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Language and Literacy Development of Children with Williams Syndrome

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Abstract

Children with Williams syndrome, a rare neurodevelopmental disorder caused by deletion of ~25 genes on chromosome 7q11.23, evidence large individual differences in both broad language and reading abilities. Nevertheless, as a group, children with this syndrome show a consistent pattern characterized by relative strengths in concrete vocabulary and phonological processing (language skills strongly related to single-word reading) and relative weaknesses in relational concepts, receptive grammar, verbal working memory, comprehension monitoring, and discourse (language skills strongly related to reading comprehension). Children with Williams syndrome who have been taught reading using a systematic phonics approach both decode and comprehend significantly better than children who have been taught using a whole-word approach. Consideration of these patterns in the context of what is known about the reading development of children in the general population provides a strong foundation for facilitating the reading development of children with Williams syndrome.

Keywords

intellectual disability; language development; literacy; neurodevelopmental disorders; phonics; reading comprehension; Williams syndrome

Williams syndrome is a neurodevelopmental disorder caused by deletion of approximately 25 genes on chromosome 7q11.23 (Hillier et al., 2003; Osborne, 2006). This syndrome is characterized by a recognizable pattern of physical characteristics, including a specific set of facial features, heart disease (most commonly supraventricular aortic stenosis), connective tissue abnormalities, failure to thrive, and growth deficiencies (Morris, 2006). Young children with Williams syndrome have developmental delay which typically leads to mild to moderate intellectual disability, although some individuals have average intelligence and a very small proportion has severe intellectual disability. Williams syndrome is associated with a specific cognitive profile including relative strengths in verbal short-term memory and (concrete) language and extreme weakness in visuospatial construction (Mervis et al., 2000) and a specific personality profile, including gregariousness, overfriendliness, and anxiety (Klein-Tasman & Mervis, 2003). The prevalence of Williams syndrome is 1/7500 live births (Strømme, Bjørnstad, & Ramstad, 2002).

Most individuals with Williams syndrome (~95%) have the same set of genes deleted; this deletion is referred to as “classic.” Despite having the same deletion, verbal ability and nonverbal reasoning ability evidence the same amount of variability for individuals with Williams syndrome as for the general population, with standard deviations of ~15 on many

standardized assessments (Mervis & Morris, 2007). Given this pattern, it is not surprising that reading ability is also highly variable. Among the adults with Williams syndrome with classic deletions who have participated in my research are not only several individuals who cannot read at all but also a woman who decodes and comprehends at the level expected for entering university students. Among the child participants with classic deletions, reading skills range from an inability to read at all to age-appropriate decoding and comprehension. Mean level of overall reading ability for children and adolescents with Williams syndrome as measured by the Reading Composite standard score on the Wechsler Individual Achievement Test-II (WIAT-II; Wechsler, 2005) is in the mild academic deficit range (Becerra, John, Peregrine, & Mervis, 2008). Once again, the standard deviation is ≥ 15 , indicating as much or more variability as for the general population. Despite this variability, a common theme is expressed by parents when we discuss their child's schooling: Whether their child cannot read at all or reads and comprehends at a similar level to general population peers, improving his or her reading is the parents' highest educational priority. Parents view reading competence as critical not only for success at school but also for increasing the likelihood of successful employment during the adult years and providing a valued leisure time activity throughout the lifespan.

Researchers studying the reading development of children in the general population, including children who read well, children who decode well but have difficulty with reading comprehension ("poor comprehenders"), children who have difficulty decoding but comprehend well what they are able to read (children who have dyslexia), and children who have difficulty with both decoding and comprehension ("poor readers") have identified several factors that contribute to reading development (see reviews in Cain & Oakhill, 2007; McCardle & Chhabra, 2004; McCardle, Chhabra, & Kapinus, 2008; Oakhill & Cain, 2007). For word reading, the primary contributors are phonological skills and vocabulary. For comprehension, both decoding and broader language abilities are important, with relative importance changing as reading ability increases (e.g., Catts, Adlof, & Ellis Weismer, 2006). Fluency, background knowledge, and motivation also are important (McCardle et al., 2008). The remainder of this manuscript is divided into three parts. In the first, I summarize the findings for children with Williams syndrome on the broad language abilities that have been reported to be important for the development of reading by children in the general population. In the second, I summarize the results of the studies that have been conducted on the reading abilities of individuals with Williams syndrome. In the third, I consider the implications of the data presented in the first two sections, in combination with what is known about reading development in the general population, for facilitating the reading development of people with Williams syndrome.

BROADER LANGUAGE

The broad language abilities that are related to reading development by children in the general population include semantics, grammar, metalinguistics, discourse-level skills, and verbal memory. Below I briefly review the literature on these aspects of language acquisition by children with Williams syndrome.

Semantics

Concrete vocabulary—Concrete vocabulary has consistently been reported as the strongest language ability for individuals with Williams syndrome. Studies of English-speaking children with this syndrome almost always use a version of the Peabody Picture Vocabulary Test (PPVT; e.g., Dunn & Dunn, 1981, 1997) to assess receptive concrete vocabulary. From early reports on Williams syndrome (e.g., Bellugi, Marks, Bihrl, & Sabo, 1988) to recent ones (e.g., Brock, Jarrold, Farran, Laws, & Riby, 2007; Mervis & Becerra, 2007; Mervis & Morris, 2007), performance on the PPVT has repeatedly yielded the highest

mean standard score for any standardized assessment. For example, Mervis and John (in press) reported a mean standard score of 81.81 ($SD = 13.55$; range: 42 – 124) for 88 individuals with Williams syndrome aged 4 – 46 years on the PPVT-4 (Dunn & Dunn, 2007). The majority (82%) scored in the normal range (≥ 70), and 6% scored at or above the general population mean (≥ 100). The same 88 participants also completed the Expressive Vocabulary Test-2 (Williams, 2007), which was co-normed with the PPVT-4. The mean standard score on the EVT-2 was 80.81 ($SD = 13.54$, range: 47 – 120), almost identical to the mean standard score on the PPVT-4. Once again, the majority of participants (82%) earned standard scores ≥ 70 ; 10% earned standard scores ≥ 100 .

Relational/conceptual vocabulary—Relational/conceptual vocabulary includes terms for both basic relational concepts (e.g., spatial, temporal, quantitative, and dimensional terms) and more advanced relational concepts such as conjunctions and disjunctions (e.g., and, although, neither ... nor). In striking contrast to their performance on concrete vocabulary measures, the performance of children with Williams syndrome on relational vocabulary measures is very low. For example, Mervis and John (2008) reported that for a sample of 92 5 – 7-year-olds, mean PPVT-III standard score was 86.73 ($SD = 13.67$, range: 59 – 118); for the same children, mean standard score on the Test of Relational Concepts (TRC; Edmonston & Litchfield Thane, 1988), which measures simple relational concepts, was 55.79 ($SD = 21.37$; range: 25 – 104). Performance on the TRC was almost as low as on the Differential Ability Scales (Elliott, 1990) Pattern Construction subtest, the signature weakness in the Williams syndrome cognitive profile (Mervis et al., 2000). The performance of 9 – 11-year-olds with Williams syndrome on the Clinical Evaluation of Language Fundamentals-IV (CELF-IV; Semel, Wiig, & Secord, 2003) Formulated Sentences subtest, which includes a number of more advanced relational concepts, was even weaker, with 12 of 29 children earning a scaled score of 1, the lowest possible (Mervis & John, 2008). Stojanovik, Perkins, and Howard (2006) also reported very poor performance on this measure. Fraser and Conti-Ramsden (2008) found that children in the general population who were having educational difficulty (primarily language and/or literacy problems) performed considerably worse on the CELF-III Formulated Sentences subtest than did children who were not having educational difficulty.

Grammar

Williams syndrome originally came to the attention of researchers in the U.S. because of reports based on the pioneering research of Ursula Bellugi (e.g., Bates, 1990; Bellugi et al., 1988) that adolescents with this syndrome correctly produced and comprehended complex syntactic constructions such as relative clauses, passives, and tag questions despite having severe intellectual disability, thus providing strong evidence of the independence of language from cognition. More extreme versions of this position were taken by several academicians who had not worked directly with individuals with Williams syndrome. For example, Jackendoff (1994, p.117) stated that despite significant intellectual disability, the language of children with Williams syndrome “is if anything more fluent and advanced than that of their age-mates.” Piattelli-Palmarini (2001, p. 887) was even more emphatic, arguing that, “... children with Williams syndrome have barely measurable general intelligence and require constant parental care, yet they have an exquisite mastery of syntax and vocabulary. They are, however, unable to understand even the most immediate implications of their admirably constructed sentences.”

