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Psycholinguistic abilities of children with Williams syndrome

Natalia F. Rossi [a,1,*], Elena Garayzábal Heinze [b,1,**], Célia M. Giacheti [a], Óscar F. Gonçalves [c], Adriana Sampaio [c]

[a] Department of Speech-Language Pathology, Philosophy and Sciences Faculty, University Estadual Paulista (UNESP), Marília, São Paulo, Brazil
[b] Department of Linguistics, University Autónoma of Madrid, 28049 Madrid, Spain
[c] Neuropsychophysiology Lab, CIPsi, School of Psychology, University of Minho, Braga, Portugal

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ABSTRACT

The objective of this study was to investigate the psycholinguistic abilities of children with Williams syndrome (WS) and typically developing children using the Illinois Test of Psycholinguistic Abilities (ITPA). Performance on the ITPA was analysed in a group with WS ($N = 20$, mean age = 8.5 years, SD = 1.62) and two typically developing groups, matched in mental (MA, $N = 20$, mean age = 4.92 years, SD = 1.14) and chronological age (CA, $N = 19$, mean age = 8.35 years, SD = 3.07). Overall, within-group analyses showed that individuals with WS displayed higher scalar scores on the visual reception and visual association subtests. When groups were compared, we observed inferior performance of the WS group on all ITPA subtests when compared with typically developing groups. Moreover, an interaction between reception and group was found, only the WS group demonstrated superior performance on the visual reception subtest when compared to the auditory reception subtest. Evidence from this study offers relevant contributions to the development of educational intervention programs for children with WS.

1. Introduction

Williams syndrome (WS) is a neurodevelopmental disorder characterised by a 1.5 Mb microdeletion on chromosome 7q11.23 that includes approximately 28 genes (Schubert, 2009). This genetic condition exhibits a cognitive profile characterised by a pattern of peaks and valleys in terms of cognitive abilities (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000). Specifically, the initial reports of WS document intellectual disability and profound impairment in visuospatial processing coupled with relatively good language abilities. Interestingly, much of the research interest in WS was fostered by this apparent dissociative pattern of neurodevelopment (Bellugi, Bihrle, Jernigan, Trauner, & Doherty, 1990), although these initial reports of excellent performance in language have been questioned (Brock, 2007; Cherniske et al., 2004; Gonçalves et al., 2004; Greer, Brown, Pai, Choudry, & Klein, 1997; Karmiloff-Smith, Brown, Grice, & Paterson, 2003; Porter & Coltheart, 2005; Sampaio et al., 2009; Stojanovik, Perkins, & Howard, 2006).

Intellectual disability is another feature of WS. Several studies use different measures of global intelligence to point out a high prevalence of intellectual disability; scores of children with WS generally fall in the interval Full Scale IQ ranging from 55 to 70 (Martens, Wilson, & Reutens, 2008, for a general review). This global intellectual functioning is associated with a

* Corresponding author at: Philosophy and Sciences Faculty, University Estadual Paulista (UNESP), Avenida Hygino Muzzi Filho, 737, CEP: 17525-900 Marília, São Paulo, Brazil.
** Corresponding author.
E-mail addresses: nataliafreitasrossi@yahoo.com.br (N.F. Rossi), elena.garayzabal@uam.es (E. Garayzábal Heinze).
[1] Share equal first authorship.

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heterogeneous profile (Porter & Coltheart, 2005), and a Williams Syndrome Cognitive Profile (Mervis et al., 2000; Pani, Mervis, & Robinson, 1999) that takes performance on specific subtests into account has been proposed.

While both overall cognitive measures and specific tests to assess verbal and nonverbal developmental trajectories have been used extensively in characterising the WS cognitive profile (Jarrold, Baddeley, & Hewes, 1998; Jarrold, Hartley, Phillips, & Baddeley, 2000), few studies have provided a detailed characterisation of psycholinguistic abilities that takes both visual and auditory information processing skills into account.

