

Evidence for a global access deficit to the meaning of numerical symbols in people with Williams syndrome



Laurence Roussele¹ & Marie-Pascale Noël²

¹ Unit of Cognitive Developmental Psychology, Cognition and Behavior Department, Faculty of Psychology and Education, University of Liège, Belgium
² Institute of Psychology (IPSY), Catholic University of Louvain, Belgium, National Fund for Scientific Research of Belgium

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 & Pylshyn, 1994; Kahneman, Treisman & Gibbs, 1992).

Lately, some authors speculate about a possible deflection 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; Roussele & 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 :

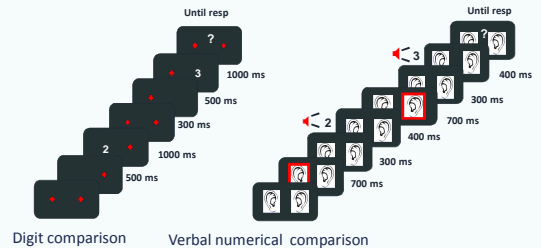
- 21 participants with WS : chronological age (CA) = 21; 11 y-o [5;5 – 52;10]
- 21 verbal-matched typically developing children (TDv): CA= 7; 6 y-o [4;6-11;8]
- 21 nonverbal-matched typically developing children (TDnv): CA= 6; 1 y-o [3;6-10;4]

- ⇒ 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	<ul style="list-style-type: none"> • Arabic digit : • Spoken verbal numerals 	<ul style="list-style-type: none"> • 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	<ul style="list-style-type: none"> • Brief presentation : 200 ms • Numerosities : 1 to 7 dots • Followed by a mask
Color naming task	Colored dots collections	<ul style="list-style-type: none"> • Same as the estimation task • 4 possible colors
Give a number Task	Tokens	<ul style="list-style-type: none"> • Numerosities : 1, 2, 3, 4, 5, 6, 8, 10, 14
Fast Counting task	Perl necklaces	<ul style="list-style-type: none"> • 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.



Results

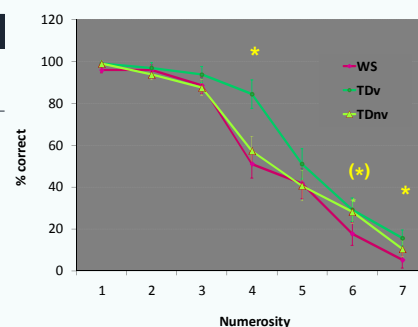
PRELIMINARY ANALYSES :

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



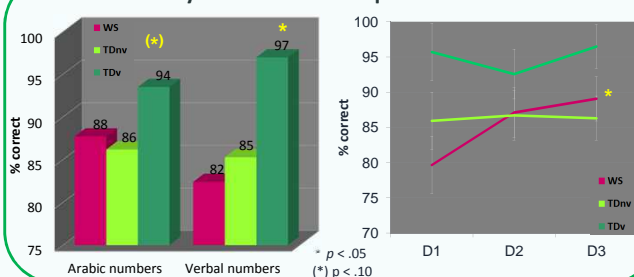
SUBITIZING ANALYSES :

- **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



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 of their visuo-spatial capacities.

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