



ELSEVIER

Contents lists available at ScienceDirect

Research in Developmental Disabilities



Analysis of speech fluency in Williams syndrome

Natalia Freitas Rossi ^{a,*}, Adriana Sampaio ^b, Óscar F. Gonçalves ^b, Célia Maria Giacheti ^a

^a Department of Speech-Language Pathology, Philosophy and Sciences Faculty, University Estadual Paulista (UNESP), Marília Campus, Avenida Hygino Muzzi Filho, 737. CEP: 17525-900, Marília, São Paulo, Brazil

^b Neuropsychophysiology Lab, CIPsi, School of Psychology, University of Minho, Braga, Portugal

ARTICLE INFO

Article history:

Received 21 April 2011

Received in revised form 22 April 2011

Accepted 3 May 2011

Available online 31 May 2011

Keywords:

Fluency

Oral narrative

Language

Williams syndrome

ABSTRACT

Williams syndrome (WS) is a neurodevelopmental genetic disorder, often referred as being characterized by dissociation between verbal and non-verbal abilities, although the number of studies disputing this proposal is emerging. Indeed, although they have been traditionally reported as displaying increased speech fluency, this topic has not been fully addressed in research. In previous studies carried out with a small group of individuals with WS, we reported speech breakdowns during conversational and autobiographical narratives suggestive of language difficulties. In the current study, we characterized the speech fluency profile using an ecologically based measure – a narrative task (story generation) was collected from a group of individuals with WS ($n = 30$) and typically developing group ($n = 39$) matched in mental age. Oral narratives were elicited using a picture stimulus – the cookie theft picture from Boston Diagnosis Aphasia Test. All narratives were analyzed according to typology and frequency of fluency breakdowns (non-stuttered and stuttered disfluencies). Oral narratives in WS group differed from typically developing group, mainly due to a significant increase in the frequency of disfluencies, particularly in terms of hesitations, repetitions and pauses. This is the first evidence of disfluencies in WS using an ecologically based task (oral narrative task), suggesting that these speech disfluencies may represent a significant marker of language problems in WS.

© 2011 Elsevier Ltd. All rights reserved.

1. Introduction

Williams syndrome (WS) is a neurodevelopmental disorder, with a prevalence of 1 in 7500 (Strømme, Bjørnstad, & Ramstad, 2002), characterized by a deletion in chromosome 7q11.23 (Ewart et al., 1993) comprising a 1.5–1.8Mb and loss of approximately 22 genes in this region (Bayés, Magano, Rivera, Flores, & Péres Jurado, 2003).

Individuals with WS are commonly characterized as displaying an intriguing neurocognitive functioning, described as the “Williams Syndrome Cognitive Profile”, in which a dissociation between cognitive domains, namely between verbal and non-verbal abilities, is described (Atkinson et al., 2001; Grant, Valian, & Karmiloff-Smith, 2002; Jarrold, Baddeley, & Hewes, 1998; Jarrold, Baddeley, Hewes, & Phillips, 2001; Mervis et al., 2000; Pagon, Bennett, LaVeck, Stewart, & Johnson, 1987; Stiles, Sabbadini, Capirci, & Volterra, 2000; Udwin & Yule, 1990). Specifically, the evidence of a superior performance in verbal tasks in WS has been proposed as being the result of an effective contribution of the working memory phonological loop in promoting retention and manipulation of verbal items and thus enhancing syntactic abilities (Jarrold et al., 2001; Nichols et al., 2004; Vicari, Bellucci, & Carlesimo, 2001; Vicari et al., 2004; Volterra, Caselli, Capirci, Tonucci, & Vicari, 2003;

* Corresponding author. Tel.: +55 14 34021324; fax: +55 14 34224797.

E-mail address: nataliafreitasrossi@yahoo.com.br (N.F. Rossi).

Wang & Bellugi, 1994). Additionally, an association between a proficient, well-articulated and grammatically correct speech fluency profile occurring in the context of impaired intellectual disabilities and an overfriendly personality, excessive use of social clichés and tendency to introduce personal experiences out of context, constituted the “Cocktail Party Speech, CPS” (Udwin & Yule, 1990).

However, this notion of spared verbal abilities was challenged and subsequent studies demonstrated that linguistic functions in WS are as impaired as other cognitive domains (Brock, 2007; Karmiloff-Smith, Brown, Grice, & Paterson, 2003; Sampaio, Sousa, Fernandez, Henriques, & Goncalves, 2008). Indeed, the number of studies disputing the hypothesis of verbal–nonverbal dissociation in WS cognitive architecture is growing and, most importantly, contributing to mounting evidence for impaired language abilities in WS, including features traditionally referred as “specific” of WS phenotype, specifically, increased fluency and vocabulary. Indeed, while some studies arguing that individuals with WS perform very well on semantic and phonological fluency tasks (Rossen, Bihrlie, Klima, Bellugi, & Jones, 1996), others have been proving inconsistent results, namely the evidence of similar and/or impaired performances in comparison with mental and chronological-age controls (Grant et al., 2002; Jarrold, Hartley, Phillips, & Baddeley, 2000; Marini, Martelli, Gagliardi, Fabbro, & Borgatti, 2010; Stojanovik & van Ewijk, 2008; Vicari et al., 2004).

