

LANGUAGE AND COMMUNICATIVE DEVELOPMENT IN WILLIAMS SYNDROME

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Williams syndrome, a genetic disorder caused by a microdeletion of ~25 genes on chromosome 7q11.23, is associated with mild to moderate intellectual disability or learning difficulties. Most individuals with Williams syndrome evidence a cognitive profile including relative strengths in verbal short-term memory and language, and considerable weakness in visuospatial construction. The syndrome has often been argued to provide strong evidence for the independence of language from other aspects of cognition. We provide a brief history of early research on the language abilities of individuals with Williams syndrome and then review contemporary studies of language and cognition in Williams syndrome, beginning with a consideration of performance on standardized assessments. In the remainder of the article, we first consider early language acquisition, with a focus on speech production and perception, vocabulary acquisition, and communicative/pragmatic development and then consider the language abilities of school-age children and adolescents, focusing on semantics, grammar, and pragmatics. We argue that rather than being the paradigm case for the independence of language from cognition, Williams syndrome provides strong evidence of the interdependence of many aspects of language and cognition. © 2007 Wiley-Liss, Inc. MRDD Research Reviews 2007;13:3–15.

Key Words: Williams syndrome; language acquisition; intellectual disability; autism spectrum disorder; modularity

LANGUAGE AND COMMUNICATIVE DEVELOPMENT IN WILLIAMS SYNDROME

Williams syndrome is a neurodevelopmental disorder caused by a microdeletion of ~25 genes on chromosome 7q11.23 [Hillier et al., 2003; Osborne, 2006]. This syndrome, which has a prevalence of 1/7,500 [Strømme et al., 2002], is associated with a recognizable pattern of physical characteristics, including a specific set of facial features, heart disease (most commonly supralvalvar aortic stenosis), connective tissue abnormalities, failure to thrive, and growth deficiency [Morris, 2006]. Individuals with Williams syndrome have developmental delay which leads to mild to moderate intellectual disability or learning difficulties, although some individuals have low average to average intelligence. The syndrome is characterized by a specific cognitive profile including relative strengths in verbal short-term memory and language, and extreme weakness in visuospatial construction [Mervis et al., 2000]. Williams syndrome is also associated with a specific personality profile, including overfriendliness, anxiety, and empathy [Klein-Tasman and Mervis, 2003].

Research on language acquisition in Williams syndrome began quietly, with two articles [Kataria et al., 1984; Meyerson

and Frank, 1987] providing evidence that the language abilities of children with Williams syndrome were well below expectations for chronological age (CA) level and a third [MacDonald and Roy, 1988] indicating that language abilities were at the same level as a group of children with other types of developmental disabilities matched for CA and standard score on the Peabody Picture Vocabulary Test-Revised [Dunn and Dunn, 1981]. As would be expected for studies of such a rare syndrome (estimated prevalence at the time was 1/25,000–1/50,000), the number of participants in these studies was small (seven in both Kataria et al. 1984 and MacDonald and Roy, 1988), and the apparent level of general interest in either these findings or cognitive or linguistic aspects of Williams syndrome more generally was quite low.

Later in 1988, reporting on a sample even smaller than the previous ones, Bellugi et al. [1988] published a chapter that, combined with conference presentations and reports of conference presentations, was to have enormous ramifications for the study of language and cognition in Williams syndrome. In their seminal chapter, Bellugi et al. argued that although individuals with Williams syndrome had severe mental retardation and were functioning in Piaget's preoperational period even as adolescents, they nevertheless had excellent language abilities. In particular, although adolescents with Williams syndrome produced drawings with parts scattered over the page such that one could only identify the item depicted if the person indicated what he or she had drawn, and although these individuals were unable to conserve either number or quantity, they nevertheless were able to comprehend and produce complex linguistic constructions such as reversible passives, conditionals, and tag questions. In addition, they had excellent vocabularies. The combination of the ability to comprehend and produce reversible passives with the inability to conserve led Bellugi et al. to argue that Williams syndrome provided strong evidence of the independence of language from cognition. The message of this chapter and related presentations was clear: Williams syndrome is a case of severe cognitive disabilities and intact language abilities; or, Williams syndrome provides a strong dem-

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onstration of the independence of language (or a language module) from cognition.

This general message quickly attracted the attention of researchers interested in the relation between language and cognition. Bellugi et al.'s [1988] relatively focused argument that the ability to comprehend and produce reversible passives in the absence of concrete operational thought provided convincing evidence that language was independent of cognition in Williams syndrome was quickly forgotten. However, Bellugi et al.'s more general claim that the language ability of individuals with Williams syndrome was intact in the face of severe mental retardation became the rallying point for researchers and theorists espousing a modularity approach. Comparisons of the language and cognitive abilities of individuals with Williams syndrome, produced by authors who had not studied Williams syndrome directly, were considerably more strident than those offered by Bellugi et al. Three examples of such statements, in chronological order, and in order of increasing stridency, are as follows:

1. "Williams syndrome individuals almost invariably show mild to moderate mental retardation Particularly severe deficits show up in tests of spatial understanding such as copying patterns of blocks. Their language, though, is if anything more fluent and advanced than that of their age-mates . . ."; [Jackendoff, 1994, pp 116–117].
2. "The genes of one group of children [children who have specific language impairment] impair their grammar while sparing general intelligence; the genes of another group of children [children who have Williams syndrome] impair their intelligence while sparing their grammar" [Pinker, 1999, p 262].
3. "For instance, 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" [Piattelli-Palmarini, 2001, p 887].

Most researchers actively studying the language development of children who have Williams syndrome do not take

a modularity perspective, and many no longer even mention modularity in their articles. A few researchers [e.g. Clahsen and Almazan, 1998; Clahsen et al., 2003; Marshall and van der Lely, 2006] argue for a limited modularity position in their discussions of the acquisition of the past tense by children and adolescents with Williams syndrome. Other researchers set out to investigate the modularity position but conclude that their data are not supportive [e.g. Lukács, 2005] and still others point out that there is a wide range of linguistic ability among individuals with Williams syndrome and that this variability needs to be taken into account when making claims about the independence of language from cognition in Williams syndrome [e.g. Stojanovik et al., 2006]. In the remainder of this article, we briefly review current research on language acquisition in Williams syndrome, including studies whose authors have taken a modularity

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perspective, studies whose authors have argued for links between cognition and language, and studies whose authors have researched language acquisition in Williams syndrome without regard to the issue of relations between language and cognition.

