DISSERTATION

# PRAGMATIC LANGUAGE PROFILES IN AUTISM AND WILLIAMS SYNDROME

Submitted by

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WE HEREBY RECOMMEND THAT THE DISSERTATION PREPARED UNDER OUR SUPERVISION BY AMY PHILOFSKY ENTITLED PRAGMATIC LANGUAGE PROFILES IN AUTISM AND WILLIAMS SYNDROME BE ACCEPTED AS FULFILLING IN PART REQUIREMENTS FOR THE DEGREE OF DOCTOR OF PHILOSOPHY.

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ABSTRACT OF DISSERTATION

PRAGMATIC LANGUAGE PROFILES IN AUTISM

AND WILLIAMS SYNDROME

The purpose of this study was to compare pragmatic language functioning in children with autism and Williams syndrome using The Children's Communication Checklist-2 (CCC-2), a standardized parent report measure. Previous research has suggested pervasive pragmatic language impairment in autism and a profile of relative pragmatic strength and weakness in Williams syndrome. The emerging literature on pragmatics in Williams syndrome has suggested a possible need for pragmatics interventions like those used in autism (Laws & Bishop, 2004). This study compared overall communication and pragmatic language functioning in school-aged children with autism and Williams syndrome and included a group of younger, typically developing school-aged children to assess the construct validity of the CCC-2. Results suggested that the ten subscales of the CCC-2 showed good internal consistency in the entire sample. As expected, the typical group demonstrated typical communication and pragmatic language functioning, while both clinical groups showed significant impairment in overall communication abilities, overall pragmatic impairment, and overall pragmatic language functioning and were indistinguishable from each other on these dimensions. However, the Williams syndrome group showed a trend of slightly better overall pragmatic functioning compared to the autism spectrum disorder group, when controlling for expressive age equivalent. Additionally, some, but not all children with Williams syndrome and autism spectrum disorder were identified as having a pragmatic language impairment using the CCC-2. However, derivation of another variable indicative of overall pragmatic language functioning suggested that almost all children in both groups evidence some degree of difficulty in overall functioning. Examination of the

profiles between autism spectrum disorder and Williams syndrome revealed equivalent performances on most of the subscales of the CCC-2, however better performance in the Williams syndrome group was found on the Stereotyped Language and Nonverbal Communication subscales. A trend of better performance in Williams syndrome was also noted on the Social Relations subscale. Further, overall pragmatic language functioning was strongly associated with the adaptive Socialization Standard Score of the Vineland Adaptive Behavior Scales/II, providing evidence for convergent validity of the CCC-2 with another tool measuring a similar construct. Clinical and research implications were provided.

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## Chapter I: Introduction

Pragmatics is the linguistic domain concerned with the appropriate use of language across a variety of social contexts that provides for a listener's accurate interpretation of the speaker's intentions and references (Berko-Gleason, 1993; Rice, Warren, & Betz, 2005). Pragmatics comprises a critical component and intersection for children's developing language competencies and social interactions. Despite its importance with respect to children's linguistic and social development, assessment of pragmatics can be difficult. Nonetheless, standardized measures for assessing pragmatics are available and studies suggest that these measures can reliably identify pragmatic language impairment in children (Bishop & Baird, 2001; Gilmour, Place,& Skuse, 2004; Guerts et al., 2004; Laws & Bishop, 2004; Norbury, Nash, Baird, & Bishop, 2004).

Some developmental syndromes place children at particular risk for pragmatic language impairment. Autism and Williams syndrome represent two examples of such syndromes (Dawson et al., 2002; Gillberg & Rasmussen, 1994; Laws & Bishop, 2004; Semel & Rosner, 2003), despite the fact that the disorders present very differently socially (e.g., asocial in autism and hypersocial in Williams syndrome). Standardized assessment of pragmatic language abilities is particularly important for these children with respect to the potential determination for service eligibility and the development and monitoring of appropriate interventions.

Rice and colleagues (2005) argue that there is a current research need for pairwise comparisons across different developmental conditions to better understand differences and

similarities observed between linguistic profiles. While pragmatic language impairments have been documented separately in both autism and Williams syndrome, the two syndromes have not been directly compared previously on this dimension. Such lack of comparison is likely relatable to difficulties with accessible assessment tools for pragmatic language impairment and difficulties matching two syndromes characteristically mismatched with respect to verbal abilities, along with the current limitations in use of this methodology in language studies (Bishop, 1998; Young, Diehl, Morris, Hyman, & Bennetto, 2005; Rice et al., 2005). Additionally, pragmatic language impairment on a standardized measure of pragmatic assessment has not been replicated in children with Williams syndrome. Further, the sample of children used in the only study looking specifically at pragmatics in Williams syndrome (Laws & Bishop, 2004) comprised a variable age range from children to young adults, spanning several different age cohorts including school-age, adolescence, and young adulthood.

Thus, the purpose of this study was to compare and describe the pragmatic language profiles of school-age children with autism and Williams syndrome, using typically developing children as a control group for understanding what is typical within the pragmatics construct, to determine whether or not pragmatic profile characteristics can differentiate between the two groups of children with opposite social presentations and pragmatic language difficulties. Such research will help to clarify some of the similarities and differences of the pragmatic language characteristics of children with autism and Williams syndrome from a school-age cohort, which is important for better understanding how an atypical social profile potentially impacts upon pragmatic language development in the school-aged years. Also, this study will further serve to underscore the importance of standardized pragmatic assessment by speech-language pathologists in these populations.

*List of Keywords*

**Pragmatic Language**—the aspect of language concerned with the appropriate use of language across a variety of social contexts that provides for a listener's accurate interpretation of the speaker's intentions and references.

**Structural Language**—the aspect of language concerned with the rules for putting words together into grammatically correct, understandable, and meaningful sentences, as well as the relations between words and what they denote (i.e., syntax and semantics).

**Autism**—a neurodevelopmental, behaviorally diagnosed pervasive disorder of development that consists of three core areas of difficulty: 1) language and/or communication delays and/or unique pragmatic features; 2) difficulties with core social relating and social relatedness; 3) restricted and/or repetitive interests.

**Williams syndrome**—a syndrome caused by a deletion on chromosome 7 that results in a characteristic behavioral phenotype that uniquely impacts upon language and cognitive abilities and results in hypersociability, as well as a cluster of maladaptive behaviors.

*Research Questions*

1. Can a standardized tool of pragmatic language abilities using parents as raters detect impairment in school-aged children with autism and Williams syndrome?

2. What are the differences, if any, in the pragmatic language profiles of school-aged children with autism and Williams syndrome?

3. Is pragmatic language impairment in children, as rated by their parents and measured by a standardized tool, related to other measures of social functioning?

*Hypotheses*

1. The pragmatic language profiles of school-aged children with autism and Williams syndrome, as determined by the Children's Communication Checklist-2 (CCC-2), will be similar, with a pragmatic language impairment, relative to structural language performance in both groups. While there may some CCC-2 items that are different for children with autism and children with Williams syndrome, as a function of how the questions parsed into subscales, both groups will show similar levels of impairment across every pragmatic subscale.

2. While the pragmatic language profiles will be able to reliably separate typical children from children with autism and children with Williams syndrome, the profiles will not clearly distinguish between children with autism and Williams syndrome. Rather, the tool will indicate commensurate levels of pragmatic language impairment across both syndrome groups, again as a function of how the items parse into the different subscales on the CCC-2.

3. Pragmatic language impairment, as evidenced by the CCC-2, will be highly associated with decreased performances on another measure of social functioning, the adaptive Socialization domain Standard Score of the Vineland Adaptive Behavior Scales (VABS, 1rst or 2nd edition).

## Chapter II: Literature Review

### *Pragmatics*

Originally, Charles Morris (1938), a prominent philosopher and academician, delineated three distinct domains which have come to characterize the field of linguistics: syntax, semantics, and pragmatics. While syntax concerns the formal relations of signs (i.e., words) to one another and rules concerning how words are organized into sentences (i.e., grammar), semantics considers the relation of signs to what they denote (i.e., meaning) and vocabulary, including word knowledge. Distinct from the more structural aspects of language (i.e., syntax and semantics), pragmatics concerns the relations between signs and their interpreters, speakers and their communicative partner[s] (i.e., appropriate use; Morris, 1938; Ninio & Snow, 1996). Pragmatics has also been described as the "branch of linguistics concerned with speech use, and studies of pragmatic development are concerned with how children acquire the knowledge necessary for the appropriate, effective, rule-governed employment of speech in interpersonal situations (p. 4; Ninio & Snow, 1996)."

Beyond a basic ability to put words together into grammatically correct, understandable sentences, pragmatics inserts the notion of *appropriateness* into the concept of language use. While a sentence may be technically correct and make sense, depending upon the context in which it is used, it may be inappropriate. For example, while it may be appropriate to say, "Look at the kitty!" to a young child in a high-pitched, sweet sounding voice, such use of language would be inappropriate in a business meeting, although it might technically "make sense." Thus, not only do pragmatic language abilities rely upon intact

structural language skills, but moreover upon a larger knowledge base of the social context, specific communication contexts, and an understanding of the communication partner as well as, general world knowledge (Martin & McDonald, 2003). That helps to explain why an atypical social trajectory might be associated with pragmatic language difficulties.

Ninio & Snow (1996) have conceptualized the scope of pragmatics as falling somewhere within the following three broader categories: 1) understanding and using the rules that govern how communication behaviors are used effectively; 2) conversation skills and; 3) engaging in long, back-and-forth exchanges, which includes adapting one's communication effectively for different audiences, and narrating or story-telling. Additionally, Ninio & Snow's (1996) categories are organized according to developmental acquisition, with the first category of behaviors representing the earliest emerging pragmatic competence. A number of supra-linguistic factors are taken into account within the scope of pragmatics, though linguistic factors (i.e., what you say) also matter to some extent. Use of eye contact, use of gestures, speech function (i.e., what a communicator is trying to accomplish with what is being said, such as requesting, directing, etc.), appropriate establishment of a conversation topic (i.e., providing the appropriate amount of background information that the particular listener requires to understand what is being said as well as keeping track of what the listener may already know), appropriate maintenance of a conversation topic (i.e., staying with and supporting an existing conversation topic), conversational repairs (i.e., repairing a breakdown when a miscommunication occurs), tone of voice, and prosody (i.e., the melody of the voice) all fall under the domain of pragmatics (Berko-Gleason, 1993).

Our current abilities to describe and assess pragmatic language abilities within typically developing children (not to mention children with developmental delays) remain

somewhat limited. For example, there remains much confusion around the construct of pragmatics with respect to what uniquely distinguishes pragmatics from other domains of language (i.e., semantics or word meanings and their interconnections may also reflect the meaning of words in different contexts; Botting & Conti-Ramsden, 1999). Difficulties in definition of the construct, in turn, make judgments and assessment of pragmatic performance problematic. Additionally, certain unavoidable biases may be introduced by cultural factors and/or co-morbid conditions (i.e., anxiety or depression; Ramberg, Ehlers, Nyden, Johansson, & Gillberg, 1996). Testing conditions pose potential challenges to the validity of pragmatic language assessment (Bishop & Adams, 1991). Furthermore, much debate continues around whether or not isolated "semantic-pragmatic" or "pragmatic language disorders" can occur separate from syndromes such as pervasive developmental disorders, complicating issues around diagnosis (Bishop, 1989; Botting & Conti-Ramsden, 1999; Norbury & Bishop, 2003; Rapin & Allen, 1998).

Despite some remaining confusion concerning scope and definitions, pragmatic language comprises a critical component of children's developing social and language competencies. While pragmatics are typically evaluated by a speech-language pathologist, this domain of language is much more difficult to assess than the more structural aspects of language (e.g., morpho-syntax, semantics and phonology; Norris, 1995; Young, Diehl, Morris, Hyman, & Bennetto, 2005). Although standardized measures have recently become available that reliably determine pragmatic language impairment in children, speech-language pathologists typically rely on observational and descriptive assessments when considering the pragmatic language abilities of children (ASHA, 2006; Bishop & Baird, 2001; Gilmour, Place, & Skuse, 2004; Guerts et al., 2004; Laws & Bishop, 2004; Norbury, Nash, Baird, & Bishop, 2004; Young et al., 2005). However, without standardized test results most speech-

language pathologists do not have the flexibility to determine service eligibility (Kamhi, 1998). Further, lacking standardized assessment makes it more difficult to monitor treatment or developmental progress.

*Assessment of Pragmatic Language Skills*

There are three main ways in which pragmatic language skills have been traditionally assessed in children: 1) standardized tests that elicit the behaviors of pragmatic language skill, 2) observational methods for identifying pragmatic language skills within semi-naturalistic settings (e.g., analysis of a laboratory and/or home conversational transcripts), and 3) behavioral ratings made by someone who knows the child (Bishop, 1998). Each method has its own unique benefits and limitations. Standardized testing provides uniform prompts and procedures, such that every child is exposed to and responds to the same set of stimuli, therefore introducing few extraneous variables to the testing condition. In the particular case of pragmatics, however, it has been demonstrated that children with pragmatic difficulties tend to perform better in situations that involve explicit instruction (as is the case with standardized testing) compared to dynamic, natural situations, as is the case with daily life (Bishop & Adams, 1991; Bishop, 1998). By definition, pragmatics represent skills that are utilized within the dynamic and natural contexts of daily life, thus necessarily precluding the use of standardized testing alone as a valid forum for consideration of this behavior.

Observational methods for identifying pragmatic language skill within natural settings have been successfully employed at both micro- and macro-analytic levels (Bishop, 1998; Siebert, Hogan, & Mundy, 1982; Stojanovik, 2006). Assessment within natural conversational contexts is partially achieved (though often in a laboratory setting), bypassing the improvement that children with pragmatic difficulties can show when provided explicit

instruction for a task (Bishop & Adams, 1991). However, the short timeframe within which pragmatic language skills are observed in the laboratory may not necessarily enable generalization to all contexts (i.e., daily life; Bishop, 1998). Further, behaviors that occur only rarely may not be present during observational timeframes, creating uncertain answers about what constitutes typical pragmatic language functioning. Finally, micro-analytic techniques, which involve language transcription, are extremely time-consuming and require extensive training to provide for acceptable levels of inter-rater reliability (Bishop, 1998).

Behavioral ratings made by someone who knows the child have been suggested to provide an efficient and reliable method for assessing pragmatics for both clinical and research purposes (Bishop, 1998). Although subjective interpretation by a rater involves some inherent disadvantages, the advantages are more numerous. They take a relatively short period of time to complete and score, relative to other measures of pragmatics. When completed by a rater who is familiar with the child, such assessments likely yield a representative overall picture of the child that is not affected by minor daily fluctuations (Bishop, 1998). Further, this method circumvents a disadvantage applicable to both of the prior two methods by providing for assessment of behaviors that may be difficult to elicit in test situations or that occur rarely (Bishop, 1998). Finally, assessment of pragmatics in this manner has been deemed adequately reliable and valid when rated by parents, teachers and therapists through standardization procedures (Bishop, 1998; Bishop, 2003).

Recently, a pragmatic language assessment tool that employs behavioral ratings, the Children's Communication Checklist-2 (CCC-2), has become available for identifying children with pragmatic language impairment. Content validity for the CCC-2 is available within the history of the instrument's development, which is described in its manual, as well as in the original paper describing the tool (Bishop, 1998; Bishop, 2003). Originally devised

as a research tool whose main objective was to identify children with so-called semantic-pragmatic disorder (Rapin & Allen, 1993) from within a larger language impaired sample, the Checklist for Language Impaired Children (CLIC) consisted of 20-items to be completed by teachers or therapists in which the rater had to determine the best fitting description among five for a particular child. Revised into the CLIC-2 following feedback from respondents regarding dissatisfaction with having to determine one best fitting description among five, the five items per question were parsed out into individual questions. A large-scale reliability was conducted on the CLIC-2 providing information with respect to the following: inter-rater reliability and preliminary construct validity to determine which items truly assisted in identifying the target group (i.e., isolated pragmatic language impairment) from language impaired children.

Out of the CLIC-2, the first Children's Communication Checklist (CCC) was developed. The items selected for the CCC consisted of those with the best inter-rater reliability and statistical criterion for internal consistency on the CLIC-2 (Bishop, 1998). Thus, the CCC consisted of 9 scales: A-Speech; B-Syntax; C-Inappropriate Initiation; D-Cohesion; E-Stereotyped Language; F-Use of Context; G-Rapport; H-Interests; and I-Social Interaction. Scales C through G combined formed into a pragmatic language composite, while scales A and B were included to cover the more structural aspects of language for the purposes of distinguishing children with pragmatic language disorder from children with structural language disorders. Scales H and I were included to index behaviors particular to autistic disorder in an attempt to further examine the clinical and research question of whether or not semantic-pragmatic disorders can exist separate from an autism spectrum disorder. Conti-Ramsden, Crutchley, & Botting (1997) initially conducted studies on 242 language-impaired 7-year olds determining an inter-rater reliability of .80 for the pragmatic

language composite, as well as construct validity data consisting of confirmation between children's school record diagnoses and their diagnostic classification on the CCC. In 1998, Bishop published a paper about this instrument leading to a surge of interest for possible wider application of the tool including as a screener for language impairment and for identifying pragmatic language impairment within children who also have psychiatric disorders. Additionally, the notion of obtaining useful information from parents, as opposed to teachers and therapists, was raised. A subsequent study suggested that while parent-teacher reliabilities were only moderate (r =.45), both sets of ratings demonstrated construct validity in terms of consistency with the child's diagnosis and psychometric data (Bishop & Baird, 2001). Feedback from users of the CCC resulted in subsequent re-organization and additions to improve the instrument's psychometric properties and revision into the CCC-2.

