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Children with Williams Syndrome: Language, Cognitive, and Behavioral Characteristics and their Implications for Intervention

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Abstract

Williams syndrome (WS) is a rare genetic disorder characterized by heart disease, failure to thrive, hearing loss, intellectual or learning disability, speech and language delay, gregariousness, and non-social anxiety. The WS psycholinguistic profile is complex, including relative strengths in concrete vocabulary, phonological processing, and verbal short-term memory and relative weaknesses in relational/conceptual language, reading comprehension, and pragmatics. Many children evidence difficulties with finiteness marking and complex grammatical constructions. Speech-language intervention, support, and advocacy are crucial.

Williams syndrome (WS) is a rare neurodevelopmental disorder caused by a microdeletion of ~26 genes on the long arm of chromosome 7 (7q11.23) (Osborne, 2012). The prevalence of this syndrome has been estimated as 1 in 7500 live births (Strømme et al., 2002). WS is associated with a characteristic facial appearance (see Figure 1), congenital heart disease, connective tissue abnormalities, and failure to thrive or growth deficiency (Morris, 2006). Recurrent otitis media is very common among young children with WS (Morris, 2006) and unilateral or bilateral mild to moderate high-frequency hearing loss is common among school-aged children, with parents often unaware that their child has hearing loss (Marler et al., 2010). Sensory modulation problems, including difficulty with auditory filtering and hypersensitivity to sound, also are very common (John & Mervis, 2010). Young children with WS have developmental delay and older children have intellectual or learning disabilities although there is a broad range of intellectual levels, from severe intellectual disability to average for the general population. WS also is associated with specific cognitive and personality/behavioral phenotypes, as described below.

Over the past two decades, WS has been featured in the public media on several occasions. The descriptions provided are typically paradoxical; individuals with WS are often described as evidencing near-normal language abilities in the face of severe intellectual disability and as being extremely sociable at the same time as evidencing limited understanding of social norms (Mervis & John, 2010a, 2010b). Research studies have provided a more nuanced portrayal of the cognitive, language, and behavioral characteristics associated with this syndrome. Below we provide a brief overview of these findings; more detailed information is provided in Mervis (2009), Mervis and Becerra (2007), Mervis and John (2010a, 2010b), Morris, Lenhoff, and Wang (2006), and Farran and Karmiloff-Smith (2012). We then provide a brief discussion of the language intervention needs for children with WS and strategies for addressing them.

Cognitive Profile

Although the average full-scale IQ for individuals with WS is in the mild intellectual disability range (Martens, Wilson, & Reutens, 2008), this overall score masks a striking pattern of relative strengths and weaknesses, best illustrated by standard scores (SSs) on the Differential Ability Scales-II (DAS-II; Elliott, 2007). Mervis and John (2010a) reported SSs for 120 children with WS. Although the mean General Conceptual Ability (GCA; similar to IQ) is in the mild intellectual disability range (65, range: 31 – 96), the mean Verbal Cluster SS (74, range: 30 – 111) and the mean Nonverbal Reasoning Cluster SS (79, range: 37 – 118) are in the borderline to low average range as is mean performance on verbal short-term memory (72, range: 40 – 102). In strong contrast, the mean Spatial Cluster SS (53, range: 32 – 81) is in the moderate intellectual disability range. About 85% of children with WS evidence a significant discrepancy between their Verbal and/or Nonverbal Reasoning Cluster SSs and their Spatial cluster SS, indicating that their intellectual abilities are not best represented by a single overall score (GCA or IQ). Instead, each of their Cluster SSs should be considered separately. Unfortunately, this pattern of strengths and weaknesses is much harder to detect on the Wechsler tests [e.g., Wechsler Intelligence Scale for Children-IV (WISC-IV)], the intellectual ability tests school psychologists most commonly use.

