RUNNING HEAD: Perceptual grouping in infants with Williams syndrome

The Development of Perceptual Grouping in Infants with Williams Syndrome

Emily K. Farran¹, Janice H. Brown², Victoria L. Cole¹, Carmel Houston-Price¹, Annette Karmiloff-Smith³

¹ School of Psychology and Clinical Language Sciences, University of Reading

² Department of Psychology, London South Bank University

³ Neurocognitive Development Unit, Institute of Child Health, University College

London

Address correspondence to: Emily Farran School of Psychology and Clinical Language Sciences University of Reading Earley Gate Reading RG6 6AL UK Tel: +44 (0)118 378 7531 Fax: +44 (0)118 378 6715 E-mail: <u>E.K.Farran@reading.ac.uk</u>

This research was funded by the ESRC

Abstract

Perceptual grouping by luminance similarity and by proximity was investigated in infants with Williams syndrome (WS) aged between 6 and 36 months (visit 1, N=29). WS infants who were still under 36 months old, 8 months later, repeated the testing procedure (visit 2, N=15). Performance was compared to typically developing (TD) infants aged from 2 to 20 months (N=63). Consistent with the literature, TD participants showed grouping by luminance at the youngest testing age, 2 months. Grouping by proximity had not previous been charted in typical development: this study showed grouping by proximity at 8 months. Infants with WS could group by luminance. Developmental progression of the WS group showed some similarities to typical development, although further investigation is required to further address this in more depth. In contrast, infants with WS were not able to group by proximity. This pattern of emergence and development of grouping abilities is considered in relation to the pattern of grouping abilities observed in adults with WS. The Development of Perceptual Grouping in Infants with Williams Syndrome

Introduction

Perceptual grouping or perceptual integration was first introduced by Gestalt psychologists. It is a low-level process in which local elements are grouped together according to shared properties or Gestalt principles. These include grouping by colour, orientation or shape similarity, by common fate (movement) and by proximity. Perceptual grouping is important for many aspects of visuo-spatial cognition. It functions to form objects for object recognition, to direct attention, and to increase the efficiency of processing by grouping redundant elements (see Gillam, 2001).

In typically developing infants, different forms of perceptual grouping emerge at different developmental time-points within the first few months of life. Grouping by luminance similarity is the earliest form of grouping, evidenced in neonates (Farroni, Valenza & Simion, 2000). Grouping by common fate has emerged by 4 months (Kellman & Spelke, 1983) and grouping by shape similarity has been observed in infants aged 3 to 4 months (Quinn & Bhatt, 2005). Furthermore, infants aged 4 months are able to perceive parts of an occluded object as one object, based on the shared properties of texture, colour similarity, and good continuation (Johnson & Aslin, 1996). It is not known when grouping by proximity emerges, although work in our lab suggests that it does not emerge within the first six months (Farran, Brown, Cole, Houston-Price & Karmiloff-Smith, submitted).

Taken together, the processes involved in perceptual grouping emerge early in development. As such, a deficit in processing any form of perceptual grouping could have cascading effects on later development. This is particularly pertinent for the

development of directly related functions such as local and global processing, selective attention and object recognition. The present study investigated perceptual grouping in infants and toddlers with Williams syndrome (WS) a rare genetic disorder in which attention and visuo-spatial processing are known to be poor (Brown et al., 2003; Farran & Jarrold, 2003).

Williams syndrome occurs in 1 in 20, 000 live births (Morris & Mervis, 1999). Individuals with this developmental disorder have an average IQ of approximately 60 (Mervis, 1999). However, this masks an uneven cognitive profile in which visuospatial cognition is markedly poorer than verbal cognition (e.g. Udwin & Yule, 1991). Visuo-spatial cognition in WS is not simply delayed, but shows a number of atypical characteristics. For example, although individuals with WS are able to perceive both the local and global aspects of an image (Deruelle, Rondan, Mancini & Livet, 2006; Farran, Jarrold & Gathercole, 2001; 2003), in some areas of cognition individuals with WS attend more to the local details of an image than to the configuration of the elements or the global percept. This is apparent in the relative strength observed in face processing in WS (e.g. Karmiloff-Smith, 1997; Karmiloff-Smith et al., 2004). However, such a processing style also accounts for the relative deficit in visuo-spatial construction and drawing in WS: solutions to drawing and construction tasks lack organisation, and show poor global cohesion (e.g. Bellugi, Sabo & Vaid, 1988; Farran et al. 2003; Landau, Hoffman & Kurz, 2006).

It has been hypothesised that other aspects of the profile of visuo-spatial abilities in WS reflect a 'vulnerable' dorsal visual stream (e.g. Atkinson et al., 1997; 2003; Braddick et al., 2003). For example, individuals with WS demonstrate impairment on a motion coherence task, which activates areas of the dorsal visual stream, relative to a form coherence task which activates areas of the ventral stream

(Atkinson et al., 1997, 2006). However, this pattern of performance is not specific to WS (Braddick et al., 2003). Furthermore, note that individuals with WS are able to perceive biological motion from the lights of a point-light walker (PLW), also a dorsal stream function (Jordan, Reiss, Hoffman & Landau, 2002). Thus, as the term 'vulnerable' suggests, dorsal stream functions and their relative levels of ability are fractionated in WS. As such, the dorsal stream vulnerability hypothesis will not be addressed in this study.

