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SPECIAL SECTION: THE DEVELOPMENT OF MATHEMATICAL COGNITION

Small and large number processing in infants and toddlers with Williams syndrome

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Abstract

Previous studies have suggested that typically developing 6-month-old infants are able to discriminate between small and large numerosities. However, discrimination between small numerosities in young infants is only possible when variables continuous with number (e.g. area or circumference) are confounded. In contrast, large number discrimination is successful even when variables continuous with number are systematically controlled for. These findings suggest the existence of different systems underlying small and large number processing in infancy. How do these develop in atypical syndromes? Williams syndrome (WS) is a rare neurocognitive developmental disorder in which numerical cognition has been found to be impaired in older children and adults. Do impairments of number processing have their origins in infancy? Here this question is investigated by testing the small and large number discrimination abilities of infants and toddlers with WS. While infants with WS were able to discriminate between 2 and 3 elements when total area was confounded with numerosity, the same infants did not discriminate between 8 and 16 elements, when number was not confounded with continuous variables. These findings suggest that a system for tracking the features of small numbers of object (object-file representation) may be functional in WS, while large number discrimination is impaired from an early age onwards. Finally, we argue that individual differences in large number processing in infancy are more likely than small number processing to be predictive of later development of numerical cognition.

Introduction

It has been suggested that precursors to numerical abilities, such as simple arithmetic computations and discrimination between numerosities, exist from infancy onwards (Dehaene, 1997) and that studying number processing abilities in young infants is crucial in the search for core knowledge and evolutionary origins of numerical cognition (Hauser & Carey, 2003) as well as for uncovering the roots of atypical development of numerical competence (Ansari & Karmiloff-Smith, 2002).

A number of studies have indicated that typical developing (TD) newborns can discriminate between small numerosities (Antell & Keating, 1983; Starkey & Cooper, 1980) and that 5-month old infants are sensitive to the outcomes of simple addition and subtraction (Wynn, 1992). However, these early studies of infant number discrimination did not control for variables continuous with number, such as area occupied by numerical stimuli or their contour length. More recent studies have shown that when these continuous variables are systematically controlled for, young infants fail to discriminate between small numerosities and are not sensitive to arithmetic transformations (addition and subtraction) of visual stimuli (Clearfield & Mix, 1999; Feigenson, Carey & Hauser, 2002; Xu, 2003; Xu, Spelke & Goddard, 2005). These findings suggest that for small number discrimination, infants attend more to variables continuous with numerosity than to numerosity itself.

More recently, infants' ability to discriminate between larger numerosities such as 8 and 16 and 8 and 12 has been tested. Converging evidence from studies across different laboratories suggests that infants as young as 6 months can discriminate between large numerosities even when continuous variables such as area and contour length are systematically controlled for (Brannon, Abbott & Lutz, 2004; Xu & Spelke, 2000). This finding indicates that the large number discrimination abilities in infants may be sustained by the same system that is thought to enable number discrimination in animals (e.g. Brannon

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& Terrace, 1998) and human adults (Moyer & Landauer, 1967). It turns out that infants' large number discrimination depends crucially on the ratio between numerosities. More specifically, Xu and Spelke (2000) found that infants can discriminate between 8 and 16, but not between 8 and 12 dots. In a similar study, 6-month-olds were able to discriminate between 16 and 32 dots, but not between 16 and 24 (Xu *et al.*, 2005). These data carry the signature of Weber's Law, by revealing that the discriminability of numerosity in infancy is dependent on the ratio of the numerosities presented. Large number discrimination in young infants has also been demonstrated using sounds and jumps, pointing to the existence of a stimulus-independent, amodal mechanism for large number processing in infancy (Lipton & Spelke, 2003; Wood & Spelke, 2005).

