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PAPER

Learning without representational change: development of numerical estimation in individuals with Williams syndrome

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Abstract

Experience engenders learning, but not all learning involves representational change. In this paper, we provide a dramatic case study of the distinction between learning and representational change. Specifically, we examined long- and short-term changes in representations of numeric magnitudes by asking individuals with Williams syndrome (WS) and typically developing (TD) children to estimate the position of numbers on a number line. As with TD children, accuracy of WS children's numerical estimates improved with age (Experiment 1) and feedback (Experiment 2). Both long- and short-term changes in estimates of WS individuals, however, followed an atypical developmental trajectory: as TD children gained in age and experience, increases in accuracy were accompanied by a logarithmic-to-linear shift in estimates of numerical magnitudes, whereas in WS individuals, accuracy increased but logarithmic estimation patterns persisted well into adulthood and after extensive training. These findings suggest that development of numerical estimation in WS is both arrested and atypical.

Introduction

Williams syndrome (WS) is a neurodevelopmental genetic disorder that affects roughly 1–1.3 in 10,000 (Strømme, Bjørnstad & Ramstad, 2002) and is caused by a contiguous hemizygous deletion of approximately 28 genes on chromosome 7 (7q11.23) (Ewart, Morris, Atkinson, Jin, Sternes, Spallone, Stock, Leppert & Keating, 1993; Peoples, Franke, Wang, Perez-Jurado, Paperna, Cisco & Francke, 2000). WS is associated with mild to moderate intellectual disability, physical abnormalities such as vascular stenoses, and atypical facial characteristics (Bruno, Rossi, Thuer, Cordoba & Alday, 2003; Martens, Wilson & Reutens, 2008; Morris & Mervis, 1999). The cognitive profile of WS is described as fractionated. Some aspects of language and face processing are relatively spared (Brock, 2007; Gagliardi, Bonaglia, Selicorni, Borgatti & Giorda, 2003; Karmiloff-Smith, Tyler, Voice, Sims, Udwin, Howlin & Davies, 1998), whereas visuospatial abilities are significantly impaired (Farran & Jarrold, 2004, 2005; Hoffman, Landau & Pagani, 2003; Nakamura, Watanabe, Matsumoto, Yamanaka, Kumagai, Miyazaki, Matsushima & Mita, 2001). The vast majority of individuals with WS demonstrate academic delays that also show evidence of variability across content areas: reading and spelling performance falls below average compared to typically developing (TD) children (Laing, Hulme, Grant & Karmiloff-Smith, 2001; Levy, Smith & Tager-Flusberg, 2003), and most mathematical skills are significantly delayed (Ansari, Donlan & Karmiloff-Smith, 2007; Howlin, Davies & Udwin, 1998; Paterson, Girelli, Butterworth & Karmiloff-Smith, 2006).

One reason math skills are significantly delayed in individuals with WS may be due to impaired representations of numeric magnitude (O'Hearn & Landau, 2007; O'Hearn & Luna, 2009). Generally, representations of numeric magnitude underlie one's ability to estimate and compare the absolute value of numeric quantities, to compare the magnitudes denoted by numerals, and to estimate the position of numbers on a number line (Dehaene, 1989; Piazza, Izard, Pinel, Bihan & Dehaene, 2004; Siegler & Opfer, 2003); proficiency in these activities also correlates positively with math proficiency (Booth & Siegler, 2008; Geary, Hoard, Byrd-Craven, Nugent & Numtee, 2007; Halberda, Mazzocco & Feigenson, 2008).

Several observations suggest that children with WS possess impaired numerical-magnitude representations. First, discriminating between large number sets (e.g. 8 vs. 16 dots) is much more difficult for WS 3-year-olds than TD 6-month-old infants (Van Herwegen, Ansari, Xu &

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Karmiloff-Smith, 2008; Xu & Spelke, 2000). Second, WS adolescents and adults have more difficulty comparing relative magnitudes of two single-digit numerals to a target than TD 6-year-olds (O'Hearn & Landau, 2007). Third, individuals with WS perform poorly on mental number line tasks, in which they are asked to determine which of two numbers is closest to a target number (O'Hearn & Landau, 2007). In addition, accuracy of numerosity estimation increases much more for TD children than WS children, with accuracy of WS adults' estimates being slightly less than that of TD 6- to 7-year-old children (Ansari *et al*., 2007). Thus, across many studies, tasks, and age groups, individuals with WS demonstrate age-inappropriate representations of numerical magnitude.

One way that numerical-magnitude representations might be impaired in individuals with WS is that magnitudes of numbers are represented as increasing logarithmically with actual value (as on a logarithmically scaled mental number line), whereas TD children can transition from logarithmic to linear representations over a relatively short period of time and with little experience necessary (Opfer & Siegler, 2007; Opfer & Thompson, 2008; Siegler & Opfer, 2003; Thompson & Opfer, 2008; for discussion, see also Barth & Paladino, 2011, and Opfer, Siegler & Young, 2011). How might previously observed errors of WS children be generated by a logarithmically scaled mental number line? Consider proficiency in number comparison, which is generally correlated positively with the distance between two numbers (e.g. 1 and 8 is more quickly and accurately compared than 6 and 8); (Krajcsi, Lukács, Igács, Racsmány & Pléh, 2009; Moyer & Landauer, 1967; Sekuler & Mierkiewicz, 1977). On a logarithmic number line, the distance between two numbers differs from the distance between the same two numbers on a linear number line. For example, on a scale from 1 to 16, the distance between 8 and 16 on a logarithmic scale is less than the distance between 8 and 16 on a linear scale. From this perspective, the fact that WS 3-year-olds have greater difficulty comparing 8 and 16 than TD children makes sense if WS 3-year-olds represent 1–16 logarithmically (where 8 and 16 seem close), whereas TD 3-year-olds represent that range linearly (where 8 and 16 seem further apart). Indeed, a switch from logarithmic to linear scaling of numerical magnitudes predicts changes in accuracy well beyond number comparison (for review, see Opfer, Siegler & Young 2011). For tasks requiring children to locate whether 8 was closer to 6 or 11 (as in O'Hearn & Landau, 2007), for example, logarithmic numerical-magnitude representations would also yield lower accuracy – and thus potentially explain why WS children's accuracy is lower than that of TD children of the same age.