The atypical profile of visuo-spatial cognition in WS, particularly the evidence relating to dorsal stream vulnerabilies emphasises that one cannot assume that cortical functioning in the WS brain relates to parts intact and parts impaired. Brain plasticity dictates that atypical functioning at the infant start-state influences subsequent brain development. Thus, both behavioural and cortical developmental pathways may not be typical in WS. One can begin to establish the nature of such pathways by examining performance from early in development. For example, research into the development of numerosity has shown that the performance of adults with WS is inferior relative to adults with Down Syndrome (DS) (Bellugi, Wang & Jernigan, 1994). However, infants and toddlers with WS show relatively superior understanding of numerosity compared to infants and toddlers with DS (Paterson, Brown, Gsodl, Johnson & Karmiloff-Smith, 1999). This suggests that the developmental pathways for the development of the concept of number are different for these two disorders, and that the underlying deficit in WS is different from the deficit observed in DS.

The present study investigates perceptual grouping in infants and toddlers with WS. To date we are only aware of a single study relating to visual cognition in early development in WS. Scerif, Cornish, Wilding, Driver and Karmiloff-Smith (2004) investigated visual search in WS toddlers aged two- to three-years using a touch

screen paradigm. Targets were determined by their size, relative to the distracters. They found that toddlers with WS, although as quick as mental age (MA) matched controls, were more likely to confuse targets with distracters than controls. Thus, at the early stages of the developmental trajectory, sensitivity to differences in local elements when determined by size appears atypical in WS. The present study investigates sensitivity to perceptual grouping by proximity and luminance at two developmental time-points. This is only the second study to investigate visual cognition in infants with WS and this is the first study to track performance across two time-points in infancy and toddlerhood in WS.

Neuroanatomical abnormalities observed in adults with WS might relate to areas involved in perceptual grouping. All forms of perceptual grouping implicate early visual areas V1 and V2 (Kapadia, Westheimer & Gilbert, 1998; Ross, Grossberg & Mingolla, 2000). Investigation of the layers of V1 in adult WS brains (Galaburda, Holinger, Bellugi & Sherman, 2002) showed increased cell packing and abnormal neuronal size, compared to control brains.

Schmitt, Eliez, Bellugi and Reiss (2002), using MRI, report increased gyrification (cortical folding) in adults with WS in the right parietal and occipital lobes, whilst Reiss et al. (2000) observed a disproportionate reduction in parietaloccipital regions and a left dominance of occipital lobe in adults with WS relative to controls. These abnormalities show consistency with activation during perceptual grouping in the typical population: in an ERP study, Han, Song, Ding, Yund and Woods (2001) demonstrated that grouping by proximity in the typical population activated from striate (V1) or prestriate cortex to medial occipital and parietal cortex, whilst grouping by shape similarity activated occipitotemporal areas. Thus, whilst our knowledge of brain activation during perceptual grouping cannot address the dorsal

stream vulnerability hypothesis, brain abnormalities at the adult end-state clearly predict that perceptual grouping in WS may be atypical. It is not clear, however, whether these abnormalities are present from the infant start-state, or are the product of atypical development.

In line with the neuroanatomical evidence above, Farran (2005) demonstrated impaired perceptual grouping abilities in older children and adults with WS. Participants were asked to determine whether a matrix of local elements were grouped into columns or rows. Six forms of perceptual grouping were employed. The performance of the WS group was compared to that of a control group of typically developing (TD) children matched by non-verbal ability. Results showed that whilst the WS group showed a similar level of performance to controls for grouping by luminance similarity, closure, and alignment, performance was significantly poorer than the controls for grouping by shape similarity, orientation similarity and proximity. This adds to our knowledge of local and global processing in WS; the grouping together of local parts into a global percept is based on common local properties. We now know that individuals with WS can perceive a global percept (e.g. Deruelle et al., 2006). However, the uneven profile of perceptual grouping abilities at the adult end-state suggests that global perception may not be typical in WS, i.e. it is possible that individuals with WS rely on their relative strengths in grouping by luminance similarity, closure and alignment to group local elements together into a global whole. Furthermore, it is possible that atypical perceptual grouping abilities contribute to the global impairments observed in image production (drawings or construction tasks) in WS.

The present study will enable us to determine whether perceptual grouping is atypical in WS from the start of the development of these processes or whether

processing becomes atypical with development. Infants will participate twice, with 8 months between visits. We aim to observe the early stages of development for two types of perceptual grouping, luminance similarity and proximity. These were chosen as they represent relatively strong and relatively weak forms of perceptual grouping in WS adults respectively, and so are likely to have different developmental trajectories. If perceptual grouping is impaired from early in the developmental trajectory, it is possible that this deficit has cascading effects on a number of developmental processes.

Method

Participants

Williams Syndrome Infants

Twenty-nine Williams Syndrome infants and toddlers were recruited from across the UK via the Williams Syndrome Foundation (WSF-UK). All individuals had received a positive genetic diagnosis for WS. Diagnosis was by a Fluorescent insitu Hybridisation (FISH) test, which checks for the deletion of elastin on the long arm of chromosome 7. Elastin is one of the twenty-four genes typically deleted in WS (Tassabehji, 2003) and is deleted in approximately 95% of individuals with WS (Lenhoff, Wang, Greenberg & Bellugi, 1997). All individuals had also been diagnosed phenotypically by a clinician. Participant chronological ages ranged from 6 to 36 months. Infants who were below 28 months of age at first test (visit 1) repeated the testing procedure 8 months later (visit 2) to assess any development of the group over time. Participant mental ages were assessed using the Bayley's Scale of Infant Development II (BSID II, Bayley, 1993). Mental ages ranged from 2 to 30 months.

