



# Geometric and featural systems, separable and combined: Evidence from reorientation in people with Williams syndrome <sup>☆</sup>



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## ABSTRACT

Spatial reorientation by humans and other animals engages geometric representations of surface layouts as well as featural landmarks; however, the two types of information are thought to be behaviorally and neurally separable. In this paper, we examine the use of these two types of information during reorientation among children and adults with Williams syndrome (WS), a genetic disorder accompanied by abnormalities in brain regions that support use of both geometry and landmarks. Previous studies of reorientation in adolescents and adults with WS have shown deficits in the ability to use geometry for reorientation, but intact ability to use features, suggesting that the two systems can be differentially impaired by genetic disorder. Using a slightly modified layout, we found that many WS participants could use geometry, and most could use features along with geometry. However, the developmental trajectories for the two systems were quite different from one other, and different from those found in typical development. Purely geometric responding was not correlated with age in WS, and search processes appeared similar to those in typically developing (TD) children. In contrast, use of features in combination with geometry was correlated with age in WS, and search processes were distinctly different from TD children. The results support the view that use of geometry and features stem from different underlying mechanisms, that the developmental trajectories and operation of each are altered in WS, and that combination of information from the two systems is atypical. Given brain abnormalities in regions supporting the two kinds of information, our findings suggest that the co-operation of the two systems is functionally altered in this genetic syndrome.

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## 1. Introduction

Abundant evidence has shown that when humans and other species become spatially disoriented, they can re-establish their orientation using a geometric representation of the surrounding space (Cheng, 1986; Cheng & Newcombe, 2005; Gallistel, 1990; Gallistel & Matzel, 2013; Tommasi, Chiandetti, Pecchia, Sovrano, & Vallortigara, 2012; Wang & Spelke, 2002). It is also known that landmarks are of great importance in both oriented navigation and in re-establishing orientation (Aguirre & D'Esposito, 1999; Epstein, 2008; Newcombe & Huttenlocher, 2000). Although representations of geometry and landmarks usually work seamlessly together at the behavioral level, evidence now suggests that the

two systems are separable, both in their contributions to spatial navigation and as they are instantiated in the brain (Bullens et al., 2010; Burgess, 2008; Doeller & Burgess, 2008; Doeller, King, & Burgess, 2008; Janzen & van Turenout, 2004; Sutton, Joannisse, & Newcombe, 2010). The learning and remembering of objects relative to the boundaries of an environment (which define its geometric shape) has been specifically linked to right posterior hippocampal activation, while learning and remembering of landmark-related locations is linked to right dorsal striatal activation (Doeller et al., 2008). In development, geometric sensitivity emerges early, and in some species appears to be independent of experience (Chiandetti, Spelke, & Vallortigara, 2014; Chiandetti & Vallortigara, 2008, 2010, but see Twyman, Newcombe, & Gould, 2012 for contrasting results with mice). By contrast, featural landmark use is highly susceptible to training and practice in children (Twyman, Friedman, & Spetch, 2007) and animals (Kelly & Spetch, 2004) and is supported by different learning mechanisms in adults (Doeller & Burgess, 2008). How information from the two systems is combined for reorientation over development still remains unknown, although fMRI evidence strongly implicates the role of

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the hippocampus in mature adults (Sutton et al., 2010). Behavioral evidence suggests that geometric representations are primary, used by children at all ages and across many contexts. Combination of geometry with a featural landmark is somewhat more variable (depending on age and size of the reorientation chamber), appearing anywhere from age 2 through 5, but stably present by age 6 (Hermer-Vasquez, Moffet, & Munkholm, 2001; Hupbach & Nadel, 2005; Learmonth, Nadel, & Newcombe, 2002; Learmonth, Newcombe, & Huttenlocher, 2001; Learmonth, Newcombe, Sheridan, & Jones, 2008).

In this paper, we probe the nature of the two systems by examining the use of geometry and features among people with Williams syndrome (WS), a genetic disorder that is characterized by a deletion of 26 genes on chromosome 7q11.23 (Hillier et al., 2003; Morris, 2006; Osborne, 2006). People with WS have a unique cognitive profile of mild to moderate mental retardation along with severe spatial impairments (Mervis et al., 2000). Across a broad range of spatial functions, they show a profile that is overall quite similar to that of TD 4–6 year-olds (Landau & Hoffman, 2012). However, perhaps the most striking aspect of the WS spatial profile is their performance on reorientation tasks. Unlike the ubiquitous pattern of geometric reorientation throughout typical human development and in all animal species studied (e.g., Cheng & Newcombe, 2005; Gallistel & Matzel, 2013; Hermer & Spelke, 1996; Tommasi et al., 2012), people with WS show severe limits in this capacity. Using the now-classic reorientation task developed by Cheng (1986) for testing rats, and adapted by Hermer and Spelke (1994, 1996) for testing children, Lakusta, Dessalegn, and Landau (2010) found that WS adolescents and adults could not reliably use geometric layout information to reorient in a rectangular room with four black walls. However, almost all were able to use a specific featural landmark (a single colored wall, hereafter referred to as a “feature”) to do so. Only 3 individuals showed use of both geometry and the feature across the two room environments.

