

SPARED OBJECT RECOGNITION WITH PROFOUND SPATIAL DEFICITS: EVIDENCE FROM CHILDREN WITH WILLIAMS SYNDROME



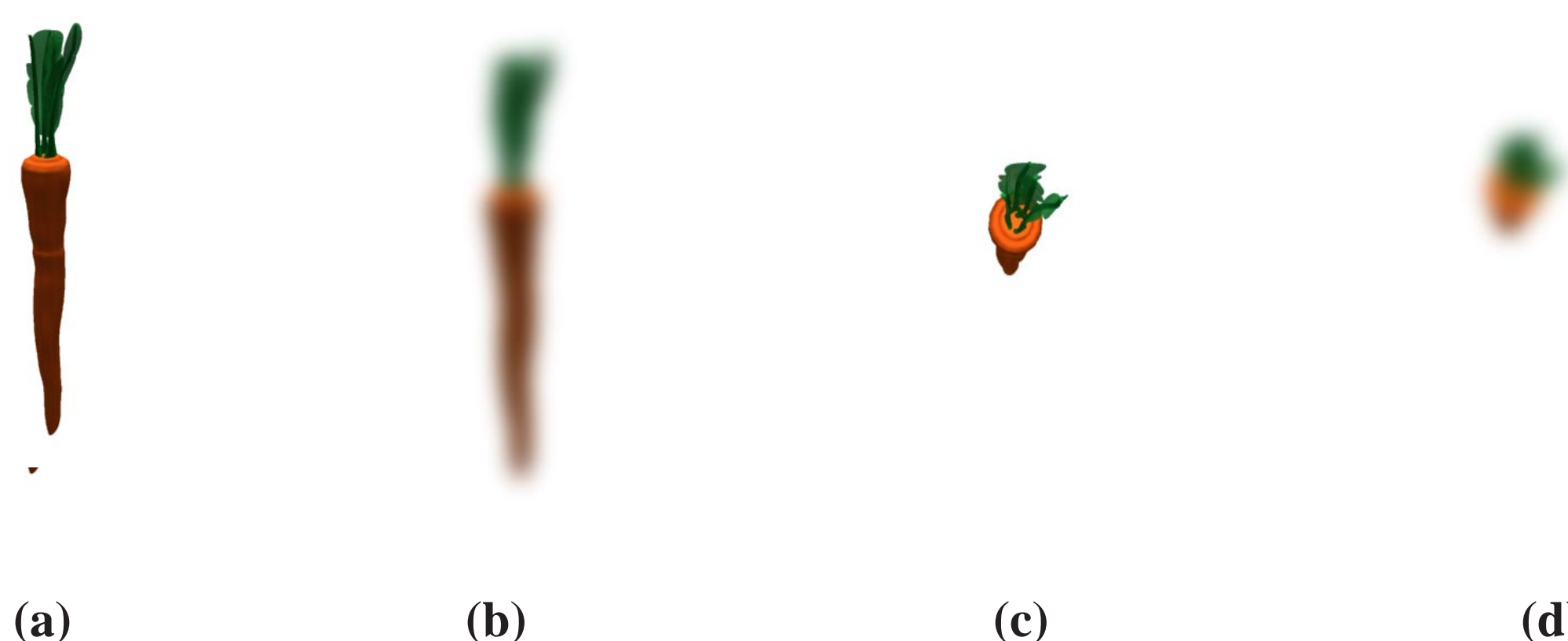
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ABSTRACT

Williams Syndrome is a rare genetic defect that causes profound spatial cognitive deficits in the context of relatively spared capacity for language. This uneven cognitive profile suggests developmental specialization across cognitive domains. In order to explore the possibility of selective breakdown within the spatial domain, we examined the capacity of WS children to recognize objects under conditions of varying difficulty. Ten WS children, ages 7-15 years, 10 normally developing children of the same mental age, and 10 normal adults were asked to name 80 different common objects presented for 500 msec per picture on a computer screen. Object were presented in one of four conditions: (a) canonical viewpoint, clear image; (b) canonical viewpoint, blurred image; (c) non-canonical viewpoint, clear image; (d) non-canonical viewpoint, blurred image.



All groups of participants showed better performance under canonical than non-canonical viewpoints, and under clear than blurred images. Moreover, the decline across conditions was similar in each group, suggesting that object recognition in WS may function normally. This finding is consistent with speculations that the spatial deficit in WS may reflect damage to the dorsal stream, leaving the ventral stream intact.

