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威廉氏症候群整體訊息處理歧異的臉部辨識神經生理證據

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過去研究文獻指出威廉氏症候群有見樹不見林的認知處理傾向，不僅在非語言範疇觀察到，近年來在語言範疇裡也得到證明，符合自閉症症候群中心連貫缺損認知處理模式，本研究旨在發掘威廉氏症候群臉部辨識能力是否也有中心連貫缺損情形，用誘發事件相關電位(event-related potentials, ERPs)尋找整體訊息處理歧異神經生理證據。研究方法讓受試者看連續呈現在電腦螢幕上的臉部圖片，第一張為模型臉(model face)，第二張為改變臉，有特徵改變(feature-changed)及整體改變(configure-changed)兩種臉，受試者需判斷連續出現的兩張臉是否相同。研究結果顯示威廉氏症候群與正常發展控制組的行為結果相同，無歧異表現，但是在腦波表現方面，當正常發展控制組在左右腦清楚區辨特徵改變臉與整體改變臉時，威廉氏症候群無法區辨這兩種臉，且以知覺特徵方式處理臉部整體改變，研究結果以神經生理證據支持威廉氏症候群臉部辨識歧異的認知處理傾向，也提供威廉氏症候群在非語言範疇的中心連貫另一缺損證明。文章也對行為與大腦不對稱表現進行探討。

關鍵詞：中心連貫缺損、威廉氏症候群、誘發事件相關電位、整體訊息偵測缺損、臉部處理

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Introduction

The major goal of this study was to investigate whether Weak Central Coherence (WCC) is syndrome-general or syndrome-specific in people with developmental disabilities. We invited people with Williams syndrome (WS) to take part in our study with the minor goal of finding evidence for the existence of an interactive specialization framework among people with developmental disorders (Johnson, 2000, 2005; Johnson, Grossman, & Cohen Kadosh, 2009). Central coherence is defined as the ability to arrange parts into a whole (Frith, 1989). The tendency of central coherence is to integrate information into meaningful representations and to use diverse information to construct higher-level meaning in a given context (Frith & Happé, 1994). There are two levels of central coherence: the semantic-conceptual level and the visual-perceptual level (Plaisted, 2001). Many studies of the visual-perceptual level have demonstrated that central coherence generates perceptual illusions, such as the Kanizsa triangle (the triangle looks whiter than the background one), Tichener circles (a circle surrounded by larger circles looks bigger than a circle of the same size surrounded by smaller circles), and Muller-Lyer figures (a line with a stretched-out arrows looks longer than a line of the same length with normal arrows). That is, people form perceptual illusions due to the drive of central coherence. If people lack the drive to form cohesiveness from context, they are described as having WCC. People with Autism Spectrum Disorders (ASD) are weak in central coherence; they prefer local units and are unaware of global configurations. It is WCC that makes it difficult for people with ASD to integrate contextual information and it is the reason they have problems understanding other people's minds. Since 1989, people with autism have been documented as being deficient in contextual information integration in both the verbal and nonverbal domains. Without direct knowledge of whether all people with developmental disorders have this deficit, it was necessary to invite people with WS to take part in this study.

People with WS have missing genes on chromosome 7q11.23; this is a rare birth disorder with a low etiology of 1 in 7500 live births (Strømme, Bjømstad, & Ramstad, 2002). The uneven cognitive profiles of preserved semantic-conceptual knowledge and the impaired visual perception of this group have been demonstrated in previous studies (Bellugi, Lichtenberger, Jones, Lai, & George, 2000; Bihrle, Bellugi, Delis, & Marks, 1989; Donnai & Karmiloff-Smith, 2000; Tyler et al., 1997). However, recent findings in our laboratory have challenged the traditional understanding of the role that uneven profiles play in the cognitive abilities of people with WS in both language and visuospatial construction. In language-related studies, we used the false memory paradigm to demonstrate that people with WS exhibit deficient concept formation (Hsu, Karmiloff-Smith, Tzeng, Tai, & Wang, 2007) and an atypical integration of propositions embedded in sentences (Hsu & Tzeng, 2011), suggesting that they have WCC at the semantic-conceptual level. Our latest study investigating causal coherence in people with WS showed delayed ability in backward and forward inferences (Hsu, 2013b), indicating relatively but not absolutely good language

ability in people with WS. Another study demonstrated delayed visual-perceptual central coherence using more concrete pictures in people with WS (Hsu, 2013a). A follow-up study using the cross-modal presentation of visual backgrounds and auditory targets revealed delayed contextual integration, suggesting the sensitivity of social relatedness in people with WS (Hsu, 2013c). Unlike the traditionally recognized uneven cognitive profile, these recent findings suggest that people with WS are neither absolutely linguistically intact nor completely impaired in their visual perception. In this study, we were interested in investigating whether the challenge also applied to facial processing, which is another previously reported preserved cognitive ability in this clinical group. We wanted to know whether WCC existed among people with WS in their visual perceptions of faces. We compared the behavioral and neurophysiological signals of feature- and configuration-related processing in people with WS using an Event-Related Potentials (ERPs) technique. Taken together with our findings on central coherence at the semantic-conceptual and visual-perceptual levels, we hope to reach a systematic cross-domain understanding of contextual integration in people with WS.

People with WS show an unusual preference for faces, compared to people with ASD and to healthy controls. This preference may be associated with the development of social skills, and related to the hyper-social behavior exhibited by people with WS. This unique preference was demonstrated in a study by Riby and Hancock (2009) that used eye tracking measurements to explore gaze patterns on faces. When viewing background scene pictures with hidden faces or scrambled pictures with embedded faces, the participants with WS fixated significantly longer on hidden faces in background scenes and embedded faces in scrambled pictures compared to people with ASD and healthy controls. In contrast to the performance pattern of people with WS, participants with ASD fixated for a significantly shorter period than the other groups. Moreover, people with WS had normal-like performance when asked to recognize holistic faces in the part-whole matching paradigm (Tager-Flusberg, Plesa-Skwerer, Faja, & Joseph, 2003). In the same study, people with WS achieved normal-like percentages when the matching faces were displayed holistically and partially, including upright and inverted orientations. Like the healthy controls, the WS participants showed a whole face advantage for upright faces and an inversion effect when the faces were presented upside down. However, these findings regarding the preference for looking at faces and the normal holistic recognition of faces do not directly address the issue of configural difficulty in face processing among people with WS.

