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Object recognition with severe spatial deficits in Williams syndrome: sparing and breakdown

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Abstract

Williams syndrome (WS) is a rare genetic disorder that results in severe visual-spatial cognitive deficits coupled with relative sparing in language, face recognition, and certain aspects of motion processing. Here, we look for evidence for sparing or impairment in another cognitive system object recognition. Children with WS, normal mental-age (MA) and chronological age-matched (CA) children, and normal adults viewed pictures of a large range of objects briefly presented under various conditions of degradation, including canonical and unusual orientations, and clear or blurred contours. Objects were shown as either full-color views (Experiment 1) or line drawings (Experiment 2). Across both experiments, WS and MA children performed similarly in all conditions while CA children performed better than both WS group and MA groups with unusual views. This advantage, however, was eliminated when images were also blurred. The error types and relative difficulty of different objects were similar across all participant groups. The results indicate selective sparing of basic mechanisms of object recognition in WS, together with developmental delay or arrest in recognition of objects from unusual viewpoints. These findings are consistent with the growing literature on brain abnormalities in WS which points to selective impairment in the parietal areas of the brain. As a whole, the results lend further support to the growing literature on the functional separability of object recognition mechanisms from other spatial functions, and raise intriguing questions about the link between genetic deficits and cognition. © 2005 Elsevier B.V. All rights reserved.

Keywords: Ventral Stream; Object Recognition; Williams syndrome

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In this paper, we report evidence that some mechanisms of object recognition can develop without impairment even while other aspects of spatial representation are severely impaired. Our evidence comes from people with Williams syndrome, a genetic disorder which gives rise to an unusual cognitive profile including severe spatial deficits together with relatively spared language¹. The striking imbalance between two major cognitive systems has suggested to some that genetic defects can have specific cognitive targets during development (Bellugi, Marks, Bihrle & Sabo, 1988; Jordan, Reiss, Hoffman & Landau, 2002; see also, Frith, 1992). Such targeting has been attributed to modular organization of cognition (Bellugi et al., 1988; Clahsen & Almazan, 1998) or specialization in streams of processing in the mind and brain (e.g. Atkinson, Braddick, Anker, Curran, Andrew and Wattam-Bell, 2003; Dilks, Landau & Hoffman, 2005). The present studies provide support for specific targeting in a genetic deficit, as they show that basic mechanisms of object recognition can be spared even though other aspects of spatial representation are severely impaired. We show that, while children with Williams syndrome cannot reproduce the spatial organization of even moderately complex figures (see Fig. 1), they can recognize a wide range of familiar objects under varying conditions of degradation.

Testing the possibility that a specific cognitive system is spared in a case of genetic impairment requires several steps. First, it requires evidence that the cognitive system in question is specialized, that is, different from other knowledge domains or functions on computational, neural, and/or psychological grounds. Such evidence is abundant in object recognition, and we discuss it below. Second, it assumes that genetic deficits can, in principal, target certain cognitive systems while leaving others either partially or fully spared. As we discuss below, this assumption is complex and currently under debate. Ultimately, the debate can only be resolved by empirical study, which we offer here. Finally, any empirical test requires sufficient fine-grained detail to examine key aspects of normal cognitive architecture. If patterns of performance are both quantitatively and qualitatively similar to normal groups, it is reasonable to conclude that basic aspects of the architecture are spared. Alternatively, detailed patterns of difference can shed light on which aspects of the architecture are robust and which are vulnerable in genetic deficit. The experiments we present will test key aspects of the object recognition system in order to determine how, if at all, performance differs from normal.

1. Object recognition as a specialized system

Thinking of human object recognition as a specialized system is not a new idea. At least since the work of Marr (1982), it has been acknowledged that one of the central computational goals of the visual system is to rapidly recognize objects under a wide range of viewing conditions. The object recognition system is designed to represent

¹ We attach no technical meaning to the term 'severe', but rather, use it to describe the hallmark pattern of Williams syndrome, in which children, adolescents, and even adults carry out visual-spatial constructive tasks such as block construction at the level of four-year-old normal children.

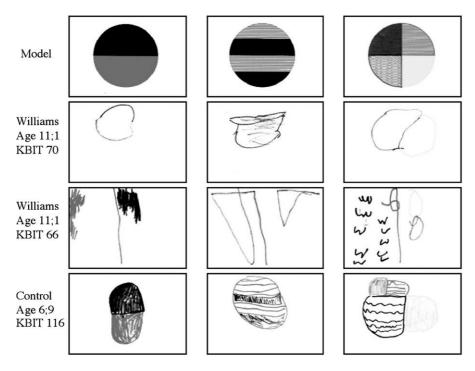


Fig. 1. Sample copies of model figures (row 1) drawn by two children with Williams syndrome (rows 2 and 3) and one normally developing child matched for mental age (row 4). KBIT scores represent IQ, not raw scores used for matching.

objects—primarily by their shapes—in a way that renders them recognizable over changes in irrelevant spatial properties such as size, translation, and viewpoint. Thus, object recognition is inherently spatial. The specific computational problems involved in visual object recognition include segmenting the image from background, parsing it into edges and/or parts, and representing the overall configuration, usually determined by the spatial relationships among parts. The resulting representation must support matching to an incoming image over changes in the object's lighting, size, viewpoint, etc. (Marr, 1982; Palmeri & Gauthier, 2004).

Arguments and evidence suggest that the computational solutions to these problems are unique to objects. For example, researchers have suggested that object shape is represented by the distributed activity of a population of neurons tuned to specific viewpoints (Logothetis & Sheinberg, 1996; Palmeri & Gauthier, 2004). Recent data from brain imaging suggests that the computations involved in recognizing objects are carried out in distributed, but constrained areas of the brain that are distinct from areas that process face recognition (Palmeri & Gauthier, 2004; Pietrini, Furey, Ricciardi, Gobbini, Wu and Cohen, 2004). Presumably, this specialized computational system has evolved to allow us to rapidly and efficiently recognize objects, perhaps even as soon as we know that an object is there (Grill-Spector & Kanwisher, 2005). A particularly difficult problem for the visual system is recognition of objects from partial, sometimes degraded images that result from viewing objects from different viewpoints, which often occlude and distort important features. Some theories suggest that the visual system constructs a single unified viewpoint-invariant representation (e.g. Biederman, 1987; Marr, 1982). However the bulk of empirical evidence indicates that we recognize objects by using viewpoint-dependent representations and interpolating new viewpoints (Bulthoff, Edelman & Tarr 1995; Tarr & Pinker, 1990; Logothetis, Pauls, Bulthoff & Poggio 1994; see Palmeri & Gauthier, 2004). The mechanisms of computing new viewpoints are not well understood, but the capacity to do so is acknowledged to be complex, and is assumed to be a hallmark of our object recognition system.

Considerations of cognitive function and brain localization also support the idea that visual object recognition is specialized. Abundant evidence shows that the human visual system is composed of separate streams which carry out different kinds of spatial processing that serve different functions. For example, beyond the primary visual cortex, the ventral stream processes information about objects and faces. Neurons in inferotemporal (IT) cortex of monkeys respond to the same object under a variety of equivalence conditions (Booth & Rolls, 1998; Logothetis et al., 1994) and to neighboring views of the same object (Tanaka, 1996). In humans, imaging studies show that homologous areas of cortex are activated during object recognition, suggesting a specific neural substrate (Kourtzi, Erb, Grodd & Bulthoff, 2003; Palmeri & Gauthier, 2004).