A subsequent study conducted by Norbury and colleagues (2004) was conducted in order to validate the use of the CCC-2 with parents as raters, as use of parents as raters for the CCC had up to that point yielded unacceptable reliability data (Bishop & Baird, 2001). In this study, 118 parents of typically- developing children, as well as children with established diagnoses of specific language impairment (SLI), pragmatic language impairments or autism spectrum disorders according to school records completed a CCC-2. Results suggested that the CCC-2 could distinguish between children who had any type of communication impairment and children who did not, based upon a summary variable, the General Communication Composite (i.e., GCC) score. While distinction between types of communication impairment (i.e., structural or pragmatic) was not possible by looking directly at the General Communication Composite or pragmatic subscales (i.e., E-H), the Social Interaction Deviance Composite (i.e., SIDC), a qualitative variable of relative strength consisting of the sum of two pragmatic and two social, "autism" summed scaled scores (i.e.,

E+F+I+J) minus the sum of the scaled structural domain scores (i.e., A+B+C+D), was devised for distinguishing between structural and pragmatic impairment in all children with a communication impairment, honoring the original goal of the instrument: to identify children who have a pragmatic language impairment from a group of larger children with varying types of language impairment. Additionally, introduction of the Social Interaction Deviance Composite variable improved the inter-rater reliability of parents as raters with professionals to an acceptable level (i.e., $r = .79, p < .001$; Norbury et al., 2004).

Reliable, standardized assessment of pragmatics is particularly important for qualification of services and assessment of children who are at an increased risk for difficulties with pragmatic language development (Bishop, 1998; Young et al., 2005). Autism and Williams syndrome represent two disorders that significantly impact upon language development, with the linguistic domain of pragmatics implicated in particular in both (Dawson et al., 2002; Laws & Bishop, 2004; Semel & Rosner, 2003). Standardized assessment of pragmatics in these populations comprises an essential component of identifying impairment, determining appropriate treatment goals for intervention and tracking treatment progress.

*Williams Syndrome*

Williams syndrome is a genetic disorder, with prevalence estimates ranging from 1:7,500 to 1:25,000 live births and presents equally in males and females (Bellugi, Lichtenberger, Jones, Lai, & St. George, 2000; Stromme, Bjornstad, & Ramstad, 2002). The syndrome typically results from a sporadic microdeletion on a portion of chromosome 7, referred to as "the Williams syndrome critical region (Ewart et al., 1993; Morris & Mervis, 1999)." Researchers have focused their efforts on disinterring a very distinctive behavioral phenotype for children with the syndrome, primarily through the study of older children and

adults with the disorder (Bellugi et al., 2000; Dykens, 2003a; Mervis & Klein-Tasman, 2000). Broadly speaking, this distinctive behavioral phenotype is characterized by a unique cognitive-linguistic profile, accompanied by an unusual and distinctive personality style, distinguished by hypersociability and a cluster of maladaptive behaviors (Bellugi, Lichtenberger, Mills, Galaburda, & Korenberg, 1999; Dykens, 2003b; Hodapp & Fidler, 1999; Mervis & Klein-Tasman, 2000).

In terms of the cognitive-linguistic profile, most individuals with Williams syndrome demonstrate mental retardation, although approximately one quarter of individuals demonstrate milder forms of cognitive difficulties in the form of learning disabilities (Mervis & Klein-Tasman, 2000). Peaks and valleys of marked strength and weakness are characteristic of the cognitive profile of children with Williams syndrome (Mervis & Klein-Tasman, 2000). An initial example of this within Williams syndrome came in the form of anecdotes relaying remarkable social and language abilities amidst clear evidence of severe impairment in other cognitive domains (i.e., visuo-spatial processing; Bellugi et al., 2000; Bellugi, Wang, & Jernigan, 1994).

While strengths within the cognitive-linguistic profile for individuals with Williams syndrome tend to fall under the domains of verbal processing, language (specifically in the peaks of expressive vocabulary, expressive grammar, and story-telling), auditory rote memory, and facial recognition skills, weaknesses have been noted in most areas of visuo-spatial processing (exception facial recognition skills) and visuo-motor functioning (Bellugi et al., 2000; Mervis, 2003; Mervis & Klein-Tasman, 2000; Semel & Rosner, 2003). Despite some areas of notable strength in language functioning, language development is typically delayed in Williams syndrome (Singer-Harris, Bellugi, Bates, Jones, & Rossen, 1997).

As some strengths are truly strengths (such that children with Williams syndrome perform similar to typical controls for example, on auditory rote memory and facial recognition skills), other "strengths" have been tempered by closer inspection of the skill within this population (Vicari, Carlesimo, Brizzolara, & Pezzini, 1996; Karmiloff-Smith et al., 2004). For example, the original remarkability of overall language skill in this population has more recently been tempered by findings noting both peaks, as well as valleys, within the understanding and use of language for children with Williams syndrome (Bellugi et al., 2000; Mervis, 2003; Stojanovik, Perkins, & Howard, 2001). Language strengths may be observed in good recall of previously heard words and phrases (Udwin & Yule, 1990), socially engaging use of prosody, discourse, and narrative skills (Bellugi et al., 1999), as well as stronger expressive morpho-syntactic skills, compared to other syndromes of mental retardation (Bellugi et al., 1994; Karmiloff-Smith, 1998; Volterra, Capirci, Pezzini, Sabbadini & Vicari, 1996). Weaknesses, on the other hand, have been noted in word-finding abilities, the understanding and use of abstract language and concepts, and generally emerge as linguistic tasks become increasingly complex (Don, Schellenberg, & Rourke, 1999; Klein & Mervis, 1999; Rossen, Klima, Bellugi, Bihrle, & Jones, 1996; Semel & Rosner, 2003). It has also been suggested that children with Williams syndrome may not understand everything that they express (Lichtenberger, Jones, Lai, & St. George, 2000), potentially resulting in unusual language use (i.e., pragmatic difficulties; Bellugi et al., 1994).

One final important aspect to understand about the cognitive-linguistic profile of this population deals with the idea that some specific subdomains within larger domains may actually be both a strength and a weakness. Diction or word choice as a specific subdomain within the larger domain of language provides a nice example of a skill that can be seen as both a strength and weakness in this population; individuals with Williams syndrome are

said to be both "sophisticated" and "slightly off the mark" in their diction (Bellugi et al., 1994; Semel & Rosner, 2003). For example, Bellugi, Wang, & Jernigan (1994) noted the use of the following sentence by a school-aged child with Williams Syndrome: "The bees abort the beehive" for "they leave the hive (p. 32; Bellugi et al., 1994)." This example provides a nice evidence of accurate—even sophisticated—structural language amidst unusual pragmatics.

In terms of distinctive personality traits, hypersociability and musicality have generated empirical data to support their role in a Williams syndrome behavioral phenotype (Semel & Rosner, 2003). Not unlike the cognitive-linguistic profile, strengths curiously accompany weaknesses. On the positive side, individuals with Williams syndrome tend to be friendly, outgoing, empathic to others, socially interested and curious, and possessing a special aptitude for music (Semel & Rosner, 2003). Less adaptive personality features have to do with overfriendliness amidst difficulties with friendships, oversensitivity, difficulty with higher-level social cognitive skills, and curiosities and interests that have the potential to intensify into obsessions and/or restricted and repetitive interests (Dykens & Rosner, 1999; Semel & Rosner, 2003; Tager-Flusberg & Sullivan, 2000). Also, individuals with Williams syndrome give abnormally positive approachability ratings to unfamiliar people compared to chronological-age and mental-age matched controls (Jones et al., 2000), which may predispose individuals to social vulnerability (Semel & Rosner, 2003).

Research into social cognition using laboratory "theory of mind" tasks has further provided some explanation to some of the personality characteristics notable of Williams syndrome. An interesting dissociation in mental state understanding has been reported in this population. Although school-aged children with Williams syndrome are able to

understand others' minds based upon social-perceptual information (e.g., facial expressions, including eye gaze and emotion), understanding another's mind proved difficult when it was necessary for children with Williams syndrome to think about others' thoughts and beliefs (Tager-Flusberg & Sullivan, 2000), a skill that is mastered by 4 or 5-years-old in typical development (Perner, Leekam, & Wimmer, 1987; Wimmer & Perner, 1983). Thus, individuals with Williams syndrome show an interesting dissociation of strength in a "perceptual" understanding of mental states (i.e., reading another's mental state based upon their eyes and facial expressions), with marked weaknesses in a "cognitive" understanding of others' minds (i.e., thinking about what and how others might think or believe in different situations; Tager-Flusberg & Sullivan, 2000). They further posited that this pattern of results may be reflected in the social style of older individuals with Williams syndrome, because of a strong social interest and empathic quality, occurring amidst difficulties with maintaining friendships and making poor social judgments (Tager-Flusberg & Sullivan, 2000).

Finally, maladaptive behaviors must be considered as they help to form a more complete understanding of the behavioral phenotype of this particular syndrome (Dykens, 2003b; Semel & Rosner, 2003). Semel & Rosner (2003) summarized these behaviors into the following six broader categories: 1) Fears and anxiety; 2) Distractibility and attentional problems; 3) Impulsivity; 4) Poor adaptability; 5) Low frustration tolerance and; 6) Atypical activity. Further subdivision within these broader categories has included specific investigations into aspects such as emotionality due to its remarkable impact upon how individuals with Williams syndrome adapt to a variety of situations (Semel & Rosner, 2003). Dykens (2003b) interviewed children with Williams Syndrome and their parents and found that compared to mental-age matched controls with other forms of developmental disability, individuals with Williams syndrome experienced significantly more anxiety, fears, and

phobias. Schreiber (2000) noted difficulties with both emotional regulation and over-emotionality (e.g., unable to hold back tears over what seems to be a minor upset) in this population.

### *Autism*

Autism is a behaviorally-diagnosed, neurobiological disorder that creates impairments in three core areas of functioning: communication, social, and restricted or repetitive interests/obsessions (APA, 2003). There are approximately four males for every one female with autism (Volkmar, Paul, Klin, & Cohen, 2005). Although previously considered a rare disorder, autism rates have risen alarmingly to approximately 1:300 to 1:500 children recently, placing it among the more common disorders of childhood (APA, 2003; Fombonne, 1999; Yeargin-Allsopp, Rice, Karapurkar, Doernberg, & Murphy, 2003). Mental retardation is highly variable and sometimes co-morbid with autism, occurring in roughly 25-80% of individuals (Chakrabarti & Fombonne, 2001; Fombonne, 1999). Unlike Williams syndrome, the exact cause remains unknown although it has been speculated that it results from a combination of both genetic and possible environmental influences upon fetal development (Bailey, Phillips, & Rutter, 1996; Dvbvik, 2004). To date, no reliable genetic markers for autism have been discovered, although chromosome 7 (the same one implicated in Williams syndrome) is under investigation (see review in Rutter, 2000).

With variability underscored in the autism behavioral phenotype, precision is compromised in limiting the disorder to one single behavioral phenotype. However, because autism is behaviorally diagnosed, the behaviors of individuals with autism have been intensely studied and distilled into the specifics that are necessary for making this behavioral diagnosis (APA, 2003). In line with other behavioral phenotype descriptions, the cognitive-

linguistic profile of autism develops in splintered fashion, with non-verbal functioning typically emerging as an area of strength, relative to weaknesses in verbal abilities (APA, 2003). Language development is usually severely delayed, although hyperlexia, or the ability to decode written words with limited understanding of the meaning, may occur (APA, 2003). While vocabulary and grammar acquisition (i.e., structural language) in autism range from normal to delayed, difficulties with understanding and appropriately *using* language (i.e., pragmatics) are universal, including difficulties with the use of nonverbal forms of communication (Dawson et al., 2002; Happe & Frith, 1996; Mundy, Sigman, Ungerer, & Sherman, 1986). Also, children with autism tend to be literal and concrete and have difficulty with abstract ideas in language and social reasoning, including difficulties with both social-perceptual and social-cognitive Theory of Mind tasks, or the ability to interpret others' thoughts based upon social-emotional cues and/or inference (Baron-Cohen, 1989; Minshew, Meyer, & Goldstein, 2002; MDOE, 1998). All of these factors combined contribute to difficulties for children with autism in interpreting environmental cues (Marcus, Garfinkle, & Woolery, 2001; MDOE, 1998).

Personality and behaviors contribute substantially to the autism behavioral phenotype. Typically described as unusual, disconnected, odd, and/or aloof, social skills difficulties comprise the hallmark deficit for individuals with autism. Difficulties with basic social relatedness and reciprocity help to comprise the overall social deficits characteristic of autism (Hobson, 1993). Of additional note, maladaptive behaviors form a significant portion of the behavioral phenotype in autism with a range of possible symptoms including: restricted and/or repetitive interests (i.e., obsessions); difficulties with attention and/or activity; aggressiveness to self and/or others; a high requirement for routine and difficulties with transitions; unusual responses to sensory stimuli; eating and sleeping abnormalities;

mood abnormalities (i.e., laughing for no apparent reason); and difficulties with anxiety and/or a lack of fear response to real dangers (APA, 2003).

Williams syndrome and autism both have known behavioral phenotypes that include language symptomology, and other investigators have pioneered the comparison of these two disorders (Gillberg & Rasmussen, 1994; Grice et al., 2001). The importance of comparisons between developmental disorders has been highlighted for understanding the commonalities across conditions, as well as "the nature of language impairments and the ways in which the human language capacity is vulnerable (p. 8; Rice et al., 2005)." While these two disorders present as polar opposites at first glance (e.g., asociability in autism and hypersociability in Williams syndrome), both syndromes are also notable for several similarities. Gillberg & Rasmussen (1994) noted the following similarities between autism and Williams syndrome: social difficulties, including isolation and difficulties with friendships; rigidity; ritual behaviors and obsessiveness; attention difficulties; and pragmatic language difficulties. See Table 1 for a comparison of the two syndromes.

The linguistic domain of pragmatics, in particular, has been implicated in both autism and Williams syndrome (Dawson et al., 2002; Gillberg & Rasmussen, 1994; Laws & Bishop, 2004). Although the two syndromes are conceptually opposite in terms of sociability (e.g., asociability in autism and hypersociability in Williams syndrome), both have been cited for specific impairment within the linguistic domain of pragmatics (Dawson et al., 2002; Laws & Bishop, 2004).

Table 1

*Comparison of the Behavioral Phenotypes for Autism and Williams Syndrome*

| *Domain* | *Autism* | *Williams syndrome* |
|---|---|---|
| Prevalence | 1:300-500 | 1:7,500-12,500 |
| Cause | Unknown; chromosome 7 has been implicated | Deletion on a "critical region" of chromosome 7 |
| Cognition | Typically variable, ranging from severe mental retardation to average; Often strong visual-spatial skills relative to verbal intelligence | Usually mild to moderate cognitive delays; Poor visual-spatial skills relative to verbal intelligence |
| Language | Delayed language development; Variable structural language skills ranging from delayed to normal; difficulties with abstract language concepts; universal pragmatic language deficits | Delayed language development; Strengths in expressive language and story-telling; difficulties in word-finding abilities, abstract language concepts, and pragmatic language profile of strength alongside marked weakness |
| Sociability | Aloof and/or unusual; may avoid others; difficulty with social reciprocity and social relatedness; difficulties with friendships | Socially interested and curious; predilection for approaching others including strangers; friendly and outgoing; highly empathic; socially vulnerable; difficulties with some aspects of reciprocity; friendship difficulties |
| Theory of Mind (TOM) | Difficulties with both social-perceptual and social-cognitive TOM tasks | Can do social-perceptual TOM tasks; difficulties with social-cognitive TOM tasks |
| Maladaptive Behaviors | Restricted and repetitive interests; attention difficulties; possible aggressiveness; unusual responses to sensory stimuli; poor adaptability and rigidity; difficulties with eating and sleeping; mood abnormalities; anxiety | Restricted and repetitive interests; attention difficulties; impulsivity; poor adaptability and rigidity; low frustration tolerance; atypical activity; mood abnormalities; fears and anxiety |

In autism, pragmatic language impairments are a well-documented characteristic of the disorder. Pragmatic impairment in school-aged children with autism can be traced to lack of communicative intent, an area where infants with autism show significant difficulties (Shulman, Bukai, & Tidhar, 2001; Wetherby, 1986). A lack of communicative intent, which is the foundation for the model of developing pragmatics, impacts upon the development of the understanding and use of the rules that govern how communication behaviors are used effectively (Nino & Snow, 1996). By the time children with autism reach school-age, their difficulties with pragmatic areas of language functioning are often pervasive, extending from the lower-level behaviors of eye contact and nonverbal communication through higher-level conversation skills and discourse.

Theoretical accounts trace the ontogeny of autism symptomology to difficulties within the social arena during infancy important for the construction of meaning and knowledge (i.e., intersubjectivity), beginning with dyadic, face-to-face interactions, which further cascade into difficulties with a number of developmental outcomes, including pragmatic language functioning (Celani, 2004; Hepburn, Philofsky, Fidler, & Rogers, under review; Hobson, 1993; Rogers & Pennington, 1991). Dawson and colleagues (2002) comment that while the structural acquisition of language may be variable in autism ranging from normal to delayed, difficulties with language use—or pragmatics—are universal in this population of children.

Many specific studies have provided examples of pragmatic language difficulty in individuals with autism over the last 30 years. In terms of receptive pragmatics (i.e., understanding how others are using language), overly literal language understanding, difficulties with understanding gestures and body language, and trouble understanding

humor have been demonstrated in autistic populations (Dewey & Everard, 1974; Happe & Frith, 1996; Hobson, 1986; Martin & McDonald, 2003; Ozonoff & Miller, 1996). While difficulties have been reported with the ability of individuals with autism to interpret emotion in others, other studies have also reported comparable aspects of emotional interpretation between children with autism and children with other developmental delays (Loveland et al., 1997; Ozonoff, Pennington, & Rogers, 1990). Likely the skill of emotional interpretation may be unusual in autism, though not altogether absent (Loveland et al., 1997).

