Magnitude processing in populations with spina-bifida: the role of visuospatial and working memory processes

Lucie Attout[1,2], Marie-Pascale Noël[2] & Laurence Rousselle[1]

[1] Research Unit "Enfances", University of Liège, Belgium

[2] Psychological Sciences Research Institute, UCLouvain, Louvain-la-Neuve, Belgium

Abstract : People with Spina Bifida usually experience difficulties with mathematics. In a series of other developmental disorders, a magnitude processing deficit was considered to be the main source of subsequent difficulties in mathematics. The processing of magnitude could be numerical (which is the larger number) or non-numerical such as spatial (e.g., which is the longer?) or temporal (which one last longer?) for instance. However, no study yet has examined directly magnitude processes in a population with Spina Bifida. On the other hand, recent studies in people with genetic syndromes have suggested that visuospatial and working memory processes play an important role in magnitude processing, including number magnitude. Therefore, in this study we explored for the first time magnitude representation using several tasks with different visuospatial and working memory processing requirements, cognitive skills frequently impaired in Spina Bifida. Results showed children with SB presented a global magnitude processing deficit for non-numerical and numerical comparison tasks, but not in symbolic number magnitude tasks compared to controls. Importantly, visuospatial skills and working memory abilities could partially explain the differences between groups in comparison and estimation tasks. This study proposes that magnitude processing difficulties in children with SB could be due to higher cognitive factors such as visuospatial and working memory processes.

207 words

Keywords: Spina Bifida; Working memory; Visuospatial skills; Basic numerical abilities; Magnitude processing.

Total number of word: 8953

What this paper adds?

> The present study conducted a systematic examination of numerical and non-numerical magnitude processing abilities using tasks with different WM load and visuospatial processing requirements in children with SB in order to gain further insights into the possible causes of their math learning disabilities. For the first time, this study demonstrated that children with SB present a deficit in basic numerical processing and that this pattern of impairment may not be the result of a defective magnitude representation, but rather of cumulative deficits impairing visuospatial processing and WM abilities.

1. Introduction

Spina Bifida (SB) is a congenital developmental disorder which is caused by a neural tube defect diagnosed prior to or at birth and which impacts the spine and brain development. This defect occurs in 1 in every 1-2000 live births (Khoshnood et al., 2015). The cognitive phenotype of people with SB is mostly characterized by fine motor and visuospatial skills deficits (Lomax-Bream et al., 2007), low working memory (WM) capacities (see for example Mammarella, Cornoldi, & Donadello, 2003) and learning disabilities, especially regarding the mathematical domain (Ayr et al., 2005; Dennis & Barnes, 2002; Fletcher et al., 2005). The visuospatial skills deficit observed in most cases with SB is characterized by globally better performance in object identification (features and gestalts) than other visuospatial representations (see for a review Dennis & Barnes, 2010). For instance, children with SB performed more poorly than a control group of children matched on chronological age, in drawing, route finding, visual pursuit, line orientation or visual figure-ground tasks, however they showed no impairment in the perception of objects in degraded pictures or face recognition (Dennis & Barnes, 2002). The deficit in WM affects visual and spatial components but also, verbal WM abilities. More specifically, deficits have been reported in visual WM (Mammarella et al., 2003), in the storage and manipulation of visuospatial information (Dennis et al., 2007), as well as in the short-term maintenance and manipulation of verbal information (English et al., 2010; Vachha & Adams, 2005).

Within the numerical domain, early difficulties are reported in preschoolers with SB with regard to knowledge of counting principles, recitation of the counting string, and object-based arithmetic (English et al., 2009). A longitudinal study found that children with SB had difficulties, compared to typically developing peers, in all mathematical tasks proposed between three and five years old, such as counting (set counting, oral counting, mastery of counting principles and object-based arithmetic over the subitizable range) except for operations (additions and subtractions) on quantities in the subitizable range (Barnes et al., 2011). Later, older children and adolescents were found to exhibit weak performance in procedural calculations such as large additions, subtractions and more complex calculations (requiring multi-step processing), compared to their chronological age (Ayr et al., 2005; Barnes et al., 2002, 2005, 2006; Coughlin & Montague, 2011). These studies have documented persistent immature strategies to solve arithmetic problems with reports of counting-based strategies in people with SB. By contrast, arithmetic facts (as required to solve multiplications) tended to be in the normal range. A recent study in nine year old children with SB, indicated weaknesses in approximate (a forced-choice approximate additions task) and standardized arithmetic measures (Woodcock-Johnson Tests of Achievement; including the calculation and math fluency subtests) when compared with a chronologically-matched group while in an exact arithmetic measure (a forced-choice exact additions task), they presented age-appropriate performance (Raghubar et al., 2015).

Altogether, these studies indicate that children with SB have lower numerical and calculation abilities compared to their peers but the origin of their difficulty is still unclear. Some neuro-behavioural assumptions have been suggested previously such as that the congenital abnormalities associated with hydrocephalus in people with SB, clearly correlated with poorer outcomes in motor and perceptual skills or that the subcortical changes in their brain could be related to mathematical difficulties (Burmeister et al., 2005; Ewing-Cobbs et al., 2003; Fletcher et al., 2005). However, the neural profile of people with SB is not so clear and tends to evolve with medical cares, as observed by a decrease of a potential impact of hydrocephalus (Copp et al., 2015). At the cognitive level, there are two major explanatory trends to account for their mathematical learning disabilities coming from the literature on children with specific mathematical learning disabilities. For some authors, mathematical learning difficulties are thought to result from non-numerical cognitive deficits (see for a discussion Fias, Menon, & Szűcs, 2013) such as WM (see for a review Allen, Higgins, & Adams, 2019 and Raghubar, Barnes, & Hecht, 2010), visuospatial abilities (see for example Ashkenazi, Rosenberg-lee, Metcalfe, Swigart, & Menon, 2013; Simms, Clayton, Cragg, Gilmore, & Johnson, 2016), phonological awareness (Jordan, Wylie, & Mulhern, 2010; Krajewski & Schneider, 2009), sensitivity to interference (Barrouillet et al., 1997; De Visscher & Noël, 2014) or order processing (Attout & Majerus, 2014; Morsanyi et al., 2018). Interestingly, a longitudinal study found that visuospatial and fine motor abilities uniquely predicted large set object-based arithmetic performance, while both phonological awareness and visuospatial ability were uniquely predictive of oral counting both in children with SB and with typical development (Barnes et al., 2011). With regard to WM, a recent study in nine year old children with SB found, with multiple mediation models, that visuospatial WM mediated the relationship between group and both math fluency and math calculation tests. Moreover, some studies have reported that verbal and visuospatial WM deficits in children and adults with SB are related to their calculation abilities (Dennis & Barnes, 2002; Raghubar et al., 2015).

