

Exploring Semantic Knowledge of Chinese Williams Syndrome: A New Approach from False Memory

The main focus of this study is to explore semantic knowledge in terms of concept formation in people with Williams Syndrome (WS). This research idea comes from the selective impairment in meaning relative to grammatical knowledge in language of Williams Syndrome patients. Two experiments related to false memory studies are provided in this research project.

In Experiment 1, a Deese-Roediger-McDermott (DRM) paradigm (1959, 1995, & 1998) is employed behaviorally and in neural correlates. Subjects are required to listen to many words which are semantically related and later on are asked to recognize them among lumped various words. It is expected that WS patients would show specific item memory effect for old items compared with lure items and new items. However, their gist memory ability would be deficit. In other words, they cannot form a concept automatically from semantically related words (lures) presented as their matched mental age controls and college students. The results of our study in behavioral data show WS patients also perform gist memory as controlled groups, suggesting a preserved semantic knowledge of mental lexicon.

In Experiment 2, an event related brain potential study on false memory was employed. It is interesting to see whether WS patients show the same neural correlates pattern as normal college students. In other words, are WS patients' semantic knowledge really normal? From event-related brain potentials study, the results showed that WS patients treated lures more like new items (i.e. semantically unrelated words) as compared to the ones of college students who treated lures much more like old items (i.e. the actual presented words). This difference

suggests that WS patients may have a distinct semantic network in terms of underlying mechanism operated.

The results of these two studies from behavior to neural correlates suggest that WS patients apply different underlying mechanism on semantic processing. Furthermore, the results also confirm an observation that behavioral performance does not imply normal processing with the same mechanism.

The Implications of This Research Project

There are three major reasons to conduct this research project. First, from literature on WS in the past, there is no study on investigating their semantic comprehension. Almost all research studies focus on their grammatical knowledge rather than semantic understanding. It is well known that WS individuals have preserved sentence structure presentation, but how good is their comprehension? Sporadic studies showed problematic semantic understanding on WS. For example, mapping error (Zukowski, 2001), which place arguments in wrong positions in a sentence, is frequently observed on WS. Second, recently researchers on WS have noticed that WS individuals have quite good verbal working memory ability and show high correlation with verbal learning and structure building (Mervis, 2003), but no one pursuits further like how they can master language so well by taking working memory advantage.

We sincerely hope that this research can be benefit to WS individuals directly and the results can be references of people who may concern this genetic disordered population like pathologists, special educationists, and parents. If they do have difficulty in understanding meaning, pathologists/teachers/parents should pay more attention on

explaining words or instructions clearer. Furthermore, we hope these research results can be useful to inventing learning instruments for WS individuals. We wish them to have a better life because of our research in the near future.

Methodology

Exp. 1: Creating False Memory on Chinese Williams Syndrome

實驗一：威廉氏症候群的記憶錯誤研究

Participants 16 WS patients join in this study. Their mean chronological age is 33.71 and mean mental age is 8.76 based on WSIC-III (for patients who are younger than 16 years of age) or WAIS-III (for patients who are older than 16 years old). Another group with matched mental age children (mean chronological age is 8.54) and fully developed college students are recruited as control groups.

Design and Materials 8 lists with 10 words each are studied items. Each list refers to a non-presented word which is the theme word of the list (in this study we call it the lure item). For example, a list containing words as following: 感冒, 癌症, 康復, 吃藥, 健保, 頭痛, 診所, 照顧, 打針, 蒼白 and the theme word related to it is 生病. The word lists for this study are given in Appendix 1. All studied words are recorded by using the software for speech synthesis, *Praat*, in a female voice and the recording rate is approximately one syllable per second. All words are disyllables and highly semantically related with each other. Stimuli are presented in blocks rather than mixed design.

As for the recognition phase, another 72 items are presented as test stimuli. There are 9 words for each list, including 3 studied words (i.e. old items), 3 non-presented words (i.e. new items)

and 3 theme words (i.e. lure items). The difference between lures and new items is semantic relatedness. Subjects are instructed to press the left side of the mouse to indicate their yes responses and right side their no responses. After each judgment by clicking a mouse, subjects are required to give a confidence values from 5 (the most confident) to 1 (the least confident). There are three dependent variables for this study, response latency, accuracy, and confidence rating.

Unlike Deese (1959), Roediger and McDermott (1995), this study is going to use matched modality presentation. Both study lists and recognition lists are presented in auditory modalities. It seems that the false alarm rates are higher for mismatched modalities, which are the traditional DRM method used (i.e. visual presentation in the study phase and visual presentation in the recognition phase or auditory presentation in the study phase and auditory presentation in the recognition phase). Though study performed by Maylor and Mo (1999) showed that visual-auditory presentation is higher than auditory-auditory (AA) presentation, in this study AA presentation is conducted because WS patients are not fluent readers. Of course, this known study-recognition modality effect will be taken into consideration in discussion.

Procedure In the study phase, a fixation point is displayed on the screen for 500 ms and followed by a disyllabic word approximately 2 second. Subjects are instructed to do nothing but to pay closely attention to the word. In the recognition phase, a fixation point is presented 500 ms on the screen and a test word followed. After subjects' response toward the word by pressing the mouse, a confidence rating is required. After a confidence value is assigned, a blank for 500 ms remained on the screen to initiate next trial.

Prediction There are 3 effects concerned: (1) specific memory effect for the comparison of

old items and lure items, (2) specific memory effect for old items and new items, (3) gist memory effect for lure items and new items.

For college students (the basic control group), it is predicted that they would show all these three effects because they can remember the old items clearly and automatically form a concept from semantically related words.

For matched mental age controlled group, they could also show all these three effects as college students. However, the false alarm rates might be lower for both lure items and new items because they are not fully developed individuals who might have incomplete semantic network as adults. Basically the patterns on these three memory effects should be obtained without doubt.

