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Cognitive and Behavioral Characteristics of Children with Williams Syndrome: Implications for Intervention Approaches

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Abstract

Portrayals of individuals with Williams syndrome (WS), a genetic disorder caused by a microdeletion of ~25 genes on chromosome 7q11.23, have reached the general public through a variety of media formats. These descriptions are often paradoxical in nature with individuals with WS repeatedly described as demonstrating near-normal language despite the presence of significant intellectual disability and as being extremely sociable and friendly in spite of their seemingly limited understanding of basic social norms. While this depiction of WS served to attract the interest of basic-science researchers, the results of subsequent studies have provided a more nuanced view. For example, rather than across-the-board "near-normal" language, children with WS demonstrate relative strengths in concrete vocabulary and verbal short-term memory, grammatical abilities at the level expected for general intellectual ability, and considerable weakness in relational/conceptual language and pragmatics (social use of language). To provide a more thorough characterization of the WS behavioral phenotype, we summarize recent findings related to intellectual ability, language development, memory development, executive function development, adaptive behavior skills, and behavior as it relates to learning by children with WS. Finally, we briefly discuss intervention approaches that may help children with WS to achieve their full potential.

Keywords

Williams syndrome; cognitive development; language development; behavior; intervention

INTRODUCTION

Williams syndrome (WS) is a complex neurodevelopmental disorder caused by a deletion of ~25 genes on one copy of chromosome 7q11.23 [Hillier et al., 2003; Osborne, 2006] with an estimated prevalence rate of 1 in 7500 live births [Strømme et al., 2002]. WS is associated with specific physical and medical characteristics including a characteristic facial appearance, congenital heart disease (especially supravalvar aortic stenosis), connective tissue abnormalities such as hernias or diverticuli of the bladder or colon, and failure to thrive or growth deficiency [Morris, 2006]. Infants and young children with WS have developmental delay and older children typically have intellectual or learning disabilities, although intellectual level ranges from average for the general population to severe intellectual disability. In addition, WS is associated with specific cognitive [Mervis et al., 2000] and personality [Klein-Tasman and Mervis, 2003] profiles.

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Portrayals of people with WS have reached the general public through a variety of media formats. Over the last two decades, a major source of information has been articlesin lay publications. The first article featuring WS in a lay magazine was published in *Discover* [Finn, 1991]. In this article, Finn stated, "People with Williams syndrome are smart and mentally retarded, gifted and inept at the same time [p. 55]." He went on to say that "People with Williams syndrome can show lots of intelligence in certain areas – language, music, and interpersonal relations, for example – and yet their IQ is typically between 50 and 70, low enough to qualify them as moderately to mildly retarded [p. 56]."

In 1994, Blakeslee, a contributor to the *New York Times*, wrote that in the case of WS, "There are severe malformations throughout the brain and heart, yet the capacity for language is remarkably unaffected. If anything, language and sociability are enriched [p. 1 of online article]." Fourteen years later, an article published in the *New York Times Magazine* offered a very similar description. Dobbs [2007] wrote that "Many with Williams have so vague a concept of space, for instance, that even as adults they will fail at six-piece jigsaw puzzles, easily get lost, draw like a preschooler and struggle to replicate a simple T or X shape built with a half-dozen building blocks. … The low I.Q., however, ignores two traits that define Williams more distinctly than do its deficits: an exuberant gregariousness and near-normal language skills. Williams people talk a lot, and they talk with pretty much anyone [p. 1 of online article]."

Increased public awareness of autism also led to increased interest in WS. In an article in *Newsweek* presenting a new theory on autism, Cowley [2003] included the following description of WS highlighting the seemingly opposite social phenotype to that associated with autism: "As fate would have it, some of the best natural readers of feelings and faces are themselves profoundly disabled. People with a rare genetic disorder called Williams syndrome are often severely retarded. Yet they're hypersocial, highly verbal and often deeply empathic [p. 50]."

A comparison of these depictions yields a recurrent theme: a fascination with how an individual could have significant intellectual disability but still have near-normal language, and could be extremely sociable and friendly yet have seemingly little understanding of basic social norms. It is this very depiction of WS that attracted the attention of basic-science researchers and encouraged them to attempt to characterize the WS behavioral phenotype systematically. In 1988, Bellugi and colleagues argued that despite demonstrating severe intellectual disability and functioning in Piaget's preoperational period, the adolescents with WS they studied nevertheless had excellent language abilities. In particular, Bellugi et al. reported that these adolescents were unable to conserve either number or quantity yet could comprehend and produce complex linguistic constructions (e.g., reversible passives, conditionals, and tag questions). They further argued that given this pattern of language strengths and cognitive limitations, WS provided strong evidence of the independence of language from cognition. This characterization of WS quickly attracted the attention of researchers interested in the relation between language and cognition and launched WS to the forefront of the debate on the modularity of language. As additional researchers began to study children with WS, experts concerned with modularity began to write about the syndrome, taking a considerably more strident position than did Bellugi and colleagues. Piattelli-Palmarini [2001] offered a particularly provocative statement: "For instance, children with WS have barely measurable intelligence and require constant parental care, yet they have an exquisite mastery of syntax and vocabulary."

Over the last two decades researchers have not only empirically evaluated these characterizations of WS but also have looked beyond these depictions, providing a more nuanced view of the overall behavioral phenotype associated with WS. Our aim in this

manuscript is to provide a summary of recent findings related to intellectual ability, language development, memory development, executive function development, adaptive behavior skills, and behavior as it relates to learning in children with WS, with the goal of providing a more thorough characterization of the WS behavioral phenotype. In addition, we briefly discuss intervention approaches that may help children with WS to achieve their full potential.

OVERALL INTELLECTUAL ABILITY

An important purpose of intellectual ability assessment is to determine IQ. This purpose is well known throughout the medical, educational, and lay communities. An equally important, but much less often considered, goal is to identify an individual's relative strengths and weaknesses as a function of type of intellectual ability. For example, as we describe below, WS is associated with relative strengths in (concrete) language, (concrete) nonverbal reasoning, and verbal short-term memory and severe weakness in visuospatial construction. WS also is associated with a range of intellectual ability, from the rare individual in the average range for the general population through the equally rare individual in the severe intellectual disability range. Most individuals have overall IQs in the borderline to moderate intellectual disability range. An assessment that is able to detect the pattern of cognitive strengths and weaknesses associated with WS across the full range of levels of intelligence associated with the syndrome would be particularly useful.

There is no mandatory set of abilities or higher-level organization of these abilities required for tests of intellectual ability. Similarly, there is no mandatory range of intellectual ability levels that must be covered. Thus, different tests measure somewhat different abilities and group these abilities in different manners. Furthermore, the subtests are normed to three standard deviations below the mean for some assessments and to four standard deviations below the mean for others. The manner in which subtests are grouped and the number of standard deviations below the mean for which subtests are normed determine how likely a particular assessment is to identify the pattern of strengths and weaknesses associated with a particular syndrome.

The most commonly used assessments of intellectual ability for individuals with intellectual disability are the Wechsler tests (e.g., Wechsler Intelligence Scale for Children [e.g., Wechsler, 2003], Wechsler Adult Intelligence Scale [e.g., Wechsler, 1981]). These assessments are normed to three standard deviations below the mean and group subtests measuring nonverbal reasoning ability and subtests measuring spatial ability together in a single composite (Performance or Perceptual Reasoning). Thus, although on average Wechsler Verbal composite standard score is ~5 points higher than Performance composite standard score for individuals with WS [Howlin et al., 1998; Searcy et al., 2004], Searcy et al. found that only 24% of individuals with WS scored significantly higher on the Verbal composite than on the Performance composite (1% scored significantly higher on the Performance composite than on the Verbal composite). Thus, for most individuals with WS, pattern of performance on the Wechsler composites does not mirror the cognitive profile associated with the syndrome. Furthermore, on some of the subtests many individuals earned the lowest possible standard score, indicating that the test is not normed low enough to accurately capture the ability levels of many individuals with WS.

