Specialization, Breakdown, and Sparing in Spatial Cognition:

Lessons from Williams syndrome

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Outline

Introduction

1.0 The Block Construction Task: Basic Facts about Performance

Introduction

It has been barely fifteen years since Williams-Beuren syndrome (WBS) first attracted serious attention from cognitive scientists interested in the architecture of the mind. The first reports of this rare syndrome offered it as a possible case of dissociation among cognitive systems responsible for language and spatial representation-- a possibility which could provide unusual insights into the organization of the mind and brain. If a genetically-based syndrome could result in selective, targeted impairment of some cognitive systems while leaving others intact, this would offer a first wedge into the ultimate problem of connecting gene and cognition.

The importance of this problem has been met by burgeoning research, which is moving quickly in new directions: As new insights are gained, the grounds for understanding cognition in WBS seem to shift. What began as a relatively simple hypothesis-- dissociation of two cognitive systems-- has developed into a set of hypotheses that address more closely whether and what kind of deficits exist in each system, and even whether the systems as a whole fall apart, or do so selectively. What seems clearest is that understanding cognitive systems in WBS-- like cognitive systems in normal individuals-- will require sustained, carefully controlled experimentation and creative theorizing to result in complete understanding of the deficit. This understanding should also shed light on the nature of normal development of mind and brain.

In this chapter, we report our recent findings, which confirm the initial notion of genetically targeted specialization. Yet these findings are tempered by the fact that the development of cognitive systems -- even development of those systems that are quite plausible candidates for specialization-- occurs at multiple levels and engages multiple mechanisms. Therefore, even one or two small missteps in the delicate sequence of developmental events can ultimately lead to cognitive performances that look quite different from the so-called "typical" profile shown by normally developing children. We believe that the critical issue is what these different performances reflect. Do they reflect a qualitatively different architecture that has necessarily evolved from altered genetic potential? Or do they reflect normal structure, which evolves from an architecture that is so highly constrained that it survives despite altered genetic potential? If the latter, then how do we explain obvious differences in performance?

In our view, close inspection of performance reveals a considerable degree of preserved cognitive structure. Differences in performance-- most often reflected in "worse" performance among WBS children than mental age-matched (MA-matched) children -- appear most strikingly when the task taxes general processes of memory and/or attention. In contrast, similarities in performance reflect intact cognitive architecture; there are often similarities in the qualitative nature of performance as well as overall performance, indicating that basic properties of cognitive systems are spared.

To us, a major key in understanding spatial cognition in Williams syndrome-- or, for that matter, in normal development-- is understanding what natural cognitive architectures are like, what functions they normally serve, and how we might engage them as directly as possible, without adding extra complexity extraneous to that system. When we examine spatial representation in this light, it becomes evident that much of its architecture is spared in Williams syndrome. This sparing reflects normal specialization of spatial cognitive systems that guides development, even under circumstances of genetic deficit.

In this chapter, we present data from our research program on spatial representation and spatial language that supports the notion that much of spatial architecture is spared in WBS. In particular, we will argue that this syndrome leaves spared a number of spatial cognitive sub-systems, including object recognition and identification, biological motion perception, and spatial language. At the same time, we will argue for the existence of complex, cascading effects of spatial cognition, which can lead from small, well-defined weakness to quite large deficits in performance. We will argue that these deficits are due *not* to abnormal architecture, but to small misadjustments that culminate in large downward spiraling performance. To illustrate these points, we will begin our chapter with the well-known profile of impaired performance in the so-called "block construction" task, often used as part of the clinical diagnosis of the Williams syndrome cognitive deficit.

1.0 The Block Construction Task: Basic Facts about Performance

The spatial deficit associated with Williams syndrome is particularly apparent in tasks that require the subject to reproduce a model by drawing it (e.g., the Test of Visual-Motor

Integration or VMI; Beery and Buktenica, 1967) or assembling parts (e.g., the Differential Ability Scales or DAS; Elliott, 1990; the Wechsler Adult Intelligence Scale-Revised or WAIS-R; Wechsler, 1981; see overview, this volume, by P. Wang). Figure 1 shows some typical drawings from our lab by children with WBS and a mental age-matched control. The WBS drawings generally preserve the colors and shapes of the component parts of the model but clearly fail to capture their global arrangement. Bellugi, Wang, & Jernigan (1994) reported similar results for the block construction task: WBS children tended to choose the correct parts but placed them incorrectly, producing "broken configurations" in which the outline shape of the completed copy did not match the model.

One attempt to explain these results proposes that people with WBS are "local processors" who perceive local details but have difficulty perceiving global structure. This is consistent with their broken configurations in block construction tests as well as their tendency to correctly draw the local level of hierarchical figures while failing to represent the global configuration—a pattern that has been described in several reports (Bellugi et al.,1994; Bihrle, Bellugi, Delis, & Marks, 1989). It is difficult, however, to unambiguously attribute errors in these tasks to a deficit in perception. Both drawing and block construction tasks require planning and working memory, and these processes could be responsible for a local bias, even if perception of global information is intact (Mottron, Belleville, & Menard, 1999). So the key to determining whether there is a deficit in perceptual processing of global spatial relationships, or in further complex operations that are carried out on the products of perception, is to dvise experiments that can separate the two.

One research group has indeed reported a local bias when WBS subjects were required to *match* two figures or judge their similarity, tasks that presumably are more diagnostic in terms of a perceptual representation (Deruelle, Mancini, Livel, Casse-Perot, & de Schoon, 1999). However, other research has failed to show a general deficit in global perception associated with WBS. For example, Key, Pani & Mervis (1998) found that broken configuration errors were relatively rare and occurred at about the same rate for WBS and control children. The most common errors involved choosing an incorrect part and placing it in an incorrect location. In addition, Pani, Mervis & Robinson (1999) found that in a visual search task, WBS were at least as sensitive as controls to global configuration effects. We showed that WBS children were comparable to MA-matched controls in the ability to see biological motion, a task that requires local element motion to be integrated into a global percept (Jordan, Reiss, Hoffman, & Landau, 2002). These results suggest that, at minimum, we will need further research to disentangle the causes of deficits in representing and/or storing global spatial information.

Aside from this, there is evidence that any difference in the representation of global spatial information is probably similar to that seen in younger, normally developing children. For example, our own analyses of copying in children with Williams syndrome indicates that, while the copies are quite abnormal for chronological age, they are qualitatively quite similar to those of normally developing children of roughly 3-4 years of age (Georgopolous, Georgopolous Kurz & Landau, 2002; Bertrand, Mervis, & Eisenberg, 1997). In our studies, the only qualitative difference in copying we observed was the tendency of WBS children (who were between 7 and 15 years of age) to create solid line figures when viewing a figure composed only of individual elements. Thus, for example, when copying a triangle made of small circles, our WBS children tended to draw a solid line triangle more often than did normally developing children matched for mental age. Interestingly, this observation goes against the idea that WBS children tend to be deficient "global processors"-- indeed, their tendency to draw the figure as a solid implies a bias *towards* global reconstruction.

The conflicting findings we have just mentioned need to be resolved, but at the very least, they suggest that we need to dissect tasks into component processes in order to attribute errors to particular processing stages. We will do this later in our chapter, when we return to the question of why block construction is so difficult. First, however, we will show that a close examination of different kinds of spatial representation—each fundamental in its own right--reveals considerable preservation in the spatial systems of people with WBS. We believe that this background of strength provides an important framework within which we can think about what goes wrong in Williams syndrome.

2.0 Specialization and Sparing of Spatial Cognitive Systems

We have examined several different kinds of spatial systems to determine whether the spatial deficit in WBS is global, or whether it shows selective breakdown along the lines of normal cognitive architecture. We here report on three components of this research.

2.1 Object Representation

One approach to understanding the patterns of preserved and impaired visual abilities in WBS depends on a distinction between the functions of the brain's dorsal and ventral visual pathways. Ungerleider and Mishkin (1982) originally proposed two systems in the mind and brain: The ventral system, thought to be responsible for representing the shape and identity of an object ; and the dorsal system, thought to represent location. More recently, Milner and Goodale (1995) have proposed that the dorsal stream is specialized for guiding action in space, and the ventral stream is responsible for conscious representation of the products of perception, which are often used in object and face perception.

The proposed functions of the dorsal stream are similar to those functions particularly impaired in WBS, which has led some to propose that WBS is a “dorsal stream deficit” (Atkinson et al., 1997; Wang et al., 1995). For example, at least anecdotally, visual-motor tasks such as reaching for and picking up small objects seem to be problematic for individuals with WBS. Additionally, other tasks that may involve the dorsal stream, such as manipulating elements of spatial arrays (e.g. object assembly, block copying, drawing) also show severe impairment (Bellugi, Bihrle, Neville, Doherty, & Jernigan, 1992; Mervis, Morris, Bertrand, & Robinson, 1999). Some have also reported deficits in certain kinds of motion perception (see Section 2.2), which have also been proposed to be dorsal functions. A natural question, then, is whether these apparent deficits reflect targeted damage to the dorsal stream with symmetrical sparing of the ventral stream.

If ventral stream functioning were preserved in WBS, we would expect to find that their ability to recognize objects is normal, and at least anecdotally, that appears to be the case. But this does not necessarily mean they have normal object recognition systems. Until recently, there was only one study that systematically examined object representation independent of other spatial functions. In this study, when the performance of people with WBS and Down syndrome was compared, they were comparable in their ability to name objects shown in a canonical view (i.e. that view usually exposing most of an object's critical parts; Wang, Doherty, Rourke, & Bellugi, 1995). However, when shown objects in unusual viewpoints, individuals with WBS performed significantly better than people with Down syndrome.

Though intriguing, these findings do little to tell us whether the object recognition system is normal. For one thing, the lack of difference between the two groups in naming objects with canonical viewpoint could point to impairment (or normalcy) in both groups. Similarly, the difference could mean that people with WBS have spared functions, or that people with Down syndrome are particularly impaired. Aside from these issues is the question of whether the object recognition system in WBS—even if it performs well under some circumstances—might break down more quickly than the normal system when put under pressure.