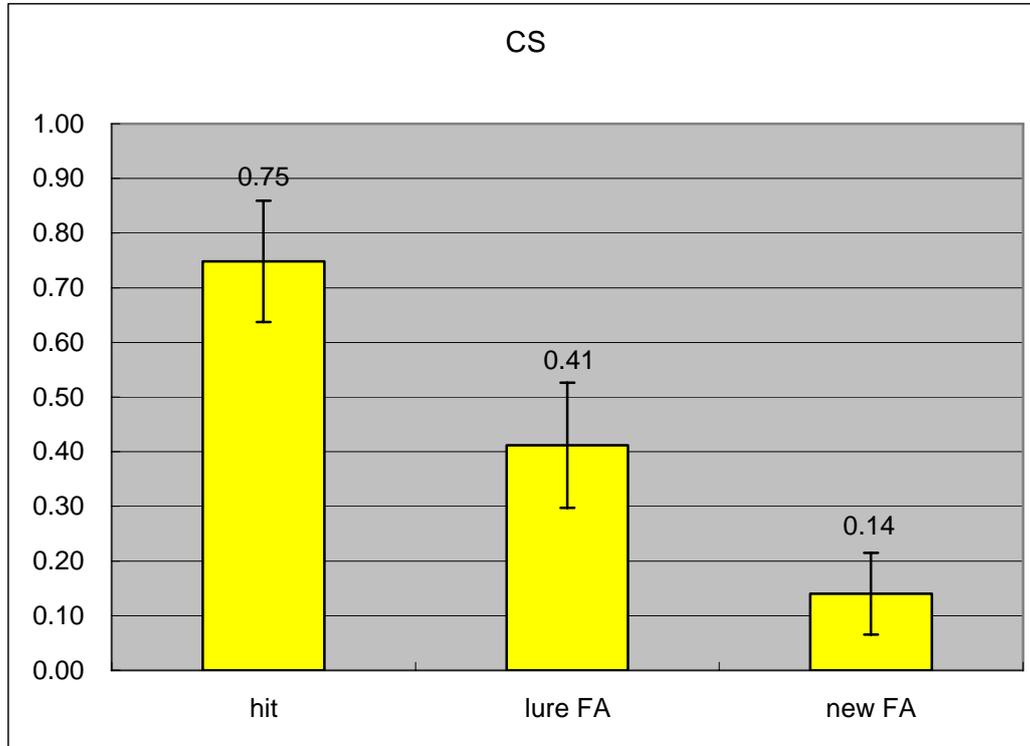
For WS patients, there are two possible predictions. First, if patients have impaired semantic knowledge in terms of concept formation, it is predicted that they would not show gist memory effect. This inability to have automatic formation comes from their dependence on verbatim memory rather than on gist memory. In other words, they can recollect the detailed information of presented words in memory correctly, but they cannot form a gist-based memory from their semantic network. Second, if WS patients have unimpaired semantic knowledge, it is predicted that they would perform same patterns as normal groups.

College Students Results

A one-way ANOVA shows that the main effect of conditions reached significance, $F(2, 8) = 93.01, p < .001$. The hit rates for old items (75%) are higher than the false alarm rates for lure items (41%), which in turn is higher than the one for new items (14%). These results

indicate an automatic semantic formation is observed on college students.

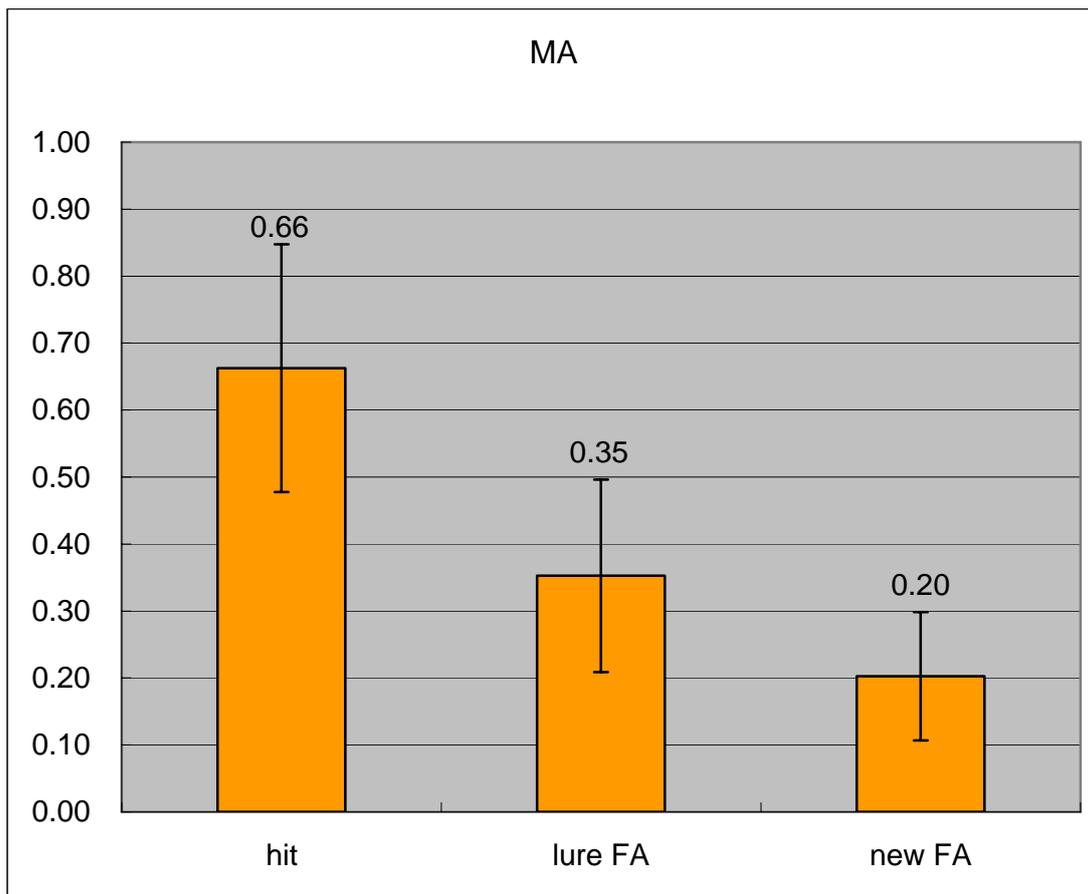
Table 1 Yes Percentage of College Students in Three Conditions (n=5)



Matched Mental Age Control Group Results

A one-way ANOVA shows that the main effect of conditions is significant, $F(2, 30) = 67.30, p < .001$. The patterns are very similar to the one of college students.

