Research in Developmental Disabilities 34 (2013) 3332–3342

Contents lists available at SciVerse ScienceDirect

Research in Developmental Disabilities

Contextual integration of causal coherence in people with Williams syndrome

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ARTICLE INFO

Article history:
Received 12 April 2013
Received in revised form 21 June 2013
Accepted 21 June 2013
Available online

Keywords:
Causal inference
Contextual integration
Central coherence
Williams syndrome

ABSTRACT

This study investigated causal coherence in people with Williams syndrome (WS). To advance our understanding of this clinical group, we examined their ability to make causal inferences, using their understanding of homonyms (words with the same spelling but distinct meanings) embedded in contexts. A minor goal was to use verbal stimuli to clarify Santos and Deruelle's (2009) findings on the knowledge of causality among people with WS. Participants were presented with two types of scenarios requiring different inference directions: backward inferences (from consequence to cause) and forward inferences (from cause to consequence). Following each scenario, they were asked a comprehension question and given three possible answers that corresponded to a figurative, literal, and unrelated meaning of the homonym embedded in the scenario. The correct answer required the participants to make a successful causal inference. People with WS aged from 13 to 29 ($n = 17$, mental age = 6–14) were able to make backward and forward inferences by selecting the context-appropriate meanings of homonyms, thus demonstrating the existence of contextual integration ability in the causal coherence of people with WS. However, as their accuracy in the figurative meaning responses was lower than that of healthy age-matched controls, suggesting the participants with WS, were delayed in the contextual integration of causal coherence. The participants with WS chose a significantly higher percentage of answers with unrelated meanings than the two control groups, indicating a certain degree of weakness in the contextual integration of homonyms in context.

1. Introduction

Williams syndrome (WS) is a rare disorder caused by a genetic deficit on chromosome 7q11.23 (Korenberg et al., 2000); the etiology is 1 in 7500 live births (Strømme, Bjømstad, & Ramstad, 2002). People with WS have an important place in the study of developmental disorders because of their unusual cognitive profile, which consists of relatively strong verbal skills and relatively weak nonverbal skills (Bellugi, Lichtenberger, Jones, Lai, & George, 2000; Mervis & John, 2010; Semel & Rosner, 2003). Since this syndrome was identified in 1961 (Williams, Barrett-Boyes, & Lowe, 1961), studies have shown that people with WS have preserved syntactic knowledge in the production of counterfactual conditionals (e.g., they correctly answered *I could fly* after the hypothetical conditional statement *If I were a bird*) and in understanding reversible passives (e.g., they

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0891-4222/$ – see front matter
http://dx.doi.org/10.1016/j.ridd.2013.06.031

accurately chose the picture corresponding to the passive sentence of *The horse is chased by the girl* from another alternative *The girl is chased by the horse*) (Bellugi, Lichtenberger, Jones, Lai, & George, 2000).

Other studies have shown that people with WS have rich lexical semantic knowledge and can generate hyponyms (e.g., dog, cat, rabbit) under superordinate categories (e.g., animal) and access both primary meanings (e.g., river) and secondary meanings (e.g., money) of homonyms (e.g., bank) (Rossen, Bihrle, Kuma, Bellugi, & Johns, 1996). A report by Tyler et al. (1997) revealed that people with WS exhibited the same monitoring priming of category taxonomies (e.g., natural type: lettuce-cabbage; artifact type: coat-hat) and functional relations (e.g., theater-play) to related pairs compared to unrelated ones as healthy controls, suggesting normal semantic association of lexical words. However, atypical semantic processing was observed in people with WS when integrating the final words into preceding contexts, e.g., *I don't go to school on Saturday or floor** (Neville, Mills, & Bellugi, 1994). While healthy controls showed a large N400 reflection, participants with WS showed compromised amplitudes to the same semantic anomaly sentences. These results indicated that people with WS integrated words into sentences in an atypical manner, as Tyler et al. suggested (1997). However, in their study of semantic integration at the sentential level, Pinheiro, Galdo-Álvarez, Sampaio, Niznikiewicz, and Gonçalves (2010) found the opposite results with regard to the performance of N400 amplitudes in people with WS. They found that incongruent conditions produced atypical brain signatures on N100 (more negative among typically developing controls in the frontal and central sites), P200 (more positive among WS participants at the central site), and P600 (more positive among WS participants at frontal and central sites). These inconsistent findings reveal the uncertainty about the contextual integration ability of people with WS.

