Contextual Coherence of Information Integration in People with Williams Syndrome: Implications for Cognitive Interventions

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Abstract

A growing body of evidence from behavioral and neurophysiological studies has shown that there is an asymmetry in the brain and behavioral performances in both verbal and nonverbal domains in people with Williams syndrome (WS). In these studies, clinical individuals were observed performing behavior that appeared normal. However, these individuals exhibited atypical neurological signatures, suggesting that they reached these typical-like performances through a distinct neurological network. This article investigates the implications of this brain and behavioral asymmetry for educating people with WS. Based the effects of computer-assisted interventions in a wide variety of individuals with developmental disabilities and neurological disorders, it is suggested that people with WS would also benefit from taking similar computer-based training programs. Through this training, it is hoped that people with WS can achieve typical behavioral performances and even function with typical neurological mechanisms.

Keywords: cognitive intervention, asymmetry of brain and behavior, Williams syndrome

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1. Introduction

Traditionally people believe that speech therapy benefits those who need articulation adjustment and speech training. The same concept could also apply to cognitive interventions for people with developmental disabilities. Several studies have reported improvements after patients took behavioral or computer-assisted cognitive training programs. The positive influences of this type of intervention have been observed in people with developmental disabilities and neurological disorders. This article suggests a possible research direction for assisting people with Williams syndrome (WS). WS is a rare disorder caused by a gene mutation on chromosome 7q11.23 that results in mental retardation, with an average IQ score of 55 (Korenberg et al., 2000). Previous research efforts have mainly focused on investigating the uneven cognitive profiles of this clinical population, who are relatively good at language and face processing but are relatively poor at visuospatial perception (Bellugi, Lichtenberger, Jones, Lai, & George, 2000; Semel & Rosner, 2003). However, little research has studied potential cognitive interventions for people with WS. Recently, a growing body of evidence has emerged that shows an asymmetry in the brain and behavioral performances of people with WS, meaning that they behave normally but exhibit atypical brain signatures both for processing language (Hsu et al., 2007) and faces (Hsu & Chen, 2014; Mills, Alvarez, George, Appelbaum, Bellugi, & Neville, 2000).

2. The Effects of Cognitive Interventions Using Computer-Assisted Programs

Bauminger (2002) conducted a 7-month cognitive behavioral intervention program on children with high-functioning autism (HFA) and evaluated the efficacy of this intervention in improving the social cognition and social interaction of these clinical individuals. As part of this intervention, the children with HFA received three different types of training that aimed to improve their social and cognitive shortcomings. First, they were trained in problem solving skills through composing stories that required cognitive reasoning. Their stories were evaluated in five categories: activity (active/passive), relevance (relevant/irrelevant), number of solutions, variety, and content. Second, they were trained to recognize complex emotions (such as guilt, embarrassment, and loneliness) and were asked to provide a specific event related to that emotion and include an audience who witnessed the display of the emotion. The third aspect of this intervention was geared toward interpersonal interaction with peers, with guidance from teachers and parents. For example, the children were taught how to initiate a conversation, how to deal with confronting a friend, and how to share with a friend. In addition, these children were also involved in peer meetings that emphasized reciprocity and continuity in communication, for instance making a phone call to practice social skills and initiate conversations. These interventions resulted in improved performances in problem solving, understanding complex emotions, and engaging in pro-social behavior. Although this study lacked a control group, the significant improvement after treatment compared to the before condition indicates that these children with HFA were able to learn about emotions and understand social cognition through this training.

In another study, people with Asperger syndrome (AS) and HFA were trained with computer-based programs to recognize emotion from faces, voices, and contexts (Golan & Baron-Cohen, 2006). This training aimed at improving the socio-emotional ability of these clinical individuals. The intervention group was trained for 2 hours a day for 10 to 15 weeks in recognizing complex emotions, such as insincerity, and mental states. The participants were evaluated before and after the intervention. Each participant received several types of software, including the Cambridge mindreading face-voice battery, a task on reading the mind in the eyes, a task on reading the mind in the voice, and a task on reading the mind in film. Two control groups were recruited: people with the same syndrome who did not receive the interventions and people with typical development. It was hypothesized that the two clinical groups would perform worse before of the intervention than the group with typical development and that the individuals with AS and HFA would improve after taking the Mind Reading training. The results confirmed these hypotheses. The group of individuals with AS and HFA who received the training using interactive, multimedia, educational software improved in their ability to recognize complex emotions and mental states, proving that computer-based intervention can help people with developmental disabilities.