Alternative explanations for the difficulties of WS individuals in numerical cognition are certainly viable. Rather than their numerical-magnitude representations failing to develop much beyond that of TD 6-year-olds, representations of WS individuals may fail to develop much beyond that of TD 2- and 3-year-olds. For example, TD 2- and 3-year-olds who count flawlessly from 1 to 10 have no idea that 6 > 4 and 8 > 6, nor know how many pennies to give an adult who asks for 4 or more, nor estimate the positions of numbers on number lines even logarithmically (Le Corre, Van de Walle, Brannon & Carey, 2006; Le Corre & Carey, 2007; Sarnecka & Carey, 2008; Young, Marciani & Opfer, 2011). A general reason to think this alternative is unlikely, however, comes from previous findings that individuals with WS undergo slow development in many areas (prominently in space), followed by arrest at the functional level of 4- to 6-year-olds, some time in adolescence (Landau, 2011; Landau & Hoffman, 2007).

The present studies

To test our theory that the logarithmic mental number line of TD children persists for many more years among individuals with WS than is typical, we asked WS children and adults to estimate the magnitudes of numbers on number lines, and we compared age-related changes among WS individuals with those previously found among TD children and adults (Experiment 1). Following previous reviews (e.g. Siegler, Thompson & Opfer, 2009; Landau, 2011), our expectation was that both WS children and adults would estimate numerical magnitudes on a 0–1000 number line to increase logarithmically with actual value, much more like TD 6-year-olds than TD 3-year-olds or TD adults.

In Experiment 2, we examined the process of developmental change in WS more closely by using the microgenetic method (Karmiloff-Smith, 1993; Siegler & Crowley, 1991), which allowed a trial-by-trial assessment of how WS participants responded to relevant experiences about the magnitudes of numbers and how their responses compared to the responses of TD individuals. In TD children, the combination of cross-sectional and microgenetic data often yields complementary information that provides an unusually clear description of the change process (Opfer & Siegler, 2004, 2007; Siegler & Svetina, 2002; Siegler, Thompson & Opfer, 2009). In WS individuals, however, use of microgenetic methods is much more rare, and cross-sectional and microgenetic data have never been obtained from the same WS individuals in previous studies. This situation is unfortunate because fine-grained observations of the change process could reveal atypical responses to experiences, which could coexist (and contribute to) the slow developmental patterns previously observed in individuals with WS.

Experiment 1: Age-related changes in WS and TD estimates of numerical magnitude

Experiment 1 had two major purposes. Our first goal was to examine whether children with WS – like TD children – estimate numeric magnitudes to increase logarithmically with actual value. This goal was important theoretically because it allowed us to distinguish between early- and late-onset differences in development, and it was important practically because it allowed us to identify children who could potentially benefit from feedback in Experiment 2. The second goal was to test whether adults with WS – like TD older children and adults – estimate numeric magnitudes to increase linearly with actual value. This goal was important because it provided the most direct test of the hypothesis that WS individuals fail to develop linear numerical-magnitude representations.

Method

Participants

WS participants were 15 children (mean age = 11.8 years, range: 6–17 years) and 15 adults (mean age = 33.7 years, range: 18–56 years) with phenotypic and genetic confirmation of WS. The mean Verbal IQ was 75.31 (range 56–101), the mean Nonverbal IQ was 66.25 (44–87), and the Composite IQ was 67.28 (45–93), based on the Kaufman Brief Intelligence Test, 2nd edition (Kaufman & Kaufman, 2004). WS participants were recruited from a Williams Syndrome National Convention and from a regional Williams Syndrome Support Group. TD participants were 160 younger (mean age = 7.9 years, range: 6.4–8.9 years) and 81 older children (mean age = 10.0 years, range: 9.0–11.2 years) who participated in our three benchmark studies (Opfer & Siegler, 2007; Opfer & Thompson, 2008; Thompson & Opfer, 2008), as well as 99 TD adults (mean age = 22.3) who were enrolled in an introductory psychology class at a large university. Consent was obtained from participants or their guardians, following institutional IRB approval.

Tasks

Each participant was administered 22 number line problems. Each problem consisted of a 20-cm line, with the left end labeled 0 and the right end labeled 1000. Appearing 2 cm above the center of the line was the number to be estimated – 2, 5, 18, 34, 56, 78, 100, 122, 147, 150, 163, 179, 246, 366, 486, 606, 722, 725, 738, 754, 818, and 938. These numbers were chosen to maximize discriminability of logarithmic and linear functions by oversampling the low end of the range, to minimize the influence of specific knowledge (such as that 500 is halfway between 0 and 1000,) and – most importantly – to allow direct comparisons between estimates of individuals with Williams syndrome in this study and estimates of TD individuals in the benchmark studies (Opfer & Siegler, 2007; Opfer & Thompson, 2008; Thompson & Opfer, 2008).

Procedure

As in our benchmark studies, participants were tested in a single session. The items within each scale were randomly ordered, separately for each child, and presented in small workbooks, one problem per page. The experimenter began by saying, “Today were going to play a game with number lines. What I’m going to ask you to do is to show me where on the number line some numbers are. When you decide where the number goes, I want you to make a line through the number line like this (making a vertical hatch mark)”. Before each item, the experimenter said, This number line goes from 0 at this end to 1000 at this end. If this is 0 and this is 1000, where would you put N?” The numbers were read aloud by the experimenter before the participants made their estimate to ensure that participants knew the number they were estimating. This procedure was the same one used in the benchmark studies with typically developing children.

Results and discussion

We first examined age differences in accuracy of numerical estimates. To measure accuracy, we converted the magnitude estimate for each number (the child’s hatch mark) to a numeric value (the linear distance from the 0 mark to the child’s hatch mark), then divided the result by the total length of the line, and multiplied the result by 1000. The magnitude of each child’s error was calculated by taking the mean absolute difference between each of the child’s estimated values and the actual values, and accuracy was calculated by subtracting the magnitude of the error from 1.

As expected, accuracy of TD estimates increased with age, $r(340) = .67$, $p < .0001$, from 78% for younger TD children to 87% for TD adults. Accuracy of WS estimates also increased with age, $r(30) = .46$, $p < .001$, increasing from 67% for WS children to 72% for WS adults. Thus, accuracy of both WS and TD individuals’ numerical estimates improved with age.

