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Spatial representation and attention in toddlers with Williams syndrome and Down syndrome

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Abstract

The nature of the spatial representations that underlie simple visually guided actions early in life was investigated in toddlers with Williams syndrome (WS), Down syndrome (DS), and healthy chronological age- and mental age-matched controls, through the use of a "double-step" saccade paradigm. The experiment tested the hypothesis that, compared to typically developing infants and toddlers, and toddlers with DS, those with WS display a deficit in using spatial representations to guide actions. Levels of sustained attention were also measured within these groups, to establish whether differences in levels of engagement influenced performance on the double-step saccade task. The results showed that toddlers with WS were unable to combine extra-retinal information with retinal information to the same extent as the other groups, and displayed evidence of other deficits in saccade planning, suggesting a greater reliance on sub-cortical mechanisms than the other populations. Results also indicated that their exploration of the visual environment is less developed. The sustained attention task revealed shorter and fewer periods of sustained attention in toddlers with DS, but not those with WS, suggesting that WS performance on the double-step saccade task is not explained by poorer engagement. The findings are also discussed in relation to a possible attention disengagement deficit in WS toddlers. Our study highlights the importance of studying genetic disorders early in development.

Keywords: Williams syndrome; Down syndrome; Toddlers; Spatial representation; Sustained attention

1. Introduction

The development of mental representations used to plan eye movements (saccades), in order to select appropriate aspects of the environment to attend to, is of major importance during development. Before motor control has developed sufficiently to allow infants to explore their environment by touching or grasping, visual exploration allows them to interact with their world, and to begin to exert control over their responses to it. But the planning of saccades is no simple task. Successful spatial orientation relies on accurate perception of the physical self in relation to the environment, and adaptation to the changes both within that environment and of the position of the body. Such spatial knowledge is represented by frames of reference, a coordinate system used to code positions in space that can then be used to monitor stimuli and plan actions within the environment. The particular type of mental representation used for spatial orientation will dictate how efficiently different aspects of visual stimuli can be processed and integrated.

Attention also plays a role in the development of visual cognition, and thus in the infants ability to plan eye movements. The infant must attend to objects in the real world, and shift attention appropriately, either when an object has been fully processed, or when a new object appears in the environment. Thus, individual differences in attention levels will have an impact on infants ability to process visual stimuli.

But what happens if visual exploration or attention is impaired from early infancy onwards? In this paper, we examine this question with respect to two genetic disorders—Williams syndrome (WS) and Down syndrome (DS)—and compare these to two groups of typically developing children.

Williams syndrome is caused by a sub-microscopic deletion on chromosome 7q.11.23, and occurs in approximately 1 in 20,000 live births. Clinical features include several

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physical abnormalities that are accompanied by mild to moderate mental retardation and a specific personality profile. The interest of WS to cognitive neuroscientists stems from the very uneven profile of cognitive abilities, with spatial cognition seriously impaired and language and face processing relatively proficient (for full details, see [9,14,45]). Our knowledge of the spatial problems in the WS adult end state is relatively advanced. In contrast, we know relatively little about the development of spatial cognition in infants with WS. Furthermore, while there is some anecdotal evidence that attention is poor in children and adults, little is known about the development of attention in infants with WS.

Down syndrome is the most common chromosomal abnormality, and one of the leading causes of mental retardation, with a prevalence of 1 in 600–800 live births [13]. It is caused by extra material on chromosome 21. Trisomy 21, in which all cells have an extra chromosome 21, is the most common form of DS and accounts for 90–94% of cases. Unlike the uneven cognitive profile found in WS, the pattern of cognitive abilities in DS is usually somewhat more uniform. Both spatial cognition and attention are typically reported as problem areas in adults and children with DS (e.g. [8,19,43]). Prior to examining spatial representation and attention in these clinical groups, we provide details of the normal course of early development in these domains.

2. Spatial representation in normal development

In typically developing infants, increasingly complex representations of spatial information are used throughout infancy and then childhood, before adult representations are ultimately formed. At around the age of 6 months, children tend to use body-centred representations, i.e. they use body- or head-centred coordinates (e.g. [10,11]), while younger infants tend to rely on retinocentric representations. Gilmore and Johnson [20,21] investigated 3- and 7-month-old infants frames of reference used to plan saccadic eye movements in a double-step saccade paradigm [2,7,17,25,26,28]. In this task, participants are encouraged to look at sequences of visual targets that flash briefly in a dark visual field; the second stimulus appears and disappears before a saccade is made to the first stimulus. Subjects cannot use retinal position relative to the fovea to plan a response to the second target, because the first eye movement shifts the center of gaze, and with it, the second targets position on the retina (see Fig. 1B). Accordingly, to make accurate saccades to the location of both targets, subjects must plan the saccade to the second target by combining retinal position with information about current eye position or planned eye movements. Gilmore and Johnson [20,21] showed that 6–7-month olds were able to do this, but not 3-month olds, indicating that the ability to plan body-centred saccades emerges some time between 3 and 7 months of age.

The double-step saccade task also allows the study of the averaging of saccade signals or vector summation. In the sub-cortical route for saccade planning, it is known that if cells in the superior layers of the superior colliculus are stimulated by the presence of two adjacent targets, there results a saccade which is the sum of each stimulated cells retinal error or movement vector [35]. In other words, the resulting saccade takes a trajectory mid-way between the two targets. Johnson et al. [30] used vector sum saccades, as measured by end-points of saccades made in response to two