The Differential Ability Scales assessment [DAS; Elliott, 1990 and DAS-II; Elliott, 2007] was designed to identify individuals' patterns of strengths and weaknesses. This measure provides separate composite (cluster) standard scores for Verbal, Nonverbal Reasoning, and Spatial abilities. Verbal short-term memory is assessed by a supplemental subtest that is not included in any of the clusters. The DAS-II subtests are normed to four standard deviations

below the mean. Examination of the pattern of mean standard scores (Table I) indicates that at the group level, the DAS-II accurately captures the pattern of strengths and weaknesses previously reported for individuals with WS, with performance on the Verbal, Nonverbal Reasoning, and verbal short-term memory measures at about the same level and performance ~20 points lower on the Spatial measure. The pattern of significant weakness in spatial abilities is also captured at the individual level; 86% of children performed significantly better on either the Verbal or Nonverbal Reasoning cluster than on the Spatial cluster. Two children (2%) scored significantly higher on the Spatial cluster than on the Verbal cluster. Examination of the standard deviations indicates that the DAS-II is normed low enough to capture the abilities of even low functioning children with WS.

The Mullen Scales of Early Learning [Mullen, 1995] may be used to assess the intellectual abilities of very young children with WS. This measure also includes separate measures of nonverbal reasoning (referred to as Visual Reception) and spatial ability (referred to as Fine Motor). As indicated in Table I, the same pattern of relative strengths in nonverbal reasoning and verbal abilities and severe weakness in visuospatial construction abilities is apparent even for 2-year-olds with WS. Unfortunately, the Mullen is only normed to three standard deviations below the mean, so this assessment does not accurately capture the abilities of lower functioning children with WS.

In collaboration with Karen Berman's research group at the National Institute of Mental Health, our research group has conducted neuroimaging studies comparing adults with WS to groups of individuals in the general population matched for gender, age, and IQ. We have identified an area of reduced gray matter and sulcal depth in the intraparietal sulcus [Kippenhan et al., 2005; Meyer-Lindenberg et al., 2004, 2006]. Results of functional neuroimaging (fMRI) studies indicated that this area served as a roadblock to dorsal stream information flow in a two-dimensional analogue to the DAS Pattern Construction subtest (the hallmark visuospatial construction weakness in WS). The convergence of behavioral and neuroimaging results strongly suggests that one or more genes in the WS region, in transaction with other genes and the environment, contributes to the development of visuospatial construction skills.

LANGUAGE ABILITIES

For the past 20 years, the modal topic for behavioral research studies of individuals with WS has been language development. This pattern reflects the fact that behavioral researchers initially were drawn to the study of WS to address questions of modularity, in particular, whether language ability was independent of cognitive ability. Until recently, most studies of language ability focused on language content (vocabulary) and structure (grammar). Although studies of these aspects of language development have continued, as the question of similarities and contrasts between WS and autism or autism spectrum disorders (ASDs) has become a major focus, studies of the socio-communicative aspects of language ability (pragmatics) have become much more common. Below we briefly review results of studies focused on early language development followed by findings from studies of vocabulary, grammar, literacy, and pragmatic development.

Early Language Development

The onset of language acquisition by children with WS is almost always delayed. Masataka [2001] has argued that this delay is due to specific motor delays. In particular, he has argued that rhythmic hand banging provides the motor substrate for canonical babble and that without canonical babble, word production is for the most part impossible. In a longitudinal study of 8 children with WS, Masataka [2001] found that the onset of rhythmic hand banging was considerably delayed relative to expectations for TD children. Nevertheless, the

pattern of correlations among abilities shown by children with WS was consistent with that for TD children, with the onset of rhythmic hand banging strongly correlated with the onset of canonical babble, which in turn was strongly correlated with the attainment of a 25-word expressive vocabulary. Mervis and Bertrand [1997] reported similar findings for the 2 children in their longitudinal study who were not producing canonical babble at the start of the study. Velleman et al. [2006; see also Mervis and Becerra, 2007] analyzed the phonological repertoires of 6 18-month-olds with WS and found that their babble was considerably delayed relative to that of the age-matched TD comparison group. Consistent with Masataka's [2001] argument that the production of canonical babble is critical for word production, Velleman et al. found that the children whose language was the most advanced had the most normal babble histories and that the child whose language was the most delayed had not met the criterion for canonical babble even at age 36 months.

Although speech perception skills are critical for spoken language development, only two studies of these abilities in young children with WS have been reported. Nazzi et al. [2003], in a study of 17 children with WS aged 17 – 47 months (mean age 33 months), found that the children were able to segment words with a strong-weak stress pattern (the predominant pattern in English) from ongoing speech but could not reliably segment words with a weak-strong stress pattern. Nazzi et al. argued that this combination of results suggested that young children with WS were using prosodic cues (which are adequate to identify words with a strong-weak stress pattern) rather than distributional information (which is needed to identify words with a weak-strong stress pattern) to identify words in ongoing speech. Cashon et al. [2009] studied the speech segmentation skills of 10 9 – 20-month-olds (mean age 14 months) with WS using an artificial language in which all syllables were equally stressed, so that "words" could only be identified based on distributional properties. Results indicated that the children were able to use distributional properties to segment words from continuous speech in the absence of prosodic cues. In both studies, the youngest children with WS were older than the ages at which these abilities are shown by TD infants, so data are not available to address the question of whether speech perception development is delayed for children with WS.

As expected given delays in the onset of canonical babble, the onset of word production also is delayed. Mervis et al. [2003] followed 13 children with WS longitudinally and found that age at acquisition of a 10-word expressive vocabulary was below the 5th centile (the lowest centile included in the norms) for the MacArthur-Bates Communicative Development Inventory (CDI) [Fenson et al., 1993] for all of the children. Age at acquisition of 50-and 100-word expressive vocabularies was also below the 5th centile for 12 of the 13 children. Despite these delays, the underlying categories for the early object labels comprehended and produced by children with WS were similar to those for both TD children and children with Down syndrome (DS), indicating that in contrast to prior claims that for individuals with WS, language was independent of cognition [e.g., Bellugi et al., 1988], the early cognitive development and early language development of children with WS in fact were closely linked [Mervis and Bertrand, 1997; Mervis and Becerra, 2007]. For example, similarly to TD children and children with DS, the extension of the early object labels of children with WS corresponded to their play patterns with the objects (e.g., the children rolled a wide variety of approximately spherical objects whether or not they were balls; they also comprehended and produced the word "ball" in relation to the same set of objects)[Mervis and Bertrand, 1997; Mervis and Becerra, 2007].

The expressive vocabularies of young children with WS and young children with DS have been compared in several studies. Results indicated that the mean expressive vocabulary size of young children with WS was significantly larger than that of children with DS when the children were matched for chronological age (CA) [Mervis and Robinson, 2000] but when

the two groups were matched for developmental level, the mean expressive vocabulary size of the DS group was almost the same as that of the WS group[Vicari et al., 2002]. At the same time, the grammatical abilities and verbal memory abilities of the WS group were considerably more advanced than those of the matched DS group [Vicari et al., 2002; see also Volterra et al., 2003].

Vocabulary Development

Receptive concrete vocabulary (comprehension of labels for objects, actions, and descriptors) has consistently been identified as an area of relative strength for individuals with WS. As indicated in Table I, the highest mean standard score for individuals with WS is on the Peabody Picture Vocabulary Test (in this case, the 4th edition) [PPVT-4; Dunn and Dunn, 2007]. Furthermore, 83% earned a standard score of at least 70 (the bottom of the "normal" range) and 8% earned a standard score of at least 100 (the 50th percentile for the general population). Similar results have been reported for previous editions of the PPVT as well [e.g., Bellugi et al., 1988;Brock et al., 2007,Mervis and Becerra, 2007]. The finding that receptive concrete vocabulary is a relative strength is not unique to WS, however. Glenn and Cunningham [2005] reported that pattern for individuals with DS and Facon et al. [1993] reported similar findings from a meta-analysis, for studies in which mean IQ was <70. Participants in these studies had a wide range of etiologies.

Expressive concrete vocabulary as measured by the Expressive Vocabulary Test-2nd edition [EVT-2; Williams, 2007] also is a relative strength for individuals with WS. As indicated in Table I, mean EVT-2 standard score was 2 points lower than mean PPVT-4 standard score, and similar to the findings for the PPVT-4, 83% of individuals with WS earned a standard score of at least 70 on the EVT-2 and 6% earned a standard score of at least 100.