To determine whether age-related improvements in accuracy were associated with the hypothesized logarithmic-to-linear shift, we next compared the fit of the logarithmic and linear regression functions to participants’ median estimate for each number. Among TD children, younger children’s median estimates were better fit by the logarithmic function ($R^2 = .96$) than by the fit of the linear function ($R^2 = .75$), whereas older children’s median estimates were better fit by the linear function ($R^2 = .99$) than by the logarithmic ($R^2 = .73$). Similarly, WS children’s median estimates were also better fit by the logarithmic regression function ($R^2 = .78$) than by the linear function ($R^2 = .38$; Figure 1A). Unlike TD older children and adults’, however, WS adults median estimates were better fit by the logarithmic regression ($R^2 = .80$) than by the linear ($R^2 = .47$; Figure 1B). Thus, although both TD and WS estimates were initially fit by the logarithmic function, there was no evidence of

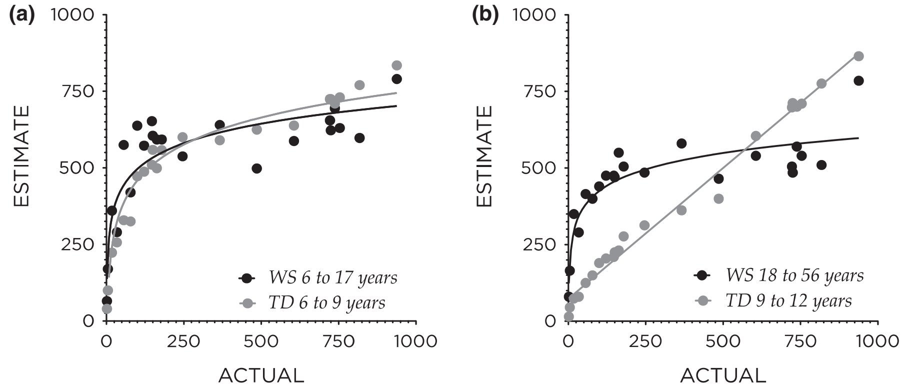

Figure 1 *Experiment 1: Age-group differences in numerical estimates. Among younger children (panel A), median estimates of both WS and TD children increased logarithmically with value. Among older children and adults (panel B), median estimates of TD children increased linearly with value, whereas estimates of WS adults increased logarithmically. Estimates of WS adults were lower than WS children.*

the logarithmic-to-linear shift in participants with WS, suggesting that development was arrested roughly at the level of typically developing 6-year-olds.

To ensure that analyses of group medians reflected individual performance, we also regressed individuals' estimates on each task against the predictions of the best fitting linear and logarithmic functions. We assigned a 1 to the model that best fit each participant's estimates and a 0 to the other model. The function that provided the better fit to individual participant's estimates varied in ways that mirrored the findings with the group medians, $X^2(3) = 102.5$, $p < .0001$. That is, the logarithmic function provided the best fit for the large majority of WS individuals (80% of WS children, 93% of WS adults) and 80% of TD younger children, whereas the logarithmic function provided the better fit for only 16% of TD older children. Thus, the individual patterns of estimates – like the group data – supported the hypothesis that WS individuals fail to develop linear numerical-magnitude representations.

If *linearity* of numerical estimates among WS individuals failed to increase over a period of 50 years (i.e. from ages 6 to 56), then why might estimation *accuracy* nevertheless improve with age? Comparison of Figures 1A and 1B suggests a clue: although both WS children and WS adults increase their estimates of numerical magnitude logarithmically with number, estimates of WS children appear systematically higher than those of WS adults and further from the ideal linear function, $y = x$. Indeed, a paired t-test of estimates confirms this inspection, $t(22) = 5.74$, $p < .0001$, with 20/22 of WS children's median estimates being greater than those of WS adults.

These results suggest that WS individuals might improve their estimates with age and experience – not by adopting linear representations of number, as TD children did in our benchmark studies – but by reacting to error signals (i.e. feedback regarding the direction and/or magnitude of the estimation error) with piecemeal (downward) adjustments in estimation behavior. Put another way, by relying on a logarithmic mental number line, both WS and TD children's errors may be biased toward overestimation (e.g. placing 150 where 720 would go, not where 150 would go), and in everyday life, adult reactions to these overestimates could provide an error signal (e.g. "that's too many!" when children return with all the possible spoons when asked for just 15 spoons). To improve future accuracy, children could revise their estimates to increase linearly with actual magnitude (as older TD children appeared to have done) or lower their estimates overall (as WS adults appeared to have done). If true, this observation is important theoretically because age-related lowering of estimates – without increasing linearity – is certainly an atypical developmental path; it is, however, also an effective strategy for improving accuracy of the early logarithmic estimates depicted in Figure 1A.

In summary, Experiment 1 yielded three main findings. First, as in TD children, estimation accuracy of children with WS improved with age, with the initial inaccuracy of estimates by WS children and TD children tied to their use of logarithmic representations of numerical value. Second, unlike older TD children and adults, WS adults continued to rely on logarithmic representations, even after many years of age and experience. Finally, we were able to identify a large number of WS individuals whose estimates were best fit by a logarithmic function, which allowed us to directly examine the impact of feedback on their estimates in Experiment 2 and thus test our hypothesis about how WS and TD children respond differently to the same experiences.

Experiment 2: Short-term changes in WS and TD estimates of numerical magnitude

Experiment 2 was designed to compare short-term developmental changes in WS and TD individuals in terms of four key dimensions of cognitive change: the source, rate, path, and variability of change. These dimensions have been proposed as central aspects of change and have proved useful in describing cognitive change in a wide variety of contexts (for reviews, see Siegler, 2006). To test the idea that feedback is an important *source of change*, we compared the amount of improvement in estimation between the feedback and no-feedback groups. To examine the *rate of change*, we measured how many feedback problems children required in each condition before they adopted a linear numerical-magnitude representation. To learn about the *path of change*, we tested whether children showed an abrupt shift from a logarithmic pattern to a linear pattern of estimates or whether they progressed from a clear logarithmic pattern to a pattern intermediate between the two functions to a clear linear pattern. Finally, to enhance understanding of the *variability of change*, we examined the relation between children's IQ and estimation performance.