With respect to expressive pragmatics (i.e., how language is used) in autism, some characteristics are lacking while others comprise unusual additions. Difficulties consistent with lacking have included a limited use of facial expressions and gestures; a lack of reciprocity in conversations with others, including difficulties with topic establishment and maintenance; an impaired, lacking prosody (i.e., melody of the voice); a lack of cohesive ties in the conveying of information to others; as well as difficulties with the use of pronouns, suggesting problems with referential skills (Baltaxe, 1977; Dewey & Everard, 1974; Freeman & Dake, 1996; Happe & Frith, 1996; Lord, Rutter, & LeCouteur, 1994; Martin & McDonald, 2003; Szatmari, Bartolucci, & Bremmer, 1989). Characteristic difficulties more relatable to unusual additions have included the use of uninhibited, socially inappropriate comments; an over-use of stereotyped utterances and the use of tangential language; as well as an increased use of neologisms (i.e., novel, made up words for things) and idiosyncratic language (Dewey & Everard, 1974; Lord, Rutter, & LeCouteur, 1994; Volden & Lord, 1991). By contrast, an area of possible preserved pragmatic functioning may lie in the ability to repair communication breakdowns as nine subjects with autism were comparable in employing a range of repair strategies to a language matched control group in a study

designed to consider this behavior, although off-topic comments were also significantly more common in the autism group for attempted repairs beyond the initial one (Volden, 2004).

Research on narrative skills (i.e., story-telling) in autism has been somewhat limited and conflicting, however initial findings seem to support a possible strength at the local level of story-telling with more difficulties at the global level in this population (Loveland, McEvoy, Kelley, & Tunali, 1990; Loveland & Tunali, 1993; Norbury & Bishop, 2003; Tager-Flusberg & Sullivan, 1995). Tager-Flusberg & Sullivan (1995) reported no differences among children and adults (range 6-22 years) with autism; mental-age matched, developmentally delayed controls; and mental-age matched, younger, typically developing controls on local story measures including number of different words used, number of events talked about, and syntactic complexity. Further, children with autism have been reported to re-tell the same number of story events as children with Down syndrome (Loveland, McEvoy, Kelley, & Tunali, 1990; Loveland & Tunali, 1993) with similar results replicated in comparison to typical controls on a local measure of amount of information provided by the story-teller (Norbury & Bishop, 2003).

However, children with autism may show more global difficulties in re-telling a linked, coherent sequence of events—rather simply commenting on each individual and seperate story event on a page (Loveland & Tunali, 1993). Conversely, Norbury & Bishop (2003) reported no differences between younger typical controls and children with autism in terms of a global story structure measure, though the authors suggested that the younger typical controls may not have been far enough along developmentally for a distinction to have been evident on the particular measure employed. Errors with referencing, resulting in ambiguity, and the addition of bizarre, irrelevant statements may negatively impact upon the

overall coherence of stories told by children with autism (Loveland & Tunali, 1993). Norbury & Bishop (2004) also noted an unusual, frequent use of ambiguous nouns in their sample of children with autism in re-telling a pictureless story book echoing the difficulties with referencing in narration (Loveland & Tunali, 1993; Norbury & Bishop, 2004).

A few different studies have considered the overall pragmatic language profiles on a standardized measure of pragmatics in individuals with autism determining significant impairment. A study of 15 school-aged children with autism suggested difficulties with every pragmatic subdomain of the Children's Communication Checklist (CCC). Among the particular CCC subdomains where children with autism demonstrated difficulty included: Inappropriate Initiation, Coherence, Stereotyped Language, Use of Context, and Rapport, reflective of findings from other studies of pragmatic functioning in autism (Bishop & Baird, 2002; Martin & McDonald, 2003). In a different study and group of 42 children with high-functioning autism that also employed the CCC, significant impairment in every pragmatic subdomain resulted, replicating the earlier findings of Bishop and Baird (2002; Geurts et al., 2004).

*Pragmatics in Williams Syndrome*

Research on pragmatics in Williams syndrome, conversely, remains patchy and incomplete with the vast majority of information in this area based upon observational and anecdotal reports from parents and researchers (Gillberg & Rasmussen, 1994; Laws & Bishop, 2004; Semel & Rosner, 2003). Furthermore, given such limited research, the presence of a friendly personality and "hypersociability" in Williams syndrome has led some to suggest that social communication- or pragmatics- may not be an area of weakness in this population (Jones et al., 2000; Reilly, Klima, & Bellugi, 1990; Rice et al., 2005), though others have noted deficiencies in pragmatics in Williams syndrome (Gillberg & Rasmussen,

1994; Laws & Bishop, 2004; Stojanovik, 2006). Accordingly, the pragmatic language characteristics of school-aged and adolescent children with Williams syndrome show both areas of strengths and areas of difficulty. While pockets of strength in pragmatics in Williams syndrome generally emerge from skills that are tied to emotional connectedness and story telling, difficulties are evident in conversation skills, likely relatable to limitations in cognition, social reciprocity, and maladaptive behaviors (Hepburn et al., under review; Semel & Rosner, 2003).

Some areas of pragmatic strength have been addressed with respect to Williams syndrome. Many children with Williams syndrome can introduce themselves in an appropriate manner, although they may act overly familiar to a virtual stranger (Semel & Rosner, 2003). Pragmatic aspects involving eye contact and using nonverbal gesturing to evidence interest in and maintain the interest of their conversational partners are also reported (Schreiber, 2000; Semel & Rosner, 2003). Further, strength is observed in reading the emotional expressions of others (Schreiber, 2000). With respect to conversation, while there is definite variation, many can make relevant comments to maintain a topic, as well as make conversational repairs (Semel & Rosner, 2003).

Notable among the strengths in pragmatic functioning in this population, individuals with Williams syndrome derive enjoyment from entertaining an audience (Schreiber, 2000). Children with Williams syndrome are able to re-tell stories from wordless picture stories demonstrating many of the major components of good story re-tell: coherence, structure, an orientation of the setting and characters, and a problem with a resolution (Bellugi, Wang, & Jernigan, 1994; Reilly, Klima, & Bellugi, 1990; Semel & Rosner, 2003). In a study of thirty 5-10-year-old children with Williams syndrome, matched by chronological age and gender to typical controls, the children with Williams syndrome significantly exceeded control children

in their use of elaboration of character's emotional states and social engagement devices (i.e., sound effects, phrases, and exclamations that capture the listener's attention) when re-telling a wordless picture book (i.e., "Frog, Where are You?"), despite significantly more structural language errors (Jones et al., 2000). However, children with Williams syndrome also used significantly fewer cognitive evaluation devices (e.g., inferences of causality, etc.) compared with typical controls (Jones et al., 2000), possibly reflecting findings from Theory-of-Mind tasks in Williams syndrome (Tager-Flusberg & Sullivan, 2000). Others have reported similar results in comparison to children with Down syndrome, emphasizing the tendency for children with Williams syndrome to excel at the expression of affect through changes in pitch, volume, and stress in story-telling, matching the level of expression seen in typical 9-10-year-olds when telling a story to pre-schoolers (Reilly et al., , 1990). These devices used by children with Williams syndrome serve to capture and delight the listening audience (Jones et al., 2000; Reilly et al., 1990).

Other parts of pragmatics present difficulty with respect to incessancy in Williams syndrome. Over-talkativeness, in general, including "cocktail party chatter" (i.e., superficial talking that lacks meaningful content) is problematic for children with Williams syndrome and is considered a hallmark of the syndrome (Semel & Rosner, 2003; Udwin & Yule, 1990; Udwin, Yule, & Martin, 1987). Contributing to the cocktail party effect of the chatter of individuals with Williams syndrome is an over-use of learned mature-sounding phrases (e.g., "Lo and Behold!"; Bellugi,et al., 1994; Schreiber, 2000; Udwin & Yule, 1990). Further, Semel & Rosner (2003) describe constant, inappropriate requests for attention and greeting behaviors. For example, children with Williams syndrome tend to enthusiastically greet just about everyone with whom they come into contact (Semel & Rosner, 2003). Asking the same question over and over again is frequently problematic for these children; however, this

behavior may be related to anxiety-invoking situations, such as changes in the routine or trying to understand a peer's social rejection (Schreiber, 2000; Semel & Rosner, 2003). Finally, indiscriminate flattery (e.g., complimenting another's clothing, etc.) has been described as a means for initiating and maintaining the attention of others in conversation (Levine & Wharton, 2000).

Children with Williams syndrome also tend to struggle with certain aspects of conversation. Limitations in conversational skills relate to difficulties with "giving up the floor," making irrelevant and tangential comments causing difficulties with topic maintenance, and perseveration or inappropriate and frequent mentioning of the same thoughts and ideas (Levine, 1993; Meyerson & Frank, 1987; Semel & Rosner, 2003; Udwin & Yule, 1998; Udwin & Yule, 1990). Further, despite use of nonverbal expressions and the ability to read the emotional expressions of others, understanding and interpreting the nonverbal communicative body language and gestures of others ironically presents challenges for many with Williams syndrome (Schreiber, 2000). This may be linked to difficulties that individuals with Williams syndrome experience in reasoning about other's beliefs about situations based upon their behaviors, without any difficulty in the interpretation of intentions and emotion from facial expressions and eye gaze (Tager-Flusberg & Sullivan, 2000).

Although empirical analyses have been limited, Stojanovik (2006) analyzed the conversational structure of five school-aged children with Williams syndrome by coding 100-150 utterances obtained from conversations generated by looking at pictures of familiar situations (i.e., a birthday party, vacation, and children playing together) , in comparison to typical development and children with Specific Language Impairment (SLI). Results suggested that the conversations of children with Williams syndrome were more immature

and inappropriate, in comparison with both other groups of children. Further, while children with SLI only demonstrated one area of pragmatic difficulty (with significantly more difficulties in structural language), the children with Williams syndrome displayed predominantly pragmatic language difficulties, tending to provide too little information for the conversational partner, to over-rely on the conversational partner's leads and contributions, and to demonstrate over-literal misinterpretations of the communication partner. However comparable performance between children with SLI and Williams syndrome were evidenced in topic maintenance, language structure, and turn-taking. As a function of comparison with SLI, the paper's author concluded that the pragmatic difficulties in Williams syndrome could not be attributed to structural language difficulties but rather to the impaired general cognitive abilities characteristic of the syndrome (Stojanovik, 2006).

Another recent study revealed a striking finding of pervasive impairment in the pragmatic language profiles of school-aged and adolescent children and adults with Williams syndrome. Laws & Bishop (2004) administered the Children's Communication Checklist (CCC) to the parents of 19 individuals with Williams syndrome between the ages of six and 22 and determined that these children, in fact, demonstrated significant impairment in pragmatic language functioning, relative to typical, Down syndrome, and structurally language disordered (i.e., Specific Language Impairment—SLI) controls. Further, they cited inappropriate initiation of conversation and the use of stereotyped language as two areas of pragmatic competence that were "particularly affected" in their sample. They concluded that the pragmatic impairment in Williams syndrome is significant enough to warrant interventions in pragmatics and social skills like those offered to children with autism (Laws & Bishop, 2004).

Based upon the Laws & Bishop (2004) study, the pragmatic language profiles of individuals with Williams syndrome, like the profile for individuals with autism, suggested relative difficulty with every pragmatic subdomain of the CCC when compared with typical individuals. This finding is particularly interesting given the differences in types of pragmatic difficulties demonstrated by each syndrome. Conversely, when compared to controls with Down syndrome, relative difficulties were noted in only two areas: Inappropriate Initiation and Stereotyped Language. While this study provided an initial specific look at the pragmatics language profiles of individuals with Williams syndrome, the study sample was relatively small and the age sample was highly variable, such that 4-year-olds and 22-year-olds were considered within the same study. Arguably, the differences, in terms of pragmatic environment and demands between early childhood, school-age, adolescence, and adulthood are quite significant, likely tempering the findings of this study. Additionally, use of a control group of children with only Down syndrome is limiting considering that individuals with Down syndrome have their own unique linguistic profile and have been suggested to evidence relative strength in their pragmatic language skills (Chapman & Hesketh, 2000; Johnston & Stansfield, 1997). Future studies should not only attempt to replicate pragmatic impairment in this population and include pair-wise comparisons with other populations, but consider examining groups of individuals with Williams syndrome who are within a similar age cohort and are thus functioning within more similar pragmatic language environments.

Based upon reviewing the pragmatic language characteristics of individuals with autism and Williams syndrome, a number of similarities and differences emerge. See Table 2 for a brief summary of these findings. Among the more salient distinctions in pragmatics between the two syndromes lies in the notion of asociability in autism and hypersociability in Williams syndrome. Impairment derives from both types of atypical personality with regard

Table 2

*Comparative Summary of Pragmatic Language Behaviors in Autism and Williams syndrome*

| *Pragmatic Behavior* | *Autism* | *Williams syndrome* |
|---|---|---|
| Unusual use of eye contact (Happe & Frith, 1996; Lord et al., 1994; Semel & Rosner, 2003) | + | - |
| Difficulty reading emotion in facial expression (Hobson, 1986; Ozonoff,, Pennington, & Rogers, 1990; Schreiber, 2000; Tager-Flusberg & Sullivan, 2000) | + | - |
| Difficulty understanding communicative, nonverbal gestures (Happe & Frith, 1996; Lord et al., 1994; MDOE, 1998; Schreiber, 2000) | + | + |
| A lack of use of communicative, nonverbal gestures (Happe & Frith, 1996; Schreiber, 2000) | + | -/+ |
| Difficulties with conversational repair (Semel & Rosner, 2003; Volden, 2004) | - | - |
| Lacking prosody (i.e., voice melody) (Baltaxe, 1977; Lord et al., 1994; Happe & Frith, 1996; Reilly et al., 1990; Semel & Rosner, 2003) | + | - exaggerated |
| Difficulties with topic maintenance (Dewey & Everard, 1974; Freeman & Dake, 1996; Schreiber, 2000; Semel &Rosner, 2003; Stojanovik, 2006) | + | -/+ |
| Over-talkativeness (Schreiber, 2000; Semel & Rosner, 2003) | - | + |
| Difficulties with conversational reciprocity (Dewey & Everard, 1974; Ozonoff & Miller, 1996; Screiber, 2000; Semel & Rosner, 2003) | + | + |
| Perseveration (Laws & Bishop, 2004; Lord et al., 1994; ; Screiber, 2000; Semel & Rosner, 2003) | + | + |
| Difficulties with topic coherence (Freeman & Dake, 1996; Laws & Bishop, 2004; Schreiber, 2000; Szatmari et al., 1989) | + | + |
| Use of tangential language (Dewey & Everard, 1974; Semel & Rosner, 2003) | + | + |

to pragmatic language functioning. Likely, pragmatic difficulties impact upon social functioning or else deviant social trajectories impact upon developing pragmatics. Children with autism show marked impairment at all levels of pragmatic language functioning, from body language through conversation and discourse, though pockets of skill may be observed in the ability to make a conversational repair and certain aspects of story-telling (Dawson et al., 2002; Dewey & Everard, 1974; Happe & Frith, 1996; Loveland & Tunali, 1993; Volden, 2004). Children with Williams syndrome, on the other hand, demonstrate patchy difficulties with conversation skills and a remarkable strength in discourse or story-telling abilities (Semel & Rosner, 2003). Uncovering this pattern of difficulties in Williams syndrome might suggest that many of their difficulties lie in the reciprocal, back-and-forth elements of pragmatics, including conversation. Adopting the Ninio & Snow (1996) model of developing pragmatics, while children with autism likely show difficulties at the first stage of developing pragmatics (i.e., understanding and use of rules that govern how communication behaviors are used effectively) which likely impacts upon all later stages of pragmatics development, children with Williams syndrome may be effective with this initial developmental pragmatics stage, but show difficulty at the next developmental stage, conversation. Thus, strength may be observable in story-telling where the child with Williams syndrome is the only person talking.

## Chapter III: Method

### *Design*

A quantitative methods approach was implemented to explore the research questions. Initially, reliability analyses were applied to understand how items on the CCC-2 were related into subscales within the sample. Next, the overall communication and pragmatic language summary variables of children with autism and Williams syndrome were compared, with an attempt to control for the effects of expressive language age. Further, it was determined whether or not a standardized assessment measure could accurately identify pragmatic language impairments in children diagnosed with either autism or Williams syndrome, who were likely evidencing difficulties in this area. Scaled Scores for the subdomains of the CCC-2 (i.e., pragmatic language profiles) were next compared for determining where group differences lie between autism and Williams syndrome in terms of specific subscales of pragmatic language functioning. Subsequently the individual questions comprising the subscales where group differences were found were compared between children with autism spectrum disorders and children with Williams syndrome. Finally, relationships between pragmatic language variables and adaptive socialization skills on the Vineland Adaptive Behavior Scales (1$^{rst}$ or 2$^{nd}$ edition) were explored.

### *Participants*

Participants in this study were recruited from two larger studies concerning the developing behavioral phenotypes of autism and Williams syndrome (NIH# U19 HD35468-

07and MOD# 12-FY03-47, respectively). All of the data for the autism and typical groups were collected, along with other measures not reported here, as a part of an ongoing longitudinal research project concerning the emerging behavioral phenotype of autism under the procedures outlined by that grant (NIH# U19 HD35468-07). The remaining participants used in those two groups had already been recruited and data were collected within the following six months, according to that study's timeframe. Data were already collected for 3 eligible participants with Williams syndrome who met criteria for this study under the procedures outlined by that grant (MOD# 12-FY03-47). Four additional participants in the Williams syndrome group were children who had been seen for the larger Williams syndrome study, but did not qualify for this study at the time that their initial data were collected. These families were re-contacted and participated in this study. Thus, these four families agreed to fill out some additional updated paperwork in order to participate in this study.

Additional recruitment of participants with Williams syndrome was necessary to obtain an adequate sample of individuals for this study. This included advertisement through mailings reaching educators, professionals, parents, and teachers of children with disabilities in the community, through recruitment in talking with regional Williams syndrome parent group representatives and asking for word-of-mouth referrals, as well as through advertisement on the Williams Syndrome Association (WSA) website listserve, a national organization committed to the collection and dissemination of resources for families affected by Williams syndrome.