The contrast between the failures in small number discrimination and the successes in large number discrimination provide support for invoking two systems which may form the basis of the human's basic 'number sense' (Dehaene, 1997; Feigenson, Dehaene & Spelke, 2004). A first system is thought to be an approximate number estimation system. A second system, the parallel individuation or object-file system, is thought to underlie the tracking and reasoning about small numbers of individual objects. According to this model, each individual object is represented by an opened 'file' in memory. The one-to-one correspondence function between the object files stored in memory and the objects being presented results in either a mismatch or match, which enables discrimination between small numerosities. This mismatch could be numerical (difference in the number of objects), but it could also be non-numerical (difference in the stimulus area). Against this background, it has been suggested that infants' performance in small (but not large) number discrimination studies is based on an object-based attention system rather than a system dedicated to the processing of numerosity (Feigenson *et al.*, 2004; Xu, 2003).

The establishment of two separate systems supporting number discrimination in typically developing infants is important for the understanding of numerical impairments in developmental disorders. It is possible that the impairment of number processing in children with developmental disorders arises as a function of an impaired small or large number discrimination system or a combination of the two.

Williams syndrome (WS) is one developmental disorder in which numerical impairments have been detected in older children and adults (Udwin, Davies & Howlin, 1996). It is a neurodevelopmental genetic disorder with a specific physical, behavioural and cognitive profile, which is estimated to have a prevalence between 1 in 7,500 and 1 in 20,000 live births and is caused by a microdeletion on the long arm of chromosome 7, affecting some 28 genes (Donnai & Karmiloff-Smith, 2000; Rosner & Semel, 2005; Tassabehji, 2003). During the last decade WS has received a lot of attention from researchers, mainly because of the uneven cognitive profile, including particular problems with number skills.

Previous studies have shown that older children and adults with WS have an impaired estimation system (Ansari, Donlan & Karmiloff-Smith, 2007; Paterson, Girelli, Butterworth & Karmiloff-Smith, 2006). Moreover, Paterson, Brown, Gsodl, Johnson and Karmiloff-Smith (1999) found that infants with WS could discriminate between two and three visually presented objects. However, thus far no studies have contrasted small and large number systems underlying number processing in the *same* children with WS. It is possible that the impairments of estimation and number comparison found in children and adults with WS are the consequence of an atypical developmental trajectory, starting with impaired large number processing in infants with WS and that small number discrimination reflects a non-numerical system for tracking the object properties of small sets, rather than reflecting a number-specific system (Uller, Carey, Huntley-Fenner & Klatt, 1999). To investigate this hypothesis, the present study used a within-subject design to explore for the first time small (2 vs. 3 dots, Experiment 1) and large number discrimination (8 vs. 16 dots, Experiment 2) in infants and toddlers with WS.

Experiment 1

Methods

Participants

Fourteen infants and toddlers with WS participated in this study. They were recruited through the Williams Syndrome Foundation, UK. All of the children had been diagnosed clinically as well as by means of a FISH test for the deletion of one copy of the elastin gene. The participants were assessed on the Bayley Scales of Infant Development (Bayley, 1993) either in London or, for two participants, as part of another study at Reading University. For the analysis of the experimental results, five participants with WS were excluded due to fussiness or parental interference with their behaviour, leaving nine participants with WS for this experiment (mean Chronological Age (CA): 35 months; range: 13 to 53 months and mean Mental Age (MA): 22 months; range: 7 to 38).

Apparatus

The participants were seated in a high-chair 60 cm away from a 97 × 56 cm monitor screen, surrounded by off-white curtains with dimmed light from above in an otherwise darkened room. Their mothers were seated on a stool behind their child and instructed to look at their child's head and refrain from talking or interfering during testing. A camera was focused from under the monitor screen on the upper part of the infant's face. This camera was connected to a VCR and monitor screen where the experimenter, who was in the same room but behind curtains, monitored the child's looks. A 'picture-in-picture'

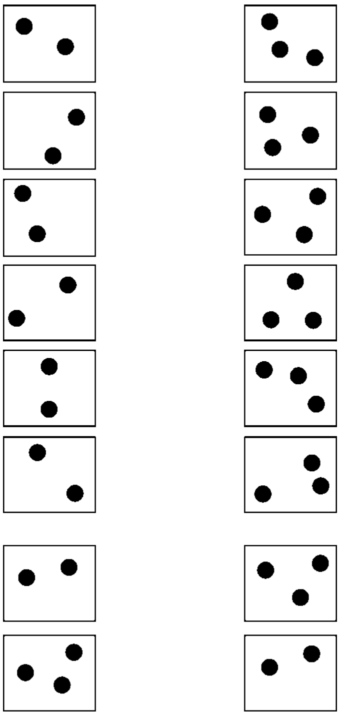

Figure 1 *Schematic representation of six of the nine familiarization and the first test trial displays for Experiment 1.*

tool showed the displays of the infant's monitor screen in the right corner of the experimenter's monitor for subsequent coding purposes.