The results of more recent research have offered a more nuanced picture. First, the majority of individuals with Williams syndrome have intellectual abilities in the borderline to mild disability range; severe intellectual disability, rather than being the norm, is quite rare (e.g., Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Mervis & John, in press; Mervis &

Morris, 2007; Meyer-Lindenberg, Mervis, & Berman, 2006; Searcy, Lincoln, Rose, Klima, & Bavar, 2004). Second, although Bellugi and her colleagues' finding (e.g., Bellugi et al., 1988, 2000; Bellugi, Wang, & Jernigan, 1994) that the grammatical abilities of adolescents with Williams syndrome are considerably more advanced than those of CA- and IQ-matched adolescents with Down syndrome has been replicated by other research groups (e.g., Klein & Mervis, 1999; Mervis, Robinson, Rowe, Becerra, & Klein-Tasman, 2003; Vicari, Caselli, Gagliardi, Tonucci, & Volterra, 2004), these results most likely reflect the inordinate difficulty that most individuals with Down syndrome have with grammar, rather than that individuals with Williams syndrome have particularly good grammatical ability relative to their cognitive ability. Studies comparing the grammatical abilities of children and adolescents with Williams syndrome to either those of CA- and IQ-matched individuals with forms of intellectual disability other than Down syndrome or to typically developing children matched for mental age (MA) have consistently indicated that the syntactic abilities of the Williams syndrome group are at or slightly lower than that of the comparison group. This finding has held across English (Grant et al., 2002; Mervis et al., 2003; Udwin & Yule, 1990; Zukowski, 2004), German (Gosch, Städing, & Pankau, 1994), Hungarian (Lukács, 2005), and Italian (Volterra, Capirci, Pezzini, & Vicari, 1996; Volterra, Caselli, Capirci, Tonucci, & Vicari, 2003). Studies examining the morphological development of children and adolescents with Williams syndrome acquiring languages that have more complex morphology than English have indicated that morphological ability is at or slightly below the level of MA-matched typically developing children. This pattern has been reported for French (Karmiloff-Smith et al., 1997), Hebrew (Levy & Hermon, 2003), and Hungarian (Lukács, 2005; Lukács, Racsmany, & Pléh, 2004).

Researchers focusing on grammatical comprehension have administered a version of the Test for Reception of Grammar (TROG; Bishop, 1989 or TROG-2; Bishop, 2003) or a translation of this measure to individuals with Williams syndrome. These measures test understanding of a wide range of constructions ranging from simple positive statements to sentences containing center-embedded clauses. Mervis and Becerra (2007) reported a mean TROG-2 standard score of 70.25 ($SD = 16.33$, range: 55 – 111) for 110 individuals aged 5 – 18 years. The modal standard score was 55, the lowest possible score, indicating that many individuals with Williams syndrome have considerable difficulty with grammatical comprehension, especially of complex constructions. Karmiloff-Smith et al. (1997) reported a similar pattern of findings for a smaller sample. Lukács (2005) found that the performance of children, adolescents, and young adults with Williams syndrome on the Hungarian version of the TROG followed the same pattern as for MA-matched typically developing children, although the Williams syndrome group performed more poorly overall. Volterra et al. (1996) reported similar findings based on a comparison of the performance of children and adolescents with Williams syndrome on the Italian version of the TROG to that of MA-matched typically developing children. Individuals with Williams syndrome found the same types of grammatical constructions difficult to comprehend whether they were learning English, Hungarian, or Italian. Oakhill, Cain, and Bryant (2003; Oakhill & Cain, 2007), reporting the results of a longitudinal study of children in the general population, indicated that after taking into account vocabulary ability and IQ, performance on the TROG was not related to reading comprehension for 7 – 8-year-olds but was related to reading comprehension a year later.

Metalinguistics

Metalinguistics refers to the ability to consciously manipulate components of language, whether phonemes, words, or sentences (syntactic structures). Two metalinguistic abilities that are closely related to reading are phonological awareness and comprehension monitoring.

Phonological Awareness—The Differential Ability Scales-II (DAS-II; Elliott, 2007) includes a supplemental subtest, Phonological Processing, normed for ages 5 – 12 years, that measures phonological awareness. Four types of skills are assessed: rhyming, blending, deletion, and phoneme identification & word segmentation. The subtest yields an overall scaled score (T score). For the general population, mean T score is 50 ($SD = 10$). My lab (Mervis, unpublished data) has administered this subtest to 55 children with Williams syndrome aged 6.03 – 12.90 years (mean CA = 8.89 years, $SD = 2.15$). Mean T score was 40.24 [$SD = 13.28$, range: 10 (lowest possible) – 62]. Fourteen children (25%) scored at or above the mean for the general population. The correlation between CA and Phonological Processing T score was -0.01 , indicating that within this age range, the phonological awareness abilities of children with Williams syndrome relative to their CA-peers do not vary as a function of CA. These findings indicate considerable variability in the phonological processing abilities of children with Williams syndrome, with some children having extreme difficulty and others performing very well. Findings from studies that have related the phonological awareness abilities of individuals with Williams syndrome to their reading abilities are discussed in the Reading section.

Comprehension monitoring—Comprehension monitoring involves the ability to reflect on what one has heard or read, including the abilities to determine if one has understood the content and if one did not, to know what to do to fix the situation. There have not been any studies of text comprehension monitoring by individuals with Williams syndrome, and there has only been one study of comprehension monitoring in the oral modality involving individuals with this syndrome. John, Rowe, and Mervis (2009) examined the performance of 57 6 – 12-year-olds in the listener role of a referential communication task modeled after Abbeduto et al. (2008). In this task, children were asked to place one of several pictures into a scene based on the researcher's instructions. Although children performed very well when they understood the instructions and the required picture was present, they had considerable difficulty when the researcher's message was inadequate, letting the researcher know that there was a problem only 45% of the time; the remaining 55% of the time, the child placed one of the pictures into the scene even though he/she either did not have the correct picture or needed additional information to identify which picture had been requested. Indication that there was a problem was most likely if the requested picture was not among the available choices and the word used by the researcher to identify the referent was in the child's vocabulary, next most likely if the researcher used a word the child did not understand, and least likely if the request was ambiguous. Even when the child did indicate there was a problem, he/she often did not identify the correct problem. Performance was related to first-order Theory of Mind ability and CA. The performance of the children with Williams syndrome was considerably worse than that of the typically developing 3 – 6-year-old group in Abbeduto et al.'s study. This lack of comprehension monitoring by children with Williams syndrome even in a situation where it should have been obvious that the child could not comply with the researcher's request suggests that most children with Williams syndrome are likely not to monitor if they have understood what they have read and, even if they did realize that they had not understood, they likely would neither try to figure out the problem themselves nor seek help.

Discourse-level skills

Discourse-level skills that have been identified as important for reading comprehension include narrative understanding, narrative production, inference making, understanding story structure (including understanding sequencing of events), understanding anaphor, and using context to establish meaning (Oakhill & Cain, 2007). The few studies of these topics involving individuals with Williams syndrome are described below.

Narrative structure and inference making—Bellugi and her colleagues (Losh, Bellugi, Reilly, & Anderson, 2000; Reilly, Losh, Bellugi, & Wulfeck, 2004) have examined the structure of children's narratives of the wordless picture book, *Frog, Where Are You?* (Mayer, 1969) and the inferences included in these narratives. Reilly et al. compared a group of 4 – 12-year-old children with Williams syndrome to groups of CA-matched typically developing children and CA-matched children with specific language impairment. Losh et al. compared a group of 5 – 10-year-old children with Williams syndrome to three groups of typically developing children: one matched for CA, one matched for vocabulary age, and one matched for spatial age. Reilly et al. reported that the Williams syndrome group included significantly fewer story components than either control group and was considerably less likely to refer to the goals and motivations of the main character or to link his actions to his goal of finding his pet frog. In both studies, the Williams syndrome group was significantly more likely to use evaluative devices than any of the control groups. Furthermore, the Williams syndrome groups used predominantly social engagement devices whereas the control groups used predominantly cognitive inferences.

Other discourse-level skills—The Children's Communication Checklist-2 (CCC-2; Bishop, 2002), a parental report measure for use with individuals who are talking at least in short sentences, includes several items addressing discourse-level skills. Peregrine, Rowe, and Mervis (2004) considered parental ratings of 30 children with Williams syndrome aged 6 – 12 years and 35 typically developing siblings in the same age range on the CCC-2. The two groups differed significantly on all 10 scales ($p < .001$). The largest difference ($t > 8.0$) was for the Coherence scale (Cohen's $d = 2.12$; note that $d > 0.8$ is considered a large effect), which measures discourse-related skills such as sequencing events and appropriate use of anaphor. Further evidence of difficulty in event sequencing is provided by the inclusion of basic sequencing goals and/or goals addressing the comprehension and appropriate use of temporal terms such as before/after in the Individualized Education Programs (IEPs) of almost all children with Williams syndrome in preschool or elementary school and for many children in middle school.

Verbal memory

Studies of the verbal memory abilities of individuals with Williams syndrome have focused on verbal short-term memory, verbal working memory, and phonological memory. Although definitions of these terms vary (see the articles in Pickering, 2006), there is some consensus on basic distinctions. Verbal short-term memory refers to immediate memory for something that was just said, in the order in which it was said (verbatim recall). Verbal working memory requires active manipulation of the items in immediate memory (and in some cases, integration of material from long term memory with material in immediate memory), rather than simple verbatim recall. Phonological memory refers to immediate memory for the sounds of language, usually measured by verbatim recall of nonsense words or syllables that follow the phonotactics rules of the language in which the child is being assessed. The findings for each of these types of memory for individuals with Williams syndrome are summarized below (see Rowe & Mervis, 2006 for a review).