Fifty-six subjects were included in this study comprising three groups: Williams syndrome (WMS; $n$=21), Autism Spectrum Disorder (ASD; $n$ =22), and Typically-developing (TYP; $n$=13). An additional four parents of children with Williams syndrome were

interested in participating in this study; however, their child did not meet the minimum expressive language level on the Vineland Adaptive Behavior Scales-II (VABS-II) and could not be enrolled. Data were also collected on 6 additional TYP participants who were removed from the sample as a function of being above the VIQ cutoff score of 120. The addition of the TYP group of children assisted in establishing the validity of this tool in a group of school-aged American children, such that children with typical development should not be showing any pragmatic language difficulties on the measure that was used to compare and determine whether or not children with Williams syndrome and autism demonstrate pragmatic language impairment.

Inclusion criteria for this study were as follows:

1. Previous diagnosis of (a) Williams syndrome via chromosomal analysis or genetic testing as endorsed by a parent to be qualified into the WMS group or (b) a diagnosis on the autism spectrum (i.e., Autistic Disorder, Pervasive Developmental Disorder, Not Otherwise Specified, or Asperger's Syndrome ) via clinical opinion and endorsement of symptoms on the Autism Diagnostic Observation Schedule (ADOS) and Autism Diagnostic Interview-Revised (ADI-R) and/or Social Communication Questionnaire (SCQ) to be qualified into the ASD group or (c) absence of a delay on a standardized measure of cognitive functioning and no known current developmental diagnoses to be qualified into the TYP group.

2. An overall verbal IQ (i.e., VIQ) under 120 in the ASD group and an overall verbal IQ above 85 and below 120 in the TYP group.

3. Chronological age between the ages of 5 years; 11 months and 12 years; 11 months old.

4. Verbal fluency or the ability to speak in full sentences and score an age-equivalent of 3 years, 11 months or older on the Expressive subdomain of the VABS or VABS-II. The VABS-II was administered to all children who were enrolled into the study after 9/1/05.

5. Parents who are willing to complete and return a checklist concerning pragmatic language abilities about their child, as well as complete a 20-30-minute standardized VABS-II interview in person (if local) or over the phone.

See Table 3 for this study's participant characteristics. While all of the children in the ASD group received a diagnosis on the autism spectrum as a function of endorsement on the ADOS and ADI-R or SCQ, as well as by the clinical opinion of a psychologist with a specialty in autism spectrum disorders, specific participant DSM-IV diagnoses broke down as follows: 13 Autistic Disorder, 7 Pervasive Developmental Disorder- Not Otherwise Specified (PDD-NOS), and 2 Asperger's Disorder.

Gender data are presented by group in Table 3. While the WMS groups demonstrated relatively even gender distributions reflective of the larger population of individuals with Williams syndrome, the ASD group is over-represented by males. However, this is relatively consistent with the larger population of children with autism who show similar patterns of male over-representation (Volkmar et al., 2005). Conversely, the TYP group was over-represented by females despite relatively even gender distribution in the larger population.

### *Group Differences*

Using one-way analyses of variance (ANOVA) with the between groups factor diagnosis and the within groups factors CA and expressive language equivalent on the VABS/II, mean group comparisons were analyzed. When differences were detected post-hoc analyses, Tukey's test, was run, unless the homogeneity of variance assumption was violated, in which

Table 3
*Participant Characteristics*

| | Autism Spectrum Disorder (ASD) | Williams Syndrome (WMS) | Typical (TYP) |
|---|---|---|---|
| *N*=56 | *n* = 22 | *n* = 21 | *n* = 13 |
| Diagnostic status | 100% endorsement of an autism spectrum diagnosis on both ADOS and ADI-R or SCQ and by expert clinical opinion | 100% parent endorsement on Medical Records Checklist | N/A |
| Gender<br>N (%Male) | 19 (86%) | 11 (52%) | 4(30%) |
| CA (M[SD])<br>in yrs., mos. | 9,7 (1,5) | 9,1 (2,1) | 6, 9* (0, 8) |
| Range | 7,0-12,9 | 6,2-12,4 | 5,11-8,4 |
| Expressive Language Age Equivalent on the VABS/II<br>CA(M[SD] )<br>in yrs., mos. | 6, 8** (1,3) | *n* = 20<br>5,6** (1,0) | 8,7** (1,6) |
| Range | 4,4-8,9 | 3,11-7,4 | 5,6-11,3 |
| VIQ (ASD and TYP groups only) | | | |
| M (SD)<br>Range | *n*= 21<br>88.67 (19.1)***<br>53-118 | Not collected | 107.3 (9.7)<br>89-126 |

*Note.* VABS/II data not available for one child with WMS. VIQ data were not available for one child with ASD. * indicates a significant difference: TYP<WMS=ASD. ** indicates significant between group differences: TYP>ASD>WMS. *** indicates significant difference: ASD<TYP.

case the Games-Howell post-hoc procedure was alternately chosen (Field, 2000). Given the communication impairments associated with both ASD and WMS, it was not possible to find a typically developing group of children matched on both CA and expressive language age equivalent on the VABS/II to the clinical groups. Thus, children with ASD and WMS were equated on CA, though the TYP group was significantly younger $F(2,53) = 12.4, p < .001$. However the TYP group was equated on expressive language age equivalent to the CA of both WMS and ASD groups, $F(2,53) = 1.2, p = .32$. Further, while the CA of the TYP group matched the expressive language age equivalent of the ASD group, the WMS expressive language age equivalent was significantly younger than that for the ASD group or when compared to TYP CA, $F(2,52) = 7.6, p = .001$. Finally, the TYP group had a significantly higher VIQ than the ASD group, $F(1,32) = 5.7, p = .008$.

*Group Matching*

Chronological age (CA) was used for matching the clinical groups, and not VIQ or expressive language age equivalent on the VABS/II as a function of the following. Initially, the relationship between VIQ, expressive language age equivalent, and the outcome variables were explored. Pearson's correlations were run in order to determine whether or not the pragmatics variables used in this study (i.e., General Communication Composite and Social Interaction Deviance Composite) were associated with either the expressive language age equivalent or verbal intelligence (i.e., VIQ), as a means for justifying the lack of VIQ data in the WMS group and the variability within VIQ of the ASD group, in particular. See Table 4 for a correlation matrix of VIQ and outcome study variables in the ASD and TYP groups.

While no significant relationships were found between VIQ and either pragmatic outcome variable (i.e., General Communication Composite and Social Interaction Deviance Composite) in either group, a medium-sized trend was noted in the TYP group, such that as

VIQ gets larger, so does General Communication Composite (i.e., overall communication functioning). Additionally, a moderate negative association was noted in the TYP group such that as VIQ gets larger, the Social Interaction Deviance Composite tends to get smaller (i.e., more pragmatic language impairment), though not significant.

Table 4
*Correlation Matrix of VIQ and Expressive Language Age Equivalent on the VABS/II to Outcome Variables*

| | | General Communication Composite | Social Interaction Deviance Composite |
|---|---|---|---|
| ASD group | VIQ | -.03<br>$p = .92$ | -.01<br>$p = .98$ |
| $n$=21 | expressive language age equivalent | .00<br>$p = .99$ | .12<br>$p = .63$ |
| TYP group | VIQ | .33<br>$p = .27$ | -.39<br>$p = .19$ |
| $n$=13 | expressive language age equivalent | .27<br>$p = .38$ | -.70**<br>$p = .008$ |
| WMS group<br>$n$ = 20 | VIQ<br>expressive language age equivalent | NC<br>.16<br>$p = .51$ | NC<br>.09<br>$p = .72$ |
| Entire Sample | | | |
| $n$ = 55 | VIQ<br>expressive language age equivalent | NC<br>.58**<br>$p < .001$ | NC<br>-.07<br>$p = .60$ |

*Note.* Characteristics of the pragmatic outcome variable (i.e., Social Interaction Deviance Composite) can be found later in this paper in Table 19. NC = Not collected. VIQ data were not available for one child in the ASD group and expressive language age equivalent was not available for one child in the WMS group. ** indicates significance at the .01 level.

Next, expressive language age equivalent on the VABS/II was considered. While expressive language level on the VABS/II was important for determining that children in the

sample were using language at an age that was developmentally appropriate for the assessment of the pragmatics language skills being measured, the WMS sample was significantly younger than the ASD group on this variable despite equivalent chronological ages. Expressive language age equivalent was not related to either outcome variable in the clinical groups; however a large negative association was found in the TYP group, such that as expressive language age equivalent gets larger, the Social Interaction Deviance Composite values gets smaller. Additionally, there was a large positive association in the entire sample, such that as expressive language age equivalent gets larger, so does the General Communication Composite value. While this makes sense because both variables are measuring a type of communicative functioning, it also suggests that expressive language age equivalent will need to be controlled for during subsequent analyses (because the groups are not equated on this variable), where the General Communication Composite is the outcome variable.

*Procedures*

Data collected for the ASD and TYP groups, as well as three participants from the WMS group, were done so according to the procedures outlined by their respective grants. Generally speaking, autism symptoms and cognitive measures were collected by experienced master's or doctoral level clinicians at the lab at the University of Colorado at Denver and Health Sciences Center. Usually, parents were administered the VABS/II by a second clinician in an adjacent room, while the child was participating in autism diagnostic, cognitive, or other measures not described here. CCC-2 protocols were mailed with instructions in a packet to parents, along with other measures not reported here, prior their child's first lab session. Parents typically returned the CCC-2, with the packet, by the end of their child's lab sessions.

In terms of new study recruits, once participants (parents of a child with Williams syndrome) were screened for eligibility over the phone and agreed to participate in the study, study consents were discussed. Enrolling participant's parents were mailed a CCC-2 and *Medical Records Checklist*, along with highlighted consent and HIPPA forms (i.e., federal forms disseminated to emphasize the importance of privacy and discretion by the researcher with the data collected) for signatures. The forms were then returned in a pre-addressed envelope. Instructions were discussed in the eligibility phonecall, as well as either written on each standard protocol or included as a part of an included letter, which also served to encourage additional phonecalls and/or e-mails to the investigator should any confusion persist around completion and return of the forms (i.e., CCC-2, consent and HIPPA forms, and *The Medical Records Checklist*). Additionally, an appointment either over the phone or in person to complete the VABS-II occurred between a parent and the investigator, typically before the packet was sent out.

*Measures*

The measures used for this study were either an inclusion measure or an outcome measure. Inclusion measures for this study included two diagnostic measures for autism symptoms for the ASD group only and one of three cognitive tests for the ASD and TYP groups only. The VABS/II served as an inclusion measure for all participants to qualify the language abilities of all of the children in this study to be sure that children were communicating at a level that was developmentally appropriate for assessing the pragmatics skills measured by the CCC-2. Outcome measures included the CCC-2, a parent-rated pragmatic language checklist, and the Socialization domain Standard Score of the VABS/II. All measures were administered in a standardized fashion by trained examiners. All autism diagnostic sessions were videotaped and fidelity of administration and scoring reliability was

established from the beginning of the study and maintained in at least 20% of measurements throughout the duration of the study. Further, all administrators of the autism diagnostic sessions, including this paper's author, were trained to 80% reliability on the ADOS by a certified ADOS trainer, either Dr. Sally Rogers or Dr. Susan Hepburn, both of whom had completed ADOS trainer workshops with the developer of the tool, Dr. Catherine Lord. See Table 5 for a list of the inclusion and outcome measures selected for this study.

*Inclusion Measures: Autism Spectrum Diagnosis*

*Autism Diagnostic Observation Schedule (ADOS; Lord, Rutter, DiLavore, & Risi, 2002).* The ADOS is a standardized, semi-structured interview that employs developmentally appropriate social and toy-based probes into a 30-45 minute interaction between an examiner and the child to elicit possible symptoms of autism in four areas: social interaction, communication, play, and repetitive behaviors. The ADOS comes in four modules with module selection dependent upon a child's language level. Module 3 ADOSes were used for this study because that is the module available for "verbally fluent" children, a criterion necessary for study participation. On the Module 3 ADOS, 28 behavioral dimensions (e.g., Conversation, Stereotyped Behaviors, Social Responsiveness, etc.) are scored on a 3-point scale, with higher scores indicating more evidence for abnormality of a particular behavior (i.e., 0 = no evidence of abnormality related to autism; 1= some evidence for autism-related abnormality; 2=definite evidence, etc.). A total symptom score from the algorithm of the ADOS was used for qualification purposes, with score totals above the "Autism spectrum cutoff" necessary for qualifying subjects into the autism spectrum disorder group. The reliability and validity of the ADOS are generally strong (Lord, Rutter, DiLavore, & Risi, 2002).

Table 5
*Inclusion and Outcome Measures Used in This Study*

| *Construct measured: Measurement tool* | *Inclusion* | *Outcome* |
|---|---|---|
| Autism Diagnosis: ADOS and ADI-R or SCQ | ASD group only: Positive for autism spectrum diagnosis on both measures | |
| Cognition: WISC-IV, WASI or DAS | ASD and TYP groups only: Below 120 in verbal reasoning domain for ASD group, and above 85 and below 120 for the TYP group | |
| WMS diagnosis: *Medical Records Checklist* | WMS group only: Written parent endorsement of formal WMS diagnosis via FISH testing | |
| Language proxy and social skills: VABS/II | All groups: Expressive age equivalent at or above 3 years, 11 months | All groups: Socialization Standard Score of VABS/II |
| Pragmatic language impairment: CCC-2 | | All groups: General Communication Composite and Social Interaction Deviance Composite variables, mean Scaled Scores for each CCC-2 pragmatic subscale |

*Autism Diagnostic Interview – Revised (ADI-R; Lord, Rutter, & LeCouteur, 1994).* The ADI-R is a structured, standardized parent interview designed to consider the presence and severity of autism symptoms in children across all three main symptom areas involved in autism: social relatedness, communication, and repetitive behaviors. An algorithm has been established that differentiates autism from other developmental disorders. A total autism symptom

score from the algorithm of the ADI-R was used for qualification purposes, with scores above the "Autism spectrum cutoff" necessary for qualifying subjects into the autism spectrum disorder group. Reliability and validity data for this instrument are generally good to excellent (Lord, Rutter, & LeCouteur, 1994). Many participants have completed a full ADI-R through their participation in Dr. Hepburn's longitudinal study of behavior, funded by NICHD. Due to time and cost constraints, subjects recruited since 2004 have received a shortened version of this interview, the Social Communication Questionnaire. Dr. Hepburn's personal communication with Dr. Lord, developer of both tools, supported this decision.

*Social Communication Questionnaire (SCQ; Berument, Rutter, Lord, Pickles, & Bailey, 1999).* The SCQ, also sometimes called the Autism Screener Questionnaire (ASQ), is a 40-item parent interview for autism symptoms developed to be a parent report measure based on the ADI-R. The tool may be used with individuals of any age or IQ level. It has good discriminative validity (.88) for the separation of autism spectrum diagnoses (autism/Asperger/PDD) from other diagnoses and specificity (.75), although the tool may have some difficulty distinguishing between autism diagnoses (i.e., autism vs. PDD-NOS) and be considered conservative for not picking up some high-functioning individuals on the spectrum (Berument, et al., 1999).

*Inclusion Measures: Cognition to Confirm Inclusion for TYP and ASD Groups*

*Wechsler Intelligence Scales for Children-IV (WISC-IV; Wechsler, 2003 ).* The WISC-IV is a standardized test of intelligence for children ages 6-16. It examines both verbal and nonverbal intelligence performance and provides age equivalents and standard scores for each. An abbreviated version of the test can be given using four subscales which yield three scores: Verbal Composite Index (VCI), also referred to as VIQ for the purposes of this

study, Perceptual Composite Index (PCI), and Full Scale IQ (FSIQ). The Vocabulary and Similarities subtests make up the abbreviated VCI, while the Block Design and Matrices subtests make up the abbreviated PCI. The VCI is the variable of interest for this study to determine that subjects are within or above the average range in the TYP group (i.e., above 85) and below 120 in both the ASD and TYP groups. The WISC-IV generally demonstrates good psychometric properties and shows acceptable convergent reliability with both the WASI and DAS (Wechsler, 2003).

*Wechsler Abbreviated Scales of Intelligence (WASI; Wechsler, 2002).* The WASI is a standardized test of intelligence for children ages 6-16. For the purposes of this study, the verbal intelligence quotient (i.e., VIQ) was used to qualify participants and is made up of two subtests (i.e., Vocabulary and Similarities) and takes approximately 10-15 minutes to administer. The WASI shows acceptable convergent reliability with both the WISC-IV and DAS (Wechsler, 2002).

*The Differential Abilities Scale (DAS; Eliot, 1990).* The DAS is a standardized test of intelligence with versions suitable for children from ages 2 ½ to 18 years. For the purposes of this study, six core subtests of the School-Aged version were administered which yield a verbal performance, nonverbal performance, and spatial performance as well as an overall score termed the General Conceptual Ability (GCA). While subtests provide T scores ($M$ = 50, $SD$ = 10), the verbal, nonverbal, spatial, and GCA scores are reported as standard scores, with a mean of 100 and standard deviation of 15. Verbal performance (i.e., VIQ) is the variable of interest for this study to determine that subjects are within the average to above average range in the TYP group (i.e., above 85) and below 120 in the ASD and TYP groups. In terms of its psychometrics, the DAS manual reports good to excellent properties in terms

of both validity and reliability and shows acceptable convergent reliability with both the WISC-IV and WASI (Eliot, 1990).

*Inclusion Measures: Parental Confirmation of a Williams Syndrome Diagnosis*

*Medical Records Checklist (Fidler & John, 2002).* The *Medical Records Checklist* is a one-page form that takes about 5 minutes for a parent to complete. The form asks if and when their child received a formal diagnosis of Williams syndrome and includes a Y/N checklist of common, concomitant medical conditions (e.g., heart anomalies, dental abnormalities, etc.) that are sometimes associated with a diagnosis of Williams syndrome.

