The effect of visual arrangement on visuospatial short term memory-insights from children with 22q11.2 deletion syndrome

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Recent models of visuospatial (VSSP) short-term memory postulate the existence of two dissociable mechanisms depending on whether VSSP information is presented simultaneously or sequentially. However, they do not specify to what extent VSSP short-term memory is under the influence of general VSSP processing. This issue was examined in people with 22q11.2 deletion syndrome, a genetic condition involving a VSSP deficit. The configuration of VSSP information was manipulated (structured vs. unstructured) to explore the impact of arrangement on VSSP short-term memory. Two presentation modes were used to see whether the VSSP arrangement has the same impact on simultaneous and sequential short-term memory. Compared to children matched on chronological age, children with 22q11.2 deletion syndrome showed impaired performance only for structured arrangement, regardless of the presentation mode, suggesting an influence of VSSP processing on VSSP short-term memory abilities. A revised cognitive architecture for a model of VSSP short-term memory is proposed.

Keywords: Short-term memory; visuospatial processing; 22q11.2 deletion syndrome

Introduction

The visuospatial (VSSP) component of short-term memory, which involves the temporary retention (without manipulation) of VSSP information has been much less investigated than verbal short-term memory. According to Logie's (1995) model, the VSSP sketchpad consists of a visual store, named the visual cache, and a rehearsal mechanism, known as the inner scribe. About fifteen years ago, more systematic explorations led to further specifications of the architecture of VSSP short-term memory components in the *Continuity model* (Cornoldi & Vecchi, 2003), implementing the dissociation between visual and spatial short-term memory components (Mammarella, Pazzaglia, & Cornoldi, 2008) and within the latter, a distinction between simultaneous and sequential processes (Lecerf & De Ribaupierre, 2005; Mammarella, Borella, Pastore, & Pazzaglia, 2013).

Consistent with this model, double dissociations suggesting that simultaneous and sequential VSSP short-term memory components rely on different processing mechanisms have been reported in patients with visual neglect (Wansard et al., 2015) and in children with nonverbal learning disabilities (Mammarella et al., 2006). However, the nature of the mechanisms that contribute to these processes in VSSP short-term memory remains under-specified in the model. We could naturally postulate that sequential VSSP short-term memory tasks somehow require processing of the serial order of the items as the presentation order has to be kept in memory. Moreover, a series of studies suggest consistent and probably, bidirectional interactions between VSSP short-term memory processing and VSSP abilities. Indeed, difficulties with VSSP short-term memory processing has been noted in a number of developmental and acquired disorders, such as a representational neglect (Wansard, Meulemans, & Geurten, 2016). In some contexts such as in Williams syndrome, 22q11.2 deletion syndrome and in individuals with nonverbal learning disabilities, these VSSP short-term memory disabilities are associated with a cognitive profile characterized by weaker performance in VSSP tasks than in verbal task (De Smedt et al., 2007; Mammarella &

Cornoldi, 2005; Wang, Woodin, Kreps-Falk, & Moss, 2000). Moreover, in both typically developing children and adults, the VSSP span in short-term memory was larger when the patterns to be memorized followed a regular, structured configuration (i.e. a configuration respecting the gestalt principles of symmetry, repetition and continuation) than when the VSSP arrangement was unstructured, regardless of the presentation mode (i.e. simultaneous or sequential) (Imbo, Szmalec, & Vandierendonck, 2009; Kemps, 2001; Pieroni, Rossi-Arnaud, & Baddeley, 2011; Rossi-Arnaud, Pieroni, Spataro, & Baddeley, 2012). According to these studies, the arrangement effect may reveal the role played by long-term VSSP knowledge of structure on the retention of VSSP information in short-term memory, allowing chunking of information into familiar or regular structures.

