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Explorative function in Williams syndrome analyzed through a large-scale task with multiple rewards

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ABSTRACT

This study aimed to evaluate spatial function in subjects with Williams syndrome (WS) by using a large-scale task with multiple rewards and comparing the spatial abilities of WS subjects with those of mental age-matched control children. In the present spatial task, WS participants had to explore an open space to search nine rewards placed in buckets arranged according to three spatial configurations: a cross, a 3×3 matrix and a cluster composed by three groups of three buckets each. The findings demonstrate that WS individuals were impaired in efficiently exploring the environment and in building cognitive spatial maps. In exploring the three spatial configurations, they performed worse than control subjects on all parameters analyzed. In fact, WS individuals took more time to complete the task, made more errors, performed a reduced number of error-free trials, displayed lower search efficiency, exhibited shorter spatial spans, showed a higher number of no-visits and displayed marked tendencies to perseverate and to neglect some buckets. Furthermore, WS individuals showed disorganized explorative patterns in comparison to control children. WS influenced performances differentially as a specific effect of the susceptibility of the configurations to being explored in a principled way. In the cross configuration that had strong spatial constraints, both groups exhibited their worst performances. In the matrix configuration, the altered explorative strategies of the WS subjects primarily affected their central exploration. The performances in the cluster configuration indicated that chunking was a strategy of strength in both TD and WS groups. In conclusion, WS individuals' deficits exhibited in the present explorative test may be considered an index of their difficulties in spatial orientation and motion perception displayed in the real world. The marked impairment in spatial information processing is discussed in neuro-anatomical alterations reported in WS.

1. Introduction

Williams syndrome (WS) is a rare genetic condition resulting from a microdeletion in chromosome 7q11.23 featuring a profile of mild to moderate retardation and selective impairment in spatial function. In fact, while certain aspects of

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language development are relatively proficient, visuo-spatial processing, planning and implicit learning abilities are severely impaired (Atkinson et al., 2001; Bellugi & St George, 2001; Klein & Mervis, 1999; Vicari, Bellucci, & Carlesimo, 2001; Vicari, Verucci, & Carlesimo, 2007). Yet, despite an overall impaired level of ability, WS individuals have a dissociation of visuo-spatial abilities. In fact, they exhibit specific difficulty in maintaining visuo-spatial information in working memory and in performing long-term memory tasks but exhibit a relatively preserved perception of the visual characteristics of objects (colors and shapes) (Atkinson et al., 2003; Vicari, Bellucci, & Carlesimo, 2005; Vicari, Bellucci, & Carlesimo, 2006). Furthermore, WS individuals have difficulties on other spatial tasks including remembering locations on a screen (Paul, Stiles, Passarotti, Bavar, & Bellugi, 2002), keeping track of moving objects (O'Hearn, Landau, & Hoffman, 2005) and judging spatial relations between stimuli presented simultaneously (Farran & Jarrold, 2005; Landau, Hoffman, & Kurz, 2006).

In addition to the different components of spatial function (short/long term spatial memory, declarative memory or procedural knowledge) tapped by the different spatial tasks, the characteristics of the environment in which the subjects move weigh on spatial performance. Namely, WS individuals have a reportedly poor ability to use intrinsic, object-based representations based on local landmarks to find a target within a spatial array (Nardini, Atkinson, Braddick, & Burgess, 2008). This deficit was more evident when the participants had to rely on the spatial configuration of the array than when they determined locations relative to the position of their body. Impairment in the use of different frames of reference are consistent with everyday navigation and way-finding difficulties reported in WS individuals and may also contribute to their problems with basic localization. There are few studies of navigation in WS subjects even though parents anecdotally report that WS individuals often meet with difficulties in moving into new environments, get lost or disorientated in unfamiliar places and have difficulties with wayfinding (Atkinson et al., 2001; Farran, Blades, Boucher, & Tranter, 2010). Furthermore, WS individuals have an impaired ability to search for objects in a room-sized space (Smith, Gilchrist, Hood, & Karmiloff-Smith, 2006) and weakened performance in "large-scale" route-learning (Farran et al., 2010) and spatial re-orientation tasks (Lakusta, Dessalegn, & Landau, 2006). Recently, to analyze spatial competencies in WS, we used a "large-scale" Radial Arm Maze (RAM) in a real environment that allowed the participants to be physically active within the maze (Mandolesi et al., 2009). By using the RAM, we distinguished between deficits in the acquisition of spatial procedural competences and deficits in spatial memory processes in WS individuals. Another large-scale study addressed the route-learning topic in WS individuals (Farran et al., 2010), demonstrating that they were able to learn a 1-km route through a natural environment and improve their route knowledge by verbal coding, but they had poor relational knowledge between landmarks. However, both researches did not allow an investigation of the exploratory capacities of WS individuals. In fact, in the RAM the searching strategies are forced by the preset number of alternative routes that thus constrain and limit searching behavior, while in the route-learning task an efficient performance is not linked to explorative but to mnesic capacities.

Thus, it is seemed interesting to investigate whether the repeatedly described spatial impairment exhibited by WS individuals affected their searching strategies and how their eventual explorative deficits were reflected in organizing principled search patterns and in remembering visited locations and representing the entire setting.

To this aim, we tested WS individuals in a spatial task in which the structural affordances of the search space were manipulated to make subjects' explorative strategies emerge. In fact, when multiple rewards are distributed in unconstrained environments, the exploratory behavior has to adapt to the environmental features so that spatial structure of the search space influences the economy and organization of searching behavior. Thus, structural affordances of the environment influence the construction of search strategies and the information about where the rewards are located. In the present spatial task with multiple rewards, WS participants had to explore an open space to search nine rewards placed in buckets arranged according to three spatial configurations. They had to keep track of the visits made over time in the absence of any constrained trajectory and of physical traces left from choices they had already made. Searching for multiple rewards arranged according to different spatial configurations involved the use of explorative strategies, visuo-spatial abilities, spatial memory and cognitive mapping. Efficient explorative strategies required the processing of extra-maze (allothetic), intra-maze and idiothetic stimuli (Nardini, Jones, Bedford, & Braddick, 2008). The processing of the extra-maze cues that were kept stable throughout the task required the encoding of the spatial relationships between landmarks. The processing of intra-maze cues required the encoding of the spatial relationships specified by the buckets themselves. The idiothetic cues were derived from the subject's own movements. These clues encompassed vestibular, proprioceptive, and somato-sensory information provided by the sequence of movements performed by the subject; efference copies of motor commands; and external motion-related information such as optic flow (Hartley, Maguire, Spiers, & Burgess, 2003; Mittelstaedt & Mittelstaedt, 2001). Visuo-spatial abilities were required to plan actions to move into the environment, to direct attention towards relevant environmental elements, to recognize objects, to memorize either the single details (for example, the buckets) or the entire environmental setting and, finally, to build the cognitive mapping of the environment.

The spatial configurations were a cross, a 3×3 matrix and a cluster composed of three groups of three buckets each. Before the spatial task, participants were given verbal information only about how to play the "game" they were going to experience. No verbal information was provided about how to search rewards or about the spatial setting. Because verbal cognition is retained as a relative strength in WS individuals, we used this procedure to prevent them from compensating for their performance in weaker areas of cognition.