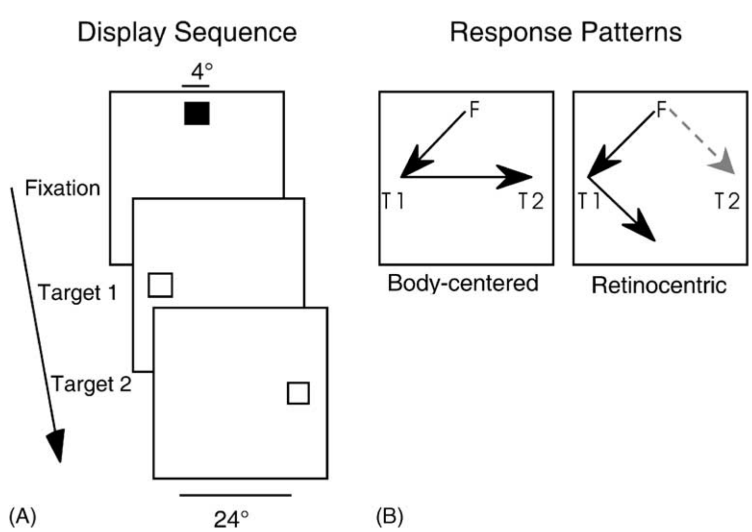

Fig. 1. Design of infant double-step saccade experiment. (A) A trial began with the presentation of a fixation stimulus consisting of a sequence of colored shapes that was followed by the brief appearance of two identical targets that flashed sequentially. Fixation positions varied in a pseudo-random order, between four possible locations: centre top (as shown), centre bottom, centre left, and centre right. (B) Two types of responses were of critical concern: a body-centred sequence consisting of two saccades, one to each target location; and a retinocentric sequence consisting of two saccades, each equivalent to the positions of the targets at the time of presentation relative to the fovea. The dashed line to target 2 represents the direction of the second saccade in the retinocentric sequence, if no saccade had been made to the first target, and mirrors the direction of the actual second saccade in the retinocentric sequence.

simultaneously presented stimuli, as a marker of sub-cortical control, and demonstrated that there is a decrease in such saccades from 2 to 6 months in healthy infants, indexing increasing cortical control during this period.

It has been well established in the neuropsychological literature that visual processing in the cortex involves two pathways, one ending in the occipito-parietal region, usually referred to as the "dorsal stream", and the other pathway ending in the occipito-temporal region, known as the "ventral stream" [3,23,46]. One of the primary functions associated with the dorsal stream is its involvement in visually-guided action, particularly when actions are governed by spatial representations [1,12]. An area of intense neurophysiological investigation in this respect has been the role of neurons in parts of the parietal cortex in establishing "body-centred" frames of reference for action, i.e. integrating information about head, eye, and body position with respect to a target in the environment [1,15,22,42].

3. Attention in normal development

Spatial attention, as defined by Posner and Peterson (e.g. [37]) consists of three phases: engage, disengage, and shift. Similar components are also found in attention in infancy. Lansink and Richards [34] describe three phases of infant attention: stimulus orienting, sustained attention, and attention disengagement.

The first phase of attention, stimulus orienting, involves the direction of attention toward a spatial location or object of interest. This usually (but not always) involves eye and head movements. The development of orienting is thought to progress from sub-cortical control in new-borns, to more cortical mechanisms by the age of 2–3 months. New-born infants orient more readily towards stimuli in the temporal visual field, which is thought to input to the sub-cortical visual pathway from the eye to the superior colliculus [29]. By the age of 2 months, infants are more able to orient towards stimuli in the nasal visual field, indicative of cortical control of orienting. Speed of orienting also changes over the first few months; 1-month olds will take longer to orient to peripheral stimuli than 3-month olds, and will also display more directional errors [4].

The second phase, sustained attention, is a period during which the infant is engaged with processing the stimulus. This phase may involve the enhancement of information processing and learning, and can be associated with a characteristic decrease in heart rate [34]. The traditional approach toward the measurement of sustained attention has been the observation of childrens interactions with toys. These interactions are typically videotaped, and subsequently coded by trained coders. The general trend in the development of sustained attention is one of greater sustained attention, and decreased latency to enter periods of sustained attention with increasing age, which may be due to improved motor control [40].

Finally, the third phase, disengagement, involves a process of decreasing attention to the foveated object or location, so that attention can then be oriented elsewhere. This is a necessary component within development, as having completed processing the current object of attention, it allows infants to move on to, and learn about, other environmental stimuli [40].

4. Spatial representation and attention in atypically developing children

The aim of the present study is to examine both spatial representation and sustained attention in toddlers with WS and DS, as well as mental age-matched (MA) and chronological age-matched (CA) healthy controls. Spatial representation will be tested in Experiment 1, using the double-step saccade paradigm employed by Gilmore and Johnson [20,21], to determine whether the visuo-spatial impairments found in adults with WS [9,14,48] have precursors in early childhood, in the form of deficits in spatial frames of reference used to guide visual action. Other work has already suggested the possibility of a dorsal stream deficit in WS [5]. It will also be possible to examine vector summation using this task. If the WS group exhibit more vector summation than other groups, this may indicate a tendency to rely on sub-cortical saccade planning processes. Finally, the double-step saccade task also provides a measure of orienting of attention. Eye movements to the first target can be used to indicate accuracy of orienting to the target location.

Sustained attention will be examined in Experiment 2. Anecdotal reports suggest that attention is a problem area in both clinical groups of interest here. However, there has been little empirical work on attention in WS, and studies of attention in DS have tended mainly to focus on school-age children and adults [33,38,43], rather than the early stages of development (although see [24,32]). Three issues are of interest: the first is to determine whether any differences found between groups on the saccade planning task might be accounted for by impairments of attention. The second issue concerns whether the patterns of attention claimed to be present in adults and older children with WS and DS are present from infancy, or if these problems develop later. Finally, if infants with WS do show deficits in this area, are they qualitatively similar to those exhibited by infants with DS?

5. Experiment 1: saccade planning

It is proposed that toddlers with WS will be impaired on the double-step saccade task, and will make more errors of saccade planning that require body-centred frames of reference, relative to typically developing controls. A secondary prediction of Experiment 1 is that toddlers with WS will differ from typically developing controls in terms of vector summation, which would appear as looks to the central position on the first saccade, and which would indicate delayed maturation of the cortical control mechanism. Finally, it is predicted that orienting of attention, as indicated by accuracy of target localisation on the first eye movement, will be poorer for the toddlers with WS.

