

# **Errors in reorientation**

# A study of typically developing children and individuals with Williams syndrome



Katrina Ferrara\* and Barbara Landau, Language and Cognition Lab Department of Cognitive Science, The Johns Hopkins University

\* corresponding author and IGERT trainee: kferrar3@jhu.edu

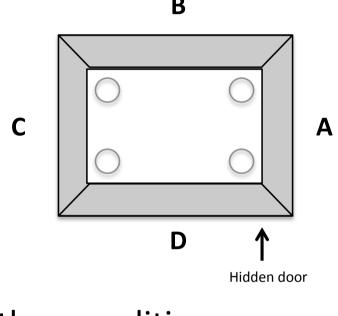
#### **Abstract**

When an organism has become disoriented, it must find a way to reestablish the relationship between itself and the environment. A wide range of research has demonstrated that both humans and animals achieve reorientation by forming a representation of the surrounding space based upon its geometry (e.g., the lengths of surfaces and their angles of intersection). However, research with one population has shown that many members do not reorient in accord with the geometry of a room (1). Williams syndrome (WS) is a disorder marked by the deletion of approximately 25 genes on chromosome 7, which presents with a cognitive profile that includes severely impaired spatial understanding and strikingly strong language (1, 2, 3). We tested reorientation in people with WS, as well as typically developing (TD) children. We found that many WS participants were able to successfully use geometry to locate a hidden target. These data enrich our understanding of the navigational abilities of the members of this population; some do possess a sensitivity to geometric relationships early in life. In terms of performance in the rooms with one red wall, the collective results of previous research and the present study indicate that successful landmark use develops at a rate that is much slower than in the typically-developing case.

Participants 16 individuals with WS (M = 16 yrs, range = 5.6 - 32.6 yrs) 16 TD children (M = 3.9 yrs, range 3.5 - 4.5 yrs)

#### **Method: Room reorientation task**

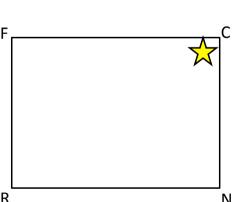
**Stimuli** A diagram of the reorientation chamber is shown on the right. White circles indicate the locations of plastic opaque containers. In the small room, walls A and C were 4 ft. across, and walls B and D were 6 ft. across. In the large room, walls A and C measured 8 ft. across, and walls B and D were 12 ft. across.



In one condition all 4 walls of the chamber were black, and in another condition one wall was covered with red felt. Thus, 4 testing environments were used.

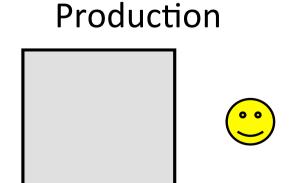
**Task** Participants were blindfolded and disoriented by turning around in a circle several times. Upon opening their eyes, they were asked to recover a hidden sticker. Subjects completed 16 trials total, 4 within each testing environment.

**Data Analysis** For each trial, each search was coded as correct (C), rotational equivalent (R), near the target (N), or far from the target (F). Target is indicated by a star.

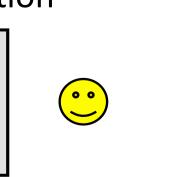


#### **Additional Measures**

#### Spatial Language Task

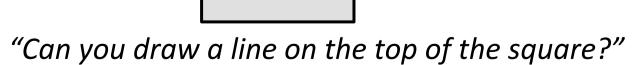


"Where is the smiley face to the square?"









Comprehension

#### Standardized assessments

To assess the cognitive profile of the WS participants, 3 standardized tests were used:

- Kauffman Brief Intelligence test (K-BIT II) (4)
- Average IQ of 76.69 (SD, 13.31; range, 49-91) DAS II: Pattern Construction (5)
- Average performance at the 5<sup>th</sup> percentile for chronological age (range, 0.01% 29%)
- DAS II: Digit Span (forward) (5)

Average performance at the 11<sup>th</sup> percentile for chronological age (range, 0.01% - 66%)