Typically developing (TD) infants

Forty-three TD infants and toddlers were recruited from the infant research database held at the School of Psychology and Clinical Language Sciences, University of Reading. The majority of the infants on the database were initially recruited from local maternity wards in Reading, Berkshire. These consisted of four age groups: 8 months (N=11), 12 months (N=11), 16 months (N=10) and 20 months (N=11). Mental age was also assessed in this group, again using the BSID II (Bayley, 1993). Mental ages ranged from 8 to 24 months, and did not differ significantly from CA (t(42)=0.29, p=.77).

In addition to the TD infants detailed above, data is also included from infants from another study (Farran et al., submitted). These infants were part of a cohort of sixty-one infants who had taken part in a longitudinal study which, among other tasks, involved completing the same grouping tasks administered here. The infants included in the present analysis were the first 10 2-month-old infants who contributed a full set of data (three infants were excluded using exclusion criteria described below, and a further four had incomplete datasets). The same selection criteria were employed to identify 10 4-month-olds (six infants were excluded using exclusion criteria described below and a further two due to incomplete datasets), with the additional restriction that infants who had been selected at 2 months were exempt from selection at 4 months. This formed two independent groups (2 months, N=10; 4 months, N=10). As these infants were assessed for a different study, mental ages are not available, thus we assume that MA did not differ from CA.

Due to fussiness, not all infants completed all four grouping experiment tasks. In addition, some data was excluded from analysis due to infants demonstrating a side bias (>95% of looking time was to one side), a bias toward either vertical or horizontal stripes at baseline (>95% of looking time was to vertical or horizontal

stripes), or overall disinterest in the task (determined as spending less than 15% of available time attending to either test stimulus). The resulting WS and TD participant numbers for each grouping task at each age are shown in Table 1. For clarity, the WS data and the TD data at 8, 12, 16 and 20 months are exclusive to this study, whilst the TD data at 2 and 4 months are also presented as part of another study (Farran et al., submitted).

Table	1	about	here

Stimuli

All stimuli were equated for luminance (all had an overall luminance of 50 to 55 cd/m²). Examples of familiarisation stimuli are shown in Figure 1. Each familiarisation stimulus was a matrix of local elements which subtended a visual angle of 14.5 degrees squared and contained local elements which were grouped either horizontally or vertically. The luminance stimulus displayed 16 circles in a 4-by-4 formation. In order to give the best opportunity for grouping to occur, circles were black or white (the highest possible luminance contrast). The proximity stimulus displayed 24 squares in a 4-by-6 formation. The choice to include 6 elements in the more proximal dimension was dictated by the number of elements that could be fitted within the square of the matrix, while keeping the size of each element consistent with the luminance stimulus. Local elements subtended a visual angle of 2.2 degrees squared. Test stripe stimuli subtended an overall visual angle of 14.5 degrees squared, each stripe subtending 14.5 by 2.2 degrees of visual angle (Figure 2). The dishabituation stimulus was a set of varying grey concentric circles on a black background, subtending overall visual angle of 14.5 degrees.

Figures 1 and 2 about here

Design and Procedure

Infants were tested using a preferential looking laboratory, seated on their parent's lap, within a viewing booth. They viewed stimuli back projected onto a large screen, at a distance of 100cm. The infants aged 2 months and 4 months had a viewing distance of 50cm. Note that this dictated slightly different visual angles (matrices of elements, test stripes and dishabituation stimulus: 16.5 degrees squared; local elements and strip width: 24 degrees squared). Attention noises (a voice recording of "look!") were played from a small speaker above the centre of the screen at the onset of each stripe stimuli presentation, and at the onset of the first familiarisation stimulus presentation. This was in order to ensure attention towards the screen. The testing session included two other experiments not reported here (covert attention, and attention shifting tasks) and took no longer than 20 minutes overall, depending on individual habituation times. The order of the three experiments was fixed, although this was subject to change, dependent on infant temperament. In most instances, infants viewed an attention task, the perceptual grouping task, followed by a second attention task. The order of the attention tasks was counterbalanced, as was the order of presentation of grouping types within the perceptual grouping task. Breaks were given between grouping types or experiments if the child became agitated. In addition, horizontal/vertical stimuli arrangement was counter-balanced across infants. 50% of infants started with horizontal presentations, and the other 50% started with vertical presentations.

For each grouping type, a baseline preference for vertical versus horizontal stripes was established by measuring infant looking times to two test stimuli presented simultaneously to the left and right of the screen. One test stimulus depicted a set of vertical stripes, and the other a set of horizontal stripes (Figure 2). These were

presented for a ten second duration, followed by a left-right reversal for a further ten seconds. Participants were then presented with a familiarisation stimulus (Figure 1), composed of a matrix of local elements, which could be grouped vertically (columns) or horizontally (rows). Each presentation of the familiarisation stimulus was five seconds in duration (10 seconds at 2 months and 4 months), and was presented repeatedly until habituation towards the stimulus occurred. This was determined by online coding of infant looking behaviour, using a 50% decrement habituation procedure. Specifically, habituation was assumed when looking time to three consecutive 5-second presentations (10 seconds: 2 months, 4 months) of a stimulus had reduced by 50% of the infant's total looking time on the first three 5-second stimuli presentations.