Verbal short-term memory—Verbal short-term memory has been measured by forward digit recall (repeating a string of numbers in the same order as the examiner provided them) or the initial trial of list recall. Results have consistently indicated that individuals with Williams syndrome perform significantly better than CA- and IQ-matched individuals with either Down syndrome (Edgin, 2003; Klein & Mervis, 1999; Vicari et al., 2004, Wang & Bellugi, 1994) or intellectual disability of unknown or mixed etiology (Devenny et al., 2004; Udwin & Yule, 1991).

Verbal working memory—The verbal working memory abilities of the participants in the Becerra et al. (2008) study were measured using the DAS-II Recall of Digits-Backward subtest (Mervis, unpublished data). On this supplemental subtest, the child is instructed to repeat strings of numbers presented by the examiner in reverse order. Performance is measured by a T score (general population mean = 50, $SD = 10$). The participants' mean T score was 30.25 [$SD = 10.56$, range: 10 (lowest possible) – 48], indicating a mean level of performance at the 2nd percentile for the general population, with as much variability as for the general population (Mervis, unpublished data). Five children (11%) scored at floor on this measure.

Three of the studies described above (Devenny et al., 2004; Edgin, 2003; Wang & Bellugi, 1994) also assessed backward digit recall. In each study, the Williams syndrome group earned a higher mean score than the contrast group did, but the between-group difference was not significant.

The relation between working memory as measured by backward digit recall and vocabulary or grammatical ability in individuals with Williams syndrome has been addressed in two studies. Mervis et al. (1999) found a strong correlation between backward digit span and both receptive vocabulary and receptive grammatical ability, even after controlling for CA, for 55 individuals with Williams syndrome. Robinson, Mervis, and Robinson (2003) considered the performance of 39 children with Williams syndrome aged 4 – 16 years and found that variance in backward digit recall ability accounted for the largest amount of variance in receptive grammatical ability as measured by the TROG. The correlation between backward digit recall and TROG performance was significantly higher for the Williams syndrome group than for a group of typically developing children matched to the Williams syndrome group for number of blocks correct on the TROG.

Phonological memory—The relation between phonological memory (as measured by nonword repetition) and vocabulary or grammatical development has also been considered. Grant et al. (1997), examining the performance of 17 children and adults with Williams syndrome (mean age 18 years), found a significant relation between ability to repeat low-wordlike nonword items and performance on the British Picture Vocabulary Scale (BPVS; the British version of the PPVT-R) that remained even after controlling for performance on the TROG. The pattern was similar to that for 4-year-old typically developing children (Gathercole, 1995) but differed from that for typically developing 5-year-olds in a manner that suggested that the Williams syndrome group was relying more heavily on memory and less on semantics in acquiring vocabulary; the authors considered this a less mature pattern. Robinson et al. (2003) found that performance on nonword repetition was not significantly related to receptive vocabulary (PPVT-R) for either the Williams syndrome group or the typically developing contrast group, after CA was partialled out. (Grant et al. did not control for CA.) However, Robinson et al. did find that nonword repetition ability accounted for a significant amount of variance in performance on the TROG even after the effects of CA, verbal short-term memory, and verbal working memory were taken into account. The relation between nonword repetition ability and receptive grammatical ability, after controlling for CA, was stronger for the Williams syndrome group than the contrast group, but the p value (.09) did not reach the conventional level of statistical significance.

Summary

Children with Williams syndrome evidence considerable variability in their broad language abilities. This variability is apparent both as a function of type of ability and, within a particular ability, as a function of the individuals tested. As a group, children with Williams syndrome evidence relative strengths in receptive concrete vocabulary and phonological

processing, with mean level of performance in the low average range for the general population. Relative to other groups with similar levels of intellectual disability, individuals with Williams syndrome show a relative strength in verbal short-term memory. At the group level, weaknesses for children with Williams syndrome include relational vocabulary, grammatical comprehension, verbal working memory, comprehension monitoring, and discourse-level processing. This pattern suggests that for individuals with Williams syndrome, the abilities which studies of children in the general population have implicated in single word reading are considerably stronger than those that have been implicated in reading comprehension. At the same time, almost every broad language ability discussed evidenced considerable variability. On most standardized measures, variability among individuals with Williams syndrome was at least as great as among individuals in the general population. This pattern suggests that there is likely to be considerable variability among children with Williams syndrome in both single-word reading ability and reading comprehension ability. In the next section, I describe the results of the studies that have been conducted on the reading abilities of individuals with Williams syndrome.

READING

In the initial studies of the reading abilities of individuals with Williams syndrome, researchers examined performance on standardized measures of reading and IQ. Factors beyond IQ that may have affected reading were typically not addressed. More recent studies have focused on the relation between reading abilities and other abilities (beyond IQ) that have been identified based on research with children in the general population as being important for reading development.

Early studies

The first formal reports of the reading ability of individuals with Williams syndrome were published in the late 1980s. Pagon, Bennett, LaVeck, Stewart, and Johnson (1987) assessed nine children and adolescents with Williams syndrome aged 10 – 20 years for both intellectual ability and reading ability. Full-scale IQs on the Wechsler Intelligence Scale for Children-Revised (WISC-R; Wechsler, 1974) ranged from <40 – 75. Four participants had a full-scale IQ of <40 (the lowest possible IQ). Reading was assessed using the Peabody Individual Achievement Test (PIAT; Dunn & Markwardt, 1970), which includes Reading Recognition and Reading Comprehension subtests. The authors reported the higher of the two reading standard scores for each participant; these ranged from 41 – 99. Grade equivalents ranged from K.5 – 9.1 for Reading Recognition and from K.5 – 9.2 for Reading Comprehension. All but the oldest participant was able to decode and comprehend at least at the mid-1st grade level; median grade level was mid-2nd grade. The child scoring at the 9th grade level was in fact in 9th grade, so decoded and comprehended at grade level. The authors argue that for all participants, the higher of the two reading standard scores was above the person's full-scale IQ, therefore exceeding expectations based on IQ. This claim assumes that if the child is reading at the level expected for IQ, reading standard scores should be the same as IQ. In fact, for children with IQs < 100, predicted reading standard scores are well above IQ. For example, for a child with a WISC-III full-scale IQ of 60, predicted reading standard scores for the WIAT-II are 74 for Word Reading, 79 for Pseudoword Decoding, and 73 for Reading Comprehension (Wechsler, 2005). Nevertheless, the discrepancy between full-scale IQ (or Verbal IQ) and highest reading standard score was large enough to be considered significant for 6 of the 9 participants.

Udwin, Yule, and Martin (1987) assessed the intellectual and reading abilities of 44 children with Williams syndrome aged 6 – 16 years. The WISC-R full-scale IQs of 10 children were <40. For the remaining children, full-scale IQ ranged from 40 – 89, with a mean of 54.5. Reading ability was measured with the Neale Analysis of Reading (Neale, 1966), which

includes Reading Accuracy and Reading Comprehension subscales. Only 22 children (mean CA 12.0 years; range: 6.0 – 15.9 years) obtained at least a basal score on the Reading Composite. For these children, mean Reading Accuracy age equivalent was 7 years 10 months (range: 74 – 137 months) and mean Reading Comprehension age equivalent was 7 years 9 months (range: 75 – 144 months). The group that could read was significantly older and had significantly higher IQs than the group that could not read.

Udwin, Davies, and Howlin (1996) retested 23 of the 44 participants in the Udwin et al. (1987) study an average of 8.83 years later. This group had a mean CA of 12.92 years ($SD = 1.90$ years) at initial study and 21.75 years ($SD = 1.90$ years) at follow up. Reading was assessed with the Wechsler Objective Reading Dimensions test (WORD; Rust, Golombok, & Trickey, 1993), which includes Basic Reading and Reading Comprehension subtests. Fourteen participants attained at least a basal score on the Neale; 13 of them plus 4 additional participants obtained at least a basal score on the WORD. The authors note that the two reading tests are not directly comparable and that it is easier to obtain a basal on the WORD than on the Neale. Comparison of grade equivalent scores indicated a small and nonsignificant increase for decoding and a small but significant decrease for comprehension. (For a discussion of problems with using grade and age equivalents and of comparing these types of scores across different assessments, see Mervis & Robinson, 2005.)

Howlin, Davies, and Udwin (1998) considered the reading abilities of 62 adults with Williams syndrome (mean CA = 26.49 years, range: 19 – 39 years). This group included the 23 participants in Udwin et al. (1996). Mean WAIS-R (Wechsler, 1981) full-scale IQ was 60.85 ($SD = 5.94$). Forty seven participants scored above basal on the WORD. For this group, mean Basic Reading age equivalent was 8.65 years ($SD = 0.2$ years; range: 6.0 – 18.0 years) and Mean Reading Comprehension age equivalent was 7.16 years ($SD = 2.13$ years; range: 6.0 – 11.0 years). The Readers and Non-Readers did not differ significantly in CA. However, reading ability was related to IQ: Although no participant with an IQ < 50 was able to read, 78% of participants with IQs of 50 – 69 and all participants with IQs ≥ 70 could read.

