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Electrophysiological correlates of semantic processing in Williams syndrome

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ABSTRACT

Williams syndrome (WS), a genetic neurodevelopmental disorder due to microdeletion in chromosome 7, has been described as a syndrome with an intriguing socio-cognitive phenotype. Cognitively, the relative preservation of language and face processing abilities coexists with severe deficits in visual-spatial tasks, as well as in tasks involving abstract reasoning. However, in spite of early claims of the independence of language from general cognition in WS, a detailed investigation of language subcomponents has demonstrated several abnormalities in lexical-semantic processing. Nonetheless, the neurobiological processes underlying language processing in Williams syndrome remain to be clarified. The aim of this study was to examine the electrophysiological correlates of semantic processing in WS, taking typical development as a reference. A group of 12 individuals diagnosed with Williams syndrome, with age range between 9 and 31 years, was compared with a group of typically developing participants, individually matched in chronological age, gender and handedness. Participants were presented with sentences that ended with words incongruent (50%) with the previous sentence context or with words judged to be its best completion (50%), and they were asked to decide if the sentence made sense or not. Results in WS suggest atypical sensory ERP components (N100 and P200), preserved N400 amplitude, and abnormal P600 in WS, with the latter being related to late integration and re-analysis processes. These results may represent a physiological signature of underlying impaired on-line language processing in this disorder.

1. Introduction

Williams syndrome (WS), a rare neurodevelopmental disorder characterized by a deletion on chromosome 7 q11.22-23, has called the attention of researchers due to its intriguing phenotype in which an apparent preservation of language and face processing seems to coexist with severe intellectual deficits in other cognitive domains such as visual-spatial processing and executive functioning (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; see Martens, Wilson, & Reutens, 2008, for a review). This syndrome was initially proposed as a paradigmatic example of cognitive dissociation and of a modular organization of the brain and, more specifically, an example of the independence of language from general cognition (Bellugi,

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Bihrle, Jernigan, Trauner, & Doherty, 1990; Bellugi, Bihrle, Neville, Jernigan, & Doherty, 1992; Bellugi, Marks, Bihrle, & Sabo, 1988; Pinker, 1994).

In fact, when compared with other developmental disorders such as Down syndrome (DS) there is evidence for increased verbal production in WS (e.g., Mervis & Robinson, 2000). Other aspects of language production, such as affective prosody and the use of audience hookers, seem to be not very different from typically developing chronological age matched controls (Gonçalves et al., 2004, 2010; Jones et al., 2000; Reilly, Klima, & Bellugi, 1991). Additionally, WS verbal abilities seem to develop at a faster rate than their nonverbal abilities (Jarrold, Baddeley, & Hewes, 1998).

However, contrary to initial claims of modular language preservation, recent studies show that language abilities are below age-appropriate levels (Grant, Valian, & Karmiloff-Smith, 2002; Landau & Zukowski, 2003; Laws & Bishop, 2004; Lukács, Pléh, & Racsmány, 2004; Phillips, Jarrold, Baddeley, Grant, & Karmiloff-Smith, 2004; Pléh, Lukács, & Racsmány, 2003; Sullivan, Winner, & Tager-Flusberg, 2003; Vicari et al., 2004) and follow an atypical developmental pathway (e.g., Laing et al., 2002; Mervis & Bertrand, 1997; Mervis et al., 2003). It appears that some subcomponents (receptive vocabulary, phonological short-term memory, regular morphology, production of affective prosody) are relatively more preserved than others (e.g., repetition of syntactically complex sentences, production of grammatical gender, irregular morphology, comprehension of spatial terms, comprehension of figurative language, pragmatics) (see Brock, 2007, for a review).

There is less agreement in the literature when it comes to semantic fluency. While some studies suggest a higher production of words in semantic fluency tasks in WS individuals than would be expected for their average mental age along with an increased production of atypical category exemplars (e.g., Bellugi et al., 1990, 1992, 1988; Bellugi, Wang, & Jernigan, 1994; Rossen, Klima, Bellugi, Bihrle, & Jones, 1996; Wang & Bellugi, 1993), others suggest that this is not the case (Jarrold, Hartley, Philipps, & Baddeley, 2000; Johnson & Carey, 1998; Scott et al., 1995; Stevens & Karmiloff-Smith, 1997; Volterra, Capirci, Pezzini, Sabbadini, & Vicari, 1996).