PERFORMANCE ON STANDARDIZED ASSESSMENT MEASURES

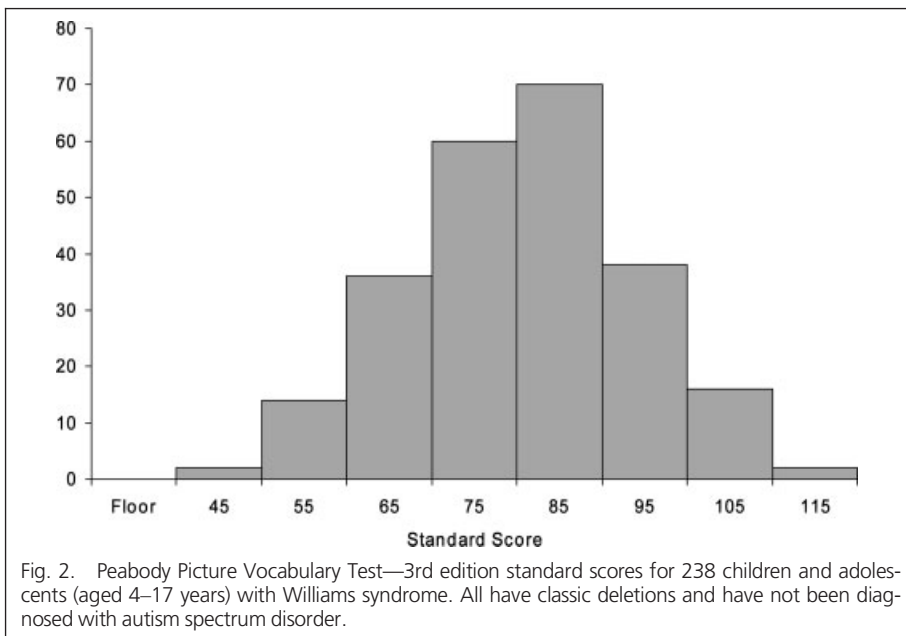
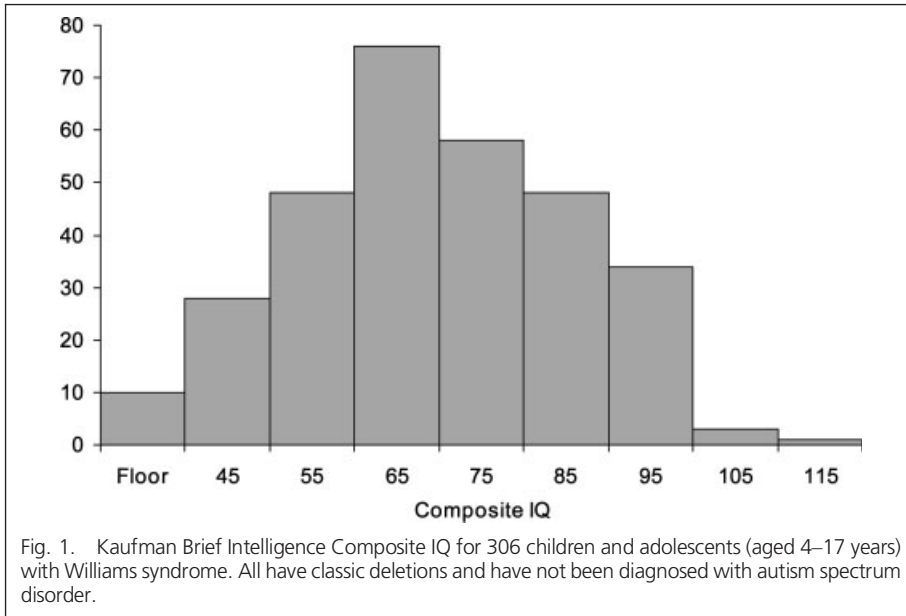
The most extreme claims about the independence of language from cognition in Williams syndrome are made by people who either have not studied the syndrome directly or who have tested very small samples. In this section, we consider the data from large samples of children and adolescents with Williams syndrome who have completed various standardized tests that provide evidence regarding the types of claims exemplified by the three quotations. All of the children included in the samples have "classic" Williams syndrome (as do

~98% of individuals with this syndrome; the remainder have either shorter or longer deletions) [Mervis and Morris, 2007] and have not been diagnosed with autism spectrum disorder (ASD). We first consider the question of whether general intelligence is "barely measurable." We then consider the claims for language being more advanced than expected for CA and for "exquisite mastery of vocabulary" by focusing on concrete vocabulary, which is the aspect of language for which children with Williams syndrome show the best performance [Mervis and Morris, 2007]. Finally, we consider the claims regarding syntax and sentence construction.

General Intellectual Ability

The Kaufman Brief Intelligence Test (KBIT) [Kaufman and Kaufman, 1990] is the most commonly used measure of general intellectual ability in studies of English-speaking individuals with Williams syndrome. Our laboratory has tested 306 children aged 4–17 years on this test. The distribution of standard scores is shown in Figure 1. As is clear from this figure, the description of general intelligence as "barely measurable" is highly inaccurate. Mean Composite intelligence quotient (IQ) is 69.32, which is at the top of the range for mild intellectual disability, with a range from 40 (the lowest possible standard score) to 112. For 47% of the participants, Composite IQ was 70 or higher. Only 3.6% earned the lowest possible IQ (40, which is at the bottom of the moderate intellectual disability range). The standard deviation was 15.36, which is similar to the standard deviation of 15 for the general population. Thus, the distribution of IQ in Williams syndrome has the same shape and variability as for the general population, but is depressed by about two standard deviations. Mean verbal IQ was 71.35 and mean nonverbal (matrices) IQ was 1 point higher, 72.47.

The KBIT does not assess visuospatial construction, the area of greatest weakness for individuals with Williams syndrome. When IQ is measured by a full-scale assessment that includes spatial ability as well as verbal ability and nonverbal reasoning ability, overall IQ is considerably lower. For example, on the Differential Ability Scales, School Age version, mean GCA (similar to IQ) for a sample of 119 children aged 8–17 years tested by our laboratory was 58.29, toward the lower end of the mild intellectual disability range [Mervis and Morris, 2007], with a standard deviation of 12.77 and a range from 24 (the lowest possible GCA using the extended norms) to 94. Despite the 10–



point difference between KBIT composite IQ and DAS GCA, the mean standard scores for the DAS Verbal Cluster (70.18) and Nonverbal Reasoning cluster (67.43) were similar to the Verbal and Nonverbal IQs for the KBIT; the primary reason for the lower GCA was very weak performance on the Spatial cluster (mean standard score: 55.54). Mean GCA was not a valid indicator of intellectual ability for the majority of children with Williams syndrome. For 80%, Verbal cluster standard score, Nonverbal Reasoning cluster standard score, or both was significantly higher than expected for GCA [Mervis and Morris, 2007; see also Meyer-Lindenberg et al., 2006]. Once again, “general intelligence” is clearly measurable.

Concrete Vocabulary

Studies of English-speaking children with Williams syndrome almost always assess concrete vocabulary using a version of the Peabody Picture Vocabulary Test (PPVT), which measures receptive vocabulary with a focus on names for objects, actions, and descriptors. From the early studies [e.g. Bellugi et al., 1988] to the present [e.g. Mervis and Morris, 2007], performance on this test has consistently yielded the highest mean standard score for any assessment. Our laboratory has tested 238 children (aged 4–17 years) with Williams syndrome on the third edition of the PPVT (PPVT-III) [Dunn and Dunn, 1997]. The distribution of standard scores is shown in Figure 2. Mean standard score

was 79.85 (in the low average range), with a standard deviation of 13.63 and a range from 40 (lowest possible standard score using the extended norms is 20) to 118. The majority of children (78%) scored at least 70, and 8% scored at least 100. Nevertheless, performance on this measure clearly was not above the level expected for children in the general population and is not consistent with “exquisite” mastery of vocabulary. It is also worth noting that even for individuals with Down syndrome, who are generally characterized as showing either a flat cognitive profile or a relative strength in nonverbal cognition and relative weakness in verbal ability, standard score on the PPVT is typically the highest standard score earned [e.g. Glenn and Cunningham, 2005].