Single-word reading and its correlates

Studies that included typically-developing contrast groups—Most of the recent studies of the reading abilities of individuals with Williams syndrome have focused on single-word reading and factors affecting this ability. Four research groups have compared the performance of individuals with Williams syndrome to that of considerably younger typically developing children. In this paragraph, I describe the participants studied by each research group. In the following paragraphs, the results of the studies are compared and contrasted. Laing, Hulme, Grant, and Karmiloff-Smith (2001; Laing, 2002) compared the reading abilities of 15 individuals with Williams syndrome (mean CA = 15.08 years, range 9 – 27 years) to those of 15 typically developing children (mean CA = 6.75 years, range: 5.0 – 9.17) individually matched for reading age on the British Ability Scales (BAS; the British version of the DAS) test of single-word reading and verbal mental age on the BPVS. The Williams syndrome group had a mean GCA (similar to IQ) on the BAS of 43.8 ($SD = 5.12$, range: 39 – 54). This mean GCA is considerably lower than the 58.29 ($SD = 12.77$, range: 24 – 94) reported by Mervis and Becerra (2007) for 119 8 – 17-year-olds with Williams syndrome on the DAS, suggesting that the participants in Laing et al.'s study had more limited intellectual abilities than is typical for Williams syndrome. Menghini, Verucci, and Vicari (2004) compared the single-word reading abilities of 16 Italian-speaking individuals with Williams syndrome (mean CA = 17.58 years, range: 10.9 – 30.2 years) to those of 16 typically developing children aged 6.2 – 8.6 years group-matched to the Williams syndrome group for MA as measured by the Stanford-Binet Intelligence Scale Form L-M (Terman &

Merrill, 1973). Mean MA was 7.0 years for the Williams syndrome group and 7.7 years for the typically developing group. Although the mean MAs of the two groups did not differ significantly ($p > .10$), the groups were not well matched (see Mervis & John, 2008, for a demonstration of the impact of how well groups are matched on the outcome of between-group comparisons). Garayzabal Heinze & Cuetos Vega (2008) studied 12 Spanish children with Williams syndrome (mean CA 12.45 years, range: 8 – 15 years) who had a mean WISC IQ of 50.6. The Williams syndrome group was matched to a group of typically developing children whose CA (7.9 years) matched the Williams syndrome group's mean age equivalent on the Spanish version of the PPVT (7.8 years). Temple (2006) compared the performance of eight children with Williams syndrome with a mean MA of 6.58 years (range: 5.58 – 7.75) to that of 12 typically developing children (mean CA = 6.42 years, range: 5.5 – 7.33). CA information was not reported for the Williams syndrome group.

The results of all four studies indicated that on average, the single-word reading abilities of individuals with Williams syndrome are quite limited, although there was considerable variability. Laing et al. (2001) reported a mean WORD word recognition (Basic Reading subtest) test age of 82.2 months ($SD = 31.3$). Three participants were not able to read at all. Menghini et al. (2004) and Temple (2006) reported that single-word reading was at the level expected for MA. Garayzabal Heinze & Cuetos Vega (2008) reported that single-word reading was significantly lower than for the matched group of young typically developing children.

To address possible correlates of the reading abilities of individuals with Williams syndrome, Laing et al. (2001), Menghini et al. (2004), and Garayzabal Heinze and Cuetos Vega (2008) included measures of phonological awareness. Laing et al. found a significant difference between the Williams syndrome and control groups only for phoneme deletion. However, the mean for the control group was higher than the mean for the Williams syndrome group for every phonological awareness measure administered. Performance on all the phonological awareness measures, on nonword reading, and on speeded naming was significantly correlated with single-word reading ability for both groups. However, after controlling for CA and intellectual ability, the only correlations that remained significant were between nonword reading and single-word reading and between speeded naming and single-word reading. For the Williams syndrome group, both BAS Nonverbal Reasoning abilities and BAS Spatial abilities were significantly correlated with single-word reading abilities; correlations of BAS Verbal abilities and BPVS abilities with single-word reading abilities were considerably weaker. Laing et al. also used a paired associate procedure in which real words which varied in imageability were paired with consonant trigrams that varied in their phonetic similarity to the real word to address the importance of phonological and semantic processes in learning to read single words. The Williams syndrome group showed a strong effect of phonetic similarity but no effect of imageability; in contrast, the typically developing group showed strong effects of both factors. The authors argued that these findings provide further evidence that for individuals with Williams syndrome, the process of learning to read depends disproportionately on phonological skills with only a very limited effect of semantics. However, some studies of typically developing children have failed to find semantic effects using this paradigm (e.g., McKague, Pratt, & Johnston, 2001), suggesting the importance of further research to address the question of the relative importance of phonological and semantic skills for individuals with Williams syndrome in learning to read.

Menghini et al. (2004) also found that the control group performed significantly better than the Williams syndrome group on all measures of phonological awareness included in their study, with significant differences for syllable deletion and rhyme detection and a nonsignificant difference for syllable segmentation. Syllable deletion was significantly

correlated with both single-word reading and nonword reading for the Williams syndrome group. MA was significantly correlated with nonword reading and homophone reading for the Williams syndrome group; for the typically developing group, correlations with MA were significant for word, nonword, and homophone reading. Garayzabal Heinze and Cuetos Vega (2008) found that the Williams syndrome group performed at the same level as the control group for syllable deletion and for nonword repetition. However, the control group performed significantly better than the Williams syndrome group on rhyming. No correlations between phonological processing measures and reading ability were reported.

The performance of the Williams syndrome and control groups on nonword reading was compared in three of the four studies, with contrasting findings. Menghini et al. (2004) reported that the Williams syndrome group performed significantly worse than the control group on nonword reading. In contrast, Temple (2006) and Garayzabal Heinze and Cuetos Vega (2008) reported that reading of nonwords was at the same level for the Williams syndrome group as for the control group.

Studies not including contrast groups—Two studies of the single-word reading abilities of individuals with Williams syndrome have been conducted that did not include control groups. Levy, Smith, and Tager-Flusberg (2003) considered the abilities of a group of 20 English-speaking individuals with Williams syndrome (mean CA = 16.42 years; range: 12.67 – 20.33), and Levy and Antebi (2004) considered the abilities of a group of 17 Hebrew-speaking individuals with Williams syndrome (mean CA = 16.17 years; range: 11 – 22).

The Williams syndrome group tested by Levy et al. (2003) had a mean IQ of 57.05 ($SD = 12.99$) on the Kaufman Brief Intelligence Test (Kaufman & Kaufman, 1990). This score is considerably lower than the mean of 69.32 ($SD = 15.36$) reported by Mervis and Becerra (2007) for 306 individuals with Williams syndrome. Despite this lower-than-expected mean IQ, the participants in Levy et al. performed relatively well on the Comprehensive Test of Phonological Processing (CTOPP; Wagner, Torgesen, & Rashotte, 1999); mean standard score on Elision (syllable or phoneme deletion) was in the borderline range, and mean standard scores on Segmenting Words and Segmenting Nonsense Words were in the low average range. Three participants were not able to read any nonwords on the Woodcock-Johnson (Woodcock & Johnson, 1990) Word Attack subtest. Results of correlational analyses including the 17 individuals who could read indicated that KBIT Matrices performance was significantly correlated with nonword reading, nonword reading was significantly correlated with word reading, elision was significantly correlated with both nonword and word reading, and word segmentation was significantly correlated with nonword reading. The correlations involving elision and word segmentation remained significant even after controlling for KBIT Matrices ability. Participants were at ceiling on the rhyming measures. KBIT Composite IQ was related to nonword reading ability; all of the nonreaders had $IQ < 50$, people with Composite IQs of 50 – 70 had Word Attack standard scores similar to their IQs, and people with Composite IQs > 70 had Word Attack and CTOPP phonological awareness standard scores that were considerably higher than their Composite IQs. Performance on CTOPP Rapid Object Naming was close to floor for all participants.

Learning to read Hebrew is likely more challenging for individuals with Williams syndrome than learning to read languages that use the Roman alphabet. As Levy and Antebi (2004) describe, reading Hebrew requires integrating visuospatial information along several different axes, due to the fact that Hebrew vowels are indicated below or above the consonants, rather than along the same axis. Perhaps for this reason, the proportion of individuals who could not read was higher than in studies addressing ability to read English,

Italian, or Spanish. Four of 17 participants could not read either real words on nonwords and 2 could read real words but not nonwords. The remaining 11 participants could read both words and nonwords. Four read below the 3rd grade level, six read at the 3rd grade level, and one (CA 19 years) read at age level. For the 11 readers, phoneme deletion and expressive vocabulary were significantly correlated with single-word reading, and phoneme identification and single-word reading were significantly correlated with nonword reading.

