Symbolic numerical processing deficit in people with Williams syndrome.

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Introduction

The last decade, behavioral evidence suggested that early non verbal numerical abilities rest on two core systems (Feigenson, Dehaene & Spelke, 2004). The first one, called the *Approximate Number System* (ANS), permits the representation of large, approximate numerical magnitudes The second one allows keeping track of a small number of elements and would be responsible for the *subitizing* phenomenon, that is, the ability to rapidly enumerate up to four briefly presented items. Different lines of evidence suggest that these two systems would have strong relationships with spatial cognition as approximate numerical representation would be grounded in our ability to process non-numerical magnitudes (Walsh, 2003; Bueti & Walsh, 2009; Simon 2008) while the mental keeping track of a small number of elements would tap in our ability to assign simultaneously a small number of spatial indexes coding for the spatial coordinates of up to about 4 targets (Trick &Pylyshyn, 1994; Kahneman, Treisman & Gibbs, 1992).

Lately, some authors speculate about a possible defection of the ANS in patients with Williams syndrome (WS) considering their difficulty to process numerical magnitudes (Krajcsi et al., 2009; O'Hearn & Landau, 2007; Paterson et al., 2006). As patients with WS were always tested in the visual modality, it remains unclear whether their deficit is specific to the processing of numerical magnitude or result from their basic visuo-spatial impairment (main characteristic of the WS cognitive phenotype). A first set of results supported the second hypothesis as people with WS were shown to have lower numerical acuity only in numerical tasks with high visuo-spatial processing requirements (i.e. comparing two lengths or two arrays of elements but not when comparing two durations or two sequences of flash in a single location; Rousselle & Noël, 2013). Here, we extend our investigation by examining the access to the meaning of visual and verbal numerical symbols in these patients, asking them to compare two Arabic digits or two spoken verbal numerals.

Moreover, recent results attested for the presence of a reduced ability to keep track of a small number of element patients with WS, resulting in subitizing abilities comparable to those of 4-y-old children (O'Hearn et al., 2005, 2011). This study extend those results attempting to determine whether their subitizing ability is comparable to the one expected of their non verbal developmental age.

Method

Participants :

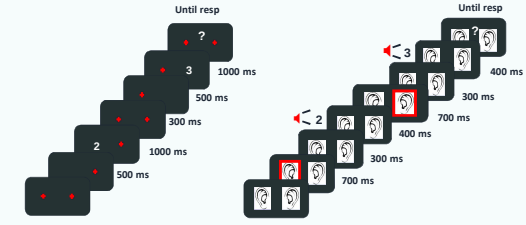
- **21** participants with **WS** : chronological age (CA)= 21; 11 y-o [5;5 – 52;10]
- **21 verbal-matched typically developing** children (TDv): CA= 7; 6 y-o [4;6-11;8]
- **21 nonverbal-matched typically developing** children (TDnv): CA= 6; 1 y-o [3;6-10;4]

⇨ WS < TDv on non verbal developmental age, visuo-spatial abilities and math abilities
⇨ WS = TDnv on non verbal developmental age, visuo-spatial abilities and most math abilities
> TDnv on verbal abilities

Tasks :

		Stimuli	Task parameters
	Symbolic Numerical comparison	• **Arabic digit :** • **Spoken verbal numerals**	• Distance 1 : pairs 2-3 and 7-8 • Distance 2 : pairs 3-5 and 6-8 • Distance 3 : pairs 2-5 and 6-9
	Numerical estimation task	Black dots collections	• Brief presentation : 200 ms • Numerosties : 1 to 7 dots • Followed by a mask
Control task	Color naming task	Colored dots collections	• Same as the estimation task • 4 possible colors
Control task	Give a number Task	Tokens	• Numerosities : 1, 2, 3, 4, 5, 6, 8, 10, 14
Control task	Fast Counting task *« Count the perls as fast as you can »*	Perl necklaces	• Numerosities : 6, 7, 8, 9, 12, 14, 16, 18 • Finger pointing on the screen authorized

In the Arabic digit comparison tasks, digits were always presented sequentially, the first one on the left side of the screen, and the second one on the right side, in order to equilibrate the working memory load with the spoken verbal comparison task.



Digit comparison Verbal numerical comparison

Results

Preliminary analyses :

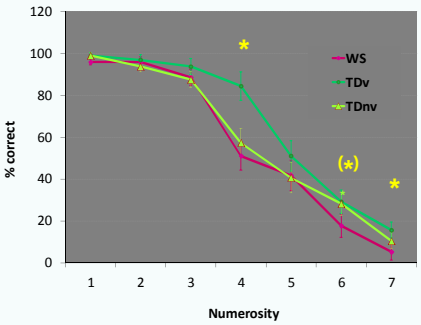
Number of participants	WS	TDnv	TDv
Color naming task OK (>50% correct)	20	21	21
Subitizing 1 dot (at least 2/6 correct)	16*	21	21

*$\chi^2(2) = 10,9$; $p = .004$

- ***Give a number* Task:**
  19 WS patients can give consistently at least 5 tokens

- **Fast Counting Task :**
  20 WS participants can count :
  - Performance reached at least 4/8 correct
  - 82% of the errors distributed on numerosities > 10

Numerical estimation task



Subitizing analyses :

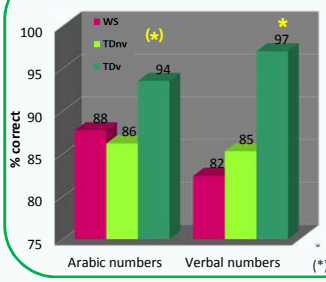
Anova Numerosity (4) x group (3) :
- Group : p = .05: WS < TDv & WS= TDnv
- Numerosity x group : p < .005

Anova Numerosity (3) x group (3) :
- Group : p > .10
- Numerosity x group : p > .10

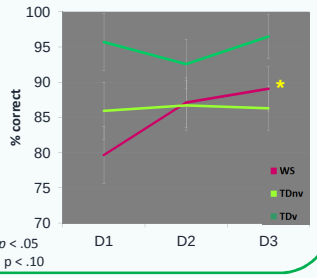
Paired comparison by numerosity
- WS = TDv for all numerosities
  ….except for the numerosity 4 (p< .005) & 7 (p< .05)
- WS = TDnv for all numerosities

Mean subitizing rank		
WS	TDnv	TDv
2.3	3 (p = .06)	3.5 (p = .001)

Symbolic numerical quantities



* p < .05
(*) p < .10



In Summary

- Participants with WS showed lower precision in accessing the numerical meaning (ANS) of numerical symbols as attested by their deficit in both symbolic numerical processing tasks. These difficulties could thus not be attributed to their visuo-spatial difficulties.
- They showed a smaller subitizing range in accordance to what would be expected on the basis or their visuo-spatial capacities.

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