Method

Participants

Individuals with WS were the same participants who participated in Experiment 1. Typically developing controls included 96 children (mean age = 7.9 years, range: 6.4–8.9 years) who participated in identical conditions from three previous microgenetic studies of number line estimation – Opfer and Siegler, 2007 (150-feedback condition, $n = 13$; no-feedback condition, $n = 15$); Opfer and Thompson, 2008 (treatment conditions, $n = 15$; control conditions, $n = 22$); and Thompson and Opfer, 2008 (treatment condition, $n = 16$; control condition, $n = 15$). More information on the controls can be found in the published benchmark studies.

Tasks

The number line task described in Experiment 1 was also used in Experiment 2.

Procedure

Immediately after Experiment 1, WS participants who did not generate linear estimates were randomly assigned to one of two groups: one group received feedback during the training phase (WS feedback group, $n = 16$ (9 children, 7 adults); average age = 23.1 years; range = 7–56 years), whereas the other group did not receive feedback (WS no-feedback group, $n = 11$ (5 children, 7 adults); average age = 22.4 years; range = 9–48 years). (Slightly different sample sizes across conditions occurred in Experiment 2 because WS participants were randomly assigned to condition before linearity of estimates could be assessed.) In our benchmark studies of typically developing children, 44 children (average age = 7.92) received feedback and 52 children (average age = 7.94) did not.

As shown in the outline of the procedure in Table 1, participants in both groups completed the number line estimation task for a pretest, three training trial blocks and a posttest. The purpose of these three phases (pretest, training trial blocks, and posttest) was to examine the course of learning prior to posttest (i.e. to examine changes from the number line pretest through posttest). On the number line pretest and posttest, children in the feedback and no-feedback groups were presented with the same 22 problems without feedback. For children in the feedback group, each training trial block included a feedback phase and a test phase. As shown in Table 1, the feedback phase of each training trial block included either one item on which children received feedback (Trial Block 1) or three items on which they received feedback (Trial Blocks 2 and 3). The test phase in all three training trial blocks included 10 items on which children did not receive feedback; this test phase occurred immediately after the feedback phase in each training trial block. Children in the no-feedback group received the same number of estimation problems, but they did not receive feedback. On posttest, children in all groups were presented with the same 22 problems without feedback as in Experiment 1. The children's estimates in Experiment 1 provided pretest data, which were used as a point of comparison for their subsequent performance and were elicited during the same session.

Feedback was administered to the individuals with WS following the same procedure used in our three benchmark studies (Opfer & Siegler, 2007; Opfer & Thompson, 2008; Thompson & Opfer, 2008). On the first feedback

Table 1 *Design of Experiment 2*

	Phase						
	Trial Block 1		Trial Block 2		Trial Block 3		
Group	Feedback[1] (1 item)	Test (10 items)	Feedback[1] (3 items)	Test (10 items)	Feedback[1] (3 items)	Test (10 items)	Posttest (22 items)
150-feedback	150	0–1000	147–187	0–1000	147–187	0–1000	0–1000
No-feedback	150, 5, or 725	0–1000	147–187, 2–42, or 723–763		147–187, 2–42, or 723–763	0–1000	0–1000

[1]During the Feedback phase, the WS participants in the no-feedback group were asked to estimate the positions of the same numbers as the WS participants in the 150-feedback group.

problem, participants were told, "After you mark where you think the number goes, Ill show you where it really goes, so you can see how close you were". After the participant answered, the experimenter wrote the number corresponding to the child's estimate (*E*) above the mark, and indicated the correct location of the number that had been presented (*N*) with a hatch mark, using a small plastic ruler that had the correct positions of numbers indicated. This allowed us to quickly indicate the number corresponding to the S's hatch mark and the correct placement of each number on the number line. The procedure took a very short amount of time (15–20 seconds at most), and we have used it successfully with TD children in several previous experiments. For example, if a child were asked to mark the location for 150 (i.e. *N*) and his estimate corresponded to the actual location of 600 (i.e. *E*), the experimenter would write the number 600 above the child's mark and mark where 150 would go on the number line. After this, the experimenter showed the corrected number line to the child. Pointing to the child's mark, she said, "You told me that *N* would go here. Actually, this is where *N* goes [pointing]. The line that you marked is where *E* actually goes". When children's answers deviated from the correct answer by no more than 10%, the experimenter said, "You can see these two lines are really quite close". When children's answers deviated from the correct answer by more than 10%, the experimenter told the child, "That's quite a bit too high/too low. You can see these two lines [the child's and experimenters hatch marks] are really quite far from each other".

Results and discussion

We organized our results into four sections: first, we report on the conditions that led to changes in numerical estimation (source of change); next, how quickly those changes occurred (rate of change); then, approaches that children used up to and following the use of adult approaches (path of change); finally, individual differences in these variables (variability of change).

Source of change

We first examined the source of change in WS estimation performance on the number line task. Specifically, we wanted to test whether the experiences that WS participants received during training improved estimation accuracy and influenced the degree to which those estimates came to follow a linear function. To find out, we performed a 2 (treatment: feedback, no-feedback) × 2 (test-phase: pretest, posttest) repeated-measures ANOVA on accuracy scores (0 to 1). As expected, we found a treatment by test-phase interaction, $F(1, 26) = 5.02$, $p < .05$. Post-hoc analyses indicated that accuracy of WS estimates improved reliably from pretest (M = 69%, $SD = .09$) to posttest (M = 72%, $SD = .08$) for the feedback group, $t(15) = 2.25$, $p < .05$, Cohen's $d = 0.37$, whereas accuracy did not increase from pretest (M = 68%, $SD = .07$) to posttest (M = 67%, $SD = .07$) for the no-feedback group, $t(10) = 1.0$, *ns*. Thus, a short session of feedback on WS estimation performance engendered similar changes in accuracy as many years of ordinary experience.

We next examined whether short-term changes in WS estimation accuracy were also accompanied by the logarithmic-to-linear shift previously observed in TD children (Figure 2). As in TD children (Figures 2C and 2F), we found that estimates of WS individuals in both the feedback group and no-feedback groups initially provided median estimates for each number that were fit better by the logarithmic regression function than by the linear one (see Figure 2, Panels A, B, D, and E). For these pretest estimates, the precision of the fit of the logarithmic function, and the degree of superiority of that function to the linear function, was nearly identical among the WS feedback (log $R^2 = .78$; lin $R^2 = .38$), and the WS no-feedback (log $R^2 = .79$, lin $R^2 = .36$) groups. Although the precision of fit was somewhat higher overall for TD children, the superiority of the fit of the logarithmic to linear functions to TD children's estimates (feedback: log $R^2 = .96$, lin $R^2 = .76$; no feedback: log $R^2 = .95$, lin $R^2 = .70$) was much like that in the participants with WS.