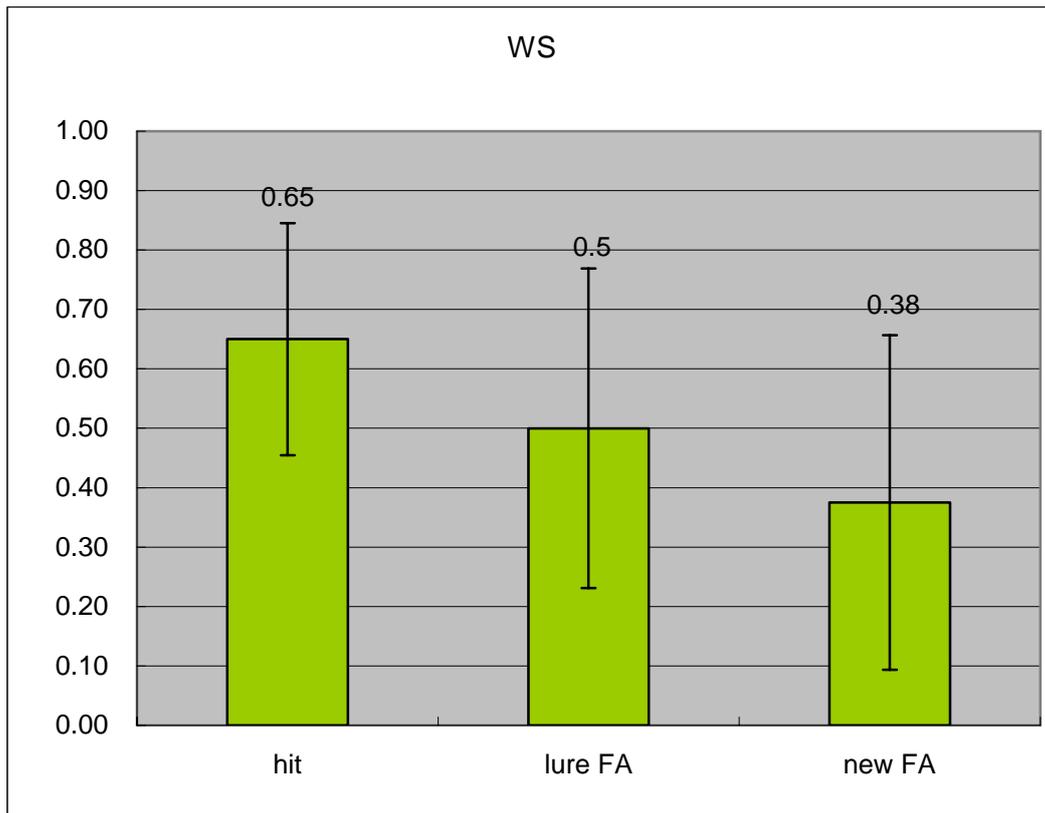
Table 2 Yes Percentage of Matched Mental Age Control Group in Three Conditions (n=16)



Williams Syndrome Patients Results

Another one-way ANOVA shows that the same patterns are obtained in WS patients [$F(2, 30) = 15.65, p < .001$]. They show high misrecognition rates (i.e. false alarm rates) for semantically related items, suggesting an unimpaired semantic knowledge in their mental lexicon. They can not only recognize the old items clearly, but also form gist memory for lure items.

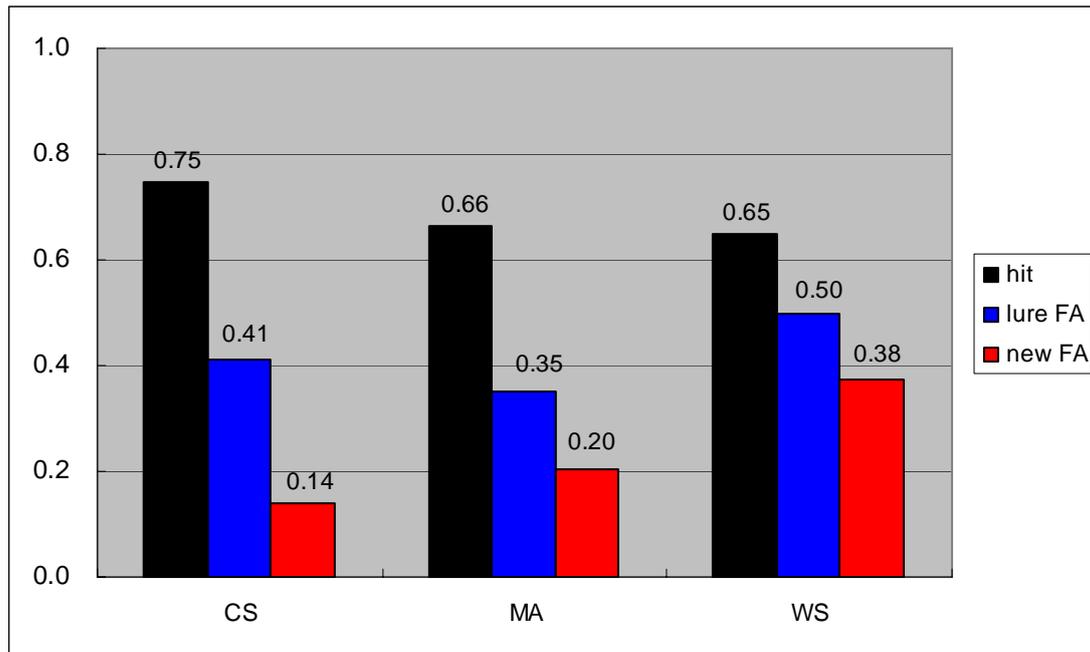
Table 3 Yes Percentage of Williams Syndrome Patients in Three Conditions (n=16)



