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Perceiving and acting in depth in Williams syndrome and typical development

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Abstract

Individuals with the neurodevelopmental disorder Williams syndrome (WS) often report difficulty processing and acting in depth, such as crossing roads or reaching for objects; however little research attention has been directed at understanding depth perception and action in depth in WS and whether deficits in depth perception have an ocular or perceptual root in this group. This study assessed the extent and relationship of deficits in stereopsis (binocular, three dimensional vision) and actions performed in depth in WS, as well as in typically developing participants (TD) matched for non-verbal ability. Stereoacuity was age-appropriate in the TD group but at the level of a TD three year old in WS; one third of the WS group did not show evidence of stereopsis. When monocularly acting in depth there was no difference between the WS and TD groups. When binocularly acting in depth the WS group that did not exhibit stereopsis were significantly poorer than the TD group and the WS group that exhibited stereopsis. When assessing the relationship between stereoacuity and action in depth, stereoacuity negatively correlated with binocular action in depth for the WS group with stereopsis, but not the TD group. Therefore, no deficits in monocular depth perception in WS were evidenced, yet significant deficits are exhibited in binocular depth perception and action. Importantly action in depth under binocular viewing may be a useful gross screening measure for stereodeficits in WS. Remediation of depth perception deficits in WS could train further understanding of monocular cues to compensate for poor stereopsis.

Perceiving and acting in depth in Williams syndrome and typical development

1. Introduction

We are continually processing and responding to cues about depth; cues can be extracted from a visual scene using one eye (monocular vision) or both eyes (binocular or stereoscopic vision), to determine the relative depth of objects. Retinal disparity allows for construction of three-dimensional representations of space; two versions of the same scene are generated (one from each eye) and features from each view are matched by fusion of these two images (Menz & Freeman, 2003); the ability to achieve this is refined from infancy up to teenage years and into adulthood (Giaschi, Narasimhan, Solski, Harrison & Wilcox, 2013). Beyond the retina, processing of depth information has been proposed to occur in two distinct pathways in the visual system (Mishkin, Ungerleider & Macko, 1983). The dorsal ("where") pathway is responsible for processing of spatial information about objects and is associated with binocular vision. Whereas the ventral ("what") pathway is used for object recognition derived from the object's appearance and physical properties, such as colour and is used in monocular depth cue processing (Fischmeister & Bauer, 2006). Both pathways become distinct at V1 in the primary visual cortex, with the dorsal stream travelling to the posterior parietal lobe and the ventral stream to the temporal lobe.

Deficits in dorsal stream functioning (behavioural and neuroanatomical) and reports of poor depth perception have been described in Williams syndrome (WS; Atkinson et al., 1997; Eckert et al., 2005). WS is a rare genetic disorder with an estimated incidence in live births of one in 20,000 (Morris et al., 1988). This disorder results from a deletion of approximately 28 contiguous genes due to a hemizygous microdeletion of 1.6Mb on chromosome 7q11.23 (Tassabehji, 2003). WS is characterised by mild to moderate learning difficulties and an unusual cognitive profile that is typified by a disparity between relatively strong linguistic ability and poor visuospatial ability (Mervis & John, 2008). Binocular depth

perception in WS may be affected by the high prevalence of ocular deficits that are known to limit binocular vision such as strabismus (in approximately 50% of individuals with WS), low visual acuity, and amblyopia (Atkinson et al., 2001). Relatively little research attention has been directed at understanding depth perception in WS but evidence suggests that stereopsis (binocular depth perception) is poor, with relatively better monocular depth perception (Atkinson et al., 1997; Van der Geest et al., 2005); this suggests that individuals with WS display deficits in dorsal but not ventral stream processing of depth information. This is in line with the dorsal stream deficit hypothesis (Atkinson et al., 1997). Indeed there are widespread structural atypicalities and reduced grey matter volumes throughout the dorsal visual stream in WS (Eckert et al., 2005).

To date only one behavioural study of depth perception using monocular cues in WS has been conducted (Van der Geest et al., 2005). Monocular vision appeared to be good in the WS group; 86% of participants could correctly judge which of two cues was larger in a computer-generated textured room. Conversely, stereopsis was poor in the WS group (49% failed the Titmus test [Stereo Optical Co.]) and participants frequently overshot when moving their hand to a target (supporting Atkinson et al., 1997). However there were no significant correlations in performance between the reaching task, stereopsis and monocular depth perception. At first glance, these results appear to suggest that monocular depth perception is typical in WS, with relatively poor binocular vision compared to a TD and a atypically developing (unknown etiology) group. However, numerous monocular depth cues were presented simultaneously, reducing task sensitivity. Furthermore, evidence from an unpublished questionnaire study from our lab (N = 23, mean CA = 19 years 1 month, SD = 10 years 4 months), suggests that individuals with WS have difficulty perceiving isolated monocular depth cues. Respondents reported significant difficulties comprehending monocular depth cues (assessed using photographs and text explanations), using relative size

(68% of participants), relative height (57% of participants) and occlusion (50% of participants). In addition respondents reported difficulty acting in depth (such as crossing roads) and using of spatial language correctly (consistent with Landau & Hoffman, 2005). Deficits in depth perception have a significant impact on daily functioning in WS, therefore it is important to quantify and understand low-level deficits in depth perception.