Another well-documented theory concerning the origins of mathematical learning disabilities considers that it could result from a basic dysfunction of numerical cognition itself. Developmental theories of numerical cognition suggest the existence of two primitive (even innate) systems for representing numerical magnitude that serve as the basis for subsequent mathematical development. The first one is an *Approximate Number System* (ANS), also called the magnitude representation, which would allow babies to represent large numerical magnitudes imprecisely (Dehaene & Changeux, 1993; Feigenson et al., 2004) and later the magnitude of numbers whatever the notation (symbolic like Arabic numbers or non-symbolic like dots) (Gilmore et al., 2014). Some authors consider that this primitive representation might be defective in dyscalculia while other postulate that the magnitude representation itself may be intact but that the connections between numerical symbols and the magnitude representation could be deficient (De Smedt & Gilmore, 2011; De Smedt et al., 2013; Landerl et al., 2004; M.-P. Noël & Rousselle, 2011). The second system is an *object tracking system*, which facilitates the exact representation of a small number of elements (Feigenson et al., 2004). This mechanism may be responsible for the phenomenon of *subitizing* (i.e. the ability to enumerate up to three or four elements very quickly and precisely, without counting them), assumed to involve the parallel deployment of a limited number of visuospatial indexes (Burr & Anobile, 2010; Trick & Pylyshyn, 1994) and to rely on visuospatial WM ability (Piazza et al., 2011). This mechanism is thought to play a crucial role in the acquisition of the cardinal value of number words (Le Corre & Carey, 2007) and was shown to be impaired in dyscalculic children (Koontz, 1996; Schleifer & Landerl, 2011).

At present, very little is known about how these two primitive systems develop in children with SB. Aside from studies which show difficulties in early numeracy skills such as counting and object-based additions and subtractions (Barnes et al., 2011), only one study on adults has shown a deficit in everyday mathematical competence tasks, such as making price comparisons (Dennis & Barnes, 2002). Furthermore, only symbolic magnitude representation (assessed with an Arabic number magnitude comparison) has been tested but non-symbolic magnitude representation has never been examined.

However, the non-symbolic magnitude representation is not so easy to assess purely. In the last few years, it has been convincingly shown that perceptual variables covarying with numerical magnitude (e.g. density, sum of perimeter, surface area, length or size) have a large influence on performance in numerical tasks using visual sets (Clayton et al., 2015; Dormal & Pesenti, 2007; Gebuis et al., 2016; Gebuis & Reynvoet, 2012a; Leibovich et al., 2017; Rousselle & Noël, 2008; Szűcs et al., 2013). There is a growing consensus that there is no pure task to measure the precision of the magnitude representation as, beyond the processing of numerical magnitude itself, number magnitude comparison tasks (task where the participant needs to say on which side the numerical magnitude was the largest) involve general cognitive processes which differ depending on the task. More specifically, when stimuli (often dots) are presented simultaneously to form a set, as in visual collections comparison, performance is largely dependent on visual perceptual dimensions and thus recruits more general visual-spatial skills. By contrast, when stimuli are presented successively, as in the comparison of two sequences of sounds, participants need to accumulate number magnitude, as the stimuli appear to create a representation of the magnitude of the whole sequence. Therefore, this task would largely be sustained by WM processes (see also the accumulator model of Meck & Church, 1983).

In line with this debate, a series of recent studies was conducted to shed light on the basic numerical processing deficit at the origin of mathematical learning disabilities in different populations with genetic anomalies. Results indicated that numerical magnitude representations and the keeping track mechanism could be differentially impaired in different genetic syndromes (Mazzocco & Hanich, 2010; Oliveira et al., 2014; Paterson et al., 2006; Simon et al., 2008; Smedt et al., 2009; Van Herwegen et al., 2008). Interestingly, the pattern of performance in the magnitude comparison task might vary depending on the presence of visuospatial and WM deficits (Attout, Noël, Nassogne, et al., 2017; Attout, Noël, Vossius, et al., 2017; Rousselle et al., 2013). More specifically, while people with Williams or and 22q11.2 deletion syndromes, with lower visuospatial skills, presented a specific impairment to magnitude tasks that involved some visuospatial processing (Attout, Noël, Vossius, et al., 2017; Rousselle et al., 2013), adults with Turner syndrome, presenting lower WM abilities, had a lower acuity only when comparing two sequences of events, that is, when the magnitude had to be accumulated over time (Attout, Noël, Nassogne, et al., 2017). These data suggest that both WM and visuospatial skills can have a direct influence on the processing of magnitudes and may determine the profile of performance in different types of magnitude comparison tasks. In other words, difficulties in WM or visuospatial skills could interfere with the processing of non-symbolic number magnitude presented sequentially or visually. Accordingly, we cannot conclude in terms of a “pure” ANS deficit in these populations. However, given that numerical magnitude are mostly apprehended visually or sequentially when presented in the auditory modality, these difficulties could lead to weaknesses at a more elaborate level of numerical cognition.

Therefore, in this study, we explored this question in another specific atypical neurodevelopmental population, presenting both, visuospatial and mathematical difficulties without any intellectual disabilities. The goal was twofold: (1) to gain further insights into the possible causes of math learning disabilities and (2) to better characterize the basic numerical deficits in children with SB in order to offer appropriate cares to these children such as training more basic level of cognition to develop mathematical abilities.

Therefore, the present study conducted a systematic examination of numerical and non-numerical magnitude processing abilities as well as of subitizing abilities (i.e. the ability to enumerate up to three or four elements very quickly and precisely, without counting them) using tasks with different WM load and visuospatial processing requirements. With regard to non-numerical magnitudes, participants were asked to compare lengths (with visuospatial demand but with low WM load, all the quantity being entirely available directly) and durations (with WM requirement due to the sequential presentation of stimuli but no visuospatial demand). With regard to numerical magnitude processing, four types of tasks were administered: (1) two sequential numerical comparisons, one with sequences of dots and the other one with sound sequences which put strong emphasis on verbal WM but which do not involve visuospatial processing; (2) one numerical comparison of visual collections which put lower emphasis on WM but which required deep visuospatial processing to extract collections; (3) one Dot estimation task which mainly put emphasis on visuospatial WM to extract a small number of dots; (4) finally, two symbolic numerical comparison tasks, one with verbal numerals and another with Arabic numerals which put lower emphasis on visuospatial and WM aspects. Moreover, an arithmetical fluency task was also proposed to assess the mathematical proficiency of the participants to ensure that participants with SB presented some difficulties in a higher level of mathematical abilities.