One question that remains open concerns the specific relation between VSSP abilities and each of the spatial short-term memory processing components. For the sake of clarity, the term *VSSP abilities* will be used here to designate the ability to form a VSSP representation of the spatial coordinate of static visual positions, that is, the processing of extrinsic and static VSSP information in keeping with the typology proposed by Uttal et al. (2013)[1]. With regard to the relationship between VSSP short-term memory and VSSP abilities, it has not been demonstrated whether sequential (one item at a time) and simultaneous (items all at once) spatial short-term memory components are equally influenced by VSSP abilities. In sequential mode, the VSSP representation has to be built from the integration of a sequence of individual *local* positions disclosed one-by-one which gives less direct access to the global VSSP

[1] In a recent discussion on the typology of spatial skills, Uttal (2013) proposed that spatial skills may be characterized along two fundamental dimensions. The first concerns the nature of the to be processed spatial information and distinguishes between intrinsic (i.e. spatial relations between the parts of an object) and extrinsic spatial information (i.e. spatial relation between the elements in a set). The second dimension bears on the task at hand and differentiates between static and dynamic information depending on whether VSSP relationships change or not during the task.

representation. By contrast, in simultaneous presentation mode, the VSSP information is presented all at once, and is thus more likely to directly elicit a *global*, configural VSSP representation. Accordingly, it may be predicted that there will be a larger effect of item arrangement on performance for simultaneous presentation compared to sequential presentation.

In line with this prediction, recent data suggested that VSSP skills have asymmetrical influence on the two VSSP short-term memory components, the effect of VSSP arrangement being more pronounced in the simultaneous than in sequential presentation mode (Pieroni et al., 2011). In developmental disorders in which individuals exhibit VSSP deficits, such as Williams syndrome, effects of arrangement on short-term memory performance have been reported with the simultaneous but not with the sequential mode of presentation (Carretti, Lanfranchi, De Mori, Mammarella, & Vianello, 2015). Moreover, people with Down syndrome with global intellectual disabilities, took less advantage of the structured configuration in simultaneous VSSP short-term memory task compared to typically developing children matched on general IQ but the two groups did not differ in the sequential VSSP short-term memory task (Carretti & Lanfranchi, 2010; Carretti, Lanfranchi, & Mammarella, 2013).

The aim of this study is to examine whether VSSP abilities contribute to performance in VSSP short-term memory and whether this influence varies with the mode of presentation, sequential or simultaneous. To this end, we tested the impact of the arrangement of the to-be-recalled VSSP configurations on both sequential and simultaneous VSSP short-term memory performance in a genetic population with lower VSSP IQ, the 22q11.2 deletion syndrome (also named velo-cardio-facial syndrome). This syndrome occurs mainly de novo with a prevalence from 1:2000 to 1:6000 (Gothelf, Frisch, Michaelovsky, Weizman, & Shprintzen, 2009). VSSP impairments have been reported in this syndrome including deficits in visualperceptual and visuomotor integration skills (Niklasson & Gillberg, 2010; Van Aken, Caeyenberghs, Smits-Engelsman, & Swillen, 2009). Adults with 22q11.2 deletion syndrome experience difficulties in perceiving VSSP information, identifying and discriminating silhouettes (Henry et al., 2002), copying a complex figure (Rey complex figure; Antshel et al., 2008), tracking objects with attention and using a scanning strategy in face recognition (Cabaral, Beaton, Stoddard, & Simon, 2012; Glaser et al., 2010; McCabe, Rich, Loughland, Schall, & Campbell, 2011). These poor VSSP skills are associated with weak VSSP short-term memory abilities for both simultaneous and sequential presentation modes (Vicari et al., 2012; Wang et al., 2000; Wong, Riggins, Harvey, Cabaral, & Simon, 2014).

In order to investigate the impact of VSSP arrangement and of the presentation mode on VSSP short-term memory, participants with 22q11.2 deletion syndrome and typically developing children, were administered VSSP short-term memory tasks in which they had to recall the position of items presented simultaneously or sequentially in a grid. The arrangement of the to-be-recalled configuration was manipulated to form structured vs. unstructured patterns. Due to their weaker VSSP abilities, patients with 22q11.2 deletion syndrome are expected to present shorter VSSP short-term memory spans and to be less sensitive to the arrangement of the to-be-recalled configurations. Moreover, if a global VSSP representations is more readily accessible from the simultaneous presentation mode, the arrangement effect is expected to be larger for simultaneous than for sequential presentation mode, both in typically developing children and in patients with 22q11 deletion syndrome.