There is a similar lack of consensus in relation to semantic priming. Some studies found the same effects of priming on reaction times of individuals with WS and controls (e.g., Tyler et al., 1997), which may suggest a normal organization of semantic system in WS. However, other studies suggested abnormal lexical knowledge in WS (e.g., Bellugi et al., 1988, 1990; Jarrold et al., 2000; Johnson & Carey, 1998; Rossen et al., 1996; Volterra et al., 1996; Ypsilanti, Grouios, Alevriadou, & Tsapkini, 2005), as well as deficits in lexical access (e.g., Bromberg, Ullman, Marcus, Kelly, & Levine, 1995; Temple, Almazan, & Sherwood, 2002; Ypsilanti, Grouios, Zikouli, & Hatzinikolaou, 2006). This finding led some authors to propose a dissociation between preserved structure of semantic memory and impairment in the integration of word meanings into on-line developing sentential representations (Tyler et al., 1997).

The impairments found in semantic memory may be explained by a lack of maturity in conceptual organization of semantic categories (e.g., Jarrold et al., 2000; Johnson & Carey, 1998) in the sense that individuals with WS can put information in semantic networks but might have difficulties in conceptually reorganising this information. This hypothesis seem to be corroborated by studies showing a decreased sensitivity to word frequency in WS and a nearly equal preference for primary and secondary meaning of homonyms (Rossen et al., 1996), a trend for the production of infrequent words towards the end of a trial in semantic fluency tasks (Rossen et al., 1996), or an atypical semantic development in the sense that older individuals with WS do not present a more sophisticated conceptual structure than younger individuals (Jarrold et al., 2000).

Together, these findings question the claim for a modular preservation of language in WS. It is possible that the idea of the intact language may have arisen from the comparisons with DS where more severe language impairment is observed (e.g., Mervis, 2003). Another contributing factor may have been the techniques used to study this genetic disorder. Most studies used behavioral measures to probe language function in WS. While these are valuable studies, they preclude observation of neural processes that underlie language performance. Both reaction times and error rates are, by definition, aggregate measures of all processes that went into making a response. As such they do not provide information about possible abnormalities at different stages of information processing prior to the response. Thus, a more comprehensive approach to the study of language in both normal and clinical population is the concurrent use of behavioral and event-related potential techniques (e.g., Hsu, Karmiloff-Smith, Tzeng, Chin, & Wang, 2007).

Event-related potential (ERP) techniques are one of the few methodologies that document real time changes in neurocognitive processes. As such they are a valuable supplement to the study of language (Kutas & Federmeier, 2000). One of the components that has been associated with language processing in numerous studies is the N400, a negative going potential that peaks around 400 ms after the presentation of visual stimuli, and around 300–350 ms to auditorily delivered stimuli (see Lau, Philipps, & Poeppel, 2008, for a review). The N400 has been found sensitive to processing semantic information regardless of the physical nature of the stimulus, i.e., it has been found to both visual and auditory language stimuli (e.g., Anderson & Holcomb, 1995; Besson & Macar, 1986; Kutas & Iragui, 1998; Niznikiewicz et al., 1997) as well as to pictures (e.g., Coch, Maron, Wolf, & Holcomb, 2002; Federmeier & Kutas, 2002; West & Holcomb, 2002; Willems, Özyürek, & Hagoort, 2008). It seems to reflect the ease with which two concepts can be linked together and as such it has been used as an index of priming and context use (e.g., Brown & Hagoort, 1993; Federmeier & Kutas, 1999a,b; Halgren, 1990; Kutas & Hillyard, 1980, 1984; Rugg, Doyle, & Holdstock, 1994; Van Petten, 1993). The N400 has been found to peak maximally at centro-parietal electrodes and is more negative going to words or pictures that do not fit well the proceed context relative to these semantic items that fit the previous context well. In addition, its amplitude can be influenced by word frequency, word type (e.g., closed vs. open class), word lexical status, or the semantic or associative relationship between words, but it is insensitive to decision-related and response-selection mechanisms (Heinze, Muente, & Kutas, 1998). Therefore, the N400 component may be a good probe to study language processes in WS.