Based on the characteristic cognitive profile of the people with SB (see above), the following predictions were made. People with SB are expected to have lower arithmetical performance than the controls, especially in the tasks that require procedural skills, that is, in the addition and subtraction tasks. Second, they are expected to have lower visuospatial skills than control participants and, consequently, to perform more poorly than controls in the length and the collection comparison tasks, which have a high visuospatial demand. Third, they are expected to have lower WM capacities and consequently, to have lower performance than control participants in tasks that load heavily on these capacities, i.e., the magnitude comparison of sequences of dots and of sounds (as suggested for Turner syndrome in Attout, Noël, Nassogne, et al., 2017). We also expect difficulties in the duration comparison task (requiring participants to accumulate an amount of information step by step, as the auditory stimuli cannot be provided all at once) and in the Dot estimation task which specifically taps into visuospatial WM performance (Piazza et al., 2011). Finally, they are expected to have no difficulties in tasks that require only minimal visuospatial skills or WM capacities, i.e., the comparison of symbolic numbers.

2. Methods

2.1. Participants

Twenty-three children with the most common type of SB, meningomyelocele, aged between seven and sixteen years old and comprising ten females participated in this study.

Participants were recruited through the department of pediatric cardiology at two University Hospitals. Eight participants did not present hydrocephalus while all the others have been shunt-treated for their hydrocephalus during the first months of their life. Of the twenty-three children with SB, seven participants had repeated one grade and three of them were in a specialized curriculum. Moreover, the majority of the group presented some difficulties with mathematical abilities but not all the group as reported by the parents, the teachers or by a specialist but not systematically based on a clinical diagnosis. The control group was composed of twenty-three typically developing children (twelve females) matched one to one with an SB child at +- four months of chronological age (CA), with no history of neurological or psychiatric disorders, hearing or visual impairments (uncorrected), or neurodevelopmental disorders. We based the number of participants on previous studies in this field (usually, twenty participants) (Attout, Noël, Nassogne, et al., 2017; De Smedt et al., 2009; Murphy & Mazzocco, 2008; Rousselle et al., 2013).

2.2. Material

2.2.1. IQ measures

Different subtests from the Wechsler intelligence scales for children were used (Wechsler Preschool and Primary Scale of Intelligence–third edition, WPPSI–III; or the Wechsler Intelligence Scale for Children– fourth edition, WISC–IV; depending on the child's age). The Block Design subtest was used to measure visuospatial abilities while the Concept identification subtest was used to estimate non-verbal intelligence. Finally, Vocabulary and Similarity subtests from were used to estimate verbal IQ. Both, raw and standard scores were computed.

2.2.2. Working memory

Using the same procedure as Attout et al. (2017), the three main components of WM were examined in tasks that did not require the recall or manipulation of numerical content.

Short-term storage of verbal information was assessed in a *forward letter span task* (three to nine letters). Sequences of letters (starting with two) were presented orally to the participant at a rate of one per second and the participant had to repeat the letter in the same order. Sequences increased in length and participants had to complete two sequences at each length before moving on.

Short-term storage of verbal information was assessed with a two-dimensional *visuospatial span task* inspired by the Corsi task including a span from two to ten and where participants had to remember the location of a series of cells in a blank grid previously indicated one at a time by the examiner, at the rate of one cell per second. The participant had to place tokens on the cells previously indicated by the examiner (two tokens were offered for span two, three for span three and so on).

Finally, a *category-span task* (M. P. Noël, 2009) was used to examine the storage and manipulation of verbal information at short-term. One-syllable words related to food and animals were presented orally at a rate of one word per second to the participants, who had to repeat them back, starting with the food words first and then the animal words. Sequences started with two words, reaching a maximum of nine words. For all tasks, participants had to succeed in two trials of the same difficulty (or number of items) to access a higher level (span +1). The tasks ended when the participant failed at two (or more) out of the three trials for a given difficulty level. Each correct response was credited with one point. The dependent variables correspond to the total of points credited.

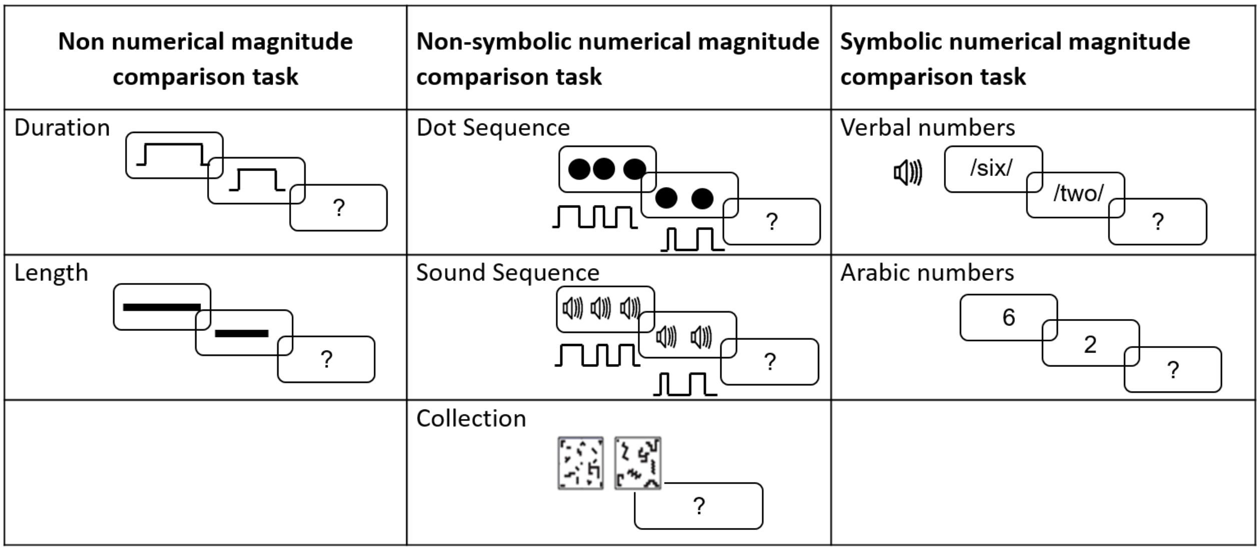
2.2.3. Mathematical level

To assess mathematical achievement, *Single-digit arithmetic fluencies* were used and consisted of three tasks of eighty-one problems each, involving additions, subtractions and multiplications. For each operation, participants were presented with one sheet containing a series of written arithmetic problems, and had one hundred and fifty seconds to give a written response to as many problems as possible. Addition and multiplication problems were drawn from all possible combinations of the integers one to nine and the set of subtractions was the exact counterpart of the addition set. In each task, the experimenter scored the number of correct responses given in the allotted time and the number of errors.

2.2.4. Magnitude comparison tasks

Seven magnitude comparison tasks were used (see Figure 1). These tasks differed in the nature of the magnitudes to be compared (numerical or non-numerical) and in the emphasis put on visual and/or spatial and STM processing. There were two non-numerical magnitude comparisons (length comparison and duration comparison) and three numerical comparison tasks with non-symbolic stimuli using different presentation modes (collection comparison, dot sequence comparison and sound sequence comparison). The two last tasks assessed symbolic numerical magnitude processing through verbal and Arabic numerical comparisons. These tasks have already been used in previous studies (Rousselle et al., 2013; Attout et al., 2017).