Behavioral Profile

The paradoxical nature of the social, personality, and psychopathology characteristics of children with WS was highlighted in one of the first publications on this syndrome. von Arnim and Engel (1964, p. 376) noted that individuals with WS, "... have a great ability to establish interpersonal contacts" but that this ability "stands against a background of insecurity and anxiety." Klein-Tasman and Mervis (2003) captured this paradox in their empirically-derived Williams Syndrome Personality Profile (WSPP) based on the Multidimensional Personality Questionnaire (MPQ; Tellegen, 1985). Parental ratings on five characteristics differentiated a group of 8 – 10-year-olds with WS from an age and IQ-matched group of children with other developmental disabilities, successfully classifying 96% of the children in the WS group and 85% of the children in the comparison group. Of these characteristics, three (gregarious, people-oriented, visible) fit with the first part of von Arnim and Engel's statement, and two (tense, sensitive) with the second. More recently, the WSPP has been shown to characterize >90% of an additional sample of 100 individuals with WS aged 5 – 20 years (Mervis & John, 2010b).

Consistent with the first three MPQ characteristics, parents of children with WS commonly report that their child never showed stranger anxiety, that he or she is widely known and well liked in their community, and that their child's smile can "light up a room." Children with WS often use these same characteristics to avoid having to put forth effort on tasks that they find difficult or frustrating (Järvinen-Parsley et al., 2008). When asked to complete a challenging task, the majority of children with WS will try to distract the adult or engage with him/her socially rather than attempting to complete the task (Mervis & John, 2010a). In the one study that addressed mastery motivation of children with WS, preschoolers with WS were found to have significantly lower mastery motivation than did age-matched children with Down syndrome (DS), even though the children with DS had significantly lower intellectual abilities. In particular, the children with WS evidenced more help-seeking behavior and less task-related behavior (Mervis & John, 2010a).

Consistent with the last two characteristics of the WSPP, the majority of children with WS evidence considerable non-social anxiety (Leyfer, Woodruff-Borden, Klein-Tasman, Fricke, & Mervis, 2006), with 54% of a sample of 119 4 – 16-year-olds meeting full criteria for a DSM-IV diagnosis of Specific Phobia (most often of loud noises; doctor/dentist and/or

blood/injection specific phobias also are common). In addition, 14% of the 7 – 10-year-olds and 23% of the 11 – 16-year-olds met DSM-IV criteria for Generalized Anxiety Disorder (GAD). Leyfer et al. argued, based on clinical experience, that the DSM-IV criteria for GAD often do not capture the nature of worrying in children with WS. In particular, children with WS worry in anticipation of (and repeatedly inquire about) both events they are looking forward to and events they expect to dislike. In addition, many children with WS are irritable and often overreact to what should be minor frustrations (Phillips & Klein-Tasman, 2009).

Attention Deficit Hyperactivity Disorder (ADHD) is also common. Leyfer et al. (2006) found that 65% of the 119 children in their study met DSM-IV criteria for ADHD. As age increases, children with WS are more likely to be diagnosed with ADHD-Predominantly Inattentive and less likely to be diagnosed with ADHD-Combined. Girls with WS are as likely to be diagnosed with ADHD as are boys.

Language

Although language is typically considered a relative strength for children with WS, there is still a clear pattern of relative strengths and weaknesses within that domain. Briefly summarized, concrete vocabulary and phonological skills are relative strengths, grammatical abilities are at the level expected for overall intellectual abilities, and relational language and pragmatics are clear weaknesses. Below we first briefly discuss the early language development of children with WS and then consider the pattern of language strengths and weaknesses of preschool and school-age children with WS.

Early Language Development

The onset of language development is nearly always delayed for children with WS. Mervis et al. (2003) used the MacArthur-Bates Communicative Development Inventory (CDI; Fenson et al., 1993, 2007) to track the early vocabularies of 13 children with WS. Age at acquisition of a 10-word expressive vocabulary was below the 5th percentile for all 13 children; age at acquisition of 50- and 100-word expressive vocabularies was below the 5th percentile for 12 of 13. Median age of acquisition of a 100-word expressive vocabulary was 37 months (range: 26 – 68 months) (Mervis & John, 2012). For a group of 22 children with WS, age at acquisition of a 10-word expressive vocabulary was highly correlated with age at acquisition of 50- and 100-word vocabularies and the onset of novel word combinations, and all of these language measures were strongly related to both Verbal IQ and Nonverbal IQ at age 4 years (Mervis & John, 2012).