Semantic recall tasks are widely used in language and neuropsychological studies particularly because they provide important cues regarding word recovery (semantic memory) taking into account a specific semantic category (e.g., names of animals) or words that begin with the same sound or letter (e.g., say words that begin with F, A or S) (Binetti et al., 1996) within a time limit. The number of lexical items that an individual is able to recall is described as an index of verbal fluency. This measure seems to be more associated with language processing within lexical–semantic domain, and less within syntactic–semantic domain. Thus, performance in standardized tasks requiring semantic recall tends to be less ecologically oriented and provide a limited characterization of verbal fluency phenotype in WS. Therefore, narrative has a considerable value in eliciting speech samples, and thus fluency assessment should include connected speech samples that can be obtained using ecologically based measures occurring naturally in children’s environments, given that narrative abilities serve as indices for assessing several developmental domains (e.g., pragmatics, emotion, cognitive inferences, etc.), and more importantly, can be considered predictors of language development and (Bajaj, 2007). Studies that privileged oral narrative contexts to assess language abilities in WS showed that their narratives are characterized by an excessive use of prosodic markers and audience engaging mechanisms that give the idea that they are truly “natural story tellers” (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Gonçalves et al., 2004), despite impairments in narrative structure, mainly in semantic coherence, have also been described (Goncalves et al., 2010; Gonçalves et al., 2011; Marini et al., 2010; Reilly, Losh, Bellugi, & Wulfeck, 2004). Disfluencies as hesitations and false starts have been interpreted as being part of their difficulties in lexical–semantic access (Karmiloff-Smith et al., 2003; Marini et al., 2010; Reilly et al., 2004), although no formal analysis of their speech fluency profile was formally presented. Preliminary data on speech fluency profile in 12 individuals with WS using a conversational (Rossi, Moretti-Ferreira, & Giacheti, 2007) and autobiographical narrative tasks (Rossi, Souza, Moretti-Ferreira, & Giacheti, 2009) evidenced an increased frequency of hesitations and word repetitions in the WS group when compared with typically developing individuals matched in mental-age, suggesting that these type of disfluencies could be related with their difficulties in lexical–semantic and syntactic access. Indeed, an emerging field of research has been consistently questioning the idea that language is an intact ability in WS (Karmiloff-Smith et al., 2003). However, even though fluency is associated with the cognitive and language profile of this syndrome, the pattern of speech fluency is indeed one of the most remarkable characteristic exhibited by individuals with WS, which has been inclusively used as a differential speech language phenotype marker regarding other genetic conditions such as Down and Fragile X syndromes.

Despite the fact that the “speech fluency” has been associated with one of the phenotypic characteristics of WS, this dimension has been relatively unexplored and studies providing a characterization of the speech fluency profile of WS (considering frequency and typology of disfluencies) were, to the best of our knowledge, not done yet. Thus, an analysis on frequency and typology of speech language in complex linguistic tasks (e.g., narrative) could reveal aspects not only related with discourse organization and planning, but also more related to language itself. Thus the objective of this study is to characterize the speech fluency profile (type and disfluencies frequency) using a narrative task (story generation) in a large group of individuals with WS in comparison with typically developing individuals matched in terms of mental age.

2. Method

2.1. Participants

The participants consist of 30 individuals with WS (15 males and 15 females), positive to *Elastin* gene deletion confirmed by FISH analysis with mean chronological age ranging between 6 and 27 years ($M = 14.70$, $SD = 4.60$) and mental age 4.8–15.1 years ($M = 8.20$, $SD = 3.10$) that were compared with 39 individuals with typically language development (19 males and 20 females, mean chronological age 7.9, $SD = 3.3$), matched in mental age. Intellectual functioning was assessed using Wechsler Intelligence Scales – preschool, children and adult versions (Wechsler, 1989, 1991, 1997) – see Table 1.

The present study was approved by the Ethic Committee of the Faculdade de Medicina de Botucatu – UNESP Campus, the second OF. 256/2006-CEP-FMB-UNESP. Each participant (or their parents) gave written informed consent for their participation in the study via consent forms, after a complete description of the study.

متن کامل مقاله

دریافت فوری ←

ISIArticles

مرجع مقالات تخصصی ایران

- ✓ امکان دانلود نسخه تمام متن مقالات انگلیسی
- ✓ امکان دانلود نسخه ترجمه شده مقالات
- ✓ پذیرش سفارش ترجمه تخصصی
- ✓ امکان جستجو در آرشیو جامعی از صدها موضوع و هزاران مقاله
- ✓ امکان دانلود رایگان ۲ صفحه اول هر مقاله
- ✓ امکان پرداخت اینترنتی با کلیه کارت های عضو شتاب
- ✓ دانلود فوری مقاله پس از پرداخت آنلاین
- ✓ پشتیبانی کامل خرید با بهره مندی از سیستم هوشمند رهگیری سفارشات