Methods

Participants

Twenty-seven children and teenagers with 22q11.2 deletion syndrome aged between 5 and 16 years old (mean = 10 years old, 11 females) participated in this study. They were recruited through the 22q11.2 deletion syndrome association and the department of paediatric cardiology of Saint-Luc University Hospital in Belgium. Diagnosis was confirmed with two-colour fluorescent in situ hybridization (FISH). Each participant with 22q11.2 deletion syndrome was paired with a participant from a group matched on chronological age (± 5 months around the patient's age) composed of 27 typically developing children (Mean = 10 years old, 14 girls).

Materials

Verbal and non verbal intelligence was briefly assessed as well as the three main components of WM defined in Baddeley and Hitch's model (Baddeley, 1986; Baddeley & Hitch, 1974), namely the central executive component and the phonological loop as descriptive measures, and the VSSP sketchpad as the measure of interest.

Intelligence

Vocabulary and Similarity subtests from the Wechsler intelligence scales for children (*Wechsler Preschool and Primary Scale of Intelligence*-3rd edition (WIPPSI-III) or the Wechsler Intelligence Scale for Children-4th edition (WISC-IV) depending on the child's age) were used to estimate verbal IQ. The Block Design subtest was used to measure visuo-constructive abilities known to be weaker in people with 22q11 deletion syndrome. Standard scores were used.

Verbal short-term memory

Phonological loop capacity was assessed in a forward letter span task. Participants were instructed to listen to a sequence of letters and to repeat them immediately afterwards in the same order. Letters were read at the rate of one per second. Sequences consisted of monosyllabic consonants with no repetition within any sequence. The first sequences included two letters and were then followed by sequences of increasing length (3 to 9 letters).

Verbal working memory

The category-span task developed by Noël (2009) was used to examine the central executive component. In this task, one-syllable words, related to either food or animal, were read at the rate of one word per second to the participants and they had to recall them category by category (starting with the food words and then the animal words). To help participants understand the instructions, pictures of a forest and a plate with cutlery were presented to support the recall of the animal and food names, respectively. Sequences started with 2 words, reaching a maximum of 9 words.

VSSP short-term memory

The VSSP short-term memory was assessed with two tasks inspired by Imbo et al. (2009): the simultaneous VSSP short-term memory task in which the elements were presented all at once, and the sequential VSSP short-term memory task in which the to-be-remembered positions were presented sequentially one at a time. In the simultaneous condition, participants were presented with a matrix of two to ten dots for 5 seconds. In the sequential condition, participants were presented with a blank matrix and were instructed to remember the positions of a series of cells indicated one by one (one per second), by the examiner. In both tasks, participants were instructed to recall the positions by placing tokens in the right location on a blank matrix. The order of the responses was not taken into account in the sequential VSSP short-term memory task so that the output modes were the same across tasks. Thus, the two VSSP short-term memory tasks differed only on the input mode, which was either simultaneous or sequential. Two conditions were contrasted within each VSSP short-term memory task: for half of the trials, the layout of the to-be-remembered positions presented no regular pattern (*unstructured* condition) while for the other half, the position of the presented elements formed a regular pattern respecting either a principle of symmetry or continuity of the path (Gestalt principles) (*structured* condition). In both conditions, the trials progressively increased in complexity as the size of the matrix and the number of positions to be remembered increased during the task. The lower level of difficulty corresponded to a 2 × 2 matrix with 2 locations to be recalled and the higher level included a 4 × 5 matrix with 10 positions to be remembered.

Experimental procedure

For all short-term memory and WM tasks, participants had to succeed on two trials of the same difficulty to access a higher level (larger matrix and/or larger span). The task ended when the participant failed at two out of the three trials for a given difficulty level. Each correct response was credited with one point. Testing was completed in two sessions at school or at home in a quiet room and tasks were offered in a Latin square order to counterbalance the presentation order. The experiment was conducted in accordance with the Declaration of Helsinki and the regional ethical committee for biomedical research approved the experimental protocol (Record number: B403201111579). All legal guardians gave informed written consent and the participants themselves assented to participation before the cognitive tests were administered.