Five-second presentations were employed to ensure that habituation was truly infant lead; longer presentation times risk habituation occurring before the minimum number of presentations needed to determine habituation, have been administered (a ceiling effect). As younger infants take longer to habituate, the risk of a ceiling effect, with the 10-second presentation times used at 2 months and at 4 months, was minimal.

After habituation had occurred, infants' preferences for horizontal versus vertical stripes were then tested again, in the same manner as at baseline. This was followed by a 5 second duration dishabituation stimulus in an attempt to remove any residual effects of habituation to one grouping type on to the subsequent grouping type. This procedure was then repeated for the remaining grouping type.

Eye movements were captured via cameras positioned at the top of the presentation screen in left, central and right positions, in order to measure looking

behaviour towards all stimuli. Video footage of all eye movements was coded frame by frame off-line and analysed using the Observer software package.

Results

The primary and secondary observer was naïve to whether a familiarity or novelty preference was anticipated. The second observer coded a random 20% of the data for each group. Inter-rater reliability showed consistent substantial agreement or almost perfect agreement (see Viera & Garrett, 2005), Cohen's Kappa: TD group, mean (SD) = 0.85 (0.07); WS, mean (SD) = 0.84 (0.08).

Exposure duration

The exposure duration of each familiarisation stimulus was infant controlled using a habituation procedure. Exposure durations for each TD age group and for the WS group at visit 1 and at visit 2 were significantly above the lowest possible duration of 25 seconds (50 seconds at 2 months and 4 months) (p < .05 for all).

Infants aged 2 and 4 months were habituated using 10-second presentations, whilst WS and TD infants aged 8 to 20 months received consecutive 5-second presentations until habituation. Due to this procedural difference, group comparisons were explored where 5-second presentations were employed only. WS visit 2 data is not included in this analysis as it is not independent of WS visit 1 data. Separate t-tests between the TD group and WS visit 1 data were carried out for grouping by luminance similarity and for grouping by proximity. Both analyses revealed significantly longer exposure durations in the WS group than the TD infants (luminance: t(58)=3.08, p<.01; proximity: t(57)=2.90, p<.01). This demonstrates that the WS infants took longer to habituate than the TD infants. Exposure durations are shown in Table 2. Separate regression analysis of the exposure duration for each

group revealed no significant relationships between cumulative exposure duration and mental age for either perceptual grouping type (WS visit 1, WS visit 2, TD: p>.05 for all).

Table 2 about here

Perceptual grouping

When an infant had viewed a familiarisation stimulus that showed vertical grouping, horizontal and vertical test stripes were coded as novel and familiar respectively. When an infant had viewed a horizontally grouped familiarisation stimulus, test stripes were coded in the opposite manner. Preferences to horizontal and vertical stripes were measured using a difference score between the looking time (msecs.) to the novel and to the familiar stripes. This was calculated by subtracting the looking time to the familiar stripes from the looking time to the novel stripes for each of the two ten second presentations. The average of these two scores was employed. Thus, a positive difference score indicates a novelty preference and a negative difference score indicates a familiarity preference. Performance at baseline and at test was compared to chance (a looking time difference score of zero msecs).

WS performance

Baseline trials indicated no spontaneous preferences for luminance similarity or proximity at visit 1 or visit 2, p>.05 for all. For test trials at visit 1 and visit 2, infants and toddlers with WS showed a marginal familiarity preference for grouping by luminance similarity at visit 1 (t(21)=-1.85, p=.08), which was significant by visit 2 (t(13)=-2.28, p=.04). This appears to indicate progression in grouping ability with development. However, within participant analysis across visits 1 and 2 (N = 10 as not all participants completed the grouping by luminance condition at both visits) did not support this, t(9)=0.964, p=.36. Grouping by proximity was not evident at visit 1

or visit 2 (p>.05 for both), and showed no within participant development (N=12), t(11)=0.15, p=.88. This is illustrated in Figure 3, and shows that infants with WS are able to group by luminance similarity, but not by proximity.

It is possible that within participant development was not apparent due to the age range of the WS group, and the overlap in mental age and chronological age between visits 1 and 2. Performance was therefore also analysed at each visit using linear regression against mental age (MA) and chronological age (CA). At visit 2, grouping by luminance similarity showed a marginal relationship with mental age and with chronological age: mental age, F(1, 12)=4.69, p=.051, $r^2=.28$; chronological age, F(1, 12)=3.41, p=.09, $r^2=.22$. This was due to a reduced familiarity effect with increasing mental/chronological age, and is illustrated in Figure 4. Interestingly, this indicates that the ability to group by luminance similarity is linked to development. However, this was not evident at visit 1 (F<1 for both).

Regression analysis for grouping for proximity was consistent with the preference data above. There was no relationship between mental age or chronological age and grouping by proximity at either visit 1 or visit 2 (p>.05 for all).