Configural processing deficit in people with WS was observed in studies using same-different judgment (Karmiloff-Smith et al., 2004), facial expression recognition (Gagliardi, Frigerio, Burt, Cazzaniga, Perrett, & Borgatti, 2003), and in studies investigating the inversion effect (Rose, Lincoln, Lai, Ene, Searcy, & Bellugi, 2007). Configural information (second-order processing) refers to eyes-distance, nose-mouth distance, and any change other than changes in facial features. In a study by Karmiloff-Smith et al., three experiments were conducted to determine whether people with WS differed from healthy controls in

processing faces with changes in features or configurations. In the first study, participants were presented with two consecutive faces, a target face and then an experimental face that was either the same as the target face or altered in features or configurations. They were asked to make a same or different judgment between two faces. Unlike the healthy controls, who showed an expertise in configural processing, the participants with WS were slower and less accurate in their responses to configure-changed faces. In the second study, aimed at investigating WS participants' ability to identify upright and inverted faces using a storybook, the clinical group did not show an increased accuracy or decreased response latency to upright faces or a progressive sensitivity to the inversion effect. This configural detection impairment was also observed in studies that used schematic faces, composed of geometric shapes. Furthermore, with regards to task-specific developmental trajectories, although the typically-developing controls improved their upright face recognition performance with age and showed a greater influence of inversing faces over time, people with WS failed to show such improvement or influence. Overall, previous studies of people with WS have shown they are deficient in the configural detection of face processing.

Recognition of emotional expressions is another method used to explore the ability of people with WS to detect configural alterations in faces, because facial expressions cause dynamic configural changes. Gagliardi et al. (2003) presented animated facial expressions that vividly mimicked human emotions (anger, disgust, happiness, sadness, fear) to individuals with WS. The results revealed that the WS participants performed as well as Mental-Age (MA) matched healthy children, but were inferior to the Chronological-Age (CA) matched healthy adults in their sensitivities to different emotional expressions. It was concluded that the limited ability of the individuals with WS to recognize facial expressions was due to a deficiency in coding configural information. The study identified a further correlation of recognition performance with the WS participants' IQs, but not with their age or Benton recognition test scores. These correlations suggested that, unlike typically developing controls, people with WS applied a deviant strategy in processing faces with spacing changes and their proficiency did not increase with age. Another face matching study by Deruelle, Mancini, Livet, Cassè-Perrot and de Schonen(1999) confirmed that people with WS had atypical recognition of expressions. The participants were presented with three photographs of faces; the face at the top of the screen was the target face and the two at the bottom were probes, shown simultaneously on the screen. The participants were asked to choose which of the probes matched the target face based on the conditions of identity, emotional expression, eye gaze, age, gender, and lip reading. The results revealed that people with WS were significantly less accurate than the CA-matches in all of the matching conditions except lip reading. Unlike the healthy controls, who had significant correlations between matching results and age in all of the conditions, people with WS only had significant correlation between age and the lip reading condition. It was concluded that the non-significant difference in matching lips between the two groups meant that people with WS applied a featural strategy to process faces. They

paid attention to local elements rather than to global configurations. People with WS are deviant in face processing.

The 'seeing the trees but not the forest' phenotype of people with WS was further demonstrated in the study by Deruelle et al. (1999) in which they measured the influence of the inversion effect, an index for the configural processing of faces. It has been demonstrated that the configural processing advantage decreases when faces are perceived upside down. Instead, the featural processing strategy takes over (Maurer, Grand, & Mondloch, 2002). Although the CA-matched and MA-matched controls showed a significant inversion effect for faces but not houses, the participants with WS failed to show any processing influence for inverted faces. The results suggested that people with WS use a featural mode of face processing related to the lack of the inversion effect typically observed in healthy controls. However, other studies have reported that people with WS performed identically to healthy controls and showed a normal-like inversion effect. Rose et al. (2007) presented upright faces with neutral expressions (upright neutral), upright faces with affective expressions (upright affective), and inverted faces without emotional expressions (inverted neutral) to two clinical groups (people with WS, people with autism) and to healthy controls. All of the participants were asked to make a same or different judgment regarding the facial identity of two consecutively presented faces. Although the results revealed that the WS individuals were less accurate than the healthy controls when identifying the inverted neutral faces, the overall pattern of the WS individuals was the same as the pattern of the healthy controls. Moreover, although the participants with autism showed significantly poorer recognition ability in relation to the upright affective faces, and had the lowest percentage of correct answers among the groups, the participants with WS correctly matched a normal-like percentage of faces carrying expressions. Hence, Rose et al. concluded that people with WS perceived configure-changed faces normally. Whether people with WS experience a configural processing deficit in their facial perceptions remains inconclusive, although more studies support impaired visual construction and an inability to organize parts as a whole.

The relative local preference of people with WS has been observed in face processing. Riby and Doherty-Sneddon (2008) explored the facial recognition ability of people with WS by presenting parts and wholes of unfamiliar faces. The participants had to decide whether two faces were the same or different based on internal features (mouth, eyes, nose), external configurations (without internal parts), or whole faces. No difference was observed between the groups in recognizing the unfamiliar faces that were presented as whole faces. However, group differences emerged when differentiating internal-feature faces and external-configure faces. While all of the healthy controls showed higher recognition rates for faces with external configurations, compared to faces with internal features, the WS participants showed the opposite pattern. When perceiving faces, internal features were more salient to people with WS compared to external configurations. It was concluded that people with WS have different face processing strategies than typically developing controls.

In addition to the behavioral difference, atyp-

ical face processing in people with WS was observed at the brain level. Grice et al. (2001) measured a gamma-band frequency (40 Hz) of around 200 ms in participants who were passively viewing inverted and upright faces displayed on the screen. The results uncovered a deficient stimuli-dependent coherence in the WS individuals. Unlike the healthy controls, who expressed larger gamma bursts when looking at upright faces than inverted faces, the clinical group showed no clear bursting reaction to any type of face stimuli. This finding suggested that people with WS have difficulty binding local features into a gestalt configuration. Another study also demonstrated atypical neural activations of people with WS (Mills, Alvarez, George, Appelbaum, Bellugi, & Neville, 2000). Participants were required to judge whether a target face matched or mismatched a prime face in upright and inverted conditions. People with WS perceived the prime faces differently than the healthy controls. The healthy controls showed larger negative amplitudes around the post-stimulus 100 ms (N100) and reduced activation about 200 ms (N200); however, the participants with WS showed a reversed pattern (small N100 and large N200). This processing difference between groups was also observed in target faces. Although the healthy controls performed the match-mismatch effect (larger responses to mismatched faces vs. matched faces) and identified around 320 ms negativity for upright faces, the WS individuals showed the same negativity to both upright and inverted faces. Together, these results suggested that, behaviorally, people with WS pay attention to local features, but not to global configurations when processing faces; neurophysiologically, WS individuals process faces differently than typically developing controls. Despite these advances in our understanding, questions still remain. No previous studies have investigated the brain modulations of people with WS when they process faces with changes in features or configurations. Therefore, we conducted this study with the hope of expanding the cross-modal understanding of central coherence beyond visuospatial construction in the nonverbal domain in people with WS. This finding further reveals whether the WCC is syndrome-general or syndrome-specific in people with developmental disabilities.