After receiving feedback, however, estimates of the WS groups differed from those of TD groups who had received the same feedback. Unlike TD controls (Figure 2C), WS children in the feedback (Figure 2A) and no-feedback (Figure 2D) groups continued to generate estimates that fit the logarithmic function better than the linear one (WS feedback: log $R^2 = .83$, lin $R^2 = .37$; WS no-feedback: log $R^2 = .67$, lin $R^2 = .20$). That is, feedback improved WS accuracy but did not engender a logarithmic-to-linear shift. If anything, feedback increased both the absolute degree to which estimates of WS participants were fit by the logarithmic function as well as the relative fit of the logarithmic function to the linear one. That this change from pretest to posttest was a response to feedback is indicated by the fact that a similar pretest-to-posttest change was not evident in the no-feedback groups (Figures 2D and 2E). Thus, although feedback led to a logarithmic-to-linear shift in TD children (Figure 2C), feedback did not lead to a similar shift in WS children (Figure 2A) or adults (Figure 2B). In fact, the observed responses to feedback were similar to the age-related changes observed in Experiment 1, suggesting atypical development of changes in both age and experience in individuals with WS.

If feedback did not improve accuracy by engendering a logarithmic-to-linear shift in WS participants, how did WS participants in the feedback group improve their estimates from pretest to posttest? Again, the explanation for short-term changes in WS estimation accuracy might be similar to the long-term changes in WS estimation accuracy. That is, WS participants in the

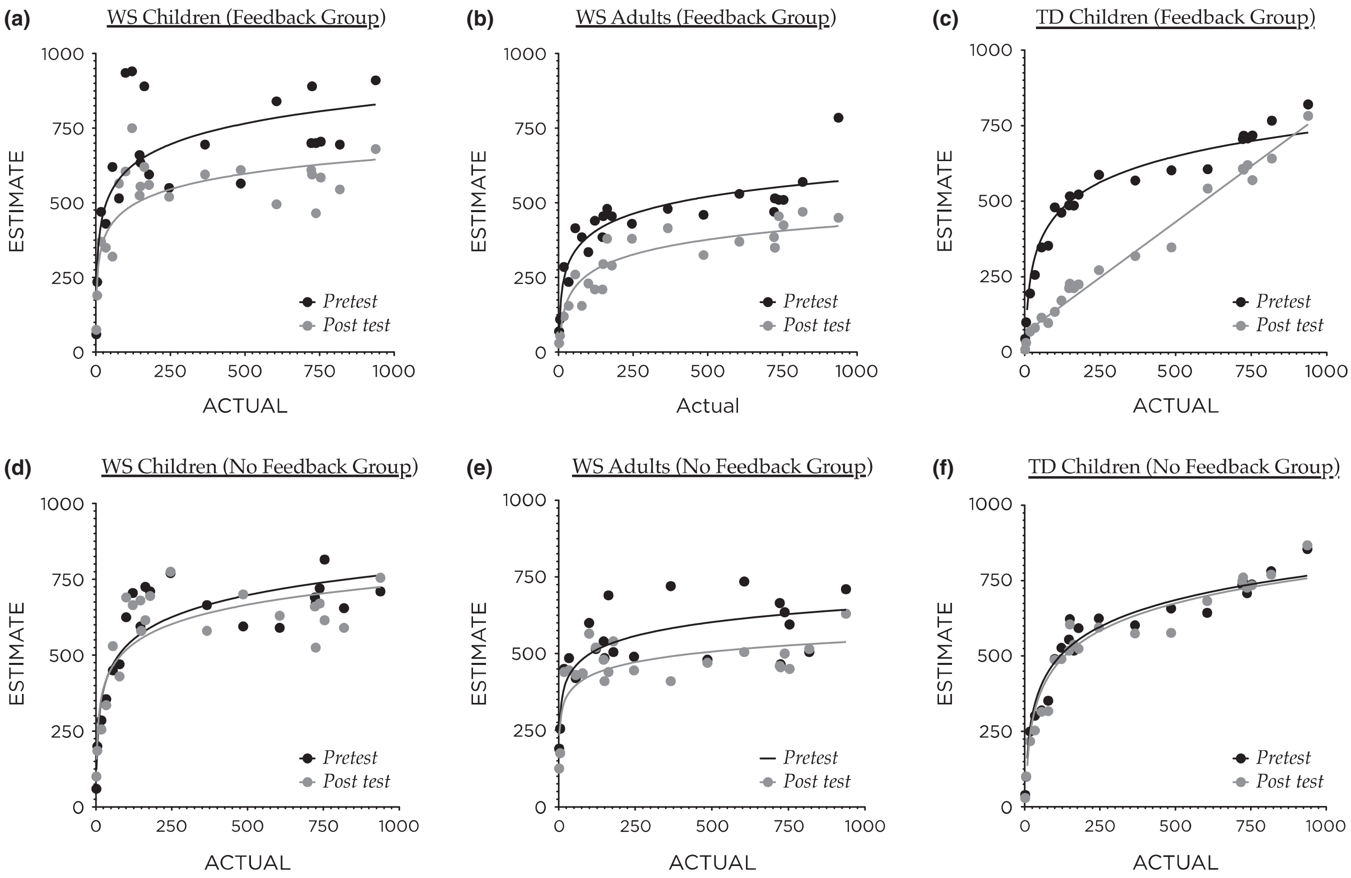

Figure 2 *Experiment 2: Pre- to posttest changes in numerical estimates. From pretest (black series) to posttest (grey series), only estimates of TD children who received feedback (panel E) were characterized by a logarithmic to linear shift.*

feedback group (like older WS individuals) could attain greater accuracy than the control group simply by lowering their estimates of numerical value across the number range. Consistent with this analysis, WS children's estimates in the feedback group were consistently lower on posttest than on pretest, $t(21) = 5.40$, $p < .0001$. In contrast, WS children's estimates in the no-feedback group were neither lower nor higher from pretest to posttest, $t(21) = 1.07$, *ns*. Thus, feedback engendered an atypical change in WS children's estimates.

