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Note

Dorsal-stream motion processing deficits persist into adulthood in Williams syndrome

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Abstract

Previous studies of children with Williams syndrome (WS) have found a specific deficit in dorsal cortical stream function, indicated by poor performance in coherence thresholds for motion compared to form. Here we investigated whether this is a transient developmental feature or a persisting aspect of cerebral organization in WS. Motion and form coherence thresholds were tested in a group of 45 WS individuals aged 16–42 years, and 19 normal adult controls.

Although there was considerable variation in the coherence thresholds across individuals with WS, the WS group showed overall worse performance than controls. A significant group × threshold condition interaction showed a substantially greater performance deficit for motion than for form coherence in the WS group relative to controls. This result suggests that the motion deficit is an enduring feature in WS and is a marker for one aspect of dorsal-stream vulnerability.
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1. Introduction

Individuals with Williams syndrome (WS) show a characteristic and unique cognitive and behavioural profile. Although there are wide variations across individuals with WS, they generally have IQ's between 50 and 90, with a relative sparing of expressive language, good visual object recognition (especially of faces), 'hypersocial' behaviour with generally 'friendly' personality traits, but poor performance on most visuo-spatial and constructional tasks (Atkinson et al., 2001; Bellugi, Bihrle, Trauner, Jernigan, & Doherty, 1990; Bellugi, Lichtenberger, Jones, Lai, & St George, 2000; Jones et al., 2000; Mervis et al., 2000). The syndrome is associated with a specific deletion on chromosome 7, and therefore provides a way to explore links between specific gene expression, brain development, and cognitive function (e.g., Bellugi et al., 1990; Bellugi, Lichtenberger, Mills, Galaburda, & Korenberg, 1999). However, while structural differences between WS and typically developing brains have been identified (Eckert et al., 2005; Mercuri et al., 1997; Meyer-Lindenberg et al., 2004; Reiss et al., 2004; Schmitt, Eliez, Bellugi, & Reiss, 2001), the brain basis of the cognitive profile is still far from fully understood.

It is now widely accepted that visual information in the primate cortex is processed through two distinct, yet interacting, processing streams (Milner & Goodale, 1995; Mishkin, Ungerleider, & Macko, 1983). From studies of non-human primates the *ventral* stream, projecting from primary visual cortex to the temporal lobe, performs the visual computations needed for the recognition of objects and faces (i.e., 'what' and 'who' tasks) and its intermediate stages (e.g.,

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area V4) show specific sensitivity to shape and colour information. The *dorsal* stream, projecting from primary visual cortex to the parietal lobe, performs computations needed to register spatial relationships relative to the observer and to provide the visual information needed for the control of spatially directed actions (i.e., 'where' and 'how' information). Its intermediate stages (e.g., area V5/MT) show sensitivity to motion and stereo information. Measures of global form and motion processing have therefore been taken as indicators of the function within extrastriate visual areas in the two streams (Atkinson et al., 1997; Braddick, Atkinson, & Wattam-Bell, 2003; Gunn et al., 2002); functional imaging results have supported this separation by demonstrating that global coherence of form and motion activate largely non-overlapping systems in posterior cortex (Braddick, O'Brien, Wattam-Bell, Atkinson, & Turner, 2000). However, these imaging studies suggest that the independent networks for form and motion both involve areas in occipital, parietal and temporal lobes, a rather different picture to the division, suggested by work with non-human primates with the ventral stream being primarily directed to the temporal lobes and the dorsal stream to the parietal lobes.

The profile of abilities in WS suggests that visual abilities subserved by the ventral stream, such as face recognition, are relatively well developed, whereas those subserved by the dorsal stream, such as visuospatial manipulation, are markedly impaired. Experimental identification of a specific dorsal stream deficit in WS was first provided by Atkinson et al., who showed that children with WS showed deficits in a visuomotor task (the 'mailbox' task) compared to a corresponding visual matching task (Atkinson et al., 1997), and in motion compared to form coherence thresholds (Atkinson et al., 1997, 2003). Since these initial results this 'dorsal-stream vulnerability' has also been found to characterise a number of other developmental disorders, including hemiplegia, autism, developmental dyslexia, and fragile X (e.g., Braddick et al., 2003; Gunn et al., 2002; Kogan et al., 2004; Spencer et al., 2000).

However, the identification of dorsal-stream dysfunction in WS children leaves open the question of the developmental course and ultimate outcome of perceptual and visuospatial skills in the disorder. Is the development of functions normally served by the dorsal stream merely delayed in WS, either because the mechanisms mature slowly or because, given time, WS individuals develop alternative neural routes for such performance? Alternatively, are dorsal-stream functions permanently impaired by an enduring difference between WS and typically developing brains in the absence of successful neural reorganization? Although the answers to these questions have yet to be found, studies have shown quite diverse levels of performance in the young WS groups (Atkinson et al., 1997, 2003), suggesting that the WS phenotype does not lead to a fixed outcome for 'dorsal' processing, but rather that alternative strategies or pathways can be developed.

Williams syndrome has aroused wide interest as an example of genetically determined anomalous cognition. However, its neurocognitive phenotype has to be understood as the result of a developmental cascade, not as a simple expression of a genetic anomaly (Karmiloff-Smith & Thomas, 2003). It is important, therefore, to examine processes of developmental change and stability in the disorder, not simply snapshots at a given stage of development.

In the present study we use global motion and form sensitivity tests with adult WS individuals to assess the developmental course of these abilities. The group tested have shown the ability to participate in wide-ranging cognitive testing, alongside similar testing of controls. They therefore provide a good and well-characterised group to examine whether a stable difference in basic dorsal-stream visual processing persists into adulthood.

2. Participants

Forty-five adults with Williams syndrome were recruited for studies at the Laboratory of Cognitive Neuroscience, Salk Institute, in co-operation with the Williams Syndrome Association. All WS participants met clinical criteria for a diagnosis of WS and obtained a score of at least three points on the WS Diagnostic Score Sheet (DSS), indicating the presence of a minimum threshold for common medical and physical characteristics associated with WS in clinical studies (Korenberg et al., 2000). Furthermore, all WS participants tested positive on a fluorescence in situ hybridization (FISH) test for the absence of one copy of the gene for elastin on chromosome 7 (AAP, 2001). Also recruited was a group of 19 typically developing age-matched controls, generally naïve concerning psychophysical testing. Control participants were screened for the existence of any developmental neurological or psychiatric conditions. Characteristics of the two groups are summarized in Table 1.

The WS participants took part in a wide range of investigations of cognitive performance in the Salk Institute's Laboratory for Cognitive Neuroscience, including administration of the Wechsler intelligence tests (WAIS-R or WISC-R), which yield both verbal and performance component scores. Control participants were assessed on the same test.

Table 1
Characteristics of WS and control groups

	N	Mean age (year)	Age range (year)	Verbal IQ score (S.D.)	Performance IQ score (S.D.)	Full scale IQ score (S.D.)
WS	45	28.3	16–47	69.2 (10.1)	63.8 (9.6)	64.8 (10.5)
Controls	19	27.5	18–41	101.7 (10.4)	98.6 (10.5)	99.8 (9.9)

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