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Chapter 3

Cross-Domain Investigation of Weak Central Coherence in People with Williams Syndrome: Asymmetrical Brain and Behavioral Performances in Verbal and Nonverbal Domains

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Abstract

Background: Central coherence is a fundamental cognitive ability concerned with integrating information from parts into a whole. Deficiencies in contextual integration, termed weak central coherence (WCC), have been reported in people with autistic spectrum disorders (Frith, 1989). This study investigated whether WCC is a syndrome-specific or syndrome-general deficit. To examine this issue, people with

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Williams syndrome (WS), a population with a genetic disorder, were recruited. The reported cognitive profile of this clinical group is asymmetrical, with good language ability but poor visuospatial ability (Bellugi et al. 2000), so a secondary aim of this study was to examine whether different levels of WCC deficits are observed in the verbal and nonverbal domains of people with WS.

Studies and Results: Two studies were conducted to examine the verbal domain: a concept formation from a semantic network study and a causal inference on ambiguities study. The concept formation study used the false memory paradigm and revealed normal-like behavioral performance but abnormal brainwave patterns in people with WS. The causal inference on ambiguities study revealed that people with WS were not deviant but delayed in making causal inferences, both backward and forward, when comprehending ambiguous words in contexts. In the nonverbal domain, a face processing study and a series of contextual integration studies using unimodal or cross-modal methods were used. The face study involved showing faces to people with WS, and then showing them the same faces with changes to the features and configurations, and the differences in processing were recorded. The results showed that people with WS exhibited similar behavioral patterns to the healthy controls, but their brainwaves differed. The controls processed faces with changes in configurations differently from faces with changes in features, while the clinical group did not exhibit this distinction neurologically.

The contextual integration studies, using different modality presentations, revealed a delay in semantic integration in people with WS. These findings challenge the traditional claim that people with WS are deficient in visuospatial construction ability. This clinical group demonstrated central coherence ability in terms of contextual integration, as long as the stimuli were socially meaningful rather than abstract block designs or pattern constructions. *Conclusion:* WCC is a syndrome-general deficit manifested in the verbal and nonverbal domains of people with WS at both the behavioral and the neuro-physiological level. The finding of brain-behavioral asymmetry confirms the neuroconstructivist view that impairment in the early stages influences later development (Karmiloff-Smith, 1998) and has implications for educational intervention. People with WS may benefit from cross-modal learning, which is related to social knowledge. Further studies may lead to further insights regarding brain plasticity in people with developmental disabilities.

Introduction

Local preference and global disinterest are characteristics of weak central coherence (WCC), which is identified as a symptom of people with autism. This asymmetrical processing is recognized as a cognitive style rather than an impairment (Happé, 1999), and is observed in verbal and nonverbal domains (Happé, 1997; Sha & Frith, 1993). In both domains, people with autism exhibit deficient processing in the global integration of local elements, or impaired organization of piecemeal cues that enable the understanding of other people's minds. This study demonstrates that WCC is a syndrome-general but not syndrome-specific phenotype by reviewing a series of studies on people with Williams syndrome (WS), with evidence from brain and behavioral findings in both verbal and nonverbal domains.

Williams syndrome (WS) was first diagnosed in 1961 (Williams, Barrett-Boyes, & Lowe, 1961). It is a rare disorder with genetic deficits on chromosome 7q11.23 and the etiology is 1 in 7500 live births (Strømme, Bjømstad, & Ramstad, 2002). Weak central coherence is observed at the visual perceptual level and in semantic conceptual level processing. Standard block design, pattern construction, and visual-motor integration tests have revealed that people with WS are impaired at the visual perceptual level (Bellugi, Lichtenberger, Jones, Lai, & George, 2000; Brock, 2007; Mervis & John, 2010). These investigations into global and local asymmetry show that people with WS display a preference toward local, rather than global, focus. For example, in the visual-motor integration test, people with WS had difficulty drawing a two-story house, and only depicted certain sections of it. This was also observed in pattern construction and block design tests, in which people with WS were unable to arrange blocks according to the displayed models. The opposite finding was reported by Farran, Christopher, and Gathercole (2001), who found normal-like performance in embedded figure tests; that is, people with WS did not identify a preference for local figures in global configurations. However, most of the findings did support an asymmetry, with local bias and global disinterest, in the visual processing of people with WS.