Typically developing performance

Performance at baseline did not differ significantly from zero for grouping by luminance similarity or grouping by proximity (p>.05 for all). Performance on test trials was analysed for each age group separately. Grouping by luminance similarity was observed as a familiarity effect at 2 months, t(9)= -2.29, p=.05. However, at 4, 8, 12, 16, and 20 months, this effect was no longer evident (p>.05 for all). Grouping by proximity was not available at 2 months and 4 months (p>.05 for both), but was observed as a familiarity effect at 8 months, t(9)= -3.24, p=.01. At 12, 16 and 20 months, grouping by proximity was no longer evident (p>.05 for all).

The preferences observed in the TD groups were both familiarity preferences. It is therefore possible that the subsequent loss of an effect of grouping reflects a linear progression from familiarity towards novelty preferences with increasing age (CA and MA did not differ in this group) (see Hunter & Ames, 1988). To investigate this, linear regression was carried on the relationship between grouping performance and CA, from the age at which a grouping preference had been observed (luminance similarity, from 2 months; proximity, from 8 months). For grouping by luminance similarity, the relationship between performance and CA was observed in the predicted direction (F(1, 56)=2.92, p=.09, $r^2=.05$). However, the effect was marginal and so cannot be said to truly support progression from familiarity to novelty preferences. Grouping by proximity was not related to CA (F<1). Performance at each age group is illustrated in Figures 5a and b.

Figures 4, 5a and 5b about here
Diaguasian

Discussion

This study demonstrated that grouping by luminance similarity emerges earlier than grouping by proximity in WS. Infants and toddlers with WS aged between 14 and 36 months (visit 2) showed evidence of grouping by luminance similarity (familiarity preference). Grouping ability was assessed across two time-points, spaced 8 months apart (visit 1, visit 2). Grouping by luminance similarity was marginally evident at visit 1, and had fully emerged at visit 2. We suggest that this difference is accounted for by a lack of grouping ability in the youngest WS participants at visit 1 (range: 6 to 36 months). Thus, we can conclude that the ability to group by luminance similarity has emerged in WS by 14 months, but not by 6 months. In addition, when present (visit 2) grouping by luminance was marginally related to mental age (MA) and chronological age (CA), which could indicate that the ability to group by luminance is, to some extent, linked to general development. In contrast, grouping by proximity was not evident in this group at either the first or second time-point and showed no progression with MA or CA. This suggests that the ability to group by proximity (at least for the stimuli employed here) develops beyond 36 months of age in WS, a substantial delay relative to typical development.

Comparisons between the infant start-state and the adult end-state can inform how that end-state transpired. For perceptual grouping, the pattern of performance observed in older children and adults with WS appears to stem from infancy. At the adult end-state, grouping by proximity is relatively weaker than the ability to group by luminance similarity in WS (Farran, 2005). Thus, both the infant data presented here and Farran's (2005) adult data suggest that, in WS, luminance similarity is a more robust form of grouping ability than grouping by proximity, and we suspect, other forms of grouping as well. The late emergence of grouping by proximity is likely not only to have a negative effect on the subsequent development of this grouping ability, as observed by Farran (2005), but may also have cascading effects on the emergence of other related abilities such as focussed attention (see Karmiloff-Smith, 1997).

In the present study, TD participants, consistent with the literature, demonstrated early emergence of grouping by luminance similarity, which compared to later emergence of grouping by proximity. The ability to group by luminance similarity was present at the youngest testing age, 2 months (familiarity), whilst the ability to group by proximity was evident only at 8 months (familiarity). Previous studies have shown grouping by luminance in neonates (Farroni et al., 2000). Indeed, luminance similarity is described as the most robust form of grouping ability in typical

development (Bremner, 1994). The emergence of grouping by proximity, before the current study, had not been charted. However, Farran et al. (submitted) found no evidence of its emergence within the first 6 months. As such, the current finding of grouping by proximity in typical development at 8 months is, itself, a novel finding. Although one must note that emergence at this age could be specific to the stimuli employed. Further experimentation could determine whether a higher ratio of vertical and horizontal proximity elicits evidence of grouping at an earlier age.

After the appearance of grouping by luminance at 2 months, subsequent age groups showed no significant evidence of grouping by luminance similarity. A possible move from familiarity to novelty preferences with development was explored, but not supported. For grouping by proximity, the TD groups showed grouping at 8 months, but not at subsequent months. One explanation for the pattern observed, and also a possible explanation for the pattern observed for grouping by luminance is that the tasks failed to capture grouping abilities at older ages due to a lack of interest rather than a lack of ability. Perhaps, in this study, the tasks were not attractive enough for the older infants employed here. This account would predict that the time taken to habituate would reduce with age. However as exposure duration was low for all TD infants, no reduction was observed.

The relatively long exposure durations in the WS group demonstrate that both tasks were set at an appropriate level to capture grouping ability in this population. This supports our conclusion that the WS group lacked the ability to group by proximity, and that they did not lack interest. Despite this, as the pattern of results of the TD group was not as expected, this dictates that the novel and interesting finding from the WS group should nevertheless be treated with caution.

The relatively early ability to group by luminance observed in WS appears consistent with typical development. This indicates that, although delayed, the developmental pathway has the potential to be typical. In turn, if it is relatively robust, it is less likely to be affected by later atypical development in other areas of cognition. In contrast, it appears from the substantially delayed emergence of the ability to group by proximity in WS, and subsequent deficit in adulthood, that the developmental pathway for grouping by proximity is likely to be atypical in WS.