*Inclusion Measures: Expressive Language Level*

*Vineland Scales of Adaptive Behavior (VABS; Sparrow, Balla, & Cicchetti, 1984).* The VABS is a standardized 291-item parent interview that determines levels of adaptive behavior in four domains for children aged birth through six (i.e., Socialization, Communication, Daily Living Skills, and Motor Skills) and three domains for children aged seven through eighteen (i.e., Socialization, Communication, and Daily Living Skills). Each domain can be further divided into subdomains. The Communication domain can be further subdivided into the subdomains of expressive, receptive, and written, and the Daily Living Skills domain is further subdivided into the subdomains of interpersonal skills, domestic skills, and community skills. While the Socialization domain can be distilled into the subdomains of interpersonal relationships, play and leisure skills, and coping skills, the Motor domain is comprised of the subdomains of gross and fine. The VABS provides norm-referenced scores based on national standardization samples of 4,800 children developing typically and atypically. This measure provides a proxy for developmental functioning in expressive language (i.e., expressive language age equivalent), a qualifying measure for the purposes of this study. The VABS has good psychometric properties, has been widely used in research

contexts and shows good convergent validity with the VAB-II (Sparrow, Cicchetti, & Balla, 2005; Sparrow et al., 1984).

*Vineland Scales of Adaptive Behavior-II (VABS-II; Sparrow, Cicchetti, & Balla, 2005).* The VABS-II is a revised and updated version of the VABS, with an expanded age range and updated items to reflect cultural changes and increased research knowledge. The VABS-II is made up of the same domains and subdomains as the original VABS (Please see description above of the VABS). The VABS-II was standardized on a representative sample of over 3,000 individuals developing typically and atypically and shows a high degree of consistency with the VABS (Sparrow et al., 2005). Like its predecessor the VABS, the VABS-II demonstrates good psychometric properties.

*Outcome Measures*

*Children's Communication Checklist-2 (CCC-2; Bishop, 2003).* The CCC-2 was selected for the purpose of assessing pragmatic language skills in children for this study. According to the manual, this measure is appropriate for children who are between the ages of 4 and 16-years-old and speaking in full sentences (Bishop, 2003). The CCC-2 employs one of the three more typical approaches to the assessment of pragmatics: behavioral ratings made by someone who knows the child. This instrument was developed to evaluate behaviors that are not easily captured within traditional clinical language assessments and has been standardized on a small to moderate-sized sample of typically developing and language disordered English-speaking, school-aged, multi-word using British children (Bishop, 2003).

Two variables of interest for this study emerge from a completed CCC-2: (1) the General Communication Composite (i.e., GCC) and (2) the Social Interaction Deviance Composite (i.e., SIDC) as described by Bishop (2003). The General Communication Composite was designed to signal children with a communication impairment, such that any

number below a cutoff score (i.e., 55) is indicative of some type of a communication impairment (i.e., structural, pragmatic, or both). Additionally, the Social Interaction Deviance Composite variable may be used only *in conjunction* with children indicated by a low General Communication Composite score (i.e., under 55) to derive a qualitative variable of relative strength (in either structural or pragmatic aspects of language) intended to signal pragmatic language impairment from a structural language impairment.

The CCC-2 is a multiple-choice, 70-item checklist that takes approximately 10-20 minutes to complete by a rater (typically a parent, teacher, or therapist) who has been familiar with the child for at least 3 months (Bishop, 2003). It considers the social and communication characteristics of children, which can be totaled and converted to a Scaled Score for one of ten subscales: A-Speech (i.e., intelligibility); B-Semantics (i.e., word finding/vocabulary access); C-Syntax (i.e, grammar); D-Coherence (e.g., making sense in conversation through the proper referencing and sequencing of events); E-Inappropriate Initiation* (e.g., indiscriminate, talks too much, doesn't initiate topics about reciprocal interests, repetitive initiating); F-Stereotyped Language* (e.g., overuse of "learned chunks" in conversations; being "precise" in communications); G-Use of Context* (e.g., use and understanding of the social rules governing communication, including politeness, sarcasm, and humor; ability to correctly interpret others, including abstract language concepts); H-Nonverbal Communication* (e.g., understanding and using nonverbal conversational cues including both gestures and affect); I-Social Relations (i.e., regard for and relationships with peers); and J-Interests (i.e., restricted and/or repetitive interests and flexibility). Four of the subscales (indicated with '*') look specifically into pragmatic areas of language and comprise the pragmatic language domain, while four are considered under the rubric of structural language (i.e., Speech, Semantics, Syntax, and Coherence) comprising the structural language

domain. These two distinct linguistic domains provide for distinctions within language disordered children between structural language disorders (e.g., syntax, semantics, and phonology) and pragmatic language disorders (i.e., use). The last two subscales (i.e., Social Relations and Interests) were included in an effort to assist in distinguishing autism from pragmatic language disorder, such that children yielding additional difficulties in these subdomains may need to seek further evaluation for an autism spectrum disorder (Bishop, 2003). These two subscales comprise a third autism/social domain. These two subscales are also used instead of two pragmatic variables (G and H) in forming the Social Interaction Deviance Composite, the variable found to reliably distinguish between SLI and more pragmatic/social communication impairment in parent raters (Norbury et al., 2004). See Tables 6-15 for a list of the questions on the CCC-2 organized by table according to their corresponding subscale. Note that Tables 6-9 comprise the structural language domain subscales, while Tables 10-13 comprise the pragmatic language domain subscales. Tables 14-15 are made up of the questions that form the two subscales that comprise the autism/social domain.

While the Coherence subscale fell under the rubric of pragmatics in the original CCC and certainly contains questions abut behaviors that many would consider to be pragmatic language behaviors, validity analyses for the CCC-2 shifted this particular subscale into structural language as a result of its inability to differentiate children with Specific Language Impairment (SLI) from children with more extreme pragmatic language difficulties. Further, syntactic deficits potentially negatively impact upon one's coherence (Norbury et al., 2004). Additionally, questions relating to narrative abilities are included under the Coherence domain, likely because some findings have argued that narrative ability is more closely related

Table 6

*CCC-2 Questions Comprising the Speech Subscale*

| *Domain* | *CCC-2 Question* |
|---|---|
| A: Speech | |
| | 2. Simplifies words by leaving out some sounds, e.g., "crocodile" pronounced as "cockodile," or "stranger" as "staynger" |
| | Pronounces words in a babyish way, such as "chimbley" for "chimney" or "bokkle" for "bottle" |
| | Leaves off beginnings or ends of words, e.g. says "roe" instead of "road or "nana" instead of "banana" |
| | Makes mistakes in pronouncing long words; e.g. says "vegebable" rather than "vegetable" or "trellistope" rather than "telescope" |
| | Mispronounces "th" for "s" or "w" for "r". E.g. says "thoap" instead of "soap" or "wabbit" instead of "rabbit" |
| | Speaks clearly so that the words can easily be understood by someone who doesn't know him/her very well |
| | Speaks fluently and clearly, producing all speech sounds accurately and without hesitation |

Table 7

*CCC-2 Questions Comprising the Syntax Subscale*

| *Domain* | *CCC-2 Question* |
|---|---|
| B: Syntax | |
| | Gets mixed up between he and she so might say "he" when talking about a girl, or "she" when talking about a boy |
| | Gets mixed up between he/him or she/her, so might say "him is working" rather than "he is working", or "her have a cake" rather than "she has a cake" |
| | Produces utterances that sound babyish because they are just 2 or 3 words long, such as "me got ball" instead of "I've got a ball" or "give dolly" instead of "give me the dolly" |
| | Leaves off past tense –ed endings on words, so might say "John kick the ball" instead of John kick*ed* the ball", or "Sally play over there" instead of "Sally play*ed* over there" |
| | Leaves out "is" and so says "Daddy going to work" rather than "Daddy's going to work". Or might say "The boy big" rather than "the boy is big" |
| | Produces long and complicated sentences such as: "When we went to the park I had a go on the swings"; "I saw this man standing on the corner" |
| | Produces sentences containing "because" such as "John had a cake *because* it was his birthday" |

Table 8

*CCC-2 Questions Comprising the Semantics Subscale*

| *Domain* | *CCC-2 Question* |
|---|---|
| C: Semantics | |
| | Makes false starts and appears to grope for the right words; e.g., might say "can I- can I- can I- can-I have an- have an ice cream" |
| | Forgets words s/he knows—e.g. instead of "rhinoceros" may say, "you know, the animal with the horn on its nose..." |
| | Mixes up words of similar meaning. e.g., might say "dog" for "fox," or "screwdriver" for "hammer" |
| | Mixes up words that sound similar. e.g. might say "telephone" for "television" or "magician" for "musician" |
| | Is vague in choice of words, making it unclear what s/he is talking about, e.g. saying "that thing" rather than "kettle" |
| | Uses abstract words that refer to general concepts rather than something you can see—e.g. "knowledge", "politics", or "courage" |
| | Uses words that refer to whole classes of objects, rather than a specific item. E.g. refers to a table, chair and drawers as "furniture" or to apples, bananas and pears as "fruit" |

Table 9

*CCC-2 Questions Comprising the Coherence Subscale*

| *Domain* | *CCC-2 Question* |
|---|---|
| D: Coherence | |
| | Uses terms like "he" or "it" without making it clear what s/he is talking about. For instance, when talking about a file might say, "he was really great" without explaining who "he" is |
| | Can be hard to tell if s/he is talking about something real or make-believe |
| | Gets the sequence of events muddled up when trying to tell a story or describe a recent event. E.g. if describing a film, might talk about the end before the beginning |
| | Doesn't explain what s/he is talking about to someone who doesn't share his/her experiences; for instance might talk about "Johnny" with explaining who he is |
| | It is hard to make sense of what s/he is saying (even though the words are spoken clearly) |
| | Talks clearly about what s/he plans to do in the future (e.g. what s/he will do tomorrow, or plans for going on holiday) |
| | Explains a past event (e.g. what s/he did at school, or what happened at a football game) clearly |

Table 10

*CCC-2 Questions Comprising the Inappropriate Initiation Subscale*

| *Domain* | *CCC-2 Question* |
|---|---|
| E: Inappropriate Initiation | |
| | Talks repetitively about things that no-one is interested |
| | Talks to people too readily: e.g. without any encouragement, starts up a conversation with a stranger |
| | It's difficult to stop him/her from talking |
| | Tells people things they already know |
| | Asks a question, even though s/he has been given the answer |
| | Keeps quiet in situations where someone else is trying to talk or concentrate (e.g. when someone else is watching TV, or during formal occasions such as school assembly or a religious ceremony) |
| | Talks to others about their interests, rather than his/her own |

Table 11

*CCC-2 Questions Comprising the Stereotyped Language Subscale*

| *Domain* | *CCC-2 Question* |
|---|---|
| F: Stereotyped Language | |
| | Says things that s/he does not seem to fully understand (may appear to be repeating something s/he's heard an adult say). So, for instance, a 5-year-old may be heard to say of a teacher, "she's got a very good reputation" |
| | Uses favorite phrases, sentences or longer sequences in rather inappropriate contexts. E.g., might say "all of a sudden" rather than "then", as in "we went to the park and all of a sudden we had a picnic". Or might habitually start utterances with "by the way" |
| | Pronounces words in an over-precise manner: Accent may sound affected or "put on", as if child is mimicking a TV personality rather than talking like those around him/her |
| | Repeats back what others have just said. For instance, if you ask, "what did you eat?" might say, "what did I eat?" |
| | Includes over-precise information (e.g. exact date or time) in his/her talk, e.g. when asked "when did you go on holiday" may say "13th July 1995" rather than "in the summer." |
| | When answering a question, provides enough information without being over-precise |
| | You can have an enjoyable interesting conversation with him/her |

Table 12

*CCC-2 Questions Comprising the Use of Context Subscale*

| *Domain* | *CCC-2 Question* |
|---|---|
| G: Use of Context | |
| | Misses the point of jokes and puns (though may be amused by nonverbal humor such as slapstick) |
| | Gets confused when a word is used with a different meaning from usual: e.g. might fail to understand if an unfriendly person was described as "cold" (and would assume they were shivering!) |
| | Ability to communicate varies from situation to situation—e.g. may cope well when talking one-to-one with a familiar adult, but have difficulty expressing him/herself in a group of children |
| | Takes in just 1-2 words in a sentence, and so misinterprets what has been said. E.g. if someone says "I want to go skating next week", s/he may think they've been skating or want to go now |
| | Is over-literal, sometimes with (unintentionally) humorous results. E.g., a child who was asked "Do you find it hard to get up in the morning" replied "No. You just put one leg out of the bed and then the other and then stand up." Another child who was told "watch your hands" when using scissors, proceeded to stare at his/her fingers |
| | Appreciates the humor expressed by irony. Would be amused rather than confused if someone said "isn't it a lovely day!" when it is pouring rain |
| | Realizes the need to be polite—would pretend to be pleased if given a present s/he did not really like, and would avoid making personal comments about strangers |

Table13

*CCC-2 Questions Comprising the Nonverbal Communication Subscale*

| *Domain* | *CCC-2 Question* |
|---|---|
| H: Nonverbal Communication | |
| | Looks blank in a situation where most children would show a clear facial expression- e.g., when angry, fearful, or happy |
| | 14. Does not look at the person s/he is talking to |
| | Stands too close to other people when talking to them |
| | Ignores conversational overtures from others (e.g. if asked, "what are you making?" does not look up and just continues working) |
| | Fails to recognize when other people are upset or angry |
| | Makes good use of gestures to get his/her meaning across |
| | Smiles appropriately when talking to people |

Table 14

*CCC-2 Questions Comprising the Social Relations Subscale*

| *Domain* | *CCC-2 Question* |
|---|---|
| I: Social Relations | |
| | 3. Appears anxious in the company of other children |
| | 7. With familiar adults, seems inattentive, distant, or preoccupied |
| | 13. Is babied, teased, or bullied by other children |
| | 16. Is left out of joint activities by other children |
| | 33. Hurts or upsets other children without meaning to |
| | 57. Shows concern when other people are upset |
| | 67. Talks about his/her friends; shows interest in what they do or say |

Table 15

*CCC-2 Questions Comprising the Interests Subscale*

| *Domain* | *CCC-2 Question* |
|---|---|
| J: Interests | |
| | When given the opportunity to do what s/he likes, chooses the same favorite activity (e.g. playing a specific computer game) |
| | Talks about lists of things s/he has memorized e.g., the names of the capitals of the world, or the names of varieties of dinosaurs |
| | Moves the conversation to a favorite topic, even if others don't seem interested in it |
| | Shows interest in things or activities that most people would find unusual, such as traffic lights, washing machines, lamp posts |
| | Surprises people by his/her knowledge of unusual words- uses terms you'd expect to hear from an adult rather then a child |
| | Reacts positively when a new and unfamiliar activity is suggested |
| | Shows flexibility in adapting to unexpected situations: e.g. does not get upset if s/he planned to play on the computer, but has to do something else because it isn't working |

to core language abilities than to pragmatic language abilities (Norbury & Bishop, 2003). Assessment of narrative abilities on a formal assessment of narrative abilities, the Strong Narrative Assessment Procedure (SNAP; Strong, 1998), did not reliably separate children with autism spectrum disorders from typically developing children (Young et al., 2005).

The manual provides a technical description of the derivation of the two additional summary scores: the General Communication Composite (GCC) and the Social Interaction Deviance Composite (SIDC). The General Communication Composite derives from the first eight subscales of the CCC-2 (i.e., A through H) and serves to indicate children with

some kind of communication impairment (i.e., structural and/or pragmatic). The tool's validation data suggest the following with respect to General Communication Composite scores: virtually all children with either specific language impairment (SLI; i.e., structural language difficulties) or autism receive General Communication Composite scores at or below 55. Conversely, the Social Interaction Deviance Composite variable attempts to tease apart children with communication impairments from children who have additional autism/social/pragmatic difficulties by creating a value which reflects a relative performance between structural language functioning (i.e., sum of scales A-D) and autism/social/pragmatic variables (i.e., sum of scales E, H, I, and J). According to the manual, the Social Interaction Deviance Composite variable is only useful for children receiving General Communication Composite scores below 55—for more closely examining the children who have communication impairments according to the CCC-2. While a positive Social Interaction Deviance Composite value suggests relative difficulties with structural aspects of language compared to pragmatic and autism/social aspects of language (i.e., SLI—not a pragmatic language impairment), a negative Social Interaction Deviance Composite value suggests the opposite (i.e., a pragmatic language impairment and autism/social difficulties and not SLI). The Social Interaction Deviance Composite serves the function of improved discrimination between these two types of communication disordered children. Note that the Social Interaction Deviance Composite variable does not include the variables associated with two pragmatic subscales (i.e., F and G) because they did not successfully discriminate children with SLI likely due to the impact of structural language on these two particular pragmatic suscales (Norbury et al., 2004). The manual suggests that Social Interaction Deviance Composite scores of nine or more tend to occur in children with

SLI, while any negative value is virtually unseen in SLI though common to children with pragmatic language impairment, including children on the autism spectrum.

Each subscale on the CCC-2 consists of a total of seven items—five items that tap into communication deficits and two items concerned with strengths. The instrument asks for a frequency judgment of the occurring behavior for each item (i.e., 0=less than once a week or never; 1=at least once a week, but not every day; 2=once or twice a day; and 3=several times [more than twice] a day or always). Scaled Scores may be derived from each subscale's total raw score. Lower Scaled Scores (4 or lower, as determined by the manual from the conversion of raw scores) on a subscale indicates greater pragmatic impairment within that domain, while a Scaled Score of 6 or more indicates typical functioning. Thus, lower Scaled Scores are more indicative of pragmatic language impairment on this tool. The manual suggests that a Scaled Score of 5 should not be of concern if it is only presenting on "one or two scales. However, if more than three scales have scores this low, this suggests the child may warrant further investigation (p. 20; Bishop, 2003)." The instrument randomly intersperses questions from all of the subscales to avoid response biases (see Tables 6-15), however it is divided and grouped into questions probing for communicative difficulty followed by questions probing for communicative strength allowing for clearer instruction to the rater (Bishop, 2003). A consistency check can be calculated, however, for the rare instances (1.6% of respondents in the standardization sample) where the respondent is confused about changing their responses from communication difficulties to communication strengths. Instances of a failed consistency check result in invalid CCC-2 results and will be followed accordingly in this study.