Methods

Participants

Individuals with WS were recruited from the Foundation for Rare Disorders in Taiwan and diagnosed in hospitals as having missing genes on chromosome 7q11.23 (n=13, 2 females/ 11 males, mean CA=18.1 years, SD=4.3, age range=13.0－26.8). All of the participants with WS took either the Wechsler Intelligence Scaling for Children (WISC-IV, Chinese version)(Chen, R. H., & Chen, C. Y., 2007) or the Wechsler Adult Intelligence Scaling test (WAIS-III, Chinese version)(Chen, R. H., & Chen, C. Y., 2002), depending on their age, with 16 years of age as the boundary. The mean MA of the WS participants was 10.8 years (SD=3.3, age range=6.2－18.6). Healthy controls were recruited and matched individually by gender and age (n=13, 2 females/ 11 males, mean CA = 17.8 years, SD=4.3, age range=11.8－25.1). The individually matched typically developing controls did not exceed the age applied to each clinical participant. The mean CA between the WS individuals and the controls was not signifi-

cantly different, $t(12)=1.4$, $p>.05$. Among the people with WS, based on their CA, 7 out of 13 participants were children (<18, mean age=14.9 years, range=13.0－17.7, SD=1.9) and 6 out of 13 were adults (>18, mean age=21.9 years, range= 19.1－26.8, SD=2.9). The number of children (46%) and adults (54%) was quite equal in our clinical group and the same distribution applied to the healthy controls due to individual matching.

We invited clinical individuals who were older than 13 years to participate in this study because they could do the ERP study wearing a cap on their heads and sitting still for at least 30 minutes without much motion. We were fortunate to have all these 13 participants with WS join us, given the extreme difficulty in finding people with rare disorders, especially on a small island such as Taiwan.

The block design test, a subtest of the WISC-IV and WAIS-III used to measure visuospatial ability, showed a difference between the controls (mean score=55) and the participants with WS (mean score=11) ($t(12)=-19.3$, $p<.001$), suggesting distinct visuospatial perception abilities between the two groups. Based on the contrast between the block design testing results for people with WS and the healthy controls, we predicted that people with WS would exhibit a deficit in the global information processing of faces. However, after conducting a behavioral study, it was not necessary to recruit the MA group for this study because our clinical individuals showed a non-significant difference from the CA-matched controls at the behavioral level. Thus, the key lay in the neurological processing observed in the participants with WS and the CA-matched controls. The results of this comparison contributed to the findings regarding brain and behavioral asymmetry in people with WS (see Discussion section). Hence, the recruitment of an MA group was not our concern in this study.

Materials and Design

We followed the study conducted by Mondloch, Grand, and Maurer (2002), which investigated the facial processing of changing features or configurations using a model face. New face images were created for this study; the new faces were appropriate for the Chinese-speaking participants with WS and addressed the concern of own-race effect (Stahl, Wiese, & Schweinberger, 2008; Tanaka, Kiefer, & Bukach, 2004). We were deeply inspired by Mercure, Dick, and Johnson (2008) to investigate ERP modulations of faces with feature or configuration changes in normal people using a same-different judgment task. We combined the paradigm with our own race-face images and applied them to people with WS.

Starting with a base female face (model face) we generated faces with alterations in features (eyes, mouth) or configurations (between-eyes distance, nose-mouth distance). To eliminate unnecessary visual cues, no hair or ears were visible in the face images. All of the face images were in 288 x 355 pixels bitmap format and presented by the psychology software Eprime. Feature-changed faces were created by replacing features with features from four other female faces without changing the configuration of the model face. The distance between features justified the same as the model face (56 mm for eyes and 16 mm for nose-mouth). The four alternatives were chosen from a pool of thirty female faces. The configuration-changed faces were created by lengthening or shortening the distances between unchanged features. For faces with configuration

changes, the features remained unchanged but the spacing between features differed. Justification was revised in two ways: (1) eyes closer or further apart by 6 mm and (2) mouth upward or downward by 3 mm. Hence, there were four combinations in the spacing of changed faces: (1) closer eyes and downward mouth, (2) further separated eyes and downward mouth, (3) closer eyes and upward mouth, and (4) further separated eyes and upward mouth. The distances were determined in accord with many trials designed to make the faces with spacing changes look natural. There were three conditions of face stimuli in the same-different judgment task: (1) model face condition (model face vs. model face), (2) feature-changed condition (model face vs. feature-changed faces), and (3) configure-changed condition (model face vs. configure-changed faces). Two blocks contained 224 trials with random presentation of the stimuli (50% for the model face condition, 25% for the feature-changed condition, and 25% for the configure-changed condition). Sample face images for each type of face stimul are given in Figure 1.

Figure 1 Sample faces used in three conditions

Procedures

A 500 ms fixation was shown on the screen, followed by the experimental faces. The model face was presented prior to one of the target faces, which was from one of three conditions. Each face was presented for 700 ms with an inter-stimulus interval ranging from 200 to 400 ms. Participants passively viewed the images and pressed a button to indicate a same or different judgment. To remind the WS individuals of the meaning of each button, two squares were shown at the bottom of each target face with different colors (green, red) and distinct symbols indexing sameness (two circles) or difference (one circle, one cross). Two stickers with corresponding colors in green or red were stuck to the appropriate keys on the keyboard (D, L) to assist the WS participants. The next trial started when a response was detected. Participants were trained to detect changes in features and configuration using photos shown on the computer screen. They received 16 practice trials before the experiment began. Their electrophysiological responses were recorded while performing the task.

EEG recording and analyses

A Geodesic sensor net with 128 channels referencing to the vertex (Electrical Geodesic Incorporated, Eugene, OR) was used. We analyzed

the data using Net Station 4.2. EEG signals that were filtered at 30 Hz and segmented to the length of 900 ms (pre-stimulus 200 ms－700 ms). Each individual datum was cleaned with artifact detection to remove eye movement, eye blink, and bad channels. A bad channel replacement was applied with a default algorithm. Each condition was averaged across the trials for each participant. The reference to the vertex was re-referenced to the average of all of the channels. The pre-stimulus of 200 ms was set as the baseline correction.