Figure 1. Illustration of the different magnitude comparison tasks.



In all tasks, participants had to compare two magnitudes and select the larger one. For all non-symbolic magnitude comparison tasks, six different ratios between the quantities to compare were used: 1/2, 2/3, 3/4, 5/6, 7/8, 8/9. In each task, the side of the correct response was counterbalanced. Two different pairs of magnitudes were presented per ratio, giving a total of forty-eight stimulus pairs in each task (two pairs x two sides x two presentations x six ratios). Importantly, the ratios increased in complexity throughout the task to determine individual sensitivity to magnitude difference in each task. The task ended when a participant performed at chance level for two out of three consecutive ratios. Six practice trials with pairs of magnitudes differing by a 1/3 ratio were proposed to check the understanding of the instructions. The global accuracy score reached was used as the performance measure.

Non-numerical magnitude comparison tasks. These tasks require participants to process the duration or the length of continuous stimuli presented respectively in the auditory (no visuospatial processing) or in the visual modality (visuospatial processing requirement). In the *length comparison task*, they had to compare the length of two successively presented white lines while in the *duration comparison task*, participants had to compare the duration of two successive sounds. They were instructed to touch the screen on the side of the longest line/sound. The reliability (odd-even method to take into account to the increasing of the task difficulty) for the length comparison task was not good (.02) and moderate for the duration comparison task (.53). However, these values need to be considered cautiously since they were based on a very small sample (twenty-three participants, corresponding to the performance of the control group only since the children with atypical development could presented more inter- and intra- variability between participants).

Numerical magnitude comparison tasks. The three non-symbolic numerical comparison tasks assessed participants’ ability to process the discrete numerical properties of sets presented either sequentially (Dot Sequence or Sound Sequence, low visuospatial requirement and high

WM demands) or simultaneously (Collection, high visuospatial processing requirement and low WM demands).

In the two sequential tasks, one in visual and the other in auditory modality, participants had to compare the numerical magnitude of the two series of flashed dots/tones presented in rapid succession and had to choose the side that ‘‘saw/heard’’ the sequence containing the most dots/sounds. In the *Dot Sequence Comparison task*, the stimuli were sequences of a single white dot (diameter: 3.5 cm) flashed rapidly in a single location on the left and then on the right side of the screen. Instructions emphasized that the duration of the sequence was not important. They were instructed not to count the flashed dots as they would not have the time to do so. In the *Sound Sequence Comparison task*, stimuli were constructed in exactly the same way as the dot sequence comparison but dots were replaced by identical tones (audio format: 44100Hz, 32 bits, Mono).

In the *Collection Comparison task*, participants were asked to compare the numerical magnitude of two collections (two white boxes containing black puzzle pieces) displayed simultaneously on the screen and had to touch the screen on the side of the box that contained more pieces as soon as they got the answer, with no time limit after stimulus disappearance. In order to control as much as possible for the influence of perceptual non-numerical dimensions on the participants’ judgment, the numerical magnitude and the cumulated black area were manipulated and counterbalanced between both conditions, congruent and incongruent. For congruent trials, the numerically larger collections have also the larger cumulated black area while for incongruent trials, the numerically larger collections have the smaller cumulated black area. The form of the puzzle pieces was manipulated in order to ensure that the variations of cumulated black area were completely confounded with those of cumulated individual perimeter (i.e. sum of individual piece perimeters) and brightness. To avoid the larger collection in number being systematically the one with the smaller elements, the area of the smaller and larger pieces was the same in both arrays to be compared. Finally, the convex hull (external perimeter formed by the most external pieces) was equated for all trials (see in Rousselle et al., 2013 for more details). The odd-even reliability was .79 for the collection comparison task, .73 for the dot sequence comparison task and .79 for the sound sequence comparison task.

Symbolic numerical magnitude comparison tasks. Participants' abilities to process symbolic magnitude were assessed using Verbal and Arabic Number Comparison tasks. Both tasks followed exactly the same procedure and participants were invited to select the larger of two numerals presented successively (Arabic numbers: Arial, 48-point font; Verbal numbers: 44100Hz, 32 bits, Mono). The stimuli varied along the same ratio and corresponded to the same numbers as those used in the non-symbolic comparison tasks. In both tasks, the side of the correct response was counterbalanced: each pair (twelve pairs) appeared four times, twice in ascending (e.g. 2-3) and twice in descending (e.g. 3-2) order. We also included forty unanalyzed filler pairs (e.g. 1-2) in order to ensure that all numerals were considered equally as the smallest and the largest of a pair. For data analyses, we considered the global median correct response times (RT) for each participant.

2.2.5. Dot estimation task

Participants were briefly (200ms) presented with arrays of one to seven dots and were asked to say out loud ‘how many’ dots were presented as quickly and accurately as they could (see Attout et al., 2017 for an extensive description of this task). This presentation was immediately followed by a mask screen for 500ms, which consisted of dots of heterogeneous size and which covered the whole surface of the screen. Each numerical magnitude was presented six times in different configurations and the task was preceded by seven practice trials.

2.3. Experimental procedure

Participants were tested individually in a quiet room. Testing was completed in two seventy-five minutes sessions approximately, depending on participants' performance and attentional level. The first session started with IQ subtests, the three WM subtests followed by tasks assessing arithmetic and subitizing abilities. The second session consisted in the computerized basic numerical comparison tasks proposed in a Latin square order.

3. Results

3.1. Descriptive statistics

Table 1 presents the results for age, IQ, WM and mathematical measures in the SB and control groups. Two-sample t-tests were run to compare the SB and the control group on the descriptive measures by applying a Bonferroni correction for multiple comparison ($\alpha/10=.005$). As expected, participants with SB did not differ from the control group on chronological age or in the verbal intelligence subtests. By contrast, they presented significantly lower performance in the block design subtest, assessing visuospatial abilities. In order to give a clearer idea of the level of performance in the SB group, raw scores for each measure were converted into standard scores using the norms provided by the Wechsler's scales. The mean and standard deviation of the standard score in the SB group was 6.48 ± 3.19 (range from 1 to 14) for the block design subtest, 8.09 ± 2.23 (range from 3 to 12) for the concept identification subtest, 10.30 ± 3.10 (range from 5 to 17) for the vocabulary subtest and 11.26 ± 2.61 (range from 7 to 16) for the similarities subtest.

The SB group also had lower capacities in verbal WM and visuospatial WM than the control group but obtained similar performance in the storage of verbal information at short term as assessed by the forward letter span task.

With regard to the arithmetical fluency tasks, the SB group had lower performance, at a marginal level, than the control group in addition and subtraction but no difference between the groups was observed for multiplication (only twenty-one children with SB and twenty-one control participants of the same age, were presented with this task, the others being too young).

Table 1. Descriptive measures and statistics in the SB and control groups.