In sharp contrast to their relative strength in concrete vocabulary, individuals with WS have a great deal of difficulty with relational/conceptual vocabulary. Basic relational vocabulary includes terms for spatial, temporal, quantitative, and dimensional concepts; more advanced relational vocabulary includes conjunctions (e.g., and, or) and disjunctions (e.g., although, however, nevertheless, neither … nor); all of these concepts are very difficult for individuals with WS. A comparison of the performance of 5 – 7-year-olds with WS on the PPVT-III [Dunn and Dunn, 1997] and the Test of Relational Concepts [TRC; Edmonston and Litchfield Thane, 1988] indicated that mean standard score on the concrete vocabulary measure was ~30 points higher than on the relational vocabulary measure [Mervis and John, 2008]. In fact, the children's performance on the TRC was similar to their performance on the DAS Pattern Construction subtest [Elliott, 1990], the signature weakness of individuals with WS. The pattern of errors indicated that children with WS had difficulty with all types of relational concepts, not just with spatial concepts. This finding is consistent with Walsh's [2003] argument that spatial, temporal, and quantitative processing are all controlled by a common magnitude system that is located in the inferior parietal cortex—the area in which Meyer-Lindenberg et al. [2004, 2006] identified a region of reduced gray matter that served as a roadblock to dorsal stream information flow, suggesting a possible common basis for the findings of extreme difficulty in both visuospatial construction and relational language for individuals with WS.

Although most individuals with WS eventually acquire basic relational concepts, they continue to have difficulty with many of the more advanced relational concepts. For example, 12 of 29 9 – 11-year-olds with WS tested on the Formulated Sentences subtest of the Clinical Evaluation of Language Fundamentals-IV [CELF-IV; Semel et al., 2003], a measure that includes both simple and advanced relational concepts, earned the lowest possible scaled score [Mervis and John, 2008]. A comparison of the performance of the children who participated in both the TRC study and, an average of 4 years later, the CELF-

IV Formulated Sentences study indicated very strong continuity in relational language ability over the age range of the two studies [Mervis and John, 2008].

Grammatical Development

Bellugi et al.'s [1998] initial report that the grammatical abilities of individuals with WS were well above those expected for their cognitive abilities was based on a comparison between CA-and IQ-matched adolescents with WS and DS. The finding that the grammatical abilities of individuals with WS are more advanced than those of matched individuals with DS has been replicated for both children whose native language is English [Joffe and Varlokosta, 2007a, 2007b; Mervis et al., 2003] and children whose native language is Italian [Vicari et al., 2002; Vicari et al., 2004]. However, these results most likely reflect the inordinate difficulty that individuals with DS have with grammatical development, rather than indicating that individuals with WS have better-than-expected grammatical abilities. In fact, when the group compared to WS is composed of either CA- and IQ-matched children with other etiologies of intellectual disability (ID) or MA-matched TD children, the grammatical abilities of the WS group are consistently at or below that of the contrast group across a variety of languages: English [e.g., Grant et al., 1997; Mervis and Becerra, 2007; Perovic and Wexler, 2007; Udwin and Yule, 1990; Zukowski, 2004], Hungarian [Lukács, 2005], and Italian [Volterra et al., 1996, 2003]. For a discussion of potential difficulties in interpreting results of studies in which individuals with ID are compared to much-younger TD children a—problem for a subset of the studies just referenced—see Mervis and Klein-Tasman [2004] and Mervis and Robinson [2005].

The most common standardized assessment of receptive grammatical ability used in studies of individuals with WS is the Test for Reception of Grammar [TROG; Bishop, 1989 or TROG-2; Bishop, 2003]; this measure has been translated into several languages. The TROG measures a variety of sentence structures, ranging from simple subject-verb constructions to center-embedded relative clauses. As indicated in Table I, mean level of performance for children and adolescents with WS is in the borderline range; 28% earned the lowest possible standard score.Karmil off-Smith et al. [1997] reported similar findings for a smaller sample. The results of studies using the Italian and Hungarian versions of the TROG have indicated that the order of difficulty of grammatical constructions for individuals with WS is highly similar to that for children in the general population acquiring the same native language [for Hungarian: Lukács, 2005; for Italian: Volterra et al., 1996]. Receptive grammatical ability as measured by the TROG is strongly related to verbal working memory ability (as measured by digits backward recall ability) for individuals with WS. Furthermore, this relation is stronger for individuals with WS than for TD children with similar levels of receptive grammar ability, suggesting that individuals with WS may rely more heavily on verbal working memory when parsing complex grammatical constructions than do TD children [Robinson et al., 2003].

Literacy

Three of the first five behavioral research papers published on children with WS included information about reading abilities. The results of these studies indicated a wide range of reading abilities, with a small proportion of children decoding (reading single words) and comprehending at grade level and a small proportion not able to read at all [MacDonald and Roy, 1988; Pagon et al., 1987; Udwin and Yule, 1987]. Udwin and Yule reported that the median reading level for a group of individuals with WS aged 10 – 20 years was 2nd grade. Presaging later findings, MacDonald and Roy noted that children who were being taught to read with phonics seemed to benefit more than children being taught to read with the "look-say" (sight-word or whole-word) method.

For TD children, phonological awareness is strongly related both to decoding and to reading pseudowords based on the phonics rules of the relevant language [see reviews in Ehri, 2004 and McCardle et al., 2008]. The relation of phonological awareness to decoding has been addressed in several studies involving individuals with WS. Levy et al. [2003], studying individuals learning to read English, found that elision (the ability to delete specified syllables or sounds from a word) was strongly related to both decoding and pseudoword reading. This same finding has been obtained for individuals learning to read Italian [Menghini et al., 2004] and Hebrew [Levy and Antebi, 2004]. Based on their findings, Levy and colleagues recommended that children with WS be taught to read using phonics [Levy and Antebi, 2004; Levy et al., 2003].

Becerra et al. [2008; see also Mervis, 2009] analyzed the reading performance of 44 9 – 17-year-olds with WS. Initial analyses considered the sample as a single group to provide information about the reading abilities of individuals with WS relative to general-population norms for children of the same CA. Mean standard scores on the Reading section of the Wechsler Individual Achievement Test-II [WIAT-II; Wechsler, 2005] were 73.00 [range: 40 (floor) – 112] for Word Reading (decoding), 78.75 [range: 0 correct – 113 (standard score)] for Pseudoword Decoding, and 64.61 [range: 40 (floor) – 102] for Reading Comprehension. Mean standard score for Reading Comprehension was significantly lower than for either Word Reading or Pseudoword Decoding. For all three subtests, the standard deviations were >15, indicating more variability than in the general population. All children could read at least a few words, but 8 (18%) could not read any pseudowords. Several participants, including one in 11th grade, read and comprehended at grade level.

In a second set of analyses, the participants were divided according to their primary method of reading instruction: phonics (n = 24) or whole word/sight word/whole language (n = 20). The children's reading standard scores were then compared to those predicted based on their DAS-II GCA, using the tables in the DAS-II manual [Elliott, 2007]. Results indicated large and significant differences as a function of reading method for all three reading subtests. Most children in the Phonics group read at or above the level expected for their GCA. In strong contrast, most children in the Whole Word group read below the level expected for their GCA. These findings are consistent with those of the meta-analyses conducted by the National Reading Panel [see summaries in Ehri, 2004 and McCardle et al., 2008], which stressed the importance of early, explicit, and systematic instruction in phonemic awareness and phonics for all children.

Pragmatics

The combination of a relative strength in the structural and concrete content aspects of language and increased sociability paired with consistent problems in making friends and sustaining friendships led researchers to hypothesize that children with WS likely have difficulty with the pragmatic aspects of language. The results of the studies examining the pragmatic abilities of individuals with are consistent with this position, documenting pragmatic difficulties across developmental stages. The emergence of joint attention in children with WS is delayed relative to both CA and language ability [Mervis et al., 2003; Mervis and Bertrand 1993; 1997]. Furthermore, although both TD children and children with DS begin to comprehend and produce pointing gestures prior to the onset of referential expressive language, children with WS do not comprehend and produce pointing gestures until well after the onset of referential word production [Mervis et al., 2003; Mervis and Becerra, 2007]. Children with WS are significantly less likely to engage in joint attention and to comprehend and produce gestures than are either mental-age matched TD children or children with DS individually matched on CA, developmental quotient, and expressive vocabulary size [e.g., Laing et al., 2002; Rowe et al., 2005]. John and Mervis [in press-a] examined the ability of preschoolers with WS and CA-matched preschoolers with DS to

comprehend communicative intent expressed by pointing gestures and eye gaze. Despite having significantly lower mean developmental quotient scores, the children with DS were significantly better at inferring communicative intent than were the children with WS; 60% of the children with DS but only 27% of the children with WS found the hidden toy at a rate significantly above that expected by chance.