The current study assessed stereopsis and action performed in depth under monocular and binocular viewing conditions in WS and typically developing (TD) non-verbal ability matched control participants. If ocular deficits are the root of poor depth perception in WS then participants should perform more poorly on binocular assessment, but inline with the TD group on monocular tasks. TD individuals that have poor binocular vision (such as strabismus), can still function well by relying on monocular cues (Helveston, 2010; Henson & Williams, 1980; Von Noorden & Campos, 1996). If a dorsal stream deficit is the root of atypical depth perception in WS, deficits should be seen when acting in depth and perceiving depth binocularly, with little difference between performing actions under monocular or binocular viewing conditions. However, based on the trends reported from our questionnaire, we predict deficits in processing of monocular and binocular depth cues for both perception and action. This would suggest an additional general perceptual deficit in implicit understanding of monocular cues to depth, which necessarily extends to action.

2. Method

2.1. Participants

This study was given ethical approval to proceed by the Ethics Committee of the Institute of Education, University of London. Written and verbal consent was provided by all participants and their caregivers. All data were collected by the lead author.

Eighteen participants with WS were recruited through the Williams Syndrome Foundation UK (eight male, 10 female). All participants had previously been diagnosed with WS by a clinician and a positive Fluorescence In Situ Hybridisation (FISH) test to ensure deletion of the elastin gene, observed in 95% of individuals with WS (de Souza, Moretti–Ferreira & Rugolo, 2007).

Eighteen TD control participants were recruited through a primary school in Berkshire, England (ten male, 8 female). Participants were individually matched on non-verbal ability using Raven's Coloured Progressive Matrices scores (RCPM; Raven, 1993). RCPM is a standardised measure of non-verbal ability and has previously been used successfully as a matching measure for visuospatial tasks in developmental disorder groups (Davies, Bishop, Manstead & Tantum, 1994; Van Herwegen, Farran & Annaz, 2011). Matching was successful as there was no significant difference in RCPM scores between groups, $t(34) = .48$, $p = .63$. There were no differences in the number of male and female participants between groups, $p > .05$.

Table 1 about here

Table 1

WS and TD Participants' Chronological Age and RCPM Scores.

2.2. Materials and Procedure

Two tasks were used to determine stereopsis and the ability to act in depth respectively. For all participants the order of tasks was randomised. Any participants that required corrected to normal vision were permitted to wear corrective lenses at all times during each task and were not excluded from the study.

2.2.1. Frisby Stereotest (Near). The Frisby Stereotest (Near) (FSN; Frisby, Davis, & McMorrow, 1996) was used to assess stereopsis. Stereoacuity can be measured between 600arcsec (expected in TD six month olds) up to just 5arcsec (expected in TD children aged beyond six years old). The task measures stereoacuity in terms of the smallest visual angle that participants can reliably detect. The FSN has good test-retest reliability (e.g. Adams, Leske, Hatt & Holmes, 2009). This task was selected as true depth in this test is not derived from stereograms or illusory depth cues. Additionally participants are not required to wear polarised glasses or coloured lenses as in the Titmus Stereo test (Stereo Optical Co.) or TNO (Alfred Poll Inc.). Therefore the task was administered under normal vision, making the task well-tolerated by the WS and TD groups.

The FSN requires participants to find a "magic ball" in one of four quadrants printed onto an acrylic plate. The circular form appears to recede from the surrounding foreground pattern. This effect is achieved by the circular area being printed onto the back of the acrylic plate, relative to the foreground in the target quadrant and the three distracter quadrants which are printed on the front face of the plate. The task provides three levels of difficulty, achieved by plates of various thickness (6mm, 3mm and 1.5mm which is the more "difficult" trial). Using the thickest plate participants were shown how to locate the "magic ball", by movingthe plate laterally to induce motion parallax to highlight the target area in participants that did not spontaneously locate it. For test trials the plates were held at varying distances away from the participant until the participant can no longer reliably detect the "magic ball" on each plate. The plates were randomly rotated for each trial to ensure that the target quadrant was not constant. Stereoacuity (arcsec) was calculated for each participant using their interpupillary distance (mm) and the greatest distance for which the task could be reliability completed (mm) recorded by the experimenter.

