# Language and Literacy Development of Children With Williams Syndrome

**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.

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Children with WS 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. 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 approximately 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 or above, 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.

## 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.

### Semantics

#### *Concrete vocabulary*

Concrete vocabulary has consistently been reported as the strongest language ability for individuals with Williams syndrome. From early reports on Williams syndrome to recent ones, performance on the PPVT has repeatedly yielded the highest mean standard score for any standardized assessment. In a 2007 study of 88 individuals with WS ages 4-46, 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 and once again, the majority of participants (82%) earned standard scores of 70 or above; 10% earned standard scores of 100 or above.

### 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. 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).

### Grammar

The majority of individuals with Williams syndrome have intellectual abilities in the borderline to mild disability range. Grammatical measures test understanding of a wide range of constructions ranging from simple positive statements to sentences containing center embedded clauses. Many individuals with Williams syndrome have considerable difficulty with grammatical comprehension, especially of complex constructions.

### 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 and word segmentation. When 55 children with Williams syndrome aged 6.03–12.90 years were tested, Mean T score was 40.24. Fourteen children (25%) scored at or above the mean for the general population. The correlation between CA (chronological age) and Phonological Processing Tscore 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.

### 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. When children with WS were asked to place one of several pictures into a scene based on the researcher's instructions they 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. They let 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. The performance of the children with Williams syndrome was considerably worse than that of the typically developing 3- to 6-year-old group in a study of Abbeduto et al. 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 anaphoric reference, and using context to establish meaning.

### Narrative structure and inference making

When compared with typically developing children at the same chronological age, the Williams syndrome group included significantly fewer story components and was considerably less likely to refer to the goals and motivations of the main character or to link his actions to his goal. But the William 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.

### 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 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.

### 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 or intellectual disability of unknown or mixed etiology.

### Verbal working memory

The verbal working memory abilities of children with WS were measured using the DAS–II Recall of Digits-Backward subtest (Mervis, 2009). On this supplemental subtest, the child is instructed to repeat strings of numbers presented by the examiner in reverse order. In each study, the Williams syndrome group earned a higher mean score than did the contrast group, 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 also been addressed, and a strong correlation was found between backward digit span and both receptive vocabulary and receptive grammatical ability. The correlation between backward digit recall and (Test for Reception of Grammar (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. A study conducted in England found a significant relation between the 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.

### 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 that 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.

### 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.

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. The results of all studies indicated that on average, the single word reading abilities of individuals with Williams syndrome are quite limited although there is considerable variability.

### Relation of primary reading instruction method to reading ability

The relation between primary reading instruction method and reading ability of children with Wil-

liams syndrome has been addressed in an ongoing study conducted in my laboratory (Becerra et al., 2008). We have used the WIAT–II to assess the reading ability of 44 children aged 9–17 years. The children's WIAT–II standard scores indicated a wide range of reading ability relative to CA matched 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 general congnitive abilities (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). After adjusting expected reading standard scores as a function of GCA the 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 all 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

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. Those 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.

### 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. 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 or her students are able to learn.**

Meta-analysis results 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 in increasing fluency (McCardle et al., 2008). 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.**

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").

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.

### Comprehension strategies

Children with WS 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 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).

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.

The 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. The ultimate level of reading achievement will be affected by a child's intellectual ability, among other factors, but 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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