5.1. Method

5.1.1. Participants

A total of 64 infants were tested, comprising a group of 13 toddlers with WS (mean CA: 29 months, range: 23–37 months), 19 toddlers with DS (mean CA: 29 months, range: 24–37 months), 17 CA-control toddlers (mean CA: 30 months, range: 23–37 months), and 15 MA-control infants (mean CA: 15 months, range: 12–21 months). MA control infants were matched to the WS and DS toddlers using the Bayley Scales of Infant Development II [6]. The majority of the WS and DS groups failed to obtain a Mental Development Index (MDI) score and were categorised as MDI < 50. Raw scores were therefore used and are reported in Table 1. The Bayley was also used to match the DS and WS participants. The CA group was included to control for length of experience. The MA group was included to control for general level of cognitive development. The DS group controlled for general mental retardation. The children with WS were recruited through the Williams syndrome Foundation, UK and all were positive on the FISH test for the elastin deletion on one copy of chromosome 7. Children with DS were recruited through the Down Syndrome Association, and all were full trisomy-21 and not mosaic. CA- and MA-control participants were recruited through a subject pool of healthy infants and toddlers. Participants were part of a broader study of cognitive and linguistic development in atypical infants and toddlers, now being carried out at the Neurocognitive Development Unit in London. All participants except one of the DS toddlers (eliminated from this visual study because she presented with nystagmus) were reported as having normal or corrected for normal vision.

5.1.2. Procedure

Participants sat in a car seat 50 cm from a 51 cm colour computer monitor controlled by a microcomputer. Those children who would not settle in the car seat were tested on the parent or experimenters lap. The sessions were recorded from a video camera mounted above the monitor, zoomed in to obtain a close-up view of the head and eyes. A time code generator was used to allow for subsequent frame-by-frame coding. Head movements were limited by padding, but the head remained relatively unrestrained in order to maintain cooperation.

Each session started with distractor stimuli that were initiated before the participant was in place, in order to engage the participants interest. This was followed by 18 calibration trials, 48 experimental trials, and 12 break trials. Distractor, fixation, and target stimuli were chosen randomly from a subset of high contrast designs each presented at 4° in width, accompanied by random auditory tones. In each trial, a fixation stimulus appeared in one of four positions on the screen (corresponding to north, south, east, and west). Following a computer key press that terminated the fixation stimulus, two targets appeared on the screen, one following the other, with no period of overlap. Pairs of targets appeared opposite one another, e.g. above right and below right of fixation, and at a distance of 17° from the middle of the fixation point. Eight combinations of fixation-target sequences were presented in pseudo-random order in each pair of blocks of experimental trials. Break trials were presented after each block of four experimental trials, in which targets were shown but durations were longer (300 ms) to reduce the possibility of participants frustration.

Target durations were selected to maximise the number of trials in which participants made sequences of two saccades. Target durations were determined from pilot studies, and were 70 ms for target 1, and 100 ms for target 2. Target 1 was always in one of the two positions diagonally adjacent to the fixation point, with target 2 always positioned opposite target 1, as in Fig. 1A. A 1.5 s response interval was allowed before the start of the next trial. On trials where the participant was not fixating, attempts were made to attract his attention to the correct area of the screen, by calling his name and encouraging him to look at the picture, or by shaking bells in the correct position behind the monitor. Continued failure to fixate resulted in the trial being terminated in order to maintain the childs interest.

5.1.3. Coding

After the testing session, two trained coders (blind to trial type) determined start times and endpoints (one of nine screen locations on a 3×3 grid) of the first and second saccades from PAL format (50 half frames/s) videotapes of the toddlers head and eyes. If the two coders disagreed on either the direction or start time for a saccade, a third coder reviewed the trial independently and rejected the assessment of one or both of the other coders. Videotape measures of this type have been shown to have sufficient accuracy in comparison with electro-oculogram (EOG) data [44]. Reliability between the coders was calculated based on the number of trials where two of the three coders/judge agreed on start times and endpoints of both saccades, as a proportion of trials completed, and is as follows: all groups: 92%, WS: 90%, DS: 91%, CA: 92%, MA: 95%.

Table 1
Mean BSID II raw scores

Group	Mean	S.D.	Range
WS ($n = 13$)	101.43	10.00	83–122
DS ($n = 19$)	100.89	9.96	88–121
CA ($n = 17$)	146.76	9.30	140–165
MA ($n = 15$)	99.44	10.81	84–123

Table 2
Mean trials completed (out of 48), valid trials and eliminations (as a percentage of trials completed) for all groups

	Trials completed	Valid trials (%)	Elim 1% (no valid first look)	Elim 2% (look before T2 offset)	Elim 3% (coder disagreement)
WS ($n = 13$)	45.77 (5.29)[a]	51.15 (15.56)	36.05 (19.25)	3.05 (3.96)	9.76 (7.92)
DS ($n = 19$)	47.74 (1.15)	59.37 (12.92)	23.96 (13.06)	7.47 (9.00)	9.25 (7.92)
CA ($n = 17$)	42.94 (10.05)	58.12 (15.75)	22.45 (11.54)	10.83 (13.60)	8.46 (7.33)
MA ($n = 15$)	45.60 (4.44)	65.33 (13.56)	24.94 (10.40)	4.84 (4.88)	5.08 (5.35)

[a] Standard deviations in parenthesis.

Trials were coded as invalid, and eliminated, on the basis of three criteria: (1) if the toddler was judged not to be looking at fixation during the presentation of the targets, failed to disengage from the fixation, looked away from the display, or the experimenter terminated a trial due to participant fussiness, that trial was eliminated on the basis of no valid first look; (2) if the first saccade began before the offset of the second target, trials were eliminated, in order to ensure that retinal and non-retinal information were separate, and; (3) if start times of saccades did not correspond to within one frame either way, or locations of the endpoints of saccades differed, as determined by two of the three coders/judge, that trial was eliminated.