Another study in the nonverbal domain, focusing on music processing, confirmed this asymmetry in people with WS (Deruelle, Schon, Rondan, & Mancini, 2005). In clinical tests, people with WS performed equally well at processing contour-violated and interval-violated excerpts whereas the typically developing control individuals showed better recognition of global-violated melodies. These findings were concluded as the precedence of global processing in the typically developing controls in contrast to the lack of this

precedence in the clinical individuals. This asymmetry in people with WS has also been shown in face processing, another non-verbal domain. Normal-like performances were shown with the Warren face recognition test (Bellugi et al., 2000). In this test, people with WS were first shown a model face and were then required to identify the same face from six differently orientated options. People with WS accurately recognized the faces. Mills et al. (2000) found that people with WS showed the same inversion effect in face processing as healthy controls in a face matching task, suggesting that people responded slower and erred more in their recognition of inverted faces. No difference was observed between clinical and control groups in recognizing upright oriented faces. This finding suggested that people with WS used a configural strategy to process faces, rather than piecemeal processing, as they exhibited good recognition of inverted faces. Another study showed normal-like performances (Hsu & Chen, 2014a) when investigating the ability of people with WS to detect faces when the configuration was changed, suggesting unimpaired global processing. In Hsu and Chen's study, participants with WS were first presented with a model face. This was followed by a further series of faces with sequential changes either to the features (eyes or mouth) or to the configurations. The participants were asked to judge whether the two consecutive faces were the same or different in identity, by pressing one of two buttons. Participants with WS and the age-matched controls recognized a similarly high percentage of the configuration-changed faces, suggesting that face processing is not deviant in people with WS. Two processing patterns in the nonverbal domain were therefore observed in people with WS. The clinical individuals failed to combine parts into wholes in visuospatial construction perception tests, but showed perceptual coherence in face processing, further demonstrating a within-domain asymmetry in people with WS.

In the verbal domain, people with WS are reported to be fluent in language production and to possess good knowledge of lexical semantics (Jones et al., 2006; Tyler et al., 1997). In Jones et al.'s spontaneous narrative study (2006), people with WS were asked to describe a picture of a boy searching for a missing frog, and a picture depicting the theft of a cookie. A high percentage of the discourse in their linguistic output was made up of connecting words and various emotional expressions. In Tyler and colleagues' study on lexical semantic priming (1997), people with WS displayed normal-like priming effects to words related in function (e.g., broom vs. floor) or taxonomy (e.g., mouse vs. hamster). Another study investigating the spontaneous generation of semantic categories within a limited time revealed that people with WS possessed a rich knowledge of lexical semantics,

compared to both healthy controls and people with Down syndrome. People with WS produced a significantly higher percentage of low frequency words than the healthy controls (Bellugi et al., 2000). A later study used a semantic conceptual task with a false memory paradigm to investigate the ability of people with WS to integrate semantically related words (Hsu, Karmiloff-Smith, Tzeng, Chin, & Wang, 2007). Participants with WS showed the same central coherence ability as healthy controls, and formed gist themes (e.g., winter) from aurally presented semantically related associates (e.g., scarf, glove, sweater, heater, snow). This successful formation of configurations toward meanings indicated that people with WS organized semantic networks when presented with semantically related words, while the recognition rates of semantically unrelated words (e.g., piano) were low, which compared well to their age-matched controls, both mentally and chronologically.

Previous studies on verbal and nonverbal domains regarding central coherence appear to support the notion of asymmetrical performances in people with WS, who exhibit good language ability but poor visuospatial construction perception. However, neurologically observed brain patterns were inconsistent with normal-like behavioral performances in both the verbal and nonverbal domains in people with WS.

Challenges to Traditional View of Asymmetrical Performances

A series of studies using pictures, instead of abstract geometric blocks, demonstrated non-impaired central coherence ability in people with WS (Hsu, 2013a, 2013c). In Hsu's study (2013a), pictures relating to social event knowledge were presented with a leading background scene for 1500 ms followed by a target object until the participants responded. There were two conditions in the study: congruent or incongruent. In the congruent condition, the background scene matched with the target, such as *a swimming pool* and *a pair of swimming goggles*. In the incongruent condition, the background and target were mismatched in semantic appropriateness; for instance, *a swimming pool* and *a skateboard*. Participants pressed corresponding buttons to indicate the congruency between the pictures. People with WS showed a congruency effect of faster responses and lower errors to matched pairs than mismatched ones, similar to the healthy controls. However, the clinical individuals' performance was the same as that of the mental age (MA) matched controls