These findings suggest selective impairment to representation of geometric layout in people with WS, but not to representation of features, a pattern which is consistent with theories and evidence suggesting that the two navigational systems are separable in terms of both behavioral mechanism (Julian, Keinath, Muzzio, & Epstein, 2014; Lee, Shusterman, & Spelke, 2006; Lee & Spelke, 2010) and underlying neural instantiation (Doeller & Burgess, 2008; Doeller et al., 2008). The WS pattern of performance differs qualitatively from that of TD individuals, for whom geometry is used from a very early point in development, and features are integrated with geometry in both small and large spaces from about age 5 onward (Hermer-Vasquez et al., 2001; Learmonth et al., 2002). Consistent with the deficit in spatial navigation, WS individuals show structural and functional abnormalities in the parietal lobe and the intraparietal sulcus (Eckert et al., 2005; Kippenhan et al., 2005; Meyer-Lindenberg et al., 2004) and also in the hippocampus (Meyer-Lindenberg et al., 2005; Meyer-Lindenberg, Mervis, & Berman, 2006; Reiss et al., 2000).

Hippocampal damage from lesions has been linked to deficits in using geometry for reorientation among rats, pigeons and chicks (McGregor, Hayward, Pearce, & Good, 2004; Tommasi, Gagliardo, Andrew, & Vallortigara, 2003; Vargas, Petruso, & Bingham, 2004). Moreover, in a fMRI study of human adults using virtual reality, the hippocampal region was found to show greater activation when participants were required to reorient according to both room geometry and a featural cue, which suggests that this region is involved in the effective combination of these sources of information (Sutton et al., 2010). People with WS show gross preservation of hippocampal volume compared to age and gender-matched controls, but consistently unusual morphology. In particular, they show local volume reduction at the posterior apex and expansion

at the anterior base (Meyer-Lindenberg et al., 2005). The converse of this pattern is observed in humans proficient in spatial navigation (i.e., London taxi drivers), who show increased posterior and decreased anterior hippocampal volume (Maguire et al., 2000). A similar result has also been found for college students, for whom size of the right posterior hippocampus predicts relative position estimation of landmarks in a real-world environment (Schinazi, Nardi, Newcombe, Shipley, & Epstein, 2013).

The evidence to date for neural and behavioral separation of the two systems in WS depends on the assumption that geometric representation of layout is severely impaired in these individuals, but that feature use remains intact. Several aspects of the original Lakusta et al. (2010) data raise questions about these assumptions, however, and suggest new questions about how the two systems function in this genetic disorder. First, although Lakusta et al. (2010) found that most individuals with WS failed to use geometry when reorienting in an all-black room (i.e., with no featural information), a few individuals may have used geometry. This suggests that sensitivity to geometry may not be entirely absent in WS. Second, use of the feature by the majority of WS individuals hints that they may have had access to some geometric layout information, even though this was not often shown in the all-black room. This is because successful use of the feature likely requires some kind of spatial representation of the layout. Participants used the colored wall as a true landmark and not as a beacon; that is, they did not search *at* the colored wall itself, but rather, used the wall to infer the target location at a specific corner. It is unclear whether spatial information was used in combination with the landmark, because it is also possible to use sense information (e.g., “when facing the red wall, search to my left”). There are hints that this spatial representation could have been geometric, at least for some people, as errors tended to accumulate in the rotationally equivalent corner to the target corner in the feature condition, which is the signature of geometric reorientation.

These observations suggest two points. First, the previous study may have underestimated the degree to which geometric representations may be constructed and used by people with WS. Second, if geometry can be more systematically observed, then it would be possible to more closely examine the degree to which both systems—geometry and features—are used together, seamlessly, as seems to be the case for TD individuals after the age of 5. Even if features are combined with geometry in WS performance, we may observe a behavioral signature for this combination that differs from TD children, and this signature could further support the idea that the two systems are neurally and behaviorally separate.

To pursue these issues, we first aimed to test geometric response among a new set of WS participants belonging to a broad age range. Lakusta et al. (2010) had tested WS individuals who were between the ages of 9 and 27 ( $M = 17$  years), with most participants being adolescents or adults. It is possible that dampened use of geometry among these individuals may be a developmental product of reliance on unique features and landmarks for many years (perhaps even being encouraged to do so by parents). This could follow if geometric sensitivity is fragile, and/or if the system for features functions robustly from early in development. Therefore, in the current study, we tested a wider age range of individuals, looking specifically for any evidence of geometric sensitivity and its potential change over age. We also introduced minor changes to the reorientation room to enhance the salience of geometric structure (see Section 2).

Second, we aimed to replicate Lakusta et al.’s (2010) finding of robust feature use, and in particular, to examine more closely how, if at all, the feature is combined with geometry by people with WS. Recall that most of the individuals tested in Lakusta et al. (2010) successfully used a featural cue (a single blue wall) to reorient

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