In contrast to the ventral stream areas and their focus on objects, the dorsal stream is thought to be responsible for processing different kinds of spatial information, including some kinds of motion and location. For example, the perception of motion coherence— which requires computing the global direction of individual motion elements—appears to be processed in area V5/MT (Newsome & Pare, 1988). Still other spatial properties and functions appear to be governed by other areas of the dorsal stream, especially those properties pertinent for action (Bridgeman, Gemmer, Forsman & Huemer, 2000; Colby & Goldberg, 1999; Livingstone & Hubel, 1988; Milner & Goodale, 1995; Ungerleider & Mishkin, 1982).

Finally, the psychological representation of objects has been shown to be dissociable from other kinds of representation that depend on spatial organization, even those within the ventral stream. For example, Moscovitch, Winocur & Behrmann (1997) reported a brain-damaged patient with severely impaired object recognition who could nevertheless recognize faces very well. Duchaine and Nakayama (2005) reported the reverse pattern, developmental prosopagnosics who could not recognize faces, but could recognize categories of objects such as tools and cars quite well. Milner and Goodale (1995) reported a patient who was severely impaired in judging a line's orientation, but could use her perception of the line to guide action (e.g. posting a letter through a slot). Importantly, patients who lose the ability to recognize objects often have damage to the ventral stream, specifically areas of inferotemporal and occipital cortex. These people often perform worse under degraded conditions such as varying illumination or novel viewpoints (see Farah, 2000), suggesting that they have lost the hallmark capacity to recognize object equivalences over such varying conditions.

As a whole, this evidence strongly suggests that the object recognition is specialized in its computational mechanisms, neural substrate, and psychological function.

2. Genetic deficits and cognitive specialization

Can a genetic deficit target certain cognitive systems while sparing others? This issue is controversial. One aspect of the controversy concerns whether or not developmental (i.e. genetic) cases can be used to make the same kinds of inferences about cognitive structure as adult cases of brain damage. Arguments in favor of selective sparing have often come from studies of brain-damaged adults, who represent the case of a mature system that sustains damage. Evidence from brain-damaged adults who can perceive faces but not objects suggests some type of specialized organization in which systems have been differentially targeted by the damage (e.g. Moscovitch et al., 1997). In contrast, genetic deficit will reflect the same kinds of division of labor as revealed by cases of damage to the mature brain. A second controversial issue concerns the relationship between genes and cognition. Genetic changes to an organism will inevitably result in local changes to the physiology of the brain. The crucial issue is whether such local changes will inevitably affect *cognitive structure*.

Views on these two aspects of the controversy lead to very different sets of predictions about the likely cognitive outcome of any genetic deficit. In one view, it is possible for a genetic defect to selectively target particular cognitive systems—either because of distinct computational requirements of the system, or because the defect selectively impairs streams of processing in the brain that normally support that system. In this view, the local effects that genes have on brain physiology interact with constraints on knowledge domains that overdetermine cognitive outcomes. Thus, cognitive domains could be expected to have normal structure. This possibility is consistent with the idea that different cognitive domains are computationally distinct, that they have different neural substrates, and that the internal constraints of the system guide development (e.g. Frith, 1992; Gallistel, Brown, Carey, Gelman & Keil, 1991; Spelke & Newport, 1998; Tager-Flusberg, Plesa-Skwerer & Faja, 2003).

Several recent studies in the domain of faces are consistent with this view. For example, Tager-Flusberg et al. reported that people with Williams syndrome show the face inversion effect usually observed in normal individuals, suggesting that the face processing mechanisms operate under normal constraints. In addition, recent reports of congenital prosopagnosia suggest it is possible to have a specific deficit in face perception (Behrmann & Avidan, 2005); additional evidence hints that there may be a genetic basis to this disorder. In this general view, one would predict that mechanisms of object recognition could be spared even in the context of severe deficits in other areas of visual-spatial cognition.

In a different view, genetic deficits are also assumed to have widespread local effects on the development of the brain, but these effects are thought to propogate up to cognitive structure, as follows. The genetic deficit is assumed to result in impaired cognitive mechanisms. Interactions between these impaired mechanisms and particular knowledge domains will yield differences in cognitive structure. Moreover, the cognitive structures that are produced will not necessarily (or even probably) show breakdown along the lines of a normal, mature architecture (Karmiloff-Smith, 1998). Although one might observe performance that appears similar to a normal profile, closer inspection will likely show meaningful differences in the underlying cognitive structures. For example, Karmiloff-Smith, Thomas, Annaz, Humphreys, Ewing and Brace (2004) argue that face processing in people with Williams syndrome is not normal, but rather, good performance is accomplished via atypical mechanisms; specifically, they argue that configural processing of faces in WS people is impaired. This general position is consistent with the idea that, despite the distinct computational requirements of different cognitive domains, genetic deficits will likely result in subtle cognitive impairments, including the recognition of objects.

These strong positions can guide our thinking, but only empirical data can decide between them. Perhaps more importantly, empirical data can provide us with the foundation for developing more nuanced theoretical positions.

3. Williams syndrome and object recognition

The case of object recognition in Williams syndrome provides an empirical forum for doing both of these. Williams syndrome (WS) is a rare genetic disorder (1:20,000 live births) which is caused by a hemizygous submicroscopic deletion on chromosome 7q11.23. Diagnosis is made on the basis of a unique phenotypic pattern that includes a characteristic facial profile, disorders of the heart, and anomalies of the viscera. It can also be verified by a screening technique (fluoride in situ hybridization, FISH) that isolates the key region of gene deletion (Ewart, Morris, Atkinson, Jin, Sternes and Spallone, 1993; Frangiskakis, Ewart, Morris, Mervis, Bertrand and Robinson, 1996; Morris, Ewart, Sternes, Spallone, Stock and Leppert, 1994).

Of most importance to us, however, is the distinctive cognitive profile of individuals with Williams syndrome, who show severely impaired spatial cognition together with relatively spared language (Bellugi et al., 1988; Mervis, Morris, Bertrand & Robinson, 1999). Although WS individuals are also moderately mentally retarded (Mean Composite IQ=55–60, Mervis, et al., 1999), their unique pattern of spatial deficit and linguistic strength sets them apart from other groups with comparable retardation, such as Down syndrome (Mervis et al., 1999). The striking cognitive profile has also motivated some of the strongest hypotheses of developmental modularity in the field (Bellugi et al., 1988; Pinker, 1994).

Selective sparing in cognitive systems can also be tested by examining differential breakdown *within* the broad domain of spatial representation. Growing evidence shows that spatial representation is not one monolithic system, but rather, is composed of different sub-systems that are specialized in their computational goals, their functional properties, and their neural substrates. One example is the contrast between perception of space and representation of space for action, as described previously. Other functionally distinct systems include the representation of space for navigation (Aguirre, Zarahn & D'Esposito, 1998; Gallistel, 1990; Hermer & Spelke, 1996; Newcombe & Huttenlocher, 2000), the multiple representations of space that encode location in different reference

systems (Andersen, Snyder, Bradley & Xing 1997; Colby & Goldberg, 1999), and the representation of different kinds of motion such as motion coherence, biological motion, and form from motion (Schenk & Zihl, 1997; Vaina, LeMay, Bienfrang, Choi & Nakayama, 1990).