Rate of change

To address the rate of change in numerical estimation, we used logistic regression to examine the relation between generation of more linear than logarithmic patterns of estimates (linear model fitting best or not) and number of trial blocks of feedback (0–4), where 0 corresponded to the trial block prior to the administration of the treatment and thus 0 trials of feedback. First, we examined the effect of trial block for the feedback group of TD children. For TD children, there was a significant positive effect of trial block for the feedback group, indicating that with each additional trial block the likelihood of generating linear estimates was 2.71 times more likely than the previous one, $z = .50$, $z = 5.03$, Wald (2, $N = 220$) = 36.9, $p < .0001$. A similar analysis found no significant effect of trial block for WS children (whether receiving feedback or not), indicating that trial blocks of feedback did not increase the odds of WS individuals generating linear estimates. As illustrated in Figure 3, the low rate of change in WS individuals who generated logarithmic estimates on pretest differed greatly from the rate of change in similarly performing TD children, whereas it did not differ from the rate of change in WS individuals who also initially generated logarithmic estimates but did not receive feedback.

Path of change

Inspection of Figure 3A suggests that one reason trial-to-trial changes in WS individuals were so modest compared to TD individuals is that TD children were better at retaining the gains made after a single trial of feedback. Consistent with this idea, the proportion of individuals whose estimates were best fit by the linear function was never higher than after the first trial of feedback, and the decline in that proportion from that first trial of feedback to the last trial block was nominally much greater in the WS feedback

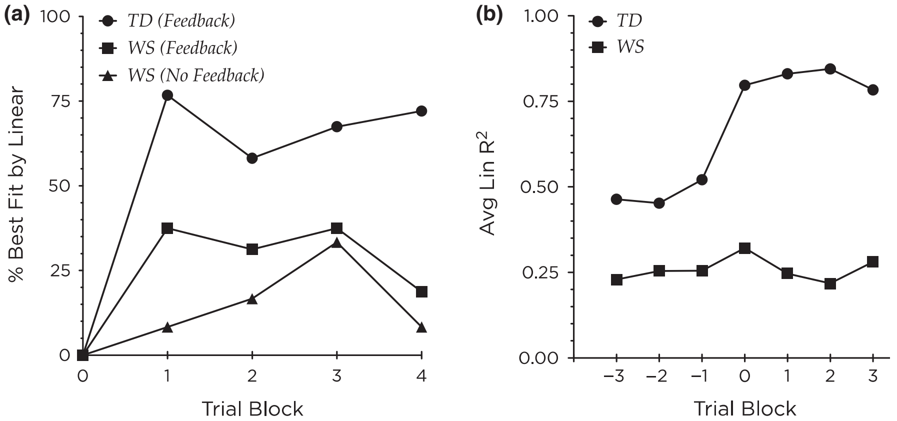

Figure 3 *Experiment 2: Rate and path of change. In Panel A, the proportion of children generating linear patterns of estimates rapidly increased among TD children, but not WS children and adults. In Panel B, the linearity of TD children's estimates increased abruptly and remained stable, whereas linearity of WS children's estimates increased little and was not stable.*

group (38% to 19%) than in the TD feedback group (77% to 72%). This observation is potentially important. If WS individuals are quite poor at retaining new information about numerical magnitudes, it could explain why so little change occurs over many years of development.

To test this idea, we examined the fit of the linear regression function to each individual child's numerical estimates as a function of the number of trials that elapsed since the linear function provided a better fit than the logarithmic (i.e. when the logarithmic to linear shift was thought to occur). To measure this, we identified the first trial block on which the linear function provided the best fit to a given individual's estimates, and we labeled it trial block 0. The trial block immediately before each child's "trial block 0" was that child's trial block −1, the trial block before that was the child's trial block -2, and so on.

These assessments of the trial block on which children's estimates first fit the linear function made possible a backward-trials analysis that allowed us to test alternative hypotheses about the path of change from a logarithmic to a linear representation (Figure 3B). One hypothesis, suggested by incremental theories of representational change (Barth & Paladino, 2010), was that the path of change entailed gradual, continuous improvements in the linearity of estimates (and thus the fit of the linear regression function to their estimates). According to this hypothesis, the fit of the linear model would have gradually increased, from Trial Block −3 to Trial Block +3. In this scenario, Trial Block 0 – the first trial block in which the linear model provided the better fit – would simply mark an arbitrary point along a continuum of gradual, trial block-to-trial block improvement, rather than the point at which children first chose a different representation.

A second hypothesis was that the path of change involved a discontinuous switch from a logarithmic to a linear representation, with no intermediate state. This would have entailed no change in the fit of the linear model from Trial Block −3 to −1, a large change from Trial Block −1 to Trial Block 0, and no further change after Trial Block 0. This second hypothesis clearly fit the data from TD children. From Trial Block −3 to −1, results of a Kruskal-Wallis test indicated no change in the fit of the linear function across these trial blocks (p's > .1). There also was no change from Trial Block 0 to Trial Block 3 (p's > .1). However, from Trial Block −1 to Trial Block 0, there was a large increase in the fit of the linear function to individual TD children's estimates, from a median R^2 = .52 to a median R^2 = .80 (difference in rank sum = 48.22, $p < .01$). Thus, rather than Trial Block 0 reflecting an arbitrary point along a continuous path of improvement, it seemed to mark the point at which TD children switched from a logarithmic representation to a linear one.

We next tested whether the path of change for WS individuals also involved a discontinuous switch from a logarithmic to linear representation. Against this hypothesis, results of a Kruskal-Wallis test indicated no change in the fit of the linear function across any of the possible pairs of trial blocks (ps > .5). Thus, even when 100% of WS individuals were best fit by the linear function (Trial Block 0), the fit of the linear function was quite small (WS, mean lin R^2 = .32; TD, mean lin R^2 = .80; Mann Whitney U = 127, $p < .0001$) and did not persist across trial blocks. This apparent lack of a discontinuous increase in linearity is another marked difference between TD and WS individuals.

Variability of change

In the previous sections, we observed that changes in numerical estimation – from pretest to posttest and from trial to trial – differed between the average WS individual and average TD child. In this section, we examined whether IQ of WS individuals might account for none, some, or all of these differences in the change trajectory.