To investigate this issue, we tested children with WBS, a group of mental age-matched controls, and normal adults (undergraduate students), who provided a benchmark for optimal performance in this task (Landau, Hoffman, & Kurz, 2002). All subjects were shown a set of 80 common objects presented in one of 4 different conditions: (a) canonical viewpoint and clear image, (b) unusual viewpoint and clear image, (c) canonical viewpoint and blurred image, and (d) unusual viewpoint and blurred image. We defined canonical viewpoint as that which exposed most critical parts and was therefore most readily recognized by normal adults. Unusual viewpoints were definitely unusual-- a stool viewed from the underside, a carrot viewed from the top end, a cheeseburger viewed straight on from the top, etc. (see Figure 2a for examples). The complete set of objects for any subject was drawn from a larger set of 320 images (80 objects X the 4 viewing conditions), with each subject viewing 80 different objects. Images were presented briefly, for 500 msec on a computer screen, and as each object appeared, participants were asked to name it ("What's that?"). All responses were coded by a rater who did not know whether he was coding a person with WBS or not.

The accuracy of naming responses was generally high, and variation across conditions was quite similar across subject groups. For the objects presented under canonical views and clear images, normal adults achieved close to 100% correct, and both WBS children and their normal controls came close, both averaging close to 90% correct. This by itself shows that, even under brief presentation, WBS children are comparable to normal children who are MA-matches, and both are performing essentially at ceiling. One critical question is whether the WBS children--despite their apparently normal performance when viewing canonical, clear images of objects--break down more rapidly or more dramatically than other groups under other presentation conditions. Especially critical are the conditions in which the objects were presented under unusual viewpoints. It has been argued that recognition of objects in non-canonical orientations is more "spatial" and may engage more activity in those areas of the brain normally responsible for spatial processing, i.e. the parietal areas (Carpenter, Just, Keller, Eddy, & Thulborn,1999; Farah, 2000). It is therefore possible that WBS individuals might have special difficulty with objects viewed from highly unusual perspectives, since this might require a mental transformation of the object into a more "normal" perspective before recognition could occur.

However, inspection of performance when objects were presented under unusual viewing conditions-- whether with clear or blurred images-- indicates that no such distinctive breakdown occurred. Adults performed better than either of the child groups under these circumstances, but there were no differences between the WBS children and their MA-matches. Even in the most difficult condition--when the objects were presented in unusual perspectives and with blurred images--all groups performed reasonably well, with adults performing at roughly 50% correct and children performing at roughly 40% accuracy.

These results suggest strong sparing of the object recognition system, at least when color and texture of objects is visible. It is still possible, however, that WBS children rely more heavily on surface properties such as color and texture than on more spatial properties such as object shape. To test this, we carried out a second experiment, in which we presented black and white line drawings of common objects for recognition and identification (see Figure 2b for examples). The same subject groups were tested using the same procedures. The results were strikingly similar to those of the first experiment: All subject groups performed very well (above 80%) when identifying objects shown in canonical views, and all subject groups did more poorly for the unusual views. Normal adults performed better than children, but there were no differences between the WBS children and their mental age matches. As in the first experiment, the quality of the naming responses was highly similar across subject groups.

Thus we found no evidence to support the notion that there is any differential breakdown in the object recognition system among children with Williams syndrome. Importantly, we documented the capacity of WBS children to recognize and identify objects under both canonical and unusual perspectives. If object representation is predominantly a ventral stream function, then this evidence would argue in favor of a spared ventral stream. However, it is also possible that recognizing and identifying objects under unusual perspectives is carried out by more dorsal stream functions, because of their spatial nature. If this is the case, then the findings would argue for some sparing of both ventral and some dorsal stream functions (see, e.g. Dilks, Landau, Hoffman, & Siegfried, 2001).

2.2 Biological Motion Perception

If the spatial deficit observed in WBS were due to widespread damage to the dorsal visual system, we would expect to see poor performance in a variety of tasks, such as motion perception and visually-guided reaching that are thought to be mediated by this stream. Atkinson and colleagues (1997,2000) were the first to investigate WBS motion perception by assessing subjects' ability to detect "motion coherence." Motion coherence tasks require subjects to detect a set of signal dots moving in the same direction, embedded in noise dots moving in random directions. Single unit research in monkeys (Newsome & Paré, 1988) as well as functional imaging work in humans (Braddick, O'Brien, Wattam-Bell, Atkinson, & Turner, 2000) indicates that detection of motion coherence relies at least partially, on a dedicated brain area in the dorsal stream (MT in monkeys, area V5 in humans).

Atkinson and colleagues (2000) determined the threshold for detecting motion coherence among WBS people by varying the number of signal dots relative to the number of noise dots (signal-to-noise ratio or S/N). Performance on this task was compared to a companion task that used static line segments to measure "form coherence" which presumably reflects ventral stream processing. They reported that WBS people appeared to fall into one of three distinct subgroups. One group performed poorly on both motion and form coherence tasks, suggesting a possible general deficit in detecting coherence. A second group performed poorly on the motion task but was comparable to controls in perceiving form coherence, a profile found in their earlier Atkinson et al. (1997) work as well as in younger normally developing children. The third group achieved normal performance on both tasks. Overall, these results show that there is a good deal of variability among WBS subjects in terms of their ability to detect motion coherence, and some WBS children, who do poorly on this task, appear to be developmentally delayed rather than selectively impaired. In any case, it does not appear that deficits in detecting motion coherence are uniformly associated with WBS.

We recently investigated whether WBS children were impaired on a motion task involving seeing form-from-motion (FFM), namely the perception of so-called "biological motion" stimuli. These are produced by filming a person moving in the dark with lights attached to the head and limbs. Viewing these displays leads to a clear perception of a person walking, dancing, etc. There are conflicting predictions of how WBS children should perform on this task. On the one hand, they might find it easy, given their interest in other social stimuli, such as faces

(Mervis, et al. 1999; Tager-Flusberg & Sullivan, 2000). Alternatively, this task might be difficult because it requires subjects to integrate the local motion of dots into higher-level units such as arms legs, and ultimately a person (Johansson, 1973). As we noted earlier, some researchers believe that WBS children have difficulty in integrating local features into global configurations (Bellugi, et al. 1994; Bihrle, et al. 1989; but see Pani, et al. 1999) and therefore we might expect them to have trouble with all FFM tasks, including biological motion.

To test these predictions, we compared WBS children, mental age-matched (MA) controls, and normal undergraduates on their ability to perceive biological motion (Jordan, Reiss, Hoffman, & Landau, 2002). In our first experiment, we presented subjects with displays depicting a person engaged in various actions (e.g., slipping on a banana, doing jumping jacks, etc. [1]). Subjects in all three groups easily identified the activity. Next, we presented subjects with point-light-walker (PLW) displays showing a side view of a person walking (as if on a treadmill; see Figure 3, Top Panel). All three groups were able to discriminate the walker's heading (left or right) at or near perfect levels (Jordan et al., 2002).

While this initial work provided evidence that WBS individuals can perceive biological motion, our final study investigated whether their ability was fragile and subject to failure in noisy conditions. To do this, we again showed subjects PLW displays and asked them to indicate the direction the person was headed, but this time, the walker was embedded in different types and amounts of noise (see Figure 3, Middle and Bottom Panels). The presence of these distractor (or "noise") lights makes the task difficult because they are similar to signal dots in terms of shape, color, and motion and therefore can be mistaken for parts of the "figure" (Cutting, Moore, & Morrison, 1988).

Our research examined three types of noise conditions: static noise (static), randomly moving noise (random), and noise having motion paths similar to the signal lights (yoked). The results are shown in Figure 4. All three groups were at or near ceiling performance in the static condition, but made errors in the presence of moving noise (both random and yoked), particularly at high noise levels. In the low noise condition, both normal adults and WBS children were mildly affected by moving noise while MA-matched controls showed a sharp drop in performance (see Figure 4, Left Panel). In the high noise condition, WBS children still appeared to perform better than MA-matched controls, but this difference was not significant. In addition, normal adults were only marginally better than the WBS children (Figure 4, Right Panel). In summary, even with moving noise, WBS children performed as well or better than MA-matched controls in their ability to perceive biological motion, and in some cases, their performance was comparable to normal adults, suggesting that biological motion perception is a spared ability in WBS.

Overall, it does not appear that children with WBS are generally impaired in perceiving motion, although in some cases, they may be developmentally delayed. This finding argues against the hypothesis that WBS is associated with a general deficit in the dorsal stream. Of course, it is still possible that other areas of the dorsal system, such as parietal cortex, are responsible for the spatial deficits observed in WBS. However, our data rule out the strongest form of the hypothesis that all aspects of motion perception are unimpaired.

2.3 Spatial Language

The severe impairment in some aspects of spatial cognition coupled with strength in language inevitably leads to the question of how the spatial deficit is reflected in language. The case of spatial language in WBS provides an unusual wedge into important questions of how space and language interact in normal human architecture. It is normally assumed that, in order to talk about what we see (or hear, or feel), we must be able to represent spatial aspects of the world. If these non-linguistic representations are damaged, then one might expect severe impairment in spatial language.

The relationship between spatial language and non-linguistic representations of space is neither simple nor direct (Landau, 2001). Some aspects of spatial representation may be impaired while leaving intact some spatial language. First, abundant evidence from brain-damaged adults suggest that individual systems of spatial representation can break down while leaving others spared (for review, see Farah, 2000). The same could be true for spatial language. We have already argued that the spatial breakdown in WBS is not global; in fact, some aspects such as object recognition and some kinds of motion perception appear to be spared. These facts immediately raise the question of whether spatial language might also be a specialized system, which encodes highly selective spatial properties using formal structures that simply do not exist in non-linguistic systems (see Landau & Jackendoff, 1993; Landau, 2001, 2002a). If it is a specialized and independent system, spatial language might be acquired with impunity even in the face of severe impairments in other aspects of spatial cognition

We have been testing this hypothesis in a range of studies of spatial language, some of which we review in this chapter. First, however, we make some observations that serve to underscore the importance of considering how knowledge is being measured, i.e. the kinds of tasks that are chosen to tap into linguistic knowledge.

