

Collinear Inhibition in Williams Syndrome?

Melanie Palomares¹, Barbara Landau¹, James E. Hoffman² and Howard Egeth¹

¹The Johns Hopkins University; ²University of Delaware

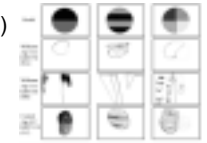
Williams Syndrome (WS) is a rare genetic disorder with a distinct cognitive hallmark: profoundly impaired visuospatial cognition and relatively spared language. To explore the contribution of "low-level" visual functions to WS deficits, we measured threshold contrast for detecting a grating with and without flankers (Fig.1-5 and Methods) in WS. We found that WS participants (10-33 years old) have contrast thresholds for detecting an unflanked grating that is as good as normal adults, and better than normal 3- to 5-year old children (Fig. 1). However, we found that WS were actually worse (i.e. inhibited) in the presence of flankers regardless of their distance and collinearity from the target (Fig. 2-5). Normal detection thresholds in WS indicate that the mechanism underlying their feature detection is intact, while the interference of flankers in WS suggests that they are impaired in tasks requiring spatial segmentation, perhaps reflecting abnormal attentional mechanisms. Failure in spatial segmentation could be partly responsible for the hallmark disorder in visuo-construction in WS (See side-panel.)

METHODS:

Gratings (3 c/deg) were presented as images in a booklet (600 dpi) at 57 cm viewing distance. Observers chose which panel, left or right, has the grating in the middle. We used an "eye chart" procedure to get threshold in which contrast decreases as trials progress. Threshold contrast was the contrast at which the observer made the first error. There were 22 trials per booklet. Threshold ratio was calculated as $\text{threshold}_{\text{flanked}} / \text{threshold}_{\text{unflanked}}$

WILLIAMS SYNDROME:

- Locus of gene deletion (7q11.23)
- Occurs in 1/20000 live births
- Elfin features
- Weak heart
- High rate of amblyopia
- High rate of hyperacusis
- Mild-moderate mental retardation (mean IQ=60)
- Poor in visuo-construction task
- Highly social, verbal
- Smaller brain volume, specifically V1, PPC, Amg. (Larger cerebellum)



Summary Table: Effect of Flankers

	NEAR	FAR	ORTHOGONAL	JITTERED
Williams Syndrome (10-33 years old)	Inhibition	Inhibition	Inhibition	Inhibition
Normal Children (3-5 years old)	Inhibition	Neither	Inhibition	Inhibition
Normal Adults (18-30 years old)	Facilitation	Neither	Facilitation	Neither

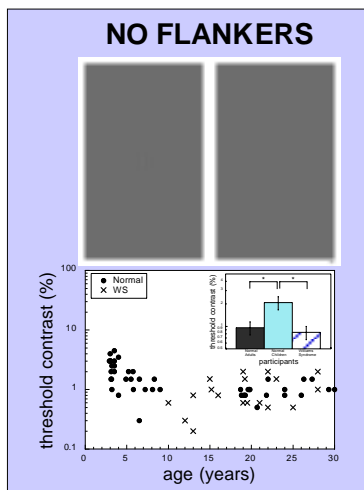


Figure 1. Top: No flanker stimuli. Target presented at 3% contrast on the left. Bottom: Threshold contrast as a function of chronological age. WS participants (n=19) have same threshold contrast as normal adults (n=18) and better than normal 3- to 5-year old children (n=17). See inset for averaged thresholds of participant groups. * indicates significant difference between groups $p < 0.05$.

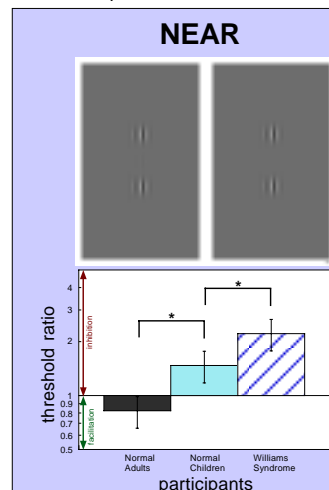


Figure 2. Top: Near collinear flanker stimuli. Target presented at 3% contrast on the left. Flankers were 3 wavelenghts away from the target (center-to-center). Bottom: Threshold ratio for participant groups. WS participants (n=19) and normal children (n=17) show inhibition while normal adults (n=18) show facilitation. * indicates significant difference between groups $p < 0.05$.

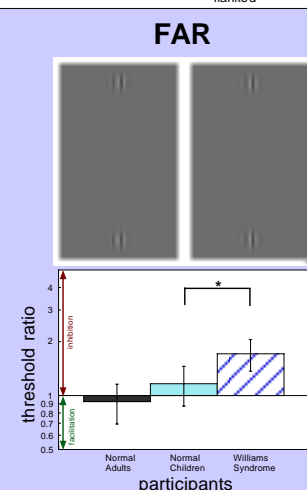


Figure 3. Top: Far collinear flanker stimuli. Target presented at 3% contrast on the left. Flankers were 9 wavelenghts away from the target (center-to-center). Bottom: Threshold ratio for participant groups. WS participants (n=19) show inhibition, while normal children (n=15) and normal adults (n=18) show no effect of flankers. * indicates significant difference between groups $p < 0.05$.

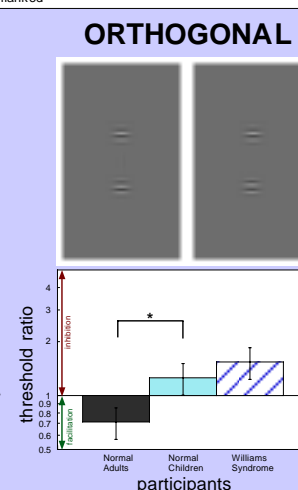


Figure 4. Top: Orthogonal flanker stimuli. Target presented at 3% contrast on the left. Flankers were 3 wavelenghts away from the target (center-to-center). Bottom: Threshold ratio for participant groups. WS participants (n=9) show and normal children (n=11) show inhibition, while normal adults (n=11) show facilitation. * indicates significant difference between groups $p < 0.05$.

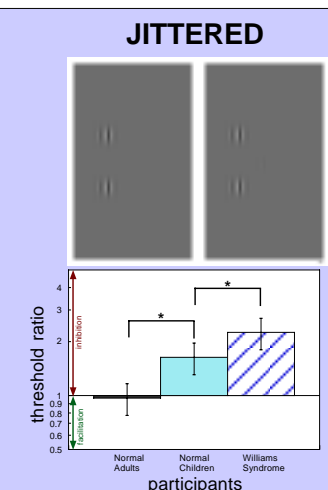


Figure 5. Top: Jittered flanker stimuli. Target presented at 3% contrast on the right. Flankers were 4.2 wavelenghts away from the target (center-to-center). Bottom: Threshold ratio for participant groups. WS participants (n=10) and normal children (n=7) show inhibition while normal adults (n=12) show no effect of flankers. * indicates significant difference between groups $p < 0.05$.