To examine whether IQ moderated pretest-to-posttest changes in number line estimation, we used WS participants' composite scores on the KBIT-2. These scores provided us with two groups to compare: 11 participants with borderline to average scores (i.e. scoring 70–99, identified as the 'higher IQ group') and 16 participants with mild intellectual disability to moderate intellectual disability (i.e. scoring 40–69, identified as the 'lower IQ group').

As is evident in Figure 4, estimates of both groups were better fit by the logarithmic than by the linear function on both pretest (higher IQ group, log R^2 = .84, lin R^2 = .36; lower IQ group, log R^2 = .81, lin R^2 = .39) and posttest (higher IQ, log R^2 = .73, lin R^2 = .26; lower IQ, log R^2 = .81, lin R^2 = .33). In addition, the proportion of individual participant's estimates best fit by the linear function on posttest did not differ across the two IQ groups (higher IQ group, 18%; lower IQ group, 13%), nor did composite (or raw) IQ scores correlate with linearity of estimates on posttest (r = .07, *ns*). Thus, among WS children, higher intelligence scores were not associated with the logarithmic to linear shift seen in TD participants.

Rather than higher intelligence scores being associated with a logarithmic to linear shift, inspection of Figure 4 suggests that higher IQ was associated simply with greater lowering of estimates. Consistent with this conjecture, paired t-tests confirmed that estimates were consistently lower on posttest than pretest for both the higher IQ group ($t[21]$ = 10.89, $p < .0001$) and the lower IQ group ($t[21]$ = 4.72, $p < .0001$), but the magnitude of change was greater for the higher IQ group ($t[42]$ = 5.60, $p < .0001$). This difference between the higher and lower IQ groups is interesting because it parallels the difference between WS adults and children in Experiment 1 and the difference between the feedback and no-feedback groups in Experiment 2. That is, with greater IQ, greater age, and greater experience, WS participants estimated the magnitudes of numbers on number lines simply as being lower. As we have seen, this lowering of estimates did improve estimation accuracy, but it did so without the logarithmic-to-linear shift that characterized TD children.

General discussion

Previous work has indicated that development of linear representations of numerical magnitudes profoundly expands typically developing children's quantitative thinking (Opfer, Siegler & Young 2011). It improves their ability to estimate the positions of numbers on number lines (Opfer & Siegler, 2007; Siegler & Opfer, 2003; Siegler & Booth, 2004), to estimate the measurements of continuous quantities (Booth & Siegler, 2006; Thompson & Siegler, 2010) and the quantity of discrete objects (Opfer, Thompson & Furlong, 2010), to categorize numbers according to size (Laski & Siegler, 2007; Opfer & Thompson, 2008), to estimate and learn the answers to arithmetic problems (Booth & Siegler, 2008), and to remember numbers encountered through stories and first-hand experiences (Thompson & Siegler, 2010; Thompson & Opfer, 2011; Young *et al*., 2011).

In this paper, we examined long- and short-term changes in the numerical-magnitude estimates of children and adults with Williams syndrome. Given persistent mathematical deficits in WS individuals (Ansari *et al*., 2007; Howlin *et al*., 1998; Paterson *et al*., 2006) and structural abnormalities in WS cortical regions associated with numerical-magnitude representations (Boddaert, Mochel, Meresse, Seidenwurm, Cachia, Brunelle, Lyonnet & Zilbovicius, 2006; Eckert, Hu, Eliez, Bellugi, Galaburda, Korenberg, Mills & Reiss, 2005; Kippenhan, Olsen, Mervis, Morris, Kohn, Meyer-Lindenberg & Berman, 2005; Meyer-Lindenberg, Kohn, Mervis, Kippenhan, Olsen, Morris & Berman, 2004), we had expected differences in numerical estimation to exist between TD and WS individuals, yet it was not clear what those differences might be and how they might manifest over development. To examine this issue, we investigated long-term changes in numerical estimation that occurred over many years (Experiment 1) and short-term changes that occurred in response to instructive experiences (Experiment 2).

Across both time scales, we found several similarities in the development of numerical estimation among

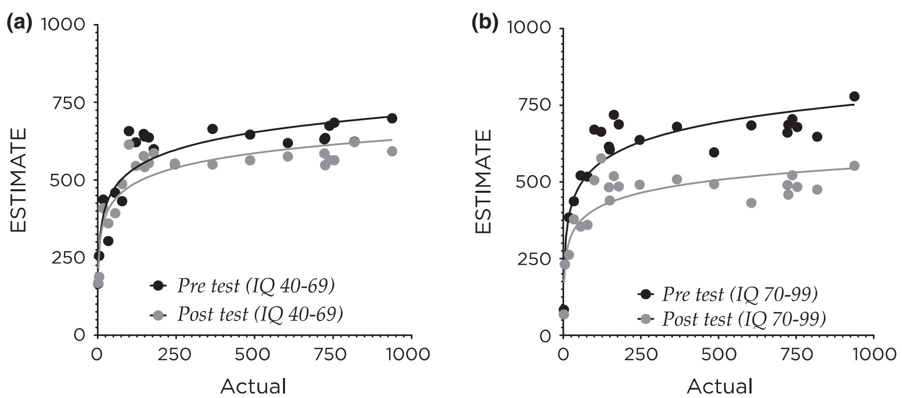

Figure 4 *Median estimates of both lower IQ (panel A) and higher IQ (panel B) WS children increase logarithmically with actual value on pretest and posttest. In higher IQ WS individuals, posttest estimates were lower than pretest.*

individuals with WS and TD individuals. In both WS and TD groups, we observed that accuracy of numerical estimates improved with age (Experiment 1) and with instructive experiences (Experiment 2). Further, we observed that estimates of younger TD and WS children were actually quite similar (Figure 1). In both groups, estimates of numerical magnitude increased logarithmically with the actual value of numbers. Further, 80% of children in each group generated estimates that were better fit by the logarithmic than linear function. Thus, the chromosomal deletions typifying WS did not appear to prevent WS children in this study from learning to improve their estimates of numerical magnitude nor to cause WS children to deviate from the normal pattern of estimating numerical magnitudes to increase logarithmically with actual value. These results suggest that, at least in the 0–1000 number line context, numerical-magnitude representations of WS individuals are more like those of TD 6- to 9-year-olds than TD 3- to 6-year-olds or TD 9- to 12-year-olds.