For typically developing children, children with DS, and children with severe developmental delay of unknown etiology, the comprehension and production of referential pointing gestures and engagement in triadic joint attention also precede the onset of referential expressive language (Mervis & Bertrand, 1997). This pattern does not hold for children with WS; for most children with WS, the onset of referential language precedes the onset of referential pointing by several months (Mervis & Bertrand, 1997). The absence of triadic joint attention is consistently raised as a concern by parents of toddlers with WS (Mervis & John, 2012). Mervis and Becerra (2007) describe several alternative strategies that the communicative partners of children with WS use to establish joint focus on an object.

Speech Development

Masataka (2001) argued that the late onset of language is due in part to motor delays, specifically a delay in onset of rhythmic hand banging, which provides the motor substrate for canonical babble. He found a strong correlation between age at onset of rhythmic hand banging and canonical babble and between the onset of canonical babble (mean age of onset

in his study: 17.7 months) and a 25-word expressive vocabulary for children with WS (mean age: 22.9 months). In a study of 6 prelinguistic children with WS followed longitudinally for varied periods of time (age range 9–39 months), Velleman, Currier, Caron, Curley, and Mervis (2006) also found delays in vocal development. In particular, the number of different consonants produced per ½-hour session and the average number of syllables per utterance were consistently below age expectations.

Between the ages of 4 and 12 years, children with WS continue to demonstrate features of motor speech disorders. Of 33 children with WS studied by Huffman, Velleman, and Mervis (2011), only one scored within normal limits on all four main speech subscores of the Verbal Motor Production Assessment for Children (Hayden & Square, 1999). General Motor Control was most often most affected (85% severe), probably due to low muscle tone. Elastin deficits as well as low muscle tone were the likely sources of deviant Speech Characteristics (especially, harsh vocal quality), which affected more than half of the children. However, motor control and planning were also affected, with 50% and 82% receiving scores reflecting at least mild deficits on Focal Oral Motor Control and Sequencing, respectively. Focal Oral Motor Control and Sequencing scores rose steadily with age in this cross-sectional study; General Motor Control and Speech Characteristics remained static. Despite ongoing notable distortions and atypical voice quality, most school-age children with WS are quite intelligible in context.

Vocabulary Development

Within the language domain, concrete vocabulary (names of objects, actions, and descriptors) has consistently been identified as the area of greatest strength. Mean SSs for both the Peabody Picture Vocabulary Test-4 (PPVT-4; Dunn & Dunn, 2007) and the Expressive Vocabulary Test-2 (EVT-2; Williams, 2007) are in the low average range for the general population (PPVT-4: 82, EVT-2: 79). In fact, >80% of children with WS earn SSs above 70 on the PPVT-4 and EVT-2 and 6 – 8% score at or above 100 (Mervis & John, 2010a). Phonological memory (nonword repetition), which is also a relative strength for children with WS, is significantly related to concrete vocabulary ability (Grant et al., 1997).

In strong contrast, conceptual/relational vocabulary is an area of considerable weakness. Performance on the Test of Relational Concepts (TRC; Edmonston & Litchfield-Thane, 1988), which measures receptive knowledge of spatial, quantitative, temporal, and dimensional concepts) averages ~30 points lower than performance on the PPVT (Mervis & John, 2008) for children with WS aged 5 – 7 years. These difficulties with relational language continue as children get older; the modal (most frequent) scaled score for a group of 9 – 11-year-olds on the Formulated Sentences subtest of the Clinical Evaluation of Language Fundamentals-4 (CELF-4; Semel et al., 2003) was 1 (the lowest possible), with a large proportion of the points lost coming from the relational concept items (e.g., “unless,” “although”). Children’s performance on the TRC was strongly related to their performance on the CELF-4 Formulated Sentences subtest four years later (Mervis & John, 2008).

Grammatical Development

The initial claims that individuals with WS evidenced near-normal grammatical abilities and that these abilities were well above what would be expected given their significant intellectual disabilities (Bellugi, Marks, Bihrie, & Sabo, 1988) were based on comparisons with age- and IQ-matched adolescents with DS. More recent research has confirmed that the grammatical abilities of children with WS are consistently above those of matched children with DS. However, the grammatical abilities of children with WS are about at the level expected for their overall intellectual abilities, and many individuals evidence considerable difficulty with complex grammatical constructions (e.g., Karmiloff-Smith et al., 1997).

