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Is the contextual effect weak in people with Williams syndrome? An investigation of information integration ability using pictures

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ABSTRACT

Previous studies have shown that deficiencies in visuospatial perception and semantic processing in people with Williams syndrome (WS) are due to deficient central cohesiveness. Unlike previous studies that used abstract stimuli, this study used pictures to determine the relative ability of people with WS to integrate contextual information with the aim of exploring the nature of central coherence in people with WS. Participants were sequentially presented with a leading background picture followed by a single-item target picture and required to assess the congruence of the two pictures. The results showed that our participants with WS performed the same pattern as controls matched by chronological age (CA) and mental age (MA), demonstrating a contextual effect between congruent and incongruent conditions. Using concrete pictures, contextual integration was successfully induced in people with WS. There were differences between groups in response latencies and accuracy percentages, suggesting that contextual integration in information processing normally develops from childhood to adulthood, but is delayed in people with WS.

1. Introduction

The cognitive profile of people with Williams syndrome (WS), a rare disorder with genetic deficits, is characterized by local preference and global impairment in information processing. This tendency to see the trees rather than the forest is compatible with the hypothesis of weak central coherence (Frith, 1989), which proposes that people with autism have difficultly integrating discrete elements into the whole. Previous studies have revealed that in WS individuals, this uneven profile exists in visuospatial and musical perception (Deruelle, Schon, Rondan, & Mancini, 2005), but does not influence language. However, studies on semantic formation (Hsu, Karmiloff-Smith, Tzeng, Chin, & Wang, 2007) and proposition integration (Hsu & Tzeng, 2011) have demonstrated atypical semantic processing in people with WS. Overall, previous studies have concluded that individuals with WS have a general impairment in integrating contextual information in both verbal and non-verbal domains. In this study, unlike previous studies that have used abstract linguistic stimuli and geometric blocks, we investigated the contextual integration ability of people with WS using pictures.

The preference for local processing strategies among WS individuals has been observed in visual perception and visual memory tasks (Bellugi, Lichtenberger, Jones, Lai, & George, 2000; Bernardino et al., 2002; Bihrle, Bellugi, Delis, & Marks, 1989). One of the standard tasks used to determine local–global processing strategy preferences is copy-drawing. When participants with WS were presented with a picture model of an object (a bicycle) and asked to copy the object, they drew

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parts of it (a petal, a basket, two wheels) rather than an organized configuration (the bicycle itself). A similar pattern of local bias was observed in block design tasks. When people with WS were shown a picture model with blocks and asked to arrange real blocks according to the model, they could only perform discrete elements, but failed to organize a global configuration. The study by Bernardino et al. (2002) confirmed the local bias of people with WS. They used the Navon task (e.g., small letter 'A's are used to form the letter 'P') (Navon, 1977) to elicit the piecemeal- or configure-focused preferences of people with WS. Participants were asked to choose from two options, the probe that was the most like a given target (same global but different local form or different global but same local form) in no-, local-, and global-rotation conditions. The results revealed that individuals with WS performed differently than controls in all of the conditions. The people in the control groups usually chose the form with the same global but different local configurations as being the most similar to the targets, whereas the participants with WS tended to choose probes with the same local but different global forms. Another study that explicitly instructed participants to choose a probe from local- or global-match alternatives when matching a target revealed significant differences between the clinical group and the control group. Unlike the people in the control group, who preferentially chose global-match probes, individuals with WS preferred local-match probes. A study by Bellugi et al. (2000) that used a visual-motor integration task (a copying version of the Navon task) confirmed the detail-focused information processing style of participants with WS. These studies have confirmed an obvious detail-focused processing tendency in people with WS.

This behavioral preference for a local bias among people with WS has been linked to atypical brain activity, specifically deficient gamma-band activity (an index of neurological processing for information integration; Grice et al., 2001). In the Grice et al. passive viewing-of-faces test, controls showed a burst of gamma-band activity when processing faces, whereas people with WS failed to show the same pattern. Thus, the lack of construction in visual perception in people with WS was evidenced as correlating with a neuro-physiological deficit. It has been hypothesized that this visuospatial deficit in the WS population is a result of the atypical development of lower-level visual processing (Grice et al., 2003). The study by Grice et al. compared brainwaves evoked by four types of visual stimuli: human faces (c.f., unanalyzed fillers), real squares, Pacmen stimuli, and the Kanizsa square (an illusory triangle perceptual image). All of the participants were required to trace the contours of the Kanizsa square with their fingers to ensure that they recognized the illusory image. The controls showed significant differences in the N1 component between the illusory square, the real square, and the Pacmen stimuli, whereas the participants with WS did not differentiate between the illusory square and the real square. This abnormality in contour perception might be the result of early atypical neural development in people with WS.

The study by Farran, Christopher, and Gathercole (2001) yielded the opposite result; people with WS had normal integrating abilities in the block design task and the embedded figure test. Unlike people with autism (Shah & Frith, 1993), people with WS benefited from the segmentation of blocks and their reaction times and number of correct responses suggested that they had the same global processing tendencies as the control group. Moreover, both groups had similar search times and number of correct responses in the task using embedded figures in larger configurations. This study also confirmed the existence of developmental delays in the participants with WS, who were matched with children of the same mental age. However, only a few studies of people with WS have found either intact or delayed abilities with geometric blocks in visuospatial construction.

Despite their piecemeal processing preference, people with WS show relative strength in language skills and have rich lexical semantics and grammatical knowledge. Bellugi et al. (2000) found that people with WS were able to name significantly more low frequency words (hippopotamus, dragon, brontosaurus) and produce a similar number of lexical items as the control group in a timed category-generating test (animal) (Bellugi et al., 2000). They also showed normal semantic priming when identifying the relationship of synonyms (hamster and mouse) and taxonomic words with functional relations (broom and floor) (Tyler et al., 1997). Similar to the people in the control group, participants with WS were able to reason counterfactually by producing the conjunction *if* with subjunctive moods and could clearly define the primary and secondary meanings of homonyms (money bank vs. river bank). Moreover, a conceptual formation study with the false memory paradigm conducted by Hsu et al. (2007) showed a normal-like formation of gist themes among WS individuals who were presented with semantically related items. Participants with WS listened to words that were associated with gist themes in a learning phase and were then required to differentiate old words from mixed lure items (non-presented semantically related associates) and new items (non-presented semantically unrelated associates) in a recognition test. The results showed no significant differences between participants with WS and the control group.

In recent years, there has been growing evidence that people with WS express atypical processing of sentential propositions at the behavioral level (Hsu & Tzeng, 2011) and abnormal brainwave signatures at the neurological level (Hsu et al., 2007; Neville, Mills, & Bellugi, 1994). In Hsu and Tzeng's study, participants with WS were presented with sentences containing various numbers of propositions from related scenarios: a maximum of three propositions in the learning phase. In the recognition phase, the participants were asked to recognize whether the presented sentences, containing up to four propositions, had been heard before. The results revealed that the adult controls recognized four-proposition sentences as old ones (presented sentences), indicating that they had automatically integrated the contextual information, whereas participants with WS were unable to recognize even one-proposition sentences. Their performance was also significantly different from the children controls who could recognize sentences with up to two propositions. These findings suggested that people with WS are impaired in the contextual integration of information processing. Hsu et al. (2007) also observed