Syntax and Sentence Construction

The Formulated Sentence subtest of the Clinical Evaluation of Language Fundamentals, 4th edition (CELF-4) [Semel et al., 2003] assesses the child’s ability to construct a sentence about a picture using a target word provided by the examiner. Sentences are considered correct if they include the target word, are grammatically and semantically correct, and bear any relation to the picture. Sentences that contain only one error receive partial credit. The subtest yields a scaled score ranging from 1 to 19, with a mean scaled score of 10 and a standard deviation of 3. Our laboratory has tested 61 individuals (aged 8–17 years) with Williams syndrome on this subtest. The distribution of scaled scores is shown in Figure 3. Mean scaled score was 4.31 (in the test authors’ range of moderate language impairment) with a standard deviation of 3.35 and a range from 1 (lowest possible scaled score) to 12. The modal scaled score was 1. The participants made both grammatical errors (e.g., deletion of tense markers or auxiliaries, failure to include a mandatory second clause for relational terms such as “if”) and semantic errors (e.g., incorrect uses of relational terms such as “third,” “when,” or “although”). Sentences involving conjunctions such as “if” or “although” often contained both grammatical and semantic errors. When parents were asked if their children commonly made these types of errors in everyday conversation involving the types of words included in this subtest, they almost always responded affirmatively. Stojanovik et al. [2006] reported Formulated Sentences scaled scores for five British children (aged 7–12 years) with Williams syndrome; mean scaled score was 3.60; errors were similar to those produced by the individuals in our sample. Sentence construction clearly is neither exquisite

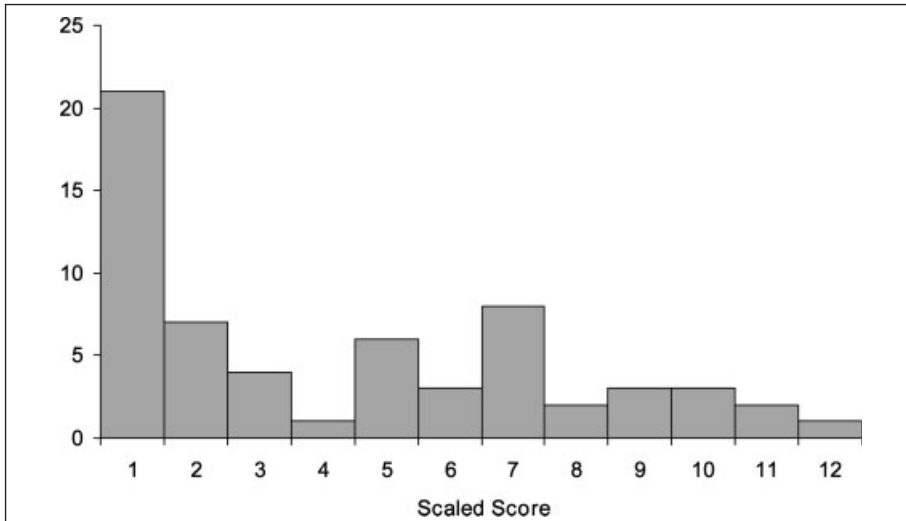


Fig. 3. Clinical Evaluation of Language Fundamentals—4th edition. Formulated Sentences subtest scaled score for 61 children and adolescents (aged 7–14 years) with Williams syndrome. All have classic deletions and have not been diagnosed with autism spectrum disorder.

nor at/above age level for most individuals with Williams syndrome.

Grammatical comprehension is most often tested using a version of the Test for Reception of Grammar (TROG). Our laboratory has tested 110 individuals (aged 5–18 years) with Williams syndrome on the second edition of the TROG (TROG-2) [Bishop, 2003]. Mean standard score was 70.25, with a standard deviation of 16.33 and a range from 55 (lowest possible standard score) to 111 [Mervis and Morris, 2007]. The modal score on this measure was also the lowest possible standard score (55). Thus, the mean score is likely artificially inflated relative to other standardized assessments for which the lowest possible standard score is 40.

Relations Among Scores on Standardized Assessments of Language and Cognition

The standardized assessment scores reported earlier are clearly not consistent with excellent language abilities. Nevertheless, scores on language measures are significantly higher than on spatial measures (e.g., DAS Verbal cluster standard score is significantly higher than DAS Spatial cluster standard score). Thus, the possibility remains that even if performance is not as strong as expected given the three quotations, performance on language measures might be independent of performance on cognitive measures, thus still providing evidence for a modularity position. We have addressed this possibility explicitly [Mervis, 1999; Mervis et al., 2004] by examining the correlations among performance on a series of language (PPVT-R, KBIT verbal, TROG), memory (forward digit span, backward digit span), and

other cognitive measures (KBIT matrices, DAS pattern construction) for a sample of 50 school-age children and adults with Williams syndrome, controlling for CA. All of the correlations were significant. Further, partial correlation analyses indicated that the significant correlations between DAS pattern construction and the language measures were mediated primarily by nonverbal reasoning (KBIT matrices) and working memory (backward digit span). These findings indicate that language abilities of school-age children and adults with Williams syndrome are not independent of their cognitive abilities.

As reviewed below, several other studies have considered the relations between language ability and memory ability for children with Williams syndrome; the authors have concluded that language ability is much more strongly correlated with memory ability for children with Williams syndrome than for typically developing children.

EARLY LANGUAGE ACQUISITION

Studies of early language acquisition by children with Williams syndrome focus on speech production and perception, vocabulary, and pragmatics/sociocommunication. Each of the three topics is reviewed below.

Early Speech Production and Perception

The onset of language acquisition by children with Williams syndrome is almost always delayed [Mervis and Klein-Tasman, 2000]. Researchers studying early speech production and perception have argued that this delay is likely due at least in part to

delays in the onset of rhythmic productions in general (both linguistic and nonlinguistic) [Masataka, 2001] or to delays in the development of the ability to segment words out of the speech stream [Nazzi et al., 2003].

To examine the relations among early motor and language milestones for infants and toddlers with Williams syndrome, Masataka [2001] conducted a longitudinal study of eight children from ages 6–30 months, with biweekly data collection (two sessions on adjacent days, every 2 weeks). All motor and linguistic milestones were delayed, with varying amounts of delay for different children. The linguistic milestones studied were canonical babble (defined as the first session in which the proportion of canonical syllables relative to all nonvegetative sounds was at least 0.2) [see Oller, 1986] and first words (defined as the first session in which the child's cumulative lexicon reached 25 words that were used appropriately although perhaps in an overgeneralized manner; words could be spontaneous or imitated). Motor milestones included rolling from stomach to back, unsupported sitting, reaching for objects, pulling to a stand, first steps, and rhythmic hand banging (defined as repetitive banging with an open hand on a horizontal surface, demonstrated in at least two of the four sessions from a 2-week period; onset was the date of the first of the two sessions). Rhythmic hand banging was the only motor milestone whose onset was significantly correlated with the onsets of the linguistic milestones. The correlation between the onsets of canonical babble and first words also was significant. Mean age of onset was 74.50 weeks for rhythmic hand banging, 76.50 weeks for canonical babble, and 98.50 weeks for first words. All children attained rhythmic hand banging either before or in the same session as the onset of canonical babble, and all children attained canonical babble at least 18 weeks prior to the first words milestone. This same pattern of correlations had previously been demonstrated for both full-term and preterm typically developing children, although at much younger ages [Eilers et al., 1993]. When the children with Williams syndrome first began to produce canonical syllables, their production was facilitated if they simultaneously produced rhythmic hand banging. Masataka argued that rhythmic hand banging provides the motor substrate for canonical babble (which is both a motor and a linguistic milestone) and that without canonical babble, the production of words is for the most part impossible. Thus, the delay in onset of rhythmic hand banging would be

expected to result in a delay in the onset of canonical babble, and the delay in the onset of canonical babble would be expected to result in a delay in the onset of first words. Consistent with Masataka's findings, Mervis and Bertrand [1997] reported that for the two children with Williams syndrome in their longitudinal study, who were not producing canonical babble when they entered the study, onset of rhythmic hand banging and canonical babble occurred in the same month.