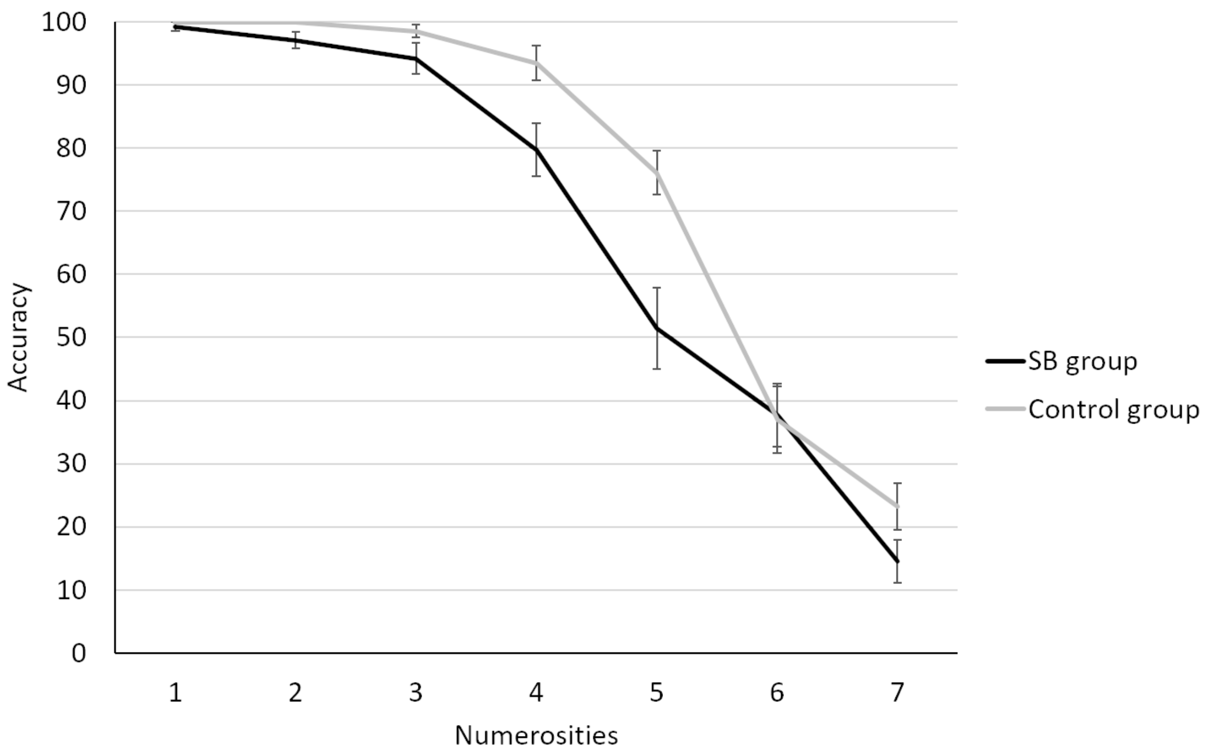
	SB group		**Control group**		**t(44)**	**η^2**	**p**
	Mean (SD)	**range**	**Mean**	**range**			
Age	145.26 (28.46)	94-199	145.65 (28.03)	98-199	-0.5	.00	.96
IQ measures *(raw score)*							
Block design (max. 68)	31.13 (13.19)	10-62	45.74 (8.92)	26-59	-4.40	.30	<.001
Concept identification (max. 28)	17 (3.45)	7-23	19.22 (2.19)	13-23	-2.60	.13	.013
Vocabulary (max. 68)	35.39 (9.17)	21-55	38.78 (7.66)	20-56	-1.36	.04	.18
Similarities (max. 44)	24.61 (4.77)	17-34	25.70 (4.66)	14-32	-0.78	.01	.44
Working memory							
Category-span (max. 16)	6.00 (1.45)	3-8	7.48 (1.08)	6-10	-3.93	.26	<.001
Visuospatial span (max. 26)	13.65 (2.48)	7-20	16.17 (1.83)	13-20	-3.93	.26	<.001
Forward letter span (max. 16)	7.61 (1.67)	6-12	8.39 (1.85)	5-12	-1.50	.05	.14
Mathematical fluency							
Addition fluency (max. 81)	21.87 (15.58)	1-81	29.96 (14.75)	11-74	-1.81	.07	.08
Subtraction fluency (max. 81)	18.13 (11.76)	0-57	26.52 (12.83)	7-66	-2.31	.12	.02
Multiplication fluency (max. 81)	19.05 (13.25)	0-51	23.48 (10.91)	8-48	-1.18	.03	.24

3.2.Dot estimation task

Performance in the Dot estimation task was analyzed in a global repeated measures ANOVA with group (SB vs control group) as a between-subject factor and quantity (collections from one to seven) as a within-subject factor. This analysis showed significant main effects of quantity ($F(6,264) = 216.97$, $\eta^2 = .83$, $p<.001$) and of group ($F(1,44) = 8.67$, $\eta^2 = .16$, $p<.01$) as well as a significant interaction between group and quantity ($F(6,564) = 4.08$, $\eta^2 = .08$, $p<.001$), see Figure 2. A Newman-Keuls post-hoc test on quantity, corrected for multiple comparisons, indicated that there was no difference between the small numerical magnitudes from 1 to 3 (all ps>.49) while there were significant differences between the large numerical magnitudes (from 4 to 7, all ps<.002), indicating, as expected, a non-linear curve with precision starting to decrease when 4 items or more had to be estimated. More interestingly, a Newman-Keuls post-hoc test on the interaction between group and quantity showed significant differences between both groups for quantities 4 and 5 ($ps < .004$). To have a clearer picture of the subitizing range difference in the two groups, the largest numerical magnitudes for which at least 5 out of the 6 trials led to correct responses was determined for each individual and considered as the subitizing range. The subitizing range of the SB group (3.83±0.83) was significantly smaller that of the control group (4.52±0.90) ($t(44) = -2.72$, $\eta^2 = .14$, $p < .01$).

In order to assess the role of visuospatial capacity in subitizing ability, the same ANOVA was conducted, controlling for visuospatial short-term abilities which are assumed to be crucial to subitize (Piazza et al., 2011). Obviously, the main effect of quantity was still significant ($F(6,258) = 5.77$, $\eta^2 = .12$, $p < .001$) but the group effect ($F(1,43) = 2.84$, $\eta^2 = .06$, $p = .09$) and the group by quantity interaction ($F(6,258) = 1.69$, $\eta^2 = .04$, $p = .12$) were no longer significant. Moreover, to test whether visuospatial short-term abilities explained a *specific* part of variance of the group effect and the interaction, the same ANOVA was conducted controlling for verbal short-term abilities. This time, the group effect ($F(1,43) = 6.14$, $\eta^2 = .13$, $p < .05$) and the group by quantity interaction ($F(6,258) = 3.33$, $\eta^2 = .07$, $p < .01$) were still significant.