The peak amplitudes of all of the channels for each participant, for faces in each type of condition, were averaged in the occipital-temporal region of both hemispheres (selected channels are shown in Figure 2). A visual inspection of the grand average waveforms in both groups identified five time-windows that were of interest: P1 (82－162 ms), N170 (174－212 ms), P2 (200－250 ms), N2 (234－338 ms), and P3 (386－698 ms). As reported in previous studies of facial processing with similar same-different judgment tasks among healthy controls (Mercure et al., 2008) and in people with WS (Mills et al., 2000), the P3 was large in the vertex area and therefore in this study, different montages were created in the vertex region in both hemispheres, as displayed in Figure 3.

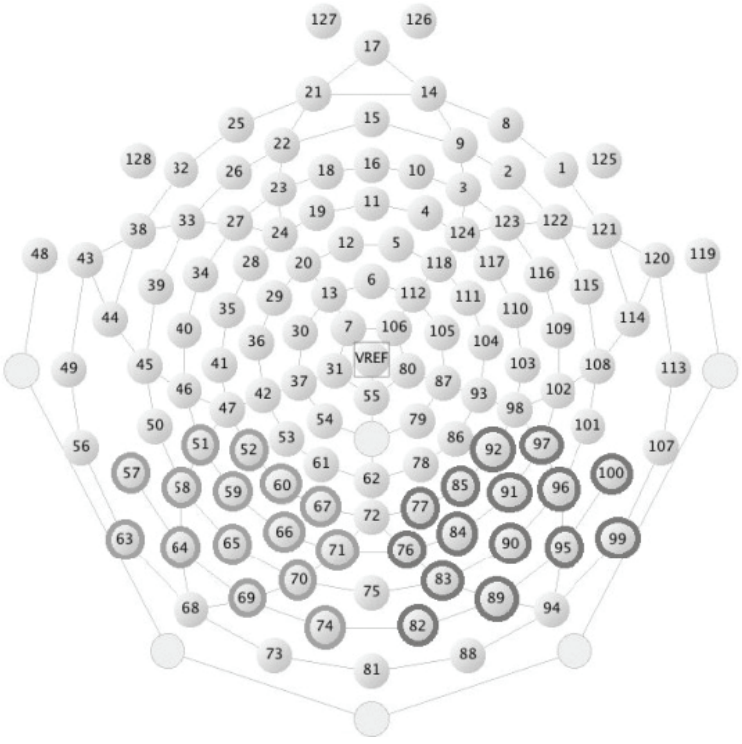

Figure 2 Selected channels in the occipital-temporal region of the brain

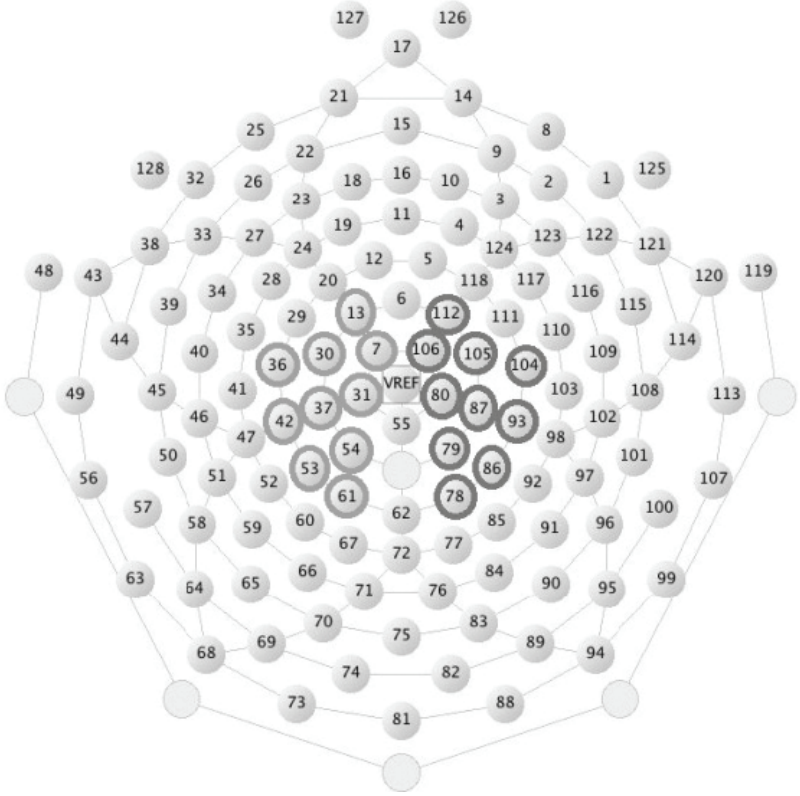

Figure 3 Created montages in the vertex region of the brain

Behavioral Results

Reaction times

Reaction times were calculated based on the correct responses to face stimuli. An interaction of face type (the within-subjects factor) by group (the between-subjects factor) was significant, $F(2, 48)=3.4$, $p=.04$ with the two-way repeated measure ANOVA. People with WS (873 ms) generally responded to faces more slowly than the healthy controls (436 ms) (main effect of group, $F(1, 24)=13.3$, $p<.001$). Faster reaction times for feature-changed faces (460 ms, SE=33) than for configuration-changed (827 ms, SE=105) and model (677 ms, SE=68) faces were observed (main effect of face type, $F(2, 48)=11.2$, $p<.001$). Although the latter two face types were different, the difference did not reach significance. Separate group analyses revealed that like the healthy controls, participants with WS responded fastest to feature-changed faces (hereafter, Features) and slowest to configure-changed faces (hereafter, Configures) as Table 1 shows. The differences between Features vs. Configures and Features vs. Model faces reached significance in both groups,

but not the difference between Configures vs. Model faces (although Configures elicited a slower response than Model faces).

Table 1 Response latency (ms) to three types of face stimuli in people with WS and controls

	Model Faces	Feature-Changed Faces	Configure-Changed Faces	*p* value
Controls	444 (41.9)	350 (29.4)	514 (73.3)	6.0, $p<.008$
WS	910 (472)	571 (215)	1139 (709)	7.5, $p<.003$

Note: Standard errors were in parentheses.
$F=(2, 24)$

The group difference in response to each type of face stimuli was significant (Model faces, $F(1, 24)=11.5$, $p=.002$; Features, $F(1, 24)=11.0$, $p=.003$; Configures, $F(1, 24)=8.9$, $p=.007$). The results indicate that although they were generally slower to respond, the WS participants showed the same pattern in detecting faces as the controls (see Figure 4).