With respect to reliability of the CCC-2, internal consistency and inter-rater reliabilities were available in the manual. Alpha values for internal consistency of the CCC-2

yielded a range from .65 to .80. Inter-rater reliabilities for each scale of the instrument were computed between parents and either teachers or speech-language pathologists with correlations ranging from .16 to .79 (Bishop, 2003). However, an alternative approach has been devised for the CCC-2 to improve subgroup discrimination through the creation of a variable called the Social Interaction Deviance Composite (SIDC), which is a composite variable comprised of the addition of the Scaled Scores of two pragmatic and two autism/social subscales minus the sum of the four structural language subscale Scaled Scores. Use of this variable significantly boosted the inter-rater reliability between parents and professionals to .79 (Bishop, 2003). What could distinguish children with structural language impairments from children with pragmatic language impairment was the relative poorness of ratings received on scales A through D (i.e., four structural aspects of language) relative to scales E, F, I and J. Scales F and G (i.e,. Stereotyped Language and Use of Context), the other two pragmatic subscales that do not comprise the SIDC variable, were not included in this discriminatory variable because children with SLI (i.e., a structural language disorder) also tended to receive low ratings from parents in these areas of pragmatic communication.

According to the manual, validity data were collected on this instrument to determine whether or not it can adequately distinguish between children who have a pragmatic language impairment and those who do not, within a sample of children with language disorders. Validation data for the CCC-2 were collected from three clinical samples: a) 74 children with a range of communicative problems who were a part of a larger research study; b) 26 children who were on the caseloads of Scottish speech-language pathologists and; c) 34 children who were referrals to an English developmental pediatric clinic. In all 3 samples, very little overlap was observed between children who were

described as having pragmatic language impairment and those who did not. In sample A, every subscale demonstrated a significant group effect ($p < .0001$) for the means obtained by the CCC-2 in an ANOVA, with Scheffe tests revealing a significant group effect ($p = .05$) between the control group and the clinical groups (Bishop, 2003). The data also suggested, however, that the CCC-2 may not be as good at distinguishing between subtypes of communication impairment, with relatively poor scores on the pragmatic composite by children with structural language impairments. Further, low inter-rater agreement between parents and professionals ($r = .237$) suggested that this instrument may not satisfactorily discriminate among subgroups of children with communication impairments when parents are the raters, simply based upon absolute levels of pragmatic impairment (Bishop, 2003). Thus, the aforementioned SIDC variable was devised.

*Outcome Measure: Adaptive Social Skills*

*VABS.* The Standard Score from the Socialization domain of the VABS ($M = 100$, $SD = 15$) is of interest in terms of its relationship to pragmatic impairment on the CCC-2 for the purposes of this study. See above for additional description of this measure under inclusion measures.

*VABS-II.* For all participants enrolled after 9/05, the updated Socialization domain Standard Score of the VABS-II was examined in relation to pragmatic impairment on the CCC-2.

## Chapter IV: Results

### *Preliminary Data Analysis*

#### *Internal Reliability of the CCC-2 within the Sample: Cronbach's Alpha*

Because the CCC-2 was standardized on a sample of British children, reliability was calculated within this sample of school-aged American children. Cronbach's alpha has been suggested to be the best methodological choice for determining internal consistency for tests that contain multiple choice Likert-type responses, like the CCC-2, which asks the parent to rate a frequency from 0 through 3 for their child on each behavior (Gliner & Morgan, 2000; Leech, Barrett, & Morgan, 2005). Prior to the analyses, reverse coding (i.e., 0's became 3's, 1's became 2's, etc.) had to be performed for the 2 items within each scale of the CCC-2 that asked the parents to rate communication strengths (as opposed to weaknesses), in order to correctly assess alpha. Appropriate levels of internal consistency were sought (i.e., above .7). See Table 16 for a summary of alphas for each of the ten CCC-2 scales for the entire sample combined. Because the variability of responses was limited for many of the questions within each individual group, use of the entire sample was warranted for this calculation.

The data shown in Table 16 present evidence that each of the ten CCC-2 scales have good (alpha > .6) internal consistency when parents were the raters for this sample of American, school-aged children, all large effects based upon Cohen (1988). While only two of the subscales had alphas just under .7 (i.e., .68 and .69), eight of the subscales demonstrated alphas above .7. Thus, all scales were retained for further analyses.

Table 16
*Internal Consistency of the CCC-2 Subscales Using Cronbach's Alpha*

| *CCC-2 Scale* | Entire sample (*N*=56) |
|---|---|
| A: Speech | .72 |
| B: Syntax | .68 |
| C: Semantics | .80 |
| D: Coherence | .83 |
| E: Inappropriate Initiation | .79 |
| F: Stereotyped Language | .69 |
| G: Use of Context | .85 |
| H: Nonverbal Communication | .79 |
| I: Social Relations | .78 |
| J: Interests | .84 |

*Intercorrelations of the CCC-2 Scales*

Pearson's correlations were run between the total raw scores of each scale of the CCC-2 in order to measure the degree of linear association between subscales of the CCC-2. See Table 17 for the intercorrelations of the ten CCC-2 scales for the entire sample.

The median correlations among all of the scales were .63, indicating that many of the scales were highly linearly associated with the others, which Cohen (1988) considers a large effect size. While a majority of the scales were strongly associated with one another, the subscales were not combined into fewer factors as a function of conceptual reasons. Overlap of all of these scales (except Speech—or articulation) theoretically aligns with models suggesting that not only do pragmatic language abilities rely upon intact

Table 17
*Intercorrelation Matrix of the 10 CCC-2 Scales (N = 61)*

| | Syn. | Sem. | Coh. | II | SL | UC | NC | SR | Int. |
|---|---|---|---|---|---|---|---|---|---|
| Speech | .58 | .52 | .42 | .50 | .47 | .52 | .37 | .41 | .44 |
| Syntax | - | .65 | .63 | .59 | .64 | .60 | .44 | .46 | .60 |
| Semantics | | - | .75 | .60 | .66 | .74 | .59 | .61 | .56 |
| Coherence | | | - | .69 | .76 | .78 | .76 | .69 | .62 |
| Inappropriate initiation | | | | - | .71 | .83 | .64 | .62 | .80 |
| Stereotyped language | | | | | - | .77 | .69 | .69 | .69 |
| Use of context | | | | | | - | .74 | .72 | .79 |
| ɔrbal ɔmmunication | | | | | | | - | .83 | .64 |
| Social relations | | | | | | | | - | .72 |
| Interests | | | | | | | | | - |

*Note.* All correlations are significant.

structural language skills, but moreover upon a larger knowledge base of the social context, specific communication contexts, an understanding of the communication partner and general world knowledge (Martin & McDonald, 2003). Based upon earlier work (Bishop, 2003; Norbury & Bishop, 2004), the provision of these ten scales will allow for a richer profile exploration of similarities and differences between the clinical groups. Additionally, the ten CCC-2 subscales theoretically measure different aspects of social and communication abilities.

*Outcome Variables of Interest*

On the CCC-2, Scaled Scores were computed either from the examiner's manual or from an Excel-based computational disk provided in the test kit for each of the ten subscales from the raw scores scored by a parent (Bishop, 2003). Additionally, the General Communication Composite and Social Interaction Deviance Composite variables were computed based upon the Scaled Scores obtained on each CCC-2 protocol. Finally, one additional summary variable was created: PRAG (i.e., sum of Scaled Scores for the four pragmatic subscales: E+F+G+H), as a measure of overall pragmatic language functioning. Exploration of PRAG demonstrated a strong relationship with expressive language age equivalent, $r(55) = .59, p < .001$, so subsequent analyses using PRAG (like General Communication Composite) will control for expressive language age equivalent, but no relationships to VIQ in ASD, $r(21) = -.01, p = .95$ or TYP, $r(13) = .24, p = .42$, by group, though a trend toward a small to medium-sized effect was noted such that as PRAG increases, VIQ increases in the TYP group. Recall that while there was an association between expressive language age equivalent and Social Interaction Deviance Composite in the TYP group only, the Social Interaction Deviance Composite variable is not a valid number for children receiving General Communication Composite scores above 55, for which all children in the TYP group did. The Social Interaction Deviance Composite variable for the TYP group will accordingly not be reported. Thus, given the lack of association seen in either clinical group between expressive language age equivalent and Social Interaction Deviance Composite, controlling for expressive language age on the VABS/II will not be necessary. All data were entered into SPSS as a person-level dataset (Singer & Willet, 2003).

## *Study Analysis*

### *Analysis of the CCC-2 Summary Variables*

Next, one-way analyses of co-variance (ANCOVAs) were conducted, with diagnosis as the between groups factor, communication outcomes (i.e., General Communication Composite and PRAG) as within groups factors, and expressive language age equivalent as the covariate, to look for mean differences between groups, controlling for expressive language level. ANCOVA affords the additional benefit of preserving power when the covariate is related to the dependent variable, which is important because of this study's relatively small sample sizes (Huck, 2004). Because there are three groups, when group differences were detected post-hoc analyses were conducted using Bonferroni's test.

Before reporting any of the analyses, the data were examined for linearity of the covariate, equality of slopes, and independence of the covariate to satisfy the necessary assumptions for ANCOVA; however both the General Communication Composite and PRAG variables failed the equality of slopes assumption with the following values for interaction variables (diagnosis x expressive language age), $F(2,52) = 54.2, p < .001$ and $F(2,52) = 61.8, p < .001$, respectively. Various transformations were performed for both the General Communication Composite and PRAG variables and the assumptions were re-tested; however at least one variable continued to violate this assumption.

However, when the TYP group was removed from the analysis (because the study's primary purpose was in comparing ASD and WMS), this assumption was no longer violated for either variable: General Communication Composite, $F(1, 40) = .34, p = .72$ and PRAG, $F(1, 40) = 3.2, p = .53$. Without the use of inferential statistics, the performance of the TYP group on the General Communication Composite and PRAG variables was a lot stronger

than for either clinical group, as expected: General Communication Composite, $M(13)$ = 87.6, $SD$ = 15.0 and PRAG $M(13)$ = 44.0, $SD$ = 8.1. See Table 18 for adjusted and unadjusted means for the General Communication Composite and PRAG variables in ASD and WMS. Every participant in the TYP group had a General Communication Composite above 55, and thus was not showing communication impairment on the CCC-2 which also incidentally invalidates the interpretation of the Social Interaction Deviance Composite variable in this group (Bishop, 2003). Further, mean PRAG scores all fell well above 24, which is the minimum cutoff for typical functioning considering it is the sum of four Scaled Scores, of which a score of 6 and above is typical functioning.

Table 18
*Adjusted and Unadjusted Means For the General Communication Composite and PRAG Covarying by Expressive Language Age Equivalent*

| | | **Unadjusted** | | **Adjusted** | |
|---|---|---|---|---|---|
| | | *M* | *SD* | *M* | *SD* |
| *WMS* | General Communication Composite | 31.8 | 12.2 | 32.6 | 2.4 |
| $n$ = 20 | PRAG | 15.7 | 5.1 | 16.9 | 1.4 |
| *ASD* | General Communication Composite | 29.1 | 14.5 | 28.7 | 2.9 |
| $n$ = 22 | PRAG | 12.7 | 6.5 | 12.1 | 1.2 |

*Note.* Recall that VABS/II data (i.e., expressive language age equivalent) were missing for one WMS participant.

There were no main effects for the General Communication Composite, $F(1, 40)$ = .337, $p$ = .72, by diagnostic group suggesting that there were no significant differences between overall communication abilities (i.e., General Communication Composite) on the

CCC-2 between ASD and WMS, when expressive language age on the VABS/II is held constant. Results using the PRAG variable approached significance, PRAG, $F(1, 40) = 3.18$, $p = .053$. Thus, there was a trend toward significance with the PRAG variable suggesting slightly stronger pragmatic language performance in the WMS group compared to the ASD group, when expressive language age is held constant. Despite the lack of a statistically significant difference, this finding has substantive value, given limitations with power in this study. Adjusted mean PRAG scores demonstrated an additional 4.8 Scaled Score points over four combined pragmatic subscales of the CCC-2 for the WMS group compared to the ASD group. Stated differently, the children with WMS scored 20% better in their overall pragmatic language functioning than the children with ASD.

Next, one-way analysis of variance (ANOVA) was conducted between the WMS and ASD groups with diagnosis as the between groups factor and Social Interaction Deviance Composite as the within groups factor. Prior to the analysis, the data were screened for normality of distribution and outliers. There were no obvious outliers and both skewness and kurtosis tests suggested no significant departures from normality (with absolute values of less than 1). Additionally, Levene's test for the homogeneity of variance was not violated for the outcome variable. See Table 19 for the clinical group means of the Social Interaction Deviance Composite, another CCC-2 outcome variable.

Table 19
*Mean Scores by Group for CCC-2 Outcome Variables*

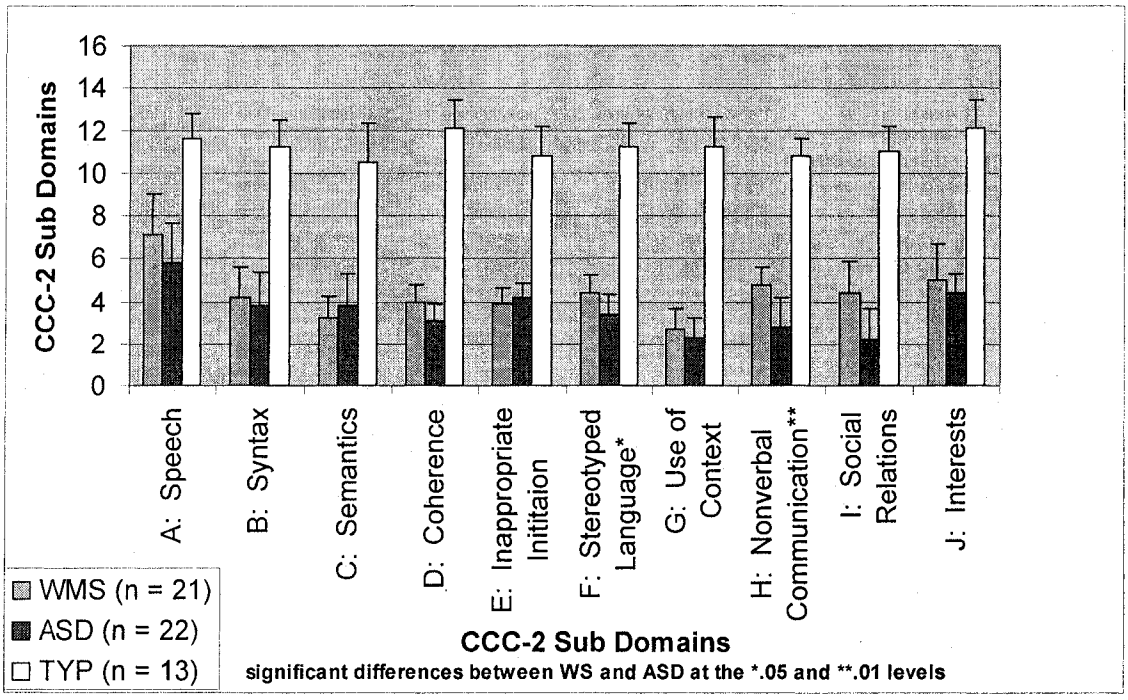
| | WMS ($n$=21) | ASD ($n$=22) |
|---|---|---|
| Social Interaction Deviance Composite M(SD) | -.57(8.0) | -2.8 (7.0) |

No mean group differences were found between WMS and ASD on the Social Interaction Deviance Composite variable, $F(1,41) = .97, p = .33$. Thus, the ASD and WMS groups evidence equivalent levels of pragmatic language impairment according to the Social Interaction Deviance Composite variable of the CCC-2.

*Comparison of ASD and WMS on Subscale Scores by CCC-2 Profile.*

Comparison of the pragmatic language profiles of the study participants by group were examined next. See Figure 1 for the mean Scaled Scores and standard deviations for each of the ten CCC-2 subscales by study group.

*Figure 1.* Scaled Scores of the 10 Subscales of the CCC-2 by Diagnostic Group




Next, an ANCOVA was run to compare the ASD and WMS groups on each of the subscale Scaled Scores of the CCC-2, to determine whether or not there were any significant differences between the two groups on any of the CCC-2 subscales, controlling for expressive language age on the VABS/II. See Table 20 for the adjusted means and *F*-values

associated with each subscale group mean comparison. While there were no significant differences on the majority of subcscales, significant differences were found between WMS

Table 20
*Adjusted Group Means and F-values Associated with CCC-2 Subscales for the WMS and ASD Groups, Controlling for Expressive Language Age on the VABS/II*

| CCC-2 Subscale | Adjusted WMS group mean | Adjusted ASD group mean | *F*-value |
|---|---|---|---|
| A: Speech | 7.7 | 5.5 | 1.4<br>$p = .26$ |
| B: Syntax | 4.0 | 3.8 | .11<br>$p = .90$ |
| C: Semantics | 3.3 | 3.6 | .38<br>$p = .69$ |
| D: Coherence | 4.1 | 3.1 | 2.0<br>$p = .16$ |
| E: Inappropriate Initiation | 3.9 | 4.1 | .67<br>$p = .52$ |
| F: Stereotyped Language | 4.8 | 3.2 | 4.6*<br>$p = .02$ |
| G: Use of Context | 2.9 | 2.2 | .62<br>$p = .54$ |
| H: Nonverbal | 5.2 | 2.6 | 5.9*<br>$p = .01$ |
| I: Social Relations | 4.6 | 2.1 | 3.1<br>$p = .06$ |
| J: Interests | 5.4 | 4.1 | 1.3<br>$p = .27$ |

*Note.* * indicates a significant between-group difference.

and ASD on two CCC-2 subscales: 1) Stereotyped Language $F(1, 40) = 4.6, p = .02$, and 2) Nonverbal Communication, $F(1, 40) = 5.9, p = .01$. Additionally, the Social Relations subscale approached a significant difference for both groups, $F(1, 40) = 3.1, p = .06$, likely

not reaching significance due to limited power and increased variability within this subscale. On all three of the aforementioned subscales, the WMS group outperformed the ASD group.

