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Concrete and relational vocabulary: Comparison between Williams and Smith–Magenis syndromes

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ABSTRACT

We compared the performance of two clinical groups, Williams syndrome (WS) and Smith–Magenis syndrome (SMS), in terms of concrete and relational vocabulary. We analyzed (a) whether the WS group had an advantage in concrete vocabulary when compared to the SMS group, as good concrete vocabulary knowledge is considered a hallmark of WS; (b) if spatial processing difficulties in WS would be reflected specifically in their knowledge of relational spatial vocabulary; (c) if a specific vocabulary profile could be outlined for SMS.

Our results show similar performances on receptive concrete and relational vocabulary in both groups. However, and as anticipated, performance on relational space concepts was significantly lower in the WS group.

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1. Introduction

Recent studies have pointed out the necessity of doing cross-syndrome comparisons between neurodevelopmental disorders, rather than relying solely on comparisons with typically developing (TD) samples, as this can better highlight uncommon developmental courses (Brock, 2007; Karmiloff-Smith, 2012; Mervis & John, 2008). Indeed, although comparisons with TD samples provide researchers with information on how atypical some populations are, exclusive contrasts with TD can only offer information on the performance of syndromic populations regarding what is expected according to their chronological age and/or mental age. This is a critical issue, particularly if we consider the different methodological procedures used across studies to measure mental age. In contrast, the comparison of clinical groups may provide a better characterization of such atypical samples in various areas of functioning, including language. This approach offers a stronger view on the cognitive functioning of populations different from what is typically expected, allowing a wider perspective of the human cognitive system. Smith–Magenis syndrome (SMS) and Williams syndrome (WS), two rare disorders of known genetic origin, emerge as relevant natural models for understanding the links between genes and language abilities.

WS is a neurodevelopmental disorder caused by a microdeletion in chromosome 7 in its long arm in the band 11.23 (Ewart et al., 1993) that includes approximately 28 genes (Schubert, 2009). The estimated prevalence of this syndrome is

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approximately 1 in every 7500 births (Strømme, Bjømstad, & Ramstad, 2002). While some propose that WS features a cognitive profile of peaks and valleys, where language is a relatively preserved skill and visual-spatial cognition is greatly impaired, there is no general agreement toward a unique and common neuropsychological profile (Bellugi, Bihrle, Jernigan, Trauner, & Doherty, 1990; Bellugi, Lichtenberger, Jones, Lai, & George, 2000; Jarrold, Baddeley, Hewes, & Phillips, 2001). Some studies found evidence for a superiority of verbal over nonverbal and spatial skills measures (Howlin, Davies, & Udwin, 1998), including longitudinal evidence that verbal abilities develop faster than non verbal ones (Jarrold et al., 2001). This pattern of cognitive functioning, described in children with WS, has also been reported to persist into adulthood, as Performance IQ remains inferior to Verbal IQ (Howlin et al., 1998). Nevertheless, the initial proposal of WS as a genetic model for studying a "language module" independent from cognition, was further challenged and subsequent studies demonstrated that language in WS is impaired along with other cognitive domains (Garayzabal Heinze, Prieto, Sampaio, & Goncalves, 2007; Gonçalves et al., 2004; Rossi, Moretti-Ferreira, & Giacheti, 2006; Sampaio et al., 2008).

Despite the lack of consistent data supporting a preservation of language, there are nevertheless specific strength areas within this domain. For instance, performance on tasks assessing concrete vocabulary, phonological processing and verbal short-term memory seem to be preserved (Bellugi et al., 2000; Bellugi, Sabo, & Vaid, 1988; Mervis & Morris, 2007). In contrast, pragmatic components, abstract vocabulary and reading comprehension are problematic areas (Mervis & Velleman, 2011).

Regarding visual-spatial functioning there is common scientific agreement that skills in this domain are severely impaired. Indeed, many studies have remarked that visual memory, visual perception, mental rotation, spatial visualization, spatial perception and drawing are particularly deficient in WS (Atkinson et al., 2003; Bihrle, Bellugi, Delis, & Marks, 1989; Farran, Jarrold, & Gathercole, 2001; Mandolesi et al., 2009; Wang, Doherty, Rourke, & Bellugi, 1995). Interestingly, little research has been conducted regarding relational vocabulary, where space concepts are integrated. Moreover, research has been mostly limited to spatial language, with studies showing that individuals with WS are more prone to errors in the use of spatial (vs. non-spatial) terms and prepositions (Bellugi et al., 2000; Phillips, Jarrold, Baddeley, Grant, & Karmiloff-Smith, 2004) and show impaired comprehension of spatial language (Laing & Jarrold, 2007). Semel and Rosner (2003) have stressed special difficulties in spatial concepts in WS, but also in temporal and quantitative terms when assessing general language abilities.