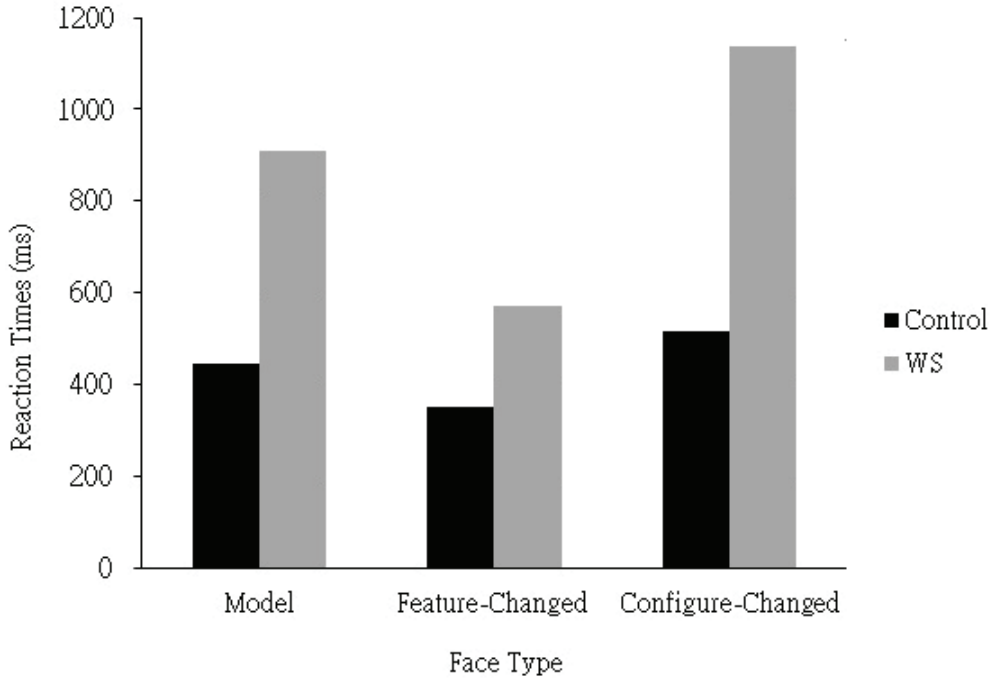

Figure 4 Response latency to the three types of face stimuli for controls and WS individuals

Accuracy

No interaction of group by face type was observed (F<1). The main effect of face types was significant, F(2, 48)=13.2, p<.001. A higher percentage of accurate responses to Features (.95, SE=.02) than to Configures (.64, SE=.06) and Model faces (.74, SE=.04) was observed. The latter two types of faces did not reach a significant difference. The individuals with WS were less accurate (.70, SE=.03) than the controls (.85, SE=.03) in responding to faces as the main group effect was significant, F(2, 24)=12.7, p=.002. Although the clinical group was less accurate in responding to all of the face stimuli types, the response patterns were similar to those of the healthy controls. The correct percentage of each face type in the two groups is listed in Table 2 and depicted in graph form in Figure 5.

Table 2 Accurate percentage (standard errors) to three types of face stimuli in people with WS and controls

	Model Faces	Feature-Changed Faces	Configure-Changed Faces
Controls	.85 (SE=.03)	.98 (SE=.008)	.71 (SE=.07)
WS	.63 (SE=.07)	.92 (SE=.035)	.56 (SE=.08)

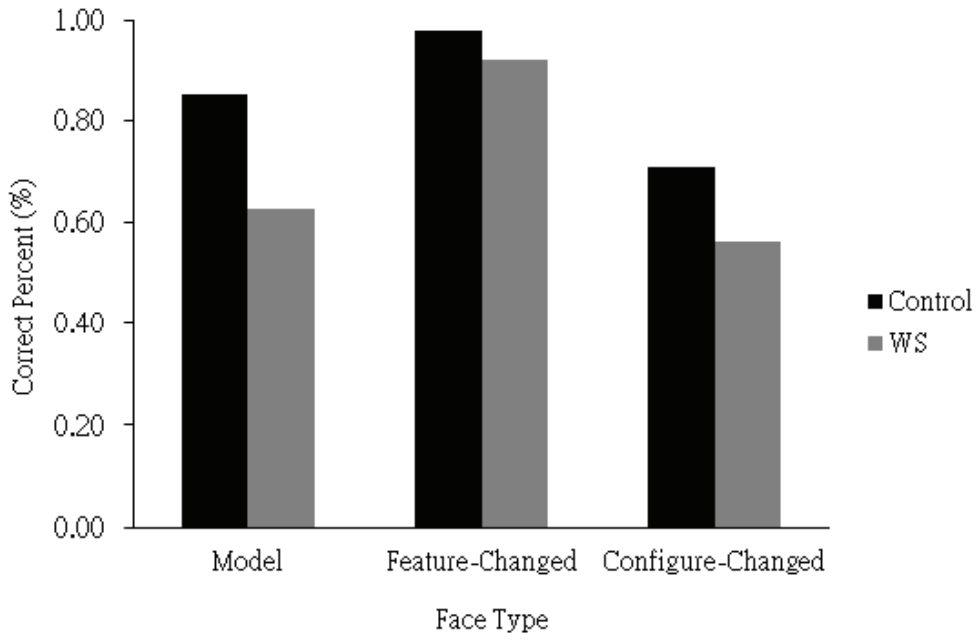

Figure 5 Correct percentages for the three types of face stimuli among controls and WS individuals

Electrophysiological Results

Two-way ANOVAs with repeated measures on brainwave peak amplitudes evoked by the event of face processing, using the two within-subjects factors of face types (Model faces, Features, and Configures) and hemispheres (left, right) were conducted on each component (P1, N170, P2, N2, P3) for each group. The major finding of hemispheric asymmetry emerged in the P3 interval (368－698 ms) in the vertex area. The statistical results for the occipital-temporal and vertex areas are given in Table 3.

Table 3 Average amplitudes evoked in interested time windows for people with WS and controls

Brain region	Occipital-Temporal					Vertex
Component	P1	N170	P2	N2	P3	P3
Face type	.15	2.85	7.23**	9.47***	.88	12.63***
Face type x group	.07	.002	0.85	.02	.65	.42
Hemisphere	6.32*	6.05*	1.93	3.20	.81	4.20
Hemisphere x group	1.72	.59	.92	1.11	.07	.64
Face type x Hemisphere	1.52	.57	1.06	.46	.93	2.09
Face type x hemisphere x group	4.37*	2.45	2.38	2.61	3.76	6.55**
Group	.09	10.96**	16.21**	14.53**	.07	2.60

Face type, F =(2, 48); Hemisphere, F =(1, 24); Face type x hemisphere, F =(2, 48); Group, F =(1, 24)
* p<.05 , ** p<.01, *** p<.001

Because the interaction of the P3 component in the vertex region reached significance, further analyses found the simple interaction of face type by hemisphere in the healthy control group (F(2, 24)=4.5, p=.02) and the WS group (F(2, 24)=4.2, p=.03). To the healthy controls, the simple main effects were from the differences among types of facial stimuli in the left hemisphere (LH, F(2, 24)=15.8, p<.001) and the right hemisphere (RH, F(2, 24)=23.4, p<.001). In the LH, the post-hoc analyses using the Bonferroni method revealed significant differences in the comparisons of Features (5.7)>Model faces (2.9) at p<.001 and Configures (4.5)>Model faces at p=.009. In participants with WS, no simple main effect was observed in the LH (F(2, 24)=1.9, p>.05), although the trend of difference revealed patterns similar to those exhibited by healthy controls (Features 4.4>Configures 3.4>Model faces 2.6). The graphs of the microvolts of each type of facial stimuli among the healthy controls and the WS group are shown in Figure 6. Taken together, in the LH the major finding between the two groups lay in the differences in processing unchanged faces (Model faces) and changed faces (Features, Configures). That is, while the healthy controls showed different responses to these two types of faces, people with WS failed to notice a difference.