Preliminary results from an ongoing study of phonological development by six children with Williams syndrome who are participating in a longitudinal study in our laboratory have recently been presented [Velleman et al., 2006]. The performance of these children at age 18 months was compared with that of a CA-matched sample of children with Down syndrome [Velleman et al., 1989] and CA-matched samples of typically developing children [Stoel-Gammon, 1989; Velleman et al., 1989; Rvachew et al., 2005]. The three groups of children produced about the same mean proportion of canonical babble syllables, with much greater variability shown by the Williams syndrome and Down syndrome groups than the typically developing group. One child in each of the syndrome groups had not yet met the criterion for the production of canonical babble, as described in the Masataka [2001] study. The two syndrome groups produced a considerably higher proportion of V-alone syllables (a more immature pattern) and a lower average number of syllables per babble (also a more immature pattern) than the typically developing group, with the Williams syndrome group evidencing considerably more variability than the other groups. The two syndrome groups also produced fewer different consonants per session. Finally, the mean Babble Level as defined by Stoel-Gammon [1989] was lower for the two syndrome groups, indicating that the babble of the typically developing children was more complex, typically including more than one true consonant per babble, whereas that of the children with Williams syndrome or Down syndrome was more likely to include only a single true consonant or in some cases, no true consonant. Consistent with Masataka's [2001] argument that canonical babble is critical for language production, the two children with Williams syndrome in this sample whose language development was the most advanced for their age had the most "normal" babble histories and the child whose language was the most delayed had not yet met criterion for attainment of canonical babble even at age 36 months.

The speech perception abilities of young children with Williams syndrome have been addressed in only one study [Nazzi et al., 2003]). These researchers argued that the language delay associated with Williams syndrome may be due in large part to difficulty segmenting words from the speech stream. Jusczyk et al. [1999] demonstrated that at age 7.5 months, typically developing infants are able to segment words with a strong-weak stress pattern (the predominant pattern in English) out of ongoing speech if they had previously been familiarized with the words in isolation. However, typically developing infants were not able to segment words with a weak-strong stress pattern out of ongoing speech until 10.5 months. At 7.5 months, infants use prosodic information as the primary cue to word segmentation and pay little attention to distributional information, leading to successful segmentation of strong-weak words but not weak-strong words.

To successfully segment weak-strong words, infants begin to weight

These findings indicate that language abilities of school-age children and adults with Williams syndrome are not independent of their cognitive abilities.

distributional information more strongly, leading to the ability to segment both strong-weak and weak-strong words from the speech stream. Nazzi et al. replicated Jusczyk et al.'s research using the youngest participants with Williams syndrome available; the resulting CA range for the 17 participants was 15-47 months (9-26 months MA as measured by the Bayley Scales of Infant Development-II; Bayley, 1993), with a mean CA of 33 months and a mean MA of 19 months. The Williams syndrome group was able to segment the strong-weak words but not the weak-strong words from the speech stream.

This study does not address the question of whether onset of the ability to segment strong-weak words from the speech stream is delayed in Williams syndrome, because the youngest child in the study was considerably older than the age at which this ability is evidenced by typically developing children. However, the data provide clear evidence that onset of

segmentation of weak-strong words from the speech stream is considerably delayed for children with Williams syndrome. The impact of this delay on early language acquisition depends in part on whether parents provide words in isolation; if the child who is just beginning to learn to talk often hears words in isolation (and in conjunction with their referents), then the ability to segment words out of the speech stream would be considerably less critical than if words are not produced in isolation. The fact that the parents of 13 of the 17 children reported that their child comprehended "balloon" (one of the target weak-strong words) suggests that these children with Williams syndrome are able to acquire weak-strong words without being able to segment them out of the speech stream. Furthermore, the rarity of weak-strong words in the early lexicons of typically developing children learning English is attested to by the fact that only 8 of the 396 words on the MacArthur-Bates Communicative Development Inventory, Words and Gestures version (CDI) [Fenson et al., 1993, 2007] fit the weak-strong pattern. Nevertheless, the delay in ability to segment weak-strong words out of the speech stream (and the possible delay in learning to segment strong-weak words out of the speech stream) means that an important route for acquiring new vocabulary is unavailable to young children with Williams syndrome, thus limiting the contexts in which new words can be acquired and likely leading to a reduction in rate of vocabulary acquisition even though the isolated-word route is still available.

Early Vocabulary Acquisition

As would be expected based on the early speech production findings, early vocabulary acquisition is almost always delayed. The results of a longitudinal analysis of the early vocabularies of 13 children with Williams syndrome who had classic deletions [Mervis et al., 2003] indicated that age of acquisition of a 10-word expressive vocabulary was below the 5th percentile (the lowest percentile provided) for the norms for the Early Vocabulary Checklist included in the CDI, Words and Sentences version [Fenson et al., 1993] for all 13 children. For 12 of the 13 children, age of acquisition of 50- and 100-word expressive vocabularies was also below the 5th percentile. Mean age of acquisition of a 100-word expressive vocabulary was 40.90 months (range: 26.24-68.05 months). In contrast, the 50th percentile for acquisition of a 100-word vocabulary is 18 months for typically

developing children and the 5th percentile is 28 months [Fenson et al., 2007].

We also have considered the expressive vocabulary growth patterns (nonlinear versus linear) of a group of children with Williams syndrome followed longitudinally [Mervis, 2004]. Although the onset of expressive vocabulary acquisition was delayed for all of the children, once vocabulary acquisition began, 15 of 17 demonstrated a nonlinear growth pattern similar to that of typically developing children. The two remaining children demonstrated very slow linear growth at least through age 48 months. Comparison of the children's intellectual abilities at 48 months as measured by the Differential Ability Scales, Preschool version [Elliott, 1990] indicated that 14 of the 15 children in the nonlinear group performed above floor on both the Verbal and the Nonverbal clusters; the 15th child performed above floor on the Nonverbal cluster only. All 15 children were able to repeat at least one digit and were producing productive (as opposed to frozen phrases only) two-word combinations. In contrast, the two children in the linear group performed at floor on both clusters of the DAS, were not able to repeat even a single digit, and were not yet producing novel word combinations. The same pattern held for the small sample of children with Down syndrome (two nonlinear, four linear) included in the analyses, indicating that nonlinear vocabulary growth is associated not only with earlier production of novel word combinations but also with growth in verbal short-term memory and in nonverbal cognition.

Two cross-sectional studies comparing the vocabulary sizes of young children with genetically-confirmed Williams syndrome to young children with Down syndrome have been conducted. Mervis and Robinson, [2000] found that 30-month-olds with Williams syndrome (mean: 132 words, range: 3–391 words) had significantly larger expressive vocabularies than 30-month-olds with Down syndrome (mean: 79 words, range: 0–324 words). Vicari et al. [2002] found that children with Williams syndrome had equivalent expressive vocabulary sizes to those of somewhat older children with Down syndrome matched for MA. However, the children with Williams syndrome had significantly more advanced grammatical ability and verbal memory skills [see also Volterra et al., 2003].