Procedure

Experiments 1 and 2 were carried out in alternating order, so that half of the children were presented with Experiment 1 first. For Experiment 1, four of the infants were familiarized with 2 dots and tested on 3 dots, while five children were familiarized with 3 dots and tested on 2 dots. Before each trial was started a Teletubbie appeared in the middle of the screen combined with a squeaky sound in order to attract the child's attention. Once the child was looking at the screen, the trial started. It ended with a first 2 seconds look away or until 10 seconds had elapsed. After nine familiarization trials, the child was presented with six test trials in which either the new or familiarized number was displayed in alternating order.

Stimuli

All dots were presented on a 17 by 19 cm white background in both the familiarization and the test displays. Infants were familiarized with nine trials showing either 2 or 3 dots (see Figure 1) which were chosen at random out of a set of six displays in which the position of the dots varied, after which they were presented in alternating order with three familiar trials and three novel trials that were randomly chosen out of six trials for each number.

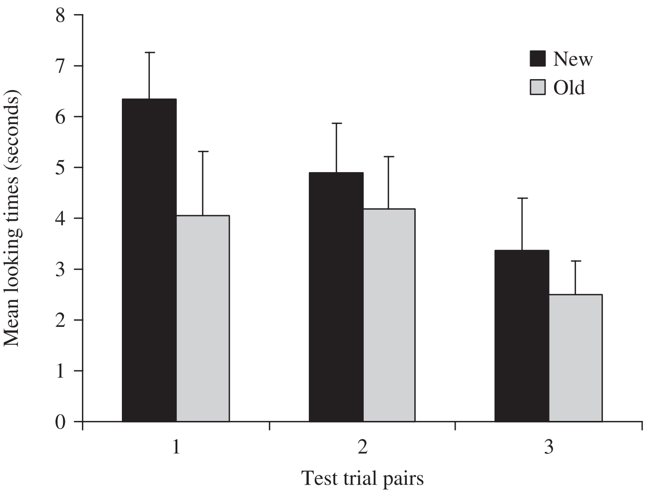

Figure 2 *Mean looking times for old vs. new test trials in Experiment 1. Error bars represent the Standard Error of the Mean.*

All trials were presented and randomized using E-prime (Psychological Software Tools, Pittsburgh, USA).

Results

Infants' looking times were coded frame-by-frame using The Observer computer programme (Noldus, Nottingham, UK) for managing and analysing data. A second person coded 42% of the trials; inter-rater reliability was high (r = .96).

To investigate the main effect of test trial pair (1, 2 and 3) and test trial type (old vs. new) a 3*2 mixed repeated-measure Analysis of Variance (ANOVA) was carried out. There was a marginally significant effect of test trial type [$F(1, 8) = 4{,}974$, $p = .056$]. There were no significant effects for test trial pair [$F(2, 16) = 2{,}964$, $p = .08$] or for the interaction between test trail pair and test trial type [$F(2, 16) = 0{,}757$, $p = .485$]. To investigate the locus of the marginally significant main effect of test trial type, post-hoc t-tests were run. These revealed a significant effect for the first test trial pair [$t(8) = 2{,}542$, $p = .035$] but not for the second [$t(8) = 0{,}543$, $p = .602$] or third [$t(8) = 1{,}325$, $p = .221$] test trial pair. Inspection of individual looking times revealed that six out of nine participants looked longer at the new number (M = 6.33 s, SD = 2.82) compared to the old number (M = 4.05 s, SD = 3.78) in the first test trial (Figure 2). Given the large individual variability, we also ran non-parametric contrasts of the looking times in the old and new test trial pairs. Consistent with the t-test reported above, Wilcoxon tests for the three test trail pairs revealed a significant effect only for the first test trial pair ($z = -1{,}960$, $p = .050$) (second pair: $z = -0{,}770$, $p = .441$; third pair: $z = -1{,}245$, $p = .231$).