Several differences between WS and TD individuals were also observed to exist in the development of numerical estimation. First, although estimates of WS and TD individuals were initially fit by the logarithmic function, we found no evidence among WS individuals of the typical logarithmic-to-linear shift over a period of 50 years (i.e. from ages 6 to 56). That is, even adults with WS continued to estimate numbers to increase logarithmically with actual value (Experiment 1), and this estimation pattern was observed in WS individuals of all ages (and IQs) even after considerable training (Experiment 2). These results are consistent with the arrested development that has been observed in many aspects of spatial representation, as noted by Landau and colleagues (Landau, 2011; Landau & Hoffman, 2007).

A second difference noted between WS and TD individuals was in their respective responses to the experience of feedback, where the responses of WS individuals followed an atypical developmental path. That is, after observing that they had estimated the magnitude of (say) 150 as being too high, WS individuals responded by lowering all their subsequent estimates *while continuing to scale the numbers logarithmically.* In contrast, TD individuals responded by lowering their subsequent estimates, too, but *spontaneously scaled the numbers linearly*, typically after a single trial of feedback. Moreover, this difference in responding to feedback could not be attributed to WS individuals' low IQ; as IQ increased in the WS group, the probability of this atypical response to feedback only increased. (Although we do not have the data to address the issue, it seems unlikely that TD children with high IQs would respond similarly.) These results suggest that the chromosomal deletions typifying WS may prevent the development of linear numerical-magnitude representations that typically occurs as school-children gain experience with the decimal system. Moreover, this atypical response to experience is apparently not unique to number line estimation. In their study of spatial reasoning, for example, Hoffman *et al.* (2003) also found individuals with WS adopting atypical strategies once they had observed their errors in a block construction task. Thus, it seems possible that atypical responses to experience more generally could play a large role in preventing individuals with WS from progressing down the typical developmental path.

How might the deficits of WS prevent the development of linear numerical-magnitude representations? Admittedly, any answer is speculative. One possibility is that WS is associated with a general (non-numeric) impairment in magnitude representations. Providing evidence against this idea, however, Farran (2006) examined discrimination of line lengths in WS adolescents and adults and found evidence of a distance effect among WS adolescents and adults that was quite similar to what Moyer (1973) found in TD adolescents and adults. Thus, it seems unlikely that visual magnitude representations are as impaired in WS as are numeric magnitude representations (e.g. O'Hearn & Landau, 2007). One way to explain this uneven developmental progression among WS individuals is that *symbolic* magnitude representations are impaired in WS. To examine non-numeric symbolic magnitude representations, Moyer (1973) showed pairs of animal names (e.g. 'COW-ANT') to TD participants and asked them to name the larger animal. Here, the psychophysical function was very close to what Moyer had observed for line length, suggesting a common magnitude representation for symbolic and non-symbolic items. An interesting question for future studies is whether a general impairment in *symbolic* magnitude representation is present in WS children and adults.

Another (somewhat complementary) possibility is that delays in numerical-magnitude representations in WS persist because structural abnormalities in parietal cortex inhibit development of linear numerical-magnitude representations that characterize TD adults. Among TD adults, for example, detecting the correct placement of numbers on number lines is strongly associated with posterior parietal activity, at least compared with detection of the correct placement of non-numerical stimuli (Kanayet, Opfer & Cunningham, 2010), and this same cortical area is broadly reported to represent numeric magnitudes (Cantlon, Brannon, Carter & Pelphrey, 2006; HoudÕ, Rossi, Lubin & Joliot, 2010; Piazza *et al.*, 2004). Further, among WS adults, structural and functional abnormalities are evident within parietal cortex, especially within the IPS region (Boddaert *et al.*, 2006; Eckert *et al.*, 2005; Kippenhan *et al.*, 2005; Meyer-Lindenberg *et al.*, 2004; Reiss, Eckert, Rose, Karchemskiy, Kesler, Chang, Reynolds, Kwon & Galaburda, 2004; Van Essen, Dierker, Snyder, Raichle, Reiss & Korenberg, 2006). Thus, although overall brain volume is reduced in WS (Martens, Wilson, Dudgeon & Reutens, 2009; Reiss, Eliez, Schmitt, Straus, Lai, Jones & Bellugi, 2000; Reiss *et al.*, 2004), compared to CA-matched controls, parietal regions in WS show a significant decrease in gray matter (Boddaert *et al.*, 2006; Eckert *et al.*, 2005; Meyer-Lindenberg *et al.*, 2004), with reduced sulcal depth noted in the IPS (Kippenhan *et al.*, 2005). Thus, previous studies with adults are consistent with the idea that an abnormally functioning posterior parietal cortex in WS individuals may cause them to fail to recognize the linear placement of numbers on number lines (such as those presented in Experiment 2) and to learn to use this linear placement in their own estimates.

Still another potential reason for the lack of learning linear numerical-magnitude representations in WS individuals is a deficiency in cognitive resources that have nothing to do with quantity at all. For example, analogical reasoning has been implicated in the ability of TD children to learn linear numerical-magnitude representations for large numbers (Opfer & Siegler, 2007; Thompson & Opfer, 2010), and verbal analogies are known to be difficult for children with WS to perform accurately (Porter & Coltheart, 2005). Indeed, in other domains where analogical reasoning plays an important role (e.g. in acquisition of an adult *living things* concept; Opfer & Siegler, 2004), individuals with WS were also reported to fail to show the pattern of conceptual change observed in TD individuals (Johnson & Carey, 1998). Although we observed no correlation between one kind of analogical reasoning (matrix completion) and learning of linear numerical-magnitude representations, it seems clear that further research is needed to evaluate this possibility as well.

In conclusion, a combination of cross-sectional and microgenetic data yielded an unusually clear and consistent description of how developmental changes in numerical estimation do and do not differ between TD and WS individuals. These findings indicate that both TD and WS individuals improve the accuracy of their numerical estimates with age and experience, but that only TD individuals show a shift from logarithmic to linear representations of numerical magnitude. Whether learning without representational change is specific to the numeric domain, to domains involving symbolic magnitude, or to domains requiring analogical insights, however, remains an important issue for future research.

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