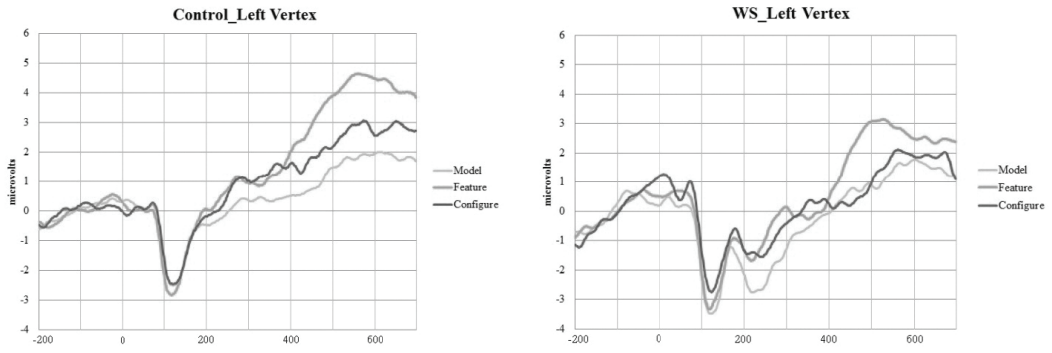

Figure 6 Amplitudes of the three types of face stimuli in the left vertex region among controls and WS participants

In the RH, followed by the significant simple interaction, the Bonferroni post-hoc comparisons among the healthy controls revealed response differences between Features (7.2) and Configures (4.8) and Features and Model faces (4.2) at $p<.001$. The amplitudes of Configures and Model faces did not differ from each other. In participants with WS, a simple main effect was observed among face stimuli in the RH ($F(2, 24)=3.8$, $p=.04$). The post-hoc comparisons using the Bonferroni method revealed a significant difference between Features (4.4) and Model faces (2.6) at $p=.001$. The differences were not significant between Features and Configures (3.4) or between Configures and Model faces, although there were differences between each comparison. The graph relating microvolts to each type of facial stimuli in each group is shown in Figure 7. In the RH, feature- and configure-changed faces prompted facial processing differences among the healthy controls, whereas participants with WS failed to exhibit such processing differences.

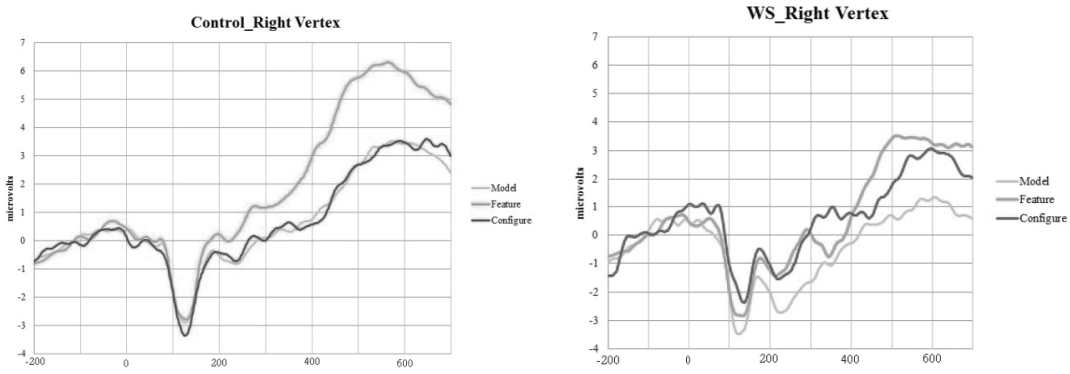

Figure 7 Amplitudes of the three types of face stimuli in the right vertex region of controls and WS participants

The simple interaction of face type by hemisphere with the P1 component in the occipital-temporal region was observed in the WS group. However, the interaction violated the sphericity hypothesis, thus a Greenhouse-Geisser correction was applied, $F(1.4, 32.5)=3.7$, $p=.05$. Post-hoc comparisons revealed no significant differences among face types in either hemisphere, and no hemispheric difference in any face type. No other interactions were noted in the healthy controls regarding this component. The main effect of hemisphere was larger responses in the RH (7.4) than those in the LH (6.5). This hemispheric difference in processing faces was extended to the N170 component (RH: 4.9, LH: 3.9), but disappeared within the P2 time window. Instead, the main effect of face type emerged. Smaller microvolts to Features than to Model faces were observed in P2 (3.9 vs. 4.9, respectively) and N2 (3.4 vs. 4.8, respectively). No group difference in facial type recognition was observed in these two components due to the non-significant interaction of face type and group. However, regarding the vertex P3 component, we observed larger responses to faces with changes in features (5.6) than to model faces (3.0). Consistently larger (more positive) voltages in the facial processing of people with WS, compared with those among the healthy controls were observed for N170 (WS: 5.8, Controls: 3.0), P2 (WS: 6.1, Controls: 2.9), and N2 (WS: 5.7, Controls: 2.7) components.The graph relating microvolts to each type of facial stimuli in each group is shown in Figure 8.

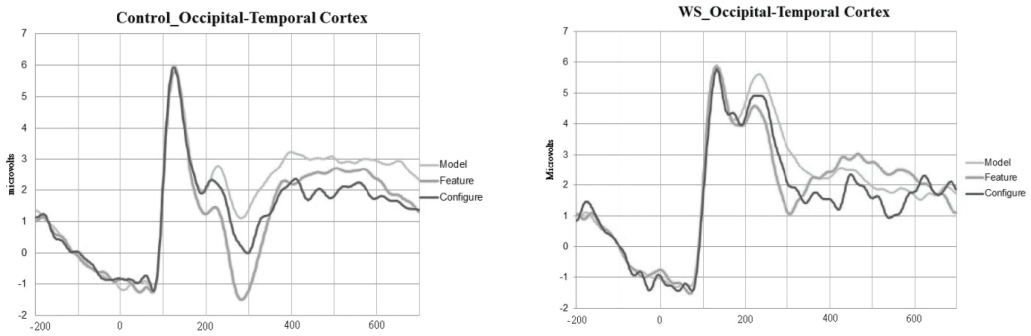

Figure 8 Amplitudes of the three types of face stimuli in the occipital-temporal region among controls and WS participants

In summary, the hemispheric asymmetry in the vertex between healthy controls and people with WS revealed an atypical processing of faces. The clinical individuals responded similarly to (but non-significantly different between) changed and unchanged faces in the LH, but they failed to detect differences between faces with changes in spacing and faces with changes in features in the RH. Hence, deviant neural correlates to processing feature- and configure-changed faces were observed in people with WS.

