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Early communication abilities of 24-month-old children with Williams syndrome as measured by the Communication Complexity Scale.

Sean B. Woods
University of Louisville

Early Communication Abilities of 24-Month-Old Children with Williams Syndrome as Measured by the Communication Complexity Scale

By

Sean Woods

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Abstract

Williams syndrome (WS) is a neurodevelopmental disorder associated with intellectual disability accompanied by a distinct cognitive profile. Despite their socially outgoing nature, children with WS exhibit delayed communication development and specific deficits across various functions of communication. The purpose of this study was to describe the range of communication complexity observed in 24-month-olds with WS and determine relations between communication complexity and other areas of cognitive development. The communication complexity of 17 24-month-old children with WS was measured using the Communication Complexity Scale (CCS), which quantifies optimal and typical communication complexity, as well as optimal communication for Joint Attention (JA) and Behavior Regulation (BR) functions. Other aspects of development were measured using the Mullen Scales of Early Learning (MSEL), MacArthur–Bates Communicative Development Inventory (CDI), and language measures derived from a naturalistic play session with the child's mother. A wide range of communication complexity was observed, from dual and triadic orientations to multiple-word phrases. Communication complexity was significantly and strongly correlated with expressive and receptive language, fine motor skills, overall cognitive development, expressive vocabulary size, and language abilities in a naturalistic setting. The findings indicated that the CCS is a valid measure of communication complexity in young children with WS and could be used as a diagnostic tool to identify intervention goals and measure the effectiveness of implementation of intervention techniques targeting communication development.

Introduction

Williams syndrome (WS) is a neurodevelopmental disorder characterized by a deletion of 25 - 27 genes on chromosome 7q11.23 (Hillier et al., 2003) with an estimated prevalence of 1 in 7,500 live births (Strømme et al., 2002). Individuals with WS typically have borderline intellectual ability to moderate intellectual disability accompanied by distinct cognitive and personality profiles (Mervis & John, 2010). Individuals with WS tend to acquire language later and more slowly than typically developing (TD) children (Mervis & John, 2012). There has been little research on the characteristics of presymbolic and early symbolic communication in young children with WS, therefore this study will address this need.

Early Communication in Typically Developing Children

Infants begin communicating pre-intentionally, using purposeful behaviors not directed toward another person (Carpenter et al., 1998). These include behaviors such as changes in eye gaze or facial expression in response to stimuli. Infants then develop intentional communication, which includes gestures and vocalizations directed toward another person as indicated by eye gaze, touch, or posture (Brady et al., 2012). By 12 months of age, TD infants are able to combine vocalizations and gestures to communicate (Salley et al., 2020). From there, children tend to begin speaking using single words, which opens the door to future communication combining these words into phrases and sentences.

Joint Attention (JA) and Behavioral Regulation (BR) are some of the reasons infants and young children communicate (Bakeman & Adamson, 1984). JA communication involves sharing emotion or experience, such as pointing with coordinated eye contact or showing a toy the child finds interesting (Salley et. al, 2020). BR communication is focused on requesting or avoiding, such as pointing to a desired toy to request it or gesturing to show disgust toward an object.

Communication complexity can be defined by the use of various forms of communication (eye contact, gestures, vocalizations, etc.) and the level of coordination between these forms of communication toward a communication partner. For example, a communication event that includes a word would be considered more complex than a vocalization or a gesture. Also, a gesture combined with a vocalization would be considered more complex than either behavior on its own. Complexity can aid in the characterization of communication quality and skill, which can allow for more sensitive measurement of subtle developmental changes in children, which can be indicative of their future developmental trajectory and may identify potential deficits from a young age (Salley et al., 2020).

Early Communication in Children with Intellectual and Developmental Disabilities

Measuring communication complexity is especially important for children with intellectual and developmental disabilities (IDD). Many IDD populations have slow and delayed communication development, as well as particular deficits in specific communication functions and other areas of development. With targeted intervention, communication complexity could be increased, which could lead to subsequent improvements across other areas of language and cognitive development.

Several studies addressing presymbolic communication have been conducted with individuals with IDD, primarily within a behaviorist framework. Many studies have demonstrated that children with Autism Spectrum Disorder (ASD) often have deficits in social communication, eye contact, gaze following, social orientation, and the use of gestural communication (Bhat et al., 2010; Wetherby et al., 2004; Zwaigenbaum et al., 2013). Children with fragile X syndrome (FXS) are considered to have deficits in social communication, reciprocity, and representational gestures (Flenthrope & Brady, 2010; Hahn et al., 2017;

Marschik et al., 2014). Children with Down syndrome (DS) are considered to develop symbolic language one to two years later than TD children but exhibit a relative strength in the use of gestures across communication functions when compared to other IDD populations (Abbeduto et al., 2007).