Our analysis focused on participants sequences of two eye movements in which the first saccade was toward the first target. In these circumstances, it is the endpoint of the second saccade that indicates whether retinocentric or body-centred information controlled the response (Fig. 1B).

5.2. Results

5.2.1. Valid trials and elimination rates

Table 2 shows the trials contributed by each of the groups, and the pattern of elimination of trials.

Separate one-way ANOVAs were carried out on the percentage of valid trials contributed by each of the groups, and on each of the elimination measures. There was no significant difference between groups on percentage of valid trials ($F(3, 60) = 2.243$, n.s.). For the elimination measures, there was a significant difference between groups on Elimination 1 ($F(3, 60) = 2.903$, $P < 0.05$), toddlers with WS scoring higher on this measure than CA toddlers (Tukey's HSD, $P < 0.05$). There were no group differences on Elimination 2 ($F(3, 60) = 2.126$, n.s.), or Elimination 3 ($F(3, 60) = 1.275$, n.s.).

5.2.2. First saccade responses

A summary of first saccade types (as a percentage of all valid first saccades) is presented in Fig. 2. The types of saccade presented are:

Target 1: looks to the position of the first target
Target 2: looks to the position of the second target
Centre: looks to the central (vector averaged) position
Other: looks to any other position on the 3 × 3 grid

Separate one-way ANOVAs were carried out on each of the first saccade response types. There was a significant difference between groups on saccades to target 1 ($F(3, 60) = 7.359$, $P < 0.001$), saccades to the centre ($F(3, 60) = 16.517$, $P < 0.0001$), and saccades to other positions on the 3 × 3 grid ($F(3, 60) = 7.911$, $P < 0.001$). Differences on all these measures were found to be between the

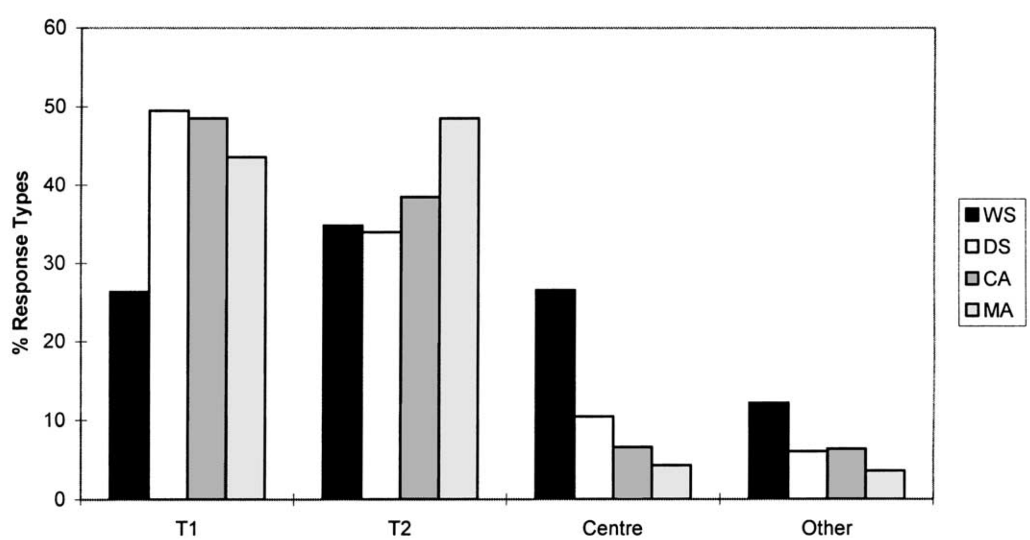

Fig. 2. First saccade responses for all groups (%). T1: target 1, T2: target 2, centre: the vector averaged position, other: any other position on the 3 × 3 grid.

