Semantic Knowledge in Williams Syndrome: Insights from Comparing Behavioural and Brain Processes in False Memory Tasks

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Abstract--- This study attempts to understand the relationship between use of context and semantic knowledge in the genetic disorder, Williams syndrome (WS). Earlier work had arrived at discrepant results, suggesting either normal semantic priming [10], or unusual lexical organization [5] and atypical sentence integration [8] in this clinical group. To address these discrepant findings, we used two methodologies with an auditory false memory paradigm, and measured functional and neurophysiological (ERP) responses from three groups: children and adults with WS, Mental-Age matched normal children, and normal adults. While the behavioural data suggested that individuals with WS revealed a similar pattern of recognition as both groups of controls for words with semantic relatedness, their neurophysiological correlates suggested a different pattern. Our findings indicate that WS proficient compensatory behaviour camouflages a deviant neural pathway in the use of contextual cues. Our results also point to neurological changes during typical development, since typically developing children showed a distinctive pattern from our adult participants. Overall, our findings suggest that semantic organization develops slowly over typical development, and atypically in the Williams syndrome.

Index Terms--- Williams syndrome, semantic knowledge, context use, false memory, event-related potentials

I. INTRODUCTION

In previous research on individuals with Williams syndrome (WS), it was shown that they have deficits in semantic knowledge [1][5][8][13]. In contrast to these findings, the results of a subsequent study [10] pointed to normal semantic priming, similar to that found in control groups. Because of these discrepancies in the literature, we selected a false memory paradigm with auditory presentation because it is believed to correlate with semantic relatedness and other non-semantic associations [2].

Based on our knowledge of the syndrome and the tasks chosen, two hypotheses were formulated: 1) if people with WS show no difference in rates of illusory recognition memory compared to normal controls, we would conclude that they have normal abilities in using contextual cues, implying typical semantic organization; and 2) if people with WS, like those with autism, show decreased illusory recognition rates compared to controls, we would conclude that they have atypical semantic organization. Furthermore, we planned a second experiment if the results from behavioural measures support the first hypothesis. In the second experiment, we would go on to explore the neural correlates of their successful behaviour, using event-related potentials.

II. BEHAVIOURAL METHODS

A. Participants

Three groups were recruited: 10 adolescents and adults with WS [2 females/8 males, mean Chronological Age (CA)=15.4 years (SD=3.4, range=9.8-29.7) and mean Mental Age (MA)=8.7 years (SD=2.6, range=6.6-14.9)], 10 Verbal Mental-Age matched children on WSIC-III [1 female/9 males, mean CA=8.0 years (SD=3.0, range=5.5-12.5), mean MA=9.0 (SD=3.3, range =5.9-15.9)] and 5 adults (4 female/1 male, CA=27.2 years, SD=2.9, range=25-32). For participants with WS, although their chronological age (CA) was quite broad, these were the totality of adolescents and adults that we could find on the island of Taiwan, which does not lend itself to further subgrouping in terms of chronological age.

B. Design

Eight word-lists of 10 Chinese words each were presented
orally during the study phase. All words were semantically related. In the test phase, nine words were tested for each word list: three semantically-related non-presented words as lures, three previously-presented words as old items, and three semantically-unrelated non-presented words as new items. All the words were bisyllabic and were recorded by a female voice using a speech production apparatus Praat (8 bit, sampling frequency=44.1KHz).

C. Procedure
In the study phase, a fixation point was displayed on a computer screen for 500ms, followed by an auditory presentation of a target via headphones. Another 500ms interval was added before each new trial. In the test phase, a fixation point (500ms) was followed by a target word orally. Participants had to pay attention to the spoken voice and make a yes/no judgment as to whether the word had been heard before by clicking the mouse. Response latency and yes/no judgment percentages were dependent variables [for full details of the procedure, see Hsu & Karmiloff-Smith et al., submitted].

D. Results
A comparison of the proportion of yes-responses as a within factor [previously presented words (Hits), new words with semantic relatedness (lure false alarm, hereafter Lure-FA) and new words without semantic relatedness (new false alarm, hereafter New-FA)] yielded an interaction with group as a between factor [two-way ANOVA, F(2,9,32)=3.84, P<.03]. The simple main effect was from the proportional difference of Hits between adult controls (.75) and the clinical group (.50). No difference was found between MA-matched children and individuals with WS. A significant main effect was found among yes-response categories [F(1,5,32)=96.11, P<.01]. A Tukey post hoc test revealed that the proportion for yes-responses was significantly greater for Hits (0.61) than for Lure-FA (0.33), which in turn was greater than for New-FA (0.13). The clinical group showed a similar pattern to normal controls with respect to semantic illusion. No interaction emerged for reaction times of yes-response categories and groups. The main effect of categories was significant [F(2,28)=7.01, P<.004], showing that response latency for Hits (1250ms) was faster than for Lure-FA (1509ms, P<.05) and for New-FA (1704ms, P<.01). The latter two categories were different from one another [for full details of the results, see Hsu et al., submitted].