Performance on the Illinois Test of Psycholinguistic Abilities (ITPA) (Kirk & McCarthy, 1961) has provided inconsistent information about the relative impairment of visual and auditory processing in children with WS (Crisco, Dobbs, & Mulhern, 1988; Gejão et al., 2007; Nakamura et al., 1999). According to Crisco et al., visual processing abilities as measured by the ITPA were significantly different in children with WS when compared with those of a control group. Specifically, individuals with WS demonstrated more difficulty performing visual reception, visual closure and visual memory tasks; visual processing abilities were generally below their cognitive level. However, Nakamura et al. (1999) described a single WS case in which scores for both the auditory and visual processing subtests were equally below normal. Finally, a Brazilian case report in which the ITPA was used to monitor language acquisition in children with WS showed that visual performance on ITPA subtests was within the normal range while cognitive difficulties were more prominent in auditory subtests (Gejão et al., 2007).

Taking into account both the inconsistent findings regarding auditory and visual information processing and the limited number of studies using the ITPA to assess cognitive abilities in children with WS, the main objective of this study was to compare psycholinguistic abilities measured using the ITPA of a group of individuals with WS (with confirmed fluorescence in situ hybridization – FISH) with those of two typically developing groups (a chronological age-matched group and a mental age-matched group).

2. Materials and methods

2.1. Participants

The Ethical Board (no. 256/2006) approved the current study. Written informed consent was obtained from all participating individuals and/or their parents prior to enrolment. The WS group included 20 individuals with confirmed genetic diagnoses (Ewart et al., 1993), with a mean age of 8.5 years (SD = 1.62 years, participant ages ranged from 5.5 years to 10.75 years) and a mean Full Scale IQ of 56.72 (SD = 10.08, range 42–75). Individuals with WS were recruited from the Brazilian Williams Syndrome Association (ABSW, $N = 12$) and from the Spanish Williams Syndrome Association (ASWE, $N = 8$). Typically developing groups consisted of a chronological age-matched group (CA, $N = 19$), with a mean age of 8.35 years (SD = 1.75; FSIQ = 96.58, SD = 2.22), and a mental age-matched group (MA, $N = 20$), with a mean age of 4.92 years (SD = 1.14; FSIQ = 97.05, SD = 3.07). Spanish and Brazilian participants in the CA and MA groups were typically developing individuals with no evidence of speech-language disorders, learning disabilities, hearing loss, or visual impairment. Mental age for the MA group was derived for each participant according to Full Scale IQ (Terman, 1916). Finally, all participants had normal hearing assessed through behavioural audiometry and/or threshold tonal audiometry as well as normal or corrected-to-normal vision. Two participants with auditory thresholds below 25dBNSat several frequencies were excluded from this study (Davis & Silverman, 1970). Demographic characteristics of the groups are shown in Table 1.

2.2. Instruments

To assess the general cognitive functioning level (Full Scale IQ – FSIQ), the Wechsler Preschool and Primary Scale of Intelligence-R (WPPSI-R) and Wechsler Intelligence Scale for Children-Third Edition (WISC-III) were used for children between 3–6 years of age and 6–11 years of age, respectively (Wechsler, 1989, 1991).

The Brazilian (Bogossian & Santos, 1977) and Spanish versions (Kirk, McCarthy, & Kirk, 1986) of the Illinois Test of Psycholinguistic Abilities (ITPA) (Kirk & McCarthy, 1961) were used in the present study. This test assesses psycholinguistic abilities that are important for communication, namely those related to the ability to understand, process, and relate visually and aurally presented stimuli. The ITPA model was developed using a language-learning model proposed by Kirk that consists of two primary input channels for communication (auditory and visual), two primary output channels (verbal and motor), three psycholinguistic processes (reception, association and expression) and two organisation levels (automatic and

Table 1
Demographic characteristics of the ws and typically developing groups.

Group	WS ($N = 20$)	CA ($N = 19$)	MA ($N = 20$)
	M (SD)	*M* (SD)	*M* (SD)
Age (mean, SD)	8.60 (1.62)	8.35 (1.75)	4.92 (3.07)
FSIQ	56.72 (10.08)	96.58 (2.22)	97.05 (3.07)
Male/female	10/10	10/9	10/10