Group Comparison Results

A two-way ANOVA which takes groups as a between factor and conditions as a within factor show that no interaction is found, $F(4, 102) = 1.861, p > .05$. The main effect of groups is significant, $F(2, 102) = 3.367, p < .05$ and the main effect of conditions is also significant, $F(2, 102) = 36.42, p < .001$. This pattern indicates that the results are quite parallel in three groups. No any group performs worse than other groups in any conditions.

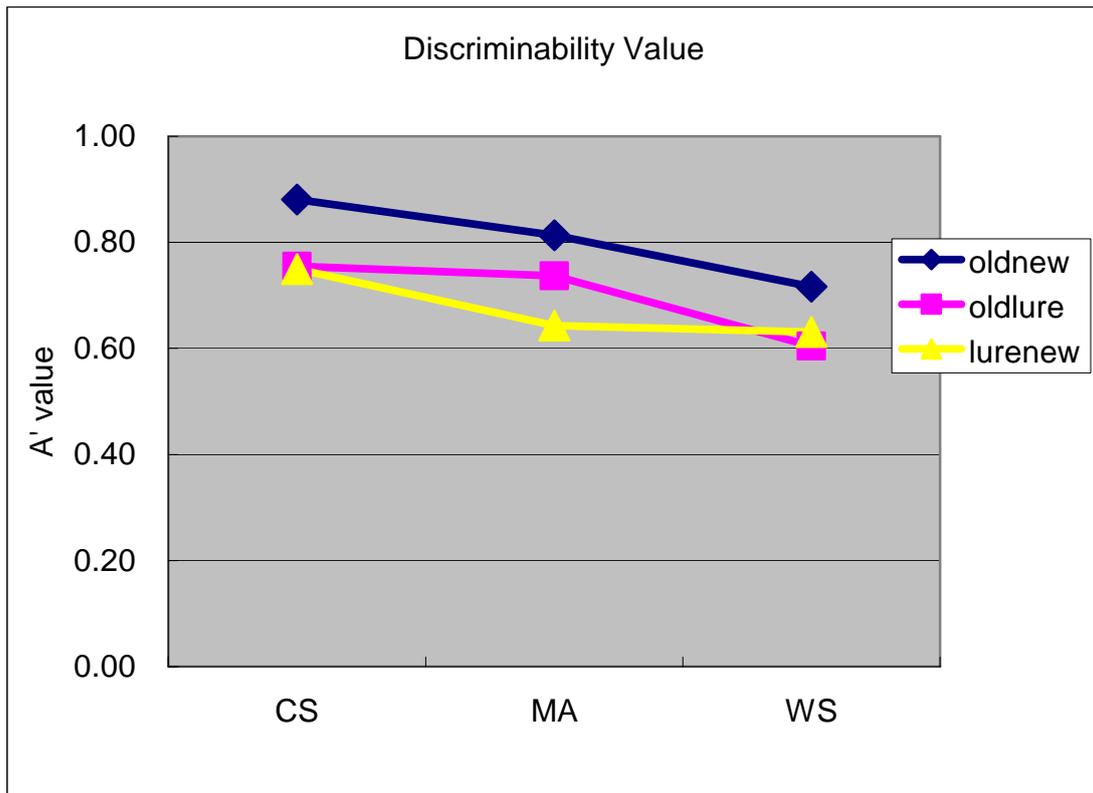
Table 4 Yes Percentage of Three Groups in Three Conditions



Individual Analysis on Discriminability Values

A two-way ANOVA shows that there is no interaction between these three groups in three conditions, $F(4, 102) = .983, p > .05$. The main effects on groups and on subjects are all significant ($p < .05$). For old and new A' value, a one-way ANOVA shows that the comparison between college students (CS) and matched mental age controls (MA) is marginally significant. But other two comparisons between MA vs. WS and CS vs. WS are significantly different. For old and lure A' value, another one-way ANOVA shows that the A' between CS and MA is not different to each other. This result suggests that these two groups use similar criteria in judging lures. However, the comparison between MA and WS is significantly different. Meanwhile, the comparison between CS and WS reaches marginally different. So, WS patients have lower criteria in judging lure items. As for lure and new items, the comparison between WS and MA is not different and the comparison between CS and MA is marginally different. The difference between CS and WS is significant.