Frequently used measures of early language and communication, such as the Receptive Language (RL) and Expressive Language (EL) scales of the Mullen Scales of Early Learning (MSEL; Mullen, 1995) and the Bayley Scales of Infant Development (BSID-III; Bayley, 2005), capture more broad elements of communication to determine the absence or presence of a delay in that specific skill, but do not differentiate between more specific levels of presymbolic communication complexity (Salley et al., 2020). Additionally, these measures do not adequately distinguish between different levels of presymbolic and early symbolic communication, leading to significant floor effects for certain IDD populations that do not reflect the true communication complexity of these individuals (Brady et al., 2012; 2018). Other scales of presymbolic and early symbolic communication complexity, such as the MacArthur–Bates Communicative Development Inventory (CDI; Fenson et al., 2007) Words & Sentences form, which focuses on expressive vocabulary size and grammatical complexity, and the Communication and Symbolic Behavior Scales (CSBS; Wetherby & Prizant, 2003) Parental Report section, which focuses on the ability of the child to initiate a specific type of communication event, rely heavily on parental report data, which may result in an overestimation of intentional communication when compared to a more objective, trained observer (Brady et al., 2012). Other measures, including the Early Social Communication Scales (ESCS; Mundy et al., 2003), which consists of a play protocol designed to elicit specific acts of verbal and non-verbal communication in young children, are administered to the child by an examiner but focus on frequency of specific communication acts rather than complexity (Brady et al., 2012). Furthermore, many of these scales are not sensitive to subtle changes in communication complexity over time, which are especially important when monitoring the effectiveness of early intervention measures in individuals with IDD (Brady et al., 2012; Salley et al., 2020). Additionally, these measures do not provide a numeric score that can describe the overall level of presymbolic and early symbolic communication complexity for individuals with IDD, making it difficult to compare such complexity to other measures of development (Brady et al., 2012; Salley et al., 2020).

All of these concerns are adequately addressed by the Communication Complexity Scale (CCS; Brady et al., 2012), making it an ideal measure to describe presymbolic and early symbolic communication in individuals with IDD. The CCS is one of the few measures available that is specifically focused on assessing the complexity of communication in individuals with limited linguistic capabilities. The CCS was designed to evaluate early communication, objectively differentiating between various levels of presymbolic and early symbolic communication, as well as between the JA and BR communication functions, in the context of a structured experimental setting. CCS scores provide a quantitative evaluation of early communication complexity, which are sensitive to subtle developmental changes and can be compared to other metrics of language and cognitive development.

Several studies have considered the concurrent validity of the CCS relative to other common measures of early language abilities for TD children. Brady and collaborators (2012) found that CCS Optimal scores were significantly correlated with MSEL Expressive Language scores ($r = .40$) and with the Preschool Language Scale-4 (Zimmerman et al., 2003) Expressive Language Scale ($r = .44$) in a sample of preschool-aged children. Salley et al. (2020) found that expressive vocabulary size, as measured by the MacArthur–Bates Communicative Development

Inventory (CDI; Fenson et al., 2007) vocabulary checklist, was significantly related to CCS JA complexity ($r = .30$), CCS BR complexity ($r = .31$), and CCS JA frequency ($r = .35$), but not to CCS BR frequency ($r = .18$) for 12-month-old TD children.

Studies addressing the concurrent validity of the CCS for children with IDD also have been conducted. Brady and collaborators (2018) found that overall CCS scores were significantly correlated with Vineland Adaptive Behavior Scales–Second Edition (VABS-II; Sparrow et al., 2005) Expressive Language scores ($r = .47$, $p < .01$) and the Communication Matrix (CM; Rowland & Fried-Oken, 2010) scores ($r = .35$, $p < .01$) in a sample of 231 individuals with IDD between the ages of 3-66 years. Additionally, Brady and collaborators (2018) found that CCS Optimal JA scores were significantly correlated with both CM scores ($r = .44$, $p < .01$) and VABS-II ($r = .50$, $p < .01$) Expressive Language scores, while Optimal BR scores were significantly correlated with VABS-II Expressive Language scores ($r = .50$, $p < .01$), but not CM scores ($r = .17$, $p > .01$). Brady and collaborators (2018) also noted high interrater reliability ($\kappa = .83$) for the CCS in evaluating early communication in individuals with IDD.