Reading comprehension

Both Laing et al. (2001) and Menghini et al. (2004) included a measure of reading comprehension in their studies. Laing et al. reported a large and significant difference in the performance of the participants with Williams syndrome on the Basic Reading (mean test age = 82.2 months, $SD = 31.3$) and Reading Comprehension (mean test age = 58.2 months, $SD = 44.0$) subtests of the WORD. Menghini et al. reported that although the Williams syndrome and typically developing control groups did not differ significantly in their single-word reading abilities, the typically developing group performed significantly better than the Williams syndrome group on the reading comprehension test.

Relation of primary reading instruction method to reading ability

Most of the articles discussed in the previous two sections ended with brief statements regarding the implications of the reported research for reading instruction for children with Williams syndrome. Based on her finding that individuals with Williams syndrome benefitted from the phonetic information but not from the (implicit) semantic information provided in the paired-associate trigram reading task, Laing (2002) raised the question of whether the methods used to teach reading to typically developing children are appropriate for children with learning difficulties or if instead different and perhaps even syndrome-specific methods are needed. No suggestions for specific methods were mentioned. Menghini et al. (2004) argued that children with Williams syndrome seemed likely to benefit from the whole word approach. This position is based on their finding that although the single-word reading ability of the Williams syndrome group was at the same level as that of the typically developing control group, the control group performed significantly better on the reading of nonwords. Menghini et al. note that even if the whole word method is used, instructors should also focus on increasing phonemic awareness, given the importance of grapheme-phoneme correspondence in learning to read. Furthermore, reading comprehension should be emphasized from the onset of reading instruction. Levy et al. (2003) stated that phonics instruction should be helpful for children with Williams syndrome, given the strong relations between their phonological awareness abilities and their reading abilities. Levy and Antebi (2004) indicated that children with Williams syndrome and, more generally, children with intellectual disability, may benefit from an explicit phonics approach rather than a whole word approach to reading instruction.

The relation between primary reading instruction method and reading ability of children with Williams syndrome has been addressed in an ongoing study conducted in my lab (Becerra et al., 2008). We have used the WIAT-II to assess the reading ability of 44 children aged 9 – 17 years (mean CA = 12.49 years, $SD = 2.61$). Mean DAS-II GCA was 63.14 ($SD = 11.58$, range: 39 – 98). The children's WIAT-II standard scores indicated a wide range of reading ability relative to CA peers in the general population, with standard deviations > 15 for all measures. Mean standard scores were 73.00 [range: 40 (lowest possible) – 112] for Word Reading, 78.75 [range: 0 correct – 113 (standard score)] for Pseudoword Decoding, and 64.61 [range: 40 (lowest possible) – 102] for Reading Comprehension. All children could read at least a few of the real words, but 8 (18%) could not read any nonwords. As was the case for the previous studies, mean standard score was considerably higher for single-word reading than for reading comprehension.

Primary reading instruction method was whole (sight) word for 20 children and phonics for 24 children. Although there was a wide range of GCAs in each group, mean GCA was significantly higher for the Phonics group (67.42, range: 49 – 98) than for the Whole Word group (58.00, range: 39 – 80). Thus, to compare the reading abilities of the two groups, a method of adjusting expected reading standard scores as a function of GCA was important. The DAS-II manual provides a table of predicted WIAT-II standard scores based on the child's GCA. To determine if there were significant between-group differences in reading ability after intellectual ability was taken into account, we computed discrepancy scores (obtained reading standard score minus predicted reading standard score) for each child. Results indicated large and significant differences as a function of group for single word-reading, nonword reading, and reading comprehension. The eight children who could not read any of the nonwords were in the Whole Word group. Most children in the Phonics group read at or above the level expected for their GCA. In sharp contrast, most children in the Whole Word group read below the level expected for their GCA.

IMPLICATIONS

In the first two sections of this article, I described the pattern of broad language strengths and weaknesses characteristic of individuals with Williams syndrome and then summarized the available data on the reading abilities of individuals with this syndrome. There is clearly considerable variability among individuals with Williams syndrome with regard to both their broad language abilities and their reading abilities; the standard deviations on standardized assessments indicate at least as much variability as in the general population, although means are almost always considerably lower for the Williams syndrome group. Children with Williams syndrome show relative strengths on the broad language abilities previously identified as being particularly important for decoding and relative weaknesses on the broad language abilities considered important for reading comprehension (see reviews in McCardle et al., 2008; McCardle & Chhabra, 2004). Furthermore, as has been repeatedly found for children in the general population (e.g., Ehri, 2004), and has also been demonstrated in several intervention studies involving children with Down syndrome (e.g., Bourassa, Cleave, & Kay-Raining Bird, 2005; Cupples & Iacono, 2002; see review in Snowling, Nash, & Henderson, in press), children with Williams syndrome who are taught to read using systematic phonics instruction learn to read significantly better relative to expectations based on IQ than do children who are taught with whole word (or whole language) methods.

In the present section, I first consider the implications of these findings for single-word reading instruction for children with Williams syndrome and then consider the implications for reading comprehension instruction. Evidence that findings from studies of children in the general population are likely to be relevant to teaching children with Williams syndrome to read come from at least two sources: First, the major meta-analysis findings held for children with high, average, and low IQs and for children from all socio-economic backgrounds. Second, the results reported by Becerra et al. (2008) indicate that the type of single-word reading method that is most effective for children in the general population (systematic phonics instruction) is also most effective for children with Williams syndrome.

Single-word reading

One of the strongest findings to emerge from meta-analyses of studies of reading development is the importance of early, explicit, and systematic instruction in phonemic awareness and phonics for children in the general population. These results are summarized in Ehri (2004) and McCardle et al. (2008). The results of the meta-analyses indicated that rather than focusing on a wide variety of phonemic awareness skills, it is better to focus on only a few, in particular blending (combining a series of separate phonemes into a word) and

segmentation (breaking a word into its segments, often accompanied by tapping, clapping, etc.). Instruction is more effective if it involves actual letters rather than just sounds or blank tokens (e.g., colored squares) and if it is delivered in a small-group setting. Phonemic awareness instruction is most effective if it is provided in kindergarten or first grade. Some children with Williams syndrome have difficulty learning or remembering letter-sound correspondences. For these children, use of mnemonic devices such as incorporating an object that starts with the letter sound into the depiction of the letter presented to the child may be helpful. Ehri provides the example of an S drawn as the body of “Sammy Snake” as in the Letterland program (Wendon, 1992).

The meta-analysis results indicated that systematic phonics instruction (teaching of all major letter-sound correspondences, including consonants, vowels, and digraphs, in a clearly-defined sequence) was more effective than either whole word (having children memorize whole words; sometimes referred to as the look-say or sight-word approach) or whole language (focusing on meaning, with letter-sound correspondences taught incidentally and in context, only as needed) approaches. This general population result has already been found to hold for children with Williams syndrome. Synthetic phonics programs, which involve teaching children to break a word into its constituent phonemes and then blend them together to form the word, have been most effective in teaching children in the general population to read. This approach should help the child to connect the letter to its sound. Systematic texts that are written so that almost all of the words involve phonics rules that the child has been taught are important. It is also important for the books not to simply focus on the most recently taught correspondences but also to provide practice on previously learned ones. Finally, phonics instruction is much more likely to be successful if delivered by a knowledgeable teacher who believes that his/her students are able to learn.

Meta-analysis results (see summary in Ehri, 2004) also indicated some teaching approaches that are less effective. In particular, it is important that phonics worksheets not be the primary method of instruction. Instead, the child needs to be actively taught, with the teacher explaining and modeling phonics principles and providing the child with practice with feedback. It is better for children to learn phonics rules by reading and writing (or forming with letter tiles or spelling orally) words that demonstrate the rule than by memorizing explicit rules. This is likely to be especially important for children with Williams syndrome; based on parental and teacher report, these children often have difficulty generalizing rules that have been explicitly taught to new material. Phonics instruction should be integrated into reading and writing instruction rather than being taught as a stand-alone topic. Children with intellectual disabilities, including Williams syndrome, will need to be taught to apply their knowledge of phonics when they encounter a new word, rather than reverting to guessing. Meta-analysis results indicated that systematic phonics instruction is most effective if it is the first reading method to which a child is exposed. Introduction of synthetic phonics after children have already been exposed to whole word or whole language methods is much less effective, perhaps because children have to learn to suppress habits such as guessing words based on context or first letter.

Reading Comprehension

Successful single-word reading is clearly important for reading comprehension, and in the early grades, single-word reading skill is often the best predictor of reading comprehension. Once children become skilled single-word readers, however, other abilities such as fluency and broad language abilities (including comprehension monitoring) become increasingly important for successful reading comprehension. Results of meta-analyses of studies of children with learning disabilities indicated that reading outcomes were best when systematic phonics instruction was combined with comprehension strategy instruction (Ehri, 2004).

