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Motion processing specialization in Williams syndrome

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Abstract

Williams syndrome (WS) is a rare genetic disorder characterized by severe spatial deficits and relatively spared language. Although initial research suggested that WS entails a generalized motion processing deficit, later work demonstrated intact biological motion perception in people with WS, reflecting a sparing of a specific motion perception system. The present study examined whether this sparing is unique to biological motion, or extends to other motion tasks as well. WS children and adults and normal controls were tested to examine developmental changes across a variety of motion tasks. Results indicated that WS individuals performed at normal levels for motion coherence and biological motion tasks but had elevated thresholds for the 2-D form-from-motion task, a profile that extended into adulthood. These findings provide evidence that a genetic impairment can lead to a selective motion processing deficit and argue against characterizing WS as including a general motion processing impairment. The nature of the motion deficit is considered, including the implications for WS dorsal/ventral processing.

Keywords: Williams syndrome; Motion coherence; Biological motion; Form-from-motion; Dorsal/ventral visual processing

1. Introduction

Williams syndrome (WS) is a rare (1:20,000 live births) congenital deficit resulting from a submicroscopic deletion on chromosome 7q11.23, a region known to help regulate elastin and LIM-kinase1 expression (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Frangiskakis et al., 1996). People with WS commonly exhibit features such as 'elfin' facial appearance, connective tissue malformations, cardiovascular problems, generally reduced brain volume, a hypersocial personality, and an overall Composite IQ (as measured using the Kaufman Brief Intelligence Test, KBIT, Kaufman & Kaufman, 1990) in the mild to moderately retarded range (Bellugi, Marks, Bihrle, & Sabo, 1988; Bellugi et al., 2000; Mervis, Morris, Bertrand, & Robinson, 1999a). What has attracted psychologists and neuroscientists to this population is their strikingly uneven cognitive profile, which consists of relative strengths in language, together with severe impairments in visuospatial abilities (Bellugi et al., 1988; Mervis et al., 1999a). This profile raises the possibility that a genetic defect might developmentally target specific domains of cognition, or brain areas that underlie these domains.

The spatial deficit in WS people is most pronounced in visuo-constructive tasks such as 'block construction' (e.g., Pattern Construction subscale of the Differential Abilities Scale, DAS, Elliot, 1990) and copying (e.g., the Developmental Test of Visual–Motor Integration, VMI, Beery & Buktenica, 1967) which require individuals to replicate the configural arrangement of a model (Bellugi et al., 1988; Bellugi, Wang, & Jernigan, 1994; Mervis et al., 1999a). In contrast, perception of objects (Landau, Hoffman, & Kurz, submitted) and faces in people with WS (Tager-Flusberg, Plesa-Skwerer, Faja, & Joseph, 2003) appears to be normal.

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Although initial studies suggested that the WS visuo-spatial deficit resulted from a failure to group local features into coherent global percepts (Bellugi et al., 1994; Bihrle, Bellugi, Delis, & Marks, 1989), subsequent studies have reported normal performance in a variety of global integration tasks (Atkinson et al., 1997, 2003; Jordan, Reiss, Hoffman, & Landau, 2002; Key, Pani, & Mervis, 1998; Pani, Mervis, & Robinson, 1999; Tager-Flusberg et al., 2003). More recent attempts (Atkinson et al., 1997, 2001, 2003; Galaburda et al., 2001; Jernigan, Bellugi, Sowell, Doherty, & Hesselink, 1993; Wang, Doherty, Rourke, & Bellugi, 1995) to account for the pattern of preserved and impaired visual abilities in people with WS invoke the distinction between 'dorsal' and 'ventral' visual streams proposed by Milner and Goodale (1995). People with WS perform normally on a variety of tasks that are thought to be carried out by ventral visual areas, including recognition of objects (Landau et al., submitted) and faces (Tager-Flusberg et al., 2003). In contrast, they are impaired in a variety of dorsal stream tasks such as visually-guided actions (Atkinson et al., 1997; Dilks, Landau, & Hoffman, submitted) and perception of some kinds of motion (Atkinson et al., 1997, 2003).

