Journal of Clinical and Experimental Neuropsychology
Publication details, including instructions for authors and subscription information:
http://www.informaworld.com/smpp/title~content=t713657736

Numerical abilities in Williams syndrome: Dissociating the analogue magnitude system and verbal retrieval

Attila Krajcsi [ab]; Ágnes Lukács [c]; János Igács [a]; Mihály Racsmány [ac]; Csaba Pléh [cd]

[a] University of Szeged, Institute of Psychology, Szeged, Hungary
[b] Department of Radiology, Harvard Medical School, Brigham & Women's Hospital, Boston, MA, USA
[c] Cognitive Science Research Group, HAS-Budapest University of Technology, Budapest, Hungary
[d] Department of Cognitive Science, Budapest University of Technology and Economics, Budapest, Hungary

First Published on: 11 July 2008

To link to this article: DOI: 10.1080/13803390802244126
URL: http://dx.doi.org/10.1080/13803390802244126

JOURNAL OF CLINICAL AND EXPERIMENTAL NEUROPSYCHOLOGY
2008, i*First*, 1–8

Psychology Press
Taylor & Francis Group

Numerical abilities in Williams syndrome: Dissociating the analogue magnitude system and verbal retrieval

Attila Krajcsi,[1,2] Ágnes Lukács,[3] János Igács,[1] Mihály Racsmány,[1,3] and Csaba Pléh[3,4]

[1]University of Szeged, Institute of Psychology, Szeged, Hungary
[2]Harvard Medical School, Brigham & Women's Hospital, Department of Radiology, Boston, MA, USA
[3]HAS–Budapest University of Technology, Cognitive Science Research Group, Budapest, Hungary
[4]Budapest University of Technology and Economics, Department of Cognitive Science, Budapest, Hungary

Two numerical systems—the analogue magnitude system and verbal retrieval—were investigated in Williams syndrome (WS) with three numerical tasks: simple addition, simple multiplication, and number comparison. A new matching technique was introduced in selecting the proper control groups. The WS group was relatively fast in the addition and multiplication tasks, but was slow in number comparison. No reverse numerical effect was observed in the comparison task, and the distance effect was stronger than that in the control groups. The findings indicate a profile with an impaired analogue magnitude system and less impaired verbal retrieval in Williams syndrome.

Keywords: Williams syndrome; Numerical ability; Analogue magnitude system; Matching by target task.

INTRODUCTION

Numerical abilities

Numerical abilities comprise several representations and mechanisms that are responsible for different numerical processes. The value of numerals is processed by a domain-specific mechanism: the analogue magnitude system. This system is involved in several numerical tasks like estimation, approximation, and number comparison (Dehaene, Spelke, Pinel, Stanescu, & Tsivkin, 1999; Spelke & Tsivkin, 2001). However, this magnitude system is also activated in numerical tasks in which the semantics of the numerals is not essential (Dehaene, 1997). This representation is associated with the activation of the intraparietal sulcus (Chochon, Cohen, van de Moortele, & Dehaene, 1999; Eger, Sterzer, Russ, Giraud, & Kleinschmidt, 2003; Piazza & Dehaene, 2004). In comparison tasks the contribution of the magnitude system is indicated by the numerical distance effect (Moyer & Landauer, 1967): The smaller the numerical distance is between the two numerals to be compared, the longer the response latency.

Exact addition and multiplication heavily rely on the overlearned addition and multiplication table (Campbell, 1994) based on verbal retrieval (Spelke & Tsivkin, 2001). It is characterized by the size effect, which reflects search time in the addition or multiplication table: The response latency is proportional to the size of the operands (Ashcraft, 1992; Groen & Parkman, 1972). The verbal retrieval also shows a tie effect in multiplication and addition: Response latencies are shorter when the two operands are the same (e.g., 3×3 or $5 + 5$)

Address correspondence to Attila Krajcsi, University of Szeged, Department of Psychology, Petőfi sgt. 30–34 H-6722 Szeged, Hungary (E-mail: krajcsi@gmail.com).

http://www.psypress.com/jcen DOI: 10.1080/13803390802244126

than when they are different (3 × 4 or 5 + 2). Besides the retrieval mechanism, arithmetical problems can be solved by several other strategies as well (Siegler, 1999). A well-known mechanism relies on rules: Adding and multiplying by 0 or 1 can be solved by simple rules like $1 \times n = n$ or $0 \times n = 0$ (Ashcraft, 1992; McCloskey, 1992). Such a detailed specification of numerical representations and processing allows us to study developmental disorders at a fine-grained level.

