ‘subitizing’ mechanisms, allowing for fast enumeration without counting.

Notwithstanding the results from the overall group analysis, the analysis of subgroups divided on the basis of their Verbal MA provided a more in-depth view of the way in which the development of cardinality understanding is driven in WS. Results showed that WS children with relatively high VMAs were also those who performed at or near ceiling on the give-a-number task, whereas those with VMAs below 4;8 years were very poor at numbers greater than three. These findings show that children with WS make the conceptual shift from “grabbers” to “counters” considerably later than typically developing children. Our correlational and regression analyses also suggest that individuals with WS may rely on their relative strength in language to bootstrap or scaffold their understanding of the cardinality principle. By contrast, visuo-spatial ability rather than language predicted success in the control group, even after controlling for CA. These findings thus suggest that in typically developing children visuo-spatial cognition drives the understanding of the cardinality principle to a greater extent than verbal competence. By contrast, in children with WS the opposite obtains, with verbal ability being significantly related to understanding of cardinality principle, while the visuo-spatial cognition is not. This points to an atypical trajectory in Williams syndrome.

The results from the hierarchical regression analyses strengthen the implications derived from the correlational analyses. In the group of children with WS, regardless of the order of entry of the variables, VMA significantly accounted for the variance in performance on the "give a number" task, whereas visuo-spatial ability did not. In the control group, VMA did account for a significant amount of variance in task performance when it was entered as the first variable. However, when visuo-spatial ability was entered first, VMA did not significantly account for any additional variance. These findings suggest that in the control group the variance in the "give a number" task accounted for by VMA is shared by the variance predicted by the visuo-spatial score. In the group of children with WS, however, the variance accounted for by VMA is unique and is not significantly shared with that predicted by visuo-spatial ability. Moreover, in the group of children with WS, when CA was entered first, it did not significantly account for the variance in the total number of marbles correctly given. In this group VMA is the only significant predictor of variance in the task. In the comparison group, however, CA was found to significantly account for variance in performance on the "give a number" task. Furthermore, after allowing for CA, only visuo-spatial ability significantly added to the variance accounted for, while VMA did not. These results suggest that in children with WS, VMA accounts for a significant amount of variance over and above CA and visuo-spatial ability. In typical development, by contrast, only visuo-spatial competence accounts for the level of cardinality understanding after controlling for CA and VMA.

A more conservative interpretation of our data could be that general intellectual ability (as reflected by VMA) is what predicts the counting ability of children with WS, and that there is nothing syndrome-specific that explains this relationship. However, not only VMA but also visuo-spatial ability is significantly correlated with chronological age in the large control group, but not the WS group. Although this strong effect of age explains the correlation between VMA and cardinality understanding, it does not account for the correlation between visuo-spatial ability and performance in the large comparison group. This is also strengthened by the results from the large comparison group showing that once CA is accounted for, visuo-spatial ability, but not VMA, accounts for performance variance. Thus, even in typically developing children, the development of cardinality appears to be driven by specific cognitive capacities rather than simply overall intellectual ability. Thus the groups rely on different cognitive competencies to guide the development of numerical cognition.

The most interesting finding to emerge from our study, then, is that although individuals with WS perform at the same level as their visuo-spatial controls, the factors that explain their performance are different from those that account for the understanding of the cardinality principle in typical development. It is important to stress, however, that the apparent compensation for the strong weakness in visuo-spatial cognition by the relative strength in WS language is imperfect, since typically developing children with the Verbal MAs at same level as those found in the WS group would already have full mastery of the cardinality principle.

Our findings show that the language strength in WS does not lead to normal development of exact number representation, as might have been predicted from adult models of numerical cognition (Dehaene, 2001; Dehaene et al., 1999). Children with WS were found to be at the level of the 3-year-old controls individually matched on visuo-spatial ability, and thus far below the level that would be expected given their verbal competence or chronological age. These results suggest that the impairment in visuo-spatial cognition in children with Williams syndrome prevents normal development of an understanding of the cardinality principle, which is not fully compensated for by their strength in language.

In terms of normal development our findings show that the understanding of the cardinality principle is driven by visuo-spatial competence to a greater extent than language ability. Thus, contrary to what might have been predicted from adult models of numerical cognition (Dehaene et al., 1999), typical development of exact number representation appears to be scaffolded, at least initially, by the development of non-verbal competencies. This is also supported by the Brannon and Van de Walle (2000) finding that knowledge of the verbal counting system is not causally related to typically developing children's understanding of ordinality. These authors found that 2- and 3-year-olds only required a minimal amount of verbal numerical knowledge to represent ordinal relations between numerosities as large as 6, suggesting that the development of early number competence is driven by non-verbal systems of magnitude representation. From these various results, we hypothesise that it is only over developmental time that non-verbal representation of number becomes integrated with verbal numerical competence to lead to a language-dependent representation of exact number. Thus, to understand the roots of problems with number representation in children with developmental disorders, it is crucial to trace the developmental trajectory. Moreover, our results again highlight the fact that equivalent behavioural scores between children with developmental disorders and their typically developing controls do not necessarily reflect the same underlying representational processes (Karmiloff-Smith, 1998).