These findings that the pragmatic abilities of even young children with WS are more limited than expected for their developmental level provided the initial basis for questioning the accuracy of the early characterizations of WS as the "opposite" of autism. The overlap between the phenotypes associated with WS and ASDs have been addressed in three studies of the performance of toddlers and preschoolers [Klein-Tasman et al., 2007; Klein-Tasman et al., 2009; Lincoln et al., 2007] on the Autism Diagnostic Observation Schedule-Generic [ADOS-G; Lord et al., 1999], a semi-structured play based interaction designed to press for behaviors central to a diagnosis of ASDs. Participants in these studies had very limited to no expressive language.

Results of these studies indicated that a large portion of the participants with WS demonstrated behaviors typically thought to be characteristic of children with ASDs. For example, approximately half of the participants reported by Klein-Tasman et al. [2007, 2009] and Lincoln et al. [2007] did not clearly integrate eye contact with their communicative partner in order to reference a desired object that was out of reach. Almost three quarters of the children did not integrate eye contact or vocalization with acts of showing objects, and nearly all of the children with WS in these studies did not spontaneously use a doll or other object as an independent agent or use objects to represent other objects. These difficulties were such that many of the children in these studies were classified on the ADOS algorithm as "autism-spectrum disorder" (38% in the Klein-Tasman et al. studies, and 5% in Lincoln et al.) and some children were classified as "autism" (10% in the Klein-Tasman et al. studies and 5% in Lincoln et al.). However, differences between the behavioral phenotypes associated with WS and ASDs also were found. Few children with WS evidenced difficulty directing vocalizations or facial expressions to other people or sharing affect, and the quality of social overtures was generally good [Klein-Tasman et al., 2007; Klein-Tasman et al., 2009; Lincoln et al., 2007].

Studies examining pragmatic abilities in older individuals with WS have shown that these difficulties continue into the school-age and adult years and are of considerable concern to parents. These studies have focused on parental responses to questionnaires addressing communicative competence, analyses of children's conversations with a researcher, and children's responses in a task measuring comprehension monitoring skills. In addition, researchers have examined children's theory of mind (ToM) ability, as ToM is considered to play a key role in pragmatics.

Four studies have addressed the general pragmatic abilities of individuals with WS using a version of the Children's Communication Checklist [CCC; Bishop, 1998; or CCC-2; Bishop, 2002], a parent-report measure. Laws and Bishop [2004] studied 19 individuals with WS (mean CA = 14.83 years) and found that 15 (79%) met the CCC cut-off for pragmatic language impairment. The WS group evidenced significant difficulties in all areas of pragmatics measured by the CCC. Relative to a DS group (mean CA = 15.92 years) and a group of children with Specific Language Impairment (mean CA = 6.00 years), the WS group evidenced particular difficulty in the use of stereotyped conversations, inappropriate initiation of conversations, and overdependence on context to interpret what was said to them. These findings have been well replicated by other research groups [Harmon et al., 2009; Peregrine et al., 2005; Philofsky et al., 2007]. Many of these pragmatic problems are evident even when the comparison group is children with autism; Philofsky et al. [2007]

reported that although children with WS earned significantly better scaled scores than CA-matched children with autism on the CCC-2 Stereotyped Language and Nonverbal Communication scales, the two groups evidenced similar impairments on the Inappropriate Initiation and Use of Context scales.

Several studies directly examining the conversational abilities of children with WS have been conducted. In the first such study, Udwin and Yule [1990] reported the results of analyses of 30-minute conversations between a child with WS and a researcher. Of the 43 children who participated (mean CA = 11.1 years), 16 (37%) met the authors' criteria for hyperverbal speech (fluent speech including an excessive number of stereotyped phrases or idioms, over-familiarity, introduction of irrelevant personal experiences, and perseverative responding). More recently, Jones et al. [2000] found that while adolescents and adults with WS (CA = 15.8 years) answered the same number of questions within a biographical interview as did CA-and IQ-matched adolescents and adults with DS and MA-matched TD children, the WS group was significantly more likely to describe affective states, make evaluative comments, and use character speech and emphatic markers than was either comparison group. Stojanovik [2006] found that, within a semi-structured conversation, regardless of whether the researcher asked for information or clarification, the responses of the WS group (mean CA = 9.17years) were less likely to be adequate than were the responses of either children with specific language impairment (mean CA = 10.58) matched for receptive vocabulary and grammatical ability or slightly younger TD children (mean CA = 8.67). In particular, the WS group was more likely to provide too little information or to misinterpret what the researcher had meant and was considerably less likely to produce a response that continued the conversation.

A person's conversational success depends, in part, on the ability to monitor whether he or she understands what the speaker has said and to request clarification when needed. John et al. [2009] examined comprehension monitoring and verbalizations of message inadequacy by 57 children with WS (mean CA = 9.24 years) using a listener-role referential communication task modeled after Abbeduto et al. [2008] in which the child and an adult were separated from one another by a barrier. The child's task was to place the picture requested by the adult into a picture of a larger scene. Although children performed very well when they understood the instructions and the required picture was available, they had considerable difficulty when the researcher's instructions were inadequate (the requested picture was not one of the referents available, the researcher's instruction was ambiguous, or the researcher's instruction contained vocabulary that the child did not understand.) Children verbally indicated that there was a problem less than 50% of the time on average and most of their verbalizations were either too vague for the researcher to understand the nature of the problem or indicated the wrong problem. Performance was related to CA and first-order theory of mind.

Successful communication between two people involves not only a mastery of the language, but also taking into account basic information about the communicative partner (e.g., his or her status, knowledge, feelings, focus of attention) and using this information to help formulate an effective message. Thus, successful communication depends at least in part on the ability to understand another person's perspective (i.e., ToM). A deficit in ToM would likely contribute to pragmatic difficulties. Studies of the development of false belief (one of the first types of ToM demonstrated by TD children) have indicated that acquisition of this concept by children with WS is considerably delayed. Tager-Flusberg and her colleagues have compared the performance of children with WS aged 4 – 10 years on false belief tasks to the performance of CA-, IQ-, and language-matched children with Prader-Willi syndrome (PWS) or nonspecific ID [Joseph and Tager-Flusberg, 1999; Tager-Flusberg and Skwerer, 2007; Tager-Flusberg and Sullivan, 1994; Tager-Flusberg and Sullivan, 2000]. Findings

indicated that the WS group did not perform better than the contrast groups, and none of the groups performed well. John and Mervis [2009] used the Unexpected Contents task (a false-belief task routinely passed by TD 4-year-olds) to study the development of ToM in children with WS aged 6 – 14 years. Only the 13-and 14-year-olds reliably passed this task; of the younger children, only 3 of 28 6 – 10-year-olds and 3 of 6 11 – 12-year-olds were successful, providing further confirmation that acquisition of even basic ToM ability is greatly delayed.

MEMORY

The pattern of relative strengths and weaknesses for individuals with WS within the memory domain is consistent with the overall phenotypic pattern. In particular, individuals with WS evidence significantly better verbal memory than spatial memory [e.g., Jarrold et al., 1999; Wang and Bellugi, 1994]. This pattern also holds in comparisons with CA-and IQ-matched individuals with other forms of ID, if the memory tasks used do not involve mental manipulation (e.g., for "rote" or "short-term" memory tasks). In particular, individuals with WS perform significantly better on measures of forward digit recall than do CA-and IQ/MA-matched groups with DS [Edgin, 2003; Klein and Mervis, 1999; Jarrold et al., 1999; Wang and Bellugi, 1994] or ID of unknown or mixed etiology [Devenny et al., 2004; Udwin and Yule; 1991]. This same pattern is observed for the first trial of word list recall [Nichols et al., 2004]. In contrast, children and adults with WS perform significantly worse than CA-and IQ-/MA-matched individuals with DS on spatial memory tasks such as forward Corsi recall [Edgin, 2003; Jarrold et al., 1999, Wang and Bellugi, 1994]. No comparisons of spatial memory with contrast groups with ID of mixed or unknown etiology have been reported.