Although the nature of spatial impairment in WS is not well understood at present, there are indications that it has 'peaks and valleys' (cf. Bellugi et al., 1988), raising the possibility of selective targeting. The most widely observed hallmark of the spatial deficit appears in so-called 'visual-spatial construction' tasks, in which people are asked to replicate an overall pattern by drawing or assembling parts. Individuals with WS perform extremely poorly in such tasks, with adolescents performing in the 1st percentile—roughly at the level of a normal four-year-old (Bellugi et al., 1988; Mervis et al., 1999; see Fig. 1 for some examples). In these tasks, the deficient performance of WS children can be traced to faulty spatial representations of the individual blocks and their relations rather than faulty executive processes (Hoffman, Landau & Pagani, 2003). In particular, children with WS have trouble discriminating the 'handedness' of the individual blocks which are often split in half by color, and have distractors that are mirror-images. Additionally, WS children make errors in the spatial arrangement of blocks, often erring on left-right locations within the model. This raises the possibility that object representation more generally—and not just mirror-image structures—might be impaired.

On the other hand, growing evidence suggests differential breakdown across other domains that involve spatial organization of elements. For example, the perception of biological motion and motion coherence is spared in WS children (Jordan et al., 2002; Reiss, Hoffman & Landau, 2005), even though perception of form from motion is impaired (Atkinson, King, Braddick, Nokes, Anker & Braddick 1997; Reiss et al., 2005). Aspects of visual-manual action appear to be impaired, even relative to mental age matched children (Atkinson, King, Braddick, Nokes, Anker and Braddick, 1997; Dilks, Landau & Hoffman, 2005). Some mechanisms of global spatial perception are unimpaired (Pani, Mervis & Robinson, 1999) and there is evidence that face perception is spared, even relative to chronological age matches (Tager-Flusberg et al., 2003). As a whole, these findings are consistent with the idea that a genetic defect can result in targeted—rather than omnibus—spatial breakdown.

4. The current experiments

The case of object recognition affords a further test of this possibility. First, it is a natural domain that presents a unique computational problem: Using patterns of light striking the retina, the brain must construct a representation that will enable the perceiver to later recognize the same object, despite changes in viewpoint and lighting that occur in natural viewing conditions. This is a task that is easily solved by human observers—even infants—but cannot yet be solved by machines. Second, evidence from cases of brain damage in adults already suggests specialization of function: People may lose the ability to perceive objects, while still being able to act on them or reason about them. Third, the object recognition system seems to be vulnerable to a variety of effects of degradation under brain damage. For example, poor lighting or line drawings produces special

difficulties for agnosics (Farah, 2000), and highly unusual viewpoints produce difficulties in patients with damage to the right parietal lobe (Warrington & Taylor, 1973). This welldocumented pattern of vulnerability within the object recognition system suggests a way to examine in detail the degree to which object recognition is spared or impaired. Recognition of objects under degraded conditions, such as poor lighting, unusual viewpoint, or line drawings would provide a strong test of sparing, but it is also possible that some conditions might prove substantially more difficult than others, providing evidence for both sparing and breakdown.

Therefore, in our first experiment, we examined the capacity of children with WS to identify objects under degraded conditions. We used two critical manipulations: Presentation of clear vs. blurred images, and presentation of objects from canonical vs. unusual viewpoints. These two methods of image degradation can be used to drive down performance of all groups—including normal children and adults—thereby allowing us to examine whether patterns of failure are similar or different between WS and normal individuals. But the manipulations are also of more particular interest with respect to Williams syndrome.

First, blurring the image is of interest because initial characterizations of the spatial deficit in WS suggested that they are 'local processors' (Bihrle, Bellugi, Delis & Marks, 1989; Deruelle, Mancini, Livel, Casse-Perot & de Schoon, 1999), i.e. they correctly perceive the features of an object but are deficient in grasping the global configuration of those features. More recent work (Farran, Jarrold & Gathercole, 2003; Pani, Mervis & Robinson, 1999), however, indicates that people with WS do correctly perceive configurations and actually have trouble focusing attention on parts that are members of larger configurations (Hoffman, Landau, and Pagani, 2003). Blurring an image would primarily affect the visibility of constituent parts (Hughes, Nozawa, and Kitterle, 1996; Morrison and Schyns, 2001) while leaving the global shape relatively unaffected. Therefore, to the extent that people with WS utilize global shape to recognize objects, they might be *less affected* by blur than control children who may use both local and global information for identification.

The second, and more crucial manipulation is canonical vs. highly unusual viewpoints. Recognizing objects from highly unusual viewpoints might present a special problem for people with Williams syndrome. Perrett, Oram & Ashbridge (1998) suggest that viewpoint effects on recognition can be understood in terms of a multiple view theory of object recognition in which the current view is matched to multiple views that are stored in inferotemporal cortex that have been laid down by previous experiences with that object. Novel views do not have a matching representation and must be recognized by partial activation of nearest neighbor views, which will be slower than matches for canonical views.

This theory, however, does not directly account for findings with highly *unusual views*. Unusual views are taken from a perspective that foreshortens the principal axis of the object and often occludes many of its salient features (Humphreys & Riddoch, 1984). This precludes the kind of automatic object recognition that seems to occur with more canonical views and instead, appears to involve a 'problem solving or executive component' (Farah, 2000) in which the observer searches the image for parts that may provide cues to its identity (Perrett et al., 1998). Given the role of the parietal lobe in

directing spatial attention (Belmonte & Yurgelun-Todd, 2003), it is perhaps not surprising that patients with damage to parietal areas are selectively impaired in recognizing these views (Warrington & Taylor, 1973). The claim that recognition of unusual views requires executive control is also consistent with the finding that in dual-task experiments, a task requiring 'central executive resources' (random number generation) produced more interference with unusual views than canonical views (Baragwanath & Turnbull, 2002). Finally, recognition of unusual views has been found to activate both prefrontal (Kosslyn, Alpert, Thompson, Chabris, Rauch and Anderson, 1994) and parietal areas (Sugio, Inui, Matsuo, Matsuzawa, Glover and Nakai, 1999) in fMRI scans, suggesting top-down control of parietal areas involved with spatial attention.

The role of the parietal lobe in recognition of unusual views suggests that WS subjects may find such views particularly challenging. Damage to the same parietal areas activated in recognition of unusual views often results in impairments in tasks such as drawing and block construction (Benton, 1967; Turnbull, Denis, Mellet, Ghaem & Carey, 2001)—just those tasks that WS people typically fail. In addition, recent neuroimaging of people with WS during performance of these spatial tasks shows a reduced activation of these parietal areas (Meyer-Lindenberg et al., 2004), and other analyses show than WS people have smaller than normal volume in superior parietal areas (Eckert, Hu, Eliez, Bellugi, Galaburda and Korenberg, 2005). These observations are consistent with the suggestion by Atkinson et al. (2003) that WS (and possibly other syndromes) is primarily a dorsal stream deficit. People with WS might therefore be expected to have a selective deficit in identifying objects from highly *unusual* viewpoints, even if their identification from *canonical* viewpoints is not deficient.