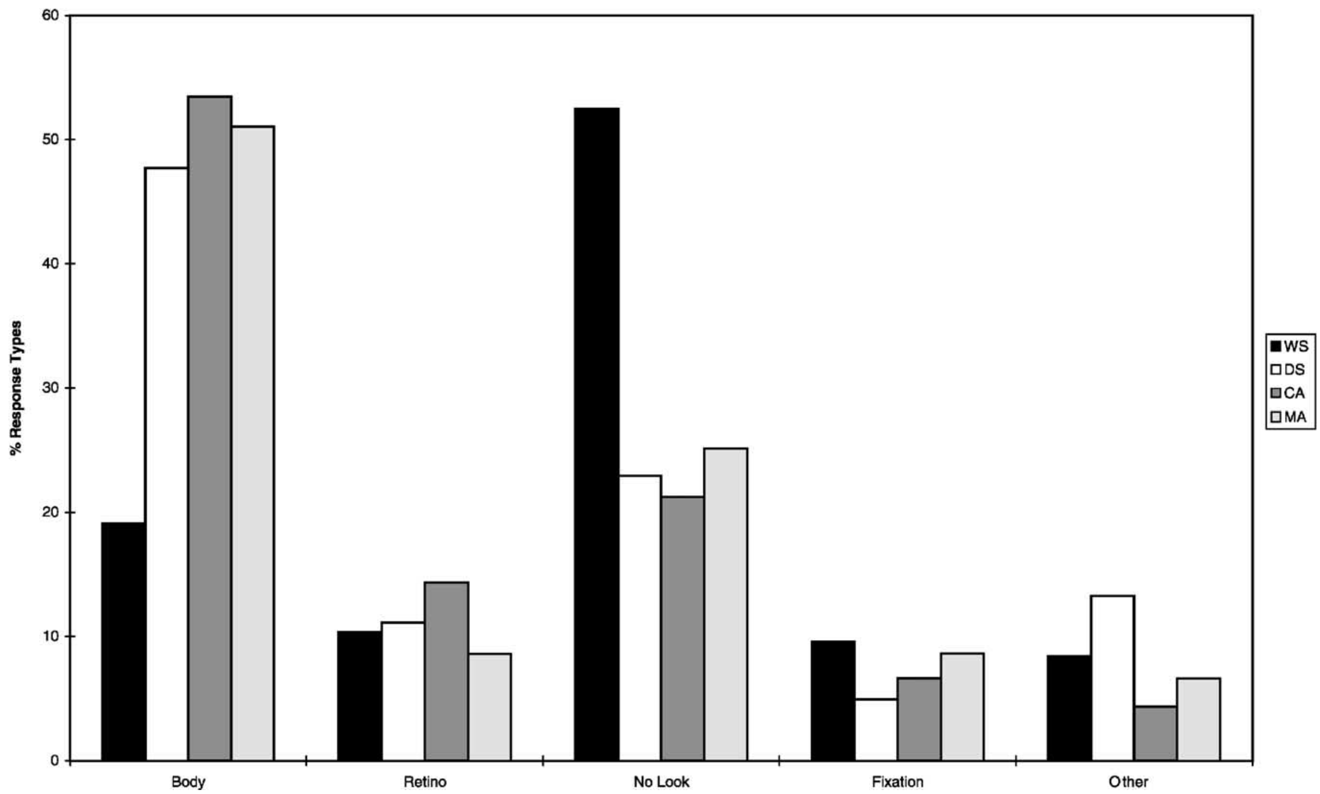

Fig. 3. Second saccade responses for all groups (%). Body: body-centred, retino: retinocentric, no look: no second saccade, fixation: saccades back to the position of the fixation point, other: any other position on the 3×3 grid.

WS group and all other groups (Tukey's HSD, $P < 0.05$), with the WS group scoring lower than other groups on looks to target 1, and higher than other groups on looks to the centre, and other positions. There were no significant group differences on saccades to target 2 ($F(3, 60) = 2.680$, n.s.).

5.2.3. Second saccade responses

Response types made in the second saccade are presented in Fig. 3. Responses presented are those made after a saccade to the first target, and response types reported are:

Body-centred: successful looks to the body-centred position

Retinocentric: looks to the retinocentric position

Fixation: looks back to the position of the fixation point

Other: looks to any other valid position on the 3×3 grid

No look: no second look made

One-way ANOVAs were performed on each of the second saccade measures. Significant differences were found on body-centred looks ($F(3, 60) = 12.55$, $P < 0.001$), and no second looks ($F(3, 60) = 8.48$, $P < 0.001$). Differences on these measures were found between the WS group and all the other groups (Tukey's HSD, $P < 0.05$). Differences at the level of retinocentric looks, looks back to fixation, and other looks were not significant.

5.3. Discussion

Overall, the results from the double-step saccade task show that young children with WS perform differently from those with DS, as well as from the chronological age- and mental age-matched, typically-developing controls. The proportion of looks to the first target by WS toddlers was lower than the other groups, including the DS group. WS toddlers also made significantly more looks to the centre location than all other groups. This supports our hypothesis that they are more frequently using vector summation to plan saccades than any of the other groups. The WS toddlers also failed to make a second look more frequently than control groups, and when they did make a second look, it tended to be less frequently directed towards the body-centred location. This indicates that they may be unable to combine extra-retinal information with retinal information to the same extent as the DS toddlers and normal controls. Finally, WS toddlers also made a higher proportion of looks to other areas on the grid than all other groups.

The pattern of results suggests that the WS toddlers are impaired relative to both DS toddlers and both CA- and MA-matched healthy controls on orienting to a target, and that they rely more than controls on sub-cortical circuits, which results in making frequent responses to the vector averaged centre location. There was no difference between groups on proportion of retinocentric responses, looks back to the fixation point, or looks to other positions. In terms of the overall pattern of second saccades made by groups, the predominant response type for the WS group was failure to make a second saccade, while for all other groups the predominant response was correct looks to the body-centred position.

These results suggest that the visuo-spatial problems reported in older children and adults with WS are present from early childhood. However, it is important to establish that group differences in levels of engagement on the task did not influence outcomes. Specifically, poor sustained attention in the WS group could have contributed to fewer looks being made in the present task.

6. Experiment 2: sustained attention

It is hypothesised that toddlers from the two clinical groups, WS and DS, will be impaired relative to control groups, both on measures of duration and number of periods of sustained attention, which would indicate that the attention problems that are reported to be present in adulthood (e.g. [19,43,52]) already exist from an early age.

6.1. Method

6.1.1. Participants

All participants from Experiment 1 took part in this second experiment.

6.1.2. Procedure

Infants were placed on a childs booster seat attached to a normal chair, and seated at a table. The parent or carer sat on the childs left, and the experimenter sat at an angle, at the corner opposite the child. A box containing the toys was to the left of the experimenter, out of the childs view. A video camera was placed directly facing the child at head level, and all sessions were recorded for subsequent coding.

The test session consisted of three parts; the warm-up, the experimental trials, and a debriefing for the parents. The warm-up consisted of two trials, in which toys were placed on the table in front of the child for 45 s each. Order of presentation of the toys was varied across participants. Parents were advised before the session that they should not talk to the child, and also that the experimenter would not talk to, or make eye contact with the child. At the end of 45 s, the toy was removed, and the next toy introduced. In cases where the child would not give up the toy, the next toy was introduced as a distracter, enabling the experimenter to remove the first toy. Experimental trials were exactly the same as in the warm-up, but different toys were used. During the debriefing, parents were asked whether the child was familiar with any of the experimental toys, and their responses recorded.

Three toys were used during the experimental trials; a corkscrew toy with revolving balls which could screw round the centre part with a suction cup base, a farmyard pop-up toy with four coloured buttons which revealed farm animals when pressed, and a ball shaped toy constructed of wooden rods and elastic which could be squashed and stretched.