Results

Descriptive measures

Two-tailed t-tests were run to compare the group of individuals with 22q11.2 deletion syndrome with the group of typically developing children for descriptive measures. As expected, there were no age differences between the patients and the typically developing groups ($t(1,52) = -.03$, $\eta^2 = .00$, $p=.98$). T-tests showed that the 22q11.2 deletion syndrome group presented significantly poorer performance than the typically developing group for standard scores[2] on the Vocabulary ($t(1,52) = 6.14$, $\eta^2 = .42$, $p<.001$), the Similarity ($t(1,52) = 4.08$, $\eta^2 = .24$, $p<.001$) and the Block Design subtests ($t(1,52) = 6.11$, $\eta^2 = .41$, $p<.001$). For the two verbal memory measures, the patient group performed slightly lower than the control group but the difference did not reach statistical significance for the verbal working memory ($t(1,52) = 1.91$, $\eta^2 = .06$, $p=.06$) or for the verbal short-term memory tasks ($t(1,52) = 1.74$, $\eta^2 = .05$, $p=.09$) (see Table 1).

<INSERT TABLE 1 HERE>

Performance in VSSP short-term memory tasks was analyzed using a 2 x 2 x 2 repeated measures ANOVA with Presentation mode (sequential vs. simultaneous) and Arrangement (unstructured vs. structured) as within-subject factors and Group (22q11.2 deletion syndrome vs. typically developing) as a between-subjects factor. The main effect of Group was significant ($F(1,52)=7.11$, $\eta^2 = .12$, $p<.05$) and indicated that participants with 22q11.2 deletion syndrome group performed worse than the typically developing group (see Figure 1). The main effects of Presentation ($F(1,52)=6.92$, $\eta^2 = .12$, $p<.05$) and Arrangement ($F(1,52)=124.52$, $\eta^2 = .71$, $p<.001$) indicated more accurate performance for simultaneous than for sequential presentation, and for structured than for unstructured patterns (see Figure 1). There was no interaction between Presentation and Group ($F(1,52)=.02$, $\eta^2 = .00$, $p=.89$), indicating that both groups were equally influenced by the mode of presentation. However,

[2] Note that for four pairs of children, the IQ battery was not the same between the pair. This was because the 22q11.2 deletion syndrome children were in the age range (6-8) that straddled the two IQ tests, WPPSI and WISC. This led to limitations for those children in completing the second battery.

there was a significant interaction between Arrangement and Group, ($F(1,52)=5.69$, $\eta^2 =.10$, $p<.05$). A Newman-Keuls post-hoc test indicated significant differences between both groups only for structured condition ($p<.001$), with the typically developing group taking more advantage of the pattern structure even if both groups presented a significant effect of Arrangement (Planned comparison for the 22q11.2 deletion syndrome group: $F(1,52)=38.48$, $\eta^2 =.43$, $p<.001$; for the typically developing group: $F(1,52)=91.74$, $\eta^2 =.63$, $p<.001$) (see Figure 1). Interestingly, there was a significant interaction between Presentation and Arrangement ($F(1,52)=28.72$, $\eta^2 =.36$, $p<.001$), indicating a more pronounced effect of the structure in the simultaneous than in the sequential presentation mode, even if a significant effect of the structure was observed in both VSSP short-term memory tasks (Planned comparison for the simultaneous mode: $F(1,52)=122.16$, $\eta^2 =.70$, $p<.001$; for the sequential mode: $F(1,52)=70.89$, $\eta^2 =.58$, $p<.001$. The interaction of Group, Arrangement and Presentation was not significant ($F(1,52)=0.14$, $\eta^2 =.00$, $p=.71$).