Atkinson and colleagues (1997, 2003) tested the hypothesis of dorsal breakdown with ventral sparing by directly comparing performance on a pair of tasks that require similar judgments but are based on information derived from each of the two visual streams. They found that although a subgroup of WS individuals performed poorly on both tasks—suggesting a general deficit in visual processing—a separate subgroup of WS individuals could successfully detect a 2-D shape that required integration of *static* oriented line segments into a global form but were impaired when perception of a form required integration of local *motion* signals. This finding is consistent with normal ventral stream function but impaired dorsal functioning because previous research in both humans and monkeys indicates that a key cortical region for detecting forms (V4, Gallant, Shoup, & Mazer, 2000; Girard, Lomber, & Bullier, 2002) lies along the ventral stream; whereas a key brain area critical for detecting motion (V5/MT, Newsome & Paré, 1988; Zeki et al., 1991) resides in the dorsal stream (see also Braddick, O'Brien, Wattam-Bell, Atkinson, & Turner, 2000).[1]

Deficits in visually-guided action and perception of motion are consistent with the hypothesis of a *generalized* deficit in dorsal stream functioning in WS (Atkinson et al., 1997, 2003); however, research both in adults with cortical lesions (Cowey & Vaina, 2000; McLeod, Dittrich, Driver, Perrett, & Zihl, 1996; Schenk & Zihl, 1997; Vaina, Lemay, Bienfang, Choi, & Nakayama, 1990) and with normal adults (Beintema & Lappe, 2002; Grossman & Blake, 1999) has revealed that motion processing is not a unitary function. Instead, it appears to be comprised of several dissociable 'classes' of motion perception (e.g., motion coherence, form-from-motion, biological motion, etc.), consistent with the existence of several motion processing 'specialists' in the normally developed brain. These behavioral dissociations have been supplemented by findings that different classes of motion perception rely on distinct neural circuitry. Specifically, even though V5/MT is a key motion processing area and appears to be the primary center for processing motion coherence, additional cortical areas have also been implicated for the other two motion classes. For example, single-cell research in monkeys as well as human brain-imaging studies report that the superior temporal sulcus (STS, which integrates information originating from both ventral and dorsal streams) responds specifically to biological motions (Allison, Puce, & McCarthy, 2000; Bonda, Petrides, Ostry, & Evans, 1996; Oram & Perrett, 1994; Vaina, Solomon, Chowdhury, Sinha, & Belliveau, 2001) and is part of a larger network (which includes the fusiform face area, Kanwisher, McDermott, & Chun, 1997) involved in the perception of biological stimuli. In contrast, perception of form-from-motion stimuli appears to be achieved through V5/MT projections to ventral stream areas such as V4 (Maunsell & Van Essen, 1983; Ungerleider & Desimone, 1986) or LO (Schoenfeld et al., 2003), which allow additional ventral stream areas such as inferior temporal cortex to complete perceptual processing (also see discussion by Milner & Goodale, 1995). In light of this evidence for multiple motion systems, the present paper asks whether there may be dissociable patterns of performance among different motion tasks in people with WS.

Recent research has, in fact, shown that at least one kind of motion perception is preserved in WS: biological motion (Jordan et al., 2002). This study demonstrated that, like mental-age-matched (MA) children and normal adults, WS individuals could easily identify various actions of a point-light character (e.g., slipping on a banana, doing jumping jacks, etc.; see Johansson, 1973). In addition, WS children were comparable to or better than MA controls in their ability to discriminate the direction of locomotion of a point-light-walker (PLW) embedded in dynamic noise.

These findings suggest that WS entails selective sparing of some types of motion perception even when they require global integration of local features (i.e., the integration of local lights into limbs and eventually a human form, Bertenthal & Pinto, 1994; Lorenceau & Shiffrar,

[1] Milner and Goodale (1995) propose that crosstalk between streams makes it difficult to conclude that visuo-spatial deficits are due solely to dorsal stream damage. For example, although V5/MT lies along the dorsal stream, the prolific connections it has with regions along both pathways make its exclusive assignment to either individual stream uncertain.