To further explore the relationship between old and new number discrimination, we contrasted the last three habituation trials with the first test trial. This was done

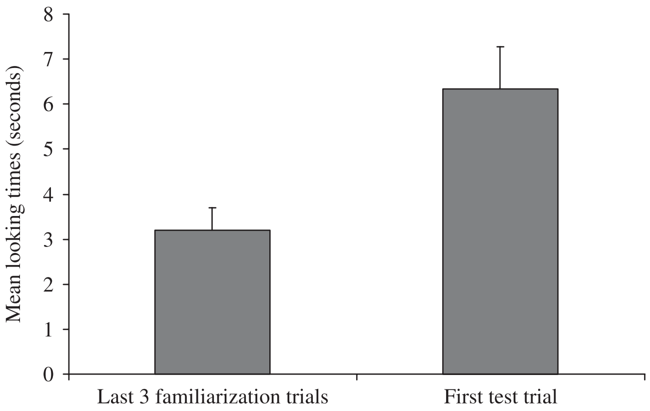

Figure 3 *Mean looking times for the last three familiarization trials and the first test trial in Experiment 1. Error bars represent the Standard Error of the Mean.*

to investigate the extent to which looking times during the end of familiarization differed from the occurrence of the first test trial (see Figure 3). A *t*-test revealed that participants looked longer at the first new test trial ($M = 6.33$ s, $SD = 2.82$) compared to the mean of the last three familiarization trials ($M = 3.21$ s, $SD = 1.47$): ($t(8) = -4{,}197$, $p = .003$). Inspection of the individual subjects' looking times revealed that eight out of nine subjects followed this trend. The result from the non-parametric Wilcoxon test converged with the above analyses: $z = 2.52$, $p = .012$.

Discussion

Analyses of variance computed over the three pairs of old and new test trials revealed a marginally significant effect for the comparison of old vs. new test trials, indicating that the infants looked longer at the new number displays on the test trials. Consistent with this, a significant effect emerged only for the first test trial pair in parametric and non-parametric post-hoc analyses of differences between old and new for each test trial pair. This finding can be explained by the fact that overall the looking times in all the test trials decreased, perhaps as a result of re-habituation. Also, the WS infants and toddlers looked significantly longer at the first test trial than the last three familiarization trials, implying that their looking times recovered significantly from habituation upon presentation of the first novel numerosity. These findings suggest that WS infants attended to the stimuli in the present experiment and were capable of noticing stimulus changes. It should be noted that due to the difficulty of finding very young participants with WS, together with the substantial drop-out rate, the sample from which these findings are drawn was relatively small. Furthermore, the absence of a significant interaction of test trial (1st, 2nd and 3rd test trial) and type (old vs. new) indicates that, while there are differences between the different test trial pairs, they did not differ from one another significantly.

The primary purpose of Experiment 1 was to establish whether infants with WS would exhibit looking time differences for the discrimination of small sets, similar to those found in the literature on typical development. In other words, Experiment 1 served as a control experiment for Experiment 2. It is thus likely that WS infants relied upon a non-numerical representation in Experiment 1. To assess whether WS infants, like typically developing 6-months-olds, could discriminate between large numerosities, the same infants were tested for their ability to discriminate between larger numerosities, a process that cannot be supported by non-numerical object-file representations.

Experiment 2

In Experiment 2, we used the same stimulus parameters as those used by Xu and Spelke (2000), and controlled for variables such as overall contour length, average brightness and overall surface covered. The only difference was that in the present study displays were presented by computer on a monitor instead of manually on a stage.

Method

Participants

To enable comparison between Experiments 1 and 2, only data from the same nine participants from Experiment 1 were analysed.

Apparatus

The same apparatus was used as in Experiment 1.

Procedure

As the order of the experiment was counterbalanced, five of the infants were presented with Experiment 2 first. For Experiment 2, five infants were familiarized with displays of 8 dots and tested on 16 dots. The other four infants were familiarized with 16 dots and tested on 8 dots. The rest of the procedure was the same as described in Experiment 1.