2.2.2. Lang Two Pencil Test. The Lang Two Pencil Test (LTPT; Lang, 1983) was used as a measure of participants' ability to act in depth using monocular and binocular depth cues. The task used two eraser-tipped pencils; one of which was held by the experimenter as a target at the participant's eye level and arm's length away and the other pencil used by the participant to touch just the tip of the eraser of their pencil to the target as quickly as possible. The task was demonstrated to the participants by the experimenter, who held both pencils to demonstrate how to correctly touch just the tip of the target. A participant passed a trial only when the end of their pencil clearly touched just the tip of the target pencil; touching of the side of the target eraser or readjustment to touch the end of the eraser was classed as a failed trial. Target locations were random points along the plane of the participant's arm-length, with the target in vertical and horizontal orientations. The horizontal LTPT is a variant of the original vertical LTPT and has a high sensitivity and negative predictive value when screening binocularity (Nongpiur & Sharma, 2010). The order of orientation of the pencil (horizontal or vertical) and the viewing condition (binocular or monocular) was randomised for each participant. Participants completed five trials per viewing condition in each orientation (20 trials in total). The experimenter recorded the accuracy of each trial (correct or incorrect).

3. Results

3.1. Frisby Stereotest (Near)

When completing the FSN, six participants with WS failed to detect the target area, suggesting a marked deficit in stereovision (binocular perception); no stereo deficits were observed in the TD group. The WS group (N=12) that passed the FSN exhibited stereoacuity at a level expected for a TD three year old (M = 67.83arcsec, SD = 83.23 arcsec); the TD group's stereoacuity was within the normal bounds for a TD six year old (M = 5.49 arcsec,

SD = 7.26 arcsec). The TD group demonstrated significantly greater stereovision than the WS group, $t(28) = 3.19$, $p = .004$. Stereoacuity was not significantly correlated with chronological age in either group, (WS: $r(N = 12) = -.19$, $p = .55$; TD: $r(N = 18) = -.34$, $p = .17$). In the TD group this is likely due to the small age-range of the group, but in the WS group this evidences lack of a developmental progression in stereoacuity (Giaschi et al., 2013).

3.2. Lang Two Pencil Test

Performance was significantly different from floor and ceiling in all conditions in both groups, $p < .05$. Performance accuracy was not related to chronological age in any viewing condition or orientation, in either group ($p > .05$ for all).

Given that a sub-group of participants in the WS group failed the FSN, the LTPT was analysed using two WS groups; these were WS that pass the FSN (N=12), and WS that fail the FSN (N=6). This was in order to differentiate individuals with WS with evidence of stereopsis from those without, as this was likely to impact the ability to act in depth.

A group (WS that pass the FSN, WS that fail the FSN, TD) by orientation (horizontal, vertical) by viewing type (monocular, binocular) analysis of variance (ANOVA) was performed on the percentage of correctly completed trials. There was no overall significant difference across groups and no effect of orientation of the pencils, or an interaction of these factors ($p > .05$ for all). Binocularly viewed trials ($M = 74.26\%$, $SE = 3.43\%$) led to significantly greater accuracy than monocularly completed trials ($M = 19.26\%$, $SE = 2.69\%$), $F(1, 33) = 165.65$, $p < .001$, $\eta_p^2 = .83$.

There was a significant interaction between the viewing type and group, $F(1, 34) = 7.07$, $p = .01$, $\eta_p^2 = .17$ (see Figure 1). This resulted from significant differences across groups in the binocular but not monocular viewing condition (monocular: $F(2, 35) = 1.89$, $p = .17$; binocular: $F(2, 35) = 3.89$, $p = .03$). On binocular trials, the WS group that failed the

FSN were significantly less accurate than the TD group (p = .009), and marginally less accurate than the WS group that passed the FSN (p = .06), with no difference between the TD and WS that pass the FSN group (p > .05). All other interactions were non-significant, p > .05 for all.

Figure 1 about here

Figure 1. The interaction of viewing type on accuracy of responses in the LTPT in WS and TD groups.

3.3. The relationship between stereovision and the ability to act in depth

In order to understand the relationship between stereovision (as measured by the FSN) and the ability to act in depth (using responses from each condition of the LTPT), correlation analysis of these two variables was conducted for participants that passed the FSN in the WS and TD groups. For the TD group there were no significant correlations between FSN scores and accuracy in any of the LTPT conditions (Vertical Binocular: r(N = 18) = -.05, p = .40; Vertical Monocular: r(N = 18) = .03, p = .91; Horizontal Binocular: r(N = 18) = -.12, p = .66; Horizontal Monocular: r(N = 18) = -.05, p = .84). In the WS group there was a significant negative correlation between stereoacuity and performance in the binocularly viewed, horizontally orientated LTPT (r(N = 12) = -.59, p = .05); thus greater stereoacuity (small arcsec value) was associated with a greater ability to act in depth. In all other LTPT conditions there was no relationship between stereovision and LTPT performance in the WS group, p > .05(Vertical Binocular: r(N = 12) = -.27, p = .40; Vertical Monocular: r(N = 12) = -.41, p = .18; Horizontal Monocular: r(N = 12) = -.34, p = .28).