6.1.3. Coding

Coding was based on that of Ruff and Lawson [40]. Two coders practised coding periods of sustained attention, using videotapes and training material provided by Holly Ruff (see [39]), until reliability within and between coders was at least 90%. The measures of interest were duration of attention and number of periods of attention. Each coding session would begin with the coder watching a complete session (one period of 45 s with one toy) in real time, to get an overall view of events during that session. Frame-by-frame coding (50 half frames per second) for each session was then performed, noting start and end times of periods of sustained attention in half frames. After coding, any periods of sustained attention that were <2 s in duration, were removed before analysis, as Ruff [39] claims that these are so short that they are not really indicative of sustained attention. One coder, who was blind as to the experimental hypothesis, coded all sessions, plus 15% of sessions a second time. A second coder also coded 15% of sessions selected at random. The intra-rater intraclass correlation was 0.986 ($n = 9$, $P < 0.001$), and the inter-rater intraclass correlation was 0.984 ($n = 9$, $P < 0.001$). These figures are comparable with the inter-observer correlations of >0.90 reported by Ruff and Lawson [40].

6.2. Results

Total duration of sustained attention recorded for each group with all three toys was analysed, to determine whether groups responded differently to individual toys. There was a significant main effect of toy ($F(2, 120) = 22.21$; $P < 0.01$), and a significant main effect of group ($F(3, 60) = 6.29$; $P < 0.05$). The interaction of toy by group was not significant ($F(6, 120) = 1.36$; n.s.), indicating that although the toys did not all elicit the same amount of sustained attention, the pattern was the same across groups. Therefore, responses from all three toys were pooled for further analysis.

6.2.1. Duration of sustained attention

Total duration of periods of sustained attention with all three toys was calculated, and average total duration for all four groups is presented in Table 3. A one-way ANOVA revealed a significant difference between groups ($F(3, 60) = 6.37$; $P < 0.05$), the DS group having significantly shorter total duration of periods of sustained attention than all other groups (Tukey's HSD, $P < 0.05$).

6.2.2. Number of periods of sustained attention

The number of periods of attention demonstrated by each infant was calculated for all toys, and the average number

Table 3
Duration and number of periods of sustained attention for all three toys, in seconds

Group	Total duration		Number of periods	
	Mean	S.D.	Mean	S.D.
WS ($n = 13$)	68.51	29.40	4.69	1.18
DS ($n = 19$)	33.19	23.15	3.32	1.29
CA ($n = 17$)	54.86	19.16	4.59	1.70
MA ($n = 15$)	58.29	24.84	5.40	2.06

of periods of sustained attention for each group is presented in Table 3. A one-way ANOVA revealed a significant difference between groups ($F(3, 60) = 5.11$; $P < 0.05$). This was found to be between the lowest number of periods of sustained attention for the DS group, and the highest for the MA group (Tukey's HSD, $P < 0.05$). There was no significant difference between the DS and WS groups.

6.3. Discussion

The results from Experiment 2 indicate that the DS group appear to have a deficit of sustained attention, in that they exhibit fewer periods of sustained attention than the younger MA group, and less total duration of sustained attention than all other groups. These results are in line with many of the studies that claim to find attention deficits in children and adults with DS (e.g. [19,43]), and are indicative of a stable deficit from infancy through to adulthood in this syndrome.

Could task engagement explain our results with the DS toddlers? Wishart and Duffy [50] suggest that factors such as task engagement and motivation may be contributory in the task performance of DS children. They found that task failure was often due to DS childrens refusal to participate, thereby failing the task by default. Children with DS, they argue, will often engage in elaborate avoidance behaviours. However, other studies claim to find no difference between typically developing infants and DS infants in terms of task persistence (e.g. [41,47]). In addition, Hasan and Messer [27] found no indication of failure to engage in six children with DS, when tested on Uzgiris and Hunt object concept and means-ends scales. Furthermore, Hasan and Messer [27] claims to find stable performance with these DS children when tested repeatedly at monthly intervals. Thus, task engagement and motivation are unlikely to explain the DS failure to engage in sustained attention.

One surprising result from this experiment is that toddlers with WS perform as well as typically developing controls with respect to sustained attention. As rates of ADHD are reportedly high in WS [18] it was predicted that toddlers with WS would perform poorly on measures of sustained attention. While ADHD is not clinically diagnosed until a child is 3 years of age or older, the task revealed a stable deficit in sustained attention in the DS group. But this is not the case for WS, despite the toddlers being matched on both CA and MA to the toddlers with DS. The fact that the toddlers with WS do not show the same deficits of attention as reported in older children and adults suggests that the attention deficit in Williams syndrome may not emerge until later, due to interactions with spatial and other cognitive deficits, and the way in which such deficits impact over developmental time on learning. However, it should be noted that the issue of attention levels in adults with WS needs empirical clarification.

7. General discussion

The results of our experiments suggest that the visuo-spatial problems found in adults with WS do have precursors in early childhood. Likewise, poor sustained attention seems to be a stable deficit over developmental time in DS.

Experiment 1 revealed that toddlers with WS perform very differently from all the other groups on each of the measures from the double-step saccade paradigm. Thus, the ability to orient accurately to target locations, and the use of body-centred spatial frames of reference to plan saccades, is impaired in this population. In contrast, the DS group demonstrated no difficulties with any of the measures from the double-step saccade task. WS performance on these measures may thus be an early precursor of visuo-spatial deficits found in older children and adults with WS.

Experiment 2 examined sustained attention, and revealed that unlike the DS group, the WS group were not impaired on this measure. Thus, the poor sustained attention reported in adults with DS appears to be present from an early age, and therefore represents a stable deficit within this population. Deficits of attention reported in adults with WS were not found in the WS toddler group, which may suggest that such deficits do not emerge until later in the WS group, but as mentioned previously, this is an area which requires further investigation.