Developmental trajectories of performance were observed using linear regression. This method is particularly relevant for the participant group employed for two reasons. First, few individuals with WS are diagnosed within the first year of life, and so the participants employed had wide age range, ideally suited for examining developmental pathways. Second, individuals with WS show large individual differences. This can be taken into account by substituting chronological age (CA) with mental age (MA) as the predictor variable.

For grouping by luminance similarity, performance in the WS group at visit 2 demonstrated a marginal linear relationship such that the familiarity preference was strongest for lower MA and CA, and became weaker with development. The same pattern was also marginally observed in the TD group, in relation to CA. This gives some further indication of relatively typical development of this grouping ability in WS, although as the results are marginal, this suggestion is tentative. In contrast, the ability to group by proximity was not related to MA or CA in WS. However, this is not surprising given that this ability had not yet emerged in the WS group.

Individuals with WS were assessed at two time-points, spaced 8 months apart. Despite some evidence, from separate analyses at each visit, that the ability to group by luminance similarity emerged within this period, a within group comparison did

not reveal any development change. Development is slower in WS than in the typical population: the fifteen individuals who took part in visits 1 and 2 increased in mental age on average by just less than 5 months. It might be that a time window of just 8 months (5 months developmental time) was not adequate to observe development, particularly given the within group variability.

The evidence for grouping is observed in this study as familiarity effects and not as novelty effects. This is surprising, given that infants were habituated to the familiarisation stimuli. This is also not consistent with previous studies which have looked at perceptual grouping in infancy, where novelty preferences are reported (e.g. Farroni et al., 2002; Quinn et al., 1993). In their model of infant attention, Hunter and Ames (1988) proposed that a familiarity preference occurs when infants require time to consolidate the familiarisation stimulus with the test stimulus. With development, consolidation requires less time, and a novelty preference is more likely to be observed. One could argue that in the present experiment, as the test stripes differed from the familiarisation stimuli, the infant noticed the similarity in organisation (horizontal or vertical), but the differences in the stimuli required further consolidation. Hence, infants showed a preference to the 'familiar' test stripes. However, this argument is difficult to support for two reasons. First, in the present study, the reduction in familiarity preferences with age, for grouping by luminance similarity, was only marginal. This does not appear to constitute a progression from familiarity to novelty. Second, novelty preferences have been reported in similar experiments, in which the familiarisation stimuli and test stimuli were different (Quinn et al., Experiment 1, 2002; Farroni et al., Experiments 2 & 3, 2000).

In addition to the perceptual grouping experiments reported here, participants also took part in an attention shifting task and a covert attention task. The stimuli in

the attention tasks were more attractive (coloured and moving stimuli) than the perceptual grouping stimuli. Thus, although habituated to the perceptual grouping stimuli, where this was preceded by an attention task, the static and dynamic changes in luminance of the attention stimuli might have had some residual effect on perceptual grouping performance. If this is the case, consolidation of the similarity between the familiarisation and test stripes might have been required, hence the resulting familiarity effects. This could explain the difference between the present results and previous studies (e.g. Farroni et al., 2000). We concede that there is uncertainty as to why familiarity preferences occurred given the habituation procedure employed. Nevertheless, it is clear that the familiarity effects observed can only result from infants' ability to perform perceptual grouping.

In summary, the profile of grouping abilities observed in adults with WS appears to relate to the emergence of perceptual grouping in infancy. In this study, the ability to group by two grouping principles was assessed at two time-points in early development. Results showed that infants and toddlers with WS can perform perceptual grouping. However, not all forms of grouping emerge at a developmental time-point predicted by typical development. Grouping by luminance similarity is available in infancy, whilst grouping by proximity emerges later. Luminance appears to be the most robust grouping type in WS as it remains a relative strength in adulthood in WS. This data can also be treated as preliminary evidence that groping by luminance might also show a similar developmental pattern in infancy as observed in typical development. The lack of evidence for the emergence of the ability to group by proximity suggests that this ability emerges relatively late in development. This could be a precursor to the poor level of ability observed for grouping by proximity in WS adults. Impairment at the infant start-state is likely to indicate an atypical

developmental trajectory, which will then have cascading effects on the development of other functions. Therefore, it is also likely, that the late emergence of grouping by proximity affects the subsequent development of related functions such as focussed attention.

References

Atkinson, J., King, J., Braddick, O., Nokes, L., Anker, S., & Braddick, F. (1997). A specific deficit of dorsal stream function in Williams syndrome. *Neuroreport: cognitive neuroscience and neuropsychology*, 8, 1919-1922.

Atkinson, J., Braddick, O., Anker, S., Curran, W., Andrew, R., Wattam-Bell, J., et al. (2003). Neurobiological models of visuospatial cognition in children with Williams syndrome: Measures of dorsal-stream and frontal function. *Developmental Neuropsychology*, 23, 139-172.