*Specific Questions within the CCC-2 that Differentiate WMS and ASD*

Upon learning about the three subscales of the CCC-2 that differentiate WMS from ASD, the responses from individual questions of the CCC-2 comprising those three subscales were compared for mean group difference using the Mann-Whitney *U* Test because raw scores from the CCC-2 are rank ordered (Gliner & Morgan, 2000). Nine questions emerged as being rated significantly different between WMS and ASD, while two questions approached significance. Five of the questions partially comprise the Nonverbal Communication subscale, four questions partially comprise the Social Relations subscale, and two questions partially comprise the Stereotyped Language subscale. See Table 21 for a summary of the eleven questions from the CCC-2 that either significantly differentiates WMS from ASD or approach a significant differentiation between the two groups. Of interest, the questions that emerged as rated significantly different by parents of children with autism spectrum disorders and parents of children with Williams syndrome within the three subscales where differences were detected primarily had to do with showing empathy, demonstrating interest in others, and seeming natural and comfortable in interactions with others.

*Identification of Pragmatic Language Impairment in WMS and ASD*

Because the Social Interaction Deviance Composite variable mean for both the WMS and ASD groups were negative, several children in these groups were evidencing pragmatic impairment, according to the CCC-2, however not all children in either group were evidencing Social Interaction Deviance Composite values indicative of pragmatic

Table 21

*Table of CCC-2 Questions, the Corresponding Subscale, and Associated U-statistic Differentiating WMS from ASD*

| *CCC-2 Subscale* | *C-2 Question* | *Mann Whitney U-value* |
|---|---|---|
| F: Stereotyped Language | 3. Pronounces words in an over-precise manner: Accent may sound affected or "put on", as if child is mimicking a TV personality rather than talking like those around him/her | $U(1,43) = -2.3$* $p = .02$ ASD>WMS |
| | 2. You can have an enjoyable interesting conversation with him/her | $U(1,43) = -2.1$* $p = .04$ WMS>ASD |
| H: Nonverbal Communication | . Looks blank in a situation where most children would show a clear facial expression- e.g., when angry, fearful, or happy | $U(1,43) = -2.6$* $p = .01$ ASD>WMS |
| | 4. Does not look at the person s/he is talking to | $U(1,43) = -2.7$* $p = .01$ ASD>WMS |
| | 0. Stands too close to other people when talking to them | $U(1,43) = -2.5$* $p = .01$ WMS>ASD |
| | 9. Fails to recognize when other people are upset or angry | $U(1,43) = -3.9$* $p < .001$ ASD>WMS |
| | 6. Makes good use of gestures to get his/her meaning across | $U(1,43) = -3.9$* $p < .001$ WMS>ASD |
| I: Social Relations | . Appears anxious in the company of other children | $U(1,43) = -2.3$* $p = .02$ ASD>WMS |
| | . With familiar adults, seems inattentive, distant, or preoccupied | $U(1,43) = -1.9$ $p = .06$ ASD>WMS |
| | 7. Shows concern when other people are upset | $U(1,43) = -2.6$* $p = .01$ WMS>ASD |
| | 7. Talks about his/her friends; shows interest in what they do or say | $U(1,43) = -1.8$ $p = .08$ WMS>ASD |

*Note.* * indicates statistical significance.

language impairment, as evidenced by the variability of this variable and proximity to zero. See Table 22 for a summary of cases in both the WMS and ASD groups who were and were

Table 22
*Summary of Cases in the WMS and ASD Groups Receiving Positive Versus Negative Values on the Social Interaction Deviance Composite Variable*

| *Valence of Social Interaction Deviance Composite score* | WMS ($n$=21) | ASD ($n$=22) |
|---|---|---|
| Negative (-) | 9 | 13 |
| Positive (+) | 9 | 6 |

*Note.* 3 children in the WMS group and 3 children in ASD group received scores of 0 on the Social Interaction Deviance Composite variable.

not showing a pragmatic language impairment, according to the CCC-2, using the variable suggested by the manual to be able to identify this type of an impairment (i.e., Social Interaction Deviance Composite), as rated by their parents. While positive values suggest structural language impairment, negative values are indicative of pragmatic language impairment (Bishop, 2003). Therefore, while the CCC-2 picked up pragmatic language impairment in some children with ASD and some children with WMS, many children were not identified as having a pragmatic language impairment using the Social Interaction Deviance Composite variable (i.e., 9 children in WMS group and 6 children in ASD group). Further, 3 children in each clinical group scored 0's on this variable, leaving an ambiguous interpretation for these participants.

Consideration of the PRAG variable (i.e., the sum of the four pragmatic subscales of the CCC-2) provided yet another chance to look at whether or not the CCC-2 was identifying pragmatic language difficulties in this sample of children. Because a score of 4 or less indicates impairment on any individual subscale of the CCC-2, a total score of 16 or less would indicate children with significant pragmatic language difficulties in the entire domain, since the domain is made up of four scales. Conversely, because a score of 6 or better indicates typical functioning on individual CCC-2 scales, a total score of 24 or more would indicate children with typical pragmatic language functioning. Scores occurring in between

impaired and normal (i.e., 17-23) will be categorized as "Borderline," for the purposes of these data. See Table 23 for a summary of pragmatic language impairment in the WMS and ASD groups as indicated by the PRAG variable.

Table 23
*Level of Functioning According to the PRAG Variable of the CCC-2*

| *Level of functioning* | WMS (*n*=21) | ASD (*n*=22) |
|---|---|---|
| Typical (24+) | 2 | 1 |
| Borderline (17-23) | 6 | 3 |
| Impaired (16 or less) | 13 | 18 |

Using the PRAG variable, a majority of children with both the WMS (i.e., 90%) and ASD (95%) demonstrated some difficulty with pragmatic language functioning, according to their parents on the CCC-2. While most children with ASD were rated as impaired on this variable, only 3 children demonstrated milder difficulties (i.e., borderline) and only 1 child was considered to behave typically within the domain of pragmatic language functioning. Conversely, more children with WMS (than ASD) fell into the borderline category of impairment (i.e., 6) and 2 children were considered typical within the pragmatic language domain of functioning. Nonetheless, more than half of children with WMS (i.e., 13) demonstrated significant difficulties with their pragmatic language functioning according to their parents on the PRAG variable of the CCC-2.

*Relationship of Pragmatic Language Variables to Other Social Variables.*

The relationships between the pragmatic language variables obtained using the CCC-2 (i.e., Social Interaction Deviance Composite and PRAG) and another measures of social functioning (i.e., VABS/II Socialization Standard Score) were next explored. Note, however, that the Social Interaction Deviance Composite values were not considered for the TYP group as previously described. See Table 24 for a summary of the relationships

Table 24

*Pearson's Correlation Matrix of Pragmatic Language Variables and Another Variables Measuring Social Functioning by Group and by Entire Sample*

| | | VABS/II Socialization Standard Score |
|---|---|---|
| ASD<br><br>$n = 22$ | Social Interaction Deviance Composite | -.02<br>$p = .92$ |
| | PRAG | .48*<br>$p = .02$ |
| WMS<br><br>$n = 20$ | Social Interaction Deviance Composite | .43<br>$p = .06$ |
| | PRAG | .51*<br>$p = .02$ |
| TYP<br><br>$n = 13$ | Social Interaction Deviance Composite | Not Valid |
| | PRAG | .81*<br>$p = .001$ |
| WMS and ASD groups only<br>$N = 42$ | Social Interaction Deviance Composite | .20<br>$p = .21$ |
| Entire sample<br>$N = 55$ | PRAG | .86**<br>$p < .001$ |

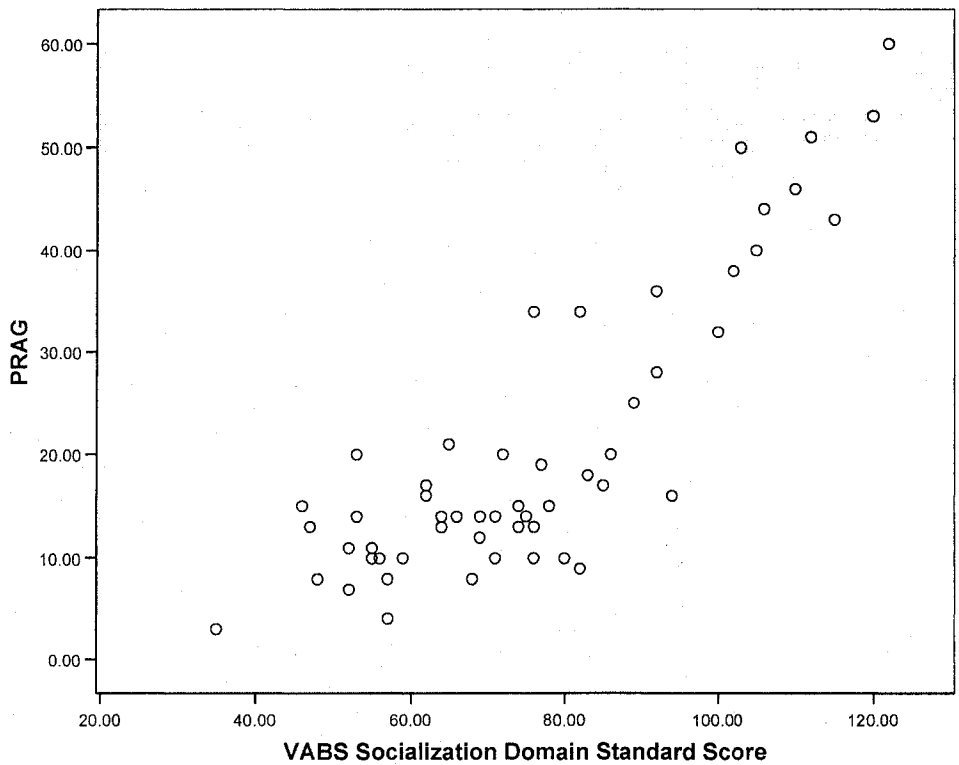
*Note.* VABS/II data not available for one participant in the WMS group. * indicates significance at the .05 level and ** indicates significance at the .001 level.

between pragmatic language outcome variables and another measure of social functioning by group, as well as using the entire sample.

While no significant associations were detected between the Social Interaction Deviance Composite variable and the other social variable (i.e., Socializations Standard Score on the VABS/II ) in the ASD group or in the ASD and WMS groups combined, a trend toward a moderate relationship was noted in the WMS group. Essentially, as the Social Interaction

Deviance Composite values get larger (i.e., less pragmatic language impairment) in the WMS group, so does the VABS/II Socialization Standard Score (i.e., better adaptive socialization). In the combined WMS and ASD groups, the trend of this relationship was small. The PRAG variable, on the other hand, showed a consistent significant relationship to the VABS/II Socialization Standard Score within each group, as well as within the entire sample. While medium to large effects were seen for the clinical groups, a very large effect was noted in the TYP group, as well as in the entire sample (Cohen, 1988). See Figure 2 for a scatterplot view of this relationship in the entire sample. Additionally, this strong relationship remained, even after controlling for expressive language age on the VABS/II, $r(52) = .80$, $p < .001$.

*Figure 2.* Scatterplot View of PRAG Plotted Against the VABS/II Socialization Domain Standard Score $r(55)=.86, p < .001$



## *Summary* of *Results*

The ten subscales of the CCC-2 showed good internal consistency in this entire sample of school-aged American children. Although the scales were highly associated with one another, they were maintained in the same subscales recommended by the tool's author (Bishop, 2003), in order to be able to more carefully compare the profiles of ASD and WMS. Both clinical groups showed significant impairment on the General Communication Composite (i.e., overall communication abilities), the Social Interaction Deviance Composite (i.e., overall pragmatic impairment), and the sum of the four pragmatic subscale Scaled Scores (i.e., PRAG—overall pragmatic language functioning) and were indistinguishable from each other on these dimensions on the CCC-2. However, the WMS group showed a trend of slightly better overall pragmatic functioning, compared to the ASD group, even when controlling for expressive age equivalent. The TYP group was significantly stronger on every variable, demonstrating typical communication and pragmatic language functioning, according to the tool.

Examination of the profiles between ASD and WMS revealed equivalent performances on 8/10 total subscales of the CCC-2, however significant differences of better performance in the WMS group (than ASD) were found on two pragmatic subscales: Stereotyped Language and Nonverbal Communication. Additionally, a trend toward a stronger performance in WMS than ASD was noted on one of the autism/social subscales, Social Relations. Ultimately, eleven questions, in particular, from the three differential subscales of the CCC-2 were rated as significantly different in WMS and ASD.

Additionally, while overall means between the clinical groups suggested impairment, some, but not all children with WMS and ASD were identified as having a pragmatic language impairment using the CCC-2. Nevertheless, derivation of another variable

indicative of overall pragmatic language functioning (i.e., PRAG) suggested that almost all children in both groups evidence some degree of difficulty with these behaviors. Yet, more children with ASD evidenced significant impairments while many children with WMS were evidencing milder (i.e., borderline) impairments.

Finally, overall pragmatic language functioning (and not pragmatic language impairment) was strongly associated with the adaptive Socialization Standard Score on the VABS/II, providing evidence for convergent validity of the CCC-2 with other tools measuring a similar construct. While a trend toward a positive relationship between the Social Interaction Deviance Composite variable (higher scores indicate less pragmatic impairment) and the Socialization Standard Score on the VABS/II was notable, the lack of an association between the variable suggested to serve as the strongest for use of parent raters of the tool (i.e., Social Interaction Deviance Composite) with adaptive socialization in the ASD group was also notable.

## Chapter V: Discussion

This study examined whether or not a standardized tool of pragmatic language abilities could detect impairment in school-aged children with autism and Williams syndrome. This study also attempted to determine what, if any, differences exist in the pragmatic language profiles of children with autism and Williams syndrome. Further, the study attempted to determine whether or not pragmatic language impairment, as rated by parents, and measured by a standardized tool, was related to other measures of social functioning. Fifty-six school-aged (ages 6-12-years-old) children with an autism spectrum disorder ($n$=22), Williams syndrome ($n$=21), and typically developing ($n$=13) were rated by their parents on a standardized measure for the assessment of pragmatic language functioning, the Children's Communication Checklist-2 (CCC-2; Bishop, 2003). When parents were the raters, American children with autism and Williams syndrome demonstrated impairment, while younger typically developing children did not, on a standardized assessment tool for pragmatic language functioning. Specifically, the CCC-2 distinguished school-aged typical children, who evidenced typical functioning, from school-aged children with autism and Williams syndrome, who demonstrated impairment on global measures of communication and pragmatic language functioning controlling for overall expressive language age equivalent. Additionally, this study yielded results to suggest that children with Williams syndrome may be slightly better at pragmatic language than children with autism spectrum disorders, according to their parents, when expressive language age is held constant. While children with autism and Williams syndrome have previously been

likened to one another in terms pragmatic language abilities (Laws & Bishop, 2004), this is the first empirical substantiation comparing pragmatic language functioning between these two populations. Additionally, while earlier studies have suggested pragmatic language impairment in Williams syndrome (Gillberg & Rasmussen, 1994; Laws & Bishop, 2004; Stojanovik, 2006), this study was the first to specifically examine this behavior in a group of school-aged children.

Reliability of the CCC-2 scales in this sample was good and consistent with previous use of the CCC and CCC-2 in children with autism and Williams syndrome, including use of parents as raters (Bishop, 1998; Bishop & Baird, 2001; Laws & Bishop, 2004; Norbury et al., 2004). Using the entire sample, high levels of internal consistency were found for all of the test questions when they were analyzed within the same subscales as suggested by the tool's manual (Bishop, 2003). Abbeduto and colleagues (1989) suggested "we know of no evidence indicating that the culture differences between Great Britain and the United States would differentially affect the course of child *language* development (p. 538; Abbeduto, Furman, & Davies, 1989)," which is further reflected by use of the CCC-2 in this sample, a British language assessment tool being rated by parents of American children.

While performance in some areas of social-pragmatic functioning suggested commensurate impairment in autism and Williams syndrome, closer inspection yielded differences in a few specific areas of pragmatic and social functioning. Specifically, children with Williams syndrome outperformed children with autism on two pragmatics subscales of the CCC-2, Stereotyped Language and Nonverbal Communication. Additionally, a trend toward stronger performance for children with Williams syndrome on the Social Relations subscale was notable. Further, while two of these subscales factor into the pragmatics domain, the third scale factors into the autism/social domain (Bishop, 2003), necessarily

employing a broader definition of pragmatic language functioning (Adams, Baxendale, Lloyd, & Aldredge, 2005; Martin & McDonald, 2003). When pragmatics are viewed as a wide grouping of communication behaviors encompassing social, emotional, and communicative aspects of social interaction (Adams et al., 2005; Bishop, 2003; Martin & McDonald, 2003), these three areas of pragmatic behavior potentially distinguish a school-aged child with Williams syndrome from a school-aged child with autism with respect to pragmatic language functioning.