PROBLEM

The profound spatial deficit shown in Williams Syndrome raises the question of whether this deficit is global, affecting many systems that involve spatial representations, or more specific to particular kinds of spatial representation, such as those important for recognizing objects. Theories of object recognition suggest that the human visual system represents objects as parts and their spatial relationships. Some theories further propose that we represent objects in terms of spatial structures which are **viewpoint invariant** (Biederman, 1987). Other theories propose that objects are more commonly recognized using **viewpoint-dependent** representations (e.g. Tarr and Pinker, 1990). Under this proposal, objects could be recognized from multiple viewpoints through experience viewing particular objects from a variety of perspectives (see, e.g. Logothetis et al, 1994). Indeed, recent evidence suggests that neurons in the temporal visual cortex of the macaque respond to multiple views of objects which had been placed in the animals' cages for a period of time (Booth & Rolls, 1998), suggesting that recognition from multiple viewpoints may develop. It may also be compromised by brain damage. Individuals who suffer damage to infero-temporal areas suffer from visual agnosia—the failure to recognize objects, despite intact low-level visual functioning (see Farah, 1990 for review). Many of these individuals show special difficulties in recognizing objects from unusual viewpoints (Davidoff & Warrington, 1999).

Does the spatial impairment in Williams Syndrome extend to the domain of object recognition? If so, this would be consistent with a relatively global spatial deficit. If not, this would suggest the possibility of selective sparing across spatial domains, in particular the sparing of the object recognition system, and more generally, the ventral stream.

Because mature object recognition depends on our capacity to recognize objects from multiple viewpoints, we tested children with Williams Syndrome on their capacity to recognize a range of common objects from “canonical” viewpoints-- defined as the viewpoint which preserves the maximum amount of object structure-- and “non-canonical” or unusual viewpoints-- ones which we would not normally encounter in our everyday interaction.

BACKGROUND

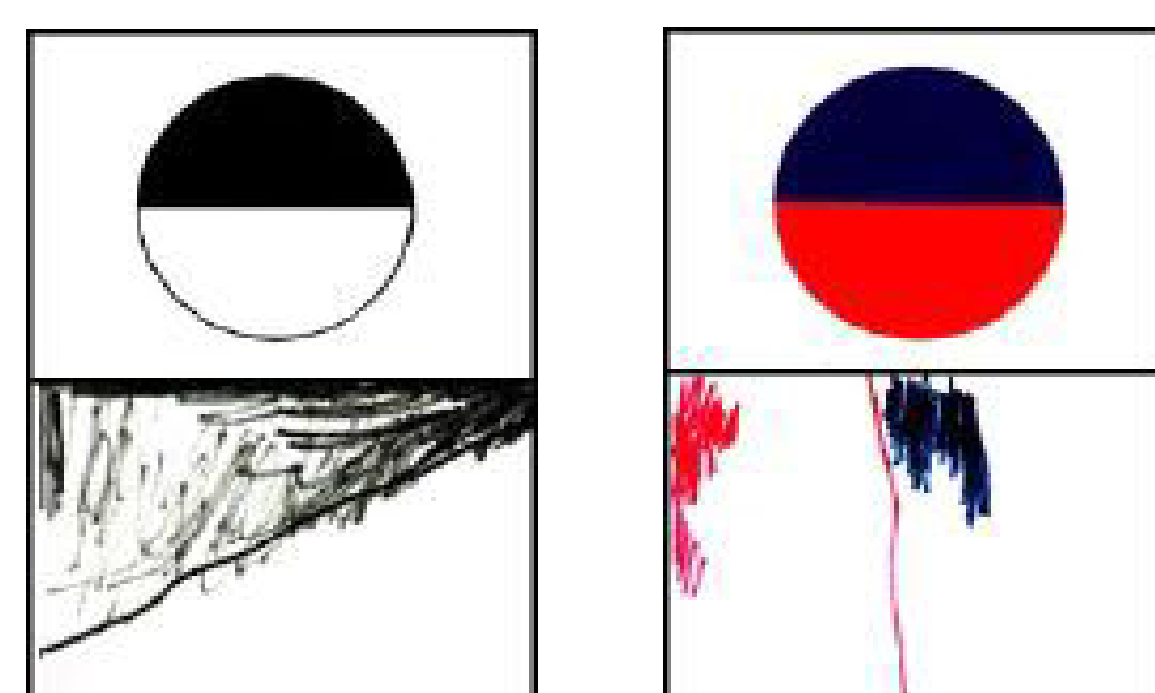


Williams Syndrome is a rare genetic defect (1 in 25,000) which is characterized by profound spatial deficits together with relatively spared language, suggesting the possibility that different cognitive systems are differentially affected.