- Atkinson, J., Braddick, O., Rose, F. E., Searcy, Y. M., Wattam-Bell, J., & Bellugi, U.
 (2006). Dorsal-stream motion processing deficits persist into adulthood in
 Williams syndrome. *Neuropsychologia*, 44, 828-833.
- Bayley, N. (1993). *The Bayley Scales of Infant Development (2nd ed.)*, San Antonio,TX: The Psychological Corporation.
- Bellugi, U., Sabo, H., & Vaid, J. (1988). Spatial deficits in children with Williams
 Syndrome. In J. Stiles-Davis, U. Kritchevshy & U. Bellugi (Eds.), *Spatial Cognition: Brain Bases and Development* (pp. 273-297). Hillsdale, New
 Jersey: Lawrence Erlbaum.
- Bellugi, U., Wang, P. P., & Jernigan, T. L. (1994). Williams syndrome: An unusual neuropsychological profile. In S. H. Broman & J. Grafman (Eds.), *Atypical cognitive deficits in developmental disorders: Implications for brain function* (pp. 23-56). Hillsdale, N.J.: Lawrence Erlbaum Associates.
- Braddick, O., Atkinson, J., & Wattam-Bell, J. (2003). Normal and anomalous development of visual motion processing: motion coherence and 'dorsal stream vulnerability'. *Neuropsychologia*, *41*, 1769-1783.

Bremner, G. (1994). Infancy. Oxford, England: Blackwell.

- Brown, J. H., Johnson, M. H., Paterson, S. J., Gilmore, R., Longhi, E., & Karmiloff-Smith, A. (2003). Spatial representation and attention in toddlers with
 Williams syndrome and Down Syndrome. *Neuropsychologia*, 41, 1037-1046
- Deruelle, C., Rondan, C., Mancini, J., & Livet, M.-O. (2006). Do children with Williams syndrome fail to process visual configural information? *Research in Developmental Disabilities*, 27, 243-253.
- Farran, E. K. (2005). Perceptual grouping ability in Williams syndrome: Evidence for deviant patterns of performance. *Neuropsychologia*, 43, 815-822.
- Farran, E.K., Brown, J.H., Cole, V.L., Houston-Price, C. & Karmiloff-Smith, A. (submitted). A longitudinal study of perceptual grouping by proximity, luminance and shape in infants at two, four and six months. *World Journal of Paediatrics*.
- Farran, E. K., & Jarrold, C. (2003). Visuo-spatial cognition in Williams syndrome: Reviewing and accounting for the strengths and weaknesses in performance. *Developmental Neuropsychology*, 23, 173-200.
- Farran, E. K., Jarrold, C., & Gathercole, S. E. (2003). Divided attention, selective attention and drawing: Processing preferences in Williams syndrome are dependent on the task administered. *Neuropsychologia*, 23, 175-202.
- Farran, E. K., Jarrold, C., & Gathercole, S. E. (2001). Block design performance in the Williams syndrome phenotype: A problem with mental imagery? *Journal* of Child Psychology and Psychiatry, 42, 719-728.
- Farroni, T., Valenza, E., & Simion, F. (2000). Configural processing at birth:Evidence for perceptual organisation. *Perception*, 29, 355-372.

- Galaburda, A., Holinger, D. P., Bellugi, U., & Sherman, G. F. (2002). Williams syndrome: Neuronal size and neuronal-packing density in primary visual cortex. *Achives of Neurology*, 59, 1461-1467.
- Gillam, B. (2001). Varieties of grouping and its role in determining surface layout. InT. F. Shipley & P. J. Kellman (Eds.), *From Fragments to Objects:Segmentation and Grouping in Vision* (pp. 247-264). London: Elsevier.
- Han, S., Song, Y., Ding, Y., Yund, E. W., & Woods, D. L. (2001). Neural substrates for visual perceptual grouping in humans. *Psychophysiology*, 38, 926-935.
- Hunter, M. A., & Ames, E. W. (1988). A multifactor model of infant preferences for novel and familiar stimuli. In C. Rovee-Collier & L. Lipsitt (Eds.), *Advances in infancy research* (Vol. 5, pp. 69-95). Stanford: Ablex.
- Johnson, S. P., & Aslin, R. N. (1996). Perception of object unity in young infants: The roles of motion, depth and orientation. *Cognitive Development*, *18*, 133-143.
- Jordan, H., Reiss, J., Hoffman, J. E., & Landau, B. (2002). Intact perception of biological motion in the face of profound spatial deficits: Williams syndrome. *Psychological Science*, 13, 162-167.
- Kapadia, M. K., Westheimer, G., & Gilbert, C. D. (1998). Spatial distribution and dynamics of contextual interactions in cortical area V1. Society for Neuroscience Abstracts, 789.786.
- Karmiloff-Smith, A. (1997). Crucial differences between developmental cognitive neuroscience and adult neuropsychology. *Developmental Neuropsychology*, 13, 513-524.
- Karmiloff-Smith, A., Thomas, M., Annaz, D., Humphreys, K., Ewing, S., Brace, N., et al. (2004). Exploring the Williams syndrome face processing debate: The

importance of building developmental trajectories. *Journal of Child Psychology and Psychiatry*, 45, 1258-1274.

