Narration and collaborative conversation in French-speaking children with Williams syndrome

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Abstract

Williams syndrome (WS) is a rare genetic disease with a specific neuropsychological profile. WS people are generally described as being “hypersociable” and as having relatively well-preserved language abilities despite cognitive retardation. Recent research into the structural aspects of their language and their sociability has found nonhomogeneous profiles (with strong and weak points) in these two areas. The goal of this study was to show that the findings are analogous for the pragmatic facet of WS language. We also looked into the source of this heterogeneity by comparing performance on two contrasted tasks: narration and collaborative conversation. Twelve native French-speaking WS children and adolescents ages 6 years 6 months to 18 years 11 months participated in the study. They were compared to children with Down’s syndrome, and to typical children of the same chronological age or the same mental age. The results showed that the pragmatic abilities of the WS children and adolescents defined a unique profile: they were proficient in some areas (narration), deficient in others (collaborative conversation), and atypical in still others (expression of subjective views or feelings).

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1. Introduction

For about 20 years now, researchers in the cognitive sciences have taken an interest in individuals suffering from Williams syndrome (WS), a rare genetic disease (1 out of every 20,000 births) caused by a microdeletion on the long arm of chromosome 7 (7q11.23) that results in the loss of about 16–25 genes (Bellugi, Lichtenberger, Jones, Lai, & St George, 2000; Karmiloff-Smith, 1998; Siegmüller & Bartke, 2004). Physiologically, persons with WS are characterized by a heart condition (supravalvular aortic stenosis) and a facial dysmorphology that makes them look elflike. From the cognitive standpoint, these persons have an Intelligence Quotient (IQ) that usually falls between 40 and 70, and they differ from other typical and atypical populations by their unique neuropsychological profile characterized by an apparent dissociation between cognition and language: language seems to be relatively well-preserved while cognitive activities (especially visuospatial construction, planning, problem solving, and numerical skills) are impaired (Bertrand, Mervis, & Eisenberg, 1997; Karmiloff-Smith et al., 2004; Klein & Mervis, 1999; Pezzini, Vicari, Volterra, Milani, & Ossella, 1999). Another characteristic of WS persons is their hypersociability (Dykens & Rosner, 1999; Gosh & Pankau, 1994; Sarimski, 1997; Tager-Flusberg & Sullivan, 1999), particularly their ease of interaction with unfamiliar persons (Doyle, Bellugi, Korenberg, & Graham, 2003; Frigerio et al., 2006; Jones et al., 2000).

Although WS persons are classically described as having relatively well-preserved structural language abilities and a high degree of sociability, recent research and analyses suggest that their skills are not homogeneous in either of these domains: they are proficient in certain areas of language structure but not in others, and some but not all aspects of sociability are well developed. This new perspective supplied the framework for the present study of French-speaking children and adolescents aimed at examining the pragmatic aspects of language, at the interface between language structure and sociability. It was hypothesized that certain pragmatic skills correspond to the strong points of persons suffering from WS, while others represent their weak points. We labelled the performance of the WS children in terms of “strong points” and “weak points” in order to bring out the nonhomogeneous profile of this population. Note, however, that the “weak” and “strong” labels were always assigned relative to the performance of a comparison group (typical children of the same chronological age or same mental age, children with Down’s syndrome (DS)).

As stated above, research on the structural aspects of WS language (Clahsen & Almazan, 1998; Grant, Valian, & Karmiloff-Smith, 2002; Karmiloff-Smith et al., 1997; Stojanovik, Perkins, & Howard, 2004) indicates a nonhomogeneous profile. For example, the lexical and semantic aspects of language appear to be less impaired than the morphological and syntactic aspects. Studies on morphosyntax in French-speaking participants (Karmiloff-Smith et al., 1997; Monnery, Seigneuric, Zalbar, & Robichon, 2002) point in the same direction: depending on the study and the task, children with WS have varying degrees of difficulty. Research on sociability in English- and Italian-speaking WSs (Doyle et al., 2003; Gagliardi et al., 2003; Jones et al., 2000; Mervis et al., 2003; Tager-Flusberg, Boshart, & Baron-Cohen, 1998; Tager-Flusberg, Plesa-Skwerer, Faja, & Joseph, 2003; Tager-Flusberg & Sullivan, 2000) also points to a nonhomogeneous profile. For instance, studies on social behavior and the expression of emotions indicate that WSs perform relatively well in these two areas while being impaired in the recognition of
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