Grammatical ability is more strongly correlated with verbal working memory ability for children with WS than for typically developing children (Mervis & John, 2010a).

The morphological abilities of children with WS are at or slightly below the level expected for their overall intellectual abilities (see review in Mervis & Becerra, 2007). Many children with WS evidence difficulties with finiteness marking. For example, Peregrine et al. (2006) found that many 6 – 11-year-olds did not reliably mark the third-person singular present and/or the past tense. Difficulty with correct use of BE and/or DO in questions was quite common. Typical errors included using intonation rather than grammar to mark a question, using BE rather than DO (e.g., “Is the bug want cookies?”), or double-marking (e.g., “Is the bug is hungry?”). Over-regularization of the past tense (e.g., “eated”) is also common.

Pragmatics Development

Despite their sociable, outgoing nature, pragmatics is an area of considerable weakness for children with WS. In fact, although WS is often contrasted with autism spectrum disorders (ASDs) in the popular press (Mervis & John, 2010a), many of the pragmatic difficulties evidenced by children with WS are similar to those of children with ASDs. For example, when preschoolers with WS who had limited language were administered the Autism Diagnostic Observation Schedule-Generic (ADOS-G; Lord et al., 2000), 48% were classified by the ADOS-G algorithm as “autism spectrum disorder” or “autism” (Klein-Tasman et al., 2007). Common problems included difficulty integrating eye contact to show an object or to request an object that was out of reach. Only a few children spontaneously used a doll or other object as an independent agent or used one object to represent another. Note, however, that most of the children would not meet the clinical-judgment standard for ASDs because they also showed important strengths that are inconsistent with ASDs. For example, the quality of the children’s social overtures was generally good and few children had difficulty sharing affect or directing facial expressions or vocalizations to other people.

These pragmatic difficulties continue into the school years. Findings from studies using parental responses to the Children Communication Checklist (e.g., CCC-2; Bishop, 2002) indicated considerable difficulties. Laws and Bishop (2004) found that the WS group evidenced significant difficulties in all areas of pragmatics measured by the CCC, with 79% meeting the CCC cut-off for pragmatic language impairment. Philofsky et al. (2007) found that although children with WS scored significantly higher than age-matched children with autism on the CCC-2 Stereotyped Language and Nonverbal Communication scales, the two groups performed equally poorly on the Inappropriate Initiation and Use of Context scales.

Significant conversational difficulties also have been identified. Udwin and Yule (1990) reported that 37% of the children with WS in their study met criteria for hyperverbal speech (excessive use of stereotyped phrases, over-familiarity, introduction of irrelevant experiences, perseverative responding). Stojanovik (2006) found that during a semi-structured conversation, when the researcher asked for information or clarification, the responses of the children with WS were less likely to be adequate than were the responses of children with Specific Language Impairment matched for receptive vocabulary and grammatical ability. The most common difficulties were providing too little information, misinterpreting what the researcher had said, or not producing a response that continued the conversation. John et al. (2009) found that in a referential communication task, when the researcher’s instructions were inadequate the children with WS verbally indicated there was a problem <50% of the time and most of these verbalizations either were too vague to communicate the nature of the problem or indicated the wrong problem. Performance was related both to age and to first-order theory of mind, an ability that is considerably delayed for children with WS.

Literacy

Studies of the reading skills of older children and adolescents with WS have demonstrated a wide range of abilities (see review in Mervis, 2009), with some individuals not able to read at all and others both decoding and comprehending at grade level. For a sample of 44 children aged 9 – 17 years, Mervis (2009) reported mean SSs of 73 (range: 40 [lowest possible] – 112) for Word Reading, 79 (range: 0 correct – 113 SS) for Pseudoword Decoding, and 65 (range: 40 [lowest possible] – 102) for Reading Comprehension on the Wechsler Individual Achievement Test-II (WIAT-II; Wechsler, 2005). Thus, as expected given the WS language profile, decoding abilities are considerably stronger than reading comprehension abilities relative to general-population expectations. Performance on the Comprehensive Test of Phonological Processing (CTOPP; Wagner et al., 1999) is typically in the borderline to low average range and correlates well with single word reading and pseudoword reading (Levy & Hermon, 2003). Given the general pattern of relative strengths and weaknesses in the language domain, one would expect that an intensive phonics approach to reading would be more effective than a sight-word/whole-word or whole language approach for children with WS. This finding has been consistently supported (Mervis, 2009).