- Kellman, P. J., & Spelke, E. S. (1983). Perception of partly occluded object in infancy. *Cognitive Psychology*, 15, 483-524.
- Landau, B., Hoffman, J. E., & Kurz, N. (2006). Object recognition with severe deficits in Williams syndrome: sparing and breakdown. *Cognition*, 100, 483-510.
- Lehnung, M., Leplow, B., Friege, L., Herzog, A., Ferstl, R., & Mehdorn, M. (1998). Development of spatial memory and spatial orientation in preschoolers and primary school children. *British Journal of Psychology*, 89, 463-480.
- Mervis. (1999). The Williams syndrome cognitive profile: Strengths, weaknesses, and interrelations among auditory short term memory, language and visuospatial constructive cognition. In R. Fivush, W. Hirst & E. Winigrad (Eds.), *Essays in honor of Ulric Neisser*. Mahwah, NJ: Erlbaum.
- Morris, C. A., & Mervis, C. B. (1999). Williams Syndrome. In S. Goldstein & C. R. Reynolds (Eds.), *Handbook of neurodevelopmental and genetic disorders in children* (pp. 555-590). New York London: The Guilford Press.
- Paterson, S. J., Brown, J. H., Gsodl, M., Johnson, M. H., & Karmiloff-Smith, A. (1999). Cognitive modularity and genetic disorders. *Science*, 286, 2355-2358.
- Quinn, P. C., & Bhatt, R. S. (2005). Learning perceptual organisation in infancy. *Psychological Science*, 16, 511-515.
- Quinn, P.C., Bhatt, R., Brush, D., Grimes, A., Sharpnack, H. (2002). Development of form similarity as a gestalt grouping principle. *Psychological Science*, *13*, 320-328

- Quinn, P. C., Burke, S., & Rush, A. (1993). Part-whole perception in early infancy: Evidence for perceptual grouping produced by lightness similarity. *Infant Behavior and Development*, 16, 19-42.
- Reiss, A. L., Eliez, S., Schmitt, J. E., Straus, E., Lai, Z., Jones, W., & Bellugi, U.
 (2000). Neuroanatomy of Williams syndrome: A high-resolution MRI study. *Journal of Cognitive Neuroscience*, 12: supplement, 65-73.
- Ross, W. d., Grossberg, S., & Mingolla, E. (2000). Visual cortical mechanisms of perceptual grouping: interacting layers, networks, columns, and maps. *Neural Networks*, 13, 571-588.
- Scerif, G., Cornish, K., Wilding, J., Driver, J., & Karmiloff-Smith, A. (2004). Visual search in typically developing toddlers and toddlers with Fragile X or Williams syndrome. *Developmental Science*, 7, 116-130.
- Schmitt, J. E., Eliez, S., Bellugi, U., Galaburda, A., & Reiss, A. L. (2002). Increased gyrification in Williams syndrome: evidence using 3D MRI methods. *Developmental Medicine and Child Neurology*, 44, 292-295.
- Tassabehji, M. (2003). Williams-Beuren syndrome: a challenge for genotypephenotype correlations. *Human Molecular Genetics*, *12*, 229-237.
- Udwin, O., & Yule, W. (1991). A cognitive and behavioural phenotype in Williams syndrome. *Journal of Clinical and Experimental Neuropsychology*, 13, 232-244.
- Viera, A. J., & Garrett, J. M. (2005). Understanding interobserver agreement: The Kappa statistic. *Family Medicine*, 37, 360-363

Table 1. Participant Details

Group	CA days	MA days	Luminance	Proximity
WS	mean (sd)	mean (sd)	Ν	Ν
Visit 1 (N=29)	754.59	401.38	22	26
	(252.35)	(183.78)		
Visit 2 (N=15)	828.20	464.00	14	13
	(225.97)	(196.68)		
TD				
Whole Group	320.29	Na	58	53
(N=63)	(197.19)			
2m (N=10)	57.10 (1.29)	Na	10	10
4m (N=10)	122.60 (3.92)	Na	10	10
8m (N=11)	245.27 (6.40)	250.91	11	10
		(20.23)		
12m (N=11)	366.82 (6.35)	346.36	9	8
		(28.03)		
16m (N=10)	487.50 (8.03)	483.00	9	7
		(49.89)		
20m (N=11)	615.72 (4.56)	627.27 (60.68)	9	8

Group	Exposure duration (sec): Mean (S.D.)	
	Luminance	Proximity
WS visit 1	35.43(8.11)	38.15(14.42)
WS visit 2	37.14 (7.52)	38.46(11.07)
TD (8 to 20 months)	30.92(4.63)	30.15(4.92)

Table 2: Exposure Durations required to Habituate

Acknowledgements

This study was supported by a grant to the authors from the Economic and Social Research Council. The authors would like to thank the infants and parents who have kindly participated in this study, the staff of Marsh and Iffley wards at the Royal Berkshire Hospital, Reading, and the Williams Syndrome Foundation, UK, for their co-operation with this work. **Figure Captions**

Figure 1: Familiarisation stimuli

Figure 2: Test stimuli

Figure 3: Mean looking time difference scores: participants with Williams syndrome (error bars represent standard error)

Figure 4: Grouping by luminance similarity, WS Visit 2: looking time difference scores plotted against mental age

Figure 5a: Grouping by luminance similarity, TD participants: mean looking time difference scores plotted against chronological age group (error bars represent standard error)

Figure 5b: Grouping by proximity, TD participants: mean looking time difference scores plotted against chronological age group (error bars represent standard error)

Figure 1



Horizontal grouping by luminance

Vertical grouping by proximity

Figure 2



Figure 3



Figure 4



Figure 5a