Figure 2. Performance in the Dot estimation task for all numerosities in both SB and control groups.



3.3.Non-symbolic magnitude representation tasks

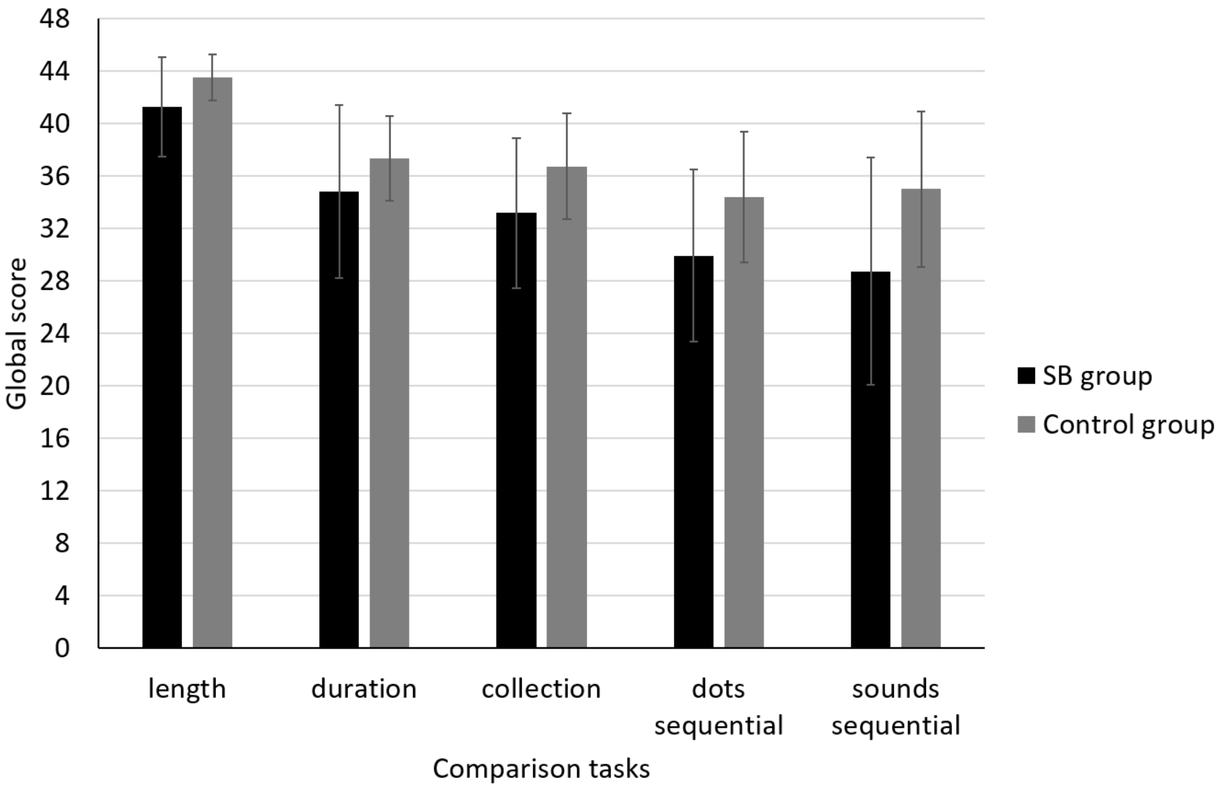
The precision of non-symbolic magnitude representations was assessed by using the global accuracy score. This measure was preferred since this index is considered as more reliable (see Inglis & Gilmore, 2014) and this should be even more true in children with developmental disorders in which the variability is greater than in healthy children.

Two ANOVAs were then conducted separately for non-numerical and numerical magnitude processing. A first 2x2 repeated measures ANOVA (contrasting the length and duration tasks in the SB and control groups) showed a main effect of task ($F(1,44) = 95.63$, $\eta^2 = .68$, $p<.001$) and a main effect of group ($F(1,44) = 4.97$, $\eta^2 = .10$, $p<.05$)[1]. More importantly, the interaction between group and task was not significant ($F(1,44) = .06$, $\eta^2 =$

[1] A post-hoc power analyses to assess the power of our main effect of group in this ANOVA showed a power (1-b err prob) of .72 with an effect size of .33 and a total sample size of 46 participants. With an effect size of this range (0.33), we would need a sample size of 56 participants to reach a power of 80%.

.00, p=.82). A second 3 x 2 repeated measures ANOVA (contrasting collection vs dot sequence vs sound sequence tasks in SB vs control groups) showed a main effect of task ($F(2,88) = 4.42$, $\eta^2 = .09$, $p<.005$), a main effect of group ($F(1,44) = 14.48$, $\eta^2 = .25$, $p<.001$)[2] and no significant interaction ($F(2,88) = .75$, $\eta^2 = .02$, $p=.49$). These results thus show poorer performance in the SB group irrespective of the task and of the fact that the magnitude processed was related to number, space or time.

Figure 3. Global scores in all comparison tasks for both SB and control groups.



[2] A post-hoc power analyses showed a power (1-b err prob) of 0.99 with an effect size f of 0.69 and a total sample size of 46 participants for this second ANOVA.

In order to see whether the differences between groups were more or less important depending on the task, while taking into account the different levels of difficulty between the tasks, z-scores were computed for each individual separately for each task, using the mean and the standard deviation of both groups considered together. Two ANOVA were then conducted separately for non-numerical and numerical magnitude processing. A first 2x2 repeated measures ANOVA (contrasting task length and duration of task in SB and control group) showed a null effect of task (due to the centering of the data) and showed a main effect of group, confirming our first result ($F(1,44) = (5.70$, $\eta^2 = .11$, $p<.05$). More importantly, the interaction between group and task was not significant, indicating a global non-numerical magnitude deficit in the SB group irrespective of the task ($F(1,44) = 0.73$, $\eta^2 = .02$, $p=.40$).

A second 3 x 2 repeated measures ANOVA (contrasting collection vs dot sequence vs sound sequence tasks in SB vs control groups) showed a main effect of group ($F(1,44) = 14.39$, $\eta^2 = .25$, $p<.001$) and no significant interaction ($F(2,88) = .05$, $\eta^2 = .00$, $p=.96$). This confirms the global numerical deficit for non-symbolic magnitude in the SB group irrespective of the task.

Finally, in order to directly test our main hypothesis of the crucial role of WM and visuospatial abilities in some numerical and non-numerical magnitude processing, we conducted a series of ANCOVAs on the previous z-scores by controlling for our variables of interest. With regard to the influence of visuospatial abilities, we conducted a first repeated measures ANCOVA considering two magnitude comparison tasks which are highly influenced by visuospatial skills, namely the length and the collection magnitude comparison tasks. This analysis allowed us to see if the main group effect disappeared after controlling for visuospatial abilities (with the Block design subtest score). This analysis showed a disappearance of the main group effect which became marginal ($F(1,43) = 3.05$, $\eta^2 = .07$, $p=.09$) and still no interaction effect ($F(1,43) = .02$, $\eta^2 = .00$, $p=.88$), suggesting that the difference between both groups in both tasks was partially due to the influence of visuospatial abilities on these tasks.