Differences in JA and BR communication function skills for children with IDD have also been addressed in two studies. Findings suggest that most children with IDD exhibit differences in JA and BR communication functions (Brady et al., 2004; McLean et al., 1991). Fleming and Brady (2019) noted that minimally verbal (expressive vocabulary of fewer than 20 words or signs) individuals with ASD had CCS Optimal BR scores that were on average 0.62 points higher than CCS Optimal JA scores compared to a 0.27-point difference for minimally verbal individuals that had other IDD. This discrepancy in score differences between JA and BR function communication was significant, demonstrating that the disparity between JA and BR function complexity was significantly greater for children with ASD than for children with other

IDD. In another study, Hahn and collaborators (2017) found that children with FXS engaged in similar proportions of JA and BR communication, but no data on the differences in JA and BR function complexity were reported.

Current research has demonstrated that the CCS is a valid measure of communication complexity for both the JA and BR communication functions for IDD populations. For this reason, the CCS allows researchers to evaluate deficits and strengths in communication complexity for children with IDD and the information obtained could then be used to determine appropriate intervention goals to improve the children's developmental trajectories.

Early Communication in Children with Williams Syndrome

The onsets of expressive language and gestural communication are well documented as being delayed for children with WS (Klein-Tasman et al., 2007; Mervis & Becerra, 2007; Mervis & John, 2012; Mervis & Robinson, 2000). Mervis and Becerra (2007) noted the average age of acquisition of a 100-word expressive vocabulary was 40.90 months, which is considerably older than the TD average age of attainment of a 100-word expressive vocabulary (18 months). Of the 13 children with WS studied, the average ages at attainment of a 10-word, 50-word, and 100-word expressive vocabularies were below the 5th percentile on the CDI norms. Becerra (2016) examined the lexical abilities of 56 24-month-old children with WS. The children in this study had a mean CDI-EV of 34.71 (*SD* = 7.15), a median of 21.00, and a range from 0 – 176 words. Of these 56 participants, 44 (78.6%) had a CDI-EV score at or below the 5th percentile on the CDI norms, while 4 scored between the 6th and 9th percentile, 5 between the 10th and 15th percentile, 2 between the 16th and 20th percentile, and 1 at the 25th percentile. Similar findings were noted at both 18 and 30 months of age.

The onset of declarative gestures is delayed for children with WS relative to TD children, as children with WS typically develop declarative gestures at a median age of 24.08 months (Becerra & Mervis, 2019) while TD children typically begin using declarative gestures at approximately 10 months of age (Fenson et al., 1997). It has also been found that referential language precedes declarative pointing gestures for children with WS, which differs from TD children, who typically use declarative pointing gestures before using referential words (Becerra & Mervis, 2019). In the aforementioned study by Becerra and Mervis, it was also found that both CDI-EV at 24 months and chronological age (CA) at onset of declarative point gestures were strong predictors of CDI-EV at 48 months ($R^2 = .71$, $p < .001$).

Specific difficulties in JA for young children with WS have been noted in several other studies. Laing and collaborators (2002) compared the communication of young children with WS (*M* = 30.90 months, *SD* = 11.5) to a mental age-matched group of TD infants (*M* = 13.50 months, *SD* = 5.04) and found that despite relatively larger expressive vocabulary sizes, the children with WS had significant difficulties initiating JA and understanding their communication partners' acts of JA communication relative to the TD group. Vivanti and collaborators (2017) studied a sample of preschool-aged children (M= 52.13 months, SD= 16.92) with WS using eye-tracking data in standardized play interactions. The findings of this study suggested that despite a strong social interest, deficits in JA comprehension hindered both the ability to understand the purposes behind JA function communication events and the understanding that targeted objects have a special status in context of the communication event. Klein-Tasman and collaborators (2007) noted that many young children with WS have significant difficulties with pointing, giving, showing, and both initiating and responding to JA.

Present Study

No previous studies have addressed the complexities of presymbolic and early symbolic communication in children with WS as measured by the CCS. Based on the validity of the CCS in characterizing early communication complexity in individuals with other IDD, it will be important to determine if this measure will appropriately characterize early communication complexity in children with WS as well. Regardless of the aforementioned delays or abnormalities in early communication for children with WS, the current literature suggests that presymbolic and early symbolic communication development is important for later language development in children with WS, which is why it is important to further address the characteristics of presymbolic and early symbolic communication in this group of children.