2.3.1 Some Observations about Linguistic Competence and Performance

Initial reports on language in people with WBS provided striking evidence for strength in vocabulary comprehension and production, and in syntax, as shown by grammaticality judgments that tap into subtle grammatical knowledge (Bellugi, Marks, Bihrle, & Sabo, 1988). More recently, careful studies have confirmed that much of the basic machinery of a mature syntax is present in children with WBS (Zukowski, 2001). This is true despite the fact that WBS children show impaired performance on standardized measures of language, such as the TROG (Test for Reception of Grammar; Bishop, 1983). We believe that, in large part, such discrepancies are due to the nature of the tasks that are used to measure grammatical competence. We take a slight detour to illustrate how certain tasks can severely underestimate a child's linguistic knowledge.

Using the TROG, researchers have reported that children with WBS are significantly impaired in processing relative clauses (Volterra, Capirci, Pezzini, Sabbadini, & Vicari, 1996; Karmiloff-Smith et al. 1997; Mervis et al. 1999). A sample item from the TROG is shown in Figure 5. The child is shown four pictures, and is asked to point to the one where "The circle the star is in is red." Note that this sentence contains a relative clause which is not marked by the lexical item "that" which commonly introduces such clauses.

Clearly, there are many ways in which performance could go awry in this task. Children could be impaired in processing the relative clauses (both subject-relatives and object-relatives), they could be impaired in mapping the syntax to the visual items, in scanning all four items, in making decisions (e.g. they might have looser or more stringent criterion for a "yes"), etc. In order to determine whether the impairment reflects a damaged system of grammatical knowledge, one would need a careful test that avoids many of these possibilities, while still tapping into the grammatical structure.

Zukowski (2001) carried out such experiments. She reasoned that accurate production of relative clauses embedded in sentences could only be generated systematically if the appropriate syntactic machinery were intact. She reasoned that the felicity (i.e. pragmatically appropriate) conditions for eliciting subject and object-relative clauses were somewhat subtle, and devised contexts that would likely elicit these clauses among normal individuals. She then tested children with WBS who had already been shown to perform very poorly on these items in the TROG. She found that all children who were tested did indeed produce grammatically correct subject and object-relative clauses, some of which were quite complex indeed. For example, in one case, a WBS subject observed an event depicting one boy sitting on a horse and another identical boy standing on a horse, followed by each boy turning a different color. When she was asked to describe the event, the subject said, "The boy that's sitting on the horse turned green, and the boy who's standing on a horse turned purple." Zukowski also found some errors of production and some of comprehension, but all of these could be explained by factors reserved to the mechanisms of production or comprehension per se-- not to the presence or absence of the grammatical mechanisms used to generate relative clauses.

Zukowski's work illustrates that, as with all questions of competence and performance, one must be careful to test for the target cognitive function in a way that gives the child his or her "best shot" at being correct. Given the degree of specificity and rich structure required to produce a grammatically correct sentence with a complex relative clause, it would be virtually impossible to generate such a sentence unless the grammatical system was functioning properly. Of course, no cognitive function can ever be completely isolated from the processes that interact with it-- we depend on performances to infer competence. This means that, depending on what task we give the child, different competencies might be inferred. Clearly, in order to discover whether a knowledge system is intact, it is crucial to tap into the system with minimal extraneous demands.

2.3.2 Spatial Language in General

Several reports have hinted that spatial language in people with WBS is impaired. The evidence from these reports is sparse. Using the TROG, Karmiloff-Smith et al. (1997) examined the items that tap comprehension of spatial and other relational terms: longer/bigger/taller, in/on, and above/below. Error for these items was 27.0% for longer/bigger/taller, 14.5% for in/on, and 27.9% for above/below (Karmiloff-Smith et al., 1997). It is unclear how to interpret these results. Compared to the extremely poor performance on "complex grammatical" items such as embedded clauses (e.g. "the book the pencil is on is red"; 67.9% errors), the spatial items look like strengths. But compared to performance on "simple" grammatical items, such as two-element combinations (e.g. "the dog is sitting"; 1.3% errors), the spatial items look like weaknesses. Aside from this problem, the spatial test items in the TROG cannot be regarded as a serious estimate of WBS spatial language, since they test comprehension of single terms in single, often ambiguous contexts, rather than systematically examining the spatial distribution of a given term's use (see Landau, 2002b).

Bellugi, Lichtenberger, Jones, Lai, & St. George (2000) reported that WBS children were more likely than normal children to switch figure and ground objects in their sentences, e.g. saying "The bowl is in the apple" rather than "The apple is in the bowl". Unfortunately, interpretations of such performance are open to the same difficulties as we observed in the case of relative clauses. Errors on figure/ground assignment could reflect misassignment of semantic roles to their syntactic positions, or they could reflect misalignment of the elements of the sentence during the process of production (see, e.g. Bock & Levelt, 1994). The former would reflect a damaged part of the linguistic system, whereas the latter would reflect missteps in the coordination of separate processes involved in production. Without systematic testing, it is impossible to tell which interpretation is correct.

Our strategy in examining spatial language has been to look both broad and deep. We have been examining spatial language broadly, by looking at the language that expresses the locations of objects (above, below, near, far, etc.) (Zukowski, Schwartz, & Landau, 1999; Landau, 2002b), the language that expresses the locations of object parts within objects (top, bottom, side, etc. (Landau & Kurz, unpublished results), and the language that expresses motion events, in which objects move through space over paths (Landau & Zukowski, 2002; Lakusta, Licona, & Landau, 2002). In the present chapter, we focus on motion events. The findings here, however, are similar to those in our other studies of spatial language: They suggest that much (if not all) of the structure of spatial language is spared, and that unusual patterns of performance reflect the operation of non-linguistic systems as they interact with language.

2.3.3 The Language of Motion Events

English, like other languages, expresses motion events using multiple pieces of structure which combine to express the entire event. According to widely cited linguistic theories, English expresses the motion event by encoding (a) the *Figure*, or object which moves, by a noun phrase (NP), (b) the *Motion* itself, usually by a verb, (c) the *Path* over which the object moves, usually by a preposition, and (d) the *Ground*, or *Reference* object for the moving object, another NP (see, e.g., Talmy, 1983). For example, in "The dog jumped off of the fence", the "dog" is Figure, the "fence" is Reference object, "jump" expresses the motion, and "off of" expresses the Path--one in which the dog moves away from the Reference object.

The acquisition of each type of component is complex. For example, expression of each component requires that the child perceive the component (object, motion, path) accurately, and then encode it correctly in language. This parsing of the event into motion/manner and path complex differs across languages, so part of the child's task is to determine just how to parse the event for his or her native language. In addition, there are spatial constraints on the expression of path. Linguistically, paths can be divided into three types: To, From, and Via paths (see, e.g. Jackendoff, 1983), and different prepositions are appropriate to each path type. Finally, within each path type, the preposition selects for the type of reference object: Some prepositions choose for "container-like" objects (e.g. in, into, etc.), whereas others choose "surface-like" reference objects (e.g. on, off of, etc.) These constraints function to narrow down which preposition is the correct one for expressing any particular path.

In sum, linguistically expressing a motion event requires accurate perception of the event, accurate parsing into the correct elements, and accurate choice of path terms on the basis of spatial and linguistic constraints.

We asked how much of this structure is available to children and adults with WBS by showing people a set of 80 motion events in which Figures underwent a variety of motions over a variety of paths, and asking "What happened?" (Landau & Zukowski, 2002; Lakusta, Licona & Landau, 2002). We found that the WBS children named both Figure and Reference objects using the same nouns as both other groups. We also found that WBS children used almost exactly the same manner-of-motion verbs as the normal controls. Table 1 shows the top 12 verbs used by each group and their proportions of use. The children accurately perceived the objects and motions, and corrected encoded them in language. Given the variety of motion types, we find it especially impressive that WBS children were able to select the correct verbs.

Overall, expression of the Path was also done accurately—the appropriate path term was chosen in most cases, and an impressive variety of path terms was produced (see Landau &

Zukowski, 2002). The selection of Path term types—To, From, and Via— among WBS children matched that of the normal controls and adults, indicating that the children correctly determined whether the path itself was most felicitously expressed in terms of its goal, source, or the intermediate (for Via paths). The only place where we found a difference between WBS children and the controls was in *whether* the Path complex was expressed: To-Paths were almost always expressed by all groups, but both Via- and From-Path types and their Reference objects were frequently omitted by WBS children. For example, if the motion event showed a girl moving past a block, the WBS child was more likely than the controls to indicate the girl and her movement, but not the block (e.g. "The girl was walking.") Or, if the event showed a block falling off of a swing, the WBS child was more likely indicate the block and its motion, but no reference object (e.g., "The block fell" or "The block fell off", but rarely "The block fell of the swing"). Thus the difference amounted to simply omitting the Path complex and/or the Reference object alone in events that displayed Via- or From-Path types.

How can we explain this difference? First, note that although the effect was quite specific, it was substantial and reliable. Second, the effect did *not* result in ungrammatical sentences, since specifying the Path complex for such events is linguistically an option. Third, all of the pertinent grammatical and semantic structure in the sentences was preserved, showing that children with WBS had control over the linguistic expression of motion events. Hence, the problem in production of From-Paths does not lie with knowledge of language.

