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Fading-figure tracing in Williams syndrome

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ABSTRACT

Williams syndrome (WS) is a neurodevelopmental disorder characterized by severe impairment of visuospatial abilities. Figure-drawing abilities, which are thought to reflect visuospatial abilities, have yet to be fully investigated in WS. The purpose of the present study was to clarify whether drawing abilities differ between WS individuals and typically developing children (TD). We compared the performance of two groups of subjects (WS, mean age 16 years; TD, 5–6 years of age) using a fading-figure tracing task that requires subjects to trace a target figure that is gradually disappearing from a PC screen. Although the TD group exhibited clearly improved performance with long fading time, the WS group did not. Moreover, the TD group exhibited poor performance for figures with more than six angles, regardless of the figure type (e.g. closed or open), whereas the WS group exhibited generally poor performance for figures with more than five angles but relatively preserved performance for open figures. These findings indicate that a combination of decreased visuospatial span associated with incomplete development of visual scanning and disproportionate development of global processing may cause drawing disabilities in WS.

1. Introduction

Williams syndrome (WS) is a contiguous gene syndrome caused by a hemizygous deletion of approximately 28 genes on chromosome 7q11.23, characterized by an uneven cognitive profile, consisting of relative preservation of expressive language and facial recognition with severe impairment of visuospatial abilities (Martens, Wilson, & Reutens, 2008; Meyer-Lindenberg, Mervis, & Berman, 2006). Although this cognitive pattern was originally construed as resulting from neuropsychological double dissociation in adult brain-damaged patients (Bellugi, Sabo, & Vaid, 1988), recent studies have demonstrated that expressive language and facial recognition are not intact in patients with WS (Karmiloff-Smith, Brown, Grice, & Paterson, 2003; Mervis, 2003).

Currently, whether the characteristic cognitive pattern of WS is a consequence of delayed (or arrested) typical development or the result of atypical development is unclear.

Figure-drawing abilities have from the beginning of cognitive research on WS been investigated as reflecting visuospatial disabilities. Bellugi et al. (1988) showed that individuals with WS were able to describe the visual features of an object (e.g. bicycle) orally but unable to draw a picture of the same object. Moreover, using hierarchical letter stimuli, Bihrle, Bellugi, Delis, and Marks (1989) showed that individuals with WS were able to copy a small letter but unable to copy a large letter consisting of small letters, and concluded that the drawing disabilities in this syndrome result from global processing impairment or local processing bias. However, the results of subsequent studies involving perceptual grouping have not supported this hypothesis (Farran, 2005; Farran & Cole, 2008; Farran, Jarrold, & Gathercole, 2003; Farran & Wilmut, 2007).

Few subsequent studies, however, have investigated the figure-drawing abilities of individuals with WS, probably because figure drawing tasks (particularly those involving drawing from memory) may include numerous cognitive processes, and because it is difficult to evaluate how and whether a drawing is "good". Most previous studies have investigated abilities to copy a geometric figure, to draw from memory (e.g. flower, human, or animal), or both. Figure copying is, along with block design, a constructive task, and can be evaluated by matching the drawn (or constructed) figure to the model (target) figure, permitting clearer evaluation of such abilities than drawing from memory. Most previous studies used the Developmental Test of Visual Motor Integration (VMI) (Beery, 1989), which includes various model figures from a simple line to complex figures and is based on standard ages at which acquisition of the ability to draw each model figure is expected. Bellugi et al. (1988) showed that the drawing abilities of adolescents and young adults with WS were at the level of a 5-year-old child; they were able

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to draw shapes or lines involving single integration and coordination. Similar results have been reported in children aged 9–10 years who had WS (Bertrand, Mervis, & Eizenberg, 1997). The individuals showed good performance with simple lines/shapes, but performance was poor when presented with intersecting lines/shapes and was similar to that seen in normally developing children aged 4–7 years. The drawing errors made by the individuals with WS were also those typically observed in normally developing preschoolers 4–7 years of age. The authors concluded that performance by WS individuals was therefore the result of delayed, rather than atypical, development. Stiles, Sabbadini, Capirci, and Volterra (2000) examined a girl with WS longitudinally from 4 to 6 years of age, and obtained findings suggesting that her drawing abilities were consistently delayed compared with her vocabulary and other non-verbal abilities. Georgopoulos, Georgopoulos, Kuz, and Landau (2004) compared the copying abilities of 6–14-year-old children with WS and mental-age-matched normal children (3–6 years old), using six model figures (line drawings of closed and open geometrical shapes (alone and in combination), crossed lines, and geometrical shapes consisting of distinct small, filled circles) from the VMI. To evaluate performance quantitatively, the authors used psychological assessment of "goodness of copy" (with respect to the template) by human adult raters, who also assessed the similarity of corresponding copies made by mental-age-matched WS and control subjects. Again, Georgopoulos et al. (2004) concluded that the drawing disabilities in WS are the result of delayed rather than atypical development, since the performance in the two groups was *overall* similar and correlated with mental age. However, some differences between the drawings by the individuals with WS and by control subjects could be observed: The individuals with WS always obtained higher scores than normally developing children for figures consisting of three crossed lines (intersecting lines). Moreover, normally developing children often exhibited errors in segmenting three lines at the center into six lines, while the individuals with WS always drew three crossed lines correctly, a finding inconsistent with the global processing disability hypothesis. The authors did not discuss why this difference was observed.