Studies of typically developing children and children who have Down syndrome have identified several links between early cognitive development and early lexical development that hold for both groups [see summary in Mervis and Bertrand, 1993]. We have used the data from our lon-

gitudinal study to determine if these links also hold for children with Williams syndrome [Mervis and Bertrand, 1997; Mervis, 2006]. The first link is that the extension of children's early object labels (as measured by comprehension or production) corresponds to the children's play patterns with these objects, with both at what Mervis [e.g. 1984, 1987] called the child-basic level. This pattern was strongly supported for children with Williams syndrome; for all 10 children, early object label extensions and play patterns corresponded to the child-basic level. For example, these children rolled a wide range of spherical objects, whether or not they were balls; they also comprehended and produced "ball" in relation to these objects). This pattern of behavior obtained even when parents tried to correct their children. The second proposed link involves the onset of spontaneous

***The nonverbal
communicative abilities of
the toddlers with Williams
syndrome were strongly
related to both their
receptive and expressive
language abilities and their
nonverbal reasoning and
visuospatial constructive
abilities.***

sorting of objects and the onset of fast mapping of object labels. Gopnik and Meltzoff [1987, 1992] argued that these onsets should happen at about the same time because they represent parallel insights: all objects belong to some category (cognitive insight) and all objects have a name (linguistic insight). For 9 of the 10 children with Williams syndrome, the onset of both spontaneous sorting and fast mapping was demonstrated in the same session. For the remaining child, the onset of fast mapping preceded the onset of spontaneous exhaustive sorting by about 2 weeks.

In contrast, a well-established link between an important early communicative ability (referential pointing) and the onset of referential language does not hold for children with Williams syndrome. For both typically developing children and children with Down syndrome, the onset of referential pointing occurs prior to the onset of

referential language [Adamson, 1995; Mervis and Bertrand, 1997]. This sequential ordering is one of the most robust findings in early lexical development and presumably occurs because the cognitive manifestation of reference (pointing) provides the child with a particularly useful way to determine the referents of words. Adults routinely use pointing to identify the referents of labels, and typically developing infants and toddlers with Down syndrome routinely use pointing to indicate interesting objects and to request labels. The pattern for almost all toddlers with Williams syndrome is very different: Only 1 of the 10 children with Williams syndrome (the child who was the most delayed) produced pointing gestures prior to beginning to produce referential language; the difference in onsets was about 3 weeks. The other nine children produced referential language prior to producing or following referential pointing gestures; referential pointing began an average of 6 months after referential language, with some children showing a delay of 12 months or more. Because joint attention to the referent is necessary for the child to acquire labels, alternative methods of establishing joint attention must be used. Mervis and Bertrand [1993, 1997] identified three such methods: (1) The communicative partner follows in to the child's focus of attention and then labels the object. (2) The communicative partner moves the object to the place where the child already was looking and then labels it. (3) The communicative partner directs the child's attention to the object by tapping it. All of these methods are used by parents of typically developing children or children with Down syndrome as well as by parents of children with Williams syndrome.

Early Communicative/Pragmatic Development

The late onset of comprehension and production of pointing gestures presages significant communicative/pragmatic difficulties for toddlers and preschoolers with Williams syndrome. Laing et al. [2002] used the Early Social Communication Scales [Mundy and Hogan, 1996] to compare the performance of a group of young children with Williams syndrome with that of an MA-matched group of typically developing infants and toddlers. Although the children with Williams syndrome had significantly larger expressive vocabularies, they were significantly less likely than the typically developing children to comprehend or produce pointing gestures or to engage in triadic joint attention (attention to both a communicative partner and an object). Rowe et al. [2005] used a similar measure, the Behavior Sam-

ple of the Communication and Symbolic Behavior Scales, Developmental Profile [Wetherby and Prizant, 2002], to compare the communicative abilities of toddlers with Williams syndrome (mean CA = 27.4 months) to those of toddlers with Down syndrome individually matched for CA, Mullen Scales of Early Learning [Mullen, 1995] composite score, and CDI expressive vocabulary size. The two groups did not differ on number of different consonants produced or number of different words produced. However, there were large differences in nonverbal communication. The children with Williams syndrome produced significantly fewer gaze shifts, engaged in significantly fewer episodes of triadic joint attention, and used significantly fewer distal gestures (including points) and significantly fewer conventional gestures. At the same time, the nonverbal communicative abilities of the toddlers with Williams syndrome were strongly related to both their receptive and expressive language abilities and their nonverbal reasoning and visuospatial constructive abilities as measured by the Mullen.

Although Williams syndrome has often been described as the “opposite” of autism, especially in the media, the types of communicative problems identified in the Laing et al. [2002] and Rowe et al. [2005] studies overlap with the difficulties associated with autism spectrum disorders. To provide a more in-depth examination of the sociocommunicative abilities and limitations of young children with Williams syndrome using a semistructured measure specifically designed to capture difficulties in sociocommunication, Klein-Tasman et al. [in press] administered the Autism Diagnostic Observation Schedule (ADOS) [Lord et al., 2000] Module 1 to 29 children aged 30–63 months (mean CA = 42 months) with Williams syndrome. Module 1 is designed for children who have very limited to no expressive language. The results confirmed and extended previous findings of sociocommunicative difficulties for young children with Williams syndrome. More than half of the children evidenced difficulties with pointing, other gestures, giving, showing, and appropriate use of eye contact. Many children also showed difficulties with initiation of joint attention or response to examiner bids for joint attention and with integration of gaze with other behaviors. These difficulties were such that 14 of the 29 children met or exceeded the ADOS algorithm cut-off for “autism spectrum disorder”; three of these children met or exceeded the ADOS algorithm cut-off for “autism.” The ADOS alone is not sufficient to make a diagnosis of autism or au-

tism spectrum disorder, and the Klein-Tasman et al. study was not intended to make this diagnosis. Subsequent to this study, however, all three children who met the ADOS algorithm for autism were clinically diagnosed with autism, two based on a combination of the ADOS, the autism diagnostic interview revised (ADI-R) [Lord et al., 1994], and clinical judgment and one based on independent clinical judgment. Klein-Tasman et al. argue that difficulties with pointing, showing, and giving are characteristics of Williams syndrome and should not be considered diagnostic for autism spectrum disorder in children with this syndrome. At the same time, however, when these difficulties are combined with difficulties directing vocalizations or facial expressions to other people and the quality of social overtures is generally poor, Klein-Tasman et al. suggest

Although Williams syndrome has often been described as the “opposite” of autism, especially in the media, the types of communicative problems identified . . . overlap with the difficulties associated with autism spectrum disorders.

that the consideration of a comorbid autism spectrum disorder may be warranted.

LANGUAGE ABILITIES OF SCHOOL-AGE CHILDREN AND ADOLESCENTS

Studies of the language abilities of school-age children and adolescents with Williams syndrome have focused primarily on semantics and syntax or morphology; recently, a few studies of pragmatics have been reported. The relations between language ability and verbal memory ability have also been studied.

Semantics

Semantic organization

Semantic organization (how a person cognitively relates the members of a category) is usually measured by word fluency tests in which a person is asked to list

as many members of a particular category as possible. The original studies of semantic organization were conducted by Bellugi et al. [1992, 1994], who compared the semantic organization of the “animal” category for six adolescents with Williams syndrome, six CA- and IQ-matched adolescents with Down syndrome, and a group of typically developing second graders. Based on the finding that the Williams syndrome group was more likely than the other two groups to name unusual (defined as low word-frequency) animals, the researchers concluded that the semantic organization of individuals with Williams syndrome was deviant. Bellugi et al. [2000] noted that at age 11 years, there is a sudden increase in the number of animal exemplars listed by individuals with Williams syndrome, with further steady increases through age 19 years and then a leveling off.