Rather, we believe that the selective omission of Path complex in Via- and From-Path types may reflect a fragility in retaining information about the event that is less salient or important to the observer. Specifically, when observers view the motion events, the most salient aspect of the event is the Figure object's motion. In cases of To-Paths, the Figure moves to end up at the Reference object, and in these cases, the children preserved the entire Path complex, producing both Path and Reference object. But in Via and From-Path types, the Figure moves away from or past the Reference object, and is not spatially coincident with the Reference object at the end of the event, when he or she must describe it. In these cases, it would seem natural that the observer would either focus only on the moving Figure object, or that he or she might even forget what the Reference object was and what Path was traversed. People with WBS are known to have fragile visual-spatial memories (Jarrold, Baddeley, & Hewes, 1999; Wang & Bellugi, 1994; Vicari, Brizzolara, Carlesimo, Pezzini, & Volterra, 1996). Therefore, it seems quite possible that they might have greater difficulty holding in mind these aspects of the event over time. Because the focus of the vulnerability seems to be "sources" (the Reference objects that are located at the beginning of From-Path events), we call it "Source vulnerability".

If our hypothesis is correct, the tendency to drop the Path complex should be correlated with weaker visual-spatial memory. Moreover, this interpretation implies that the weakness is not linguistic per se—there is nothing abnormal about the nature of the linguistic system, but only as it interacts with visual-spatial memory. Thus, we should be able to create circumstances where From-Path types are produced more easily and regularly. Finally, our interpretation places the vulnerability in the non-linguistic system that is responsible for representing and storing events. If we are right, we should see the reflex of a "source vulnerability" in non-linguistic representations of events. Such vulnerability should definitely show up in people with WBS, but may also be a characteristic of normal event representation, and if so, should appear in the performance of normal children and adults. We are now testing each of these possibilities.

2.4 Summary: Specialization and Sparing

The work we have just reviewed suggests strong sparing and preservation of structure in three domains: object representation, biological motion perception, and spatial language. In each case, we have taken pains to test people's capacity under stringent conditions which allow us to determine whether the structures underlying normal representations are spared in WBS. The findings show that, yes, structural properties of these spatial representations are intact. Given these strengths, we can now ask why people with WBS show such severe spatial breakdown in the Block Construction task.

3.0 Revisiting the Block Construction Task: Why the Deficit?

We believe it is possible to obtain poor performance in the block task for a variety of reasons. Earlier, we speculated that errors in construction tasks could be attributed broadly to two different sources: (a) executive processes that are required for planning and maintaining information in working memory and (b) spatial-perceptual processes that construct and manipulate representations of objects and their spatial relationships. Impairment in the first kind of process is a plausible source of errors in people with WBS, who may have impaired frontal lobe functions (Atkinson, et al., 2000) and are moderately mentally retarded. Impairment in the second would be expected on the assumption that WBS selectively impairs spatial capacities. The problem with the block task is that, if we examine only the final outcome of the task, these two factors are inextricably woven together. We believe that a deeper understanding of the locus of any deficits can come through a careful analysis of the task's requirements and decomposition of these into underlying mental processes. We offer the following as an example of such an analysis (see Hoffman, Landau, & Pagani, 2002 for full report).

3.1 A Model for Thinking about Construction Tasks

We assume that insight can be gained into the locus of deficits by examining people's puzzle solutions on a micro-time line--looking at when and how people move individual blocks, and where they look during this process. To understand the kinds of insight such an analysis can provide, we turn to the work of Ballard, Hayhoe, Pook, & Rao (1997) who studied eye movements of normal adults while they solved block construction puzzles such as those shown in Figure 6. In this task, people have to duplicate a *model* by moving blocks from a *parts* area to a *copy* area-- much as WBS subjects must do when confronted with the hallmark Block Assembly task (Differential Abilities Scale; DAS, Elliott, 1990). Our natural intuitions suggest that normal, healthy adults might solve such a puzzle by first glancing at the model, then memorizing the identities and locations of several blocks and then rapidly moving the corresponding parts into place. After doing this perhaps one or two times, the copy would be complete. Assuming that people are able to accurately represent the identity of each block and its location, the copy would also be correct.

Surprisingly, Ballard et al. (1997) found that observers did not solve the puzzle in this way. Rather, people tended to look at the model *each time* they moved a single block. In fact, subjects often looked at the model *twice* before placing each block--the first time, apparently, to get the *identity* of the block (then picking it up from the parts area) and then a second time to determine its *location* (then moving it into its position in the copy area). As Ballard et al. point out, subjects who use this two-prong strategy appear to be minimizing the amount of information they must hold in working memory, using the model as a kind of external source which they could continually revisit, and eliminating the need to store multiple block identities and multiple locations of blocks within the puzzle.

By examining the relationship between where people looked and the actions that they took (e.g., picking up a block in the parts area, placing it in the copy, removing a piece from the copy), Ballard et al. (1997) made otherwise covert executive processes amenable to study. We believe that their analysis also suggests that failure at copying, such as occurs among WBS people, could arise at any of a number of points during the process of constructing a copy. For example, if WBS people have faulty *executive mechanisms* (the mechanisms that might help us plan as we solve these puzzles), then many of the processes uncovered by Ballard et al. should show impairment. It is executive mechanisms that guide people's sequence of fixations on the different regions of the problem space (the model area, the parts area, the copy area) as they encode the identity and location of model parts, search the parts area, and place blocks in the copy area. Executive processes also determine when a person should check for errors (reflected in back and forth fixations between copy and model), and attempt needed repairs. These executive processes can be examined by analyzing the person's sequence of eye fixations across different regions of the problem space, both as they are placing blocks, and as they attempt to initiate any repairs.

Second, if WBS specifically targets *spatial representations*, then people with WBS might fail because they cannot create and maintain a representation of the identity and location of the pieces. This problem may be particularly acute for the kind of puzzle shown in Figure 6 in which the pieces consist of parts in various spatial arrangements. For example, the block in the upper left of the model needs to be represented as something like “ a vertical orientation with the dark part on the right”. The location of the block in the complete puzzle needs to be represented as “upper left section of the puzzle". If WBS subjects have trouble representing these kinds of *spatial* relationships, then subjects will often choose incorrect parts and/or place them in incorrect locations in the copy (Key et al. 1998). In separating these two kinds of processes, we can determine whether the deficit is due to executive mechanisms, spatial representations, or some interaction of the two.

3.2 Experimental Findings

We examined the eye fixations of WBS children, mental age-matched controls, and normal adults while they solved puzzles like those shown in Figure 6. Subjects used the computer's mouse to select and move parts into a copy area to duplicate a model that was continuously visible on the screen. The copy area contained placeholders for the blocks which "snapped into place" if they were dropped close to the correct position. Thus global shape errors or "broken configurations" were eliminated as a source of errors.

In terms of performance, we found that normally developing mental age-matched controls correctly solved more puzzles than WBS children and the difference became progressively larger as the puzzles became larger and more complex. For example, WBS children correctly solved only 15% of the most complex puzzles compared to 50% for controls. In addition, we found that the controls were more accurate on their *individual block drops* during the complex construction task. This already suggests that there are errors made during the construction process, block by block.

The low accuracy of WBS children was not, however, attributable to faulty executive processes. For smaller puzzles (two and three pieces), all three groups fixated the model equally often, showing that the WBS children looked back and forth between model and copy normally when the puzzle was relatively simple. For the larger puzzles, normal adults and control children both increased their fixations on the model relative to the simple puzzles, while WBS children showed a precipitous drop, being much less likely to consult the model-- a fact that could easily lead to poor performance. We believe, however, that this reduction in model fixation was a *result* rather than a cause of poor accuracy on these problems. First, drop accuracy was not directly related to degree of model fixation: It was approximately the same whether or not the individual drops were or were not preceded by model fixations. For example, WBS children fixated the model approximately 95% of the time prior to their first drop in the nine piece puzzles and their drop accuracy was 47% (chance was approximately 32%). On the remaining drops, their fixation rate dropped to 48% while their accuracy fell to 41%-- only a 6% difference. In other words, their accuracy was low on these puzzles regardless of the degree to which they fixated the model-- so the drops in fixation did not cause poorer performance per se. Rather, for the larger puzzles, the WBS children appear to have adopted a strategy of moving pieces randomly into the copy area in the hopes of creating a correct copy by chance-- resulting in poor performance.

We examined other executive processes by looking at the children's error detection and repair procedures. Again, it turned out that the WBS children and both control groups were quite similar, checking their *final* solutions at the same rate. So when the WBS children completed a puzzle, they knew to check their copy against the model for accuracy. Control children also frequently checked their *partial* solutions-- as they moved through the construction process-- which is a reasonable strategy, given the high probability of an error on each drop. WBS children, however, checked their partial solutions less often, consistent with the idea we suggested above, that they were more likely to attempt a random generation of the puzzle solutions, followed by checking. Finally, both groups of children were unlikely to attempt repairs on correct puzzles, showing that they knew which of their solutions were correct and which were not. For WBS subjects, 96% of their repair attempts were directed at *incorrect* puzzles. The corresponding figure for controls was 92%. Thus both groups seemed to recognize when their copies corresponded to the model.

These findings suggest that poor overall performance by WBS children is not due to impaired executive processes that guide the pickup of information from the model. First, the children accurately solve smaller puzzles using eye fixations that are similar to control children. Second, they checked complete copies and were able to determine when their copy was wrong, accurately initiating repair procedures. The only place where they differed from control children is in the degree to which they fixate the model for the most complex puzzles, and the degree to which they fixate the model as they move through the construction process, i.e. after partial solutions. Both of these differences may be due to the fact that they are aware of their own deficit, most strikingly present for the most complex and largest puzzles, and under these circumstances, they simply adopt a new strategy in which they attempt to solve the puzzle by placing pieces randomly, hoping for some accuracy.

What about deficits in their spatial representations? We carried out two experiments to evaluate these. In the *Matching* task, the four and nine piece puzzles from the first experiment were presented along with a cue (i.e. a disk in the center of a randomly chosen block in the model). Subjects were to choose the block in the parts area that matched the single cued block. Overall, WBS subjects were correct only 46% of the time compared to 81% for mental age-matched controls. Thus WBS subjects are severely impaired in a task that reduces much of the strategic complexity of the block construction task and depends primarily on representing and matching block identity.