The current study seeks to answer two major questions regarding early communication in 24-month-old children with WS:

1. What are the characteristics of presymbolic and early symbolic communication abilities of 24-month-old children with WS as measured by the CCS?
2. How do these communication abilities relate to concurrent measures of language and cognition?

It was expected that most 24-month-olds with WS would be able to speak at least a few words and to use a range of both presymbolic and symbolic communication, although there was expected to be considerable variability in both CDI-EV and communication abilities. Further, positive concurrent correlations were expected between CCS scores and measures of intellectual abilities, lexical abilities, and grammatical abilities. The current study will further contribute to the existing research on the validity of the CCS as a measure to evaluate early communicative abilities in children with IDD. This will also be the first study to evaluate communication complexity using the CCS for individuals with WS, which will establish a precedent for future studies on communication complexity in these individuals while simultaneously expanding the validity of the CCS to a different IDD population. The findings could assist researchers in better understanding how children with WS develop, which may lead to the development of better early intervention for these individuals.

Methods

Participants

Participants were 17 24-month-old children (10 girls, 7 boys) with genetically confirmed classic deletions of the WS region. Mean CA was 24.42 months (SD: 0.29, range: 24.00 – 24.87 months). All 17 participants were native speakers of English and were White, non-Hispanic, and all of their mothers had at least a bachelor's degree. The participants lived in 14 different states, representing all four United States census regions (12% Northeast, 47% South, 29% Midwest, 12% West). Data collection began in June 2017 and ended in September 2019.

Measures

Communication Complexity Scale (CCS)

Early communication complexity was measured using the CCS (Brady et al., 2015). The CCS is composed of a series of 12 standardized play interactions consisting of different tasks with various toys and scripted interactions for the experimenter. Presymbolic and symbolic communication demonstrated by the children during these tasks was coded with numerical scores ranging from 0 – 12. A "0" score indicated no response, "1"-"5" described progressions in pre-intentional communicative behavior, "6"-"10" included intentional presymbolic communication directed toward the experimenter, and "11"-"12" included intentional symbolic communication acts utilizing speech or sign language to communicate. The full scoring scale for the CCS (Brady et al., 2015) can be seen in Table 1.

Table 1

Communication Complexity Scale (CCS) Scoring

0	No response	
1	Alerting—a change in behavior, or stops doing a behavior	Preintentional
2	Single orientation only—on an object, event, or person; can be communicated through vision, body orientation, or other means.	Preintentional
3	Single orientation only + 1 other PCB	Preintentional
4	Single orientation only + more than 1 PCB	Preintentional
5	Dual orientation—shift in focus between a person and an object, between a person and an event using vision, body orientation, etc. (without PCB)	Preintentional
6	Triadic orientation (e.g., eye gaze or touch from object to person and back)	Intentional Nonsymbolic
7	Dual orientation + 1 PCB (e.g., dual focus + gesture)	Intentional Nonsymbolic
8	Dual orientation + 2 or more PCB (e.g., dual focus + gesture + vocalization, switch closure)	Intentional Nonsymbolic
9	Triadic orientation + 1 PCB (e.g., triadic + vocalization)	Intentional Nonsymbolic
10	Triadic orientation plus more than 1 PCB (e.g., triadic plus vocalization and differential switch closure)	Intentional Nonsymbolic
11	One-word verbalization, sign, or AAC symbol selection	Intentional Symbolic
12	Multi-word verbalization, sign, or AAC symbol selection	Intentional Symbolic

Note: Copyright University of Kansas, 2015. PCB: Potentially Communicative Behavior.

The highest numerical score for each task was recorded, and a categorization of either Joint Attention (JA) or Behavioral Regulation (BR) was made for each intentional communication event (score of 6 or above) based on the CCS Scoring Manual (Brady et al., 2015). Each participant received four unique CCS scores: CCS Optimal, CCS Typical, CCS Optimal JA, and CCS Optimal BR. The CCS Optimal score was determined by computing the mean of the three highest scores for the participant across the 12 tasks. The CCS Typical Score was determined by calculating the mean of the middle six scores (after removing the three highest and three lowest scores). CCS Optimal JA and CCS Optimal BR scores were determined by computing the mean for the three highest JA function and BR function scores, respectively. In the event that a participant did not have 3 JA or BR scores, the mean was calculated based the available JA or BR score(s), respectively.