### **Results: Reorientation**

#### TD Group

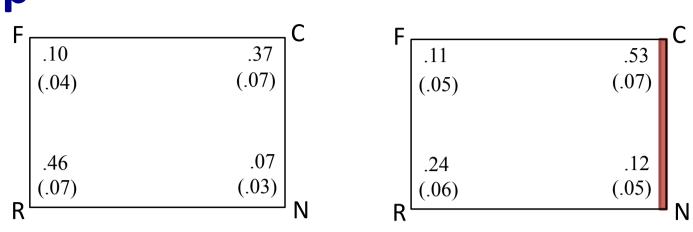




Fig. 1 - TD children follow the classic pattern (6) in the all-black room, searching the C and R corners roughly 83% of the time. In the room with one red wall, children reliably searched at the C corner more often than the R corner. (Data collapsed over 8 trials.)

#### WS Group: Was geometry used?

We found that the geometric performance of the WS participants in the black rooms closely resembled that of the TD children. 14 individuals searched more often at the C and R corners, 1 searched more often at the N and F corners, and 1 searched at the two corner types the same number of times.

### Age effect: Sensitivity to geometry?

We divided the WS participants into 2 groups: those who were 18 years and older (n = 7, M = 21.75 yrs), and those who were younger than 18 years (n = 9, M = 11.25 yrs).

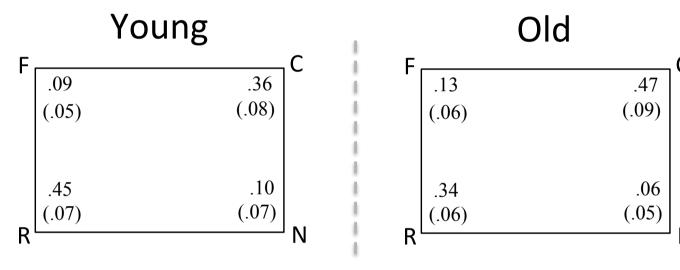


Fig. 2 - Age was not found to reliably predict the use of geometry (search at the C and R corners) in the all-black rooms.

### WS Group: Was the landmark used?

In the rooms with one red wall, 5 individuals demonstrated perfect use of the landmark (they searched at the correct corner on all 8 trials). When considering the two age groups, a clear developmental trend emerged.

## Age effect: Use of the landmark?

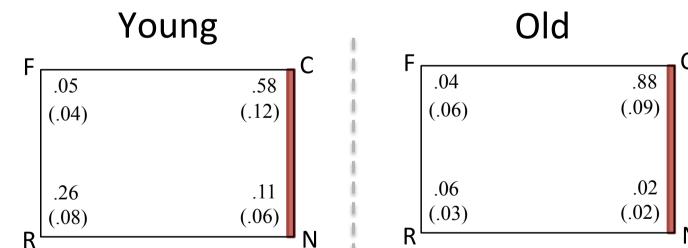


Fig. 3 - Age was found to be a reliable predictor of search at the C corner in the rooms with one red wall.

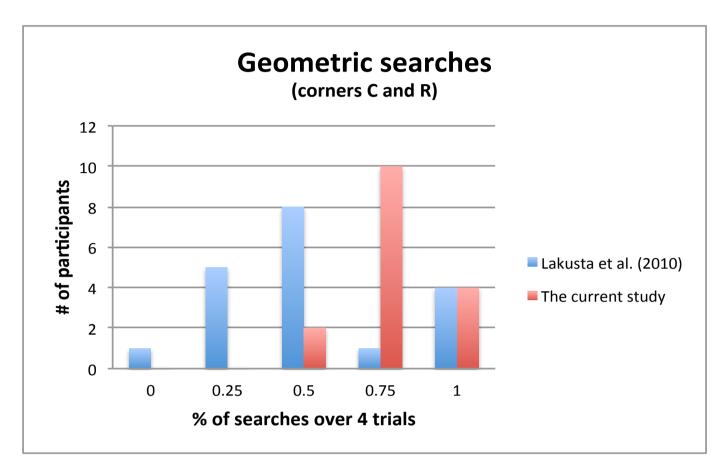
## **Additional Findings**

- The red wall is a beacon? Target proximity to the red wall did not affect performance: subjects for whom the stickers were hidden near the red wall did no better than those for whom the stickers were hidden far from the red wall (both TD and WS).
- Spatial Language Task and IQ
  - TD: Neither production nor comprehension of the terms "left/right" predicted search at C in the red-wall rooms, or search at corners C and R in the all-black rooms.
  - WS: Several factors emerged as significant predictors of geometric performance (search at C and R corners) in the all-black rooms:

Predictor	Beta	p
K-BIT II	1.48	0.001
DAS:Digit span	1.1	0.002
Comprehenion left/right	0.93	0.023
Production left/right	-1.6	0.003

### Comparison to Lakusta et al. (2010)

Previous research in our lab has suggested that many individuals with WS are unable to use geometry to help them find a hidden target. In contrast, results from this study suggest that despite their spatial deficit, some individuals do possess a sensitivity to geometric relationships. What might account for this? The difference may be the fact that two slightly different testing environments were used. In Lakusta et al. (2010), the corners of the room were concealed by panels. This may have rendered the geometry of the room less salient, in comparison to the corners that were left unconcealed in the rooms of the present study.



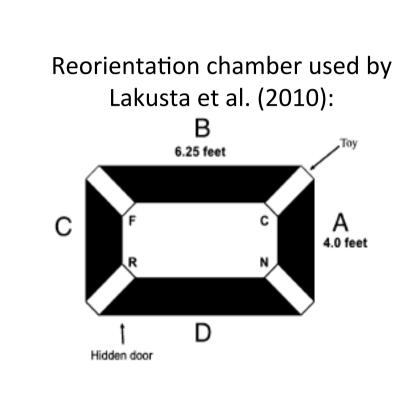


Fig. 4 - Number of subjects who searched geometrically on 0, 25%, 50%, 75%, or 100% of the trials in the room with all-black walls. More WS participants in the present study demonstrate a sensitivity to geometry.

## Conclusions

- No difference in performance was found between the large (8' x 12') and small (4' x 6') reorientation rooms, for either the TD or WS groups.
- Standardized assessment scores were found to predict geometric performance in only the black-wall rooms. This relationship to IQ suggests that different mechanisms may be recruited in the two conditions, which is also supported by research demonstrating that different neurological substrates contribute to geometric and feature-based responses in navigation tasks (7).
- The age effect of landmark use suggests that within the WS population, sensitivity to geometry remains at a fixed level, while the ability to use featural information in combination with geometry increases with age.
  - We hypothesize that older individuals with WS have developed strategies to help compensate for the difficulties they face when required to return to particular locations. For the younger participants, these compensatory strategies are not yet in place.
  - The developmental transition from purely geometric response to landmark use that takes place between the ages of 4 and 6 in the typically developing case is slowed down dramatically within this special population, such that the transition which normally occurs within the span of two years requires the input of far more life experience.

#### References

- 1. Lakusta L., Dessalegn B., Landau B. (2010). Impaired geometric reorientation caused by genetic defect. PNAS, 107, 2813-7. 2. Palomares M., Landau B., Egeth H. (2008). Visuospatial interpolation in typically developing children and in people with Williams
- Syndrome. Vision research, 48, 2439-50. 3. Tager-Flusberg, H., Plesa-Skwerer, D., Faja, S., & Joseph, R.M. (2003). People with Williams syndrome process faces holistically. Cognition, 89(1), 11-24.
- 4. Kaufman AS & Kaufman NL. (2004). *Kaufman Brief Intelligence Test*, 2<sup>nd</sup> Ed. Circle Pines, MN: American Guidance Service.
- 5. Elliott CD. (1990). Differential ABILITIES scale. San Diego, CA: Harcourt Brace Jovanovich.
- 6. Hermer L., & Spelke E. (1994). A geometric process for spatial reorientation in young children. Nature, 370, 57–59.
- 7. Jones PM, Pearce JM, Davies VJ, Good MA, McGregor A (2007) Impaired processing of local geometric features during navigation in a water maze following hippocampal lesions in rats. Behavioral Neuroscience, 121, 1258-1271.

#### **Acknowledgements**

We thank Kate Kelliher, Emma Gregory, and Michael McCloskey for their assistance with this work, which was supported by NIH grant R01 NS 050876, as well as an Integrative Graduate Education and Research traineeship through NSF grant DGE 0549379.