The results of more recent studies suggest that semantic organization in Williams syndrome is appropriate. Mervis et al. [1999] compared the responses of twelve 9- and 10-year-olds with Williams syndrome to those of a CA- and MA-matched group of children with Down syndrome, a CA-matched group of typically developing children, and an MA-matched group of typically developing children. Children were asked to list all the animals that they could. Eight measures of semantic organization were used, including measures of word frequency and representativeness; the Williams syndrome, Down syndrome, and MA-matched typically developing groups performed equivalently on seven of these measures. On the remaining measure the representativeness of the least representative category member listed, the Williams syndrome and Down syndrome groups performed equivalently to the CA-matched typically developing group.

The children with Williams syndrome in the Mervis et al. [1999] study were all younger than 11 years. Because Bellugi et al. [2000] argued that there is a steep increase in the number of items produced by children with Williams syndrome beginning at age 11 years, perhaps the differences between the two sets of findings are due to differences in the age of the participants; many of Bellugi et al.’s participants were 11 years or older. Lukács [2005] studied word fluency in 12 Hungarian-speaking children and adolescents with Williams syndrome (mean CA = 13.5 years) relative to a younger group of typically developing children matched for raw score on the Hungarian version of the PPVT. The two groups did not differ on the number of category exemplars pro-

duced or on the average word frequency of the exemplars produced. The Williams syndrome group did produce more items with a 0 word frequency. However, unlike Bellugi et al.'s [1992, 1994] findings, the exemplars produced that had 0 word frequency were not unusual but rather "pet" names for animals; such words were not produced by the control participants as they are normally produced only by very young children. The results of a study of Italian-speaking children and adolescents with Williams syndrome [Volterra et al., 1996] also indicated that the semantic organization of this group was similar to that of an MA-matched group of typically developing children. Finally, Levy and Bechar [2003] compared a group of Israeli children and adolescents with Williams syndrome to a CA- and IQ-matched group with intellectual disability of unknown etiology and found no between-group differences in semantic fluency.

Spatial language

Williams syndrome is associated with an extreme weakness in visuospatial construction (e.g., pattern construction, drawing). Because spatial language has been argued to provide important cues to non-verbal spatial representation [e.g. Bowerman, 1996], Bellugi et al. [2000] argued that individuals with Williams syndrome will have considerably more difficulty with spatial language than with other types of language. In support of this position, Bellugi et al. report the results of a study comparing adolescents and adults with Williams syndrome to a typically developing group with a mean CA of 11 years. On a test of spatial prepositions, the Williams syndrome group made significantly more errors (11%) than the typically developing group (0%). On a test in which the participant was asked to describe the spatial position of a colored object relative to a non-colored object, the Williams syndrome group again performed significantly worse than the control group, often making figure-ground reversals. Bellugi et al. interpreted these results as indicating that individuals with Williams syndrome have particular difficulty with spatial language. In contrast, Landau et al. [2006] suggested that these difficulties may instead be due to problems in the alignment of the components of the sentence during the process of sentence production.

Landau and Zukowski [2003; Landau et al., 2006] also considered the possible effect of difficulties with spatial representation on spatial language. These researchers compared the performance of 12 children with Williams syndrome (mean CA = 9.6 years) to those of an MA-matched typically developing group

(mean CA = 5.0 years) and a group of college students. Participants watched a series of 80 video clips of events containing spatial relations and described what happened. In this sample, the children with Williams syndrome only reversed figure and ground 1% of the time. The three groups tended to use the same verbs, and the most common path descriptions were the same for all three groups. However, the Williams syndrome group was significantly more likely to omit the path term (e.g., for a video of a box falling off a wall, to say, "The box fell off," rather than "The box fell off the wall"). They were especially likely to omit the path term for bounded-from paths or for via-paths, which require memory for two locations; the path term was more likely to be included for bounded-to paths, which require memory for only one location. Landau and Zukowski concluded that children with Williams syndrome have good control over much of the language needed to describe spatial events, including the semantic-syntactic mapping between spatial representation of the event and linguistic structure. They argued that the difficulty with path description is due to problems with spatial memory, which is significantly weaker than verbal memory [Rowe and Mervis, 2006]. Lukács [2005] found that children and adolescents with Williams syndrome made more errors on spatial language than typically developing children matched for Hungarian PPVT raw score, but that the pattern of errors for the Williams syndrome group was the same as the pattern for typically developing children. Lukács et al. [2004] conclude that spatial memory problems contribute to the particular difficulty that children with Williams syndrome have with bounded-from and via paths.

We have compared the performance of 86, 5–7-year-olds with Williams syndrome on the PPVT-III, which measures primarily concrete vocabulary, and the Test of Relational Concepts (TRC) [Edmonston and Litchfield Thane, 1988], which measures five types of relational concepts: temporal (e.g., before/after), quantitative (e.g., most/least), dimensional (e.g., long/short), spatial (e.g., under/over), and other (e.g., same/different). All of the children earned lower standard scores on the TRC than on the PPVT-III, with a mean difference of 29 points [Mervis and Morris, 2007]. On average, children with Williams syndrome perform worse on the TRC than on any other standardized assessment except those measuring visuospatial construction or self-help skills [Mervis and Morris, 2007]. However, a comparison of the TRC per-

formance of a group of children with Williams syndrome and a typically developing group matched for raw score on the TRC indicated no significant between-group differences as a function of type of relational concept. Thus, the relational vocabularies of the two matched groups contained similar distributions of types of relational words, indicating that children with Williams syndrome have difficulty with relational language in general, rather than specifically with spatial terms. Mervis and Morris argued that this pattern is consistent with Walsh's [2003] argument that spatial, temporal, and quantitative processing are all controlled by a common magnitude system, the posterior section of which is located in the inferior parietal cortex, a region in which Meyer-Lindenberg et al. [2004, 2006] have identified a structural abnormality for normal-IQ adults with Williams syndrome that serves as a roadblock to dorsal stream information flow. The results of a path analysis based on fMRI studies of normal-IQ adults with Williams syndrome and a CA- and IQ-matched control group of adults in the general population indicated that the only difference between the two groups was that the path from the intraparietal sulcus (the area with the structural abnormality) to the later dorsal stream region was significant only for the control group. This finding, combined with Walsh's theory, suggests that individuals with Williams syndrome should have difficulty with spatial, temporal, and quantitative concepts, consistent with the TRC findings.

Grammar

Bellugi and her colleagues [e.g. 1988, 1992, 1994, 2000] have argued that grammatical ability is a particular strength for individuals with Williams syndrome and that this ability in light of severely limited cognitive abilities provides a compelling case for the independence of language from cognition. In particular, older children and adolescents with Williams syndrome were able to produce and comprehend complex grammatical constructions such as passives, tag questions, relative clauses, and conditionals even though they were not able to conserve either number or quantity, the two types of conservation that are acquired earliest. Bellugi et al. argued that from a Piagetian perspective, comprehension and production of constructions such as reversible passives should not have been possible for individuals with Williams syndrome, since they had not yet attained concrete operations; comprehension and production of these constructions depended on reversible thought as exemplified by success on conservation tasks

[Beilin, 1975]. Ironically, even before Bellugi made this argument, Maratsos et al. [1979] had shown that Beilin's claim was not correct. In particular, Maratsos et al. found that typically developing English-speaking children are able to comprehend and produce reversible passives during the preoperational period, thus indicating that there was no link between acquisition of reversible passives and conservation during the normative process of development [see also Brooks and Tomasello, 1999]. Furthermore, results of studies of non-Indo-European languages in which passive constructions are common indicate that 2-year-olds spontaneously produce reversible passives [Allen and Crago, 1996]. Thus, the finding that adolescents with Williams syndrome were able to produce reversible passives even though they were not able to conserve did not address the question of the independence of language from cognition.