Numerical abilities in Williams syndrome

Williams syndrome (WS) is a rare (1 in 20,000) genetically based condition caused by the microdeletion of genes on the long arm of chromosome 7 (Arnold, Yule, & Martin, 1985; Monaco, 1996; Williams, Barratt-Boyes, & Lowe, 1961). Children with this syndrome are characterized by mild to moderate mental retardation and impaired spatial cognition and motor skill learning, but relatively well-functioning language and social disinhibition (Bellugi, Lichtenberger, Jones, & Lai, 2000; Brock, 2007; Mayer-Lindenberg, Mervis, & Berman, 2006). The WS IQ range is between 40 and 90 (Bellugi, Wang, & Jernigan, 1994). In contrast to relatively good linguistic abilities, spatial-visual cognition is highly impaired in people with WS (Bellugi et al., 2000; Lukács, Pléh, & Racsmány, 2004). WS persons show an impaired performance on the Block Design task of the Wechsler test (Wang, Doherty, Rourke, & Bellugi, 1995). A longitudinal study of verbal and spatial abilities revealed that the two cognitive domains develop in a divergent way, and the difference between the two abilities increases with age in these subjects (Jarrold, Baddeley, & Hewes, 1998).

Until now, there have been only a few studies investigating numerical abilities in WS. Most of the studies have shown a general deficit in mathematics without investigating numerical abilities at a fine-grained level. In a study by Howlin, Davies, and Udwin (1998) adults with WS performed at the level of 7–8-year-old children in the Wechsler Adult Intelligence Scale–Revised (WAIS–R) arithmetical task. They also found that development reaches a plateau at about 8 years of mental age. Bellugi, Marks, Bihrle, and Sabo (1988) tested 3 participants with WS on two classical Piaget tasks. Children with WS had problems with the seriation task and the conservation task.

In an experiment by Paterson and her colleagues (Paterson, Girelli, Butterworth, & Karmiloff-Smith, 2006) participants had to compare two arrays of dots. The WS group showed a somewhat reduced distance effect in reaction time (RT) than did participants with Down syndrome (DS) and two control groups matched on chronological and mental age. In another experiment of Paterson et al. (2006) participants with WS solved several numerical problems. Their performance was compared to that of a DS group. A simple counting task (counting from 1 to 20) was solved correctly, but the WS group showed impaired performance in counting from 25, counting backward, the "what comes next/before" task (e.g., say the number followed by 14), reading Arabic numerals, a seriation task (ordering four Arabic stimuli or ordering four patterns of dots), and arithmetic tasks (especially in the multiplication task).

Ansari, Donlan, and Karmiloff-Smith (2007) presented arrays of dots to WS participants and found that WS performance was delayed. Furthermore, the distribution of the answers was atypical in WS.

All of the studies presented above reveal equally weak performance in all aspects of numerical abilities or at least were unable to differentiate between relatively good and bad performing functions. However, there is some evidence that individuals with WS show selective impairment of numerical skills, which suggests possible dissociations between various components of the human numerical system.

Ansari and his colleagues (2003) studied the *How many* (counting small set of items) and *Give a number* (participants should give a certain number of marbles to a puppet) tasks in WS and a control group matched on a spatial–visual task. Counting performance was very similar in the WS and control groups. While in the control group, the best predictors of counting performance were chronological age and spatial–visual abilities; in the WS group the best predictors of counting performance were verbal abilities. This result reveals that counting performance highly relies on verbal abilities in WS, as the result of compensation for weak spatial–visual and probably impaired semantic magnitude system.

O'Hearn and Landau (2007) tested several mathematical skills in WS with the Test of Early Mathematical Ability (TEMA-2). Overall mathematical performance did not differ between WS participants and a control group matched for mental age. However, the WS group performed more poorly in tasks that rely on magnitude system, like number comparison. On the other hand, participants with WS showed an improved performance in verbal numerical tasks—for example, in reading numbers.