When verbal memory tasks require mental manipulation ("working memory"), however, differences between individuals with WS and those in the contrast group(s) are considerably reduced. On backward digit recall (verbal working memory) tasks, while groups of individuals with WS consistently demonstrate longer spans than CA-and IQ-/MA-matched groups with other forms of ID [e.g., Devenney et al., 2004; Edgin, 2003; Wang and Bellugi, 1994], these differences are not significant. In the only study that compared performance on backward Corsi recall (spatial working memory), mean span for the WS group and the DS group was almost identical [Edgin, 2003]. Finally, in contrast to initial characterizations of WS that stressed the independence of language and cognition, there is now mounting evidence that verbal memory abilities are strongly related to both grammatical and vocabulary abilities for individuals with WS [Grant et al., 1997; Mervis, 2006; Mervis and Becerra, 2007; Pléh et al., 2002; Robinson et al., 2003].

EXECUTIVE FUNCTION

Executive functioning is a blanket term referring to a set of higher-order cognitive processes associated with planning and regulatory control [e.g., Welsh and Pennington, 1988; Hughes and Graham, 2002]. These processes include working memory, inhibition, set shifting/cognitive flexibility, self-monitoring, and generativity. To date four studies of executive functioning abilities in individuals with WS have been published. Tager-Flusberg et al. [1997] compared the performance of children with PWS and children with WS aged 5 – 8 years on two executive functioning tasks, one examining the ability to verbally inhibit a prepotent response (Day-Night Stroop Task) and one examining the ability to motorically inhibit a prepotent response (Tapping Task). These tasks were quite difficult for both groups, with 56% of the PWS group and 25% of the WS group passing the verbal inhibition task and 22% of the PWS group and 17% of the WS group passing the motoric inhibition task. Atkinson et al. [2003] also tested individuals with WS aged 4 – 15 years on tasks measuring inhibition, comparing their performance to norms for TD children whose CA

matched the children with WS's vocabulary age on the British Picture Vocabulary Scale (BPVS; the British version of the PPVT). Performance was considerably better on the verbal inhibition task (Day–Night Stroop task), than on the two motor inhibition tasks (Detour Box and Pointing/Counter pointing). For many children with WS, performance on the verbal inhibition task was at or above the level expected for vocabulary age, while for most children with WS, performance on the spatial inhibition tasks was well below the level expected for vocabulary age. There was a significant correlation between performance on the Detour Box spatial inhibition task and performance on the verbal inhibition task. Rhodes et al. [in press] investigated executive functioning in teenagers and young adults with WS (mean CA = 18.08 years) relative to TD individuals matched for vocabulary age on the BPVS (mean CA = 9.25). Individuals with WS evidenced difficulty on the CANTAB [www.camcog.com] tasks of attention set-shifting (Intra-Dimensional/Extra-Dimensional), working memory (Spatial Working Memory), and planning (Stockings of Cambridge) relative to the contrast group. The validity of these comparisons depends on the assumption of similar rates of development for both the control (receptive vocabulary) and the target variable (executive functioning). No data are available to address this assumption for executive function, but Mervis and Klein-Tasman [2004] and Mervis and Robinson [2005] have shown that it does not hold for a variety of other control and target variables. If the assumption is not valid, then the prediction that two groups with identical raw scores on the BPVS but very different CAs should be expected to have similar scores on tasks of executive functioning is incorrect.

John and Mervis [in press] used parent-report measures to consider the relation between sensory modulation impairments and executive functioning. Cluster analysis identified two clusters of children with WS varying in terms of sensory symptom severity (mild abnormalities versus more severe abnormalities), with parent-reported executive functioning ability [based on the Behavior Rating Inventory of Executive Functioning (BRIEF); Gioia et al., 2000] accounting for the largest proportion of between-cluster differences (46%). Children in the severe sensory modulation impairment group were reported to have significantly more difficulty than children in the mild sensory modulation impairment group on transitioning between activities, appropriate modulation of emotional responses, initiating a task or activity, staying on task, using working memory, anticipating future events/setting goals, and monitoring their own behavior/performance. On all these activities, mean level of performance was in the clinical range for the severe sensory impairment group. Mean level of performance for the mild sensory impairment group (although significantly better than for the severe-sensory impairment group) also was in the abnormal range for the meta cognitive skills (working memory, planning, monitoring).

ADAPTIVE BEHAVIOR

An important aspect of development that brings together contributions from both cognition and personality is adaptive behavior. As defined by the American Association on Intellectual and Developmental Disabilities (AAIDD), adaptive behavior refers to "the conceptual, social, and practical skills that people have learned to be able to function in their everyday lives [AAIDD website, 2010]." In addition, adaptive ability can be considered to be the extent to which a person functions, maintains independence, and demonstrates the social responsibility expected of individuals in his or her age and cultural group [AAMR, 1992; Cicchetti et al., 1984]. As such, examination of adaptive behavior performance in individuals with WS provides insight into the impact of WS on real world functioning [Mervis et al., 2001].

Most studies of the adaptive behavior of children with WS have used the parent-interview form of the Vineland Adaptive Behavior Scales[VABS; Sparrow et al., 1984]. Gosch and

Pankau [1994] compared the performance of children with WS to a CA-and IQ-matched group with nonspecific ID and found that overall the nonspecific ID group performed significantly better than the WS group. The authors hypothesized that the difference was primarily due to the large number of items that required fine motor skills, an area of particular weakness for children with WS. Greer et al. [1997] examined the performance of children with WS aged 4 – 18 years and found that standard scores on the Socialization and Communication scales was significantly higher than standard scores on the Daily Living Skills or Motor Skills scales. Using a larger sample of participants with WS over a narrower age range (4 – 8 years), Mervis et al. [2001] found significant differences in standard scores between all pairs of scales. Performance was ordered as follows: 1. Socialization, 2. Communication, 3. Daily Living Skills, 4. Motor Skills. In this study, VABS composite standard score was not related to CA. However, Fisch [2010], in a study of 34 children with WS aged 4 – 15 years, found a significant negative correlation ($r = -.65$) between CA and VABS composite standard score, with the regression line suggesting a standard score decline of ~25 points between ages 4 and 15 years.

In Table I, we report adaptive behavior performance for 122 children with WS aged 4 – 17 years, using a different parent-interview measure, the Scales of Independent Behavior-Revised [Bruininks et al., 1996]. This measure includes four scales: Motor Skills, Social Interaction and Communication Skills, Personal Living Skills, and Community Living Skills [see also Mervis and Morris, 2007]. Performance on the Social Interaction and Communication Skills scale was significantly better than on the remaining scales [Mervis and Morris, 2007]. There was a significant negative correlation between CA and SIB-R overall standard score ($r = -.31$). For the subscales, there were significant negative correlations with CA for standard scores for Motor Skills and Community Living Skills but not for Social Interaction and Communication Skills or Personal Living Skills. Mean SIB-R composite standard score was lower than mean DAS-II GCA.

Mervis and Morris [2007] point out that adaptive behavior is perhaps the area where caregivers are most able to impact skill development. There are wide variations among parents in how much emphasis they put on acquisition of self-help skills and how much responsibility they expect their child to take for household chores. The importance parents ascribe to these types of activities is likely to have a considerable effect on their child's long-term outcome, as successful employment and (semi-) independent living are heavily dependent on adaptive skills. The adaptive skills of many children with WS are considerably lower than would be expected for IQ because they have resisted working on these skills (some of which are more difficult for them because of fine motor requirements) and family members have acceded to the child's wishes.

BEHAVIOR AS IT RELATES TO LEARNING

von Armin and Engel [1964] highlighted the paradoxical nature of the WS behavioral phenotype, describing individuals with WS as having "a great ability to establish interpersonal contacts (p. 376)" that "stands against a background of insecurity and anxiety (p. 376)." von Armin and Engel also noted that children with WS were "hypersensitive to feelings of frustration (p. 375)" and had frequent temper outbursts. In one case report, they stated that the child "would plague the other residents with interminable questions … evidently driven by an unexpected feeling of insecurity (p. 371)." These characteristics, as well as others more recently determined to be associated with WS (e.g., distractibility, impairments in sensory processing, difficulties with mastery motivation), are vital to examine as they have the potential to have a severe impact on the ability of children with WS to learn.

