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Global dot integration in typically developing children and in Williams Syndrome

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ABSTRACT

Williams Syndrome (WS) is a neurodevelopmental disorder that results in deficits in visuospatial perception and cognition. The dorsal stream vulnerability hypothesis in WS predicts that visual motion processes are more susceptible to damage than visual form processes. We asked WS participants and typically developing children to detect the global structure Glass patterns, under "static" and "dynamic" conditions in order to evaluate this hypothesis. Sequentially presented Glass patterns are coined as dynamic because they induce illusory motion, which is modeled after the interaction between orientation (form) and direction (motion) mechanisms. If the dorsal stream vulnerability holds in WS participants, then they should process real and illusory motion atypically. However, results are consistent with the idea that form and motion integration mechanisms are functionally delayed or attenuated in WS. Form coherence thresholds for both static and dynamic Glass patterns in WS were similar to those of 4–5 year old children, younger than what is predicted by mental age. Dynamic presentation of Glass patterns *improved* thresholds to the same degree as typical participants. Motion coherence thresholds in WS were similar to those of mental age matches. These data pose constraints on the dorsal vulnerability hypothesis, and refine our understanding of the relationship between form and motion processing in development.

1. Introduction

Associated with a microdeletion of about 20 genes on chromosomal region 7q11.23 (Lenhoff, Wang, Greenberg, & Bellugi, 1997), Williams Syndrome (WS) is a genetic disorder that occurs in 1 out of 7500 (Stromme, Bjornstad, & Ramstad, 2002) to 20,000 (Lenhoff et al., 1997) live births. While WS causes mild to moderate mental retardation (mean IQ of 60), its cognitive profile is uniquely uneven – with a relative strength in language and relative deficit in visuospatial abilities. Correspondingly, WS has been used as a model of atypical visuospatial development.

Early studies of drawing abilities suggest that the core impairments associated with WS lay in the global integration of local features. For example, local features such as windows and doors were drawn recognizably well, but their global configuration was drawn incorrectly (Bihrle, Bellugi, Delis, & Marks, 1989). However, drawing tasks involve not just the integration of visual information, but also the integration of visuomotor information, which have different neural requirements (Gervan, Berencsi, & Kovacs, 2011). Subsequent investigations suggest that integration in the visual domain is less impaired than integration of information from visual and motor modalities in WS (Farran & Jarrold, 2005), suggesting that integration deficits depend on task. Indeed, people with WS show typical effects of spatial configuration (Pani, Mervis, & Robinson, 1999) and typical susceptibilities to visual illusions (Palomares, Landau, & Egeth, 2008; Palomares, Ogbonna, Landau, & Egeth, 2009). Global texture segmentation in WS individuals also seems functionally intact (Farran & Wilmut, 2007), but is likely dependent on the specific characteristics of local features (Farran, 2005). Yet, they show deficiencies in basic perceptual discrimination such in orientation (Dilks, Hoffman, & Landau, 2008; Farran, 2006; Palomares, Englund, & Ahlers, 2011; Palomares, Landau, & Egeth, 2009), spatial position (Palomares et al., 2008) and size (Palomares, Ogbonna, et al., 2009). Thus even in the visual domain, there are peaks and troughs in WS abilities.

An fMRI study measuring the size of primary visual cortex, V1, in WS individuals showed typical values (Olsen et al., 2009). However, anatomical evidence suggests smaller cortical volume in occipital areas (Bellugi, Lichtenberger, Mills, Galaburda, & Korenberg, 1999) and abnormal cell size packing density in V1 (Galaburda & Bellugi, 2000). A broad framework has been proposed to explain the visuospatial deficits found in WS, which is rooted in the vulnerability to damage of dorsal visual functions (Atkinson et al., 1997, 2003; see also Reiss, Hoffman, & Landau, 2005). It is a generally accepted idea that the visual system is functionally

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segregated into dorsal and ventral visual streams, wherein the former processes motion and latter processes form (Hubel & Livingstone, 1987; Livingstone & Hubel, 1987; Mishkin & Ungerleider, 1982). The detection of coherently oriented lines within an array of random lines was reported to be relatively more intact than the detection of coherently moving dots within an array of randomly moving dots in WS, supporting the notion that dorsal visual functions were more vulnerable to damage than ventral visual functions (Atkinson et al., 2003, 2006). While it is not known whether motion direction selective neurons in V1 or in extrastriate areas such as hMT+, a motion processing area, have atypical responses in WS, parietal areas, a part of dorsal visual stream, in WS do show atypical characteristics relative to controls such as functional hypoactivation and structural reductions in grey and white matter (Thompson et al., 2005).

In the current study, we use Glass patterns (Glass, 1969; Glass & Switkes, 1976) to portray textures via orientation from locally paired dots to evaluate how global integration develops in typical and atypical maturation. We investigated how typically developing children and people with WS detect global form coherence of Glass patterns. In addition, we also presented dot stimuli with real coherent motion. If global form integration is impaired in WS individuals, then their sensitivity to global form coherence should exhibit a different pattern from those of typically developing children or adults. Furthermore, if motion mechanisms in WS were damaged, then real motion processing should be atypical from what is observed from typically developing children.