Fluency—Fluency involves reading aloud at a conversational rate and with expression. As such, it requires not only the ability to recognize words rapidly but also the ability to appropriately group words into grammatical units to provide the basis for reading with expression. Fluent reading frees cognitive resources so that the child can focus on comprehension. Fluency requires extensive practice. Round-robin reading, where each of the children in a reading group takes a turn in reading, has not been found to be effective (McCardle & Chhabra, 2008) in increasing fluency. In this method, children spend most of their time listening to other children read rather than practicing reading; good readers averaged 6 minutes a day of oral reading and poor readers averaged 2 minutes or less. Silent reading has also not been found to be effective in improving fluency. Instead, repeated reading methods in which a teacher first reads a text to the students and discusses it, using the comprehension methods described in the next section, and then the children reread the text multiple times with a parent and/or a partner and are given constructive feedback is most effective. For children having difficulty, echo-reading and/or listening to the text on tape while following along and then reading the text out loud have been found to be effective in improving fluency (McCardle & Chhabra, 2008; Stahl, 2004). Stahl (2004) provided examples of weekly lesson plans for incorporating fluency instruction and comprehension instruction.

There have been no formal studies of reading fluency for children with Williams syndrome. Informal observations of the participants in the Becerra et al. (2008) study indicated that children who comprehended well read fluently. Most of the children who did not comprehend well did not read fluently. However, a small group of children read fluently yet had considerable difficulty with comprehension. The existence of this group confirms what has been found for children in the general population: fluency is important for reading comprehension, but it is not sufficient.

Vocabulary and grammatical comprehension—Successful reading comprehension requires that the child be able to understand both the words in the text and the grammatical constructions (both within a single sentence and across sentences, e.g., as indicated by anaphoric reference) used by the author. Although children with Williams syndrome tend to have relatively good concrete vocabularies, they sometimes do not know the precise meanings of words [as exemplified by Bellugi et al.'s (1994, p. 32) example of an adolescent saying she needed to “evacuate the glass”]. These words tend to be those that fall in Beck and McKeown's (1985) third vocabulary tier, which includes words that are not high frequency but that mature language users often encounter or produce (e.g., novice, prestige). McCardle and Chhabra (2008) suggested that words in this tier are the ones for which direct instruction is likely to yield the most benefits. Recommended strategies included writing (or dictating) the words in multiple sentences, semantic mapping (graphically indicating the relation of the word to its concept and to related concepts), and word-pair charts (indicating relations between pairs of words such as same, opposite, go together, unrelated).

Children with Williams syndrome have particular difficulty with relational concepts. These terms are very important for reading comprehension; for example, they provide information regarding temporal order (e.g., before, after, until) and indicate relational links between words (e.g., and, or) or clauses and sentences (e.g., nevertheless, however, although). Learning these terms will require direct and intensive instruction in a variety of contexts. Generalization to new contexts should be tested and additional instruction provided as needed. Some of the strategies suggested above may also be helpful in teaching relational terms to older children. More complex grammatical constructions such as passives, relative/embedded clauses, or anaphoric reference also may need to be taught directly and then tested for generalization.

Verbal working memory—Verbal working memory plays a critical role in reading comprehension, providing the work space for the constructive and integrative processing necessary for the formation of a mental model of the text (Cain, 2006). Working memory holds the most recent material read, allowing the child to integrate this material into the ongoing mental-model construction. Working memory also holds material retrieved from long term memory (e.g., background knowledge), allowing that material to be integrated into the mental model. For these reasons, working memory is strongly correlated with reading comprehension. For children in the general population who are at least 8 years old, working memory capacity contributes unique variance to reading comprehension beyond that attributed to verbal IQ, vocabulary ability, and single-word reading ability (Cain, 2006; Cain & Oakhill, 2007). Fluent reading is important for freeing working memory space for mental model construction, but it is not sufficient. Although children with Williams syndrome often have relatively good verbal short term memory, their working memory is typically much more limited. These limitations adversely affect inference-making, understanding of anaphoric reference, and inferring the meaning of new vocabulary from context, all areas of weakness for children with Williams syndrome.

Cain (2006) has argued that working memory ability may be enhanced by practice at representing and manipulating linguistic information. She also noted that many of the comprehension strategies described in the next section offer ways for children to work around their limited verbal working memory ability. Combining working memory training and comprehension strategy training should facilitate the development of reading comprehension.

Comprehension strategies—Children with Williams syndrome have particular difficulty with several aspects of broad language that are critical for reading comprehension. These include comprehension monitoring and discourse components such as coherence and narrative structure, in addition to those discussed previously. The National Reading Panel (see Kamil, 2004; McCardle & Chhabra, 2008) identified seven strategies for which there was evidence that direct classroom instruction was effective in enhancing reading comprehension for children in the general population:

1. Comprehension monitoring (knowing when you have not understood what you have read, and knowing what to do to fix the situation)
2. Cooperative learning (working with another child/children to improve both comprehension and social skills related to literacy and learning)
3. Graphic organizers (use of visual representation as a memory aid for text content and organization)
4. Story structure (instruction in story components and sequences, including use of story maps)
5. Question answering (understanding what type of material is needed to answer specific questions, including when to look back at the text and when the answer requires background knowledge or needs to be inferred)
6. Question generation (posing specific types of questions to oneself to increase one's awareness of whether the material read has been understood)
7. Summarization (identification of central ideas, making inferences, and generalizing from the text).

More detailed description of these techniques and how to apply them is provided in McCardle et al. (2008). These strategies are also effective for children with poor reading

comprehension skills, although instruction for these children must be even more explicit and should include extensive practice (Gersten, Fuchs, Williams, & Baker, 2001).

The National Reading Panel also made several other important points regarding comprehension strategies (Kamil, 2004). Instruction in use of these strategies should not be restricted to reading classes; this type of instruction should be incorporated into content areas such as science or social studies as well. It is also important that children understand that the purpose of using comprehension strategies is to comprehend the text, not to show that they can apply the strategy. This point will need to be taught explicitly to most children with Williams syndrome. Comprehension strategy instruction should begin at the same time as reading instruction begins, rather than being delayed until the child is reading single words well.

CONCLUSION

Large individual differences have been documented among children with Williams syndrome with regard to broad language abilities relative to CA-matched peers, with some children performing in the average range for the general population, others performing in the severe intellectual disability range, and most performing in the borderline to moderate intellectual disability range. Nevertheless, as a group, children with Williams syndrome show a consistent pattern of relative strengths and weaknesses in broad language abilities. This pattern includes relative strengths in two areas that are strongly related to single-word reading for children in the general population: phonological processing and concrete vocabulary and considerable weakness in areas that are strongly related to reading comprehension: relational concepts, receptive grammar, verbal working memory, comprehension monitoring, and discourse-related skills. Not surprisingly given this profile, children with Williams syndrome perform significantly better on standardized assessments of single word reading than of reading comprehension.

As has been found for the general population, method of reading instruction has a strong impact on the reading skills of children with Williams syndrome: Children who are taught to read using systematic phonics instruction both decode and comprehend significantly better, relative to IQ, than do children taught with whole word methods. This finding that the most effective approach for teaching single-word reading to children with Williams syndrome is the same as for both children in the general population who are good readers and children in the general population who are having difficulty learning to read offers encouragement that the same methods of teaching reading fluency and reading comprehension that are effective for children in the general population will also be effective for children with Williams syndrome, although more explicit and extensive instruction will likely be needed. Although the ultimate level of reading achievement will be affected by a child's intellectual ability, among other factors, most if not all children with Williams syndrome should be able to learn to read if effective instruction is provided. To achieve this goal, a partnership among highly effective teachers, classroom assistants and aides explicitly trained to facilitate children's reading development, parents, and researchers is crucial.