4. Discussion

This study aimed to understand the nature of functional depth perception in WS; there is a dearth of research in this area despite frequent reports of atypical depth processing from parents and individuals with WS (such as the unpublished questionnaire from our lab discussed in the Introduction section). Using two simple tests of monocular and binocular depth perception and action we have highlighted atypical processing of depth information in WS when compared to non-verbal ability matched control participants.

One third of participants with WS demonstrated deficits in stereoacuity that prevented completion of the FSN, i.e. stereoacuity was poorer than that of a TD infant at six months. Even for the WS group that could complete the FSN, stereoacuity was at the level expected for a TD three year old despite an average chronological age of 26 years 11 months. There was no significant correlation of stereoacuity with age. Conversely the TD group's stereoacuity was age-appropriate and no TD participant failed to complete the task. Therefore the WS group showed a marked deficit in stereoacuity, as functional age was well below that of chronological age and stereoacuity did not improve with development. This is clear evidence of a deficit in the ability to extract information about depth, which is consistent with reports of functional deficits in depth in WS (Withers, 1996). However a limitation of this study is that we used low-level tests of stereopsis. Future research would benefit from studying a broader range of monocular and binocular depth cues, including assessment of participants' ability to use depth cues to act in depth, such as when walking or judging the speed of cars when crossing a road.

The ability to perform actions in depth was explicitly examined using the LTPT. Both WS and TD groups were more accurate when completing trials under binocular viewing, relative to monocular viewing; representing a typical pattern of performance. This underlines that biomechanical constraints in arm movements are unlikely to be the root of any difficulties with acting in depth. The WS group who failed the FSN showed less of a benefit

of completing the task under binocular conditions relative to the TD group and the WS group that passed the FSN. This demonstrates that for some individuals with WS there are marked deficits in binocular depth perception, and that these pervade into actions performed in depth. The WS group that failed the FSN may make use of adaptive strategies to compensate for visual deficits which differ from the strategies used by the WS group that passed the FSN. Individuals with WS with strabismus attempt to circumvent amblyopia by ignoring the image from one eye (Semel & Rosner, 2003) and thus relying on monocular cues to depth. A similar approach is adopted by TD children with stereodeficits (Von Noorden & Campos, 1996). Interestingly, in individuals with WS that passed the FSN there was a negative correlation between stereoacuity and accuracy on binocular, horizontally orientated LTPT trials. Thus there appears to be a particular sensitivity to problems with depth perceived binocularly in WS, supporting the assertions made in TD performance that the horizontal test is more sensitive to deficits in stereopsis than its vertical counterpart (Nongpiur & Sharma, 2010). Accordingly there is likely to be an association of dorsal stream deficits in poor stereopsis in WS, with individual differences in the degree of the deficit (i.e. individuals with WS that pass the FSN and those that fail the FSN). Therefore performing the binocular horizontal LTPT may be a quick and inexpensive stereopsis screening tool that would require little training to administer to identify individuals with WS with poor stereopsis.

Future research to remediate depth perception deficits in WS should be mindful of the deficits or absence (in one third of this sample) of stereopsis in WS. Individuals with WS may be able to improve depth perception with the aid of training to understand monocular depth cues. It is a likely that, if spontaneous compensation has been implemented by individuals, this strategy may be utilised in some individuals with WS with inherent poor stereopsis. Participants that may benefit from this type of intervention could be identified using the horizontal LTPT.

Acknowledgements

This research was supported by Autour des Williams, France. The authors would like to thank the members of the Williams Syndrome Foundation, UK, for their participation in this study and the staff and students at Willow Bank Infant School. We also wish to thank Anna Horwood for useful discussions at the design stage of this study.

Table 1: WS and TD Participants' Chronological Age and RCPM Scores.

	Williams Syndrome (*n*=18)		Typically Developing (*n*=18)	
	Mean(*SD*)	Range	Mean(*SD*)	Range
CA (years; months)	26;11 (11;10)	7;10-47;03	6;02 (0;08)	4;07-7;02
RCPM Score (max.=36)	18.44 (4.49)	12-27	17.67 (5.19)	11-27

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FIGURE CAPTION

Figure 1. The interaction of viewing type on accuracy of responses in the LTPT in WS and TD groups.

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