Results of more recent research studies of children with WS using parental report measures of personality provide further support for the pattern of characteristics described by von Armin and Engel [1964]. Lieshout et al. [1998], using a Dutch translation of the California Q-set [Block and Block, 1980], found that while children with WS (CA range: 2.75 – 19.5 years) obtained similar scores on Extraversion and Agreeableness in comparison to gender- and CA-matched TD children, their scores were significantly lower for Conscientiousness, Emotional Stability, and Openness and significantly higher for Irritability. Klein-Tasman and Mervis [2003] proposed an empirically supported WS Personality Profile based on the comparison of parental ratings of children with WS aged 8 – 10 years and parental ratings of CA-and IQ-matched children with ID of mixed etiologies on the parent report version of the short form of the Multidimensional Personality Questionnaire [MPQ; Tellegen, 1985]. The children with WS were rated as significantly more gregarious, people-oriented, visible, tense, and sensitive than the mixed etiology group. This combination of items correctly classified 21 of 22 children with WS and 17 of 20 children in the mixed etiology group.

Although the initial descriptions of WS did not emphasize distractibility as a characteristic of the syndrome, studies using standardized questionnaires have consistently identified difficulty concentrating as a major concern. Significantly elevated scores on the Attention Problems subscale of the Child Behavior Checklist [CBCL; Achenbach 1991] were reported for 67% of the children studied by Dilts et al. [1990] and 73% of the children studied by Greer et al. [1997]. Einfeld and Tonge [Einfeld et al., 1997; Tonge and Einfeld, 2003] reported that parents of children with WS were significantly more likely to endorse the items “overactive” and “short attention span” than were parents of the epidemiological control group for the Developmental Behavior Checklist [DBC; Einfeld and Tonge 1995]. Rhodes et al. [in press] reported that all of the children whose parents completed the Conners Rating Scale [Conners, 1997] scored in the clinical range on the ADHD Index.

Studies in which structured interviews designed to yield DSM-IV diagnoses were completed by parents of children with WS have yielded similar findings. Two studies have used the Anxiety Disorders Interview Schedule – Parent [ADIS-P; Silverman and Albano, 1996]. Kennedy et al. [2000] reported that 43% of a sample of 20 individuals with WS aged 7 – 28 years met DSM-IV criteria for ADHD, and Leyfer et al. [2006] reported that 65% of 119 children with WS aged 4 – 16 years met criteria for ADHD. In both studies, the most common ADHD diagnosis was ADHD-Predominantly Inattentive type. Dodd and Porter [2009] administered the Schedule for Affective Disorders and Schizophrenia for School-Age Children-Present and Lifetime Version [K-SADS-PL; Kaufman et al., 1997] to caregivers of 30 children with WS aged 6 – 17 years and found that 33% met criteria for ADHD, primarily for the Predominantly Inattentive type.

Other concerns and behavioral difficulties have also been associated with WS. Based on the ADIS-P interviews mentioned above, Leyfer et al. [2006] reported that 57% of children with WS met DSM-IV criteria for at least one anxiety disorder, with 54% meeting criteria for Specific Phobia, 12% for Generalized Anxiety Disorder, and 7% for Separation Anxiety. Kennedy et al. [2000], whose sample was somewhat older, found that 48% of participants met DSM-IV criteria for at least one anxiety diagnosis, with 43% meeting criteria for Specific Phobia and 24% for Generalized Anxiety Disorder. Leyfer et al. [2009] compared the prevalence of DSM-IV anxiety disorders in children with WS to those reported in an epidemiological study of children with ID[Dekker and Koot, 2003] and found that the rates for children with WS were significantly higher for Specific Phobia, Generalized Anxiety Disorder, and Separation Anxiety Disorder. Children with WS also demonstrate higher rates of fears [e.g., Dykens, 2003] and of sensitivity and tenseness [Klein-Tasman and Mervis, 2003] than CA-and IQ-matched samples of individuals with other forms of ID. Highlighting another concern raised by von Engel and Armin, some individuals with WS have been

reported to be "easily angered and reactive … over 'trivial' things (p. 119)" [Phillips and Klein-Tasman, 2009]. In line with this concern, Rhodes et al. [in press] reported that 55% of the children in their sample scored in the clinical range on the Oppositional Scale on the Conners' and 54% scored in the abnormal range (with another 9% in the borderline range on the Conduct Problems scale of the Strengths and Difficulties Questionnaire (SDQ) [Goodman, 2001].

All of the types of problems discussed in this section are commonly reported for children who have sensory integration problems [e.g., Ayres and Robbins 2005]. John and Mervis [in press-b] administered the Short Sensory Profile [SSP; McIntosh et al., 1999] to parents of 72 4-to 10-year-olds with WS to determine if children with WS demonstrate difficulties with sensory integration. Only 10% of the children were classified as having normal sensory processing overall; most children were classified as having definite abnormalities. Based on parental responses to the SSP, John and Mervis [in press-b] identified two clusters of children with WS varying in terms of sensory symptom severity. Children in the severe sensory impairment cluster demonstrated significantly poorer executive functioning, more negativity, less effortful control, and more attention-related complications and anxiety than did children in the mild sensory impairment cluster.

Individuals with WS also have been described to demonstrate an increased interest in people, a characteristic that is particularly apparent when these children encounter difficult situations that they would prefer to avoid. Mervis and colleagues [2003] were first to empirically demonstrate that infants and toddlers with WS intensely stared into the faces of strangers in situations where other children with neurodevelopmental disabilities as well as TD children would look at an object or coordinate attention between a person and an object. Järvinen-Pasley et al. [2008] reported that, when attempting to administer a task designed to elicit emotional reactions of frustration and anger, they were unable to collect data on many of the children with WS as these children focused only on the experimenter as opposed to attending to the toy behind a plastic barrier. This finding is consistent with our own observations and experiences testing children with WS. During the assessment process, the majority of children with WS attempt to distract the examiner or engage the examiner socially once a task becomes difficult, as a strategy to avoid having to attempt activities they find challenging. Therapists who work with children with WS have often reported the same types of problems.

One of the most common concerns expressed by parents of children with WS is that their child's level of motivation to attempt to complete a somewhat difficult task is extremely low. The results of the one empirical study that addressed this issue indicate that these concerns are well founded. Rowe [2007] compared mastery motivation, or the willingness to persevere on a moderately difficult task and the expression of pleasure upon mastery the task, in preschoolers with WS to that of CA-matched preschoolers with DS. Despite more limited cognitive and adaptive abilities, the DS group exhibited significantly more persistent task-related behavior and mastery pleasure on moderately challenging tasks than did the WS group. In contrast, the WS group demonstrated significantly more help-seeking behavior and non-goal oriented apparatus-directed behavior than did the DS group. This pattern of results indicates a considerably limited mastery motivation in preschoolers with WS, which is especially striking considering their significantly higher cognitive and adaptive skills when compared to children with DS. Rowe hypothesized that the limited mastery motivation evidenced by children with WS may be related to the low levels of independent living and employment success observed in adults with WS.

INTERVENTION

Over the past two decades, considerable progress has been made toward refining the WS behavioral phenotype. Although much more work needs to be done to fully understand this complex phenotype, our current level of knowledge provides a reasonable foundation for the start of intervention research. It is vital that efforts be dedicated toward the development and evaluation of practical methods of intervention. In addition to helping the children and their families, this line of research may also help to refine researchers' understanding of how the WS behavioral phenotype comes to develop over time. Research dedicated to the development and evaluation of interventions appropriate for individuals with WS has barely begun. However, intervention approaches developed for other developmental disorders that share characteristics with WS (for example, ASDs, children with learning disabilities) are likely also to be appropriate for individuals with WS and may serve as a foundation for the development of intervention approaches and studies specific to WS. In this section, we briefly outline recommendations for applied behavior analysis approaches to address both learning and behaviors that interfere with learning, social skills training, language intervention, and reading intervention. Although not addressed in this paper, other types of therapies (most notably occupational therapy and cognitive-behavioral approaches to address anxiety) also are important to help children with WS reach their potential.