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References

- Abbeduto L, Murphy MM, Kover ST, Giles ND, Karadottir S, Amman A, et al. Signaling noncomprehension of language: A comparison of Fragile X syndrome and Down syndrome. *American Journal on Mental Retardation*. 2008; 113:214–230. [PubMed: 18407723]
- Bates, E. Early language development: How things come together and how they come apart. *International Conference on Infant Studies*; Montreal, Quebec. 1990 April.
- Becerra, AM.; John, AE.; Peregrine, E.; Mervis, CB. Reading abilities of 9 – 17-year-olds with Williams syndrome: Impact of reading method. *Symposium on Research in Child Language Disorders*; Madison, WI. 2008 June.
- Beck IL, McKeown MG. Teaching vocabulary: Making the instruction fit the goal. *Educational Perspectives*. 1985; 23:11–15.
- Bellugi, U.; Marks, S.; Bihle, A.; Sabo, H. Dissociation between language and cognitive functions in Williams syndrome. In: Bishop, D.; Mogford, K., editors. *Language development in exceptional circumstances*. London: Churchill Livingstone; 1988. p. 177-189.
- Bellugi U, Lichtenberger L, Jones W, Lai Z, St George M. The neurocognitive profile of Williams syndrome: a complex pattern of strengths and weaknesses. *Journal of Cognitive Neuroscience*. 2000; 12(Supplement 1):7–29. [PubMed: 10953231]
- Bellugi, U.; Wang, P.; Jernigan, TL. Williams syndrome: An unusual neuropsychological profile. In: Broman, SH.; Grafman, J., editors. *Atypical cognitive deficits in developmental disorders: Implications for brain function*. Hillsdale, NJ: Erlbaum; 1994. p. 23-56.
- Bishop, DVM. *Test for Reception of Grammar*. Manchester, UK: Chapel Press; 1989.
- Bishop, DVM. *The Children's Communication Checklist*. 2. London, UK: Psychological Corporation; 2002.
- Bishop, DVM. *Test for Reception of Grammar, version 2*. London, UK: Psychological Corporation; 2003.
- Bourassa, DC.; Cleave, P.; Kay-Raining Bird, E. Teaching children with Down syndrome to read: An update. *Annual Meeting of the Canadian Language and Literacy Research Network Centre of Excellence*; Toronto, Ontario. 2005 June.
- Brock J, Jarrold A, Farran EK, Laws G, Riby DM. Do children with Williams syndrome really have good vocabulary knowledge? Methods for comparing cognitive and linguistic abilities in developmental disorders. *Clinical Linguistics & Phonetics*. 2007; 21:673–688. [PubMed: 17701755]
- Cain, K. Children's reading comprehension: The role of working memory in normal and impaired development. In: Pickering, SJ., editor. *Working memory and education*. Amsterdam, The Netherlands: Elsevier; 2006. p. 61-91.
- Cain, K.; Oakhill, J. Reading comprehension difficulties. In: Cain, K.; Oakhill, J., editors. *Children's comprehension problems in oral and written language: A cognitive perspective*. New York, NY: Guilford; 2007. p. 41-75.
- Catts HW, Adlof SM, Ellis Weismer S. Language deficits in poor comprehenders: A case for the simple view of reading. *Journal of Speech, Language, and Hearing Research*. 2006; 49:278–293.
- Cupples L, Iacono T. The efficacy of 'whole word' versus 'analytic' reading instruction for children with Down syndrome. *Reading and Writing: An Interdisciplinary Journal*. 2002; 15:549–574.
- Devenny DA, Krinsky-McHale SJ, Kittler PM, Flory M, Jenkins E, Brown WT. Age-associated memory changes in adults with Williams syndrome. *Developmental Neuropsychology*. 2004; 26:691–706. [PubMed: 15525565]
- Dunn, LE.; Dunn, DM. *Peabody Picture Vocabulary Test*. 4. Minneapolis, MN: Pearson Assessments; 2007.
- Dunn, LE.; Dunn, LE. *Peabody Picture Vocabulary Test-Revised*. Circle Pines, MN: American Guidance Service; 1981.
- Dunn, LE.; Dunn, LE. *Peabody Picture Vocabulary Test*. 3. Circle Pines, MN: American Guidance Service; 1997.
- Dunn, LM.; Markwardt, FC, Jr. *Peabody Individual Achievement Test*. Circle Pines, MN: American Guidance Service; 1970.

- Edgin, JO. Dissertation Abstracts International: Section B: The Sciences & Engineering. Vol. 64. US: University Microfilms International; 2003. A neuropsychological model for the development of the cognitive profiles in mental retardation syndromes: Evidence from Down syndrome and Williams syndrome; p. 1522
- Edmonston, NK.; Litchfield Thane, N. TRC: Test of Relational Concepts. Austin, TX: PRO-ED; 1988.
- Ehri, LC. Teaching phonemic awareness and phonics: An explanation of the National Reading Panel meta-analyses. In: McCardle, P.; Chhabra, V., editors. The voice of evidence in reading research. Baltimore, MD: Brookes; 2004. p. 153-186.
- Elliott, CD. Differential Ability Scales. San Antonio, TX: Psychological Corporation; 1990.
- Elliott, CD. Differential Ability Scales. 2. San Antonio, TX: Psychological Corporation; 2007.
- Fraser J, Conti-Ramsden G. Contribution of phonological and broader language skills to literacy. *International Journal of Language & Communication Disorders*. 2008; 43:552–569.
- Gathercole SE. Is nonword repetition a test of phonological memory or long-term knowledge? It all depends on the nonwords. *Memory and Cognition*. 1995; 23:83–94.
- Garayzabal Heinze E, Cuetos Vega F. Aprendizaje de la lectura en los niños con síndrome de Williams. *Psicothema*. 2008; 20:672–677. [PubMed: 18940067]
- Gersten R, Fuchs LS, Williams JP, Baker S. Teaching reading comprehension to students with learning disabilities: A review of research. *Review of Educational Research*. 2001; 71:279–320.
- Gosch A, Städing G, Pankau R. Linguistic abilities in children with Williams-Beuren syndrome. *American Journal of Medical Genetics*. 1994; 52:291–296. [PubMed: 7528971]
- Grant J, Karmiloff-Smith A, Gathercole SA, Paterson S, Howlin P, Davies M, Udwin O. Phonological short-term memory and its relationship to language in Williams syndrome. *Cognitive Neuropsychiatry*. 1997; 2:81–99.
- Hillier LW, Fulton RS, Fulton LA, Graves TA, Pepin KH, Wagner-McPherson C, et al. The DNA sequence of chromosome 7. *Nature*. 2003; 424:157–164. [PubMed: 12853948]
- Howlin P, Davies M, Udwin O. Cognitive functioning in adults with Williams syndrome. *Journal of Child Psychology and Psychiatry*. 1998; 39:183–189. [PubMed: 9669231]
- Jackendoff, R. Patterns in the mind: Language and human nature. New York: Basic; 1994.
- John AE, Rowe ML, Mervis CB. Referential communication skills of children with Williams syndrome: Understanding when messages are not adequate. *American Journal on Intellectual and Developmental Disabilities*. 2009; 114:85–99. [PubMed: 19391675]
- Kamil, ML. Vocabulary and comprehension instruction: Summary and implications of the National Reading Panel findings. In: McCardle, P.; Chhabra, V., editors. The voice of evidence in reading research. Baltimore, MD: Brookes; 2004. p. 213-234.
- Karmiloff-Smith A, Grant J, Berthoud I, Davies M, Howlin P, Udwin O. Language and Williams syndrome: How intact is “intact”? *Child Development*. 1997; 68:274–290.
- Kaufman, AS.; Kaufman, NL. Kaufman Brief Intelligence Test. Circle Pines, MN: American Guidance Services; 1990.
- Klein BP, Mervis CB. Cognitive strengths and weaknesses of 9- and 10-year-olds with Williams syndrome or Down syndrome. *Developmental Neuropsychology*. 1999; 16:177–196.
- Klein-Tasman BP, Mervis CB. Distinctive personality characteristics of 8-, 9-, and 10-year-old children with Williams syndrome. *Developmental Neuropsychology*. 2003; 23:271–292.
- Laing E. Investigating reading in atypical populations: The case of Williams syndrome. *Reading and Writing: An Interdisciplinary Journal*. 2002; 15:575–587.
- Laing E, Hulme C, Grant J, Karmiloff-Smith A. Learning to read in Williams syndrome: Looking beneath the surface of atypical reading development. *Journal of Child Psychology and Psychiatry*. 2001; 42:729–739. [PubMed: 11583245]
- Levy Y, Antebi V. Word reading and reading-related skills in Hebrew-speaking adolescents with Williams syndrome. *Neurocase*. 2004; 10:444–451. [PubMed: 15788284]
- Levy Y, Hermon S. Morphological abilities of Hebrew-speaking adolescents. *Developmental Neuropsychology*. 2003; 23:61–85.