Table 6 Discriminability Values (A') of Three Groups in Three Conditions

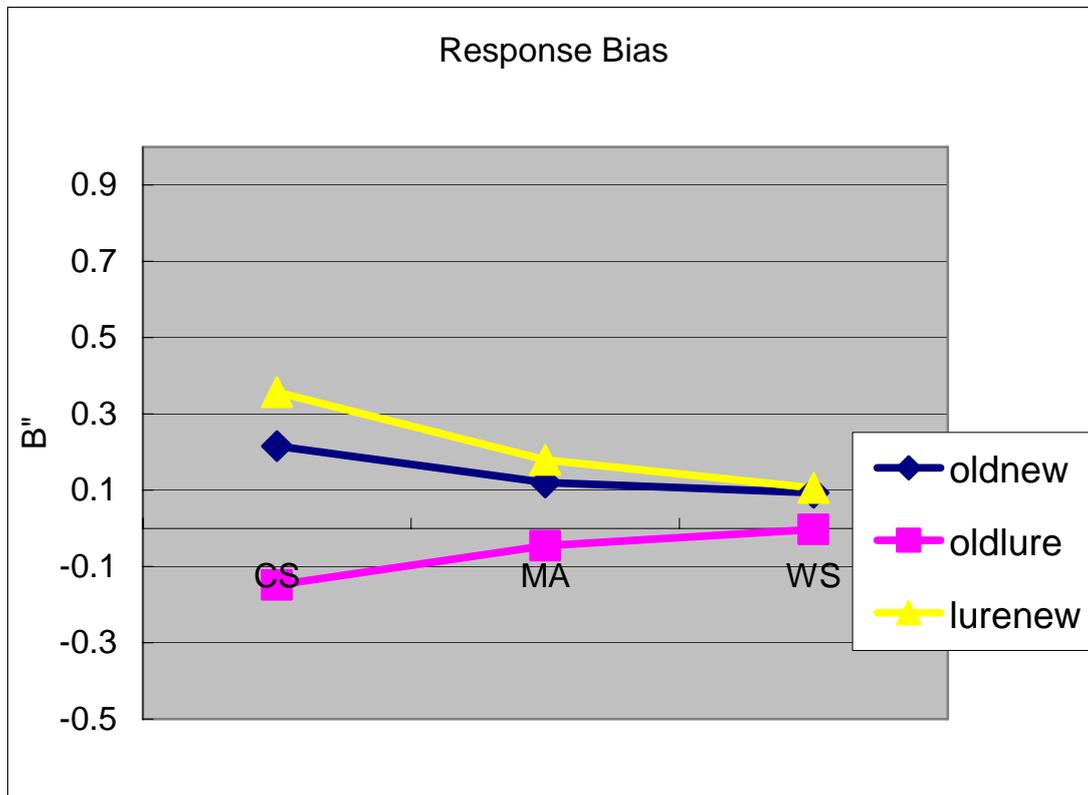


Individual Analysis on Response Bias

A one-way ANOVA shows that there is no interaction between three groups on any comparisons of conditions, $F(4, 102) = 1.01, p > .05$. The main effect of conditions reaches significance ($p < .05$), but the main effect of groups does not ($p > .05$). For college students, B'' value on the comparison between old and lure items is different from comparisons between old vs. new items and lure vs. new items, suggesting that they are more liberal to respond to lure items. It seems that they could not distinguish lure items from old items. In other words, college students treat lures much more like old items. For matched mental age children, they also show the same pattern, which the comparison between old vs. lure items is significantly different from comparisons between other conditions. This pattern observed in both college students and matched mental age children is not obtained in WS patients. None of the comparisons reaches significance. They seem to perform all responses in conservation.

They are not prone to say yes to all responses. In other words, WS patients do understand this task and follow the instruction.

Table 7 Response Bias (B'') of Three Groups in Three Conditions



Interim Summary of Exp. 1

In this study, WS patients show parallel pattern to their normal control groups, including college students and matched mental age children. They show three memory effects on specific items and constructed non-presented concepts. Based on these results, it can be inferred that WS patients do not have impaired semantic knowledge because they can form semantic network from displayed related words automatically. Their semantic network seems to be (near-) normal as typically developing adults and children.

However, questions still remain. Do their behavioral patterns come from same

mechanism operated in cognition as normal people or from superficial behavioral proficiency? In other words, do these results merely reflect their surface knowledge disguised by totally different underlying mechanism? If so, what kind of mechanism they use/apply? In order to pursuit questions further, neural correlates of gist memory formation are investigated by using event-related potentials with high temporal resolution.

Exp. 2: An Event-Related Brain Potentials Study of False Memory on Chinese Williams Syndrome

實驗二：威廉氏症候群的記憶認知功能---一個事件相關誘發電位腦波研究

This study is parallel to experiment one. According to previous studies with ERPs as face processing of WS patients (Mills, Alvarez, George, Appelbaum, Bellugi, and Neville, 2000), different underlying neuropsychological patterns are found (i.e. reversed N100/N200 complex and also N320 component for both upright and inverted faces), which are very distinctive from the ones observed in normal people. Moreover, another ERPs study on comprehension (i.e. N400) also finds different patterns from normal people. WS patients show both N400 and LPC toward semantic anomaly sentences whereas typically developing control groups do not show N400 anymore (Neville, Mills, & Bellugi, 1994). Therefore, it is interesting to see whether WS patients show different patterns of neural correlates to semantic memory as false memory paradigm employed in this study.

Participants 7 subjects with WS who are older than 10 years old (CA = 27.41; MA = 9.59) and matched mental age children (MA = 9.56) are recruited. Since these two groups of individuals are not fully developed populations, another 10 college students are included serving as a basic control group.

Design and Materials There are 16 word lists which are divided into two blocks in this study. Each block contains 8 word lists and 13 words are included in each list. Thus there are 208 words presented to each subject in the study phase. After each block, another 96 words are tested in the recognition phase. There are 12 words for each word list, including 3 presented old words (i.e. old items), 6 non-presented theme words (i.e. lure items) and 3 non-presented new words (i.e. new items). All stimuli are recorded in a female voice approximately one word a second with a sound-recorded software, *Praat*, in 44100 monotone frequency. This study is also in AA presentation which means stimuli are all presented in auditory modality in both study phase and recognition phase. The stimuli are provided in Appendix 2.

Procedure In the study phase, a fixation point is shown on the computer screen for 2500ms. During presentation of a fixation, after 500ms there is a disyllabic word presented through speakers connected to a computer displayed stimuli. A disyllabic word is approximately presented for 2 seconds. Right after this presentation, another fixation on the screen starts a new trial.

In the recognition phase, a fixation point is displayed on the computer screen for 500ms. After 500ms, a target word is presented through speakers. Subjects are required to make a judgment toward that target whether it is heard before by clicking a mouse. If the target word has been heard before, they press the left side of the mouse. If it has not been heard before, they press the right side of the mouse. After each judgment is made, a 500ms blank is shown on the screen and a sign “@” is displayed instead to initiate next trial. Subjects are asked to press the space bar to begin. This design is sort of like self-pace reading paradigm in which subjects can stop making judgment during experiment at any time. This design is especially for WS patients because they may have unexpected needs. Each target word is presented in

approximately 2 seconds. Thus a complete trial is approximately 3500ms. Before real experiment begins, practice trials are given to each subject. For WS patients, several practice sections are needed to make sure they understand requirements.