Mullen Scales of Early Learning (MSEL)

Intellectual abilities were measured using the MSEL (Mullen, 1995). The MSEL includes four subscales with mean T scores of 50 (*SD* = 10) for the general population: Visual Reception (VR; measuring primarily nonverbal reasoning), Fine Motor (FM; measuring primarily visuospatial construction), Receptive Language (RL), and Expressive Language (EL). The Early Learning Composite (ELC), which takes into account performance on all four scales, was also determined. The ELC general population mean is 100 (*SD* = 15).

MacArthur-Bates Communicative Development Inventory-Words and Sentences (CDI-W&S)

Lexical abilities were assessed using the CDI-W&S (Fenson et al., 2007), a parental report measure of language development. Lexical abilities were measured using the Vocabulary Checklist, a 680-item list of words that provides an Expressive Vocabulary (CDI-EV) score based on how many of the 680 words on the checklist the parent reported that the child could say or sign spontaneously.

Language Abilities as Measured by a Naturalistic Observation

Child-mother dyads participated in a 30-minute videotaped play session to determine language abilities in a naturalistic setting. Play sessions were transcribed using the Systematic Analysis of Language Transcripts (SALT; Miller & Iglesias, 2019). Language measures from the play session transcripts included lexical ability (Number of Different Words, NDW) and grammatical ability (Mean Length of Utterance in Morphemes, MLUm). Due to technical difficulties during her play session, one participant was excluded from the analyses that included these measures.

Procedure

All measures were administered as part of the participant's 24-month-old assessment at the Neurodevelopmental Sciences Laboratory at the University of Louisville.

Reliability Analysis

An initial analysis of CCS intercoder reliability yielded a Cronbach's α of .910, showing high intercoder consistency. Discrepancies were subsequently resolved by consensus.

Data Analysis

Data were analyzed using IBM SPSS v. 27. The distributions of CDI-EV, Mullen RL, Mullen EL, NDW, and MLUm scores were non-normal. For this reason, nonparametric Spearman correlation coefficients were used to determine the concurrent relations between CCS, MSEL, CDI-EV, NDW, and MLUm scores. For the correlation analyses, α was set at $p = .01$, 2-tailed.

Results

Early Communication Abilities

Descriptive statistics for the CCS measures are presented in Table 2. These statistics allow us to objectively quantify and describe the communication abilities of 24-month-old children with WS, addressing Research Question 1.

Table 2

Descriptive Statistics for CCS Scores

Variable	Mean	Median	SD	Interquartile Range	Range
CCS Optimal	10.65	11.00	.84	10.00-11.17	9.00-12.00
CCS Typical	9.54	9.40	1.46	8.50-10.92	6.50-11.33
CCS Optimal JA	9.74	9.50	1.22	9.17-11.00	7.33-11.33
CCS Optimal BR	10.36	11.00	1.16	10.00-11.00	7.33-12.00

Note: $N = 17$, CCS: Communication Complexity Scale, JA: joint attention, BR: behavior regulation

A wide range of Optimal CCS scores was noted. The CCS Optimal score for one individual in the sample was 12.00, indicating that she often communicated using phrases of two or more words. The minimum score in this sample was 9.00, indicating that even the lowest-scoring individual was sometimes able to communicate using a triadic orientation accompanied by a Potentially Communicative Behavior (PCB), which could include any gesture, such as pointing or giving, or a vocalization that demonstrates clear communicative intent but lacks a specific symbolic meaning. The mean Optimal CCS Score for this sample was 10.65 (SD: 0.84), which demonstrates that the average level of optimal communicative ability for this sample included spoken or signed words.

An even wider range was noted for Typical CCS scores, with values from 6.50 – 11.33 for individual participants. Four participants had Typical CCS scores of 11.00 or higher, indicating that these children typically communicated using words. At the other extreme, the child with the most limited communication typically communicated with a combination of isolated triadic orientations and dual orientations coupled with a single PCB, such as a vocalization or a gesture. Typical CCS Scores (*M*: 9.54, *SD*: 1.46) demonstrated that the average typical level of communication for this sample involved triadic orientations accompanied by one PCB.