<INSERT FIGURE 1 HERE>

In order to confirm the impact of VSSP abilities on performance observed in the VSSP short-term memory task, we conducted correlation analyses between scores in the Block Design subtest and the facilitation effect of arrangement. Block Design subtest provides a measure of visuo-constructive abilities which rely directly on VSSP abilities in the meaning considered here. Indeed, to be able to reproduce a visual configuration using blocks, participants have to process the respective spatial position of each constituent relative to each other. Arrangement facilitation effects were thus calculated for each presentation mode (simultaneous and sequential), by subtracting performance for structured and unstructured items for each subject. This question was addressed using the sample as a whole, including the typical and the 22q11.2 deletion syndrome groups, in order to maximize the variability on the variable of interest. These individual facilitation effects were found to be significantly correlated with performance in the Block Design subtest both for simultaneous ($r(54)=.27$, $p=.052$) and sequential presentation mode ($r(54)=.30$, $p<.05$), respectively. Importantly, the Vocabulary and Similarity subtests, used to estimate verbal IQ measure, did not correlate significantly with the facilitation effects whatever the presentation mode (all $rs<.23$, $p>.09$).

Discussion

The goal of this study was to examine whether VSSP abilities contribute to performance in VSSP short-term memory and whether this influence varies depending on item presentation mode. To this end, we tested the impact of the to-be-recalled VSSP arrangement in both sequential and simultaneous VSSP short-term memory in a sample of individuals with 22q11.2 deletion syndrome, a genetic condition associated with multiple VSSP processing deficits.

Our results showed significantly lower performance for the 22q11.2 deletion syndrome group in the VSSP short-term memory tasks in both simultaneous or sequential presentation mode, in comparison to children matched on chronological age. Moreover, compared to the typically developing group, the patients took less advantage of the pattern arrangement to strengthen their VSSP representation in short-term memory. Finally, we observed a significant relation between performance in the Block Design subtest and the arrangement facilitation effect across both groups.

First of all, these results support the hypothesis that weak VSSP abilities are associated with weak VSSP short-term memory performance and replicate previous studies showing weak VSSP short-term memory spans in children with 22q11.2 deletion syndrome with items presented simultaneously (Bearden et al., 2001; Campbell et al., 2010; Sobin et al., 2005; Wang et al., 2000) or sequentially (Vicari et al., 2012; Wong et al., 2014).

Second, children with 22q11.2 deletion syndrome were less sensitive to structured arrangement than typically developing children. Moreover, when considering both groups together, the arrangement effects correlated with performance in the Block Design subtest. Imbo et al. (2009) speculated that the growing effect of structure as a function of age (in a typically-developing population) could be explained by at least two mechanisms: (1) a growing ability to implement strategies, like chunking and (2) an increasing store of VSSP knowledge in long-term memory whether, associated to semantic content (i.e. cross, U, …) or not. In the present case, this lesser influence of VSSP arrangement on representation in VSSP short-term memory could be attributed to either limited pre-existing VSSP knowledge in long-term memory (such as "*this part forms a cross*" or "*this pattern is like the letter v*"), or difficulty processing extrinsic VSSP information as input in short-term memory, or to their weaker ability to use strategies to organize or chunk VSSP information in short-term memory itself (e.g., reorganization or grouping of information into familiar or regular structures). These three possibilities being not mutually exclusive.

Third, whatever the population, the arrangement effect was larger in the simultaneous than in the sequential VSSP tasks, as in previous studies (Carretti et al., 2015; Pieroni et al., 2011) which supports the view that extrinsic VSSP information may be more accessible with the simultaneous presentation than with the sequential presentation mode. In the framework of the Continuity Model (see Figure 2a), our results indicate that both spatial short-term memory components are sensitive to extrinsic VSSP information since an arrangement effect was observed in both conditions. However, this influence is larger in the simultaneous than in the sequential component in VSSP short-term memory. Accordingly, we propose that a part of the simultaneous-sequential dissociation could result from the way extrinsic VSSP information is presented and then processed. For both modes of presentation, access to a configural representation in short-term memory is facilitated when VSSP information is embedded in a regular structure (as noted by the * in Figure 2b). However, as depicted in Figure 2b, VSSP information presented simultaneously may provide more direct access to a configural, global representation in short-term memory that could be based upon long-term knowledge. By contrast, VSSP information presented sequentially may require intermediate processing coding for the local individual positions and for the order of each position in the sequence in order to construct this global VSSP representation in short-term memory, thus activating long-term knowledge less directly. Although speculative, this alternative two processing route model still accounts for the well-documented sequential-simultaneous dissociation reported in previous studies. In addition, it provides a more transparent picture of how a VSSP representation is elaborated in short-term memory depending on the input presentation mode and how order processing and long-term memory knowledge contribute to VSSP representation in short-term memory.