5. Experiment 1

5.1. Participants

Twelve children with Williams syndrome (mean age 11;0, range 7;4 to 15;3 years), 12 normally developing children who were mental-age matches for the WS group (Mean Age 5;8, Range 4;1 to 7;1 years), 12 normally developing children who were chronological age-matches for the WS group (Mean Age 11;11, Range 10;6 to 14;3 years) and 12 undergraduates participated. Children with WS were identified through the National WS Association, and had been positively diagnosed by a geneticist; all but one had also been diagnosed by the FISH test (the remaining person did not undergo the test). Normal children in the MA group were matched individually to children with WS, using the Kaufman Brief Intelligence Test (Kaufman & Kaufman, 1990), which yields a verbal and non-verbal (Matrices) score. The latter does not have many spatial items, and hence does not unfairly penalize WS children for their spatial impairment. The mean scores for the WS children were Verbal = 31.7 (SE = 2.32), Matrices = 18.58 (SE = 1.11); corresponding scores for the MA-matched controls were 29.33 (SE = 2.33) and 18.25 (SE = 1.52). Scores for the CA group were Verbal = 59.6 (SE = 1.46) and Matrices = 38 (SE = 0.99). The mean IQ scores for the three groups were 71.42 (SE=4.35) for the WS children, 116.2 (SE= 4.11) for the MA group, and 123 (SE = 2.50) for the CA controls.

In addition, the WS children and ten of the MA controls were tested on the Pattern Construction Sub-test of the Differential Abilities Scales (Elliot, 1990), which requires children to replicate a design using individual component blocks, and is the hallmark test used to diagnose the WS spatial impairment. The scores for the WS children were M= 81.83, percentile=1.83, SE=5.82; scores for the MA controls were, M=109.2, percentile=58.1, SE=6.96. The WS scores are in the range reported by other investigators (see Mervis et al., 1999). All but two WS children fell into the 1st percentile of performance, thus conforming to the reported pattern of severe spatial deficit. Note that, although the WS children were matched to normally developing children on verbal and non-verbal scores (the KBIT), they performed much worse than their MA matches on the Pattern Construction task, as would be expected. All participants signed informed consent forms.

5.2. Design, stimuli, and procedures

Participants were asked to name each of a set of 80 full color pictures of objects which were presented on a computer screen for 500 ms. The objects were drawn randomly from a set of 320 images consisting of 80 objects (listed in Table 1) in each of four conditions: (a) canonical view, clear image, (b) canonical view, blurred image, (c) unusual view, clear image, and (d) unusual view, blurred image. Canonical views were deemed those that exposed all of each object's relevant parts; many were full-face views of the object. Unusual views varied in their orientation, and included foreshortening along the primary axis, and selection of views from above or below the object. Blurred images were created by editing the clear images using the Gaussian Blur tool (radius 10) in Photoshop. Objects were drawn from two sources: The 'Object DataBank' available from Michael Tarr (http://www.cog.brown.edu/~tarr/stimuli.html) and the model set which accompanied Ray Dream Studio 5.5. Examples of objects in each of the four conditions are shown in Fig. 2 and the complete set of images can be viewed at http://hoffman.psych.udel.edu/ ObjectPicturesForWeb.pps.

Each participant saw 80 different objects with an equal number of objects presented in each of the four conditions. All objects were randomly ordered over eight lists, and individual WS subjects were assigned the same list as their controls (both MA and CA). Across lists, each object was represented equally often in each of the four presentation modes. Responses were recorded verbatim. If the participant was uncertain, the experimenter encouraged him or her to 'give your best guess'.

Responses were coded by a person who was 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 creating the objects; see Table 1) and the image of the object that the participant had viewed when producing the name. The rater used seven categories, including correct name, correct definition or use, related member of same basic category, correct superordinate category, similar shape, incorrect name, or do not know. For example, if a picture of a sunflower was shown, responses would be coded as follows: 'sunflower' (correct name), 'goes in a vase' (correct use), 'daisy' (related member of same category), 'plant' (correct superordinate), 'clock' (similar shape), 'truck' (incorrect name),

List of objects used in Experiments 1 and 2	
Anchor	Jet plane
Banana	Key
Barn	Kite
Basket	Motorcycle
Baseball bat	Mug
Bed	Pad lock
Bee	Pen
Belt	Piano
Bike	Pitcher
Binoculars	Plant
Blender	Pliers
Bottle	Pot
Bow (archery)	Pretzel
Bridge	Pumpkin
Brush	Refrigerator
Bulb (light bulb)	Ring
Cannon	Rollerblades
Carrot	Ruler
Cassette tape	Scissors
Castle	Screwdriver
Chain	Shovel
Chair	Sink
Clock	Sports car
Couch	Stool
Crayon	Stove
Cup	Sunflower
Desk	Table
Dresser	Tank
Drums	Tennis racket
Egg (fried)	Toothpaste tube
Eye glasses	Trash can
Electric fan	Truck
Fork	Trumpet
Frying pan	Turkey
Glass	Umbrella
Grill	Violin
Guitar	Wagon
Hamburger anger	Watch
Hair dryer	Windmill
-	

Table 1 List of objects used in Experiments 1 and 2

or 'do not know'. A second rater coded 20% of the responses, and reliability was 90%. Where there was disagreement, the first rater's data were used.

5.3. Results and discussion

An initial analysis examined the accuracy of participants' labels. Responses in the first four categories were considered to be correct. Any other response was scored as incorrect. Naming accuracy (percent correct) as a function of image condition and group is shown in

	Canonical Clear	Unusual Clear	Canonical Blurred	Unusual Blurred
Blender		ICO	Ŵ	IK.
Cup		0		•
Bottle		١	l	8
Stool	A		甲	

Fig. 2. Examples of objects used in each condition of Experiment 1

Fig. 3 (shown as a line graph to emphasize patterns of interaction and additivity between groups and viewpoint). Fig. 3a shows data for the clear image condition and Fig. 3b shows data for the blurred image condition.

First, it is apparent that we were effective in our manipulation of the difficulty of identifying the objects. For both clear and blurred images, participants were more accurate in naming objects shown in a canonical than in an unusual orientation. Clear images were identified more accurately than blurred ones. These effects of image quality were confirmed by significant main effects of Orientation, F(1,44)=344.7, P<0.001 and Clarity, F(1,44)=384.5, P<0.001. In addition, there was an Orientation by Clarity interaction, F(1,44)=84.7, P<0.001 indicating that the effects of combining the two distortions was greater than the additive combination of each distortion in isolation.

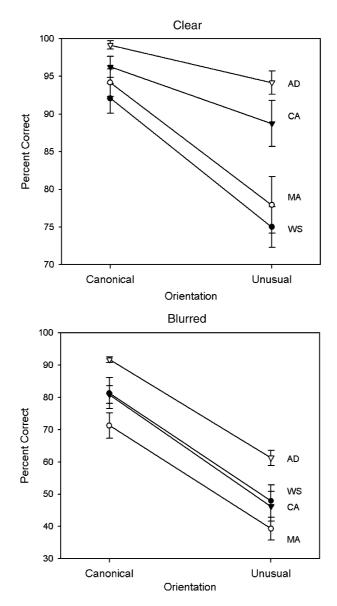


Fig. 3. (a) Experiment 1: Clear Images. Mean percent correct (S.E.) over condition and group (Ad: Adults; CA: chronological age matches; WS: Williams syndrome; MA:mental age matches). (b) Experiment 1: Blurred Images. Mean percent correct (S.E.) over condition and group (Ad: Adults; CA: chronological age matches; WS: Williams syndrome; MA: mental age matches).