A subsequent conceptual formation study used the false memory paradigm (Roediger & McDermott, 1995) and electrophysiological recordings to explore whether people with WS had the ability to integrate associated lexical items into a connected semantic network (Hsu, Karmiloff-Smith, Tzeng, Tai, & Wang, 2007). In that study, people with WS and healthy CA- and MA-matched controls first learned, from an oral presentation, wordlists containing semantically related associates (e.g., *getting a cold*, *hospital*, *cancer*, *fever*) under different themes (e.g., *getting sick*). Then, in the recognition test, participants were required to make old-new judgments about a series of presented words including previously presented old associates (e.g., *hospital*) with non-presented themes as lures (e.g., *getting sick*), and semantically unrelated new words (e.g., *piano*). If participants with WS successfully constructed semantic networks through connecting semantically related associates, the misrecognition rates (false alarms) for lures (semantically related but non-presented themes) would be high. In contrast, the false alarm rates for new words would be low if they were semantically unrelated to the constructed networks. The results revealed that people with WS misrecognized lures as old items at the same rate as healthy age-matched controls, suggesting that they had typical contextual integration.

Despite their successful behavioral performances, atypical neuro-physiological brain signatures have been observed in people with WS. Specifically, individuals with WS showed deviant brainwaves when processing semantically related lures. Although healthy CA-matched controls processed lures in a significantly different manner than they processed semantically unrelated new words (at Pz electrode site on the time window of 700–1300 ms), participants with WS failed to differentiate between these two types of words in their brainwaves. Moreover, unlike the CA controls, who showed a non-significant difference in brainwaves when processing lures and previously presented old words, people with WS had clearly distinct brainwaves during these processes. The MA controls showed partially similar pattern of neural correlates to the clinical group. The discovery of deviant brain signatures in people with WS suggested the existence of brain and behavioral asymmetry during the same cognitive task.

The strength of lexical knowledge and the deficient neurological findings on sentential integrations were compatible with cognitive profile of local preference and global impairment. Seeing the trees rather than the forest is a characteristic of some developmental disorders. It was first documented in people with autism spectrum disorders (ASD) and characterized as weak central coherence (WCC) (Frith, 1989). People with ASD failed to correctly pronounce a homograph based on its leading context (e.g., In her eye/dress there was a big *tear*) (Happé, 1997). A recent study investigating central coherence in people with WS revealed their deviant proposition integration ability (Hsu & Tzeng, 2011). In this study, Hsu and Tzeng used Franks and Bransford's paradigm (1972, 1974a, 1974b) to decompose four-proposition parent sentences into sentences with various numbers of propositions from one to four. Half of the decomposed sentences (maximally to three propositions) were orally presented to participants with WS, and to healthy CA- and MA-matched controls. The other half of the sentences with the same parent sentences were presented in the recognition test. It was hypothesized that the recognition rates would be a function of the number of propositions, because of the automatic integration of information under the central coherence drive. The results revealed that people with WS failed to show this function. Healthy adults integrated sentences with more than three propositions and healthy children integrated sentences with more than two propositions, whereas participants with WS showed bizarre semantic integration from one-proposition sentences. That is, the clinical individuals seemed unable to distinguish propositions, and mixed all of the propositions together. These findings revealed the semantic integration difficulty of people with WS when the unit of integration was larger than words.

Causal inference is an important factor in contextual integration. A successful causal inference requires making implicit connections with the representations of propositions or proposition arguments in the text (Singer & Ferreira, 1983). Singer and Ferreira gave participants reading scenarios with sentences that embedded three types of verbs (explicit, synonym, inference), then measured the response latency to comprehension questions. For instance, after reading the critical sentence *Bob burned/incinerated/threw his report into the fire* in a scenario, participants were asked *Did Bob burn the report?* Participants responded faster to sentences with explicit verbs (*burned*) than sentences with synonym verbs (*incinerated*) and sentences