<INSERT FIGURE 2 HERE>

This model can thus lead to testable predictions about how local and global VSSP processing could affect performance in VSSP short-term memory tasks (D'Souza, Booth, Connolly, Happé, & Karmiloff-Smith, 2016). For example, we could expect that people with Williams syndrome, who exhibit a dominance of local VSSP processing, would find it easier to elaborate a VSSP representation in short-term memory from sequential than from simultaneous inputs. These predictions align with existing data for Williams syndrome (Carretti et al., 2015). On the other hand, patients with Down syndrome or with nonverbal learning disabilities (Cardillo, Mammarella, Garcia, & Cornoldi, 2017), who exhibit a preference of global VSSP processing, could be more impaired with the sequential mode of presentation, which require encoding VSSP position sequentially. With respect to these predictions, our data provide no indication about a local-global VSSP preference in patients with 22q11 deletion syndrome as the three way interaction was not significant. This suggests that the arrangement effect is larger in the simultaneous mode in both groups.

One limit of this study is that the examination did not provide an extensive assessment of the nature visuo-spatial deficit of our patients with 22q11 deletion syndrome. Their VSSP skills were assessed using the Block Design subtest which is a complex measure known to tap into numerous VSSP skills including not only the processing of static extrinsic VSSP information but also local-global processing, spatial reasoning in 2 and 3 dimensions and visuo-constructive abilities. This task is thus very sensitive to VSSP difficulties but lacks specificity to differentiate among the various VSSP processes that could be more impaired in this population. A more extensive exploration is needed in the future to specify the nature of VSSP abilities that are specifically recruited by VSSP short-term memory.

To conclude, the present study confirmed that the processing of extrinsic VSSP information influences performance in VSSP short-term memory. This was supported by the observation of a global VSSP short-term memory deficit in 22Q11.2 deletion syndrome related to their general VSSP deficit, and also by their weaker ability to take advantage of the structure of the VSSP arrangement. Finally, we observed an asymmetrical influence of the VSSP arrangement on VSSP short-term memory performance depending on the presentation mode, which has implications for the modelling of VSSP short-term memory.

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Table 1. Descriptive measures for 22q11.2 deletion syndrome and typically developing groups.

Figure 1. Effect of Task x Structure of accuracy data in each group.

Figure 2. Representation of the continuity model (a) and the proposed model (b) taking into account the influence of VSSP skills on the VSSP short-term memory. * = facilitation.

Table 1. Descriptive measures for 22q11.2 DS and TD groups.

	22q11.2 DS group Mean (SD)	TD group Mean (SD)
Age (months)	121.41 (37.01)	121.67 (37.43)
Vocabulary (standard score)	6.19 (2.75)	10.19 (1.98)
Similarities (standard score)	8.04 (2.77)	11.04 (2.64)
Block design (standard score)	5.48 (3.12)	10.07 (2.35)
Verbal WM	5.22 (2.36)	6.30 (1.73)
Verbal STM	6.22 (2.14)	7.22 (2.08)

Figure 1.