When investigating numerical disabilities, a methodological issue arises regarding interpreting

RT differences and numerical effect differences between atypical and control groups. Absent or atypical numerical effects can be the sign of impaired functions in developmental disorders. More specifically, an altered distance effect as a symptom of an impairment of the analogue magnitude system is cited most frequently as the possible cause of numerical disabilities (Bruandet, Molko, Cohen, & Dehaene, 2004; Butterworth, 1999; Paterson et al., 2006). The question is what the direction of this change is when a function is impaired. According to the most radical view, impaired functions could be replaced by compensatory mechanisms; thus the effect can even reverse (Butterworth, 1999). For instance, when participants are unable to compare two numerals with the magnitude system, they can use an alternative counting strategy—for example, by choosing one number and counting on until one reaches the other number. The response latency increases as the distance between the two numbers increases, and this way this compensatory mechanism would predict a reversed distance effect. A second possibility is that impaired functions are reflected in longer response latencies (Bruandet et al., 2004; Landerl, Bevan, & Butterworth, 2004), regardless of the effects. A third position suggests that weak performance is signaled by decreased effect sizes (Paterson et al., 2006). Rethinking the second and third possibilities, we have to consider that faster RTs generally show a smaller distance effect, and slower RTs are associated with a larger distance effect. One can argue then, on the one hand, that the deficit of the magnitude system is revealed in slow comparison RTs (and thus a large distance effect) or, on the other hand, impairment can be seen in a decreased distance effect (and thus fast RTs). These two interpretations clearly contradict each other.

Our main goal in the study was to test whether there is dissociation in WS between performance on numerical tasks that rely on the verbal system and those that exploit the analogue magnitude systems. To answer this question, error rates and response latencies were collected both from tasks that require verbal retrieval (simple one digit addition and simple one digit multiplication) and from a task that requires the activation of the parietal magnitude system (one digit number comparison). Former studies showed that typically developing children use the retrieval strategy in multiplication by the end of second grade (Lemaire & Siegler, 1995). Based on previous findings on numerical abilities (Dehaene et al., 1999) and WS (Ansari et al., 2003; O'Hearn & Landau, 2007) we expected to find a relative advantage of simple addition and multiplication mediated by verbal systems, and a relative disadvantage of comparison mediated by the analogue magnitude system.

METHOD

Participants

We tested 8 participants with Williams syndrome. All of them were positively diagnosed by the fluorescence in situ hybridization (FISH) test. Participants were selected from a larger pool of all diagnosed Hungarian-speaking individuals with WS. Only those individuals who could solve the simple arithmetical tasks required by the test participated in the study. Mean age of the participants (5 female and 3 male) was 17 years 8 months (17;8; range: 12;0 to 23;1). Informed consent was obtained from participants and parents. We selected three groups of typically developing children as potential control groups from the three youngest age groups that could solve the three numerical tasks: 10 second graders (5 females, mean age 8;5), 10 third graders (5 females, mean age 9;7), 11 fourth graders (6 females, mean age 10;3). Control groups were recruited through a general primary school for typically developing children.

Matching by target task

Matching the control group on mental age would have been inappropriate for our study, as the control group would have included preschool children, who would have had less experience with numbers and arithmetical education. This is a general problem with matching by mental age: A younger group always has less experience, leading to an inevitable methodological confound. For this reason, we had to find a novel way of matching.

The matching technique used in our study relies on the aim to find dissociations between abilities. The most frequent matching method controls one ability (*control variable* on which the control group is matched), and measures another ability (*target variable*). This method allows an inference that the clinical group has a deficit on the target variable compared with the control group matched on control variable. In other words it means that the deficit in the target variable cannot be attributed to a more general deficit that is measured by the control variable. This conclusion reveals dissociations between the abilities measured by the control and the target variables.

In the present study an alternative and more effective technique of matching was used. To find a dissociation, it should be enough to show that there is no difference between the clinical and control groups in one task, while there is a difference in another task. This way, performance on one task (on which performance is similar) serves as a control variable, and performance on the other task is the target variable. The performance pattern on the two tasks can reveal a dissociation between the two tasks in the two groups. In our study participants had to solve three tasks. If there is one task among the three on which there is no difference between the clinical and control groups, then we use that specific task for matching (i.e., as a control variable). If we find a difference between the groups in the other task(s), that is a sign of a dissociation between the two tasks. On the other hand, if there is no difference between the clinical and control groups on any of the tasks, that may be the sign of a general delay (based on the age differences between the groups). In traditional matching terminology the first task serves as a control variable, and the second task is a target variable. However, the first task is also a target variable at the same time, because the task is evaluated as part of the dissociation. Matching by mental age requires an extra variable that can be used only as a control variable, while in matching by target task it is enough to use one of the target variables as a control variable. In the present study three numerical tasks are used: comparison, addition, and multiplication. To take an example, if the performance of the WS group and one control group is at the same level on the comparison task and differs on addition, we can conclude that addition dissociates from comparison.

