How does the visuo-spatial deficit impact basic numerical processing in Williams syndrome? The question of domain specificity

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Williams Syndrome : cognitive profile

Known to be highly unequal:

- IQ~60 : majority of mild mental retardation [low average to severe MR]
- VIQ > NVIQ
- Relatively spared language
- Relatively spared face processing but still peculiarities
- Deficient visuo-spatial capacities : Local > global processing
- Academic cursus : slight evolution in reading and spelling until adult age
- Rapid stagnation of math development from adolescence (Udwin et al., 1996)
About math development...

- Verbal number sequence can be retained but remains inflexible  
  Paterson et al. (2006)

- Counting procedure **OK** (overlearned, *How many, Give me n*)  
  (Ansari et al. 2003)

- Single digit Arabic number reading **OK** in WS adults  
  (Ansari et al., 2007)  
  but multi-digit Arabic number reading < Down syndrome  
  (Paterson et al., 2006)

- Some WS adults **OK** to check one-digit additions and multiplications

- Better performance for small calculations learned by root  
  (Krajcsi et al., 2009)
These studies:

- Information about the *nature* of the difficulties experienced quite late in the development

- But no information about the *origins* of these difficulties, about basic numerical processing
  (= foundation of math competence)
Basic numerical processing?

Early sensitivity to numerosities: \(\cong\) Approximate Number System

- Imprecision with the nb of elements to quantify:
  - Size, Distance and Ratio effects
  - Weber fraction: Smallest numerical change to a stimulus that can be reliably detected
    => “numerical acuity” (Halberda & Feigenson, 2008)

- Innate/precocious: Independent of learning
- Basis of subsequent math learning
ANS in WS

- **Young WS children** (CA = 35 months; DA = 22 months):
  
  Difficulty to detect the difference between collections of 8 vs 16 dots (Van Herwegen et al., 2008)

- **Older WS children and adults** (CA = 20 y-old [10-32 y] ; MA = 6;9 y [5-9y])
  
  Difficulty to determine which collections has more dots
  
  % children of the same MA (Paterson et al., 2006)
Deficit of the ANS in WS?

➡️ Premature ...

→ Only tested with visual quantities involving processing the stimuli position in space

But, WS have important visuo-spatial difficulties

➡️ How do they process quantities in tasks with no visuo-spatial processing requirement?
Study : Quantitative processing in WS

Impact of visuo-spatial deficit on quantitative processing : 2 types :

- Continuous/ Non numerical → Experiment 1
- Discrete / non symbolic numerical → Experiment 2

The focus on continuous quantitative processing?

Walsh (2003) : central magnitude system for the processing of time, space and number magnitude information.

Simon (2008, 22q11 deletion syndrome) : Spatiotemporal processing form the basis of numerical and mathematical competence: Spatiotemporal processing deficit create “suboptimal foundation for the subsequent development of numerical and mathematical competence, thereby “cascading” impairments into those more academic domains”
XP 1: Continuous quantities

- 20 SW & 20 TD children matched on verbal MA

- 2 tasks:
  - Visual
    - « Show me the longest bar? »

- Ratio of increasing difficulty = > Weber fraction

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- Hypothesis:
  - If difficulty to process continuous quantities, then WS patients should have weaker performance in both tasks
  - If their visuo-spatial deficit interferes with visual quantitative processing, they should be impaired in the visual modality only.
XP 1: Continuous quantities: Weber fraction

Wilcoxon: Group effects:
- Length: $p = .001$
- Duration: $p > .10$
XP 1: Continuous quantities: % correct responses

Wilcoxon: Group effects (ratio 7/8)

- Length: p = .02
- Duration: p > .10
XP 2: Non symbolic numerical quantities

- 20 SW & 20 TD children matched on verbal MA

- 2 conditions:
  - Spatial
  - «Who has more pieces of puzzle?»
  - 2 sec

- Ratio of increasing difficulty

- Hypothesis:
  - If difficulty to process non symbolic numerical quantities, WS patients should have weaker performance in both tasks.
  - If their visuo-spatial deficit interfere with visual quantitative processing, they should be impaired in the spatial condition only.
XP 2: Non symbolic numerical quantities: Weber Fraction

Wilcoxon: Group effects:
- Collection: p = .008
- Dot sequence: p > .10
XP 2: Non symbolic numerical quantities: % correct responses

Wilcoxon: Group effects (ratio 3/4)
- Collection: $p = .03$
- Dot sequence: $p > .10$
Conclusion

SW patients:

- Difficulty to process numerical and non numerical quantities involving visuo-spatial processing

- Much less difficulty to process numerical and non numerical quantities distributed in time, and thus involving no visuo-spatial processing

=> WS difficulties in quantitative processing task are not domain specific.
Thank you for your attention
Math Development

- Verbal number sequence can be retained but remains inflexible
  - Difficulty to count between 2 numbers (25 to 35)
  - Difficulty to count backwards (20 to 1)
  - Difficulty to give the number coming after $n$

  Paterson et al. (2006)

- Counting procedure OK: (overlearned)
  - *How many*
  - *Give me n*

  Ansari et al. (2003)

\[ \text{% Down syndrome (same MA et CA)} \]

= children matched on visuo-spatial MA
Math Development

- Transcoding:
  - Single digit Arabic number reading **OK** in adults (Ansari et al., 2007)
  - But multi-digit Arabic number reading < Down syndrome of the same MA et CA (Paterson et al., 2006)

- Arithmetic:
  - Some WS adults **OK** to check one-digit additions and multiplications
  - Better performance for small calculations learned by root (Krajcsi et al., 2009)
**XP 1 : Continuous quantities : Correct responses**

![Graph showing correct responses vs. ratio n2/n1 for Length and Duration tasks for SW and TD groups.](image)

Mann-Whitney: Group effects (ratio 7/8):

- **Length**: N = 20 : p = .02
- **Duration**: N = 16 : p > .10
Study 2: Symbolic numerical processing

- Association symbol ------ quantity
  - Difficult step for dyscalculic children
  - Also difficult for WS patients
    (Paterson et al., 2006; Ansari et al. 2007; Krajsci et al., 2009)

Ex: SW < TD children matched on MA (O’Hearn & Landau, 2007):

But! Only tested Arabic number symbols (visual decoding)
Study 2: Symbolic numerical processing

- 17 SW & 17 TD children matched on verbal MA

- 2 tasks:

  - Visual
    - « Which number is the larger? »
    - 6 9

- Hypothesis:
  - If difficulty to associate a symbol to its quantitative meaning, then WS patients should have weaker performance in both tasks.
  - If their visuo-spatial deficit interfere with visual symbolic processing, they should be impaired in the visual comparison task only.
Study 2: Symbolic numerical processing

Wilcoxon: Group effects (ratio 3/4)

- Arabic number: p < .05
- Verbal number: p < .01