The present results motivate further investigation into the atypical nature of the development of number representations in WS. In particular, they highlight the importance of investigating very basic number representations in all children suffering from impairments of numerical cognition. Whether our findings for WS are syndrome-specific, or more general to impairments of mathematical cognition in children with other developmental disorders who have, for instance, relative strengths in visuo-spatial cognition rather than language, awaits future cross-syndrome comparisons.

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References

Ansari, D., & Karmiloff-Smith, A. (2002). Atypical trajectories of number development: a neuroconstructivist perspective. *Trends in Cognitive Sciences, 6*, 511–516.

Arnold, R., Yule, W., & Martin, N. (1985). The psychological characteristics of infantile hypercalcemia: A preliminary investigation. *Developmental Medicine and Child Neurology, 27*, 49–59.

Bellugi, U., Marks, S., Bihrle, A., & Sabo, H. (1988). Dissociation between language and cognitive functions in Williams syndrome. In D. Bishop, & K. Mogford (Eds.), *Language development in exceptional circumstances* (pp. 177–189). London: Churchill Livingstone.

Beuren, A. J. (1972). Supravalvular aortic stenosis: a complex syndrome with and without mental retardation. *Birth Defects, 8*, 45–46.

Brannon, E. M., & Van de Walle, G. A. (2000). The development of ordinal numerical competence in young children. *Cognitive Psychology, 43*, 53–81.

Briars, D., & Siegler, R. S. (1984). A featural analysis of preschoolers' counting knowledge. *Developmental Psychology, 20*(4), 607–618.

Dehaene, S. (2001). Precis of the number sense. *Mind & Language, 16*, 16–36.

Dehaene, S., Spelke, E., Pinel, P., Stanescu, R., & Tsivkin, S. (1999). Sources of mathematical thinking: behavioural and brain-imaging evidence. *Science, 284*, 970–974.

Donnai, D., & Karmiloff-Smith, A. (2000). Williams syndrome: from genotype through to the cognitive phenotype. *American Journal of Medical Genetics, 97*, 164–171.

Elliot, C. D., Smith, P., & McCulloch, K. (1996). *British Ability Scales*. Windsor, UK: NFER-Nelson.

Ewart, A. K., Morris, C. A., Atkinson, D., Jin, W., Sternes, K., Spallone, P., Dean Stock, A., Leppert, M., & Keating, M. T. (1993). Hemizygosity at the elastin locus in a developmental disorder, Williams syndrome. *Nature Genetics, 5*, 11–16.

Fuson, K. C. (1988). *Children's counting and concepts of number*. New York: Springer-Verlag.

Gelman, R., & Gallistel, C. R. (1978). *The child's understanding of number*. Cambridge, MA: Harvard University Press.

Greenberg, E. (1990). Introduction to special issue on Williams syndrome. *American Journal of Medical Genetics Supplement, 6*, 85–88.

Karmiloff-Smith, A. (1998). Development itself is the key to understanding developmental disorders. *Trends in Cognitive Sciences, 2*, 389–398.

Mervis, C. B., Robinson, B. F., Bertrand, J., Morris, C. A., Klein-Tasman, B. P., & Armstrong, S. C. (2000). The Williams syndrome cognitive profile. *Brain and Cognition, 44*, 604–628.

Paterson, S., Girelli, L., Butterworth, B., & Karmiloff-Smith, A. (2002). Are numerical difficulties syndrome specific? Evidence from Williams Syndrome and Down's Syndrome. Unpublished manuscript.

Tassabehji, M., Metcalfe, K., Fergusson, W. D., Carette, M. J., Dore, J. K., Donnai, D., Read, A. P., Proschel, C., Gutowski, N. J., Mao, X., & Sheer, D. (1996). LIM-kinase deleted in Willimas syndrome. *Nature Genetics, 13*, 272–273.

Thomas, M. S. C., Dockrell, J., Messer, D., Parmigiani, C., Ansari, D., & Karmiloff-Smith, A. (2002). Naming in Williams syndrome. Submitted.

Udwin, O., Davies, M., & Howlin, P. (1996). A longitudinal study of cognitive and education attainment in Williams syndrome. *Developmental Medicine and Child Neurology, 38*, 1020–1029.

Wynn, K. (1990). Children's understanding of counting. *Cognition, 36*, 155–193.

Wynn, K. (1992). Children's acquisition of the number words and the counting system. *Cognitive Psychology, 24*, 220–251.