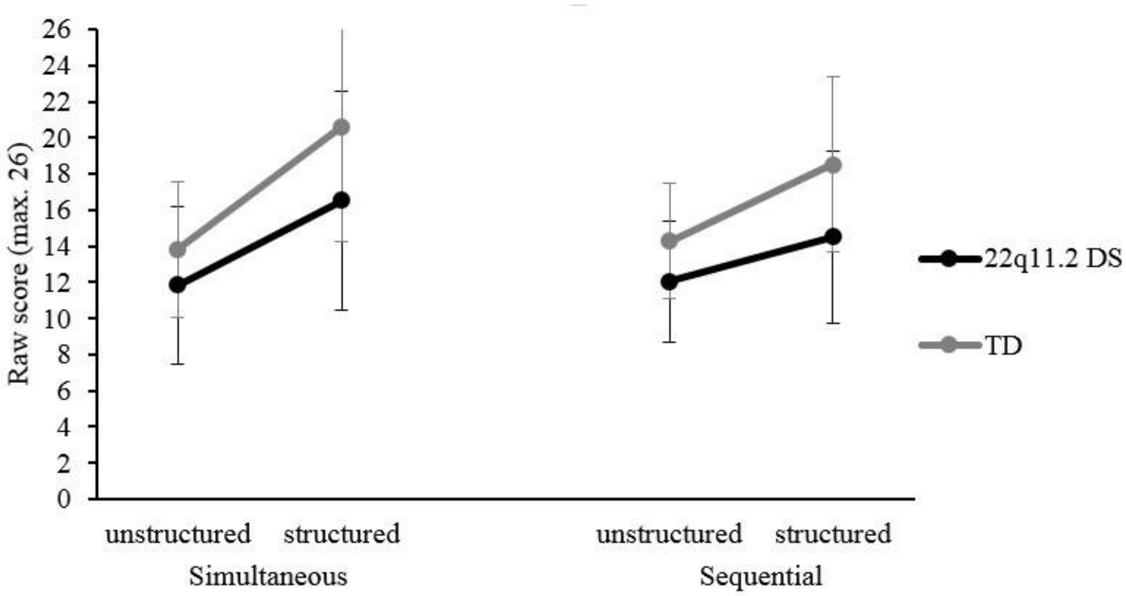
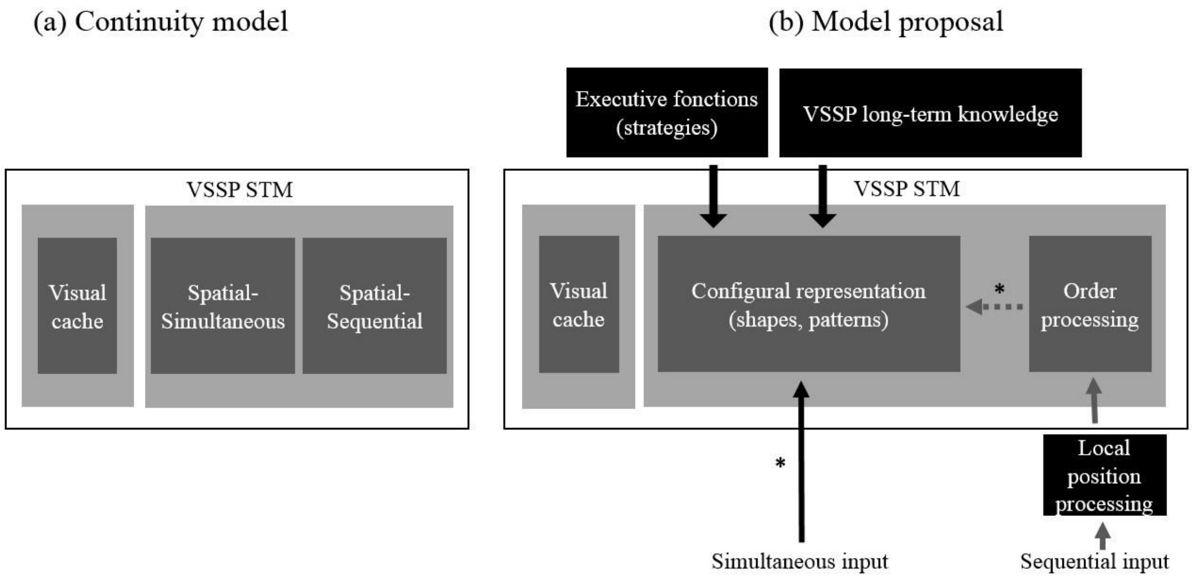

Raw score (max. 26)
26
24
22
20
18
16
14
12
10
8
6
4
2
0
unstructured
structured
Simultaneous
unstructured
structured
Sequential
22q11.2 DS
TD

Figure 2.


(a) Continuity model
VSSP STM
Visual cache
Spatial-Simultaneous
Spatial-Sequential
(b) Model proposal
Executive fonctions (strategies)
VSSP long-term knowledge
VSSP STM
Visual cache
Configural representation (shapes, patterns)
*
Order processing
*
Local position processing
Simultaneous input
Sequential input