Matching by target task has another advantage compared to matching by mental age, as it is easier to find a control group: Any of the target tasks can be used as a control task as none of them has a distinguished role. However, in matching by mental age only mental age (e.g., nonverbal IQ) could be the control task. As control groups are found post hoc in both the mental age control method and target task control method, and there are several possible candidates for control tasks in the target task method, it is less time consuming to find appropriate control participants with the latter. Furthermore, if mental age is not required for matching, we can also get around the problem of finding the appropriate test to measure mental age validly.

For the above reasons, we believe that the matching technique used in our study fits the aim of finding dissociations between abilities better than matching on some sort of mental age: (a) The comparison of target variables is more direct than that using an indirect control variable, and thus the measurement of dissociation is more valid; (b) the technique of matching does not require the frequently debated measurement of mental age; (c) more specifically, in our study the control group could be older than preschool children are.

Stimuli

In the *comparison* task, participants had to decide which one of two Arabic numerals had a larger value. The two numerals appeared simultaneously on the left and right sides of a computer screen. Numbers were between 1 and 8. The stimulus pairs contained all the possible combinations with a distance of 1, 3, 4, 6, and 7 between the two numerals and all possible pairings with the larger number both on the left side and on the right side. All number pairs were presented three times in a task, resulting in 114 trials in the comparison task altogether.

In the *addition* task the participants had to decide whether an addition with its result was correct or not—for example, 5 + 3 = 7. The operands were between 0 and 8, and thus the results were between 1 and 16. The task contained all the possible combinations of the operand (the 0 + 0 tie problem excluded) with the greater operand in the first position. There were two trials for each addition: one with the correct, and one with an incorrect result. In the incorrect addition trials the result differed from the correct result by one. The task included 88 trials.

In the *multiplication* task participants also had to decide whether a multiplication with a result was correct or not. The operands were again between 0 and 8. The task contained all the possible combinations of the operand (the 0*0 tie problem excluded) with the greater operand in the first position. There were two trials for each multiplication: one with the correct, and one with an incorrect result. In the incorrect multiplication trials the result was a number in a neighboring cell to the correct result in the multiplication table. The task included 88 trials.

In the addition and multiplication tasks all trials were tagged according to the following criteria: (a) Trials with same operand are *tie trials* (e.g., 3 × 3); (b) trials with an operand 0 are *operand 0 trials* (e.g., 4 + 0); (c) trials with an operand 1 are *operand 1 trials* (e.g., 1 × 8); (d) trials with both operand smaller than 6 and not tie, operand 0, or operand 1 trials are *small trials* (e.g., 3 + 5); (e) trials with at least one operand with 6 or larger and not tie, operand 0, or operand 1 trials are *large trials* (e.g., 6*3).

Procedure

All participants solved the simple comparison, simple addition, and multiplication tasks in this order. The order of the three tasks was fixed, but trials in a task were randomized within that specific task. All stimuli appeared in white fonts on a black background on a standard computer monitor. In each task after the instruction the participants had to indicate the correct answer by pressing one of the two response buttons on the keyboard. The stimuli were visible until the response button was pressed. No feedback was given. There was a 1,000-ms delay between pressing the response button and the beginning of the next trial. Presentation of the stimuli and measurement of RT were managed by Presentation software (Neurobehavioral Systems, 2003).

RESULTS

The error rates were relatively low (less than 20%) in all groups and in all conditions, and the pattern of error rates was similar to response latency patterns. For this reason only the reaction time analysis is presented here. The response latencies of the four groups in the three tasks are shown in Figure 1. Individual performance of every single participant with WS is presented in Table 1. Three between-subject analyses of variance (ANOVAs) revealed that in all of the three tasks the differences between the groups were significant, $F(3, 35) = 6.34$, $p = .002$ in comparison, $F(3, 35) = 6.19$, $p = .002$ in addition, and $F(3, 35) = 5.52$, $p = .003$ in multiplication. Post hoc pairwise comparison revealed that in the comparison task, the WS group was slower than any other control groups, in the addition task 4th graders were faster than any other three groups, and in the multiplication task 4th graders were faster than the 2nd and 3rd graders.