Across these data, the main effect of Group was significant, F(3,44) = 8.8, P < 001, but this was due to the adults' superior performance. Tukey post-hoc tests showed no significant differences among the three children's groups (WS vs. MA, P=0.72, WS vs. CA, P=0.62, and MA vs. CA, P=0.12). The adult group, however was more accurate

than both the WS group (P < 0.002) and MA controls (P < 0.001), and marginally better than the CA controls (P < 0.056). In addition, there was a Clarity X Group interaction (F(3, 44) = 5.66, P < 0.002. To isolate this interaction, we analyzed the difference between clear and blurred images as a function of group. Tukey tests revealed that the WS group showed smaller effects of blur than either the MA or CA controls (P < 0.01 and 0.03, respectively) and were indistinguishable from adults (P > 0.98).

The data in Fig. 3 also suggest that the effects of viewpoint and image clarity depended on group. In order to evaluate the effects of viewpoint, we analyzed the data separately for clear and blurred images. For clear images (Fig. 3a), there is a striking similarity between the CA and adult groups as well as between the MA and WS groups. The adults and CA controls were more accurate across the two viewpoints (Ms=0.96, 0.93) than the other two groups (Ms=0.86, 0.83) and also showed a smaller effect of viewpoint. An analysis of variance on these data revealed a main effect of Group (F(3, 44)=6.61. Tukey tests on the Group effect revealed that the WS group was less accurate than both the CA and adult groups (P<0.05 and 0.001, respectively) but did not differ from MA children (P=0.87). The MA group only differed from the adults (P<0.01).

There was also a significant Group X Viewpoint interaction (F(3,44) = 5.62, P < 0.002). Planned comparisons of the three children's groups showed that the effect of orientation was larger for WS compared to CA, t(22) = 2.38, P < 0.02 and MA compared to CA, t(22) = 2.37, P < 0.02; The WS and MA groups did not differ, t(22) = 0.18, P > 0.85. It is possible that the smaller effect of view in the CA group is due to their performance being at ceiling in the Canonical condition. However, this seems unlikely because performance of all three groups is quite good in this condition, with all means above 90% (Ms = 0.99, 0.96, 0.94, 0.92 for adults, CA, MA, and WS groups). A separate ANOVA on these data revealed no significant difference among groups, F(2,33) = 1.41, P > 0.25.

Blurring the image produced a different pattern of results (Fig. 3b). The WS subjects show a relative improvement in their standing compared to the other groups, and this appears to be mainly attributed to the larger impact of blur on the CA controls. In fact, the WS group is now indistinguishable from the CA group. In addition, blurring resulted in similar effects of orientation for all groups. There were main effects of Group (F(3,44)=9.07, P<0.001 and Orientation (F(1,44)=280.41, P<0.001 but no interaction (F<1). Tukey post hoc tests showed that the main effect of group was due to the adults performing more accurately than all three groups of children (P's < 0.05) which were not different from each other. (WS vs. MA, P=0.12, WS vs. CA, P=0.99, and MA vs. CA, P=0.20).

These results show two main patterns. First, the WS children have very high performance for canonical views and clear images and their performance drops for unusual views. Their pattern of accuracy across all conditions is remarkably similar to MA controls and the performance of both of these groups is worse than that of normal adults. The CA group appears to occupy an intermediate position. For canonical views of clear images, they are no different from WS children; for unusual views, they show less impact of viewpoint, with performance better than both WS children and MA controls.

Second, once images were blurred, the WS children perform as well as the CA children. In fact, all three groups of children showed comparable accuracy and all groups, including adults, showed the same relative decline in performance with unusual views. Why the different effects of blur? Showing *clear* objects from unusual viewpoints impaired performance in all of our groups, and in this case, the CA and adult controls were less affected by unusual views than the WS and MA groups. This advantage disappeared when images were blurred, however, suggesting that a key ingredient in deciphering unusual views may be related to analysis of internal features which is weakened by blurring. This result is consistent with results reported by Lawson & Humphreys (1999) who studied the effects of foreshortening of the main axis of an object presented as either a line drawing or a silhouette. Both line drawings and silhouettes preserve the 'occluding contour' of the object and therefore allow the observer to extract the object's main axis of elongation. Silhouettes, however, eliminate the possibility of using internal details and features for identification. Their results showed that extreme foreshortening, similar to the unusual views in Experiment 1, reduced identification accuracy for both kinds of objects but particularly for the silhouettes, suggesting that internal features become particularly important clues to object identify when axis information is no longer available.

The greater effect of unusual views on MA and WS subjects compared to CA controls in the clear condition suggests that the latter group is better at utilizing the internal details of objects to identify them from unusual viewpoints. We might expect then that if these features were more difficult to perceive—as in the blur condition—the CA's advantage would disappear and that is what we observed. When objects were blurred, all three groups of children were comparable. Interestingly, of the three groups of children, the WS group was the least affected by image blur suggesting that they may rely on the occluding contour of the object more than control children, even for clear images. This is consistent with other evidence suggesting that people with WS may have trouble focusing attention on subparts of a larger pattern. For example, Hoffman et al. (2003) reported that in a variation of the block construction task, WS children were unable to ignore nearby blocks when they were cued to attend to a single block in the model.

The foregoing evidence relies on measures of accuracy, and thus might miss some finegrained qualitative differences in the way that WS children label objects. Accordingly, we analyzed the distribution of responses across the seven response categories for the four groups (see Fig. 4). The number of responses in each response category for each subject was entered into a χ^2 test (SPSS Crosstabs). The only significant interaction of Group with a Response Category was for Correct Definition or Use (X^2 (15)=28.9, P < 0.02), reflecting a slightly greater tendency for MA controls to label pictures in terms of their definition or use. Otherwise, the distribution of responses across the various categories was similar for all four groups (χ^2 s p values ranged from 0.11 for the 'Do not Know' category to 0.68 for the 'Same Category' response).

A final set of analyses examined whether the four groups differed in which objects they found difficult to identify. We computed the average accuracy for each of the 80 objects, collapsed across the 4 image quality conditions, for each group. We then computed the correlation between all pairs of groups for accuracy on the 80 objects. The correlations were significantly greater than zero (P < 0.001 for all 6 correlations) and ranged from 0.67 (WS vs. adult groups) to 0.76 (WS vs. MA groups). None of the pairs of correlations showed reliable differences (all Ps > 0.17), suggesting that there was a high degree of similarity among all four groups in terms of which objects were easy or difficult to identify. We also asked whether object identification accuracy depended on object

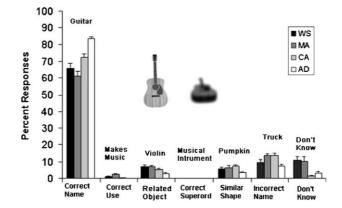


Fig. 4. Experiment 1: Distribution of response types (*M* percent, S.E.) over group (Ad: Adults; CA: chronological age matches; WS: Williams syndrome; MA:mental age matches).

complexity. Two independent raters rank ordered the entire set of 80 objects in terms of their complexity (correlation between raters =0.83). These complexity ratings were not reliably correlated with average accuracy of each object for any of the groups (correlations = 0.09, 0.10, 0.21, 0.17 for the WS, MA, CA, and adults, respectively, alpha = 0.05, two-tailed test). Both of these analyses suggest similarity across groups in the relative difficulty of individual objects.