- Levy Y, Smith J, Tager-Flusberg H. Word reading and reading-related skills in adolescents with Williams syndrome. *Journal of Child Psychology and Psychiatry*. 2003; 44:576–587. [PubMed: 12751849]
- Losh M, Bellugi U, Reilly J, Anderson D. Narrative as a social engagement tool: The excessive use of evaluation in narratives from children with Williams syndrome. *Narrative Inquiry*. 2000; 10:265–290.
- Lukács, A. Language abilities in Williams syndrome. Budapest, Hungary: Akadémiai Kiadó; 2005.
- Lukács, A.; Pléh, C.; Racsmány, M. Language in Hungarian children with Williams syndrome. In: Bartke, S.; Siegmüller, J., editors. *Williams syndrome across languages*. Amsterdam: John Benjamins; 2004. p. 187-220.
- Mayer, M. *Frog, where are you?*. New York, NY: Dial Press; 1969.
- McCardle, P.; Chhabra, V. *The voice of evidence in reading research*. Baltimore, MD: Brookes; 2004.
- McCardle, P.; Chhabra, V.; Kapinus, B. *Reading research in action: A teacher's guide for student success*. Baltimore, MD: Brookes; 2008.
- McKague M, Pratt C, Johnston MB. The effect of oral vocabulary on reading visually novel words: a comparison of the dual-route-cascaded and triangle frameworks. *Cognition*. 2001; 80:231–262. [PubMed: 11274984]
- Menghini D, Verucci L, Vicari S. Reading and phonological awareness in Williams syndrome. *Neuropsychology*. 2004; 18:29–37. [PubMed: 14744185]
- Mervis CB, Berra AM. Language and communicative development in Williams syndrome. *Mental Retardation and Developmental Disabilities Research Reviews*. 2007; 13:3–15. [PubMed: 17326109]
- Mervis CB, John AE. Vocabulary abilities of children with Williams syndrome: Strengths, weaknesses, and relation to visuospatial construction ability. *Journal of Speech, Language, and Hearing Research*. 2008; 51:967–982.
- Mervis, CB.; John, AE. Williams syndrome: Psychological characteristics. In: Shapiro, BK.; Accardo, PJ., editors. *Neurobehavioral disorders*. Baltimore, MD: Brookes; (in press)
- Mervis, CB.; Morris, CA. Williams syndrome. In: Mazzocco, MMM.; Ross, JL., editors. *Neurogenetic developmental disorders: Variation of manifestation in childhood*. Cambridge, MA: MIT Press; 2007. p. 199-262.
- Mervis, CB.; Morris, CA.; Bertrand, J.; Robinson, BF. Williams syndrome: Findings from an integrated program of research. In: Tager-Flusberg, H., editor. *Neurodevelopmental disorders*. Cambridge, MA: MIT Press; 1999. p. 65-110.
- Mervis CB, Robinson BF. Designing measures for profiling and genotype/phenotype studies of individuals with genetic syndromes or developmental language disorders. *Applied Psycholinguistics*. 2005; 26:41–64.
- Mervis CB, Robinson BF, Bertrand J, Morris CA, Klein-Tasman BP, Armstrong SC. The Williams Syndrome Cognitive Profile. *Brain and Cognition*. 2000; 44:604–628. [PubMed: 11104544]
- Mervis, CB.; Robinson, BF.; Rowe, ML.; Berra, AM.; Klein-Tasman, BP. Language abilities of individuals who have Williams syndrome. In: Abbeduto, L., editor. *International Review of Research in Mental Retardation*. Vol. 27. Orlando, FL: Academic Press; 2003. p. 35-81.
- Meyer-Lindenberg A, Mervis CB, Berman KF. Neural mechanisms in Williams syndrome: A unique window to genetic influences on cognition and behavior. *Nature Reviews: Neuroscience*. 2006; 7:380–393.
- Morris, CA. The dysmorphology, genetics, and natural history of Williams-Beuren syndrome. In: Morris, CA.; Lenhoff, HM.; Wang, PP., editors. *Williams-Beuren syndrome: Research, evaluation, and treatment*. Baltimore, MD: Johns Hopkins University Press; 2006. p. 3-17.
- Neale, MD. *Neale Analysis of Reading*. London, UK: Macmillan; 1966.
- Oakhill, J.; Cain, K. Introduction to comprehension development. In: Cain, K.; Oakhill, J., editors. *Children's comprehension problems in oral and written language: A cognitive perspective*. New York, NY: Guilford; 2007. p. 3-40.
- Oakhill JE, Cain K, Bryant PE. Dissociation of single-word reading and text comprehension skills. *Language and Cognitive Processes*. 2003; 18:443–468.

- Osborne, LR. The molecular basis of a multisystem disorder. In: Morris, CA.; Lenhoff, HM.; Wang, PP., editors. Williams-Beuren syndrome: Research, evaluation, and treatment. Baltimore, MD: Johns Hopkins University Press; 2006. p. 18-58.
- Pagon RA, Bennett FC, LaVeck B, Stewart KB, Johnson J. Williams syndrome: Features in late childhood and adolescence. *Pediatrics*. 1987; 80:85–91. [PubMed: 3601523]
- Peregrine, E.; Rowe, ML.; Mervis, CB. Pragmatic language issues in children with Williams syndrome. World Congress on Chromosome Abnormalities; San Antonio, TX. 2004 June.
- Piattelli-Palmarini M. Speaking of learning: how do we acquire our marvellous facility for expressing ourselves in words? *Nature*. 2001; 411:887–888.
- Pickering, SJ., editor. Working memory and education. Burlington, MA: Academic Press; 2006.
- Reilly J, Losh M, Bellugi U, Wulfeck B. “Frog, where are you?” Narratives in children with specific language impairment, early focal brain injury, and Williams syndrome. *Brain and Language*. 2004; 88:229–247. [PubMed: 14965544]
- Robinson BF, Mervis CB, Robinson BW. Roles of verbal short-term memory and working memory in the acquisition of grammar by children with Williams syndrome. *Developmental Neuropsychology*. 2003; 23:13–31. [PubMed: 12730018]
- Rowe, ML.; Mervis, CB. Working memory in Williams syndrome. In: Alloway, TP.; Gathercole, SE., editors. Working memory and neurodevelopmental conditions. Hove, England: Psychology Press; 2006. p. 267-293.
- Rust, J.; Golombok, S.; Trickey, G. WORD: Wechsler Objective Reading Dimensions. London, UK: Psychological Corporation; 1993.
- Searcy YM, Lincoln AJ, Rose FE, Klima ES, Bavar N. The relationship between age and IQ in adults with Williams syndrome. *American Journal on Mental Retardation*. 2004; 109:231–236. [PubMed: 15072519]
- Semel, E.; Wiig, EH.; Secord, WA. Clinical Evaluation of Language Fundamentals. 4. San Antonio, TX: Harcourt Assessment; 2003.
- Snowling MJ, Nash HM, Henderson LM. The development of literacy skills in children with Down syndrome: Implications for intervention. *Down Syndrome Research and Practice*. (in press).
- Stahl, SA. What do we know about fluency? Findings of the National Reading Panel. In: McCardle, P.; Chhabra, V., editors. The voice of evidence in reading research. Baltimore, MD: Brookes; p. 187-211.
- Stojanovik V, Perkins M, Howard S. Linguistic heterogeneity in Williams syndrome. *Clinical Linguistics & Phonetics*. 2006; 20:547–552. [PubMed: 17056485]
- Strømme P, Bjørnstad PG, Ramstad K. Prevalence estimation of Williams syndrome. *Journal of Child Neurology*. 2002; 17:269–271. [PubMed: 12088082]
- Temple CM. Developmental and acquired dyslexias. *Cortex*. 2006; 42:898–910. [PubMed: 17131596]
- Temple CM, Almazan M, Sherwood S. Lexical skills in Williams syndrome: a cognitive neuropsychological analysis. *Journal of Neurolinguistics*. 2002; 15:463–495.
- Terman, LM.; Merrill, MA. The Stanford-Binet Intelligence Scale: Third revision form L-M. Boston, MA: Houghton-Mifflin; 1973.
- Udwin O, Davies M, Howlin P. A longitudinal study of cognitive abilities and educational attainment in Williams syndrome. *Developmental Medicine and Child Neurology*. 1996; 38:1020–1029. [PubMed: 8913183]
- Udwin O, Yule W. Expressive language of children with Williams syndrome. *American Journal of Medical Genetics Supplement*. 1990; 6:108–114. [PubMed: 2118771]
- Udwin O, Yule W. A cognitive and behavioural phenotype in Williams syndrome. *Journal of Clinical and Experimental Neuropsychology*. 1991; 13:232–244. [PubMed: 1864913]
- Udwin O, Yule W, Martin N. Cognitive and behavioral characteristics of children with idiopathic infantile hypercalcaemia. *Journal of Child Psychology and Psychiatry*. 1987; 28:297–309. [PubMed: 3584299]
- Vicari S, Bates E, Caselli MC, Pasqualetti P, Gagliardi C, Tonucci F, Volterra V. Neuropsychological profile of Italians with Williams syndrome: An example of a dissociation between language and

- cognition? *Journal of the International Neuropsychological Society*. 2004; 10:862–876. [PubMed: 15637777]
- Volterra V, Capirci O, Pezzini G, Sabbadini L, Vicari S. Linguistic abilities in Italian children with Williams syndrome. *Cortex*. 1996; 32:663–677. [PubMed: 8954245]
- Volterra V, Caselli MC, Capirci O, Tonucci F, Vicari S. Early linguistic abilities of Italian children with Williams syndrome. *Developmental Neuropsychology*. 2003; 23:33–59. [PubMed: 12730019]
- Wagner, RK.; Torgesen, JK.; Rashotte, CA. *Comprehensive Test of Phonological Processing*. Austin, TX: PRO-ED; 1999.
- Wang PP, Bellugi U. Evidence from two genetic syndromes for a dissociation between verbal and visual-spatial short-term memory. *Journal of Clinical and Experimental Neuropsychology*. 1994; 16:317–322. [PubMed: 8021317]
- Wendon, L. *First steps in Letterland*. Cambridge, UK: Letterland Ltd; 1992.
- Wechsler, D. *Wechsler Adult Intelligence Scale-Revised*. New York, NY: Psychological Corporation; 1981.
- Wechsler, D. *Wechsler Individual Achievement Test-II Update 2005*. San Antonio, TX: Harcourt Assessment; 2005.
- Wechsler, D. *Wechsler Intelligence Scale for Children-Revised*. New York, NY: Psychological Corporation; 1974.
- Williams, KT. *Expressive Vocabulary Test*. Circle Pines, MN: American Guidance Service; 1997.
- Woodcock, RW.; Johnson, MB. *Woodcock-Johnson Psycho-Educational Battery-Revised*. Itasca, IL: Riverside; 1990.
- Zukowski, A. Investigating knowledge of complex syntax: Insights from experimental studies of Williams syndrome. In: Rice, M.; Warren, S., editors. *Developmental language disorders: From phenotypes to etiologies*. Cambridge, MA: MIT Press; 2004. p. 99-119.