Similarly, the *Location* task was designed to evaluate the ability of children to represent the location of blocks in the model without the complexity of the full puzzle. Models were presented with a cue on just one of the blocks, subjects were given a single block, and they had to place this into the copy location corresponding to the cued location in the model. Overall, WBS subjects were correct on 80% of the trials compared to 97% for controls. This result suggests that WBS subjects are also impaired at representing the location of the block in the model.

These two experiments indicate that WBS children are impaired at representing spatial relationships that are important for *identifying* the blocks in the model and coding their *locations*. These impairments appear to be sufficient to account for much of the observed deficit in block construction because people appear to solve these puzzles “one block at a time” by coding the identity and location of individual blocks. The fact that WBS children appeared to be good at recognizing whether their copies were correct suggests that-- contrary to some reports-- their perception of global shape may be intact.

In summary, the results of our micro-analysis of the block construction task show, first, that the executive processes used to search for information, check copies against the model, and initiate repairs, are all intact in children with WBS. Where they differ dramatically from normal mental age-matched control children is in the quality of their representations of the blocks themselves and the spatial relationships among them in the model. Their inability to choose the correct matching block, and their difficulties placing even a single block in the copy space point to real fragility in the spatial representations that must be engaged during this highly complex task.

This fragility needs to be more precisely characterized, a task in which we are now engaged. One question that arises is why the children perform so poorly in the simple block matching task, if they do so well on demanding object recognition and identification tasks (see

Section 2). One possibility is that the complexity of familiar everyday objects is different from the complexity of the blocks used in the construction task. In particular, the key property of the blocks and their distractors is asymmetry: The *target* blocks in the standard construction task are vertically, horizontally, or diagonally asymmetrical by color (e.g. split right/left, top/bottom, or top-left/right-bottom, etc. by color). The *distractor* blocks in the full construction task, and in the matching task, are also vertically, horizontally, or diagonally asymmetrical by color. Thus, accuracy on the matching task requires that the subject note the color relationships of the target (i.e. how the block is split, and which side has which color), and match it to another, disregarding blocks that are split differently and those that are split the same, but whose colors lie in a mirror-image relationship to the target.

Inspection of all children's errors in the Matching task shows that their errors are overwhelmingly choices of the mirror-image mates to the target. For example, given a black/white block split horizontally (black on top, white on bottom), all children will erroneously choose the black/white horizontal split with reversed colors-- but rarely the vertically or diagonally split blocks. However, WBS children make more such errors than MA-matched controls. In addition, the errors were predominantly made on diagonally-split blocks, followed by both horizontals and verticals in equal proportion. Again, WBS children make more such errors, but the error types are the same as in normally developing mental age-matched children.

Possibly, one locus of deficit in WBS lies in a highly specific characteristic of object recognition which engages the capacity to differentiate mirror-image reversals. Add to this the fact that even our simple matching task requires some degree of visual-spatial memory (as one moves from the model to the block choices), and we suggest that the deficit taps into fragile mirror-image relationships which are further weakened over time.

The fragility in placing single blocks into the copy raises yet other questions. Why should this be a problem, given the strength seen in other recruitment of spatial relationships, such as the description of spatial motion events? We do not yet have a resolution to this puzzle, but ongoing experiments in our lab are designed to provide greater detail on how spatial relationships are represented by people with WBS. Some evidence suggests that, like the case of objects and blocks, there may be specific kinds of relationships that are particularly hard to represent, and that these will undergo rapid decay whenever the task requires any retention in memory.

In sum, we have placed the burden of the deficit in the block construction task onto spatial representations, and not on the executive processes that control the viewer's selection and use of information. We should also point out that the two aspects of cognition interact in a clear but complex way. We have already noted that the executive processes appear remarkably intact for simple puzzles, but that, with larger (more complex) puzzles, the children with WBS appear to adopt a different kind of strategy. Specifically, they no longer consult the model as frequently as with simple puzzles, and appear to merely place blocks in the puzzle with no rhyme or reason, hoping for a solution to appear. This strategy also appears for normal children with more complex puzzles, but the difference is that (a) the overall accuracy for individual blocks is much higher for normal children, and (b) normal children check partial solutions more frequently than WBS children. The upshot is that the normal children are able to correct more frequently along the way, and when they do, they are more often accurate in their corrections, leading to overall performance that greatly outstrips the children with WBS. Thus, although the executive processes of the WBS children are normal, they are derailed when the puzzles becomes too difficult. In this way, the complexity of the full block construction puzzle and the striking deficits seen in WBS performance, reveal a complex sequence in which a persistent deficit in representing spatial relationships can derail other mechanisms, leading to downward spiraling performance.

4.0 Clinical Implications

Although our research is not applied in nature, we believe that our findings have important implications for the treatment and education of people with WBS. One clear implication of our results is that WBS is not characterized by monolithic spatial impairment. We have found remarkable sparing of spatial representational capacities in three separate domains: Object representation, biological motion, and spatial language. To the extent that clinicians can build on spared aspects of spatial cognition, they may find ways to help WBS people compensate for their impairments. The result would be more effective treatment and education of people with WBS.

Our findings may also help in understanding the exact nature of the WBS impairment, further elucidating treatment and education possibilities. Like other researchers, we found striking shortcomings among WBS children in the hallmark task of block construction. However, we were also able to distinguish between the executive (i.e. planning) processes that are used in the task and the spatial representations that people use when they identify blocks and place them in locations. We argued that only the latter are severely impaired in WBS, but that this impairment interacts with executive processes as the puzzles become more complex: Children are aware of their difficulties, and so they shift executive strategies to essentially abandon what they recognize to be futile strategies of information search. Furthermore, we emphasized that the block task taps many spatial functions, including visual memory for objects and locations, planning ability, motor control. It also requires overall maintenance of a an overall "plan" which allows periodic checking between a partially constructed model and the final production. As we have pointed out, things can go wrong for the WBS patient at any point along the route to final puzzle solution; and failure does not necessarily indicate failure at all levels. This puts their failure in a new perspective, and suggests that the focus for researchers should be discovering more precisely what characterizes the deficit in representing certain kinds of spatial relationships

Understanding the nature of spatial strengths and deficits in WBS should have implications for possible treatment and intervention. For example, perhaps we can take advantage of the plasticity of brain connections to provide targeted training that would gradually produce improvement in those brain circuits that are deficient in WBS. Tallal et al. (1996) showed rapid improvement in Language-Learning Impaired Children by requiring them to discriminate temporal information in synthetic speech. This training was given in the context of a computer game in which the temporal information was easy to discriminate but became progressively harder over time. Presumably, perceptual learning produced changes in neural networks responsible for processing the rapid changes that occur in human speech and these changes were manifest as improvements in comprehending speech. Subsequent applications of this approach to dyslexia (Temple et al., 2003) resulted in improved reading performance and the authors observed training-induced changes in language related brain areas using fMRI.

This approach depends on having a good understanding of the underlying causes of any given deficit, which would allow a targeted training regime to be designed. Therefore, an important goal in research on people with WBS is to achieve a detailed understanding of the nature of their spatial deficits as well as strengths.

5.0 Perspectives

. We believe that the next ten years of research should emphasize three themes. One is the study of highly specialized spatial and cognitive systems, to gain a greater understanding of what is spared. We would especially like to see serious study of domains that -- like space-- are likely to reflect inherent characteristics of the human brain and mind. Such domains include knowledge of objects, causality, number, and music. If basic structural facts about the architecture of these human knowledge domains are shown to be spared in WBS, then we will have ample evidence for the idea that this syndrome does not compromise the qualitative nature of knowledge. Aditionally, the fact that aspects of spatial cognition survive in WBS is testament to its robust nature, and to the likelihood that these aspects of cognition emerge naturally in development despite genetic deficit. To the extent that other, non-spatial domains follow this pattern, we will have stronger evidence that genetic syndromes need not impinge on basic cognitive architecture. These facts would be important for studies of WBS, and equally important, for our scientific theories of human cognition.

The second theme is the need to determine just what goes wrong in WBS cognition. We believe that this may be a very difficult task, especially given the inherent complexity of any cognitive task in any domain. However, by investigating fundamental representations and mechanisms -- such as visual-spatial memory and attention-- we might be able to develop a model that speaks to both the strengths and weaknesses of the WBS cognitive profile. To use spatial cognition as an example, we have shown that the cognitive processes that are specific to functions such as object recognition and biological motion perception (for example) are preserved. Yet, it is quite possible that the WBS cognitive system might break down quite rapidly as pressure from memory and/or attentional requirements increases. Such breakdown would reflect greater fragility under stress, but not a different cognitive architecture.

The final theme is the importance of studying learning itself in WBS. Until now, researchers' major task has been to document the pattern of strengths and weaknesses, in an attempt to firmly establish the nature of the cognitive profile. Crucial questions about this profile may be answered by studying how people with WBS learn. Because learning has both theoretical and applied ramifications, this theme may be the most important of all.

References

Atkinson, J., Braddick, O., Anker, S., Curran, W., Andrew, R., Braddick, F., & Wattam-Bell, J. (2000). *Neurobiological models of visuo-spatial cognition in children with Williams syndrome: Measures of dorsal-stream and frontal function.* Unpublished Manuscript.

Atkinson, J., King, J., Braddick, O., Nokes, L., Anker, S., & Braddick, F. (1997). A specific deficit of dorsal stream function in Williams' syndrome. *NeuroReport, 8*, 1919-1922.

Ballard, D.H., Hayhoe, M.M., Pook, P.K., & Rao, R.P.N. (1997). Deictic codes for the embodiment of cognition. *Behavioral and Brain Sciences,* 20, 723-767.

Beery, K.E. & Buktenica, N.A. (1967). Developmental Test of Visual-Motor Integration. Cleveland: Modern Curriculum Press.

Bellugi, U., Bihrle, A., Neville, H., Doherty, S., & Jernigan, T. L. (1992). Language, cognition, and brain organization in a neurodevelopmental disorder. In M. Gunnar & C. Nelson (Eds.), *Developmental behavioral neuroscience: The Minnesota Symposia on Child Psychology*. Hillsdale, NJ: Lawrence Erlbaum Associates.