To summarize the results of response latency analysis, the WS group performed extremely poorly in the comparison task, lagging significantly behind even 2nd graders, although they performed at the level of 2nd and 3rd graders in the addition task, and they performed at the level of the other control groups in the multiplication task. These results show that based on the addition task, 2nd and 3rd graders can be the control groups of the WS group. Based on the multiplication task, any of the typically developing groups in our study could serve as a control group of our WS group. Matching this way on

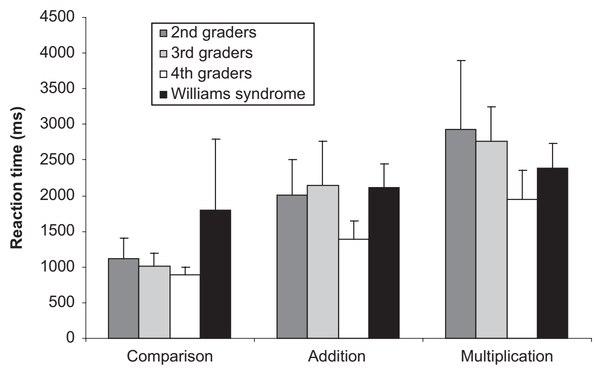

Figure 1. Mean response latency of four groups in comparison, addition, and multiplication tasks. Error bar represents standard deviation.

TABLE 1
Error rate and response latency of the 8 individuals with Williams syndrome in the three tasks

	Mean error rate			*Median reaction time (ms)*		
Participant	*Comparison*	*Addition*	*Multiplication*	*Comparison*	*Addition*	*Multiplication*
1	0.18	0.13	0.28	1,346	2,022	2,304
2	0.32	0.30	0.44	547	2,802	2,411
3	0.35	0.11	0.14	2,052	2,128	1,913
4	0.04	0.03	0.02	2,802	2,055	2,359
5	0.05	0.09	0.16	1,993	2,243	3,085
6	0.02	0.08	0.09	3,512	2,160	2,228
7	0.18	0.17	0.25	1,214	1,792	2,217
8	0.03	0.18	0.20	964	1,662	2,589

either addition or multiplication (which are not dissociated) shows that comparison is a function of a distinct system, which seems to be impaired in WS, as performance on this task lags behind control groups matched on the other two tasks.

Numerical effects

As mentioned above, a developmental deficit of numerical abilities can also manifest itself in the distortion of effects associated with numerical processing. Paterson et al. (2006) found smaller distance effect in WS, and Butterworth (1999) found a reverse distance effect.

In the comparison task we measured the distance effect, while in the addition and multiplication tasks the tie effect, size effect, and rule effects were tested (see Introduction and Method for a description of these effects). Figure 2 shows response latencies in the three tasks as a function of numerical effects. Three 5 (numerical effect) × 4 (groups) ANOVAs were performed in all three tasks. The levels of the numerical effect in the comparison task are the distances between the two numerals. The levels of the numerical effects in addition and multiplication tasks were (a) trials with operand 0 and (b) trials with operand 1 (rule effect), (c) tie problems (tie effect), and (d) trials with small result and (e) trials with large result (size effect). (Although the five effects of addition and multiplication originate in two different cognitive modules, i.e., verbal retrieval and rule production, statistically it is appropriate to handle these trial types together.) Numerical effects had an effect on response latency in all tasks: $F(4, 140) = 16.2$, $p < .001$, in comparison, $F(4, 140) = 32.2$, $p < .001$, in addition, and $F(4, 140) = 29.02$, $p < .001$, in multiplication. The group had a significant effect in all three tasks as it was also revealed in the previous analysis. The interaction between numerical effects and groups was significant only in the comparison task, $F(12, 140) = 2.63$, $p < .001$ (revealing a greater distance effect in the WS group), while there was only a tendency in the other two tasks, $F(12, 140) = 1.66$, $p = .08$ in addition, and $F(12, 140) = 1.72$, $p = .07$ in multiplication. To ensure that the distance effect of the comparison task is present in the control groups (as the interaction and the increased distance effect in WS group could mean the lack of distance effect in the control groups) another 5 (distance) × 3 (control groups) ANOVA was run. The distance effect was again significant, $F(4, 112) = 11.0$, $p < .001$. The three groups showed small differences, $F(2, 28) = 3.3$, $p = .053$, without interaction of the effect and groups.