A study investigating the executive function of 4-year-old children with attention deficit hyperactive disorders (ADHD) showed improvements in working memory and reasoning after training with computer games (CogMed). Similar benefits were observed in a selective attention task in 4- and 6-year-old children. These participants also showed improvements in two of three inhibition tasks. Other interventions that have been shown to be effective in improving executive function include aerobic exercises, traditional tae-kwon-do, mindfulness training (e.g., yoga), and appropriate curriculum design for children with specific needs (Diamond & Lee, 2011). This qualitative change in cognitive behavior after the use of technology-based training programs can be observed not only in people with developmental disabilities but also in people with neurological disorders. A study of people with Alzheimer’s disease (AD) using computer-assisted training programs showed domain-specific effects after 6 weeks of intensive practice (Barnes et al., 2009). These clinical participants received intervention assignments, including listening, reading, and playing a visuospatial-oriented computer game, to recover their brain plasticity. Their memory (and/or learning), language (and/or visuospatial cognition), and attention (and/or executive function) were tested before and after the training to access the effects of this intervention. The results showed that these participants with AD showed improvement in memory/learning compared to a control group but exhibited less improvement in visuospatial cognitive function (except for the spatial span test). Although further studies are needed to clarify why certain domains are amenable to training whereas others are not, these results suggest that computer-based interventions are effective for people with neurological disorders. For instance, another study on cognitive rehabilitation for people with AD showed that training improved the neuropsychological functioning of the participants in attention, memory, perception, visuospatial cognition, language, and nonverbal intelligence (Cipriani, Bianchett, & Trabucchi, 2006). During the 16-day training program spread across 4 weeks, the participants were evaluated on their performances before and after the intervention based on their total score, time taken, and the number of errors that they made. Another group of people with mild cognitive impairment (MCI) was also recruited in this study. The results showed that both people with AD and those with MCI benefited from receiving the computer-based neuropsychological training program.

Another neurocognitive disorder, schizophrenia, has also been shown to benefit from computer-assisted remediation (Kurts, Seltzer, Shagan, Thime, & Wexler, 2007; Bellucci, Glaberman, & Haslam, 2002). In Kurts and colleagues' study (2007), the patients undertook a 100-hour, yearlong intervention aimed at improving their attention, verbal memory, non-verbal memory, and language processing in both visual and auditory modalities. Their pre- and post-training assessments were compared to assess the effects of the intervention. The results showed that the working memory of the patients with schizophrenia was significantly improved, particularly in the digit span and arithmetic assessments. This finding provides more support for the use of computer-assisted remediation for people with neurocognitive deficits. Additionally in Bellucci and colleagues' study (2002), the negative symptoms of patients with schizophrenia were largely reduced after participating in the computer-based rehabilitation for cognitive functioning.

3. Asymmetry of Brain and Behavioral Performances in People with Williams Syndrome

People with WS show local preference but global ignorance in processing perceptions, including visuospatial construction, emotional identification, and facial recognition. For instance in visuospatial construction, people with WS drew only parts of the objects shown in pictures, such as a bicycle and a swimming pool, in a copying task (Bellugi et al., 2000). People with WS also focused on parts rather than wholes in block design tasks. Their arrangements generally lacked the form of configurations. In terms of their emotional recognition, Gagliardi, Frigerio, Burt, Cazzaniga, Perrett, and Borgatti (2003) presented animated facial expressions that mimicked human emotions, such as anger, disgust, happiness, sadness, and fear, to individuals with WS. Their study showed that the participants with WS performed as well as the mental age matched healthy children, but were inferior to chronological age matched healthy adults in their sensitivities to different emotional expressions. It was concluded that the limited ability of individuals with WS to recognize facial expressions was due to a deficiency in coding configural information. Moreover, although the healthy controls showed a correlation between age and the recognition of emotional expressions, the clinical individuals failed to show this pattern. Instead, their intelligence, rather than their age, was correlated with their correctness of recognition, suggesting that their ability to recognize expressions would not improve with age. These studies showed that people with WS exhibit an atypical pattern of processing facial expressions.