During this experiment, EEG is recorded simultaneously. The electrode sites are especially interested on the left parietal area and the right frontal area, which are reported to be relevant to false memory.

Prediction

For normal participants, the old new effect is supposed to be observed. According to the literature, the brainwaves for old items are more positive going and the ones for new items are more negative. For WS patients, they are predicted to show same patterns as their normal control groups.

EEG Recording

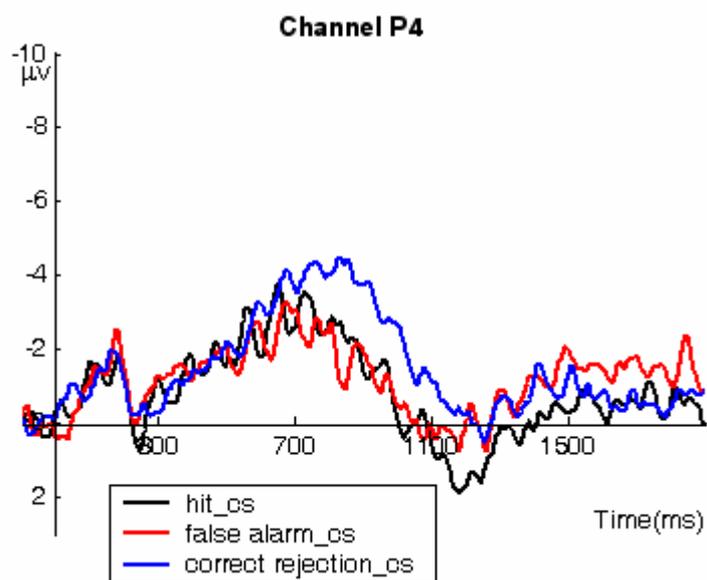
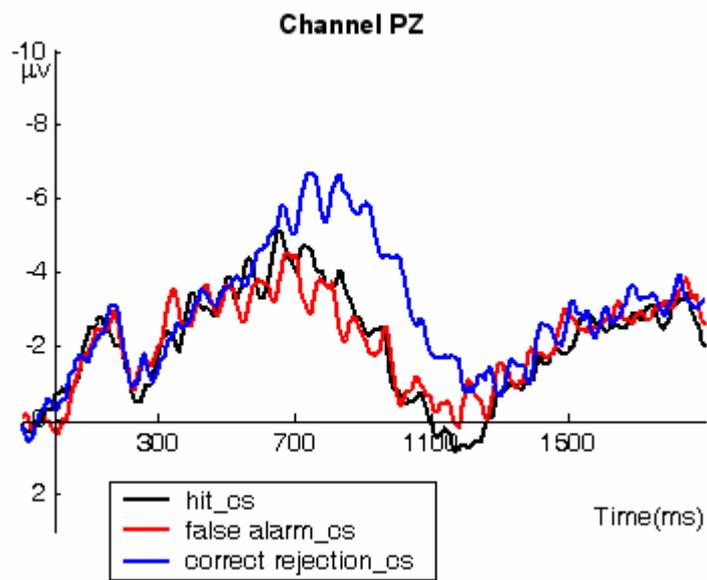
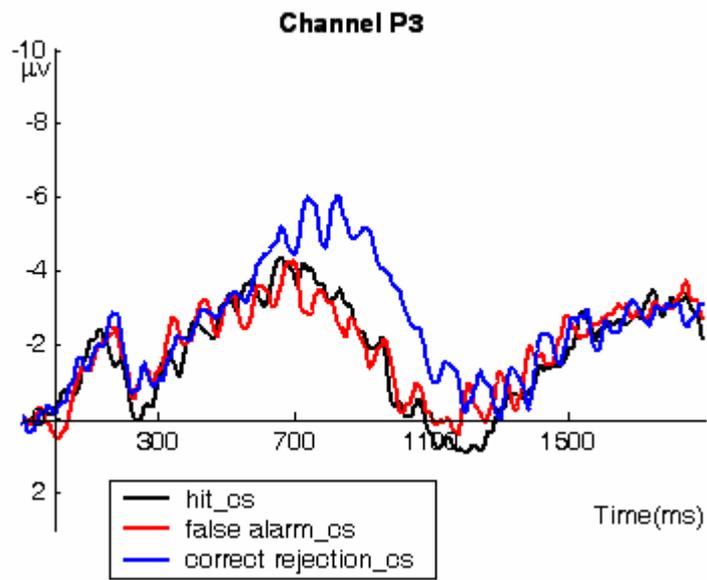
A 64 channel electrode cap is used in recording EEG on college students and WS patients whereas another 32 channel electrode cap is used in recording matched mental age students' EEG. Equivalent electrode sites are compared no matter which channel numbers are used. Recordings are taken from all channels over the scalp including standard 10-20 system locations. The EEG was amplified with a bandpass of .01 to 100 Hz. ERPs are averaged off-line. The averages include only trials that are free from artifact to which the subject responded correctly.