On average, the optimal level of communication for the individuals in this study was nominally higher level when utilizing the BR function (M = 10.36, SD = 1.16) as opposed to the JA function (M = 9.74, SD = 1.22). However, this difference was not statistically significant, $t(16) = -1.87$, $p = .080$.

Performance on Measures of Early Cognitive and Language Assessments

Descriptive statistics for performance on the MSEL are shown in Table 3. MSEL scores showed a wide array of abilities across all scales.

Table 3

Descriptive Statistics for MSEL Scores

Variable	Mean	Median	*SD*	Interquartile Range	Range
MSEL VR	32.71	34	7.27	28.0-39.0	20[a]-43
MSEL FM	30.82	32	6.89	24.0-36.0	20[a]-40
MSEL RL	30.24	24	10.81	20.0[a]-37.5	20[a]-56
MSEL EL	31.47	30	8.80	25.0-37.0	20[a]-51
MSEL ELC	66.12	65	11.96	55.5-73.5	50-89

Note: $N = 17$. [a]Lowest possible T-score. MSEL: Mullen Scales of Early Learning, VR: Visual Reception, FM: Fine Motor, RL: Receptive Language, EL: Expressive Language, ELC: Early Learning Composite.

Table 4 provides descriptive statistics for CDI-EV and the measures computed from the play session transcripts. CDI-EV scores demonstrated that the participants present considerable variability in lexical abilities. The mean CDI-EV of the sample is consistent with the mean reported in a larger study on language and cognitive abilities in 24-month-olds with WS utilizing a separate sample (Becerra & Mervis, 2019), suggesting that the present sample is likely representative of the general population of 24-month-old children with WS. Based on parental report (CDI-EV), all participants had begun to produce words spontaneously. All but two participants produced at least one word spontaneously during the 30-minute play session. Most children had not yet begun to combine words.

Table 4

Descriptive Statistics for CDI-EV and Play Session Measures

Variable	Mean	Median	*SD*	Interquartile Range	Range
CDI-EV	43.06	18.00	63.61	3 – 47	0 – 236
MLUm	0.94	1.00	0.38	1.00-1.16	0.00 – 1.33
NDW	9.06	4.00	13.07	1 – 14	0 – 42

Note: *N* =17 for CDI-EV and 16 for Play Session variables. CDI-EV: MacArthur-Bates Communicative Development Inventory-Words and Sentences Expressive Vocabulary score, MLUm: mean length of utterance in morphemes, NDW: number of different words.

Spearman correlations were computed to determine the relations between CCS variables. CCS Optimal performance and CCS Typical performance were strongly related, *rho* = .84, $p < .001$. In contrast, the correlation between BR Optimal performance and JA Optimal performance was not statistically significant, *rho* = .37, $p = .144$. Table 5 lists Spearman correlations between the four CCS measures and the five MSEL measures, CDI-EV, and the two play session measures. These correlations allow us to examine the relations between communication complexity as measured by the CCS and the other measures of language and cognitive ability, addressing Research Question 2. Both CCS Optimal and CCS Typical scores were significantly correlated with MSEL RL, MSEL EL, MSEL ELC, CDI-EV, and NDW values, demonstrating significant and strong relations between communication complexity as measured by the CCS and other common measures of early language and cognitive development. CCS Optimal JA Scores were significantly correlated only with MSEL EL, CDI-EV, MLUm, and NDW, demonstrating significant and strong relations between JA function complexity as measured by the CCS and measures of expressive language abilities. CCS Optimal BR scores were significantly correlated with MSEL RL, MSEL ELC, and NDW, demonstrating significant and strong correlations between BR function complexity as measured by the CCS and some common measures of receptive language, overall cognitive abilities, and expressive language.

Table 5

Spearman Correlations Between CCS Performance and Measures of Language or Cognitive Abilities

Measure	MSEL VR	MSEL FM	MSEL RL	MSEL EL	MSEL ELC	CDI-EV	MLUm	NDW
CCS Optimal	.52	.55	.65*	.67*	.67*	.62*	.60	.74*
CCS Typical	.52	.70*	.69*	.79**	.77**	.74*	.69*	.81**
CCS Optimal JA	.47	.52	.50	.66*	.60	.70*	.65*	.65*
CCS Optimal BR	.45	.51	.61*	.59	.62*	.49	.56	.68*

Note: CCS: Communication Complexity Scale, JA: joint attention, BR: behavior regulation, MSEL: Mullen Scales of Early Learning, VR: Visual Reception, FM: Fine Motor, RL: Receptive Language, EL: Expressive Language, ELC: Early Learning Composite, CDI-EV: MacArthur-Bates Communicative Development Inventory-Words and Sentences Expressive Vocabulary score, MLUm: mean length of utterance in morphemes, NDW: number of different words.
*$p<.01$ (2-tailed), ** $p< .001$ (2-tailed).