The distance effect in the comparison task was not reversed for any participants including the WS group. The ANOVA revealed similar numerical effects in all four groups; however, it is worth noting that only 8 WS individuals participated in the study, and possibly more subtle differences in addition and multiplication tasks could only be found with greater statistical power.

DISCUSSION

The present study investigated numerical abilities in WS at a more fine-grained level. The main question was whether a specific dissociation

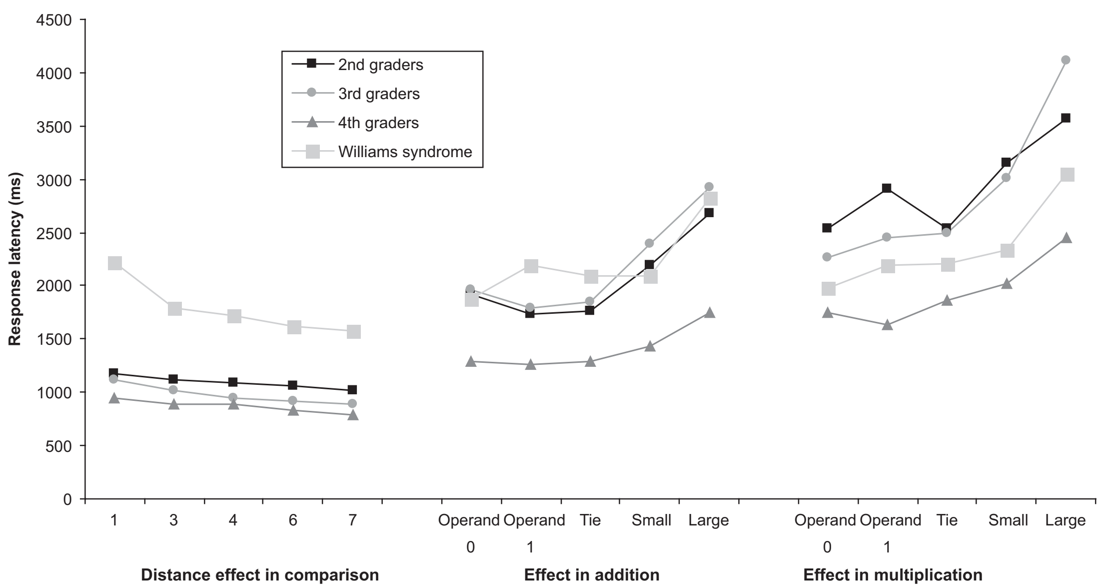

Figure 2. Numerical effects in comparison, addition, and multiplication tasks. Error bar represents standard deviation.

between different numerical functions can be observed. The data presented above show poor WS performance in the comparison task (worse performance than that in 2nd graders) compared to relatively better performance in addition and multiplication tasks (performance at the level of 2nd, 3rd, and partly 4th graders). These results reflect an impaired numerical magnitude system associated with the intraparietal sulcus (Dehaene et al., 1999; Eger et al., 2003; Piazza & Dehaene, 2004) with a less impaired verbal retrieval system (Siegler, 1999; Spelke & Tsivkin, 2001). This conclusion is in line with previous non-numerical results of people with WS, contrasting a relatively good verbal performance and a bad spatial function located in the parietal lobe. The present conclusion is in line with the results of a more general investigation completed by O'Hearn and Landau (2007), showing relatively intact number reading and a relatively impaired number comparison. These results confirm the distinct functions of the analogue magnitude system and verbal retrieval in numerical tasks. The data reveal a pattern of peaks and valleys of numerical abilities in WS, instead of a generally weak overall mathematical performance. Knowing the exact nature of numerical abilities could serve as a starting point in training mathematical skills in WS. The verbal system could be the base of a compensatory mechanism; however, it is still unknown how effective such a substitute process could be in different types of numerical tasks. The addition and multiplication performance of the WS group was better than that reported in the study of Paterson et al. (2006). Educational differences between the two samples could be a reasonable cause of the discrepancies.

