Brain and Cognition 70 (2009) 21–30

Contents lists available at ScienceDirect

Brain and Cognition

journal homepage: www.elsevier.com/locate/b&c

Orientation perception in Williams Syndrome: Discrimination and integration

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ARTICLE INFO

Article history:
Accepted 20 November 2008
Available online 23 February 2009

Keywords:
Williams Syndrome
Development
Perceptual organization
Orientation perception
Dorsal visual pathway
Ventral visual pathway
Grouping
Contour integration

ABSTRACT

Williams Syndrome (WS) is a rare neurodevelopmental disorder, which stems from a genetic deletion on chromosome 7 and causes a profound weakness in visuospatial cognition. Our current study explores how orientation perception may contribute to the visuospatial deficits in WS. In Experiment 1, we found that WS individuals and normal 3–4 year olds had similar orientation discrimination thresholds and had similar prevalence of mirror-reversal errors for diagonal targets (±45 deg). In Experiment 2, we asked whether this immaturity in orientation discrimination would also be reflected in a task requiring integration of oriented elements. We found that sensitivities of WS individuals for detecting orientation-defined contours were higher than sensitivities of normal 3–4 year olds, and were not significantly different from sensitivities of normal adults. Together, these results suggest that orientation discrimination and orientation integration have different maturational trajectories in normal development and different susceptibilities to damage in WS. These may reflect largely separate visuospatial mechanisms.

1. Introduction

Orientation perception is a fundamental property of the visual system, which may limit several aspects of visuospatial functions. In order to identify objects correctly, one must be able to detect and integrate orientation information. For example, the letters **N** and **Z** have similar configurations, but have orthogonal orientations. Failure to discriminate among horizontals, verticals, right obliques and left obliques would result in the perception of novel symbols or wrong letters. Moreover, combination of orientation information is necessary for formation of contours and objects. When local orientations are aligned (i.e. collinear), global contours (Field, Hayes, & Hess, 1993) are more easily detected.

Although much is known about the mechanisms involved in detecting and discriminating oriented elements, little is known about how information from individual oriented elements is integrated into contours, particularly in development. Understanding the characteristics of orientation processing in immature and abnormal systems may elucidate the developmental mechanisms underlying visuospatial processing and their role in higher-level functions such as spatial cognition and object recognition. Thus, we examined the relationship between orientation discrimination and integration of orientation information in typically developing children and in individuals with Williams Syndrome (WS), a genetic disorder resulting in severe visuospatial impairments (Bellugi, Lichtenberger, Mills, Galaburda, & Korenberg, 1999).

1.1. Williams Syndrome

WS is a developmental disorder associated with a microdeletion of about 20 genes on chromosomal region 7q11.23 (Lenhoff, Wang, Greenberg, & Bellugi, 1997). WS occurs in 1 out of 7500 live births (Stromme, Bjornstad, & Ramstad, 2002). It causes mild to moderate mental retardation (mean IQ of 60), but is typically associated with relative strength in language despite severe weakness in visuospatial tasks (Bellugi et al., 1999).

1.1.1. Orientation discrimination

The study of orientation discrimination in people with WS is particularly relevant because one of the hallmarks of their spatial deficit is severely impaired representation of orientation, at least as measured so far. Using the Benton judgment of line orientation task (JLOT), early studies showed that many WS participants failed even the pretest, which requires two consecutive correct answers out of 5 relatively easy items (Wang, Doherty, Rourke, & Bellugi, 1995). By contrast, these same early studies showed that WS performance in the Benton face recognition task is very close to normal adult performance (Bellugi et al., 1999; Wang et al., 1995), even though it requires recognition of the same face over different orientations. This suggests that representation of orientation in WS may be relatively normal for their chronological age under some circumstances (e.g. face recognition) and not others (e.g. line orientation discrimination).

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doi:10.1016/j.bandc.2008.11.007

Recent evidence suggests that floor performance of WS individuals in JLOT may be due to the complexity of the task. The Benton JLOT requires that a person discriminate the orientations of two different lines, and match them to their identity from a set of 11 choices. Although people with WS fail even the pretest of this task, simpler tasks of line orientation have shown that judgments in WS adults are comparable to those of typically developing children (Farran, 2006). In Farran's study, thresholds were measured by adjusting the number of possible choices (2–10 lines instead of 11 in the standard JLOT) or percent correct was measured in a same-different task. The results showed that the *precision of tilt discrimination* (e.g. discriminating between lines tilted 0 and 2 deg) is similar in WS adults and normally developing children matched for mental age (who were on average 5–6 years old). This suggests that orientation tuning in people with WS is not completely absent, nor is it qualitatively different from a normal system (as suggested by Scerif & Karmiloff-Smith, 2005), but rather, that it is functionally delayed or arrested at the level of a 5- to 6-year-old normally developing child. This is consistent with the fact that sensitivity to orientation differences is a foundational visual skill, which is present in young infants (e.g. Slater, Morison, & Somers, 1988) and a range of species, including even lower vertebrate species such as the goldfish (Volkmann, Zametkin, & Stoykovich, 1974).