1.1. Global integration in typical development

Random-dots are widely used stimuli to study global integration of local cues because the global structure can be incrementally manipulated from organized to random without altering the structure of the local features. In these hierarchical dot stimuli, large receptive fields in extrastriate cortex detect the global structure, while small receptive fields in early visual cortex detect the local structure. An example of dot stimuli that requires global integration of form is Glass patterns, which are moirés from randomly distributed dots and their spatially transformed copies (Glass, 1969; Glass & Switkes, 1976). Spatial summation for these stimuli occurs at a much larger area than receptive fields in primary visual cortex, V1 (Mandelli & Kiper, 2005). Dot pair separation affects local and global structure of Glass patterns differentially (Kurki, Laurinen, Peromaa, & Saarinen, 2003; Palomares, Pettet, Vildavski, Hou, & Norcia, 2010). Visual evoked potentials also show different scalp topographies for the local and global structure of Glass patterns (Palomares, Ales, Wade, Cottereau, & Norcia, 2012). Global structure of Glass patterns elicits responses in the ventral visual pathway, particularly in the lateral occipital complex, (LOC; Ostwald, Lam, Li, & Kourtzi, 2008). Notably, the orientation of parallel structure in Glass patterns has been modeled in V1 (Smith, Bair, & Movshon, 2002), likely involving mechanisms outside of the classical receptive fields.

Spatial integration of local features into global shapes and textures occurs at a very early age. Newborns are able to distinguish the global orientation of squares that are grouped by columns or rows (Farroni, Valenza, Simion, & Umilta, 2000). Infants (4–6 months of age) can detect a concentric texture pattern made up of lines or spatially proximal dot pairs (Palomares et al., 2010). However, degrading the local orientation signal by removing 2 central pixels or presenting larger inter-dot separations diminished responses, suggesting that the recognition of global Glass pattern structure exhibits fragility early in development. Glass pattern sensitivity was reported to be adult-like after 6 years of age (Lewis et al., 2004), which would suggest a concise developmental trajectory relative to other visual tasks such as contour integration (Kovacs, Kozma, Feher, & Benedek, 1999; see also Palomares, Landau, et al., 2009).

1.2. Illusory motion from Glass patterns

Notably, Glass patterns induce a perception of implied or illusory motion when a series of these patterns are presented sequentially and rapidly; commonly coined as "dynamic". These dynamic Glass patterns interact with real motion psychophysically (Ross, 2004) and physiologically as they have shown some sensitivity in the motion processing area of hMT+ (Krekelberg, Dannenberg, Hoffmann, Bremmer, & Ross, 2003; Krekelberg, Vatakis, & Kourtzi, 2005). Directed attention also modulated VEP responses to dynamic Glass patterns in hMT+ (Palomares et al., 2012). Contrary to the separation of form and motion, these studies suggest that these mechanisms interact (Burr, 1980; for review of motion mechanisms, see Burr & Thompson, 2011; Geisler, 1999). In typical adults, coherence thresholds to dynamic Glass patterns are lower than thresholds to static Glass patterns (Burr & Ross, 2006; Or, Khuu, & Hayes, 2007) indicating that illusory motion enhances global integration of orientation signals. It is unknown whether static and dynamic Glass patterns would show the same relationship in immature or atypical visual global integration systems, such as in WS (Palomares et al., 2008). If dynamic Glass patterns activate motion processes, it is possible that participants with WS might have atypical sensitivities to these kinds of stimuli since it has been reported that WS individual have difficulty processing motion stimuli (Atkinson et al., 1997, 2006). It had been posited that visual functions generally mediated by the dorsal visual stream (Atkinson et al., 1997, 2003, 2006) are vulnerable to damage in WS – a "dorsal stream vulnerability hypothesis".

The current objectives of this study are threefold: (1) To test the predictions of the dorsal stream vulnerability hypothesis (Atkinson et al., 2003, 2006) – the relative sparing of form processing compared to motion processing in WS, (2) to track the developmental trajectory of detecting form and motion coherence in typical development, and (3) to determine the relationship of form and motion processing in illusory motion from dynamic Glass patterns by characterizing patterns in typical development as well as in WS participants.

2. Methods

2.1. Participants

We recruited 83 participants: 16 typical adults (range, 19.49–27.37 years of age), 50 typically developing school-aged children (range, 4.67–15.04 years of age) and 17 individuals with WS (range, 8.28–35.75 years of age). Children and WS participants received monetary compensation while adults received class credit for participation. The University of South Carolina Institutional Review Board (IRB) approved this research.

A geneticist, according to the known genetic marker, positively diagnosed our WS participants. They were given the Kaufman Brief Intelligence Tests, Second Edition (KBIT-2; Kaufman & Kaufman, 2004), an intelligence test that measured both by vocabulary (verbal) and non-verbal analytical skills (matrices). The WS group had mean raw scores of 60 (range, 32–83) for Verbal and 25 for Matrices (range, 12–36) on the KBIT-2. These correspond to the scores of typical 10-year-olds (Verbal) and 7-year-olds (Matrices) at the 50th percentile. The mean Full Scale IQ of the WS group was 73 (range, 49–90), which falls in the same range as in other studies (Mervis et al., 2000).

Performance of WS participants was also compared to a subset of typically developing children as mental age matches ($n = 8$) or