III. NEUROLOGICAL METHODS
A. Participants
Seven of the behaviourally-tested individuals with WS participated in this study (7 males, mean CA=17.8 years, SD=3.6, range=12.3-21.1, mean MA=9.8 years, SD=3.2, range=5.0-14.9) as well as 7 MA-matched children based on verbal IQ on WSIC-III (7 males, mean CA=9.6 years, SD=1.3, range=8.4-11.6, mean MA=10.8 years, SD=1.8, range=8.2-12.9). An additional 17 young adults from National Yang Ming University were recruited as a normal adult control group (CA=20.5, SD=1.2, range=18-23).

B. Design
Sixteen word lists, consisting of 13 bisyllabic Chinese words per list, were presented via headphones in two blocks (8 lists per block) to participants during the study phase. A recognition test followed after the presentation of each block. Three studied words were selected, mixed with 6 semantically-related words as lures and 3 semantically-unrelated words as new items per list. Stimuli were recorded by a female voice in Praat software (8 bit, 44.1KHz).

C. Procedure
ERPs were recorded during both phases. In the study phase, a fixation point (500ms) was displayed on the computer screen to keep participants’ eyes stationary in order to reduce eye movement artifacts while EEG data were being recorded. After 500ms, a word (average=806ms, SD=90ms) was presented orally to the participant. In the recognition phase, an auditory bisyllabic word was presented (average=807ms, SD=92ms). Participants had to decide if this word had been heard before during the study phase by clicking the mouse connected to the computer to indicate yes/no judgment. After judgment, a symbol “@” was displayed on the screen for 500ms. The inter-stimulus interval was 500ms. The entire duration was 2.80 minutes for the study phase (1.40 minutes for each block) and 5.78 minutes for the test phase (2.89 minutes for each block). The averages included only trials to which the participant responded correctly and that were free from artifacts.

D. Electrophysiological recording
EEG was recorded continually from 32 sintered Ag/AgCl electrodes mounted on an electrode, with bilateral mastoid as the reference [for full details, see Hsu et al., submitted].

E. Results
1) Behavioural Findings
An interaction between groups and response categories emerged in the proportion of yes-responses [two-way ANOVA, F(2.5,32.8)=6.78, P<.003]. For the difference of proportion, adult controls (.76) showed higher recognition rates to previously presented words (P<.01) than the clinical group (.43). A higher proportion of overall response categories for adults (.46) and MA-matched children (.43) compared to people with WS (.29) was observed [main effect of group, F(2.26)=5.09, P<.02], revealing a difference between normal controls and people with WS (P<.005 for adults, P<.05 for MA). The proportion of Hits (.60) was higher than the one for Lure-FA (.36), which was in turn higher than the one for New-FA (.21) [main effect of response category, F(1.3,32.8)=75.38, P<.01]. The clinical group, like normal controls, showed different recognition
proportions among yes-response categories [one-way ANOVAs: adults, \( F(1.4,20.6)=173.95, \ P<.01 \); MA, \( F(1.0,5.2)=6.32, P=.051 \); WS, \( F(2,12)=11.05, P<.003 \)].

No interaction was found for yes-response categories and groups in response latency. The main effect of response categories showed that the response latency for Hits (1193ms) was faster than other categories (1311ms for Lure-FA, 1366ms for New-FA), \[F(1.6,40.3)=6.3, P<.008\]. The latter two response categories were not different from each other. While the response latency for MA-matched children (925ms) was faster than adult controls (1317ms, \( P<.02 \)) and the clinical group (1628ms, \( P<.002 \)), the difference between the latter two groups was marginal (\( P=.05 \)) [main effect of group, \( F(2,25)=7.49, P<.004 \)].

2) Electrophysiological findings

Figure 1 displays the grand average for yes-response in three categories (hit for old items, correct rejection for new items, and false alarm for lure items) of the participating groups at the Pz site (for adults and clinical group) and Cz site (for mental-age matched children). The included trial numbers for Hit, New Correct Rejection, Lure False Alarm for adults were 34, 36, 18 and for WS were 20, 39, 12 and for MA were 21, 26, 12.

The temporal window for the ERP component of interest was defined as recollection retrieval (700-1300ms).

The dependent variable was the mean amplitude of the time window from each electrode in comparison with three response categories: Hits, Lure-FA and New-CR (correct responses toward new words without semantic relatedness). One-way ANOVAs with within-subject factor of response categories on each electrode found that the left parietal electrode site (P3) and the central parietal electrode site (Pz) for separate groups were significant by applying Greenhouse-Gessier correction for non-sphericity where necessary in all ANOVAs. Since the pattern found in these two electrode sites was the same, the results of Pz are reported here. For MA-matched children, the results were taken from an alternative electrode site (Cz), since no effect was found on parietal electrode sites [further details of the ERP results can be found in Hsu et al., submitted].