This finding was particularly interesting with respect to Table 2, which presented a literature review-based comparison of the similarities and differences in pragmatic language functioning between individuals with autism and individuals with Williams syndrome. While a number of areas evidenced similarity between the two disorders (i.e., difficulties with conversational reciprocity, perseveration, topic coherence, and use of tangential language), other areas evidenced differences (i.e., use of eye contact, talkativeness, lacking prosody, reading emotions in others, and use of conventional gestures). See Table 2 for complete list and references. While differences in Nonverbal Communication and Social Relations between the two groups seem to map nicely onto the differentiating behaviors of autism spectrum disorders and Williams syndrome observed in Table 2, the finding of a significant difference in Stereotyped Language served as a bit of a surprise because children with both disorders have difficulties with perseveration, potentially suggesting similar difficulties for both groups of children in this pragmatic language area.

Inspection of all of the individual questions that differentiate children with Williams syndrome from children with autism is insightful with regard to how pragmatic language functioning in Williams syndrome is *not* like autism (see Table 21), while inspection of the other questions (see Tables 6-15) may be more informative as to how Williams syndrome is

like autism (Laws & Bishop, 2004). Accordingly, the broader CCC-2 autism/social subscales where children with autism and children with Williams syndrome were indistinguishable were Inappropriate Initiation, Use of Context, and Interests. Within the subscales where difficulties were found (i.e., Stereotyped Language, Nonverbal Communication, and Social Relations), while more than half of the seven questions in each of the Nonverbal Communication and Social Relations subscales differentiated the two subgroups, only two questions in the Stereotyped Language subscale served such a function. Closer inspection of where the differences lie within the Stereotyped Language subscale between the two groups suggested differences on two questions relating to prosody and the ability to have an "interesting" conversation with others. Within the Stereotyped Language subscale, it was these two questions, in particular, that most significantly influenced the overall improved performance of the children with Williams syndrome compared to the children with an autism spectrum disorder. Because these two behaviors were classified into the Stereotyped Language subscale on the CCC-2, parents rated children with Williams Syndrome as significantly better than autism spectrum disorders in this area.

Consistent with the behavioral phenotypes associated with both disorders, children with autism demonstrated impairment across all domains of social-pragmatic functioning, while children with Williams syndrome demonstrated a few areas of relative strength compared with autism. Essentially, parents rated their children with Williams syndrome as significantly better than children with autism on items asking about prosody, being able to have an interesting, enjoyable conversation, use and understanding of affective expressions, social responsiveness and empathy to others, and social relatedness with others. This finding is consistent with findings in younger pre-school aged children with Williams syndrome who demonstrate relative strength in emotional responsivity, including increased

mimicry and affect imitation, compared to a group of mental-age matched children with other developmental delays (Fidler, Hepburn, Most, Philofsky, & Rogers, under review).

Additionally, these particular skills are consistent with a perception of "hypersociability" in Williams syndrome, yet leaves room for impairment in overall pragmatic language functioning in William syndrome as a function of the many other behaviors necessary for comprising typical pragmatic language functioning. Similarly impaired functioning for both autism and Williams syndrome were detected in the quality of initiations with others, use and understanding of humor, including abstract language concepts; and in the overall quality and variety of the child's interests. Difficulties in these additional areas could account for the reason that children with Williams syndrome have difficulty with friendships (Tager-Flusberg & Sullivan, 2000). Likely a large part of developing friendships in childhood is mediated through pragmatic language, including developing a wide range of interests and conversation skills in order to relate to and demonstrate interest in other children. Social-pragmatic language skills provide a critical foundation for developing friendships. Despite a clear interest in having friendships, children with Williams syndrome lack some of the necessary pragmatic language tools necessary for developing and nurturing them.

The origins of differential pragmatic deficit in autism and Williams syndrome may present early, when dyadic (i.e., face-to-face) and triadic (i.e., face-to-object-to-face) social relations would be expected to develop according to continuity theories of typical development (Celani, 2004; Hepburn et al., under review; Hobson, 1993; Rogers & Pennington, 1991). Relative strength and intensity of dyadic (i.e., face-to-face) interactions have been reported in infants and toddlers with Williams syndrome (Fidler et al., under review; Hepburn et al., under review; Laing, Butterworth, & Ansari, 2002; Mervis et al.,

2003), which might theoretically result in the relative strength observed in children with Williams syndrome on most of the questions in the Nonverbal Communication and Social Relations subscales and a few of the questions in the Stereotyped Language subscale of the CCC-2. Yet, these children's relative difficulties to typical development in these social/pragmatic behaviors may be related to the intensity and unusual features that are equally salient in young children with Williams syndrome during these early dyadic interactions (Fidler et al., under review; Hepburn et al., under review; Mervis et al., 2003). Conversely, young children with autism have been noted to lack many forms of dyadic interaction (Celani, 2004; Hepburn et al., under review; Rogers & Pennington, 1991), corresponding with their relative weaknesses in Nonverbal Communication and Social Relations, as well as cascading deficits in all areas of pragmatics (Rogers & Pennington, 1991).

Difficulties with triadic interactions (i.e., face-to-object-to-face), including joint attention behaviors, have been reported in young children with both syndromes (Hepburn et al., under review; Laing et al., 2002; Wetherby, Prizant, & Hutchison, 1988), providing some interesting insight into possible origins for later difficulties with other pragmatic/social behaviors including Inappropriate Initiation, Use of Context, and Interests. It is certainly plausible that early limitations in focusing attention with others to outside entities (i.e., joint attention) could have an impact on the later development of a diversity of ways to initiate interactions with others; the understanding of humor and abstract language concepts, a higher-level, complex semantic skill involving shared cultural knowledge; as well as a diverse and dynamic set of interests. Such difficulties were equally apparent in school-aged children with autism and Williams syndrome.

Despite the tool's ability to detect pragmatic language impairment in both children with autism and Williams syndrome, not all children with either disorder were identified using the variable that the manual recommended for this purpose (i.e., Social Interaction Deviance Composite; Bishop, 2003). Thus, while this variable functioned appropriately as an overall means used for the purpose of determining group membership (i.e., impaired vs. not impaired), the clinical utility of this tool for the few children who were not identified is concerning. In fact, there may be some children with these disorders who do not evidence pragmatic language impairment. It certainly is possible that only some children with Williams syndrome evidence an impairment because we are only beginning to understand the strengths and difficulties in pragmatic language functioning in Williams syndrome. However, there is strong evidence that pragmatic language impairment is a hallmark feature of autism spectrum disorders (Dawson et al., 2003; Tager-Flusberg, Paul, & Lord, 2005; Young et al., 2005). Recall that data from the CCC-2 manual suggested that the CCC-2 may not be as good at distinguishing between subtypes of communication impairment (i.e., structural impairment vs. pragmatic impairment), since relatively poor scores were observed on the pragmatic composite by some children who also have structural language impairments (Bishop, 2003).

While understanding why the Social Interaction Deviance Composite variable did not function entirely as expected in this sample is only speculative, one consideration pertains to the structural language abilities of children in this sample. Many of the children in the autism sample were considered "high-functioning," given IQ's within the typical range, which also suggests typical structural language functioning in *some* children. However a significant minority could be expected to be showing a profile like that of Specific Language Impairment (SLI), with significant difficulties in structural language, in addition to

pragmatic impairment (Kjelgaard & Tager-Flusberg, 2001; Norbury et al., 2004). While structural language skills were not additionally measured for this study (beyond what is captured in the CCC-2), it is possible that this sample of children with autism spectrum disorders was heavily represented by children with the SLI profile, which might possibly confound the ability for the Social Interaction Deviance Composite variable to identify pragmatic language impairment in these children. Further, comparable structural language abilities have been reported for school-aged children with SLI and children with Williams syndrome (Stojanovik, 2006). Recall that the Social Interaction Deviance Composite variable is comprised of the difference of the sum of two pragmatic scaled scores and two social scaled scores minus the sum of the four structural language scores. If the summed structural language scaled scores are low, relative to the pragmatic/autism/social sum, then the Social Interaction Deviance Composite variable would tend to identify structural language impairment (rather than pragmatic language impairment). This possibility cannot be ruled out for some children with autism spectrum disorders, who also evidence an SLI-like structural language profiles (Kjelgaard & Tager-Flusberg, 2001), as well as children with Williams syndrome who may also demonstrate significant structural language difficulties (Semel & Rosner, 2003; Stojanovik, 2006).

Nonetheless, the addition of an easily calculable number assisted greatly in better understanding the overall pragmatic language functioning of these children. This new, critical variable was created by adding up the Scaled Scores of the four pragmatic language subscales of the CCC-2 (i.e., PRAG). However, given the inclusion of the Social Interaction Deviance Composite variable for boosting the reliability of parent raters (Bishop, 2003), the limitations of the use of the Social Interaction Deviance Composite in this study remains concerning in terms of clinical use.

As has been recommended by a recent ad-hoc committee put together by the American Speech-Language Hearing Association (ASHA) around the guidelines for best practice for children with autism spectrum disorders, the CCC-2 may provide one useful clinical tool, along with other forms of assessment (i.e., observation, direct testing, etc.), in determining overall pragmatic language functioning, as well as to monitor and identify treatment goals for children with autism spectrum disorders (ASHA, 2006). The CCC-2 provides yet another useful and valid tool among a battery of assessment practices necessary for capturing the richness and complexity of pragmatic language functioning in children with autism spectrum disorders (ASHA, 2006) and Williams syndrome. Given that pragmatic language behaviors have historically been somewhat clinically elusive, with norms lacking even for typical development in the school-age years, the provision of another tool for consideration of these behaviors is critical (Norris, 1995).

While the CCC-2 was able to provide evidence for "typical" functioning in the typical group and impaired functioning in the two clinical groups, the performance of the TYP group was very high across every CCC-2 subscale, given that a score of six indicates typical functioning; the TYP group averaged scores above ten on every CCC-2 subscale. These findings suggest that typically developing children appear to readily master the structural and pragmatic language skills measured by the CCC-2 by the early elementary school years. Perhaps the performance of this group of typically developing children is biased due to an over-representation of females.

Additional support for the construct validity of this tool was provided with the strong relationship observed between the four summed Scaled Scores of the pragmatic language scales (i.e., PRAG) and the adaptive Socialization Standard Score of the VABS/II. This would suggest that these scales of the CCC-2 are measuring a construct very similar to

adaptive socialization: pragmatics. However, the lack of an association between the Social Interaction Deviance Composite variable with the Socialization Standard Score of the VABS/II in the autism disorder spectrum group was concerning in this sample. Perhaps this impairment variable would rather show a relationship with other direct assessments of pragmatic language functioning, such as the *Test of Pragmatic Language* (TOPL; Phelps-Teraski & Phelps-Gunn, 1992) or *Comprehensive Assessment of Spoken Language* (CASL; Carrow-Woodfolk, 1999). These findings further strengthen an argument for the utility of the PRAG variable with the CCC-2, as opposed to the Social Interaction Deviance Composite variable despite its prominence in the tool's manual (Bishop, 2003).

*Potential Limitations*

Difficulties in capturing the construct of interest (i.e., pragmatics), including selection of an instrument that could adequately identify children with difficulties in this area, posed limitations. A number of different authors speak to difficulties with the construct of pragmatics in terms of definition, scope, and measurement (Bishop, 1988; Ninio & Snow, 1996; Ramberg et al., 1996; Young et al., 2005). Further, validity data on the instrument chosen to measure pragmatics in this study remains somewhat limited, considering difficulties with the construct, relative newness of the tool, and potential limitations for use of the tool in clinical samples. Additionally, because Williams syndrome is a relatively rare genetic condition, the sample sizes were relatively small, limiting the power of the findings. Using parents as raters of their child's language functioning posed additional possible limitations as a result of the subjectivity inherent in a parent's rating of his/her own child's language abilities.

Another potential limitation concerned whether or not to match the groups on a third control variable (e.g., verbal or nonverbal mental age). By not matching the groups on a third control variable, limitations presented in terms of the study's ability to attribute pragmatic language difficulties to diagnostic group, rather than to some other variable, such as nonverbal mental age (Mervis & Klein-Tasman, 2004). However, group matching is complicated when comparing autism with Williams syndrome. The cognitive-linguistic profiles are starkly different in autism and Williams syndrome, such that while nonverbal cognition is a relative strength compared to language functioning in autism the opposite situation presents in children with Williams syndrome (APA, 2003; Mervis & Klein-Tasman, 2000). Further, while autism is an extremely heterogeneous group with cognitive levels ranging from the range of mental retardation into superior levels of intellectual functioning, children with Williams syndrome tend to cognitively present within the range of mental retardation (Chakrabarti & Fombonne, 2001; Fombonne, 1999; Mervis & Klein-Tasman, 2000). Thus, matching the children on nonverbal mental age would have limited the study of children with autism to those who were functioning within the range of mental retardation creating the extra difficulty of locating that subset of children with autism who also speak in full sentences. Further complicating this issue, Tager-Flusberg (2004) reported that most verbal children with autism have normal nonverbal intelligence. Furthermore, pragmatic performance may not be assumed to be linked to cognitive functioning when one considers that children with autism present with difficulties in pragmatics, even when cognition is in the average to above average range, while children with Down syndrome, who typically present with mental retardation, demonstrate performance in pragmatics that is not indicative of impairment (Dawson et al., 2003; Laws & Bishop, 2004; Young et al., 2005). Further, VIQ did not demonstrate significant associations to any pragmatic outcome

variable in either the ASD or TYP groups despite a trend toward a small negative association between VIQ and one pragmatic outcome variable (i.e., Social Interaction Deviance Composite) in the group of typically-developing children.

In the current study, children were qualified for the study based upon diagnosis, age, and gross level of language functioning (i.e., talking in full sentences), which has been the precedent for research comparing different diagnostic groups on pragmatics using the CCC and CCC-2 (Bishop & Baird, 2001; Botting, 2004; Botting & Conti-Ramsden, 1999; Gilmour, Hill, Place, and Skuse, 2004; Guerts et al., 2004; Laws & Bishop, 2004; Norbury et al., 2004). Further, Burack and colleagues (2003) cite one useful strategy for studying groups that are difficult to match on measures of general developmental functioning that has to do with matching groups "on a measure that is related to the ability that is being tested (p. 35; Burack, Pasto, Porporino, Iarocci, Mottron, & Bowler, 2003)." For this study's purposes, a child's chronological age serves this particular purpose. Presumably, age itself and getting older results in an amassment of opportunity, practice, and skill with social-communicative behaviors which result in pragmatic language development. The over-representation of females in the sample of typically developing children posed a possible limitation. Finally, reliance on parental confirmation of a Williams syndrome diagnosis potentially introduced a limitation.

## *Conclusions and Implications*

### *Clinical Implications*

This study has some important implications for speech-language pathologists, educators, and interventionists working with children with autism and Williams syndrome. Understanding of the pragmatic language phenotypes for individuals with these disorders may be useful to educators and interventionists working with these populations in terms of

the implementation of targeted therapies (Hodapp & Fidler, 1999). Additionally, a closer look at the pragmatic functioning of these two syndrome groups allows for a richer understanding of what constitutes atypical pragmatic language functioning in school-aged children. Given our current weaknesses in understanding what constitutes typical pragmatic language functioning in school-aged children, delineating the parameters of atypicality further assists in our understanding of pragmatic language development.

With respect to assessment, this study provided data to support the use of a relatively new tool for the assessment of pragmatic language impairment in school-aged American children with syndromes affecting social development, as one tool within a battery, for understanding the pragmatic language functioning of a child. However, use of the PRAG variable (i.e., the sum of the four pragmatic language scales) of the CCC-2 (rather than the Social Interaction Deviance Composite variable) would be recommended for use in populations who are also at-risk for structural language difficulties. Use of a measure like the CCC-2, along with other methods for looking at pragmatic language (i.e., direct assessment, observation, etc.), is currently considered best practice for assessing pragmatic language functioning in these populations (ASHA, 2006). Given the significant and sharp rise in prevalence of autism over the past decade children with the disorder are likely to be increasing members on therapist caseloads. Therefore, information about valid and useful clinical tools is critical to the speech-language pathologists working with these children.

In terms of intervention, borrowing ideas for pragmatics treatments from autism may be a necessary place to start with selecting programming for children with Williams syndrome. However, an understanding of the Williams syndrome phenotype could allow for appropriate and sensitive alterations in intervention techniques better suited to a child with Williams syndrome. Whereas targeted social skills/pragmatics training in autism tends to be

cognitive and rule-based, use of feelings and how different behaviors affect others may be more effective for teaching a child with Williams syndrome about appropriate social behavior, given their relative strength in Nonverbal Communication and Social Relations. Thus, knowledge of the behavioral tendencies of a child with Williams syndrome would inform the therapist as to possible techniques for altering an existing intervention protocol. Finally, use of valid tools for the assessment of pragmatics may aid therapists in developing treatment plans and monitoring pragmatic language gains in these populations (Young et al., 2005).

*Research Implications*

This study provided a more refined understanding of the similarities and differences of the pragmatic language behavioral profiles of school-aged children with autism and Williams syndrome. This pairwise comparison of two disorders affecting social development provided for a better understanding of possible outcomes, including a better understanding of the vulnerabilities of language and social development (Rice et al., 2005). While this study suggested that both school-aged children with autism and Williams syndrome show impairment, children with Williams syndrome demonstrated relative strength in a few certain areas of pragmatic functioning.

A number of implications for future work have been highlighted in this study for both autism and Williams syndrome. Future studies should consider replicating these findings in a larger sample and attempting to understand the reasons for variability in pragmatic language functioning in both disorders, as well as better understand whether or not every child with Williams syndrome is impacted by pragmatic language impairment. The establishment of norms in typical school-aged populations would also further work in our ability to characterize and describe atypicality in children (Norris, 1995). Studies to consider

the efficacy of treatments designed to target different areas of pragmatic language impairment in these populations will be critical (Adams et al., 2005). Finally, future studies should examine relations between the Social Interaction Deviance Composite variable and other measures of pragmatic language functioning, such as the *Test of Pragmatic Language* (TOPL; Phelps-Teraski & Phelps-Gunn, 1992) or *Comprehensive Assessment of Spoken Language* (CASL; Carrow-Woodfolk, 1999).

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