As a whole, the results suggest that object recognition is surprisingly good in children with WS, certainly much better than might be expected on the basis of their performance in other areas of spatial cognition such as block construction. Indeed, on the basis of their standardized verbal scores, they were exactly where one would predict they should be— comparable to MA controls in every condition. From this perspective, it is surprising that they were no different from CA controls in identifying objects presented as clear canonical images, or those presented as blurred images in either canonical or unusual viewpoints. The WS performance is particularly striking, in light of the fact that recognizing objects under these circumstances requires that they match an image that they likely never encountered with a stored representation of the object. This matching likely involves comparing overlap between two shapes, even though the unusual orientations obscure aspects of the objects' shapes.

It would be tempting to conclude that WS children are capable of representing and extracting an object's part structure from partial information. However, because the stimuli were full color renditions of objects, it is always possible that object recognition in WS depends more on surface features (such as color and texture) than the object's spatial structure (e.g. Tanaka & Presnell, 1999; Tanaka, Weiskopf & Williams, 2001). Moreover, the fact that recognizing objects in unusual orientations can sometimes activate more parietal areas (Sugio et al., 1999) raises the question of whether a more difficult version of the task might reveal additional differences between WS children and the normal groups. In order to test this possibility, we carried out a second experiment, using line drawings of objects. In these cases, objects can only be recognized by their shapes, not by surface information such as color or texture.

6. Experiment 2

6.1. Participants

Children were the same as those who participated in Experiment 1, with the exception of one MA control child, who replaced a child who could no longer participate in our studies. The new mental age-matched group was still well matched by KBIT verbal (WS Mean = 33, S.E. = 2.08, control Mean = 33, S.E. = 2.26) and Matrices scores (WS Mean = 18, S.E. = 1.06, control Mean = 19, S.E. = 1.30). The DAS scores of the WS children had a Mean in the 1st percentile, while the MA matched controls' Mean was in the 55th percentile. All participants were tested after Experiment 1.

6.2. Design, stimuli, and procedures

These were identical to Experiment 1, except that the objects were converted to black and white line drawings (see Fig. 5). Only the clear versions were shown, resulting in a total of

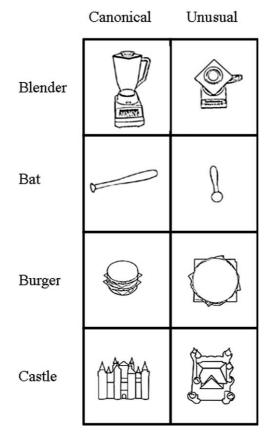


Fig. 5. Examples of objects used in both conditions of Experiment 2.

160 stimuli (80 objects X 2 views). Each subject saw all 80 objects with the view (canonical or unusual) chosen randomly under the constraint that there was an equal number of canonical and unusual views. Eight lists of objects in a random order were created and subjects were randomly assigned a list, matching WS children and their controls on the same list. Across the lists, each object was shown equally often from the two viewpoints.

6.3. Results and discussion

As in Experiment 1, responses were scored as correct if they were in any of the first four response categories (correct name, correct definition or use, related category member, or correct superordinate category); all other responses were scored as incorrect. Fig. 6 shows the percent correct for the four groups as a function of orientation (canonical or unusual). Consistent with what we observed in Experiment 1 using full color solid object images, identification was severely reduced when objects were shown in a unusual orientation, F(1,44) = 524.7, P < 0.001. In addition, there was a main effect of Group, F(3,44) = 20.9, P < 0.001) as well as an Orientation X Group interaction, F(3,44) = 7.3, P < 0.001 that reflects a similar pattern to the one we observed in Experiment 1 for the clear image condition. Post-hoc Tukey tests revealed two homogeneous subsets of the Group variable. WS and MA matches formed one set while the other set consisted of CA matches and Adults. Members of one subset differed significantly from each member of the other set, (P < 0.001) but not from each other. (WS vs. MA, P = 0.81; CA vs. AD, P = 0.78).

Separate analyses were also carried out for the canonical and unusual view conditions, with a similar result. For both conditions, the WS and MA matches formed one subgroup and the CA and AD subjects formed another. For canonical views, all groups were above

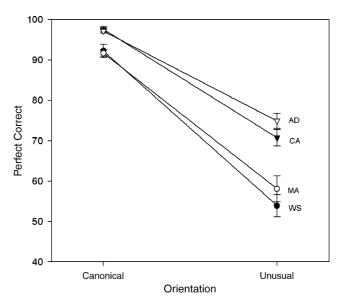


Fig. 6. Experiment 2: M percent correct (S.E.) over condition and group. (Ad: Adults; CA: chronological age matches; WS: Williams syndrome; MA:mental age matches).

90% accuracy (Ms=0.92, 0.92, 0.98, 0.97 for WS, MA, CA and adults respective). The WS and MA groups did not differ (P=0.97) nor did CA and AD groups (P=0.92). Members of each of the subgroups, however, differed from members of the other group (all P's < 0.003). For unusual views, WS and MA children were comparable, (Ms=0.54, 0.58, respectively, P=0.65) as were the CA and adult groups (Ms=0.71, 0.75, respectively, P=0.65). Members of each of the subgroups differed from members of the other groups (all P's < 0.001).

The Orientation X Group interaction was further analyzed using the difference between canonical and unusual views for each group. Post-hoc Tukey tests revealed that the WS group differed from the CA (P < 0.02) and AD (P < 0.001) groups but not from MA controls (P=0.59) and the MA group only differed from the Adults (P < 0.03). One might worry that the larger effect of orientation on WS subjects compared to CA controls is due to the CA controls being close to ceiling in the canonical view condition. However, the WS group was above 90% accuracy in this condition, as in the canonical clear condition of Experiment 1. Although the interaction should be viewed with caution, the findings here of the larger orientation effect among WS children than CA controls replicates the findings of Experiment 1. This suggests there is a real relative weakness for unusual orientations among WS children relative to CA, but not MA matched children.

These results show that when subjects must rely purely on 'shape' information to label objects, WS children are remarkably good at identifying objects—over 90% with canonical viewpoints. They are remarkably similar to the MA controls in identification accuracy, and this holds even when objects are shown in unusual views that produce substantial deficits in the performance of normal adults. These results are similar to the clear image condition of Experiment 1 which showed that CA subjects were more accurate than WS subjects and less affected by unusual views.