Pz: The results showed that adult controls had clearly differentiated brainwaves among response categories [main effect, \( F(2,32)=5.22, P<.02 \)]; the same held for our clinical group [main effect, \( F(2,12)=10.29, P<.003 \)]. However, post-hoc tests with Tukey revealed different patterns. For the adult controls, the mean amplitude of Hits brainwave (-2.49) was no different from the amplitude of Lure-FA brainwave (-2.40). By contrast, the clinical group showed a difference between them (4.09 for Hits, -0.56 for Lure-FA, \( P<.02 \)). In addition, while a difference between Lure-FA and New-CR (-4.14) was found for adult controls (\( P<.03 \)), no difference was found between them for people with WS (-1.22 for New-CR, \( P=.61 \)). Both adults and the clinical group showed the difference between the brainwaves of Hits and of New-CR (\( P<.008 \) for adults, \( P<.009 \) for people with WS).

Cz: A main effect yielded significance for MA-matched children [\( F(2,12)=9.76, P<.004 \)]. The difference was found between brainwaves of Hits (-2.07) and of Lure-FA (-5.22) (\( P<.002 \)) and also the difference between brainwaves of Hits and of New-CR (-4.95) (\( P<.02 \)). This pattern suggests that MA-matched controls have a partially similar pattern of neural correlates to people with WS, but in distinct brain loci.

IV. SUMMARY OF FINDINGS

Behaviourally, all three groups made a distinction between previously studied words and semantically-unrelated new
words. Adult controls could also differentiate semantically-related lure words from semantically-unrelated new words, and this pattern was also found in the clinical group. Our neurophysiological results, however, seem to point to a somewhat different picture. Adult controls showed a difference in brainwaves between misrecognition of semantically-related lure words and semantically-unrelated new words, whereas no such difference emerged in the clinic group. In other words, the brains of the adult controls treated semantically-related lure words in the same way as previously-presented words in the study phase, while the brains of individuals with WS treated all semantically-related words in the same way as semantically-unrelated words. In addition, while adult controls showed no difference in brainwaves between previously-presented words and semantically-related lure words, the brains of people with WS marked such a difference.

Interestingly, the behavioural results of the MA child controls yielded a distinctive pattern compared to both the adult controls and the clinical group. Besides the similar finding of a difference between previously studied words and semantically-unrelated words, the MA-matched children had higher mean amplitudes in the frontal area over the entire task.

V. CONCLUSION

Our aim in this study was to compare behavioural and electrophysiological measures of semantic abilities in individuals with Williams syndrome and child and adult controls. Our findings of normal behavioural effects, but some abnormal brain processes, may go some way towards elucidating the seemingly conflicting findings between [8] and [10]. Indeed, decreased use of contextual cues reflected in our results points to abnormal semantic processing at the neuronal level rather than better discrimination of presented and non-presented words. Our atypical ERP results in WS add to previous electrophysiological findings on WS face processing [7] and on WS visuo-spatial perception [3]. Furthermore, previous behavioural studies have pointed to less sophisticated semantic organization compared to normal controls such as in generalization fluency of subcategory items within a category [5]. Moreover, very young children with WS have also been found to be atypical in failing to use verbal cues for categorization [9]. These various findings of abnormal semantic organization is likely to influence the capacity of individuals with WS to make use of contextual cues.

It is also important to stress another implication that derives from the present study, i.e., the choice of methodology when attempting to study atypical cognition. When data solely concern behavioural measures, this runs the risk of making erroneous conclusions. In our view, it is only the combination of both behavioural and neurophysiological measures that can elucidate whether overt behaviour is normal or is underpinned by atypical brain processes. We have consistently stressed the importance of trying to trace full developmental trajectories from infancy through to adulthood, for each domain, because starting states of cognition are not necessarily identical to the end states in terms of various cognitive domains [6]. For example, in language, researchers [11][12] found that infants with WS were as seriously delayed as those with Down syndrome, with both disorders being significantly worse than their MA-matched controls. In contrast, by adulthood, individuals with WS outstrip their counterparts with DS. This indicates that the language fluency of adults with WS may be the result of a different developmental trajectory compared to healthy controls and DS.

In the current study, we also showed that although the normal children looked similar to adults behaviourally, their brain signature was different, suggesting a protracted course of normal brain development. Although electrophysiological studies yield stronger temporal than spatial findings, our results do suggest that this gradual change from childhood to adulthood in the way in which the brain processes semantic categories does not occur in the same way in people with Williams syndrome. Further research is needed to settle this question, but our results strongly highlight the necessity to look beyond behavioural similarities to verify underlying neurophysiological processes that underpin such behaviours.

REFERENCES