The precision of tilt discrimination is one aspect of orientation representation. But another aspect that could be subject to abnormality in people with WS is the *accuracy in the direction of tilt* (i.e. discriminating lines tilted left or tilted right). WS individuals often make mirror-reversal errors in block construction (Hoffman, Landau, & Pagani, 2003), a task in which observers copy a global target model using individual blocks. When copying the global pattern, a WS participant must select individual blocks that are replicas of the ones used in the target model. They often make confusions such as selecting a diagonally split block whose split runs from upper left to lower right to stand in for one whose split runs from upper right to lower left. Young normally developing children also make such errors frequently in the block task. As a whole, these studies suggest that the WS pattern of selecting mirror image blocks in the block copy task might reflect developmental delay or arrest in this aspect of orientation representation.

1.1.2. Orientation integration

If people with WS and young normally developing children have difficulty representing orientation (and especially mirror-images), one might expect that these problems would show up in tasks requiring the integration of oriented elements, as well as simple discrimination tasks. Indeed, Kovacs, Lukacs, Feher, Racsmany, and Pleh (2001) found that children with WS perform quite poorly in tasks requiring the integration of oriented elements (Kovacs et al., 2001). In their task, observers were asked to detect collinear contours embedded among randomly oriented noise elements, and thresholds were measured by changing the relative density between the contour and noise elements. Their WS participants could not perform at the same level as typical adults. The failure to use orientation for integration tasks such as this is consistent with the idea that people with WS have a deficit in global processing (e.g. Bellugi et al., 1999). However, the general hypothesis of a global processing deficit has been challenged. For example, WS individuals perform much like normal adults in tasks that require using grouping cues to accelerate visual search (Pani, Mervis, & Robinson, 1999), and to segment textures (Farran & Wilmut, 2007), They also perceive visual illusions to the same extent as normal adults, suggesting that their perceptual *integration* of elements may be normal (Palomares, Ogbonna, Landau, & Egeth, in press).

In the current study, we examined orientation perception in WS from two different perspectives: Discrimination of oriented elements, and integration of these elements into a global whole. Because people with WS appear to have some deficit in the perception and representation of oriented lines, but have strong mechanisms of visual-spatial integration (at least in some circumstances), it is possible that these two functions may be separable in this population. Moreover, because there are hints that the visual-spatial deficit in WS has remarkable parallels with the profile observed in young normally developing children around age 4 (Landau & Hoffman, 2007), it is possible that two separate profiles may be observed in normal children of this age. If separate profiles for orientation discrimination and integration of oriented elements are observed in both people with WS and young normally developing children, this would suggest two separate mechanisms with different developmental trajectories.

Therefore, In Experiment 1, we asked how people with WS and normally developing children aged 3–4, 5–6, and 7–9 years perceive and discriminate fine orientation differences (i.e. precision of tilt), and whether they make mirror-image reversal errors (i.e. direction of tilt). In Experiment 2, we asked how participants integrate oriented elements, specifically, how they detect an orientation-defined contour embedded among randomly oriented noise elements.

The locus of orientation processing is thought to be principally in early visual cortex, such as V1 (e.g. Nauhaus, Benucci, Carandini, & Ringach, 2008), an area shown to be disproportionately smaller in WS individuals relative to normal controls (Bellugi et al., 1999; Chiang et al., 2007). If orientation discrimination and orientation integration were largely limited by the same mechanisms in V1, then we would expect that they would have the similar vulnerabilities in atypical development and similar normal developmental trajectories. Alternatively, if different profiles emerge for the two tasks, then this would suggest the possibility that these different tasks engage different areas of the brain, which develop on different timetables and are differentially susceptible to neurological damage in Williams Syndrome.

Indeed, we found that orientation *discrimination* became adult-like after the age of 6 years in typically developing children, and was at the level of 3- to 4-year-old children in our group of WS individuals, whose mean chronological age was 18 years. We also found that orientation *integration* became adult-like after the age of 4 years—earlier than discrimination, and was at the level of typical adults in WS individuals. These results indicate that orientation discrimination and integration are likely mediated by largely separate mechanisms beyond early visual cortex. The immaturity of orientation discrimination in WS might be due to abnormalities in the dorsal visual pathway (see Section 4.1), cortical areas found to be vulnerable to damage in WS (e.g. Bellugi et al., 1999; Meyer-Lindenberg et al., 2004).

2. Experiment 1: Discrimination of oriented gratings

Performances of WS individuals in the Benton line discrimination task (Bellugi et al., 1999; Wang et al., 1995) and block construction tasks (Hoffman et al., 2003) indicate that WS individuals may have deficits in orientation selectivity. In Experiment 1, we measured orientation discrimination thresholds to gratings in WS individuals, normal children and adults. The Benton line discrimination task requires matching of two target lines from a set of 11 lines, and may require substantial attentional resources to select one line from a crowded display. The current task is much simpler, requiring only a match of one target grating to one of four gratings.

2.1. Methods

2.1.1. Participants

Fifty-nine people participated in this experiment. These included 11 WS individuals (mean age = 18 years; 7 months, minimum age = 11 years; 10 months, maximum age = 24 years; 5