One can note that small sample size (8 participants with WS) results in low statistical power, and thus subtle differences cannot be shown. We agree; however, our point is to show that the impairment in comparison task is so severe that it is significantly detectable even with this limited statistical power.

Our results have a methodological consequence regarding the effect of impairment on response latencies and RT patterns. Results revealed that the usual numerical effects (like the distance effect, the size effect, and the tie effect) are present in the WS group, showing a similar pattern of RTs to that observed in the control groups. No reverse effect was found in our sample. To our knowledge the only reverse effect was described by Butterworth (1999), and even he noted that this case is rather extreme (Butterworth, 2005). WS participants showed greater response latency in the comparison task, and the distance effect was bigger than the distance effect found in the control group. This result is inconsistent with the results published by Paterson et al. (2006) where the distance effect was smaller than the effect in the control groups. At the same time, there were only 8 WS individuals in both studies. Furthermore, the RT pattern of WS individuals with high error rate is noisier than that of WS participants with low error rate. Thus, because of the relatively instable RT pattern of lower functioning WS participants, response latency data might be less reliable.

The results have another methodological implication. The usual method of matching by mental age (based on IQ) would have been inappropriate in the present study, as mental-age controls would have been too young to be able to solve arithmetical tasks. Instead, we used control groups whose performance was similar to the performance of the WS group in one of the specific tasks used in the study (matching by target task). As the WS group performed in the addition task at the level of 2nd and 3rd graders and in the multiplication task at a level of 2nd, 3rd, and 4th graders, any of these two tasks could serve as control variables, and thus the addition and multiplication tasks are target and control tasks in the same time. The worse performance of the WS group in the comparison task than that of all control groups suggests dissociation between the three tasks (comparison vs. addition and multiplication) without the usage of a further control variable—for example, nonverbal intelligence. One might point out that the missing control variable makes the results invalid. However, the use of a target task as control variable makes inferences more direct and thus more valid. Finding a dissociation is more direct when comparing the performance of two abilities (e.g., the analogue magnitude system measured by comparison task vs. verbal retrieval measured by simple addition and multiplication) omitting the intermediate control variable like general intelligence. Furthermore, the concept of mental age is problematic as there is no steady agreement about which part of general intelligence measurement should be used (e.g., verbal, performance, etc.). This problem is even more prominent in developmental disorders where the peaks and valleys of abilities show an atypical pattern. Matching by target task thus provides us a more valid and direct method of finding a dissociation in developmental disorders.

Original manuscript received 18 January 2008
Revised manuscript accepted 2 June 2008
First published online day month year

REFERENCES

Ansari, D., Donlan, C., & Karmiloff-Smith, A. (2007). Atypical and typical development of visual estimation abilities. *Cortex: Special Issue on Selective Developmental Disorders, 6,* 758–768.

Ansari, D., Donlan, C., Thomas, M. S. C., Ewing, S. A., Peen, T., & Karmiloff-Smith, A. (2003). What makes counting count? Verbal and visual-spatial contributions to typical and atypical number development. *Journal of Experimental Child Psychology, 85*, 50–62.

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

Ashcraft, M. H. (1992). Cognitive arithmetics: A review of data and theory. *Cognition, 44*, 75–106.

Bellugi, U., Lichtenberger, L., Jones, W., & Lai, Z. (2000). The neurocognitive profile of Williams syndrome: A complex pattern of strengths and weaknesses. *Journal of Cognitive Neuroscience, 12*, 17–29.

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). Edinburgh, UK: Churchill Livingstone.

Bellugi, U., Wang, P. P., & Jernigan, T. L. (1994). Williams syndrome: An unusual neuropsychological profile. In S. Broman & J. Grafman (Eds.), *Atypical cognitive deficits in developmental disorders: Implications for brain function* (pp. 23–56). Hillsdale, NJ: Lawrence Erlbaum Associates.

Brock, J. (2007). Language abilities in Williams syndrome: A critical review. *Development and Psychopathology, 19*, 97–127.

Bruandet, M., Molko, N., Cohen, L., & Dehaene, S. (2004). A cognitive characterization of dyscalculia in Turner syndrome. *Neuropsychologia, 42*, 288–298.