College Students Results

The brain waveforms of these three conditions are provided in Figure 1 to 3 below. The examples shown here are in central electrode sites, C3, Cz, and C4. The old new effect is

clearly observed in college students, i.e. the waveforms of hit responses are significantly different from the ones of correct rejection responses. The differences are in the time window of 700ms to 1250ms in left and central cortexes. The main effect of condition is $p = .0037$ and the main effect of hemisphere is $p = .0087$. The difference of the later mainly comes from the difference between central hemisphere and left hemisphere ($p < .05$) and also the difference between central hemisphere and right hemisphere ($p < .05$). The interaction of hemisphere and cortical area reaches significance, $p = .0014$ and the simple main effect comes from the difference between central and parietal cortex in left hemisphere, $F(1, 27) = 5.736$, $p = .024$. The left hemisphere has greater neural activities (-4.802) than the right hemisphere (-2.053) and so does the central hemisphere (-4.753) than the right hemisphere. Meanwhile, the simple main effect of the difference between these two cortical areas in the central hemisphere reaches marginal significance, $F(1, 27) = 3.570$, $p = .069$. Furthermore, the simple main effect of hemispheres in central cortex is significant, $F(2, 36) = 11.60$, $p = .0001$. The simple main effect of hemispheres in parietal cortex is not significantly different.

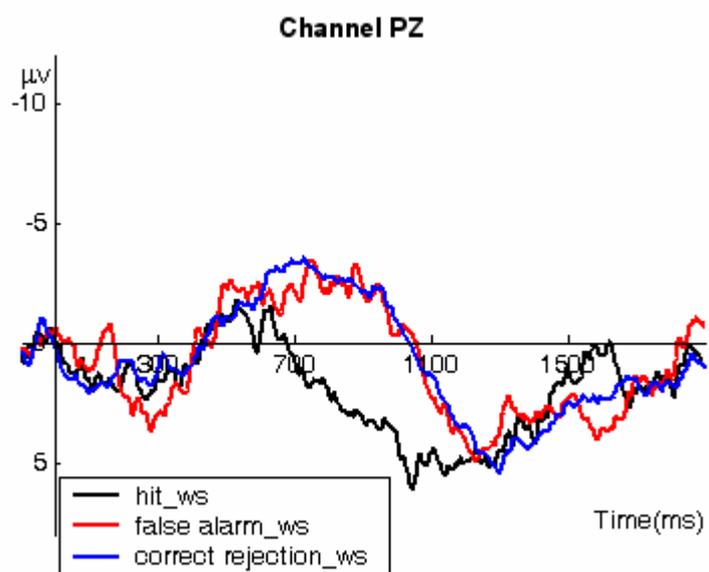
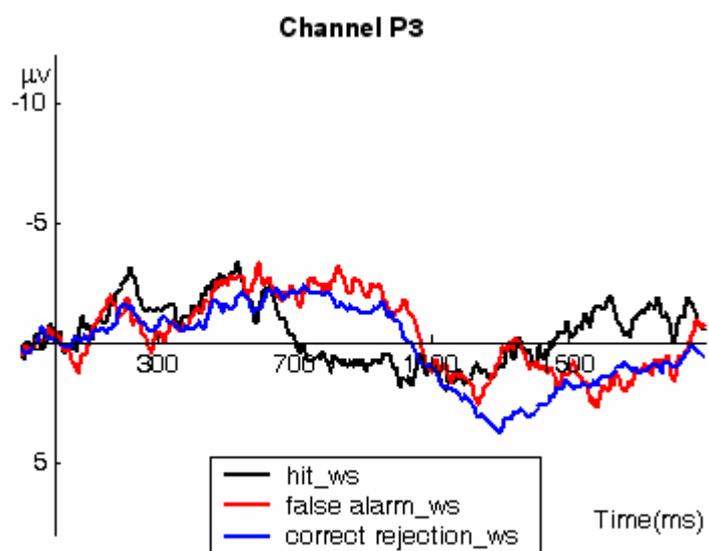
A two way interaction of hemisphere and cortical area is significant, $p = .029$. The simple main effect comes from the larger activation of right frontal area (-3.638) than the right central area (-2.053). This difference is marginally significant, $F(1, 27) = 4.195$, $p = .05$. Compared to central area, frontal area has similar activation in left hemisphere and central hemisphere (-4.802 and -4.753 for the former; -4.979 and -4.349 for the latter). There is no difference between these locations.

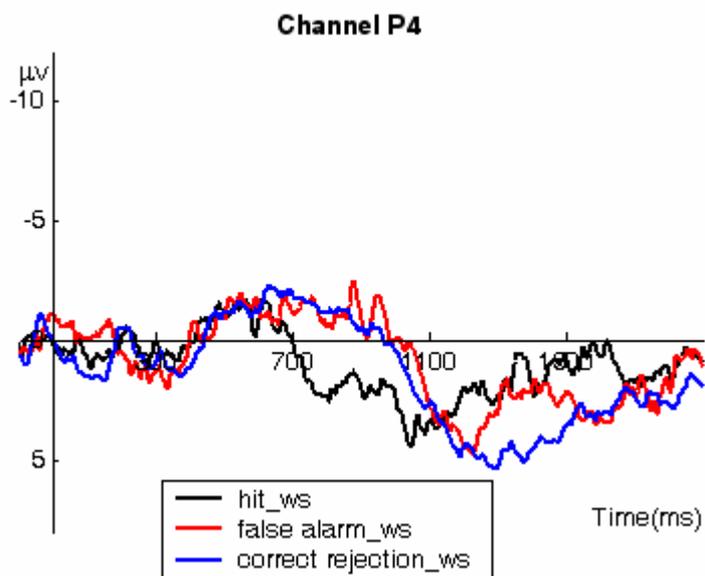


Williams Syndrome Results

From the brain waveforms of WS patients, it seems clear that old new effect is obtained.

However, due to the limitation of subject numbers (n=6), the difference is not significant.





