



Williams syndrome: A surprising deficit in oromotor praxis in a population with proficient language production

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ABSTRACT

Williams Syndrome (WS) is a neurodevelopmental disorder of known genetic origin, characterized by serious delays in language onset yet relatively verbose, intelligible and fluent speech in late childhood and adulthood. How do motor abilities relate to language in this group? We investigated planning and co-ordination of the movement of the speech articulators (oromotor praxis) in 28 fluent-speaking individuals with WS, aged between 12 and 30 years. Results indicate that, despite their fluent language, oromotor praxis was impaired in WS relative to two groups of typically-developing children, matched on either vocabulary or visuospatial ability. These findings suggest that the ability to plan, co-ordinate and execute complex sensorimotor movements contribute to an explanation of the delay in expressive language early in development in this neurodevelopmental disorder. In the discussion, we turn to more general issues of how individual variation in oromotor praxis may account for differences in speech/language production abilities across developmental language disorders.

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1. Introduction

Williams Syndrome (WS) is a neurodevelopmental disorder caused by a hemizygous submicroscopic deletion of some 28 contiguous genes on chromosome 7q11 ± 23 (Ewart et al., 1993; Donnai and Karmiloff-Smith, 2000). Although original estimates of the prevalence of WS were around 1:20,000 (Kaplan et al., 2001; Morris et al., 1988; Donnai and Karmiloff-Smith, 2000), a more recent study rates prevalence at close to 1:7,500 (Strømme et al., 2002). WS is associated with cardiac problems, distinctive facial morphology and slow physical growth. The linguistic profile of individuals with WS is typified by relatively verbose, fluent speech from late childhood onwards, a characteristic all the more striking given the fairly considerable delay in language development over infancy and toddlerhood (Singer-Harris et al., 1997; Paterson et al., 1999). However, very little is known about oromotor praxis (that is, the ability to plan and co-ordinate movements of the speech articulators) in WS, a motor ability that is particularly important for speech and language development. In the current study, we investigate oromotor praxis in a group of 12–30-year-olds with WS to establish whether oromotor ability is typical or atypical in this unusual neurodevelopmental disorder.

We first review studies that indicate that expressive language is a relative strength in the cognitive profile of WS, we then discuss why motor skills may be relevant to language development in this group, and finally, we outline the measures used in the present study.

1.1. Expressive language in WS

Individuals with WS present with an uneven and unusual cognitive profile. In adulthood, the language abilities of individuals with WS are usually better than their spatial cognition skills (Donnai and Karmiloff-Smith, 2000; Jarrold et al., 1998). Udwin and Yule (1990) studied conversational exchanges of 43 school-age children with WS. Eighty-four percent of these children were classified as having fluent, articulate speech. In a direct comparison of children with WS to children with specific language impairment (SLI) or Down Syndrome (DS), Laws and Bishop (2004) found that children with WS between the ages of 6–15 years outperformed the other two disorder groups on the speech sub-scale of the Childhood Communication Checklist. Other evidence for expressive language strength comes from studies of oral narrative production in WS, where the stories of individuals with Williams syndrome were more descriptive and engaging than the stories of those with DS (Reilly et al., 1990).

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Within language, phonological skill generally, and phonological short-term memory more specifically, are considered to be strengths in WS (Vicari et al., 1996; Nichols et al., 2004). Relative to children with DS, children with WS performed better on tasks relying on phonological short-term memory such as digit span (Wang and Bellugi, 1994; Jarrold et al., 1999), word span (Vicari et al., 2004), or verbal repetition (Vicari et al., 2002). This was the case despite similar or poorer performance by individuals with WS on visuo-spatial memory tasks such as the Corsi block span. While reliance on phonology was originally hypothesized to be unusual in WS (Vicari et al., 1996), more recent studies suggest that phonology is a relative strength but not atypical (Majerus et al., 2003; also reviewed in Brock, 2007). Indeed, phonological abilities are comparable to those of typically developing children matched on verbal or nonverbal skill (Brock, 2005; Grant et al., 1997; Laing et al., 2005).