This processing preference was also observed in facial identification. Hsu and Chen (2014), asked individuals with WS detect faces. A model face was displayed on the screen. This was followed by a target face that sometimes had changes in its features (such as the eyes and mouth) and configurations. The participants had to judge whether each target face was the same as or different from the target face. This study showed that these clinical individuals recognized feature-changed faces fairly easily compared with the controls but had difficulty recognizing faces in which the configuration had been changed. This normal-like behavioral finding in relation to face detection is similar to the observations in a study of face identification (Mills et al., 2000). In this study, two faces were shown consecutively with one face as the prime and the other as the target. The target faces were presented in upright or inverted orientations. Because inverted faces lose configuration information but keep featural cues (Karmiloff-Smith et al., 2004), it was hypothesized that the normal participants would have difficulty processing the inverted faces. The results showed that people with WS exhibited longer response times and a greater number of errors in processing the inverted faces compared to the controls, suggesting that this clinical group has a normal-like face processing strategy. However, the neurological signatures recorded from event-related potentials between people with WS and the controls were different. In Hsu and Chen's study (2014), although the control group exhibited a difference in the left hemisphere when processing changed faces (feature-changed faces, configure-changed faces), the participants with WS did not show this pattern. Additionally, the control group processed the two types of changed faces differently in their right hemispheres, whereas the clinical individuals did not show a significant difference in their brainwaves when processing these two different types of changed faces. This asymmetry between brain and behavior performances in facial processing for people with WS was also demonstrated in a study by Mills et al. (2000). The difference was observed at the very beginning of the processing task. Although the healthy controls responded to prime faces with a large N100 and a small

N200, the participants with WS showed a small N100 and a large N200. The same difference was observed when the target faces were shown to the two groups. In addition, the controls showed a N320 negativity to upright faces, whereas the participants with WS showed this negativity when viewing both upright and inverted faces. Beyond behavioral performances, these neurophysiological findings show that people with WS process faces atypically.

This unique processing pattern of people with WS was also observed in a verbal conceptual formation study with a false memory paradigm (Hsu et al., 2007). The behavioral performance of people with WS was the same as the healthy controls, who showed a high number of false positives to non-presented semantically related items (i.e., lures). This suggests that both groups misrecognized the lures as old items that were presented in the acquisition stage. However, the brain signatures in the two groups showed distinct patterns. The clinical individuals differentiated old from new items in a similar way as the healthy controls, but they responded differently to the lure items. The healthy controls processed lures as old items whereas the participants with WS recognized the lures as new items. In both the verbal and non-verbal domains, individuals with WS exhibit an asymmetry between the seeming normality of their behavior and the difference in their brainwaves compared to healthy controls. These results indicate that people with WS achieve a normal-like performance (compared to chronological age matched or mental age matched controls) via a distinctly different brain mechanism. The asymmetry shown in these studies demonstrates that people with developmental disabilities, such as WS, may be able to perform like typically developing controls through the use of cognitive interventions such as the computer-assisted training programs that have been shown to be effective for individuals with developmental and neurological disorders.

Based on this review of the cognitive impairments in visuospatial construction and emotional recognition in the non-verbal domain for people with WS, a possible intervention direction might be the use of a computer-based training program to improve the cognitive abilities of this clinical group to reach a normal-like behavioral level. Furthermore, based on these findings related to verbal and face processing in nonverbal domains, this computer-assisted educational software may benefit people with WS who exhibit an asymmetry between their typical behavior and atypical neurological signatures compared to healthy controls. It is hoped that these clinical individuals might develop similar neurological mechanisms to typically developing individuals. Computer-assisted intervention training takes only a short time and can yield a great deal of improvement in the cognitive abilities of people with developmental disabilities (such as high-functioning autism and attention deficit hyperactive disorders) and neurological disorders (such as Alzheimer’s disease and schizophrenia). People with WS may benefit from similar cognitive training and thus an effective rehabilitation of these individuals might be reached in the near future.

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