The syndrome is caused by a hemizygous submicroscopic deletion of chromosome 7q11.23, which includes the gene for elastin (ELN), the gene for protein LIMK1 (Frangiskakis et al., 1996), and others (Meng et al., 1998). The gene LIMK1 is thought to be implicated in the spatial disorder, as it is expressed pre-and post-natally in the brain, whereas ELN is not (Frangiskakis et al., 1996). Diagnosis is made through phenotypic characteristics and/or a fluorescent in situ hybridization (FISH) test, which can confirm the existence of the relevant deletion (Morris et al., 1994). The phenotypic characteristics of WS individuals include a distinctive set of facial features, certain malformations of connective tissue and heart malfunction.

Of principal relevance for studies of language and cognition, WS individuals show a characteristic cognitive pattern that includes overall moderate mental retardation (Mean IQ= 55–60) as well as a unique profile of profound spatial impairment together with relatively spared language capacities (Bellugi et al., 1992; Mervis et al., 1996).

The **SPATIAL DEFICIT** has been observed most often in the drawings of WS individuals, and their reproduction of spatial designs using block construction tasks such as are found in the WISC or the DAS (Differential Abilities Scale). Adults with WS perform in the lowest percentile on such tasks (Bellugi et al., 1992; Mervis et al., 1999). Below, we show samples of copied spatial designs from children with WS who were tested in our lab. Typically, the copy distorts spatial properties while preserving other aspects of design such as color.



SAMPLE DRAWINGS BY CHILDREN WITH WILLIAMS SYNDROME

METHOD

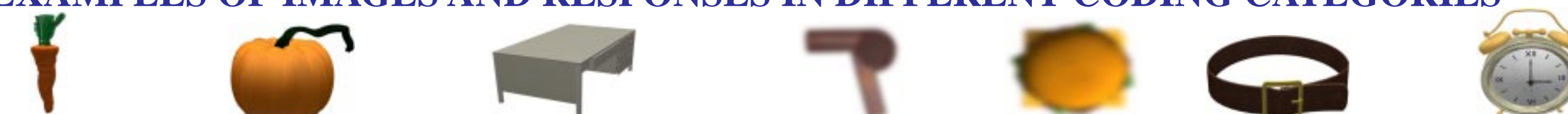
PARTICIPANTS Twelve children with Williams Syndrome between the ages of 7;5 and 15;3 (Mean= 11;1) participated, as well as twelve normally developing children matched for mental age (Mean age= 5;10, Range= 4;1-7;1) and twelve normal adults. The WS children were recruited through the Williams Syndrome Association, and had all been diagnosed by a geneticist, using phenotypic characteristics, and in most cases, FSH tests. All children were tested initially using a standardized intelligence test, the Kaufman Brief Intelligent Test (Kaufman and Kaufman, 1990). The WS children were well matched to the normally developing children on both the Verbal component (Ms = 31, 29, Ranges= 20-46, 20-48, respectively) and on the non-verbal component, Matrices (Ms= 18, 18, Ranges= 12-24, 10-29, respectively). The adults were undergraduates at the University of Delaware.

DESIGN AND PROCEDURES Participants were asked to name a total of 80 objects, each presented on a computer screen for 500 msec. The 80 target objects were drawn randomly from a set of 320 images, which included each of the 80 objects in each of four conditions:

(a) Canonical view, clear image, (b) canonical view, blurred image, (c) non-canonical view, clear image, (d) non-canonical view, blurred image. Each participant saw 20 different objects presented in each of the four image conditions. As the participant named each object, responses were recorded verbatim.

CODING All naming responses were coded by a person blind to participant group. Each response was presented individually on a computer screen along with the target name (i.e. the name that was used in the object creation program) and the image of the object that the participant had been viewing when he or she produced the name. A second rater coded 20% of the responses. Reliability was 89%.

EXAMPLES OF IMAGES AND RESPONSES IN DIFFERENT CODING CATEGORIES



TARGET NAME	RELATED CATEGORY	SUPERORDINATE CATEGORY	SIMILAR SHAPE	SIMILAR OTHER	PART	INCORRECT
CARROT	PLUM	FURNITURE	GUN	SUN	BUCKLE	GLASSES