Despite later strengths in expressive language, during the infant and toddler years, individuals with WS present with very clear delays in language development. The onset of the first words is delayed in infants with WS and tends to occur between 18 and 24 months of age (Masataka, 2001). Parental questionnaires indicate that infants with WS have similar levels of word production and comprehension as infants with DS (Singer-Harris et al., 1997). Furthermore, they produce fewer manual gestures (such as pointing) than infants with DS (Laing et al., 2002; Singer-Harris et al., 1997). In experimental studies, infants with WS have shorter looking times to named objects relative to chronological age-matched controls and their performance resembles that of children with DS (Paterson et al., 1999). Nazzi et al. (2003) observed that although infants with WS could segment words with a strong-weak stress pattern in fluent speech, they were delayed when they had to extract words with a weak-strong stress pattern from fluent speech. Therefore, it is clear that infants with WS have early delays in lexical and phonological development. Delays in abilities relevant to language continue at later stages of development, for example, toddlers with WS are impaired in triadic joint interaction as well as comprehension and production of pointing (Laing et al., 2002). Differences in language development are observed even in the preschool years, for instance, preschoolers with WS are slower at word learning than their typically developing peers (Havy et al., 2010). Vicari et al. (2004) show that the strengths in receptive vocabulary and sentence repetition typically associated with WS only emerge by late childhood/ adolescence. It remains unclear why these initial delays arise in language development and how children with WS overcome them to become relatively proficient language producers later in development.

1.2. *Links between language and motor abilities*

In other neurodevelopmental disorders where speech and language deficits have been identified, concomitant motor difficulties are frequently observed. For example, Brookman et al. (2013) have reported poorer imitation of body postures and hand movements in SLI (also see Hill, 2001). Fine motor ability in the early years has been found to predict later speech fluency in children with autism (Gernsbacher et al., 2008; LeBarton and Iverson, 2013). Leonard and Hill (2014) have suggested that genetic disorders like Williams syndrome offer an opportunity to understand relationships between motor and language abilities through the lifespan. Yet, in contrast to the increasing literature on motor abilities in behaviourally-defined developmental disorders like autism (Torres et al., 2013), relatively little is known about oromotor abilities in WS.

A motor ability that we refer to as 'oromotor praxis' is an index of an individual's ability to imitate and sequence complex oral movements. Oromotor praxis relates to language development at

ages beyond the measures of motor control taken in infancy. In typically developing children of around 21 months of age, oromotor praxis is associated with scores on language production, comprehension and grammatical complexity (Alcock and Krawczyk, 2010). Further, our own research has identified links between oromotor praxis and nonword repetition, one which lasts through the school years (Krishnan et al., 2013a) and suggests that this relationship taps into the reliance of both tasks on planning and coordinating oral movements. Even in atypically developing children, oromotor praxis appears to be associated with language outcomes. For instance, a link between oromotor praxis and phonological skill is seen in specific language impairment (Stark and Blackwell, 1997) and Elliott et al. (1990) report deficits of oromotor praxis in DS. Given that speech fluency is considered a characteristic strength in this neurodevelopmental disorder (Rossi et al., 2011), it is of particular interest to establish whether oromotor praxis ability relates to verbal ability in WS. In particular, this would allow us to explore whether the emergence of relatively good oromotor skills could influence the improvement in language proficiency.

While a handful of studies indicate that infant motor milestones are delayed in WS (Lenhoff et al., 1997; Masataka, 2001; Tsai et al., 2008), very little is known about speech motor ability or oromotor praxis in children, adolescents and adults with WS. To date, one unpublished study indicates that fine motor control of the speech articulators is affected (Mervis and Velleman, 2011). However, it has not been established if oromotor ability is related to the strengths in verbal ability. As the discrepancy between verbal and visuospatial ability only appears to develop over time (Vicari et al., 2004), it is possible that strengths in oromotor ability may only be apparent at the same time or slightly earlier than strengths in verbal ability. Furthermore, strengths in oromotor praxis may only emerge over time. As children with WS have a proclivity for social interaction, their interest in conversation may lead them to imitate words and sentences more than other children with developmental disorders. It is plausible that greater experience producing speech (relative to other children with neurodevelopmental disorders) could contribute to the improvement in oromotor praxis, as children gain increased practice with sequencing and coordinating articulators to produce sounds and words in their own language. Additionally, developmental improvements in phonological proficiency might also shape and change oromotor co-ordination for speech. Currently, it is not known what levels of oromotor ability individuals with WS attain by the time verbal strengths are apparent. In addition to the previously described strengths in spoken language, speech fluency is a characteristic strength in this group and this strength is apparent by relatively early childhood (Rossi et al., 2011). Therefore, in the current study, we have focused on oromotor skills in older individuals with WS who would be likely to show the relative strengths in spoken language and speech fluency. Consequently, we expected to see concomitant strengths in oromotor skills for individuals with WS with verbal mental ages approximating those of 7–12 year olds.

1.3. *The present study*

In this study, we compare oromotor praxis in individuals with WS to both vocabulary age-matched and visuospatial age-matched controls. Given reported strengths in speech fluency in childhood in WS, we expected that oromotor praxis would be at a par with typically developing children of similar verbal ability and better than typically developing with similar visuospatial ability.

In addition to the comparison of oromotor praxis across groups, we explore whether potential group differences will be reflected across other manual, oral and verbal tasks (visuomotor imitation,

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