Reference

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Appendix 1 DRM word list in Exp. 1

	Study List 1	Study List 2	Study List 3	Study List 4	study list 5	study list 6	Study List 7	Study List 8
theme	心情	學校	交通	工作	生病	飲食	音樂	婚姻
words	快樂	上課	車子	薪水	感冒	三餐	旋律	結婚
	難過	考試	捷運	賺錢	癌症	品嚐	演奏	夫妻
	憂鬱	作業	馬路	過活	康復	嘴巴	鋼琴	嫁人
	苦澀	黑板	事故	糊口	吃藥	咀嚼	節拍	成親
	沉悶	書本	規則	養家	健保	美味	唱片	歸宿
	不安	筆記	阻塞	辛苦	頭痛	米飯	歌曲	姻緣
	寂寞	用功	流量	溫飽	診所	吞嚥	古典	倚靠
	煩惱	演講	混亂	渡日	照顧	腸胃	樂器	幸福
	無聊	簿子	擁擠	貧窮	打針	牙齒	樂團	圓滿
	孤獨	小抄	安全	乞丐	蒼白	口腔	爵士	月老
	study list 1	study list 2	study list 3	study list 4	study list 5	study list 6	study list 7	study list 8
old item1	快樂	上課	車子	薪水	感冒	三餐	旋律	結婚
old item2	寂寞	用功	流量	溫飽	診所	吞嚥	古典	倚靠
old item3	煩惱	演講	混亂	渡日	照顧	腸胃	樂器	幸福
lure1	心情	學校	交通	工作	生病	飲食	音樂	婚姻
lure2	開心	學生	運輸	維生	醫生	吃飯	藝術	家庭
lure3	開朗	數學	秩序	做工	護士	消化	調子	安定
new1	陰天	老師	警察	充飢	腹瀉	喝水	廣播	永遠
new2	灰色	認真	黑暗	遊民	嘔吐	喉嚨	喇叭	女人
new3	枯燥	鉛筆	繁忙	節省	睡覺	甘甜	天王	溫暖

Appendix 2 ERP DRM word list in Exp. 2

theme word	環境	個性	節慶	睡覺	食物	新聞	明星	行李
	清潔	濫馴	元宵	清醒	飢餓	主播	演員	料理
	打掃	粗暴	喜慶	床鋪	蔬菜	報紙	舞台	收拾
	整齊	衝動	歡喜	棉被	營養	記者	漂亮	包裹
	骯髒	體貼	鞭炮	枕頭	新鮮	氣象	燈光	負責
	垃圾	冷靜	喧鬧	疲倦	健康	偏見	歌迷	旅行
	美觀	紳士	吵雜	熬夜	牛奶	抹黑	藝人	打算
	舒服	有禮	舞獅	呵欠	冰箱	媒體	崇拜	乾淨
	街道	冷漠	廟會	打呼	超市	消息	美麗	細心
	明亮	好人	人群	毛毯	可口	雜誌	表演	準備
	回收	激烈	繁華	平靜	過期	攝影	好看	出門
	玻璃	恭敬	夜市	滿足	冷凍	頭條	掌聲	上路
	涼爽	猛烈	冷清	安詳	腐爛	大事	賺錢	預備
	寬敞	君子	安靜	催眠	丟掉	資訊	有名	一切
lure1	環境	個性	節慶	睡覺	食物	新聞	明星	行李
lure2	乾淨	善良	過年	休息	水果	電視	偶像	安排
lure3	衛生	脾氣	熱鬧	作夢	魚肉	報導	電影	整理
old1	清潔	濫馴	元宵	清醒	飢餓	主播	演員	料理
old2	街道	冷漠	廟會	打呼	超市	消息	美麗	細心
old3	陽光	激烈	繁華	平靜	過期	攝影	好看	出門
new1	髮圈	紅燈	缺點	幫忙	鈔票	建築	夜景	迷信
new2	自由	電話	溝通	加油	國王	傳真	老闆	工廠
new3	和尚	薪水	喉嚨	大樓	香菸	香皂	森林	讚美
related new1	路面	淑女	放假	夜晚	海鮮	晚報	花瓶	離開
related new2	社區	溫柔	習俗	中午	宵夜	獨家	美女	打點
related new3	都市	老實	團聚	藥丸	好吃	颱風	閃亮	出發

theme word

保姆

煮菜

離婚

溫暖

癱瘓

制服

壽星

生病

奶媽
小孩
慈祥
幼兒
母親
搖籃
奶瓶
女人
耐心
餵奶
育嬰
和藹
尿布
保姆
嬰兒
愛心
奶媽
奶瓶
和藹
警察
結婚
油漆
娃娃
照顧
媽媽

火候
廚師
烹飪
美食
爐子
剛好
瓦斯
雞湯
中藥
味覺
功夫
時間
鹽巴
煮菜
廚房
爐火
火候
雞湯
味覺
手腳
新書
山谷
美味
煎藥
煮飯

婚姻
愛情
盡頭
不幸
分手
破裂
珍惜
失戀
挽回
後悔
失敗
婚嫁
談判
離婚
夫妻
感情
婚姻
失戀
後悔
杜鵑
地震
神仙
回頭
補救
遺忘

春天
衣服
陽光
毛衣
棉被
天氣
暖爐
冬天
溫泉
幸福
小手
被窩
熱茶
溫暖
寒冷
太陽
春天
冬天
幸福
電線
面具
教會
美滿
家庭
高溫

病人
輪椅
殘障
復健
老人
車禍
可憐
脊椎
生病
僵硬
健康
不動
辛苦
癱瘓
中風
麻痺
病人
脊椎
僵硬
開燈
科學
流淚
無助
沒力
病床

規定
學校
便服
服裝
一致
藍色
黑白
僵化
呆板
國中
高中
校規
背心
制服
學生
整齊
規定
僵化
國中
插嘴
奇怪
天空
上學
顏色
紀律

快樂
鬍子
爺爺
拜壽
年齡
蠟燭
禮物
高壽
紅蛋
慶祝
重視
舞會
唱歌
壽星
生日
蛋糕
快樂
高壽
慶祝
約會
蚊子
血管
健壯
大壽
長大

感冒
癌症
康復
吃藥
健保
頭痛
診所
昏沈
打針
蒼白
掛號
流行
發燒
生病
醫生
護士
感冒
診所
照顧
分配
蝴蝶
敵人
腹瀉
嘔吐
排隊

lure1
lure2
lure3
old1
old2
old3
new1
new2
new3
related new1
related new2
related new3