Although most previous studies construed the drawing disabilities in WS to be a consequence of delay in normal development, studies of visual processing in WS have provided some evidence of atypical processing, such as impaired judgment of line orientation (Bellugi et al., 1988), partially impaired perceptual grouping (Farran, 2005; Farran & Wilmut, 2007; Farran et al., 2003), impaired recognition of objects from unusual viewpoints (Landau, Hoffman, & Kurz, 2006), and difficulty using salient cues on a mental rotation task (Sinton, Farran, & Courbois, 2008). These findings suggest that drawing disabilities more likely reflect atypical processing, and that drawings by individuals with WS need to be more carefully examined in diverse fashions not limited to copying.

Nagai, Iwata, Matsuoka, and Kato (2001) investigated copying, tracing and drawing from memory in WS using single geometric figures including pentagons and hexagons. Individuals with WS could trace a model figure correctly, while they could neither copy nor draw it from memory. In addition, they detected a characteristic error of production of excessive numbers of angles during copying and drawing, especially in the case of pentagons and hexagons, which they termed "angulation error". The finding that they traced figures correctly indicates that the drawing disabilities in WS are not simply due to motor difficulties, as Bellugi et al. (1988) reported. At least four functional steps are believed to be required for copying but not for tracing: (i) feature detection of the target figure, (ii) decision regarding the drawing space, (iii) coordinate transformation of the target figure onto the new drawing space, and (iv) visuospatial working memory during drawing behavior. There appears to be disturbance of some of these steps in WS, although this cannot be detected with typical copying tasks.

In this study, based on the previous findings that individuals with WS can trace figures, we devised a novel tracing task requiring a subject to trace fading single figures on a PC display (fading-figure tracing task: FFT task), and then compared performance between individuals with WS and typically developing children (TD). In performing this task, decisions regarding the drawing space and coordinate transformation are not required, since the target figure (or the trace of the faded figure) must be copied in the same space. Therefore, using variable fading time and several target figures, ability to detect features of the target figure and visuospatial working memory can be examined during drawing behavior.

The FFT task was implemented with short and long fading times. Generally, it is more difficult to draw a figure with a shorter fading time because of increased demand on working memory. This is particularly true in the case of young children who require much time for the development of sufficient visuospatial working memory (Pickering, 2001). Impairment in visuospatial working memory in young adults and adolescents with WS has also been reported (Vicari, Bellucci, & Carlesimo, 2003). Therefore, we predicted both groups would exhibit poor performance with shorter fading times. In a study of typically developing children, the abilities for visual scanning (e.g. counting dots) and tracing a figure in full were found to develop by 4 years of age, while perceptual abilities (e.g. length, orientation, and location of lines), representational abilities (e.g. recognition of complex figures, detection of embedded figures, mental construction), and executive function were found to develop from 4 to 8 years of age (Del Giudice et al., 2000). Therefore, based on visual scanning ability, 5–6-year-old children may exhibit better performance when presented with longer fading times (fading time effect). On the other hand, unlike TD children, individuals with WS exhibit impairment of visual scanning (Hoffman, Landau, & Pagani, 2003; Montfoort, Frens, Hooge, Lagers-van Haselen, & van der Geest, 2007) and would be expected to show only a weak fading time effect. TD children may exhibit better performance when presented with figures with fewer angles, such as squares (figure effect) which are relatively less complex shapes. If WS individuals have different feature detection abilities than do TD children, the effects noted here may differ; for example, the figure type presented (i.e., open or closed) may also affect the performance of WS individuals. Consequently, we predicted that individuals with WS may show a weak fading figure effect and a figure effect that is distinct from that observed in TD children. The purpose of this study was to explore whether patterns of performance differ between individuals with WS and TD children in terms of such effects.

2. Methods

2.1. Participants

Informed consent was obtained from 20 subjects. This study was approved by the ethical committee of the Tokyo Women's Medical University and all experiments were performed according to standards established in the declaration of Helsinki in 1964. The original WS group included five males and five females. Genetic studies of WS by fluorescent *in situ* hybridization (FISH) with five probes have indicated a 1–2 Mb deletion of chromosome 7q11.23 which involves at least 16 genes including the elastin (ELN), syntaxin1A (STX1A), LIM-kinase 1 (LIMK1), and human frizzled homolog of the Drosophila wnt receptor (FZD3), which causes the typical phenotype of WS, including its specific facial features, associated cardiovascular diseases, and mental retardation. One woman with WS was excluded because her age (40 years) was significantly greater than that of the other WS individuals; therefore, the final WS group included five males and four females (age range