Bellugi, U., Lichtenberger, L., Jones, W., Lai, Z., & St. George, M. (2000). The neurocognitive profile of Williams syndrome: A complex pattern of strengths and weaknesses. *Journal of Cognitive Neuroscience, 12: Supplement*, 7-29.

Bellugi, U., Marks, S., Bihrle, A., & Sabo, H. (1988). Dissociation between language and cognitive functions in Williams syndrome. In D. Bishop & K. Mogford (Eds.), *Language development in exceptional circumstances* (pp. 177-189). Hillsdale, NJ: Lawrence

Erlbaum.

Bellugi, U., Wang, P. P., & Jernigan, T. L. (1994). Williams Syndrome: An unusual neuropsychological profile. In S. H. Broman & J. Grafman (Eds.), *Atypical cognitive deficits in development disorders: Implications for brain function* (pp. 23-56). Hillsdale, NJ: Erlbaum Press.

Bertrand, J., Mervis, C., & Eisenberg, J. (1997). Drawing by children with Williams syndrome: A developmental perspective. *Developmental Neuropsychology,* 13(1), 41-67.

Bihrle, A. M., Bellugi, U., Delis, D., & Marks, S. (1989). Seeing either the forest or the trees: Dissociation in visuospatial processing. *Brain and Cognition, 11*, 37-49.

Bishop, D. (1983). Test for reception of grammar. UK: Medical Research Council.

Bock, K., & Levelt, W. (1994). Language production: Grammatical encoding. In M. Gernsbacher (Ed.), *Handbook of psycholinguistics*. San Diego, CA: Academic Press.

Braddick, O. J., O'Brien, J. M. D., Wattam-Bell, J., Atkinson, J., & Turner, R. (2000). Form and motion coherence activate independent, but not dorsal/ventral segregated, networks in the human brain. *Current Biology, 10,* 731-734.

Carpenter, P.A., Just, M.A., Keller, T.A., Eddy, W.K., & Thulborn, K. (1999). Graded functional activation in the visuo-spatial system with the amount of task demand. *Journal of Cognitive Neuroscience, 11*, 9-24.

Cutting, J. E., Moore, C., & Morrison, R. (1988). Masking the motions of human gait. *Perception & Psychophysics, 44,* 339-347.

Deruelle, J., Mancini, M.O., Livel, M., Casse-Perot, C., de Schoon, S. (1999) Configural and local processing of faces in children with Williams syndrome. *Brain and*

Cognition, 41, 276-298.

Dilks, D., Landau, B., Hoffman, J. E., & Siegfried, J. (2001, April). *Selective impairment of dorsal stream functions in Williams syndrome?* Poster session presented at the annual meeting of the Cognitive Neuroscience Society, New York, NY, CA.

Elliot, C.D. (1990). *Differential Ability Scales.* San Diego, CA: Harcourt, Brace, Jovanovich.

Farah, M. J. (2000). *The cognitive neuroscience of vision*. Malden, MA: Blackwell.

Georgopolous, M. Georgopolous, A., Kurz, N., & Landau, B. (2002) Copying figures in Williams syndrome. Submitted manuscript, University of Minnesota.

Hoffman, Landau, & Pagani (2002) Spatial breakdown in spatial construction: Evidence from eye fixations in children with Williams Syndrome. *Cognitive Psychology.*

Jackendoff, R. (1983). *Semantics and Cognition*. Cambridge, MA: MIT Press.

Jarrold, C., Baddeley, A., & Hewes, A. (1999). Genetically dissociated components of working memory: Evidence from Down's and Williams syndrome. *Neuropsychologia, 37*, 637-651.

Johansson, G. (1973). Visual perception of biological motion and a model for its analysis. *Perception & Psychophysics, 14,* 201-211.

Jordan, H., Reiss, J.E., Hoffman, J.E., & Landau, B. (2002). Intact perception of biological motion in the face of profound spatial deficits: Williams syndrome. *Psychological Science, 13(2),* 162-167.

Karmiloff-Smith, A., Grant, J., Berthoud, I., Davies, M., Howlin, P., & Udwin, O. (1997). Language and Williams syndrome: How intact is "intact"? *Child Development, 68*(2), 246-262.

Key, A. F., Pani, J.R., and Mervis, C.B. (1998). Visuospatial constructive ability of people with Williams Syndrome. Paper presented at The Sixth Annual Workshop on Object Perception and Memory. Dallas, TX.

Kramer, J, J., Bulusewicz, M.J., Kaplan, E., & Preston, K.A. (1991). Visual hierarchical analysis of block design configural errors. *Journal of Clinical and Experimental Neuropsychology, 13(4)* 455-465.

Lakusta, L., Licona, R., & Landau, B. (2002, November). *Interaction between spatial representation and spatial language: The language of events*. Paper presented at the Boston University Conference on Language Development, Boston, MA.

Landau, B. (2001). Perceptual units and their mapping with language. In T. F. Shipley & P. J. Kellman (Eds.) . *From fragments to objects: Segmentation and grouping in vision* (pp. 71-118). Amsterdam, New York: Elsevier.

Landau, B. (2002a). Spatial Cognition. In Ramachandran, V. (Ed.), *Encyclopedia of the human brain*. San Diego, CA: Academic Press.

Landau, B. (2002b). *Breakdown of reference systems in spatial cognition and spatial language: Evidence from Williams syndrome*. Manuscript in preparation, The Johns Hopkins University.

Landau, B., Hoffman, J.E., and Kurz, N. (2002) Spared Object Recognition with Profound Spatial Deficits in a Genetic Disorder. Submitted manuscript.

Landau, B. & Jackendoff, R. (1993). "What" and "where" in spatial language and spatial cognition. *Behavioral and Brain Sciences, 16*(2), 217-265.

Landau, B. & Zukowski, A. (2002). Objects, motions, and paths: Spatial language in children with Williams syndrome. In C. Mervis (Ed.), *Developmental Neuropsychology: Special Issue on Williams Syndrome*.

Mervis, C. B., Morris, C. A., Bertrand, J., & Robinson, B. F. (1999). Williams Syndrome: Findings from an integrated program of research. In H. Tager-Flusberg (Ed.), *Neurodevelopmental disorders: Developmental cognitive neuroscience* (pp. 65-110). Cambridge, MA: MIT Press.

Milner, A. D., & Goodale, M. A. (1995). *The visual brain in action.* New York: Oxford University Press.

Mottron, L., Belleville, S., & Menard, E. (1999). Local bias in autistic subjects as evidenced by grapic tasks: Perceptual hierarchization or working memory deficit? *Journal of Child Psychology and Psychiatry and Allied Disciplines, 40(5), 743-755.*

Newsome, W. T., & Paré, E. B. (1988). A selective impairment of motion perception following lesions of the middle temporal visual area (MT). *The Journal of Neuroscience, 8*, 2201-2211.

Pani, J. R., Mervis, C. B., & Robinson, B. F. (1999). Global spatial organization by individuals with Williams syndrome. *Psychological Science, 10*, 453-458.

Tager-Flusberg, H., & Sullivan, K. (2000). A componential view of theory of mind: Evidence from Williams syndrome. *Cognition, 76,* 59-89.

Tallal, P., Miller, S.L., Bedi, G., Byma, G., Wang, X., Nagarajan, S., Schreiner, C., Jenkins, W.M. and Merzenich, M.M. (1996) Language comprehension in language-learning impaired children improved with acoustically modified speech. *Science, 271*, 81-84.

Talmy, L., (1983). How language structures space. In H. Pick & L. Acredolo (Eds.), *Spatial orientation: Theory, research, and application* (pp.225-282). New York: Plenum.

Temple, E., Deutsch, G.K., Poldrack, R.A., Miller, S.L., Tallal, P., Merzenich, M.M. (2003) Neural deficits in children with dyslexia ameliorated by behavioral remediation: Evidence from functional MRI. *Proceedings of the National Academy of Sciences of the United States of America, 100(5)*, 2860-2865.

Ungerleider , L.G. and Mishkin, M., (1982). Two cortical visual systems. In D. Ingle, M. Goodale, and R. Mansfield (Eds.), *Analysis of Visual Behavior.* Cambridge, MA: MIT Press.

Vicari, S., Brizzolara, D., Carlesimo, G. A., Pezzini, G., & Volterra, V. (1996). Memory abilities in children with Williams syndrome. *Cortex, 32*, 503-514.

Volterra, V., Capirci, O., Pezzini, G., Sabbadini, L., & Vicari, S. (1996). Linguistic abilities in Italian children with Williams syndrome. *Cortex, 32*, 663-677.

Wang, P. P., & Bellugi, U. (1994). Evidence from two genetic syndromes for a dissociation between verbal and visual-spatial short-term memory. *Journal of Clinical and Experimental Neuropsychology, 16*(2), 317-322.

Wang, P., Doherty, S., Rourke, S.B., and Bellugi, U. (1995). Unique profile of visuo-perceptual skills in a genetic syndrome. *Brain and Cognition, 29*, 54-65.

Wechsler, D. (1981). Wechsler adult intelligence-revised. New York: Psychological Corporation.

Zukowski, A. (2001). *Uncovering grammatical competence in children with Williams syndrome*. Unpublished doctoral dissertation, Boston University, Boston.

Zukowski, A., Schwartz, D., & Landau, B. (1999, November). *Spatial language and spatial cognition: Evidence from Williams syndrome*. Paper presented at the Boston University Conference on Language Development, Boston, MA.

Table 1 Twelve Most Frequent Specific Verbs of Motion: Percents of Use

Adults		Control Children		WS Children	
Verb	%	Verb	%	Verb	%
fall	26.6	fall	31.0	fall	28.1
jump	14.7	jump	17.2	jump	12.7
fly	9.1	fly	11.3	fly	11.5
hop	7.1	hop	7.5	hop	5.6
walk	5.6	walk	4.5	walk	7.3
roll	4.4	roll	5.5	roll	4.2
drive	4.2	drive	1.5	drive	1.9
slide	3.8	slide	1.9	flip	4.4
make a turn	2.6	run	1.4	turn	3.8
spin	1.5	go in “L”	1.5	zigzag	3.5
back/up	2.3	ride	1.4	run	2.9
bounce	1.8	bump	1.4	bounce	1.9
Totals	83.7		86.1		87.9

Figure Legends

Figure 1. Sample drawings by WS children, and one normally developing child who is matched for mental age. Matching is done using the Kaufman Brief Intelligence Test (KBIT).

