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Perceiving and acting in depth in Williams syndrome and typical development

Kerry D. Hudson *, Emily K. Farran

Department of Psychology and Human Development, Institute of Education, UK

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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.

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 and 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

* Corresponding author at: Department of Psychology and Human Development, Institute of Education, University of London, 24 Woburn Square, London WC1H 0AA, UK. Tel.: +44 020 7612 6013; fax: +44 020 7612 6304.
E-mail addresses: k.hudson@ioe.ac.uk, kerrydhudson@gmail.com (K.D. Hudson).

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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, Demsey, Leonard, Dilts, & Blackburn, 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 aetiology) 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 (8 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 (10 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).