As a second part of their argument, Bellugi et al. [1992, 1994, 2000] showed convincingly that adolescents with Williams syndrome had considerably stronger grammatical abilities than CA- and IQ-matched adolescents with Down syndrome. Recent studies comparing the grammatical abilities of both English-speaking [Mervis et al., 2003] and Italian-speaking [Vicari et al., 2004] children with these two syndromes have confirmed Bellugi et al.'s initial findings. However, rather than demonstrating that the language abilities of individuals with Williams syndrome are much stronger than expected for level of cognitive ability, these results most likely reflect the inordinate difficulties that individuals with Down syndrome have with grammar. Studies comparing the grammatical abilities of individuals with Williams syndrome with either CA- and IQ-matched children with forms of intellectual deficiency other than Down syndrome or younger MA-matched typically developing children consistently indicate that the syntactic abilities of the individuals with Williams syndrome are at or slightly lower than the level of the comparison group. This finding has been shown to hold for individuals acquiring English [Udwin and Yule, 1990; Grant et al., 2002; Mervis et al., 2003; Zukowski, 2004], German [Gosch et al., 1994], Hungarian [Lukács, 2005], and Italian [Volterra et al., 1996, 2003]. Although most of these studies have focused on relatively young children, two [Grant et al., 2002; Zukowski, 2004] focused on adolescents or young adults and their ability to produce complex constructions such as relative clauses; the pattern of results was the same as for the studies of younger chil-

dren and simpler constructions. Lukács focused on older children, adolescents, and young adults and found that the pattern of performance on the Hungarian version of the TROG was the same as for younger typically developing children, although the Williams syndrome group performed more poorly overall. Furthermore, the constructions that were difficult for the Hungarian Williams syndrome group were the same as those that had been previously identified as difficult for Italian-speaking [Volterra et al., 1996] and English-speaking [Karmiloff-Smith et al., 1997] samples of individuals with Williams syndrome on the Italian and English versions of the TROG.

Studies of the morphological abilities of English-speaking individuals with Williams syndrome have focused primarily on the acquisition of the past tense. Researchers agree that by late childhood, most individuals with Williams syndrome reliably mark the past tense correctly on regular verbs but often over-regularize the past tense of irregular verbs. Little research on the developmental pattern of reduction of over-regularization over a broad age range has been conducted. Recently Peregrine et al. [2006] used the past tense section of the Rice/Wexler Test of Early Grammatical Impairment (TEGI) [Rice and Wexler, 2001] to examine age-related changes in past tense usage by individuals with Williams syndrome ages 6–46 years. Results indicated that of the children ages 6–8 years who produced at least one irregular past tense form, 62% produced more over-regularized forms than correct irregular forms and 20% produced only over-regularized forms. The corresponding figures for 9–11-year-olds were 28 and 8%; for 12–46-year olds they were 8 and 0%. Thus, there is a dramatic reduction in the production of over-regularized past tense forms with age, although over-regularization continues into adulthood for many individuals with Williams syndrome. The interpretation of the English past-tense findings is a subject of vigorous debate between researchers who consider the data to support a dual-mechanism model of language that is compatible with the existence of an independent grammar module [Clahsen and Almazan, 1998; Clahsen et al., 2003; Marshall and van der Lely, 2006] and those who consider the same data to support a single-mechanism model [Thomas et al., 2001; Thomas and Karmiloff-Smith, 2003]. The resolution of this debate is likely to depend on longitudinal studies of past tense acquisition.

Several studies of morphological development have been conducted using participants with Williams syndrome

acquiring languages that have more complex morphology than English. Results of these studies for French [Karmiloff-Smith et al., 1997], Hebrew [Levy and Hermon, 2003], and Hungarian [Lukács et al., 2001, 2004; Lukács, 2005] indicate that morphological ability is similar to or less advanced than that of younger typically developing children matched for MA. For a summary of these results, see Mervis [2006].

Pragmatics

Pragmatics continues to be an area of particular difficulty during the school years and into adulthood, and even very capable individuals with Williams syndrome commonly (and appropriately) have IEP goals targeting such aspects of pragmatics as turn taking, conversational and topic maintenance, and appropriate use of eye gaze [Mervis, 2006]. Nevertheless, there has been very little research on the pragmatic abilities of individuals beyond preschool age. Two studies have been conducted using the Children's Communication Checklist (CCC) [Bishop, 1998] or the revised version (CCC-2) [Bishop, 2003]; both are questionnaires completed by parents or other caregivers. Laws and Bishop [2004] used the CCC to study the pragmatic abilities of 19 children and young adults with Williams syndrome. Fifteen of the 19 met the CCC cut off for pragmatic language impairment. The Williams syndrome group evidenced significant difficulties in all five areas of pragmatics measured by the CCC, with particular difficulty in the use of stereotyped conversations, inappropriate initiation of conversations, and overdependence on context to interpret what was said to them. Peregrine et al. [2005] compared the CCC-2 standard scores of 53 6- to 12-year-olds with Williams syndrome to those of their siblings in the same age range. The children with Williams syndrome earned significantly lower scaled scores on all 10 of the scales included in the CCC-2. Because the CCC-2 does not include a cutoff for pragmatic language impairment, the findings could not be directly compared with those of Laws and Bishop. However, the types of problems identified were similar. These types of difficulties likely figure strongly in the problems that children with Williams syndrome have in forming and maintaining friendships with peers, despite a strong desire for such relationships.

Recently, the findings of a small-sample study comparing the conversational abilities of five children with Williams syndrome (mean CA 9.2 years) to those of eight children with specific language

impairment (mean CA 10.6 years) matched on TROG [Bishop, 1989] and British Picture Vocabulary Scale (the British version of the PPVT-R) performance and nine typically developing children (mean CA 8.75 years) were reported [Stojanovik, 2006]. Each participant engaged in a semi-structured 15–20 min conversation with a researcher. The children with Williams syndrome were less likely than the children with specific language impairment but more likely than the typically developing children to make grammatical or semantic errors. All three groups of children almost always answered the interlocutor's questions. However, the responses of the children with Williams syndrome revealed a number of pragmatic weaknesses. Regardless of whether the interlocutor asked for information or for clarification, the responses of the Williams syndrome group were more likely than the responses of the other groups to be inadequate. In particular, the children with Williams syndrome were more likely to provide too little information or to misinterpret what their conversational partner had meant and were considerably less likely to produce a response that continued the conversation. The latter problem was particularly striking; the child with Williams syndrome, who was most likely to produce responses that successfully continued the conversation, produced these responses at almost exactly the same rate as the child in each of the other groups who was least likely to produce conversational continuations.

Grammatical Development and Verbal Memory

As indicated earlier in this article, during the preschool period, verbal memory ability is associated with the early stages of grammatical development. In particular, 48-month-olds who could repeat at least one digit also produced novel word combinations [Mervis, 2004]; those who could not repeat at least one digit did not produce novel word combinations. Preschoolers with Williams syndrome had both more advanced verbal memory abilities and more advanced grammatical abilities than preschoolers with Down syndrome matched for MA and vocabulary size [Vicari et al., 2002; Volterra et al., 2003]. Vicari et al. [2002] also found that Italian CDI expressive vocabulary size was significantly and strongly correlated with sentence repetition ability for preschool and early school-age Italian-speaking children with Williams syndrome. Comparison of the performance of older children and adults on standardized assessments of verbal memory, vocabulary, and grammar also indicates strong relations between both forward and

backward digit span and vocabulary and grammatical ability [Mervis, 1999]. There have also been several studies of the relations between verbal memory and language (typically grammar) ability for school-age children with Williams syndrome. These studies are briefly reviewed in this section.

