Impaired behavioral and neural sensitivity to boundary cues in Williams syndrome

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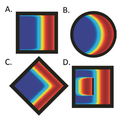
Introduction

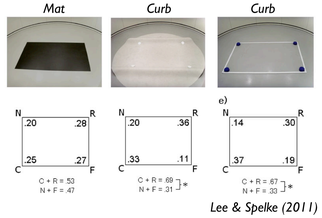
• Research highlights the importance of boundaries for the purpose of navigation

- Boundary cells fire when an animal is facing a specific direction and at a specific distance from a boundary(Burgess & O'Keefe, 2011)



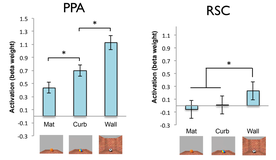
Lever et al. (2009)

- Children reorient by the shape of arrays formed by surfaces that are low enough to see and step over (e.g., a curb), but not 2D arrays such as a flat mat or tape on the floor (Lee & Spelke, 2008; 2011)



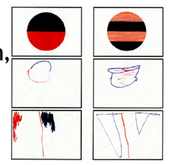
Lee & Spelke (2011)

- fMRI with healthy adults show that the PPA is sensitive to the presence of a 3D boundary cue using both univariate and MVPA (Ferrara & Park, VSS 2013; SfN 2014)



• The case of Williams Syndrome (WS)

- Williams syndrome (WS) is a developmental disorder characterized by a set of approximately 25 genes missing on chromosome 7q11.23 (Morris, 2006). (incidence estimate = 1 in 7,500 (Strømme et al., 2002)).
- Associated with mild to moderate mental retardation, accompanied by a unique cognitive profile that includes severe impairment in a range of spatial functions compared to Typically Developing (TD) Chronological age & gender matches (CA control)



Research Question

- Do WS have similar sensitivity to boundary cues as typically developing controls?
- Is there a link between the behavioral reorientation and neural representation of environmental boundaries?

Experiment 1: Behavioral Reorientation

Reorientation task:
Find a toy in one of the corners after reorientation

Design:
Within-subject design
4 trials per condition

Condition:

		Mat	Curb	Wall
TD 4-year-olds N=18		.25 .21 / .25 .29	.15 .32 / .35 .18	N: .11 .39 / .44 .06
geometric	C + R =	.46	.67 *	.83 *
non-geometric	N + F =	.54	.33	.17
TD 6-year-olds N=18		.17 .33 / .29 .21	N: .18 .33 R / .39 .10	.08 .46 R / .40 .06
geometric	C + R =	.62 *	.72 *	.86 *
non-geometric	N + F =	.38	.28	.14
CA control Mean age = 22.02 N=12		.05 .40 / .48 .07	N: .05 .47 / .43 .05	.00 .45 / .52 .03
geometric	C + R =	.88 *	.90 *	.97 *
non-geometric	N + F =	.12	.10	.03
WS Mean age = 22.37 N=15		.23 .22 / .30 .25	N: .27 .21 / .24 .28	N R: .08 .40 / .47 .05
geometric	C + R =	.52	.45	.87 *
non-geometric	N + F =	.48	.55	.13

✓ 4-year-olds reorient geometrically by Curb, but not Mat

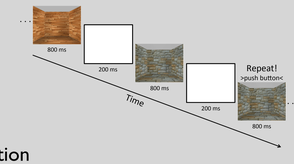
✓ 6-year-olds & CA can use all types of boundary cues (with Step-wise sensitivity)

✓ WS can't use geometry defined by curb, which 4-year-olds can

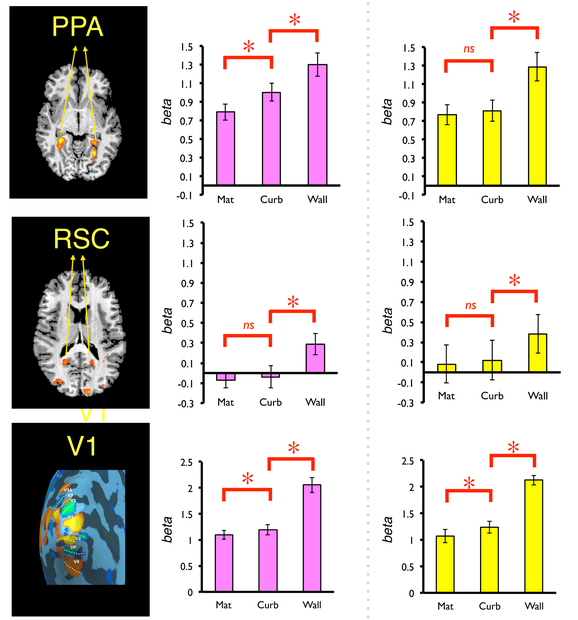
Experiment 2: fMRI

Stimuli:

Task:
1-back repetition detection



- CA control N=12; WS N=12
- Blocked design (12 s block); 24 blocks per condition
- TR=2s; 3x3x3 voxels
- Individual Scene localizer (Scenes-Objects) and Retinotopic localizer
- Number of voxels matched for CA and WS



- Exceptional sensitivity to Curb in CA's PPA
- WS don't show sensitivity to Curb! Curb is just like Mat
- RSC of both CA and WS only responds to Wall
- Previous finding suggests RSC's sensitivity to functional affordance of a boundary
- Low-level visual difference of Mat and Curb are represented in WS similar to CA

✓ Exceptional sensitivity to the Curb condition in the PPA found in CA control, but not in WS

Summary

- WS show impaired behavioral and neural sensitivity to the presence of 3D boundary cue
- Sensitivity to different types of environmental boundaries important for geometric reorientation