Figure 2a. Samples of full color objects used in object identification studies. Objects are shown in the four experimental conditions: (a) canonical, clear image, (b) unusual, clear image, (c) canonical, blurred image, and (d) unusual, blurred image. Objects are presented on a computer screen for brief duration (500 msec) and people are asked to identify them by name.

Figure 2b. Samples of outline drawings of objects used in object identification studies. Objects are shown in the two experimental conditions: (a) canonical image, (b) unusual image.

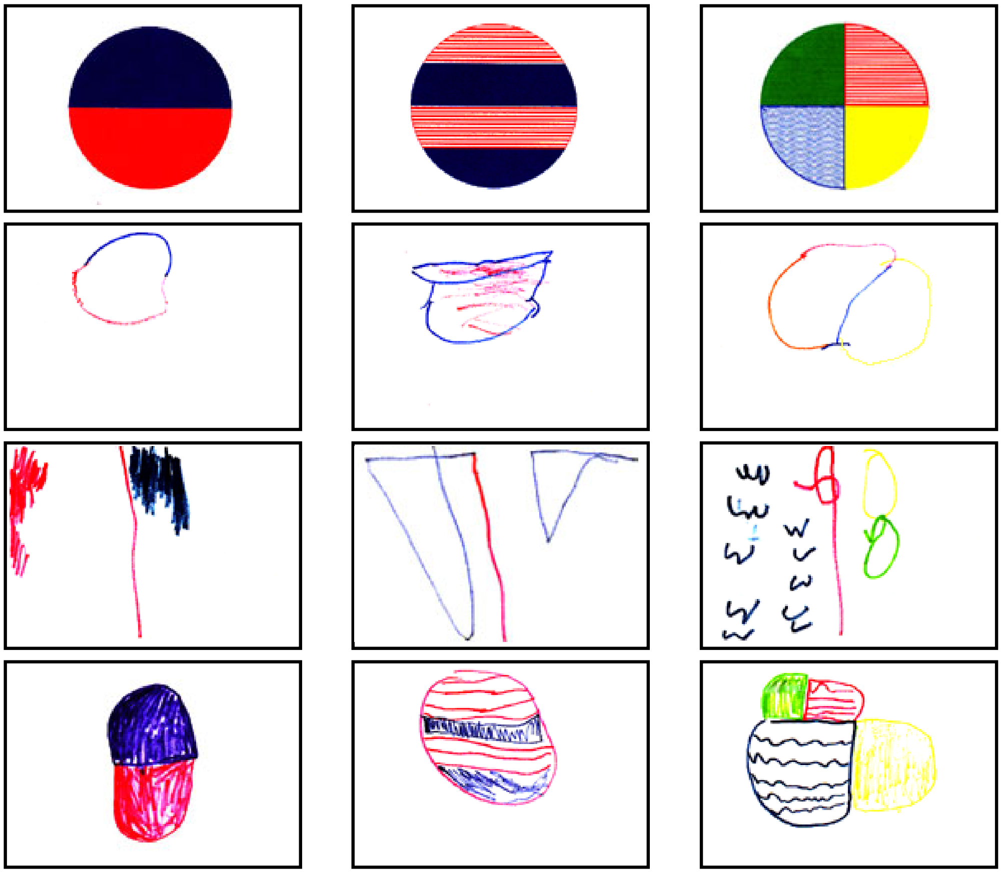
Figure 3. Scale illustrations of the biological motion displays used in pilot work (top), as well as low (middle) and high (bottom) noise conditions (Jordan et al., 2002). In all cases, the signal and noise lights were white against a black background. People viewed moving displays of these dots, and were asked to indicate whether the human figure represented by dots was moving to the right or left. Reprinted with kind permission of Blackwell Publishing, Osney Mead, Oxford OX2 0EL, UK.

Figure 4. Mean percentage accuracy of reporting walking direction in biological motion displays for each of three noise conditions (static, random, and yoked) and three subject groups (undergraduates, WS children, and mental-age-matched children; Jordan et al., 2002). Results are shown separately for low (left) and high (right) noise conditions. Chance performance lies at 50%. Reprinted with kind permission of Blackwell Publishing, Osney Mead, Oxford OX2 0EL, UK.

Figure 5. Sample item from the T.R.O.G. (Bishop, 1983) The child is asked to point to the one where "The circle the star is in is red".

Figure 6. Example of a block construction puzzle including the Model (upper left), the Parts bin (lower panel) and the Copy area (upper right)

Figure 1



Model

Williams
Age 11;1
KBIT 70
(RA)

Williams
Age 11;1
KBIT 66
(BR)

Control
Age 6;9
KBIT 116
(LC)

Figure 2a


Canonical
Clear
Unusual
Clear
Canonical
Blurred
Unusual
Blurred
Blender
Cup
Bottle
Stool