To establish whether visuospatial abilities explained a specific part of the variance of the group effect, we conducted the same repeated measures ANCOVA by controlling for another unrelated IQ task, vocabulary. We observed that this ANCOVA showed still a group effect ($F(1,43) = 9.53$, $\eta^2 = .18$, $p<.01$), meaning that visuospatial abilities explained a significant part of the performance in length and collection magnitude comparison tasks.

With regard to the influence of WM abilities, we conducted a first ANCOVA considering three magnitude comparison tasks which are assumed to be highly influenced by WM, namely the duration task and the two sequential magnitude comparison tasks (dot and sounds). This analysis allowed us to see if the main group effect disappears after controlling for WM abilities (with the verbal WM task considered to be the best reflection of working processes and probably the most solicited in the sequential tasks). This analysis showed a disappearance of the main group effect ($F(1,43) = 1.80$, $\eta^2 = .04$, $p=.19$) and no interaction effect ($F(1,43) = .91$, $\eta^2 = .02$, $p=.41$), suggesting that the difference between both groups in both tasks was partially due to the influence of WM abilities on these tasks.

To test whether WM abilities explained a specific part of variance of the group effect, we conducted the same ANOVA by controlling for another verbal task which was not presumed to have an impact on performance, the vocabulary measure. We observed that this ANCOVA showed still a group effect ($F(1,43) = 7.39$, $\eta^2 = .15$, $p<.01$), meaning that visuospatial abilities explained a significant part of the performance in length and collection magnitude comparison tasks.

3.4. Symbolic magnitude representation tasks

Two-sample t-tests were performed on the median correct response times (RT) as a function of accuracy in each symbolic magnitude comparison tasks (i.e. accuracy / median

RTs). There was no difference between groups in either the Arabic numerals comparison ($t(44) = -.53$, $\eta^2 = .00$, $p = .60$) or the Oral numerals comparison tasks ($t(44) = -.85$, $\eta^2 = .02$, $p = .40$), indicating similar symbolic magnitude processing in the SB group and the control group (see Table 2).

Table 2. Descriptive statistics of both groups in symbolic magnitude comparison tasks.

	SB group		**Control group**	
	Mean	**SD**	**Mean**	**SD**
Arabic numerals (ACC)	95.56	5.23	97.37	4.36
Arabic numerals (RT)	1497.10	370.71	1430.71	307.89
Arabic numerals (ACC/RT)	.07	.02	.07	.02
Oral numerals (ACC)	93.57	8.61	98.37	2.26
Oral numerals (RT)	1656.76	904.49	1439.49	466.38
Oral numerals (ACC/RT)	.07	.03	.07	.03

ACC= Accuracy; RT= Response Time

4. Discussion

The aim of this study was to gain a more in-depth understanding of the origin of mathematical learning disabilities in children with SB. Therefore, a systematic examination of basic number processing, including magnitude representation and subitizing abilities, was carried out in children with SB. The specific influence of higher cognitive factors, namely, visuospatial skills and WM abilities on these basic number magnitude processing skills was assessed using tasks involving different WM load and visuospatial processing requirements.

Relative to a control group matched on chronological age, children and adolescents with SB demonstrated a deficit in all continuous and discrete non-symbolic numerical magnitude comparison tasks and this, regardless of the visuospatial or WM demands of the different tasks. Moreover, they showed a reduced subitizing range (range of 3 rather than 4 for the control group) but also difficulties in estimating large numerical magnitude (from 5 to 7). Importantly, we demonstrated that the deficit in magnitude processing could be partially accounted for by the participants' visuospatial skills and WM abilities. Indeed, the deficit in magnitude comparison tasks with a high visuospatial requirement tended to disappear when we controlled for visuospatial abilities, but not when we controlled for verbal abilities. Moreover, the deficit in magnitude comparison tasks with a high WM demand disappeared when we controlled for verbal WM but not when we controlled for verbal abilities. However, they presented similar performance in symbolic number comparison tasks. Finally, children with SB presented the usual profile of a discrepancy between calculations retrieved directly from long-term memory and procedural calculation.

First, this study replicates previous studies on people with SB by showing a deficit in calculation abilities, particularly for subtractions, that is, calculations requiring procedural strategies such as counting and/or decomposition processing (Ayr et al., 2005; Barnes et al., 2002, 2005, 2006). This pattern is therefore in line with previously observed profile of children with SB showing preserved calculations when using long-term memory retrieval (i.e., mainly small addition and multiplication facts) but difficulties when procedural calculations are required (i.e., more complex additions and subtraction problems). It is important to note that in this study and also in previous studies, we used a written arithmetical timed test. It is possible that the slowness observed to solve calculations could to some extent be due to other general difficulties reported in children with SB such as their poor eye hand coordination (Lomax-Bream et al., 2007). However, even if this could overstate the mathematical impairment, this cannot explain the dissociation between procedural and long-term memory retrieval strategies observed. Interestingly, this study proposed that children with SB also have some difficulties in processing magnitude information at a very basic level. Indeed, they experienced some difficulties in comparing non-numerical magnitudes, visually and auditorily, but also in comparing non-symbolic numerical magnitudes such as the numerical magnitude of simultaneous or sequential sets/events. Moreover, we also demonstrated that children with SB presented difficulties in estimating large numerical magnitude (higher than 4), indicating that the difficulties were not restricted to the design of the tasks that is, the comparison process. While deficits in basic mathematical abilities has already been demonstrated, e.g. in counting, in the knowledge of counting principles and in object-based arithmetic in preschoolers children with SB (Barnes et al., 2011; English et al., 2009), no study until now has clearly explored the representation of numerical magnitude *per se*.

One previous longitudinal study following children with SB from 3 to 5 years old (Barnes et al., 2011) showed difficulties for all early mathematical tasks except in the small set object-based arithmetic task which entails processing quantities in the subitizing range. Even if our results showed a deficit in the extraction of numerical collections in the subitizing range, the children with SB adequately extract numerical magnitude up to the quantity 3, corresponding to the range used in the Barnes et al.'s study. Therefore, our data brings new and more precise information about the subitizing process in children with SB by demonstrating that the use of subitizing is possible, although limited to a smaller range. Our analyses thus clearly established a significant deficit in subitizing in the SB group. This deficit may be explained by their weakness in visuospatial WM abilities and not due to more general differences in WM between both groups. However, this assumption needs to be considered cautiously since to see in what extent the subitizing deficit was explained by the visuospatial short-term abilities, we used a visuospatial WM measure that also involves fine motor skills (to grasp the tokens an place them in the correct positions), abilities which are generally impaired in children with SB (Lomax-Bream et al., 2007). Therefore, even if a previous paper had already demonstrate the importance of the visuospatial WM performance in the subitizing abilities (Piazza et al., 2011), fine motor skills could also account for some part of the variability.