The findings from Experiment 2 were used to determine whether differences in the ability to sustain attention could have contributed to the results from Experiment 1. As the WS toddlers did not show poorer sustained attention, their performance on the double-step saccade task is not likely to have been affected by differences in level of engagement. If anything, the DS group should have performed poorly in Experiment 1, since Experiment 2 demonstrated lower levels of sustained attention in this group. Yet, the DS toddlers performed well in the first experiment.

The comparison of Experiments 1 and 2 would therefore seem to suggest that poor attention levels do not contribute to the impaired performance on the double-step task by the WS toddlers. However, it should be noted that the two tasks were quite different, in that the first task required passive viewing of the stimuli, while the second was more active in that the child was required to play with the toys. Thus, the demands of the tasks were different. The difference in trial times between the two tasks also meant that attention was required for very short (Experiment 1), or relatively long (Experiment 2), durations.

An alternative interpretation of the findings of these experiments concerns the issue of attention disengagement. The results from Experiment 1 could be accounted for, in part at least, by an inability in the WS group to disengage from the stimuli. If WS toddlers had an attention disengagement problem, this would result in fewer looks being made after fixation by this group. When second saccade performance is considered, the predominant response by the WS group was failure to make a second saccade. Again, this could be accounted for by a failure to disengage after the first saccade was made. Although attention disengagement was not directly tested, the results from Experiment 2 add some initial support for this claim. Periods of sustained attention were longest in the WS group, although only significantly longer than the DS group. If the WS participants were less able to disengage from the toys in Experiment 2, then we might expect longer periods of attention. Deficits of attention disengagement have been demonstrated in several clinical groups, including children with ADHD [51], children with developmental coordination disorder [49], and adults with Alzheimer's disease [16]. Thus, problems with attention disengagement are not uncommon in clinical populations, and this is an area that requires further direct investigation in WS.

The specific impairments identified in Williams syndrome differ substantially from those found in another genetic disorder, Down syndrome, with similar overall cognitive level of functioning at the same chronological age. While the underlying cause of the impaired performance displayed by the toddlers with WS and the role of sustained attention and attention disengagement require further investigation, there is no doubt that these young children experience significant problems early in life in visuo-spatial representation. This disadvantage is likely in turn to impact on their development in other areas of cognition. Our experiments highlight how important it is to examine genetic disorders early in development [31,36], rather than solely in middle childhood and adulthood.

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References

[1] Andersen RA, Snyder LH, Li CS, Stricanne B. Coordinate transformations in the representation of spatial information. Current Opinion in Neurobiology 1993;3:171–6.
[2] Aslin RN, Shea SL. The amplitude and angle of saccades to double-step target displacements. Vision Research 1987;27:1925–42.
[3] Atkinson J. The where and what or who and how of visual development. In Simion F, Butterworth G, editors. The development of sensory, motor and cognitive capacities in early infancy: from perception to cognition. Hove (England): Psychology Press/Erlbaum, 1998:3–24.
[4] Atkinson J, Hood BM, Wattam-Bell J, Braddick OJ. Changes in infants ability to switch attention in the first three months of life. Perception 1992;21:643–53.
[5] Atkinson J, King J, Braddick O, Nokes L, Anker S, Braddick F. A specific deficit of dorsal stream function in Williams syndrome. Neuroreport 1997;8:1919–22.
[6] Bayley N. The Bayley scales of infant development. second ed. San Antonio (TX): The Psychological Corporation; 1993.
[7] Becker W, Jürgens R. An analysis of the saccadic system by means of double step stimuli. Vision Research 1979;19:967–83.
[8] Bellugi U, Bihrle A, Jernigan T, Trauner D, Doherty S. Neuropsychological, neurological, and neuroanatomical profile of Williams syndrome. American Journal of Medical Genetics 1990;6:115–25.
[9] Bellugi U, Wang PP, Jernigan TL. Williams syndrome: an unusual neuropsychological profile. In: Broman S, Grafman S, editors. Cognitive deficits in developmental disorders: implications for brain functions. Hillsdale (NJ): Lawrence Erlbaum Associates; 1993.
[10] Bremner JG. Egocentric versus allocentric spatial coding in nine-month-old infants: Factors influencing the choice of code. Developmental Psychology 1978;14:346–55.
[11] Bremner JG. Spatial errors made by infants: inadequate spatial cues or evidence of egocentrism? British Journal of Psychology 1978;69:77–84.
[12] Chen LL, Nakamura K. Head-centred representation and spatial memory in rat posterior parietal cortex. Psychobiology 1998;26:119–27.
[13] Dolk H, De Wals P, Gillerot Y, Lechat MF, Aym S, Beckers R, Bianchi F, Borlee I, Calabro et al., The prevalence at birth of Down syndrome in 19 regions of Europe, 1980–86. In: Fraser WI, editor. Key issues in mental retardation research. London: Routledge; 1990.
[14] Donnai D, Karmiloff-Smith A. Williams syndrome: from genotype through to the cognitive phenotype. American Journal of Medical Genetics: Seminars in Medical Genetics 2000;97:164–71.
[15] Duhamel JR, Colby CL, Golberg ME. The updating of the representation of visual space in parietal cortex by intended eye movements. Science 1992;255:90–2.
[16] Filoteo JV, Delis DC, Roman MJ, Demadura TL. Visual attention and perception in patients with Huntingtons disease: comparisons with other subcortical and cortical dementias. Journal of Clinical and Experimental Neuropsychology 1996;17:654–67.
[17] Findlay JM. Global visual processing for saccadic eye movements. Vision Research 1982;22:1033–45.
[18] Finegan J, Sitarenios G, Smith M, Meschino W. Attention deficit hyperactivity disorder in children with WS: preliminary findings. In: Proceedings of the Williams Syndrome Association Annual Professional Conference, San Diego, 1994.
[19] Fisher L. Attention deficit in brain damaged children. American Journal of Mental Deficiency 1970;74:502–8.
[20] Gilmore RO, Johnson MH. Body-centred representations for visually-guided action emerge during early infancy. Cognition 1997;65:B1–9.
[21] Gilmore RO, Johnson MH. Egocentric action in early infancy: spatial frames of reference for saccades. Psychological Science 1997;8:224–30.
[22] Goldberg ME, Colby CL. Oculomotor control and spatial processing. Current Opinion in Neurobiology 1992;2:198–202.