Applied Behavior Analysis

As maybe seen from the literature reviewed above, there are several aspects of the WS behavioral phenotype which can affect overall functioning in adulthood including delays in communication, delays in adaptive functioning, difficulties with peer relations and interacting with others, as well as higher rates of anxiety and difficulties transitioning from one activity to the next. Unfortunately, research addressing the impact of intervention on outcome in children with WS has been extremely limited. Klein-Tasman and colleagues [2009] have argued that given the fact that most current treatment plans are based on phenotype rather than etiology, many children with WS would benefit from treatment programs initially designed for other neurodevelopmental disorders such as ASDs.

One of the most widely known types of intervention recommended for children with ASDs is applied behavior analysis (ABA), a scientific approach devoted to understanding a behavior and how it is affected by the environment. ABA uses behavioral principles including classical and operant conditioning to treat undesirable behaviors. This technique was originally established in the 1950s as a method of evaluating and changing human behavior using the principles of operant conditioning. ABA itself was not designed specifically to help children with ASDs or other neurodevelopmental disorders, but rather provides a framework upon which multiple approaches are based. According to the basic principles of ABA, there is some stimulus (i.e., antecedent) that serves as a cue for the child to respond. Once the child responds he or she receives a consequence or feedback regarding his or her behavior. Accordingly, challenging behavior is viewed as a functional communicative act reinforced by the child's environment and a systematic assessment is conducted to determine what feedback/reinforcement is encouraging or maintaining the behavior. Once this relation is determined, the ABA therapist focuses on teaching the child socially acceptable means of achieving the same function.

For many people, ABA is equivalent to Lovaas' discrete trial intervention approach [Lovaas, 1987], and their immediate response is to reject the possibility of using ABA. In reality, Lovaas' method is only one of several ABA intervention programs. Most programs used today are based on ABA principles combined with a strong emphasis on the use of naturalistic environments and tools to promote success and generalization. These programs are grounded in up-to-date understanding of child development and include empowering

children to become active participants in the world around them as a goal. This type of ABA approach will likely also aid children with WS. Examples of this type of ABA-based intervention approach include DIR/Floor time [e.g., Wieder and Greenspan, 2005], Relationship Development Intervention [RDI; Gutstein and Sheely, 2002], SCERTS [Prizant et al., 2006], Pivotal Response Training [PRT; Koegel et al., 1987; Schreibman and Koegel 2005], and milieu teaching [e.g., Warren and Yoder, 2003; Yoder and Warren, 2001].

Recently Dawson, Rogers, and colleagues [2009] conducted the first randomized, controlled trial of intervention for toddlers with ASDs evaluating the efficacy of the Early Start Denver Model (ESDM). ESDM is a comprehensive developmental behavioral intervention based on the original Denver Model therapy approach [Rogers et al., 1986], models of autism proposed by Rogers and Pennington [1991] and Dawson and colleagues [2004], and PRT, a teaching approach based in ABA [e.g., Koegel et al., 1987; Schreibman and Koegel,2005]. The aim of ESDM is to reduce the severity of autism symptoms and accelerate children's developmental rate in all domains, particularly within the cognitive, social-emotional, and language domains. ESDM focuses on building language development within a social context and building up complex behaviors and eliminating or ameliorating problem behaviors that interfere with learning. ESDM uses an interdisciplinary approach and systematically individualizes the program based on the child's individual learning needs, preferences, and interests. Family values and needs are incorporated into the child's objectives and parent's training. Results of the 2-year trial indicated that the children in the ESDM group demonstrated significantly greater progress as measured, for example, by Mullen DQ (17 point vs. 7 point increase), and VABS adaptive behavior composite (<1 point decline vs. 10-point decline), than did the children who received community intervention. These findings are particularly impressive as the community in which the intervention took place is known for providing exemplary intervention for children with ASDs.

As described above, there is considerable overlap between the behavioral phenotypes associated with ASDs and WS, and a comprehensive therapeutic approach similar to ESDM would both be appropriate for children with WS and likely effective for improving outcomes. Carefully conceived and implemented intervention studies, similar to the one conducted by Dawson et al. [2009] are important to fully evaluate the efficacy of early behavioral intervention for children with WS.

Social Skills Training

While individuals with WS have an interest in interacting with other people, their deficits in social skills and limited social competence make it very difficult for them to navigate interactions with others. Given the significant difficulties in establishing and maintaining relationships experienced by individuals with WS, it is very important that attempts be made to improve their social behavior. Althoug that present there have been no studies directly aimed at improving the social skills of children with WS, some information is available regarding strategies for improving social skills in other populations of children with similar difficulties (e.g., ID of mixed etiologies, ASDs, children with emotional and behavioral problems).

The primary purpose of Social Skills Training is to increase the child's ability to perform key social behaviors that are important in achieving success in social situations [Herbert, 1996; White et al., 2007]. Programs typically target a variety of skills including improving conversation skills (e.g., appropriate physical distance, how and when to interrupt, asking questions when you do not understand, topic maintenance), cooperative play skills (e.g., joining others in play, sharing, taking turns, compromising), friendship management (e.g., respecting personal boundaries, getting attention in positive ways, sharing a friend), and emotion management skills (e.g., recognizing feelings, problem solving, empathy, conflict

management). Within Social Skills Training, each target skill is addressed using multiple methods (e.g., instruction, modeling, behavioral rehearsal, feedback, reinforcement). Most children with WS demonstrate considerable difficulty with many of the behaviors targeted by SST; teaching children with WS skills in multiple setting and using a variety of instructional techniques should increase the likelihood that the skills learned will be generalized to their everyday interactions.

Language Intervention

The onset of language is delayed for all children with WS, and most continue to have significant delays or difficulties with at least some aspects of language throughout the school years. Thus, language therapy is critical for older infants, toddlers, and preschoolers and continues to be important throughout the school years, although the focus is likely to change over time. During early development, children with WS would benefit from therapy focused on all aspects of language, with intensity determined by the child's level of delay. School-age children with moderate to severe ID and/or moderate to severe language disability would benefit from intensive language therapy focused on all aspects of language. Language intervention also is important for older children and adolescents whose intellectual ability in the mild disability to low average range, especially with regard to conceptual/relational language and pragmatics.

Many early intervention therapists use the onset of referential communicative gestures as the indicator that a child is ready for language therapy aimed at vocabulary acquisition. Thus, the pattern evidenced by young children with WS of starting to produce referential language prior to using (or even comprehending) referential gestures often leads to significant delays in the onset of language therapy. Similarly, if a child is not referred to an early intervention program until after he or she has started to talk, the presumption is often made that the child has mastered basic referential gestures Both of these presumptions are incorrect. Children with WS are ready to begin therapy aimed at vocabulary acquisition well before they begin to produce referential pointing gestures, and young children with WS who talk well almost always still have difficulty with pragmatic aspects of language, including referential gesture comprehension and production.

For preschool and school-age children, a full assessment of all aspects of language and communication, including extensive observation of the child's linguistic interactions with teachers and peers, both in the classroom and on the playground, is critical to determining if an individual child would benefit from language intervention. All too often, as soon as the child's articulation is clear and he or she no longer makes consistent grammatical errors, speech/language therapy is discontinued, even though he or she continues to have considerable difficulty with both conceptual/relational language and pragmatics. These difficulties impact both the child's academic performance and his or her social interactions with other children, negatively affecting peer relationships. These difficulties are best addressed by a coordinated multidisciplinary approach including the language therapist, classroom teachers, special educators, and aides, as well as the child's parents; as the child gets older and more aware of these difficulties, he or she should also be included in goal selection and implementation. Carefully conceived and well-designed intervention studies targeting the efficacy of particular methods of language intervention (including, among other methods, either music therapy or the use of music within more traditional language therapy) for children and adolescents with WS, the relative advantages and disadvantages of individual or group therapy, and optimum therapy intensity (e.g., amount of time per week and the number of sessions into which that time should be divided) for particular types of language issues would provide crucial input for the design of an educational environment that would allow children and adolescents with WS the opportunity to reach their full academic and social potential.