Discussion

This study aimed to identify the brain response patterns of people with WS to faces with

changes in features or configurations. After being presented with a model face, participants were required to make same or different judgments on target faces that were either altered from or the same as the model face. While they performed the task, participants' brain responses were recorded. Given the local processing bias experienced by people with WS, we predicted that the clinical individuals would fail to show the same process ing patterns when viewing configure-changed faces as the healthy controls at both the behavioral and neurological level. This prediction was partially confirmed. The results reveal an asymmetry in the behavioral and brain levels. The WS group showed distinct brainwave responses compared to the healthy controls. In the behavioral findings, the response of the WS participants was similar to the healthy controls; among all of the face stimuli types, the longest response latency and the lowest accuracy occurred in the response to configure-changed faces. The participants in both groups detected changes in features faster and more accurately than configure changes. However, the neurophysiological findings revealed significantly distinct brain activations in the 368 ms to 698 ms range between the two groups when they processed faces. The healthy controls processed configure-changed faces differently than feature-changed faces in both hemispheres, whereas the WS individuals did not show any difference between the two types of face stimuli. The results suggest that people with WS failed to distinguish between faces with configuration alterations and those with feature changes. Hence, deviant neuronal responses to configure-changed faces were identified in people with WS.

Our neurological findings of deficient configuration processing in people with WS are consistent with previous visuospatial construction tests, such as the block design task. People with WS performed worse on this task, suggesting impaired configuration arrangement. A study by Bellugi et al. (2000) reported that people with WS had difficulty in arranging blocks as an organized whole. The same difficulty was observed when individuals with WS were asked to copy a picture of an object. Instead of drawing a complete object, they drew parts of it, without integrating them into a coherent configuration. Similar observations were made when they were asked to copy a picture of a bicycle or a house. The local preference of people with WS was also observed in the Navon (1977) paradigm, a test of perceiving local or global information. When asked to draw a large H-shape composed of many small s-shapes, the WS participants wrote many s-shapes but ignored the global shape (Bihrle et al., 1989). This global processing impairment in people with WS has been observed in other nonverbal domains, such as music perception (Deruelle, Schön, Rondan, & Mancini, 2005). In the study by Deruelle et al., the control participants detected contour-violated melodies (global-changed condition) more accurately than interval-violated melodies (local-changed condition), whereas the WS participants detected both contour- and interval-violated conditions equally well. Deruelle and colleagues concluded that unlike controls, people with WS lack global precedence before local focus. Thus, a local bias and global deficit processing are perceptual characteristics of people with WS. In addition to behavioral findings, in this study we identified the deviance at the neurological level.

Brain and behavioral asymmetry seems to be

a syndrome-general phenotype, as demonstrated in people with Prader-Willi Syndrome (PWS) (Halit, Grice, Bolton, & Johnson, 2008). Despite the similar normal-like behavioral performances of the two subtypes of people with PWS (origin nated paternally or maternally), the two groups' brain responses to faces were differentiated by different orientations and gaze directions. The paternally originated PWS participants showed no difference between face-types stimuli, whereas the participants with maternally originated PWS yielded larger amplitudes to inverted faces, with averted gazes on the N170 component. Together, these results have highlighted the importance of neurophysiological exploration beyond behavioral performance in developmental disorders. It is essential to determine the different neural mechanisms that underlie cognitive tasks, which may generate similar behavioral performances in people with developmental disabilities as typically developing controls. This asymmetry has been consistently observed in the verbal domain in people with WS. In a study by Hsu et al. (2007) on semantic concept formation using ERPs, the participants with WS processed semantically related words differently than the healthy controls. Unlike the healthy controls successfully integrated semantically associated words into gist themes and processed the themes as in the previously presented words, the WS participants processed gist themes as semantically unrelated words, which were not presented to them before recognition. In contrast, the behavioral performance of the WS participants showed concept formation ability as the healthy control through a high recognition rate of non-presented semantically related associates. Hence, the asymmetry between the brain and behavioral levels in people with WS has been demonstrated in processing both verbal and non-verbal stimuli.

The N170 component did not specifically reflect featural or configural changes in either typically developing controls or the WS participants. Our results are compatible with the findings of Mercure et al. (2008), who explored the neural correlates of the modulation of features and second-order configurations on healthy controls. As our results showed, no main effect or interaction was observed influencing the N170 amplitudes. The results of this study and the study by Mercure et al. do not reveal any sensitivity to the detection of face changes in features or configurations on this component. They suggested that this was the result of the prototypical face effect. Thus, no sensitivity was obtained on the N170.

The deficient configuration detection in the face processing of people with WS identified in this study is consistent with the WCC (Frith, 1989). Previous studies have used WCC to explain the cognitive phenotype of people with ASD, such as their superior performance in the block design test (Shah & Frith, 1993) and their impaired contextual integration to access correct meanings of homographs (Happé, 1997). We propose that people with WS are as deficient in central coherence as people with ASD on verbal and nonverbal domains. A recent behavioral study revealed deficient proposition integration in people with WS, indicating atypical cohesiveness in semantic comprehension (Hsu & Tzeng, 2011). These studies have demonstrated that both the visuospatial domain and the semantic-conceptual domain are weak in central coherence in people with WS.