Relational concepts are functional words that generally form a finite category; that is, they do not have a proper meaning unless they are embedded within a linguistic context, so they are considered dependent units (Miller, 1999). They are categorized as relational because they link at least two elements, thus providing sense to what it is heard/said. Relational vocabulary is very important to properly establish syntactic relationships between the elements of a sentence, but also to establish relationships between syntax and semantics and to give accurate information, which otherwise could be quite imprecise. Thus, relational concepts have little semantic content and are responsible for the syntactical relationships between the elements that assemble the syntactic structure by extending relations among objects to concept relations, that is, including relations among objects, attributes or concepts (Jackendoff, 2002; Schwanenflugel, 1991). Relational concepts can be organized into several categories such as space, time or quantity.

In the only study that focused on the assessment of relational concepts using a specific task, the Test of Relational Concepts – TRC – Mervis and John (2008) found that relational vocabulary is an area of difficulty in WS. However, when the authors looked for specific differences among the types of relational concepts (e.g. temporal, quantitative, dimensional), performance on spatial concepts was no worse than on the other categories. Despite these interesting results, replication is greatly needed, particularly given previous studies stressing that spatial language, language abilities in general, and non-linguistic spatial cognition walk hand in hand in WS (Laing & Jarrold, 2007; Landau & Hoffman, 2005; Mervis, Robinson, Rowe, Becerra, & Klein-Tasman, 2003).

SMS (Smith, McGavran, Waldstein, & Robinson, 1982) is also a neurodevelopmental disorder caused, in most patients, by a microdeletion in chromosome 17 in its short arm in the band 11.2 that includes the RAI1 gene. Although under diagnosed, SMS has an estimated prevalence of at least 1 in 25,000 births (Greenberg et al., 1991).

To date, relatively little research has been conducted in general about SMS, but even less regarding their language skills. It is known that they have a homogenous cognitive profile with intellectual disabilities (mainly moderate, although some patients reach a mild IQ) where attention, impulsivity, short-term memory, sensorial integration and sequential processing are areas of particular difficulty (Dykens, Finucane, & Gayley, 1997; Osório et al., 2012; Udwin, Webber, & Horn, 2001). Meanwhile, long-term memory, perceptual closure, gestalt and spatial perception are areas of relative competence (Garayzábal-Heinze et al., 2011; Gropman, Duncan, & Smith, 2006; Sarimski, 2004; Udwin et al., 2001; Wolters et al., 2009). Regarding language, it is important to note that persons with SMS display more difficulties with spoken language than with language comprehension (Dykens et al., 1997; Edelman et al., 2007; Gropman et al., 2006; Howlin & Udwin, 2002; Udwin et al., 2001), in part due to ear-nose-throat problems and speech intelligibility, especially during infancy (Solomon, McCullagh, & Krasnewich, 2002; Sonies, Solomon, Ondrey, McCullagh, & Greenberg, 1997; Zalewski, Solomon, McCullagh, & Smith, 2003). To date, few studies have focused on specific language levels in SMS, and most gave an overall view of language in the context of a general cognitive assessment (Dykens et al., 1997; Madduri et al., 2006; Martin, Wolters, & Smith, 2006; Osório, Garayzábal Heinze, Villaverde, & Sampaio, 2013; Udwin et al., 2001). Therefore, little is known regarding specific areas of language, such as concrete vocabulary or relational concepts in SMS. Regarding concrete receptive vocabulary in Spanish patients with SMS, Garayzábal-Heinze et al. (2011) showed important limitations and difficulties concerning comprehension that account for low linguistic competence, but no further studies have assessed this domain.

The syndromes we compare in this study share similar genetic mechanisms, as well as some features of their cognitive, behavioral and linguistic phenotypes and, above all, offer important possibilities to examine the links between specific