Stimuli

The familiarization displays consisted of either 8 or 16 black dots, which were presented on an 18 × 19 cm white background. Six of the nine familiarization trials had exactly the same measurements as those used by Xu and Spelke (2000). The three remaining trials were at random, selected from the previous six trials. All were presented and randomized by E-prime software. Dot size and position were randomized across trials. The displays were controlled for average brightness, display density, element size and display size. This means that the density in the

8 dots was the same as the 16-dot displays and that the individual surface of each dot in the smaller amount (mean dot diameter 1.83 cm, range 1.06–2.37 cm) was double the size of each display with 16 dots (mean 1.30 cm, range 0.75–1.67 cm). The overall size of the elements, the overall brightness and the average contour length of the dots was the same across all trials (see Xu & Spelke, 2000, B5).

Element density (0.035 dots/cm) and display height (19 cm) were the same as those in the familiarization trials. The size of the displays varied from the familiarization trials as well between the 16-dots (24 cm) and 8-dots (12 cm) test trials, and the size of the individual elements was equated (1.5 cm in diameter). Thus, continuous variables were equated across the two conditions as well as across the test displays.

Results and analysis

Before performing data analyses, looking times were coded and excluded in the same way as described in the first experiment. A correlation of $r = .96$ for the ratings of the two coders was found.

A 3*2 mixed repeated-measure ANOVA, testing the effects of test trial pair (1, 2 and 3) and test trial type (old versus new), revealed no significant effects for either test trial pair [$F(2, 16) = 0.720$, $p = .502$], test trial type [$F(1, 8) = 0{,}523$, $p = .490$] or the interaction test trial pair and test trial type [$F(2, 16) = 1{,}482$, $p = .257$]. Furthermore, no significant results were found in paired t-tests that compared the looking times for the new stimuli to the old stimuli for each test trial pair separately (first pair: $t(8) = 1{,}463$, $p = .182$; second pair: $t(8) = 0{,}345$, $p = .739$ and third pair: $t(8) = -0.841$, $p = .425$). Similarly, non-parametric Wilcoxon tests revealed no significant differences between old and new numerosities for each of the three test trial pairs.

In contrast to Experiment 1, a dependent t-test comparing the mean of the last three familiarization trials to the first test trial with new number was not carried out, because the size of the displays of the familiarization trials and test trials differed. This design by Xu and Spelke (2000) was undertaken to ensure that when comparing old and novel test trials, infants could not rely on density as a cue. This restricts the contrast between old and new stimuli to the test trials.

Discussion

In contrast to findings in TD infants (Brannon *et al.*, 2004; Xu & Spelke, 2000; Xu & Arriaga, in press; Xu *et al.*, 2005), and despite the fact that the WS infants were older than the TD infants reported in such studies, no evidence for large number discrimination was found in infants with WS. An inspection of Figure 4 suggests a non-significant difference between old and new numerosities for the first test trial pair. This non-significant difference may represent a trend towards a discrimination, which should be further investigated in future work with larger samples of infants. A comparison of Figures 4 and 2 clearly shows that infants looked consistently longer at the novel compared to the old test in Experiment 1, but not in Experiment 2.

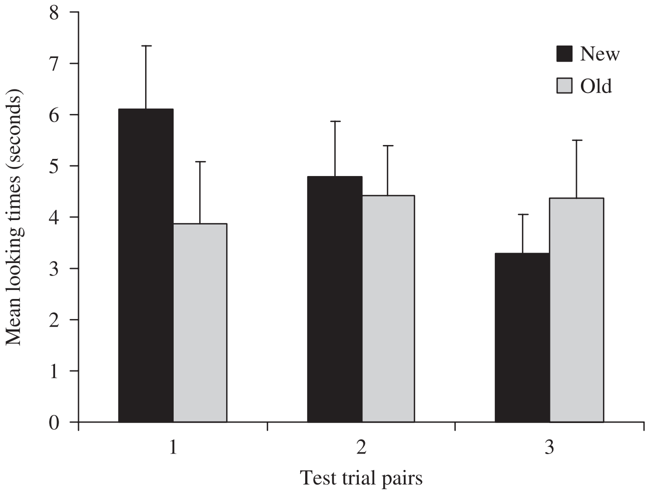

Figure 4 *Mean looking times differences for old and new number in the test trials in Experiment 2. Error bars represent the Standard Error of the Mean.*