Discussion

The purpose of this study was twofold: to characterize presymbolic and early symbolic communication abilities of 24-month-old children with WS using the CCS and to determine the relations between communication complexity as measured by the CCS and other measures of early language and cognitive development. The Discussion will focus on addressing both purposes of the study, comparing the results to the findings of previous studies, discussing the implications of these findings, explaining specific limitations of the current study, and providing direction for future research.

Language Abilities of 24-month-olds with Williams Syndrome

Because most intervention focuses on developing new skills and generalizing them to different situations, it is especially important to consider both CCS Optimal and CCS Typical scores when measuring communication complexity. As was expected based on the previous research in children with WS (Becerra, 2016; Becerra & Mervis, 2019; Klein-Tasman et al., 2007; Mervis & Becerra, 2007) using other measures, this sample of 24-month-old children with WS exhibited a wide range of communicative abilities based on results from the CCS. CCS Optimal scores indicated all children in the sample were able to pair a triadic orientation with 1 or more PCBs, 12 out of 17 children (71%) were on the cusp of shifting or had already shifted from presymbolic to symbolic communication, and 4 of 17 children (24%) could use word combinations to communicate. CCS Typical scores indicated that all individuals in the sample were typically communicating using at least some combination of triadic orientations unaccompanied by a PCB or dual orientations with one or more PCBs, 13 of 17 children (76%) were typically combining PCBs with triadic orientation, and 8 of 17 children (47%) were using symbolic communication in typical communication settings. These results demonstrate that all participants were utilizing intentional communication in typical settings and were at least starting to utilize either gestures or vocalizations, which is also consistent with previous studies (Becerra, 2016; Becerra & Mervis, 2019). This transition to symbolic communication is also confirmed by results from the CDI-EV, which showed that all but one child in this sample had an expressive vocabulary of more than one word (12 of 17 had at least 5), a pattern that is consistent with other studies analyzing different samples of children with WS at the same CA (Becerra, 2016; Becerra & Mervis, 2019; Mervis & Becerra, 2007).

CCS Optimal JA scores indicated that all participants could engage in JA function behavior with at least a dual orientation coupled with one or more PCB, while the participants with the most advanced communication skills could engage in JA using one or more words. Further examination of CCS Optimal JA scores indicated that at least 15 of 17 children (88%) in the sample could communicate using triadic orientation with one or more PCBs when communicating with JA function, and 7 of 17 children (41%) were using words or signs in JA function communication.

CCS Optimal BR scores indicated that all participants in the study could engage in BR function behavior with at least a dual orientation coupled with one or more PCBs in an optimal setting, while the most advanced participants could engage in such communication with multiple words. Further analysis of CCS Optimal BR scores indicated that 10 of 17 children (59%) were able to communicate with words and 15 of 17 (88%) children in this study were at least combining triadic orientations with both gestures and vocalizations for BR function communication.

Relations of Early Communication Complexity to Measures of Language and Cognitive Development

Both CCS Optimal and CCS Typical scores were significantly correlated with MSEL RL, MSEL EL, MSEL ELC, CDI-EV, and NDW. The significant correlations with MSEL RL, EL, CDI-EV scores were expected, as these scores measure receptive and expressive language abilities, and the CCS is also a measure of language abilities. These findings provide evidence of the validity of the CCS for assessing communication skills of 24-month-olds with WS. The correlations between both CCS Optimal and Typical scores with NDW, as well as between CCS Typical scores and MLUm suggest that communication complexity as measured by the CCS is related to both vocabulary usage and grammatical complexity in a naturalistic setting. The strong correlations between both CCS Optimal and Typical scores and MSEL ELC scores, as well as between CCS Typical and MSEL FM scores, further suggest that communication complexity as measured by the CCS is related to overall developmental ability and to early fine motor skills as measured by the MSEL. Overall, these correlations suggest that the CCS is an effective measure for the diagnostic evaluation of early communication abilities in children with WS.

CCS Optimal JA Scores were significantly correlated with MSEL EL, CDI-EV, MLUm, and NDW, which indicates that JA function communication complexity is related to the expressive language abilities of children in this population as measured in a variety of ways, including experimental, parental report, and naturalistic assessment.