rather than their chronological age (CA) matched controls, suggesting a delayed but not deviant central coherence ability. A follow up study, replacing the target pictures with auditory stimuli, was conducted to investigate whether the concreteness or social relatedness of stimuli played a role in the congruency effect of people with WS (Hsu, 2013c). It was predicted that social relatedness would be an essential factor in the judgments of semantic appropriateness. The results confirmed the prediction, showing a similar congruency effect in people with WS when using auditory targets. Thus, unimodal or cross-modal studies of visual primes elicited unimpaired or delayed central coherence in the nonverbal domain in people with WS. To systematically examine the contextual effect in people with WS, further investigations of auditory primes with visual or auditory targets were conducted (Hsu, 2014b). People with WS and healthy controls showed the same semantic priming effect to congruent pairs compared to incongruent ones, but people with WS had longer reaction times and higher error rates, indicating developmental delay but not deviant performance. The results demonstrated a systematic contextual effect in people with WS, with both unimodal and cross-modal presentation. A modality effect was observed for both visual and auditory primes, in both healthy controls and clinical individuals. Cross-modal presentations elicited better performances from people with WS, with faster responses and higher accuracy rates to semantically matched conditions than to mismatched conditions. Together, these studies suggest a not-completely-impaired central coherence ability in people with WS: their performance differed from that of CA-matched controls, but not from that of MA-matched controls. These studies challenge the traditional view of completely impaired or deviant processing in the visuospatial construction perception of people with WS. This indicates developmental delay rather than deviance. A further implication for educational intervention concerning people with WS is that cross-modal studies (i.e., visual-auditory or auditory-visual presentation) improve learning, as they draw on existing strengths by using socially related stimuli. This implication is compatible with the correlation of sociability and emotionality with musicality in people with WS (Ng, Lai, Levitin, & Bellugi, 2013), which suggests combining social and emotional aspects via music therapy.

Similar challenges have been observed in the verbal domain. In addition to a study of word-level semantic integration (i.e., the semantic conceptual formation study with a false memory task by Hsu and colleagues in 2007), Hsu and Tzeng (2011) conducted a study on the sentential integration of embedded propositions using proposition integration paradigm (Franks & Bransford,

1972). People with WS listened to several sentences with various numbers of propositions in the learning phase, and in the later recognition phase were required to judge whether the shown sentences had been previously learned. The results revealed that people with WS had difficulty integrating propositions in comprehension. While CA-matched controls successfully integrated sentences with more than three propositions, MA-matched controls showed an integration effect concerning sentences with more than one proposition. The age-matched control groups misrecognized sentences with more propositions (i.e., ≧2 for children, ≧3 for adults), whereas our clinical group could not integrate sentences with one proposition. They showed a deviant pattern from their control groups and had difficulty in integrating larger units such as sentential propositions.

A causal inference study using homonym comprehension showed the same delayed development in people with WS as the MA-matched individuals (Hsu, 2013b). There were two tasks: backward inference and forward inference. In the backward inference task, participants had to make causal links from consequences to causes; in the forward inference task, participants had to make causal inferences from causes to consequences. A similar design was used in the two tasks. People with WS listened to a short narration depicting familiar cartoon characters in different scenarios. Participants were asked a comprehension question and given three alternative responses to choose from. The key was to understand the embedded homonyms. Each homonym had two interpretations: either a figurative meaning or a literal meaning. For example, a homonym 潑冷水 (po1, 'to dampen'; leng3, 'cold'; and shuei3, 'water') contains the figurative meaning *to dampen one's enthusiasm* and the literal meaning *to pour cold water on someone*. An example of a test scenario is: *Sponge Bob would like to eat the candies on a shelf. He asked for help from Squidward Tentacles. But Squidward Tentacles dampened Sponge Bob's enthusiasm* (the original text in Chinese was printed in Hsu 2013b, p. 3336). A comprehension question followed: *What did Squidward Tentacles do?* Three options relating to the figurative, literal, and unrelated meanings of the homonym were read aloud to participants. In this example, participants heard *Squidward Tentacles poured cold water on Sponge Bob* as the interpretation based on the literal meaning; *Squidward Tentacles did not help Sponge Bob to get the candies* as the figurative interpretation; and *Sponge Bob likes to take a bath with cold water* as the unrelated interpretation. After the three options were read, participants were asked for the correct interpretation based on the context of the narration. The results of figurative comprehension showed that people with WS responded

less accurately than CA-matched controls, but did not perform significantly differently from the MA-matched controls. From the three groups, a higher percentage of clinical individuals chose unrelated meanings. Together, these studies challenge the traditional view that people with WS possess good linguistic ability, suggesting that they are deviant in the central coherence of proposition integration and delayed in backward and forward causal inference.