Once again, we analyzed the distribution of different error types to determine whether the four groups differed in the kinds of errors they made. These data are shown in Fig. 7

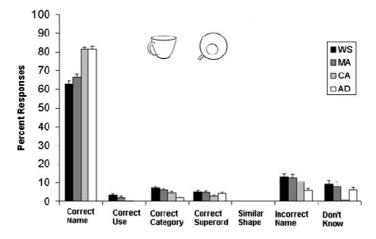


Fig. 7. Experiment 2: Distribution of responses types (*M* percent, S.E.) over group (Ad: Adults; CA: chronological age matches; WS: Williams syndrome; MA:mental age matches).

and suggest that all four groups have roughly similar profiles across the different response categories. The number of responses in each response category for each subject was entered into a χ^2 test (SPSS Crosstabs) which resulted in significant interactions between Group and the following categories: Correct name (Pearson X² (81)=105.6, P < 0.05), Correct Definition or Use (X^2 (15)=31.3, P < 0.01), Related Category member (X^2 (27)= 55.9, P < 0.01). The interaction with the 'Do not Know' category was marginally significant (X^2 (48)=64.2, P < 0.06). This pattern of significance reflects the higher use of correct names for objects in the CA and Adult groups compared to WS and MA children and may partly reflect more sophisticated vocabularies (as shown by their KBIT scores) or relatively less difficulty in retrieving the correct names. When this analysis was restricted to just the WS and MA groups, none of the interactions approached significance (all P's> 0.14), suggesting that these two groups has a similar profile of responses across the various categories.

A final analysis examined whether groups differed in terms of which objects they found difficult to identify. Here we were only able to compare the WS and MA groups because accuracy for many individual objects was at 100% for the CA and AD groups. We computed the average accuracy for each of the 80 objects, collapsed across the 2 orientation conditions. The correlation between the WS and MA groups across the 80 objects was 0.74, which is significantly different from 0, (t(78)=9.76, P<0.001), suggesting that WS children and their MA controls were similar, not only in their overall accuracy of identification but also in terms of which objects they found easy or difficult to identify. We also examined whether accuracy depended on the rated complexity of the objects for these two groups. For WS children, the correlation was 0.13 and for the MA group, it was 0.14, neither of which was significant (WS: t(78)=1.16, P>0.05 and MA: t(78)=1.25, P>0.05).

It is important to note that the WS and MA groups, who were relatively less accurate than the CA and adult groups, were nonetheless quite accurate in this task in an absolute sense. On average, they correctly recognized approximately 74% of the line drawings compared to 85% for the CA and AD groups. Even in the most difficult condition, in which objects were shown in unusual orientations, WS and MA children correctly recognized 54 and 58% of the line drawings, respectively. This is quite high, given that the random probability of guessing the correct object name is far lower than 1 in 40, which would be chance in the extremely unlikely event that the subject knew the database for the objects. Some have conjectured that identifying objects under highly unusual viewpoints may require a form of cognitive problem-solving, as the visual system may not be designed to automatically compute these extreme situations (Farah, 2000). Under the circumstances of severe degradation (line drawing and unusual orientation), this requirement may have been accentuated. If so, the difference between groups is understandable.

Comparison with the results of Experiment 1 shows that removing color and texture cues impaired subjects' performance, with the largest decrement seen when objects were presented in an unusual view. This is consistent with the notion that multiple mechanisms may be at work in recognition of unusual views and that other cues to object identity, such as color or texture, become particularly important when shape information becomes sufficiently degraded (Farah, 2000). In order to see whether the decrement from unusual views was larger for line drawings than for full-color objects, an additional analysis of

variance was conducted comparing the corresponding conditions across the two experiments (i.e. the data from Experiment 2 vs. those from the clear image condition of Experiment 1). In this analysis, the Group X Experiment X View interaction was not significant (F(3,44) < 1), suggesting that the performance decrement associated with unusual views for each group was similar for solid full color objects and line drawings.

7. General discussion and conclusions

The present experiments used full color pictures and line drawings of common objects to examine object identification in children with WS compared to normally developing children of the same mental (MA) and chronological age (CA). The difficulty of object identification was manipulated by blurring objects (full color objects) and portraying them in unusual orientations (color objects and line drawings). Overall, WS and MA children were remarkably similar under all conditions, both in terms of accuracy and in various fine-grained details of performance such as which objects they found easy or difficult to identify. Moreover, the WS children were close to CA matches in their identification of objects presented in canonical orientations, with differences of 4% to 6% (Experiments 1, 2, respectively). This strong performance by WS children in object recognition stands in marked contrast to their deficits in certain other spatial tasks. For example, in block construction (Hoffman et al., 2003), multiple object tracking (O'Hearn, Landau & Hoffman, 2005), and visual-manual action tasks (Dilks, Landau & Hoffman, 2005; Atkinson et al., 2003), they perform reliably worse than MA controls. The combination of strength and weakness shows that their spatial deficit is highly selective.

In contrast to their excellent performance for canonical objects, both WS children and MA matches were less accurate than the CA group when viewing clear image objects shown in unusual views—whether they were presented as line drawings or full-color objects. Previous research suggests that identifying objects in unusual views emphasizes the use of internal features over other sources of identity information such as global configuration, color, texture, etc. (Lawson & Humphreys, 1999). Consistent with this, we found that blurring the colored objects eliminated the advantage of the CA group, resulting in complete comparability of the WS children to their CA matches in this condition. Again, this finding of strength stands in stark contrast to the WS pattern of weakness in other kinds of spatial tasks. Indeed, given the large difference between the WS children and their CA matches on the standardized vocabulary measure (KBIT), and, of course, on their IQ measures (Ms of 71 vs. 123), it is quite surprising the WS children would perform as well as CA matches in these contexts.

These results suggest the following characterization of object identification processes in WS. First, children with WS perform close to ceiling in identifying either full color or line drawings of objects in canonical orientations. This suggests that some of the most basic mechanisms of object recognition are intact in children with WS.

Second, children with WS show developmental delay or arrest compared to CA controls in their ability to identify objects in unusual orientations. In these conditions, they are indistinguishable from younger, normally developing children of the same mental age. This suggests that, relative to CA matches (but not MA matches), the unusual orientation

condition is disproportionately difficult for WS children. The difference between CA and MA matches further suggests that the ability to recognize objects under unusual orientations increases between the ages of 6 and 12 in *normally* developing children, pointing to the possibility that recognition under unusual orientation engages different mechanisms from recognition under canonical orientation.

Third, showing objects in a blurred format decreased object identification for all participants, but of the three groups of children, it had the least impact on the WS group. We speculate that WS children may rely more on 'global' or low frequency information to identify objects, even when they are presented in clear format, so blurring has less of an effect. A weakness in attending to internal detail in WS participants is consistent with other findings as well. For example, Pani et al. (1999) found in a visual search task that WS participants had more difficulty than controls in switching from a global level of organization to a local one. Similarly, Hoffman et al. (2003) found that WS children, unlike MA controls, continued to process to-be-ignored objects even when they were physically segregated from a cued target object.

The overall pattern suggests both sparing and compromise in Williams syndrome across different mechanisms of object recognition. Canonical object recognition in WS appears to be spared in the sense that performance was close to ceiling, indistinguishable from MA controls and, although statistically different from CA controls, very close in an absolute sense. The qualitative similarity to the control groups further emphasizes that the WS children are not likely to be carrying out the task in some different manner, using different strategies to recognize the objects.

