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Evidence for atypical categorical speech perception in Williams syndrome

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ABSTRACT

Although language processing has been described as being a relatively 'spared' cognitive function in individuals presenting with Williams syndrome (7q11.23 microdeletion; WS), some studies suggest possible alterations at the level of input speech processing. We explored categorical speech perception and non-speech auditory perception in six participants with WS, as well as in chronological age-matched or reading age-matched control groups. Categorical speech perception was explored for the b-d speech sound continuum, where speech sounds vary in spectral acoustic features, and for the d-t speech sound continuum, where speech sounds vary in temporal acoustic features. Non-speech perception was explored for sine-wave analogues of the b-d continuum, which are not identified as speech stimuli. We observed a significantly increased ability in WS participants to detect subtle acoustic changes between adjacent stimuli of the b-d or the d-t continuum outside the phoneme boundary, when control participants showed absence of discrimination. This is the first study to provide evidence for an atypical sensitivity towards subtle acoustic variations during speech and non-speech auditory analysis in WS. Implications for phonological processing and reading acquisition are discussed.

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1. Introduction

Williams syndrome (WS; also known as Williams-Beuren syndrome or infantile hypercalcaemia) is a neurodevelopmental disorder caused by a microdeletion at the level of chromosome 7q11.23 (Korenberg et al., 2000). This relatively rare syndrome (incidence: 1:25,000 life births; Greenberg, 1990) has received considerable research interest in the domain of cognitive neuroscience, since oral language processing has been described to be relatively well preserved relative to other cognitive domains such as visuo-spatial processing, numerical processing and executive control (Bellugi, Lichtenberger, Jones, Lai, & St George, 2000; Bellugi, Wang, & Jernigan, 1994; Jarrold, Baddeley, & Hewes, 1998; Paterson, Girelli, Butterworth, & Karmiloff-Smith, 2006). This genetic syndrome has been put forward as providing neurodevelopmental evidence for the modularity of cognitive processing, given the very good verbal abilities relative to poor spatial abilities. However, recent data provide a more nuanced view, suggesting that language processing, although globally more preserved than spatial processing, is nevertheless atypical. For example, in the lexical and semantic domain, vocabulary knowledge is relatively well developed, with a standardized score beyond 70 for 50% of persons with WS, i.e., higher than expected based on general IQ estimates (overall intellectual functioning is typically situated in the mild to moderate mental retardation range, IQ < 70, Mervis et al., 2000). At the same time, lexico-semantic representations may be learned and stored in an abnormal manner, as indicated by diminished lexical frequency effects, looser semantic representations, and abnormal word categorization abilities (e.g., Böhning, Starke, & Weissenborn, 2004; Nazzi & Karmiloff-Smith, 2002; Rossen, Klima, Bellugi, & Bihrle, 1996; Temple, Almazan, & Sherwood, 2002). The present study provides further evidence for atypical language processing in WS, by targeting the perceptual-phonological level of processing, which has been considered as being particularly well developed.

A number of studies suggest that performance on a range of phonological processing tasks is at mental age or even chronological age appropriate levels. First, clinical descriptions of individuals with WS generally report fluent and well-articulated speech without any phonological or phonetic deformations (e.g., Bellugi et al., 2000; Reilly, Klima, & Bellugi, 1990; Volterra, Capirci, Pezzini, Sabbadini, & Vicari, 1996). Some individuals might even present quite remarkable abilities to acquire the phonology of foreign languages (Barisnikov, Van der Linden, & Poncelet, 1996). Second, experimental investigations focused mainly on phonological fluency, phonological awareness and nonword repetition as estimators of phonological and metaphonological abilities. In phonological fluency tasks (e.g., the participant is asked to produce as many words as possible starting with a target phoneme), children and adults with WS generally perform at the same level as mental or even chronological age-matched controls (Barisnikov et al., 1996; Pezzini, Vicari, Volterra, Milani, & Ossella, 1999; Volterra, Capirci, & Caselli, 2001; Volterra et al., 1996; Volterra, Longobardi, Pezzini, Vicari, & Antenore, 1999). However, it must be noted that phonological fluency tasks can also be performed on the basis of acoustic similarity between the target phoneme and the onset of the words. Studies investigating phonological awareness abilities, such as rhyme judgment, observed less consistently preserved performance. While Laing, Hulme, Grant, and Karmiloff-Smith (2001) observed similar performance between a group of WS participants and a control group matched for reading age on a set of measures of phonological awareness (rhyme awareness, spoonerisms) except for phoneme deletion measures. Majerus, Barisnikov, Vuillemin, Poncelet, and Van der Linden (2003) observed impaired performance on phonological awareness tasks in 4 children with WS, in comparison to chronological age-matched and vocabulary age-matched control groups.

Other studies explored performance in nonword repetition tasks, showing mental age or chronological age appropriate performance in some cases (Barisnikov et al., 1996; Böhning et al., 2004; Majerus et al., 2003). Interestingly, Majerus et al. (2003) varied the phonological familiarity of the nonwords to be repeated, by creating nonwords containing phoneme combinations that are frequent in the native phonology of the WS participants (high phonotactic frequency nonwords) or less frequent (low phonotactic frequency nonwords). Mental age and chronological age appropriate performance was observed for nonwords of low phonotactic frequency but not for nonwords of high phonotactic frequency. Furthermore, the 4 children with WS participating in this study presented either absent or inverted phonotactic frequency effects, suggesting an unusual organization of sublexical phonological representations in WS, relative to the standard phonotactic frequency effects observed in typically