[23] Goodale MA, Milner AD. Separate visual pathways for perception and action. Trends in Neurosciences 1992;15:20–5.
[24] Green JM, Dennis J, Bennets LA. Attention disorder in a group of young Down syndrome children. Journal of Mental Deficiency Research 1989;33:105–22.
[25] Groll SL, Ross LE. Saccadic eye movements of children and adults to double-step stimuli. Developmental Psychology 1982;18:108–23.
[26] Hallett PE, Lightstone AD. Saccadic eye movements towards stimuli triggered by prior saccades. Vision Research 1976;16:99–106.
[27] Hasan PJ, Messer DJ. Stability or instability in early cognitive abilities in children with Downs syndrome. British Journal of Developmental Disabilities 1997;43:93–107.
[28] Honda H. Perceptual localization of visual stimuli flashed during saccades. Perception and Psychophysics 1989;45:162–74.
[29] Johnson MH. Cortical maturation and the development of visual attention in early infancy. Journal of Cognitive Neuroscience 1990;2:81–95.
[30] Johnson MH, Gilmore RO, Tucker LA, Minister SL. Vector summation in young infants. Brain and Cognition 1996;32:237–43.
[31] Karmiloff-Smith A. Development itself is the key to understanding developmental disorders. Trends in Cognitive Sciences 1998;2:389–98.
[32] Krakow JB, Kopp CB. Sustained attention in young Down syndrome children. Topics in Early Childhood Special Education 1982;2:32–42.
[33] Landry SH, Chapieski ML. Joint attention and infant toy exploration: effects of Down syndrome and prematurity. Child Development 1989;60:103–18.
[34] Lansink JM, Richards JE. Heart rate and behavioural measures of attention in 6-, 9-, and 12-month old infants during object exploration. Child Development 1997;68:610–20.
[35] Mays LE, Sparks DL. Saccades are spatially, not retinocentrically coded. Science 1980;208:1163–5.
[36] Paterson SJ, Brown JH, Gsodl MK, Johnson MH, Karmiloff-Smith A. Cognitive modularity and genetic disorders. Science 1999;286: 2355–7.
[37] Posner MI, Peterson SE. The attention system of the human brain. Annual Review of Neuroscience 1990;13:25–42.
[38] Randolph B, Burack JA. Visual filtering and covert orienting in persons with Down syndrome. International Journal of Behavioral Development 2000;24:167–72.
[39] Ruff HA. Individual differences in sustained attention during infancy. In: Colombo J, Fagan JW, editors. Individual differences in infancy: reliability, stability, and prediction. Hillsdale (NJ): Lawrence Erlbaum Associates; 1990. p. 247–70.
[40] Ruff HA, Lawson KR. Assessment of infants attention during play with objects. In: Schaefer CE, Critlin K, Sandgrund A, editors. Play diagnosis and assessment. New York: Wiley, 1991:115–29.
[41] Ruskin EM, Kasari C, Mundy P, Sigman M. Attention to people and toys during social and object mastery in children with Down syndrome. American Journal on Mental Retardation 1994;99:103–11.
[42] Stein JF. The representation of egocentric space in the posterior parietal cortex. Behavioral and Brain Sciences 1992;15:691–700.
[43] Tomporowski PD. Sustained attention in mentally retarded persons. In: Fraser WI, editor. Proceedings of the Eighth Congress of the International Association for the Scientific Study of Mental Deficiency on Key Issues in Mental Retardation Research. London: Routledge, 1990.
[44] Tucker LA, Csibra G, Johnson MH, The accuracy of videotape analysis of saccadic reaction time as compared to EOG measures in 6-month-old infants. In: Proceedings of the Xth International Conference on Infant Studies 19. Providence: RI, 1996. p.476.
[45] Udwin O, Yule W, Martin N. Cognitive abilities and behavioural characteristics of children with idiopathic infantile hypercalcaemia. Journal of Child Psychology and Psychiatry and Allied Disciplines 1987;28:297–309.
[46] Ungerleider LG, Mishkin M. Two cortical visual systems. In: Ingle DJ, Goodale MA, Mansfield RJW, editors. Analysis of visual behavior. Cambridge (MA): MIT Press, 1982.
[47] Vietze P, McCarthy M, McQuiston S, MacTurk R, Yarrow L. Attention and exploratory behavior in infants with Down syndrome. In: Field T, Sostek A, editors. Infants born at risk: perceptual and physical processes. New York: Grune and Stratton, 1983:251–68.
[48] Wang PP, Doherty S, Rourke SB, Bellugi U. Unique profile of visuo-perceptual skills in a genetic syndrome. Brain and Cognition 1995;29:54–65.
[49] Wilson PH, Maruff P, McKenzie BE. Covert orienting of visuospatial attention in children with developmental coordination disorder. Developmental Medicine and Child Neurology 1997;39:736–45.
[50] Wishart JG, Duffy L. Instability of performance on cognitive tests in infants and young children with Downs Syndrome. British Journal of Educational Psychology 1990;60:10–22.
[51] Wood C, Maruff P, Levy F, Farrow M, Hay D. Covert orienting of visual spatial attention in attention deficit hyperactivity disorder: does comorbidity make a difference? Archives of Clinical Neuropsychology 1999;14:179–89.
[52] http://www.williams-syndrome.org.uk/about_ws/diagnos.htm.