Asymmetry of Brain and Behavioral Performances

Recent neurological findings on central coherence in verbal and nonverbal domains have revealed brain and behavioral asymmetry in people with WS. Behaviorally, as with the false memory task (Hsu et al., 2007), people with WS formed gist themes after listening to semantically related associates, and misrecognized non-presented theme lures as old items that had been previously presented in the learning phase, as did healthy controls. They also correctly rejected non-presented, semantically unrelated words. However, people with WS showed different brain signatures from their healthy controls. Unlike the CA-matched controls, who showed similar neurological responses when processing presented old items and non-presented semantically related theme lures, people with WS showed different response to these two types of words. While the neurological processing of the CA controls was different for the non-presented theme lures and the non-presented semantically unrelated words, people with WS did not show any distinction between the two types. The clinical individuals showed deviant brain processing from the typically developing controls. Hence, a unique pattern of brain and behavioral asymmetry in semantic conceptual formation was observed in people with WS. That is, brain signatures and behavioral performances in the verbal domain were mismatched in people with WS.

The brain and behavioral mismatch was also observed in the nonverbal domain of face processing in people with WS (Hsu & Chen, 2014a). In Hsu and Chen's study, behaviorally people with WS showed a normal ability to detect faces with changes in configurations and faces with changes in features, as did the healthy controls. However, people with WS showed distinct brain processing from the typically developing controls. While the typically developing controls responded differently to configuration-changed faces and feature-changed faces in the vertex area from 390–698 ms in both

hemispheres, people with WS did not show a distinction between these two types of faces in the same cortical region. Thus, the asymmetry between brain and behavioral performances is supported by the findings in the nonverbal domain. In summary, cross-domain asymmetry is evident at both the brain and the behavioral level in people with WS.

Weak Central Coherence of Contextual Integration in People with WS

The aim of this study was to investigate whether WCC is a syndrome-general or syndrome-specific phenotype. A series of studies in the verbal and nonverbal domains were reviewed. The investigation began by examining the asymmetry between local bias and global inattention in visuospatial construction perception, and extended to other nonverbal domains such as musical perception and face processing. Processing asymmetry is identified in the nonverbal domain, with deviance in visuospatial construction perception but normal-like face processing. Similar asymmetry is observed in processing in the verbal domain of people with WS. Although this clinical group exhibits deviant central coherence in sentential proposition integration, there is no evidence of an impairment but delay in causal inference. These cross-domain studies of people with WS challenge the traditional view that people with WS possess good linguistic abilities but poor visuospatial construction abilities. Together, these studies confirm that WCC is a syndrome-general phenotype in people with developmental disabilities.

A secondary goal was to examine whether different levels of WCC in the verbal and nonverbal domains could be identified in people with WS. Behaviorally, people with WS show delayed development when comprehending causal narrations and contextual integration of social event knowledge. They perform similarly to typically developing controls in concept formation and face matching, in both upright and inverted orientations. It appears that WCC is identified in people with WS to varying degrees. However, further neurological studies exploring the brain signatures associated with central coherence have revealed deviant performances in both verbal and nonverbal domains in people with WS, suggesting that there is a common underlying cognitive mechanism behind their normal-like behavioral performances. Asymmetry between brain and behavioral performances is found in people with WS. The importance of investigating neurological

responses in addition to conducting behavioral studies is therefore highlighted, as this can uncover the nature of the cognitive abilities of people with developmental disorders.

This series of studies on WCC has practical educational implications for people with WS. The results of the causal inference and proposition integration studies can assist the parents and teachers of people with WS. Repeating the tests concerning the meanings of homonyms, interpreting these within contexts, and linking propositions embedded in sentences will strengthen the semantic understanding of people with WS. The findings of the concept formation study can inform parents and teachers of people with WS concerning the connections between semantically related words, to enable the successful building of semantic networks. The results of the contextual integration of socially related stimuli studies illustrate that cross-modal presentation has more benefits for people with WS than unimodal presentation. These studies also imply that the social relatedness of stimuli is a more influential factor than concreteness per se. Furthermore, the observations concerning face detection and configuration changes suggest that parents or teachers of people with WS should concentrate on the focus configurations of facial features. These methods of teaching should be carried out slowly, with repeated explanations, as people with WS are delayed in many aspects of cognitive development. The findings of brain and behavioral asymmetry in verbal and nonverbal domains confirm the neuroconstructivist view that impairment at an early stage leads to devastating effects in later development (Karmiloff-Smith, 1998, 2007).

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