Intervention

Given the delays in language onset and the continuing difficulties over time especially in relational language, and pragmatics, speech/language therapy is critical for children with WS. In this section we begin with a brief consideration of the infant/toddler period and then discuss the preschool/school-age period.

Early Intervention

Given the finding that age at acquisition of 10- and 50-word expressive vocabularies is strongly correlated both with the onset of novel word combinations and with Verbal and Nonverbal IQ at age 4 years, it is critical that all older infants and toddlers receive speech therapy, with the intensity determined by the child's level of delay. It is very common for developmental therapists to use the onset of referential communicative gestures as the primary indicator that a child would benefit from language therapy. This assumption is highly problematic for children with WS, who typically begin to produce referential language prior to producing referential communicative gestures. Similarly, if a child with WS is not referred to an early intervention program until he or she is already talking, the presumption is often made that basic referential gestures have been mastered. This assumption is also problematic; it is likely that even toddlers who are talking well do not have pragmatic abilities at the level expected for their vocabulary skills. Thus, careful assessment (including extensive observation of the child interacting with his/her caregivers and other children) is necessary to determine the child's language intervention needs.

Because children with WS often have failure to thrive, many receive feeding therapy, often from an SLP. This therapy is critical but in addition, the child needs to receive language therapy at the same rate and intensity as he/she would have received it had feeding therapy not been needed. Appropriate non-speech oral-motor therapy has its function for children with dysphagia, but there is no evidence that it will positively impact speech production (Watson & Lof, 2008). With respect to speech production, special attention should be paid to consonant variety and the production of words that are more than one syllable in length. Basic sign language is often useful for launching early conventional communication when speech is slow to develop. (See Davis & Velleman, 2008 for further suggestions about stimulating early speech.)

Intervention for Preschool and School-Age Children

For preschool and school-age children, a full assessment of all aspects of language and communication, including extensive observation of the child's interactions with teachers and peers in both formal and informal settings, is critical to determining if the child would benefit from language therapy and the level of intensity needed. Frequently, once the child's speech is understandable and he/she no longer makes consistent grammatical errors, speech/language therapy is discontinued, even though the child continues to have considerable difficulty with both conceptual/relational language and pragmatics. Both of these are areas for which effective formal assessments, especially for older children, are not available, leading to a lack of formal documentation. Nevertheless, these areas are critical for both academic performance and peer relationships. In addition, difficulties with metalinguistics and inference-making lead to further problems (e.g., with reading comprehension; in determining whether one understood what the teacher said and if one did not, in being able to formulate an effective question about it; in working with other students on a team project) for which nearly all children with WS would benefit from language therapy. Further therapy recommendations are provided by Mervis and John (2010a) for pragmatics and social skills and by Mervis (2009) for reading comprehension.

Conclusion

The very strengths demonstrated by children with WS – especially certain interpersonal skills and the ability to learn concrete vocabulary – may mislead the SLP about the children's profiles and needs. This misperception may be exacerbated by the facts that overall IQ scores mask uneven patterns of strengths and weaknesses and that developmental milestones, especially the relation between referential gestures and the onset of referential language, may not proceed in the typical order. Early communication therapy is vital for getting children started in the right direction. Ongoing intervention emphasizing relational language, pragmatics, grammar, phonics, and reading comprehension will maximize the ability of children with WS to participate in and benefit from educational opportunities. Thus, the SLP's role in treating this population and in elucidating the strengths and weaknesses of children with WS for other members of academic and intervention teams is critical.

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Figure 1.
Three children who have Williams syndrome, aged 3 years, 6 years, and 10 years.