Reading Intervention

To date, only one study has considered the effect of type of reading instruction method on the performance of children with WS on standardized reading assessments [Becerra et al., 2008; Mervis, 2009]. Results indicated the importance of using a systematic phonics approach rather than a whole word or whole language approach. Data from this study suggest that only about half of the children with WS (and only a very small proportion of those with mild to moderate ID) are being taught reading using a systematic phonics approach. Use of this type of approach has been endorsed by the National Reading Panel for all children [McCardle et al., 2008]. Furthermore, as has been demonstrated for children in the general population [e.g., Ehri, 2004] and children with DS[e.g., Bourassa et al., 2005; Cupples and Iacono, 2002], children with WS who are taught to read using a systematic phonics approach read significantly better than do children who are taught with a whole-word approach, relative to expectations based on IQ. It is particularly important that phonics be used from the beginning of reading instruction; children who are initially taught using a whole word (sight-word) or whole language approach quickly develop a strategy of guessing a word based on its overall shape or first letter (or by looking at the accompanying pictures). Even if systematic phonics instruction is provided later in the child's education, the guessing strategy will have been so well established that it will be difficult to overcome, especially given the low levels of frustration tolerance and mastery motivation evidenced by most children with WS.

When working on phonemic awareness skills with children with WS, it is important to focus on only a few skills at a time. Of particular importance are the ability to blend (combine a series of separate phonemes into a word) and to segment (break a word into its segments, often accompanied by tapping, clapping, etc.). In addition, use of actual letters rather than just sounds or blank tokens (e.g., colored squares), delivery within a small group setting, and providing phonemic awareness instruction in kindergarten or first grade increases the effectiveness of the instruction method [see review in Ehri, 2004; McCardle et al., 2008]. For children with WS who also have difficulty learning or remembering letter-sound correspondences, use of mnemonic devices (e.g., incorporating an object or a photograph of an object that starts with the letter sound when presenting the letter to the child) may be helpful.

As discussed by Mervis [2009], children learn best when they are being actively taught, that is with the teacher both explaining and modeling phonics principles while also providing the child with practice in which they also receive feedback. Worksheets completed "independently" by children with WS are not an effective means of teaching phonics. In addition, as children with WS often have difficulty generalizing explicitly taught rules to new material, it is better for them 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. As such, phonics instruction should be integrated into reading and writing instruction as opposed to being taught as a stand-alone topic. Children will need to be taught and encouraged to apply their knowledge of phonics when they encounter a new word, rather than reverting to guessing.

The ability to read single words is clearly necessary for reading comprehension. However, other skills are important as well, particularly fluency and broad language abilities such as comprehension monitoring and drawing inferences. Meta-analyses of studies of the reading development of children with learning disabilities have indicated that reading outcomes were best when systematic phonics instruction was combined with comprehension strategy instruction [Ehri, 2004].

Fluency depends on not only the ability to read single words well, but also the ability to appropriately group words into grammatical units to provide the basis for reading with expression. When a child can read fluently, cognitive resources are freed so that he or she may focus on comprehension. Fluency requires extensive practice. Suggestions for increasing the fluency of children with WS are provided in Mervis [2009].

Although concrete vocabulary is a relative strength for most children with WS, relational/conceptual vocabulary is a clear weakness and grammatical comprehension may be limited. Success in reading comprehension requires comprehending the words and grammatical structures used in the text. These skills should be address both in speech/language therapy and in reading instruction. Beyond addressing these skills, children with WS require instruction in specific comprehension strategies, for example in comprehension monitoring, in graphic organizers, in question-generating (self-questioning), and in summarization (including identification of central ideas, making inferences, and generalizing from the text). McCardle et al. [2008] provide detailed descriptions of these techniques and their application. For all these skills, children with WS will require explicit instruction and extensive practice, including opportunities to generalizing the skills they are learning.

SUMMARY AND CONCLUSION

Although media portrayals of WS have changed very little over the past two decades, considerable progress has been made in behavioral research, yielding a more nuanced cognitive and behavioral phenotype for this syndrome. WS is most commonly associated with borderline to mild intellectual disability, although the range of ability extends from severe intellectual disability to average intelligence. Average level of ability is not constant across the intellectual domains commonly sampled on intelligence tests. The most common cognitive profile for WS is characterized by relative strengths in (concrete) language, verbal short-term memory, and (concrete) nonverbal reasoning accompanied by considerable weakness in visuospatial construction. Within the language domain, a characteristic profile also has emerged, with a relative strength in concrete vocabulary, grammatical abilities at about the same level as general intellectual ability, and considerable weakness in both relational/conceptual language (with performance at about the same level as for visuospatial construction) and pragmatics. Within pragmatics, considerable overlap with the autism spectrum has been noted. Although much more research is needed, the pattern of strengths and weaknesses identified in the few studies of memory and executive function is broadly consistent with that for the overall cognitive profile, with verbal memory significantly stronger than spatial memory (at least when mental manipulation is not required) and performance on executive function tasks stronger for verbal inhibition measures than for motor inhibition measures. The pattern of strengths and weaknesses in adaptive behavior also mirrors the overall cognitive profile, with relative strengths in domains that depend heavily on verbal skills (e.g., social interaction and communication) and considerable weakness in domains that depend on visual-motor integration or spatial skills (self-help skills or community living skills).

Despite frequent lay characterizations of WS as the “opposite” of autism, a number of shared characteristics with the ASDs have emerged, including difficulties with pragmatics, anxiety, attention, and behavior. Sensory problems also are common for both groups. These shared characteristics provide a starting point for identifying intervention strategies (e.g., ABA-based approaches, Social Skills Training) that when applied systematically are likely to help ameliorate the social and behavior problems that frequently characterize children with WS. Other more traditional types of intervention such as speech/language therapy and occupational therapy also are important, as is treatment for anxiety disorders. Development of treatments to address the unusually low level of mastery motivation evidenced by

children with WS is critical. Research on intervention for academic skills is just beginning Nevertheless the important finding that children with WS who are taught to read using a systematic phonics approach typically read at least as well as expected for IQ while children who are taught with more holistic methods (whole whole, sight word, whole language) typically read below the level expected for IQ, combined with the likely impact of pragmatics difficulties such as comprehension monitoring on reading comprehension, provides a starting point for recommendations regarding reading intervention. Considerably more research is needed both on basic-science aspects of cognition and behavior in children with WS and on intervention strategies. The ultimate goal, however, remains the same: to provide a sound research basis for the development of the educational, social, and behavioral interventions needed for children with WS to have the opportunity to reach their full potential.

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Table I

Descriptive Statistics for Standardized Assessment Performance of Children and Adolescents with WS

Measure	N	CA range (in years)	Mean Standard Score (SS)	SD	SS Range
Differential Ability Scales-II					
GCA (similar to IQ)	120	4.01 – 17.71	64.56	12.33	31 – 96
Verbal Cluster SS	120	4.01 – 17.71	74.06	16.41	30* – 111
Nonverbal Reasoning Cluster SS	120	4.01 – 17.71	78.89	15.44	37 – 118
Spatial Cluster SS	120	4.01 – 17.71	54.82	11.27	32* – 81
Recall of Digits—Forward SS	120	4.01 – 17.71	72.06	15.71	40* – 102
Mullen Scales of Early Learning					
Early Learning Composite	144	2.01 – 4.96	61.45	11.31	49* – 96
Visual Reception T	144	2.01 – 4.96	29.51	3.21	20* – 58
Fine Motor T	144	2.01 – 4.96	21.18	9.68	20* – 41
Receptive Language T	144	2.01 – 4.96	29.45	9.58	20* – 55
Expressive Language T	144	2.01 – 4.96	32.60	11.31	20* – 56
Scales of Independent Behavior-Revised					
Broad Independence	122	4.02 – 17.77	55.11	15.45	24* – 95
Motor Skills	122	4.02 – 17.77	57.82	15.13	24* – 88
Social Interaction and Communication Skills	122	4.02 – 17.77	73.16	14.72	30 – 110
Personal Living Skills	122	4.02 – 17.77	61.22	14.53	24* – 98
Community Living Skills	122	4.02 – 17.77	57.35	17.20	24* – 96
Peabody Picture Vocabulary Test-4	129	4.01 – 17.71	81.84	15.04	20* – 124
Expressive Vocabulary Test-2	129	4.01 – 17.71	79.43	14.83	20* – 120
Test of Relational Concepts	92	5.00 – 7.95	55.79	21.37	25* – 104
Test for Reception of Grammar-2	170	5.02 – 17.71	74.55	17.73	55* – 116

Note: For the general population, mean = 100 (SD = 15) for SS and mean = 50 (SD = 10) for T scores.

*lowest possible SS or T for the relevant assessment