In contrast, the results for unusual viewpoints suggest a much weaker sense of sparing, and the possibility that some of the mechanisms engaged for this kind of recognition reflect breakdown in WS. Here, the WS children also showed strength in object recognition, with performance far exceeding chance, and with *qualitative* similarity to normal CA matches and adults. However, their performance decrement relative to canonical views was greater than that of CA matches or adults, though it was the same as that observed among MA matched children. This suggests that the specific mechanisms used for recognition under unusual perspectives may be somewhat impaired relative to chronological age, but not mental age. This contrasts strongly with the pattern for the hallmark spatial deficit (i.e. the DAS block task) where our WS participants performed at the level of four-year-olds, and shows that object recognition—even under the difficult conditions of unusual viewpoint—is a distinct strength.

The general finding that unusual viewpoints are relatively difficult echoes a robust pattern found in the object recognition literature (Palmieri & Gauthier, 2004). The appearance of this pattern among all of our groups suggests that there is a similar functional division between known viewpoints and highly unusual ones across the groups. This conjecture of similarity in architecture is further supported by the detailed findings of similarity across groups in response types and in the particular objects that people found more or less difficult to identify. Over objects, there was considerable variation in part structure, and in occlusion of parts under unusual viewpoints. It is not surprising, therefore, that there were differences in how difficult or easy the objects were to accurately identify. However, to the extent that these factors mattered, they appear to have mattered to the same degree for all groups.

How can we explain the finding of special difficulty for WS and MA matches for unusual orientations? The relative difficulty of unusual viewpoints among WS individuals is significant for several reasons. First, some studies have found that unusual viewpoints tend to activate more parietal regions of the brain (Sugio et al., 1999), and people with parietal lobe damage can have difficulty recognizing objects from these viewpoints (Warrington & Taylor, 1973). The relative difficulty among WS children of identifying unusual viewpoint objects is consistent with the hypothesis that WS is primarily a dorsal stream, or a parietal lobe deficit that may also interact with frontal lobe executive functions (Atkinson et al, 2003; Reiss et al., 2005). Although several studies of object recognition in adults and monkey have found some sensitivity to viewpoint invariance in the IT cortex (Booth & Rolls, 1998), the bulk of current research suggests that this part of cortex computes viewpoints close to experienced ones (Palmieri & Gauthier, 2004). The viewpoints that we presented were quite unusual and in some cases rather extreme (e.g. cheeseburger viewed from directly above, a stool from directly underneath). Identifying objects under such conditions may require visual attention to segment and search for diagnostic object parts (Perrett et al, 1998) which would require participation by the parietal areas of the brain, even if object recognition is in most cases carried out by the ventral stream. If so, the similarity of the WS children to their mental age matches would suggest that these parietal functions are not normally mature until after 6 years of age, and that in WS, they do not mature until adolescence or later, if at all. Interestingly, the same selective deficit in recognizing unusual views is seen in older subjects (Dror & Kosslyn, 1998) and may be due to the degradation in frontal lobe functioning that occurs with age.

The strengths in the object recognition system in children with Williams syndrome provide another piece of the puzzle concerning the nature of their spatial deficit. Previous findings have shown that there is strength in face recognition (Bellugi et al., 1988; Tager-Flusberg et al., 2005), perception of biological motion and motion coherence (Jordan et al., 2002; Reiss et al., 2004), and certain aspects of spatial language (Lakusta & Landau, 2005; Landau & Hoffman, 2005; Landau & Zukowski, 2003). The present results suggest that fundamental mechanisms of object recognition are also a distinct strength in WS, but that recognizing objects from highly unusual viewpoints is either delayed or permanently impaired. We emphasize that this delay or impairment is quantitative rather than qualitative, as the details of error patterns were highly similar across groups.

The success seen in identifying common objects in the present studies contrasts sharply with WS children's performance in tasks of spatial construction (Bellugi et al., 1988; Hoffman, Landau & Pagani 2003; Mervis et al., 1999). Impairment in these tasks appears to be due to impairments in at least two aspects of spatial representations—mirror image structure of individual blocks and spatial relationships *between* blocks (Hoffman et al., 2003). For example, children with WS perform much more poorly than mental-age matched children when they must match blocks that are split by color to their identical mates in the context of blocks that are mirror images of the targets. Errors in this task reveal that WS children have great difficulty in retaining block 'handedness'. Importantly, the object recognition system is relatively insensitive to such handedness (Baylis & Driver, 2001), which is more reliably encoded by more parietal functions (Priftis, Rusconi, Umilta & Zorzi, 2003), suspected to be part of the WS deficit. Given the importance of being able to identify the same object *across* varying viewpoints, it seems plausible that

success at object recognition can co-exist with impaired representation of spatial relationships such as mirror-image relationships.

The emerging picture of strengths and weaknesses within the spatial representational systems has suggested to some that the WS spatial impairment is primarily a dorsal stream deficit (Atkinson et al., 1997; Brown, et al., 2003; Wang, Doherty, Rourke & Bellugi, 1995). Recent brain imaging evidence shows that people with WS show an absence of parietal activation during visual-construction tasks, an abnormally low level of parietal activation while matching complex geometric stimuli (Meyer-Lindenberg et al., 2004) as well as volume reduction in the superior parietal lobes (Eckert et al., 2005). The relative difficulty experienced by WS children in recognizing objects under unusual viewpoints is consistent with these findings. In addition, the strengths in object recognition found in our experiments are consistent with the idea that functions of the ventral stream-including face and object recognition and perception of biological motion-have the same cognitive structure as in normal individuals.

Overall, our results suggest sparing of the object recognition system, whose specialty is recognition of objects under canonical or neighboring—but not unusual—viewpoints. Over both experiments, performance of WS children was remarkably strong, and crucially, indistinguishable from normal people in its qualitative nature. The two areas of quantitative difference we found in WS children were reduced effects of blurring and increased effects of unusual viewpoint. We have speculated that the reduced effects of blur likely reflect a tendency to carry out object recognition by emphasizing global spatial structure over specific spatial features. But we emphasize that this is a matter of degree, which does not appear to change the fundamental nature or structure of the object recognition system.

The differences in unusual viewpoint point to a different matter. If recognition of unusual views engages the parietal areas, and these areas are less well-activated in people with WS, then this would explain their relative weakness in recognizing objects from unusual views. Once again, however, we emphasize that this is a matter of degree, since the WS children were comparable to MA matches, who are considerably younger. We speculate that *normal* developmental processes might result in more powerful parietal functions between the ages of 6 and 12, and if so, the weakness of WS children might reflect arrest of these functions at an early functional age of around 6.

Interestingly, the idea of differential sparing and damage over the ventral and dorsal streams in development is pleasantly consistent with two rather different ideas about the consequences of genetic deficit. It is consistent with the idea that genetic deficits have molecular effects on development, which could plausibly interact with timing to produce damage to different regions of the brain. But it is also consistent with the idea that cognitive systems with different computational and functional properties might undergo different degrees of sparing and damage due to developmental brain damage. Thus the findings contribute to our growing understanding of the complex cognitive consequences of genetic variation (see, e.g. Marcus & Fisher, 2003).

Most importantly, we believe, our findings suggest a functional separation between basic mechanisms of object recognition and other aspects of spatial representation, which we have here observed to be the consequence of a developmental and genetic impairment. The mechanisms by which such functional separation emerges are not known at present. However, the converging evidence suggesting both ventral and parietal mechanisms of object recognition shows that there may be ways for a genetic deficit to have local effects on brain development as well as more molar consequences for the development of different cognitive systems.

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