At first glance, our data are consistent with the ANS hypothesis which posits the existence of a specific amodal system dedicated to number magnitude processing. Indeed, we observed a global deficit in discriminating numerical magnitude regardless of the input modality (auditory or visual). In line with the present results, Barth et al. (2005) attempted to demonstrate that preschool children, like adults, possess approximate number representations that are independent of the modality or format of the stimuli to be processed, by showing similar performance for both kind of tasks, a comparison of two arrays of dots and a comparison between dot arrays and tone sequences. Similarly, Anobile et al. (2018) found significant correlations between three estimation tasks where children had to verbally produce the estimated number of dots sets, sequences of flashed dots or sequences of tones. Our results might suggest that mathematical difficulties come from a defect in this basic amodal magnitude representation. However, conflicting with this view, the groups did not differ on symbolic magnitude representation. In the ANS theory, the meaning of symbolic numbers is built through the connections established with non-symbolic number magnitude representation. Numerous studies have proposed that the deficit observed in mathematical learning disability could be explained by an access deficit from symbolic numbers to their corresponding magnitude representation (see Noël & Rousselle, 2011). This is clearly not the case for the sample of children with SB considered here, as their symbolic numerical magnitude processing showed no sign of impairment, despite their global non-symbolic magnitude deficit. Moreover, Gilmore and colleagues demonstrated that even if in adults, different measures of ANS did not correlate together, in children, performance across symbolic and nonsymbolic comparison or approximate addition, was related (Gilmore et al., 2014, 2011), suggesting that SB children did not demonstrate a more general weakness to process magnitude.

Instead, the main cause of magnitude processing deficit in children with SB seems to come from deficits in higher cognitive skills, such as WM and visuopatial abilities. Indeed, when we controlled for working memory abilities, we observed that the difference between the children with SB and controls became non-significant for tasks with an important WM demand. These data provide evidence for the dominant role of WM, as initially proposed in the accumulator model (see Meck & Church, 1983 as a seminal study proposing this model, see also Attout, Noël, Nassogne, et al., 2017), in accumulating magnitude and in comparing the accumulated result to numerical knowledge in long-term memory, at least when stimuli are presented sequentially.

On the other hand, another important factor in processing numerical magnitude, as underlined in the Introduction section, is visuospatial skills. Our results are in accordance with several studies showing that the processing of perceptual dimensions is clearly engaged in the construction of number magnitude representations. Indeed, when the individual differences in terms of visuospatial skills were controlled for, we no longer observed a main effect of group for magnitude comparison tasks with a high visuospatial requirement. In the same way, the difference between groups, as well as the group by numerical magnitude interaction in the Dot estimation task disappeared when we took participants' visuospatial WM abilities into account. This leads us to speculate that the group differences in the estimation and subitizing range were partially accounted for by visuospatial skills. These results add to the body of evidence supporting the influence of perceptual variables on number magnitude processing in children, as well as in adults who continue to be sensitive to perceptual dimensions in their numerical judgments (Gebuis & Reynvoet, 2012b; Szűcs et al., 2013). Even if the majority of studies currently try to control for the influence of perceptual variables when assessing non-symbolic magnitude processing (see for example Halberda & Feigenson, 2008), it is illusory to think that number magnitude acuity could be measured in a “pure” way, i.e., independently from non-numerical perceptual dimensions.

We therefore argue that the pattern of performance of children with SB in basic number processing tasks is related to a double deficit of higher cognitive factors. First, they present a visuospatial deficit which influences their numerical magnitude abilities in the same way as observed for people with 22q11.2 deletion syndrome or Williams syndrome (Attout, Noël, Vossius, et al., 2017; Rousselle et al., 2013). As in these syndromes, poorer processing of visuospatial cues may accumulate, leading to poorer precision of the resulting number magnitude representation. Likewise, other individuals with low visuospatial skills but with no associated genetic disorders, such as children with non-verbal learning disabilities, were found to show atypical number magnitude representation (Bachot et al., 2005; Crollen & Noël, 2015). Second, children with SB present a WM deficit which impacts the precision of the numerical magnitude representation of sequences of events such as in adults with Turner syndrome (Attout, Noël, Nassogne, et al., 2017). However, it is important to note that the profile of the two populations is not exactly the same since in adults with Turner syndrome, the deficit relates more to the passive storage of verbal information, while children with SB present a deficit in storage and manipulation of verbal information at short-term. This study confirms and strengthens findings reported in genetic syndromes with poor visuospatial and poor WM abilities. In the present case, the two deficits appeared to be additive, resulting in a global magnitude processing impairment.

At a more practical level, this study gives also new insights for a more adapted rehabilitation, by stimulating numerical cognition already at a very basic level and a more adapted teaching, by adopting alternatives strategies decreasing the intervention of WM and visuospatial abilities in the learning of mathematical notions. For example, in basic numerical processing learning, a multimodal approach (not exclusively visual, as usually provided) but with also other modalities (e.g., tactile approach, as when using tokens or abacus for example) could be a good way to reduce respectively the visuospatial and WM abilities requirement to construct a solid basic numerical processing. With regard to the use of a multimodal approach in very basic numerical processing, a recent study gives promising results by showing a larger increase of arithmetic performance after a multisensory training (visual and haptic modalities) than a visual training (corresponding to the same training but presented exclusively visually) or a control group (stories comprehension) (Crollen et al., 2020). At a higher level, for calculation, strategies putting emphasis on the arithmetical facts, without counting (as the compensation strategy for example, by rounding 89 to 90 and writing the borrowing), could be taught explicitly as a systematic way to solve numerous complex operations. Moreover, a feed-back could be given by a simple and systematic estimation of the solution, capacities usually preserved in these children. Future studies should explore concrete programs and practices based on these suggestions that may be effective for children with SBM and this very early, as preventive programmes given the very basic and early numerical processes involved.

5. Conclusions

Our data in children with SB indicated for the first time that they present a deficit in basic numerical processing which impairs non-symbolic magnitude representation and their ability to subitize small sets of elements. The pattern of impairment may not be the result of a defective magnitude representation, but rather of cumulative deficits impairing visuospatial processing and WM abilities. This defect may also impact the processing of non-numerical dimensions such as length and duration. The present study thus points towards multiple basic weaknesses to account for the emergence of math learning disabilities in children with SB, combining poor visuospatial and WM abilities with a lower precision in number magnitude processing and reduced subitizing abilities. Even if this study clearly states that basic numerical processing deficits could be due to primary visuospatial processing and WM abilities impairments, further studies need to investigate longitudinally the development of these basic numerical processes in younger children to see the specific involvement of these higher cognitive functions in the different basic numerical process and at which developmental stage.

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