General discussion

A growing body of studies suggests that two different representational systems may underlie small and large number discrimination in typically developing infants (Clearfield & Mix, 1999; Feigenson *et al.*, 2002; Xu, 2003; Xu *et al.*, 2005). While TD infants fail to discriminate between small numerosities when variables such as stimulus area, contour length and density are controlled for, they succeed in discriminating between large numerosities, even when these other continuous variables are controlled for.

This difference between systems for processing small and large numerosities in infancy is interesting from the point of view of precursors for both typical and atypical number development. It appears that large numerosity discrimination in infancy may be more ‘numerical’ in nature than small number processing, since typically developing infants are able to discriminate large but not small numerosities when numerosity is disconfounded from all continuous variables (area, density, etc.). This raises the possibility that individual differences in large number processing in infancy are more likely to be predictive of later development of numerical cognition than is the case for small number discrimination.

The first experiment investigated small number processing abilities of infants with WS. The results showed a significant difference for the looking times within the first test trial pair, suggesting that the infants and toddlers with WS were able to process the difference between the familiarization and test stimuli. Since variables such as brightness, overall contour and density were not controlled for in the first experiment, these results merely indicate that infants and toddlers with WS can distinguish between the visual features of small numerosities based on variables continuous with number, or a combination of number and the stimulus variables confounded with it. The present finding replicates the results reported by Paterson *et al.* (1999) in revealing small numerosity discrimination abilities in infants and toddlers with WS. However, when we combine the findings of Experiments 1 and 2 in the same infants, this now challenges the interpretation that the earlier difference in looking time during the presentation of old and new small numerosities reflected a sensitivity to number. Indeed, whereas the infants in Experiment 1 were successful, the same infants failed in Experiment 2 as no evidence for large number discrimination ability emerged. These findings clearly point to deficits in large number processing in this clinical population.

Notwithstanding, it should be acknowledged that the sample tested here was small, as is often the case with rare syndromes. In future studies larger samples of groups of infants with different genetic developmental disorders should be tested. Furthermore, the direct comparison with more groups of typically developing infants would help to further understand the specific ways in which the behaviour of infants with genetic developmental disorders in numerosity discrimination deviates from that of typically developing infants. Finally, to make sure that experiments were brief enough for atypical infants, we decided to use familiarization rather than habituation in the present study. To ensure complete similarity with Xu and Spelke's (2000) original study, future studies should try to use an infant-controlled habituation methodology, which is more sensitive to inter-individual differences in the time it takes to habituate to a particular stimulus variable (e.g. numerosity).

To the best of our knowledge, this is the first study which has simultaneously investigated small and large number processing in infants and toddlers with a genetic developmental disorder, one that is known to significantly affect number processing in later development and adulthood (Paterson *et al.*, 2006; Ansari *et al.*, 2007). The present findings indicate that number processing deficits in Williams syndrome are rooted in magnitude estimation deficits in infancy and suggest that our earlier findings of WS infant success at small number discrimination (Paterson *et al.*, 1999) may in fact have had little to do with number and more to do with the tracking of individual objects non-numerically. Future work should make comparisons similar to our earlier work between infants with WS and those with Down syndrome, with the hypothesis that because individuals with DS are less impaired numerically in adulthood than those with WS (Paterson *et al.*, 2006), they will succeed on magnitude comparisons despite failing on small number discrimination, with the infants with WS showing the opposite pattern. In fact, such cross-syndrome results would further validate the notion that two very different systems underlie the development of number. Our current results with Williams syndrome are encouraging and provide a strong impetus for tracing back to infancy full developmental trajectories of developmental deficits in all syndromes (Annaz, Karmiloff-Smith & Thomas, in press; Karmiloff-Smith, 1998). Our findings also highlight the need for early, syndrome-specific intervention strategies during the period of maximal cortical plasticity of the developing brain.

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