CCS Optimal BR scores were significantly correlated with MSEL RL, MSEL ELC, and NDW. The correlation with MSEL RL scores suggests that BR communication complexity as measured by the CCS is related to receptive language abilities. The correlation with MSEL ELC suggests that BR communication complexity as measured by the CCS is related to the overall cognitive development of the child as measured by the MSEL. The correlation with NDW scores suggests that BR communication complexity as measured by the CCS is also strongly related to expressive language abilities in a naturalistic setting.

Implications

This study is the first to characterize the communication complexity of children with WS using the CCS. The results provide an initial estimate of the complexity of communication in 24-month-old children with WS, differentiating between various levels of presymbolic and early symbolic communication. As such, this was also the first study to correlate communication complexity scores from the CCS with other measures of early language and cognitive development for children with WS, providing some initial insights into how they are related. Based on the findings of this study, the CCS is a valid diagnostic tool for the evaluation of early communication abilities in 24-month-olds with WS.

These insights could assist in the implementation of targeted intervention methods for individual children with WS. By targeting deficits in communication complexity, intervention could lead more broadly to improvements in early cognitive development for these children. As Becerra and Mervis (2019) noted, communication measures at 24 months of age are significant predictors of language and cognitive abilities at 48 months. Initial evaluations of communication complexity could be performed using the CCS as a diagnostic tool, identifying key deficits that could be targeted in intervention. Given the impact of language abilities on future cognitive development, intervention targeting the improvement of communication skills could make meaningful impacts in the lives of these children. Due to the nature of JA function deficits in children with WS, JA communication complexity should be a special point of emphasis in intervention. Early learning strategies that focus on initiating and reinforcing triadic social communication behavior and JA function communication between the child and other people may be helpful in alleviating the effects of these deficits.

More generally, this study suggests that intervention should focus on reinforcing more complex communication, especially triadic over dual orientation, as well as the use of symbolic communication in the form of words and/or signs, depending on the abilities of the child, which is consistent with the findings of Becerra and Mervis (2019). By building a more interactive and communicative environment for the child while reinforcing the targeted social communication, communication complexity—and by extension other areas of overall cognitive development—could be improved.

Limitations of Current Study

Several key limitations impacted this study. One major limitation was the sample size. Given the fact that the CCS is a relatively new measure, the Neurodevelopmental Sciences Laboratory at the University of Louisville has only been able to administer the CCS to 17 children within the targeted age group. Given the relative rarity of children with WS, recruiting large samples for such studies with specific age groups presents a significant challenge. Even still, we were able to accumulate a sample of 17 children who were all within a 1-month age range, a large sample size for this disorder for a narrow age range. Unfortunately, the relatively small sample size limited the statistical power of the study. With a larger sample size, it is likely that additional significant relations would have been found between CCS variables and other language and cognitive measures. The lack of racial, ethnic, and socioeconomic variability among the participants limits the generalization of the findings.

Another limitation of the study was the lack of frequency reporting. While the frequency of communication events can also be recorded while conducting the CCS (Brady et al., 2018), frequency was not accounted for in this study, nor is it included in the scoring protocols of the CCS manual. Studies examining both the complexity and frequency of communication in this population may have provide additional insights into the nature of communication in children with WS and how both complexity and function differ across function, setting, and age.

Directions for Future Study

Future studies should address communication complexity in younger children with WS (12-month-olds and 18-month-olds) using the CCS with larger samples. Larger, more representative samples of children with WS of various races, ethnicities, nationalities, native languages, and socioeconomic statuses could allow researchers to generalize trends in communication complexity to the population of children with WS. By covering additional, younger age groups, more specific trends in communication complexity could be observed and documented, potentially identifying sensitive periods for more targeted intervention. The CCS is unlikely to be an effective measure of communication complexity in children with WS at or over 30 months of age, as most of these children are communicating with words (Becerra, 2016; Becerra & Mervis, 2019). Both longitudinal and cross-sectional studies examining the changes in communication complexity in children with WS could also be extremely beneficial in both understanding typical developmental trajectories for children with WS and evaluating the effectiveness of intervention. CCS scores could also be compared to other metrics of early language and cognitive development to demonstrate the efficacy of the CCS in comparison to these various measures. These parameters could be applied in the study of communication complexity of other IDD populations as well, providing even more information on the early development of children with IDD.

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