Figure 2b

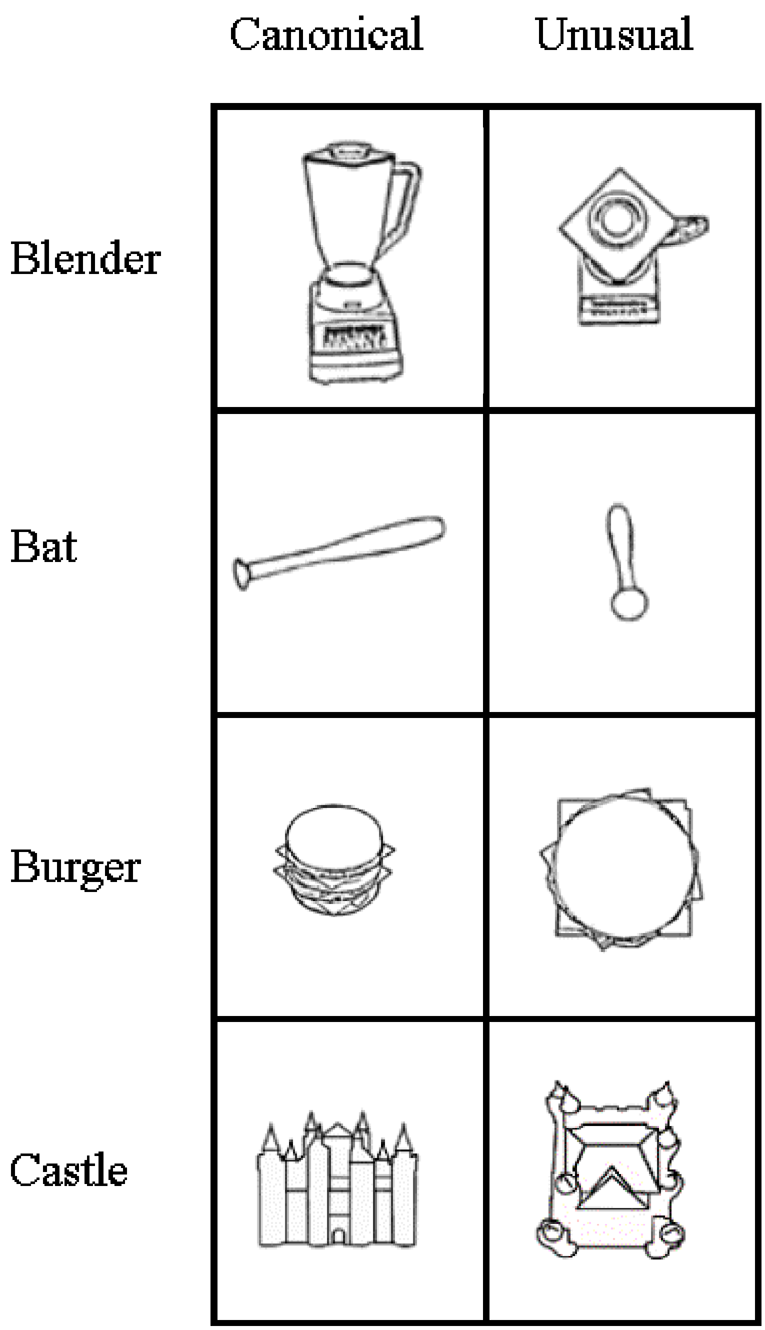


Figure 3

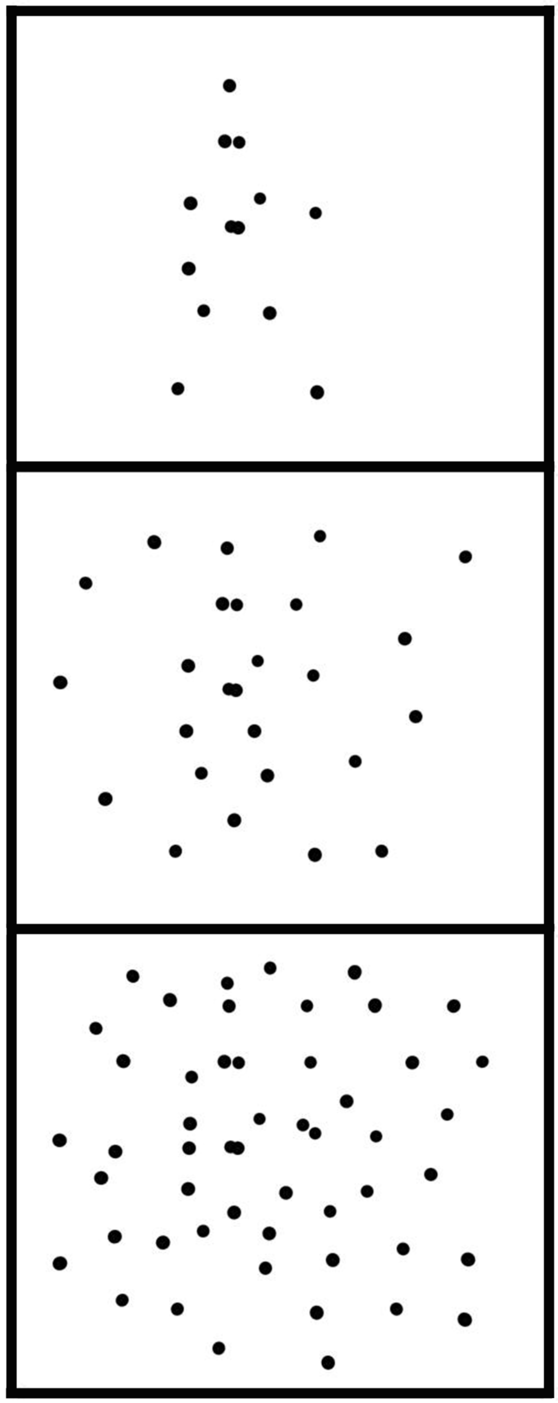
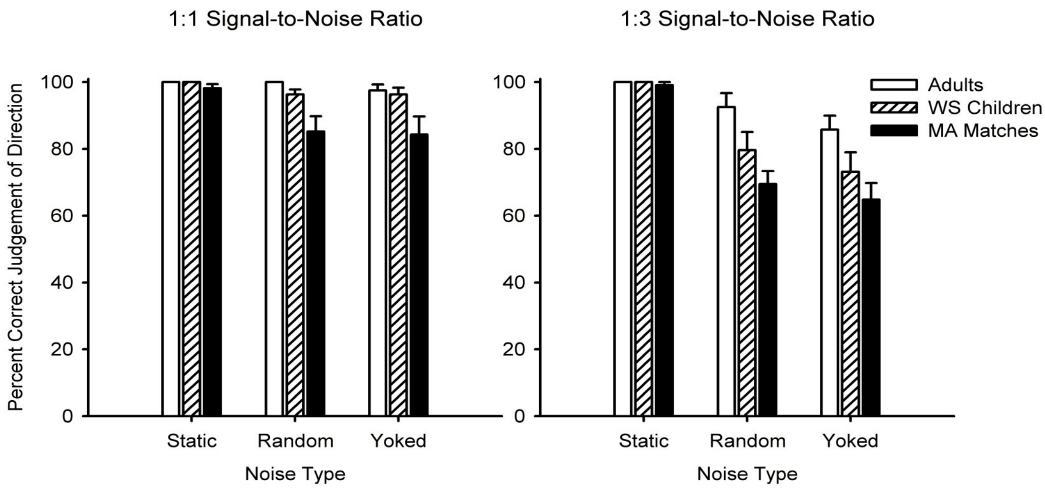


Figure 4



1:1 Signal-to-Noise Ratio
1:3 Signal-to-Noise Ratio
Percent Correct Judgement of Direction
100
80
60
40
20
0
Static
Random
Yoked
Noise Type
Adults
WS Children
MA Matches

Figure 5

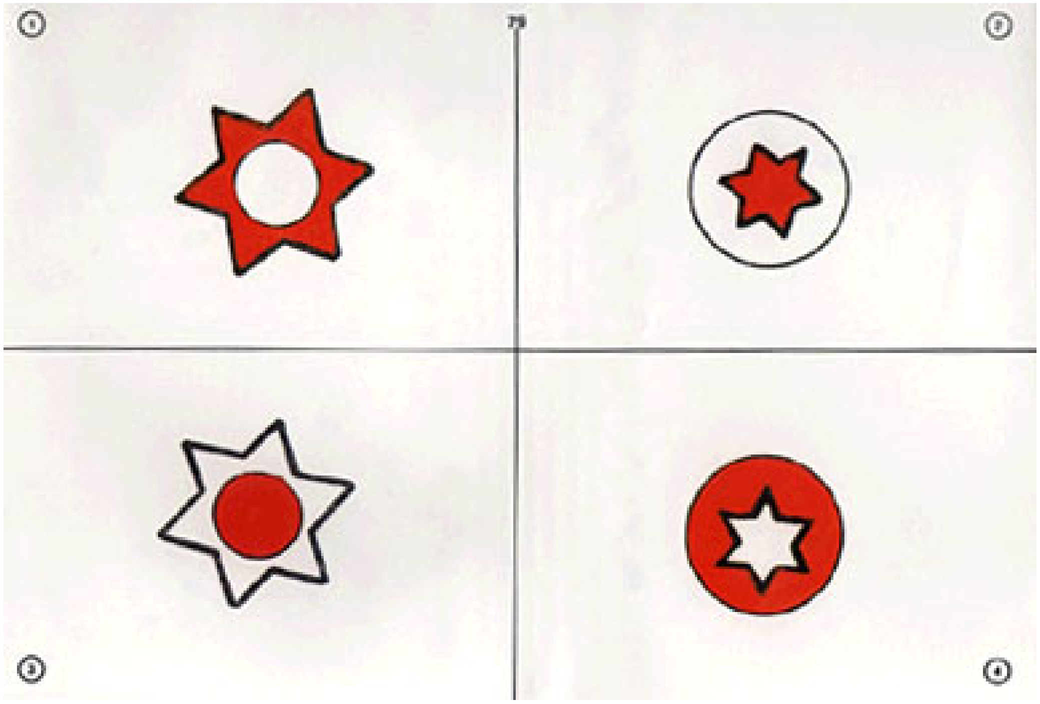
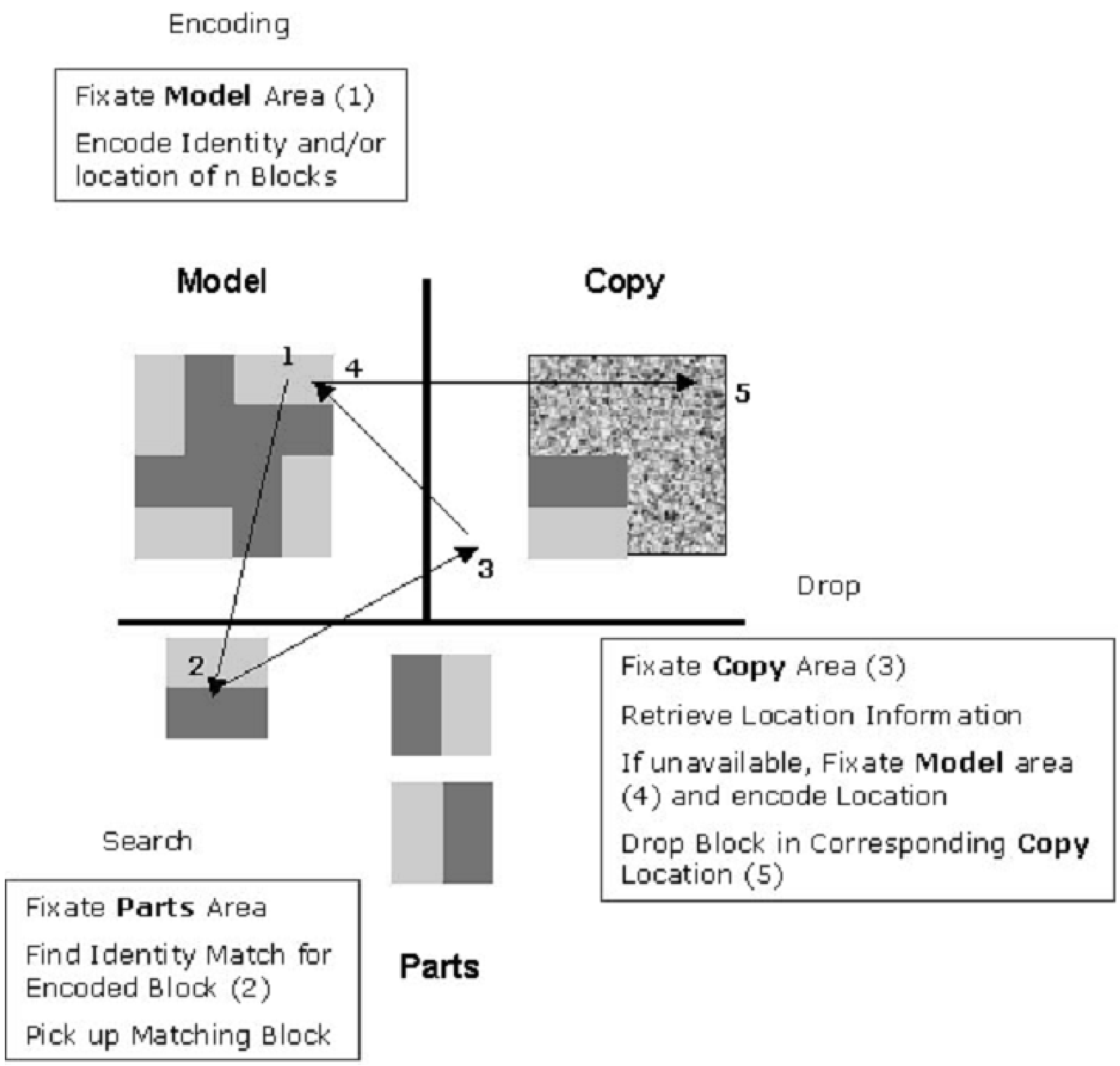


Figure 6


Encoding
Fixate **Model** Area (1)
Encode Identity and/or location of n Blocks
Model
Copy
1
4
5
3
2
Drop
Fixate **Copy** Area (3)
Retrieve Location Information
If unavailable, Fixate **Model** area (4) and encode Location
Drop Block in Corresponding **Copy** Location (5)
Search
Fixate **Parts** Area
Find Identity Match for Encoded Block (2)
Pick up Matching Block
Parts

Footnote

[1] An example of the stimuli used in this experiment can be found on the web at http://hoffman.psych.udel.edu/research/emdemo/WilliamsPage.htm.