Butterworth, B. (1999). *The mathematical brain*. London: Macmillan.

Butterworth, B. (2005). Developmental dyscalculia. In J. I. D. Campbell (Ed.), *Handbook of mathematical cognition*. New York: Psychology Press.

Campbell, J. I. D. (1994). Architecture for numerical cognition. *Cognition, 53*, 1–44.

Chochon, F., Cohen, L., van de Moortele, P. F., & Dehaene, S. (1999). Differential contribution of the left and right inferior parietal lobulus to number processing. *Journal of Cognitive Neuroscience, 11*, 617–630.

Dehaene, S. (1997). *The number sense: How the mind creates mathematics*. New York: Oxford University Press.

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

Eger, E., Sterzer, P., Russ, M. O., Giraud, A.-L., & Kleinschmidt, A. (2003). A supramodal representation in human intraparietal cortex. *Neuron, 37*, 719–725.

Groen, G. J., & Parkman, J. M. (1972). A chronometric analysis of simple addition. *Psychological Review, 79*, 329–343.

Howlin, P., Davies, M., & Udwin, O. (1998). Cognitive functioning in adults with Williams syndrome. *Journal of Child Psychology and Psychiatry, 39*, 183–189.

Jarrold, C., Baddeley, A. D., & Hewes, A. K. (1998). Verbal and nonverbal abilities in the Williams syndrome phenotype: Evidence for diverging developmental trajectories. *Journal of Child Psychology & Psychiatry & Allied Disciplines, 39*, 511–523.

Landerl, K., Bevan, A., & Butterworth, B. (2004). Developmental dyscalculia and basic numerical capacities: A study of 8–9 year old students. *Cognition, 93*, 99–125.

Lemaire, P., & Siegler, R. S. (1995). Four aspects of strategic change: Contributions to children's learning of multiplication. *Journal of Experimental Psychology: General, 124*, 83–97.

Lukács, Á., Pléh, Cs., & Racsmány, M. (2004). Language in Hungarian children with Williams syndrome. In S. Bartke & J. Siegmüller (Eds.), *Williams syndrome across languages* (pp. 187–220). Amsterdam: John Benjamins Publishing Company.

Mayer-Lindenberg, A., Mervis, C. B., & Berman, K. F. (2006). Neural mechanism in Williams syndrome: A unique window to genetic influences on cognition and behaviour. *Nature Reviews Neuroscience, 7*, 380–393.

McCloskey, M. (1992). Cognitive mechanisms in numerical processing: Evidence from acquired dyscalculia. *Cognition, 44*, 107–157.

Monaco, A. P. (1996). Dissecting Williams syndrome. *Current Biology, 6*, 1396–1398.

Moyer, R. S., & Landauer, T. K. (1967). Time required for judgement of numerical inequality. *Nature, 215*, 1519–1520.

Neurobehavioral Systems. (2003). Presentation (Version 0.76) [Computer software]. Available from http://www.neurobs.com/.

O'Hearn, K., & Landau, B. (2007). Mathematical skill in individuals with Williams syndrome: Evidence from a standardized mathematics battery. *Brain and Cognition, 64,* 238–246.

Paterson, S. J., Girelli, L., Butterworth, B., & Karmiloff-Smith, A. (2006). Are numerical impairments syndrome specific? Evidence from Williams syndrome and Down's syndrome. *Journal of Child Psychology and Psychiatry, 47*, 190–204.

Piazza, M., & Dehaene, S. (2004). From number neurons to mental arithmetic: The cognitive neuroscience of number sense. In M. Gazzaniga (Ed.), *The cognitive neuroscience* (3rd ed.). Cambridge, MA: MIT Press.

Siegler, R. S. (1999). Strategic development. *Trends in Cognitive Sciences, 3*, 430–435.

Spelke, E. S., & Tsivkin, S. (2001). Language and number: A bilingual study. *Cognition, 78*, 45–88.

Wang, P. P., Doherty, S., Rourke, S. B., & Bellugi, U. (1995). Unique profile of visuo-perceptual skills in a genetic syndrome. *Brain and Cognition, 29*, 54–65.

Williams, J. C. P., Barratt-Boyes, B. G., & Lowe, J. B. (1961). Supravalvular aortic stenosis. *Circulation, 24*, 1311.