Impaired Visuospatial Processing in Young Adult Male Fragile X Premutation Carriers

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What is visuospatial processing?

- Adult fXPCs exhibit intact function in pathways involved in color and object recognition
- Impaired function in pathways involved in motion perception, detection of spatial location, and visuomotor coordination (Kéri & Benedek 2009, 2010, 2012)
- Adult male fXPCs have intact perception, but impaired visuospatial performance (Hocking et al, 2012)
- Where in the processing stream is the root of impairment?
Aims of the Current Study

- **Aim 1**: Investigate whether adult male fXPCs asymptomatic for FXTAS exhibit impaired visuospatial functioning or visuospatial attention
  - Results might provide a basis for a biomarker for later neurodegeneration

- **Aim 2**: Examine whether genetic “dosage” (e.g. CGG repeat length, gender) modulates behavior
  - Compare male and female performance

- **Aim 3**: Examine whether performance declines w/age
Study Design

- Participants included 48 adults (aged 18 - 45)
  - 21 male fXPCs
  - 27 HCs

- 5 Behavioral tasks
  - Psychomotor speed (manual and oral)
  - Magnitude Comparison (distance effect)
  - Enumeration (numerical spatial attention)
  - Spatial cuing
Psychomotor Speed Task

“Press the button as soon as you see the alien”

- HCs replicate published population values, as do male fXPCs

- Adult male fXPCs, unlike females show similar psychomotor speed compared to HCs

Magnitude Comparison Task

“Which of the two blue bars is longer?”

- Male fXPCs are slower
- Performance relates to CGG
Enumeration Verbal Task

“How many green items do you see in the red square?”

- Male fXPCs are slower in counting

**Median RT / Oral Motor RT ± S.E.**

**Percent Error ± S.E.**

**Numerosity**

1 2 3 4 5 6 7 8

**Subitizing Range**

(< 100 ms / item)

**Counting Range**

(> 250 ms / item)

**Age (in yrs)**

20 25 30 35 40

**CGG Repeats**

50 100 150 200

**Subitizing Range**

(< 100 ms / item)

**Counting Range**

(> 250 ms / item)

**RT (in ms)**

500 1000 1500

**Number of Items**

1 2 3 4 5 6 7 8

**Expected Performance**

Spatial Cuing

"Use the clues to find the target"

Male fXPCs perform similarly to HCs

HC (n = 17)

fXPC (n = 16)

Cue effect

r = .05

r_{HC} = -.31

r_{fXPC} = -.01
Summary of Results

- Adult male fXPCs do not show enhanced psychomotor speed, unlike female fXPCs.
- Adult male fXPCs are slower in magnitude comparison and numerical spatial attention.
- Magnitude comparison performance worsens with increased CGG.
- Results suggest visuospatial processing is impacted in fXPCs, but attentional orienting is not affected.
Implications & Future Plans

- Results support previous findings of impaired performance in visuospatial tasks in fXPCs.
- They add to a growing body of literature characterizing the phenotypic spectrum produced by $FMR1$ gene dosage.
- Lack of robust effects or associations may be due to limited age and CGG sampling range. (c.f. Hocking et al, 2012 and Cornish et al, 2011)
- Data from fXPC boys will help determine whether performance progressively declines or remains stable.
Thank you

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