Recent studies have revealed another WCC pattern exhibited by people with WS; that is, delayed performance. A study in the nonverbal domain using pictures revealed delayed but not deviant central coherence ability in people with WS (Hsu, 2013a). Unlike previous studies, which have suggested that people with WS have difficulty with visual-spatial construction, given more concrete stimuli such as pictures, people with WS had the same pattern of central coherence as healthy controls. Participants were presented with a background setting (e.g., a swimming pool) and a congruent object (e.g., goggles) or an incongruent object (e.g., a skateboard). Response latency and accuracy were measured as dependent variables. People with WS showed the congruency effect in responding to pictures eliciting semantic integration. They performed similarly to the MA-matched controls, although the clinical group had longer response latencies and higher error rates. People with WS used social schema of event knowledge to respond to pictures as the controls. Hence, we concluded that concreteness of stimuli facilitates the contextual integration exercised by people with WS as in the study on people with ASD (López & Leekam, 2003). A follow-up study using the cross-modal presentation of visual backgrounds and auditory targets to test the contextual integration exercised by people with WS, the results confirmed the semantic priming effect as the unimodal study (Hsu, 2013c). People with WS showed non-significantly different response latency and accuracy than the healthy MA-matched controls, but delayed performance compared to the healthy CA matches. Due to the less concrete nature of auditory stimuli, we concluded that social relatedness rather than concreteness of stimuli per se influenced the contextual integration exercised by people with WS. To test the validity of our conclusion, other cross-modal presentation patterns (e.g., auditory backgrounds vs. visual targets, auditory backgrounds vs. auditory targets) were included as other projects in our laboratory. The results replicated our conclusion. Further finding of modality effect was observed, showing faster responses to visual targets compared to auditory ones. Hence it is suggested that cross-modal learning can benefit more on people with developmental disabilities comparing to unimodal studing for future intervention.

In addition to the nonverbal domain, another study pursuing the central coherence abilities of people with WS in the verbal domain has confirmed the delayed pattern of WCC (Hsu, 2013b). The participants were presented with short scenarios describing causes followed by consequences (backward causal inferences) or consequences followed by causes (forward causal inferences). After listening to the scenarios, the participants were asked to choose the correct answer from three alternatives related to the key homonym (the same spelling with different meanings) embedded in each scenario. For instance, participants heard a narration about Er3 Bian1 Feng1 (耳邊風, literally means *ear*, *side*, *wind*) in the following scenario (original texts were presented in Chinese and translated into English): *Daxung failed the exam this time. His mother often reminded him to study hard, but Daxung was inattentive to his mother's reminders* [cause]) and *Daxung regretted not paying attention to his mother's reminders* [consequence]). Later, the participants were asked, *What did Daxung do?* and required to select an answer based on their contextual interpretation of the

figurative meaning (*Daxung did not take his mother's words seriously*), the literal meaning (*There was a wind blowing by Daxung's ears*), and an unrelated meaning (*Daxung's ears were itchy because of the blowing wind*). Both the literal and unrelated meanings shared the same two syllables of the homonym (e.g., Er3 [ear] and Feng1 [wind]), but not the figurative meaning. The results showed that participants with WS chose the significantly lowest percentage of figurative meanings among groups, suggesting delayed causal inference ability in contextual inte gration. Moreover, the clinical individuals showed a significantly higher percentage of unrelated and literal meanings as correct answers compared with the healthy age-matched controls, implying a deviant comprehension in the contextual integration of central coherence. Taken together, people with WS were found to pay more attention to features during contextual integration. Hence, it is certain they were delayed in the verbal and nonverbal domains of WCC. As this study on facial processing revealed a neurophysiological failure to differentiate between feature- and configure-changed faces among people with WS, we are highly interested in examining whether there is a deviant neurophysiological mechanism behind the superficial delayed behavior in a future study.

Understanding development is the key to understanding developmental disorders (Karmiloff-Smith, 1998, 2007). A tiny mutation in a gene has a huge influence on later developments. Our current findings suggest that explanations based on adult neuropsychology are not accurate for people with genetic deficits. With age, deviant neurophysiological mechanisms may produce normal-like (or delayed) behavioral outputs. Hence, we have to be careful in explaining any results obtained for people with developmental disabilities. This study contributes to the understanding of the role of brain functions in differentiating the configural detection of face processing in people with WS. Likewise, it provides support for the argument that there is syndrome-general weak central coherence in people with developmental disabilities, although our wide definition of weak central coherence includes delayed and atypical performances. As mentioned in the introduction, a cross-domain understanding at the behavioral and neurological levels regarding central coherence in people with WS has been gradually achieved. This study recruited thirteen participants with WS to do the configuration detection task. Although people with WS are rare disorders in Taiwan, we still hope to invite more participants to join our study in the future. Moreover, mental age matched controls can be recruited as another control group in the future study to observe developmental changes in face processing. With hopes, we can move forward and make improvements.

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Deviant Neural Correlates of Configural Detection in the Facial Processing of People with Williams Syndrome

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ABSTRACT

Purpose: This study investigated the facial processing strategies of Williams syndrome (WS) patients, who exhibit genetic deficits on chromosome 7q11.23. Because of this deficit, this clinical group has been unsuccessful in detecting configural or global information in previous behavioral studies such as the standardized block design test. However, no neurophysiological evidence has been reported regarding this impairment; thus, the event-related potentials (ERPs) technique was used to address this deficit. **Methods:** Female faces were manipulated (changing the features or configurations) as facial stimuli. The images used to change the features (the eyes or mouth) or configurations were based on other female faces. WS patients (n=13) and their chronological-age matched controls (n=13) participated in this study. The participants assessed the similarities or differences among consecutively presented faces from a set of models, some of which had altered features (feature-changed faces) or configurations (configuration-changed faces). The faces were randomly presented and no duplicates were displayed. **Findings:** Regarding response latencies and accuracy rates, the behavioral results of WS patients were similar to those of the healthy controls. Both groups demonstrated rapid detection and high accuracy rates when assessing the feature-changed faces, but responded slowly and erred considerably when assessing the configuration-changed faces. However, the groups presented distinct brainwave responses to the configuration-changed faces. The healthy controls processed the configuration-changed faces differently compared with the feature-changed faces in the vertex areas of both hemispheres, whereas the clinical group failed to differentiate these 2 types of facial stimuli. **Conclusion:** In this study, we discovered

neural evidence for a configuration detection deficit among WS patients when processing faces. The results further identified a weak central coherence among WS patients, suggesting a syndrome-general but not syndrome-specific deficit in people with developmental disabilities. **Implications:** WS patients demonstrated asymmetric brain and behavioral performances during facial processing. This asymmetry was reported in a verbal study that used ERPs and a false memory paradigm. WS patients exhibit genetic deficits that cause atypical development during the early stages of life. These findings were consistent with those of our previous studies pursuing contextual competence, which is defined as the ability to integrate the meanings of words into a contextual theme by using appropriate social knowledge and semantic comprehension; this has been considered a major deficiency among those with autism or right-hemisphere brain damage. Our findings confirmed a deviant central coherence among this clinical group. Neuroconstructivists claim that a small gene mutation during the initial developmental stages can yield devastating effects in long-term development. The deficient configuration detection performance of the WS group provides evidence supporting central coherence deficiency, proving that the interaction between genes and cognition is a dynamic process.

Keywords: weak central coherence,Williams syndrome (WS), event-related potentials (ERPs) , configural detection deficit, facial processing

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