Verbal working memory is associated with both vocabulary acquisition [Gathercole and Baddeley, 1989, 1993] and grammatical ability [Kemper et al., 1989; Norman et al., 1992] for children and adults in the general population. Inordinate difficulty on the nonword repetition task, a measure of phonological memory, has been proposed as a phenotypic marker of specific language impairment [Bishop et al., 1996]. Similar relations have been found for children and adolescents with Williams syndrome.

Robinson et al. [2003] considered the relations among verbal rote memory

Children and adolescents with Williams syndrome rely more heavily on verbal working memory to successfully comprehend complex grammatical constructions than do typically developing children.

(forward digit recall), verbal working memory (backward digit recall), and phonological memory (nonword repetition) for 39 children and adolescents with Williams syndrome (mean CA = 10.2 years, range: 4.5–16.7 years) and 32 younger TD children matched for receptive grammatical ability as measured by raw score on the TROG. Results of a regression analysis indicated that for the Williams syndrome group, phonological memory contributed uniquely to variance in grammatical ability even after the effects of CA, rote memory span, and working memory span were taken into account. This result is consistent with Grant et al.'s [1997] finding that phonological memory is significantly related to vocabulary ability for individuals with Williams syndrome. Robinson et al. [2003] found that verbal working memory accounted for the largest proportion of variance in grammatical ability for individuals with Williams syndrome, accounting

for an additional 10% of variance after CA, verbal rote memory, and phonological memory were controlled.

The correlation between verbal working memory and grammatical ability was significantly stronger for the Williams syndrome group than for the typically developing group, even after controlling for CA. This finding suggests that children and adolescents with Williams syndrome rely more heavily on verbal working memory to successfully comprehend complex grammatical constructions than do typically developing children.

Although for typically developing children much of the process of extracting meaning from context is effortless and therefore not limited by verbal working memory capacity, for individuals with Williams syndrome who have deficits in many of the domains needed for the processing and integration of nonlinguistic cues with linguistic cues, more time and effort are needed for meaning extraction, likely increasing the importance of verbal working memory for language acquisition.

Karmiloff-Smith et al. [2003] have raised the more general possibility that because the beginning state of the Williams syndrome brain is not the same as the beginning state of the typically developing brain, the processes by which language is acquired may not be the same for individuals with Williams syndrome and typically developing children, even if the path followed appears to be normal but delayed. The increased importance of verbal memory in language acquisition for individuals with Williams syndrome is consistent with this possibility.

Klein and Mervis [1999] compared the performance of a group of 9- and 10-year-olds with Williams syndrome to a CA- and MA-matched group of children with Down syndrome. Although the two groups performed equivalently on the verbal scales of the McCarthy Scales of Children's Abilities [McCarthy, 1972], the Williams syndrome group performed significantly better on the verbal memory measures. Consistent with this finding, 9 of the 13 children with Williams syndrome but only 4 of the 13 children with Down syndrome spoke in complete, grammatical sentences. These children were a subset of a larger sample of 9- and 10-year-olds; in that sample, 19 of 23 children with Williams syndrome but only 4 of 25 children with Down syndrome spoke in complete, grammatically correct sentences [Mervis, 2006].

Two studies addressing the relation between the verbal memory and morphological abilities of Hungarian-speaking individuals with Williams syndrome have been conducted. Pléh et al. [2002] divided

a sample of 15 Hungarian-speaking children and adolescents with Williams syndrome (mean CA = 13.2 years, range: 5.9–19.6 years) into two groups based on a median split of digit span. The longer-span group performed significantly better than the shorter-span group on both regular plural and regular accusative nouns (97% versus 77%) and irregular plural and irregular accusative nouns (90% versus 61%). The memory span effect remained after controlling for CA. Digit span also was significantly negatively correlated with number of errors on both regular and irregular forms. Racsmany [2004; reported in Lukacs, 2005] correlated performance on regular and irregular plural and accusative noun forms with forward digit span for a group of 14 children and adolescents with Williams syndrome (mean CA = 14.1 years, range: 7.3–19.0 years) and a younger group of typically developing children matched for raw score on the Hungarian PPVT. Partial correlations (controlling for CA) were significant for the Williams syndrome group for both regular and irregular forms; for the typically developing group, partial correlations were significant only for the irregular forms; performance on the regular forms was at ceiling. Paralleling Robinson et al.'s [2003] findings, correlations between morphological ability and memory were stronger for the Williams syndrome group than the typically developing group.

CONCLUSION

For most individuals with Williams syndrome, language is a relative strength and visuospatial construction is a clear weakness. However, this pattern is not synonymous with either “intact” language or the independence of language from other aspects of cognition. It is rare to find an individual with Williams syndrome whose language ability is at the level expected for his or her CA. At the same time, it is rare for an individual with Williams syndrome to have severe intellectual disability. Instead, most individuals with Williams syndrome have mild intellectual disability, and their grammatical abilities are typically at the level expected for overall cognitive ability. Concrete vocabulary is usually at a somewhat higher level and relational/conceptual vocabulary is at a considerably lower level, similar to that of visuospatial construction ability. Communicative abilities are also considerably weaker than concrete vocabulary ability. Furthermore, language abilities and cognitive abilities are strongly related. For example, verbal working memory ability is considerably more important for grammatical comprehension for children and adoles-

cents with Williams syndrome than for typically developing children, and spatial memory limitations have been hypothesized to be important in difficulties in spatial/relational language. The relation between language (vocabulary or grammar) ability and visuospatial ability is mediated by nonverbal reasoning and verbal working memory. Early communicative abilities such as joint attention and gesturing are strongly related both to language abilities and to nonverbal reasoning and visuospatial construction abilities.

The finding that language ability is not at the level expected for CA has important implications for language intervention. Almost all children and adolescents with Williams syndrome would benefit from language therapy. Children who are either very young or who have significant intellectual disability would benefit from intensive language intervention focused on all aspects of language. For older children and adolescents who have intellectual ability in the mild disability to low-average range, language intervention targeting areas of specific weakness is important but all too often not provided.

The patterns of strengths and weaknesses, both between language and nonlinguistic cognition and within language, also have important implications for language intervention. First, the onset of referential communicative gestures is often used as an indicator that children are ready to acquire language; at this point, language or developmental therapy aimed at vocabulary acquisition may begin. Similarly, if children do not come to the attention of intervention agencies until after they are already talking, the presumption may be made that they have mastered the referential gesture system. Neither of these assumptions is appropriate for children with Williams syndrome. They are ready to begin therapy aimed at vocabulary acquisition well prior to the onset of referential gestures, and children with Williams syndrome who speak fluently almost always still have difficulty with pragmatic aspects of language. Thus, a full assessment of all aspects of language and communication is critical to determining the goals for language therapy for individual children with Williams syndrome. All too often, children with Williams syndrome do not receive language therapy once they are no longer making grammatical errors. These same children likely would benefit from therapy aimed at both relational/conceptual/figurative language and pragmatics. Unfortunately, there has been no research targeting either the efficacy of particular methods of language intervention (including, among other methods, either music therapy or

music within more traditional language therapy) for children and adolescents with Williams syndrome or more generally the impact of language intervention on their language and communicative abilities. Such research would provide crucial input for the design of an educational environment that will allow individuals with Williams syndrome the opportunity to reach their full potential. ■

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