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Jenna N. Tinnell
University of Louisville

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Early Mathematical Abilities of 48-Month-Old Children with Williams Syndrome

By

Jenna Tinnell

Submitted in partial fulfillment of requirements for Graduation *summa cum laude*

And

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and the Department of Anatomical Sciences and Neurobiology

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Abstract

Williams syndrome (WS) is a genetic neurodevelopmental disorder associated with relative strengths in concrete vocabulary, nonverbal reasoning, and verbal short-term memory and considerable weaknesses in visuospatial construction and relational language. While the cognitive profile of WS has been studied extensively, there have been few studies of the early mathematical abilities of children with WS and the cognitive predictors of these abilities. The purpose of this study was to describe the early mathematical abilities of 48-month-olds with WS and determine the concurrent cognitive predictors of these abilities. The Differential Ability Scales—second edition (DAS-II) was used to determine cognitive and mathematical abilities for seventy-two 48-month-old children with WS. The DAS-II Early Number Concepts subtest was administered to determine the mathematical abilities of the participants. The cognitive predictors for mathematical abilities were DAS-II Verbal Ability standard score (SS), DAS-II Nonverbal Reasoning Ability SS, and DAS-II Spatial Ability SS. The results showed that the majority of the participants understood the concept of “one,” could count by rote to 10, and comprehended the relational concepts “big” and “little”. Significant delays in mathematical abilities, including counting with one-to-one correspondence, cardinality, and understanding of relational concepts, were identified. Regression analysis indicated that verbal ability contributed significant unique variance to individual differences in mathematical ability for 48-month-olds with WS. Implications of these findings are discussed.

Early Mathematical Abilities of 48-Month-Old Children with Williams Syndrome

Williams syndrome (WS) is a rare neurodevelopmental disorder caused by a microdeletion of approximately 26 genes on chromosome 7q11.23. The prevalence is 1 in 7,500 live births (Strømme, Bjørnstad, & Ramstad, 2002). Individuals with WS typically have mild to moderate intellectual disability and tend to be highly sociable and friendly (Mervis et al., 2000; Mervis & John, 2010). The WS cognitive profile is characterized as having relative strengths in concrete vocabulary, nonverbal reasoning, and verbal short-term memory and considerable weaknesses in visuospatial construction and relational language (Mervis et al., 2000; Mervis & John, 2011). The few studies that have addressed early mathematical abilities of preschool children with WS have shown these abilities to be delayed (Van Herwegen & Simms, 2020). The aim of the present study was to characterize the mathematical abilities of 48-month-old children with WS and to determine the relations between early mathematical abilities and other cognitive abilities for these children. Providing an understanding of the nature and cognitive predictors of mathematical abilities in young children with WS will offer research-based evidence for developing interventions to support the development of mathematical skills of children with WS.

Early Mathematical Abilities of Preschool-aged Typically Developing Children

Early mathematical skills correlate with later academic success and ensure a solid foundation for later more advanced mathematical skills (King & Purpura, 2021). A solid foundation for mathematics is necessary to understand certain life skills that are utilized daily such as understanding time and counting money. Certain numeracy skills are expected to be developed at various points in a child's life going from simpler to more complex abilities as laid out by the National Council of Teachers of Mathematics and the National Math Panel (Clements & Sarama, 2010).

It is expected that by the age of 3 years, typically developing (TD) children will be able to recite the number sequence up to 10 (Clements & Sarama, 2010). Fuson (1988) reported that 83% of her participants, aged 3 years 6 months – 3 years 11 months, could recite the number sequence correctly to at least 10. One-to-one correspondence, the skill of pairing each object counted with a number word, is the first quantitative reasoning skill acquired by children, and this underlies the concept of cardinality, a measure of the number of elements of a set, and later the ability to understand all quantitative reasoning, such as multiplication and division (Torbeys, Gilmore, & Verschaffel., 2015). In one study of preschoolers, 50% of 4-year-olds were able to demonstrate one-to-one correspondence between sets with as many as five or six items (Becker, 1989). A separate experiment reported that 75–88% of 3–4-year-olds answered correctly and appropriately explained the correct answer on questions assessing one-to-one correspondence of sets containing 2–5 items (Gelman, 1982). In an experiment including 48 children with a mean age of 4 years, 76% were able to do a point-while-counting exercise involving 4–6 objects, establishing their understanding of one-to-one correspondence (Fuson, 1988).

Children learn very early on the difference between number-words and other words (Fuson, 1988). Throughout years of research on over 500 children, not one 3–5-year-old ever used anything but number words when asked questions pertaining to counting (Fuson, 1988). In a study of twenty 2–3-year-olds, all children knew the cardinal meaning of the word “one” such that they were each able to understand that the word “one” represented exactly one object (Wynn, 1992). Children learn the meaning of the number words one, two, and three in that order (Espinás & Fuchs, 2022). Children know early on that counting-words refer to the number of items in a set but do not learn how to assign an exact number to a particular set of items, a

concept known as cardinality, until they are between 3 and 4 years old (Wynn, 1992). By the age of 4 years, TD children are able to demonstrate an understanding of cardinality of up to 5 items (Clements & Sarama, 2010). Fuson (1988) reported that 44% of 2 year 8 month – 3 year 11 month-olds and 73% of 4-year-olds were able to demonstrate the cardinality principle when asked “how many,” when 4–6 objects were presented. Some research argues that around 3 years of age a TD child begins to understand the concept of cardinality by realizing that counting establishes numerosity (Wynn, 1990) and that visuospatial ability is correlated with success on cardinality tasks (Ansari et al., 2003).

An understanding of core mathematical vocabulary terms, such as relational terms (e.g., more, few), is central for expressing mathematical knowledge (Schleppegrell, 2007). In a study of eighty-four 3- to 5-year-olds, the participants who were worked with over a 4-week period on a book featuring core mathematical terms (e.g., more, few) outperformed the control group on tasks assessing number knowledge including one-to-one correspondence (Purpura et al., 2021). The age of acquisition for relational concepts varies such that the acquisition of simple terms (e.g., big, little) is earlier than comparative and superlative forms of the same terms (Edmonston & Thane, 1999). Comparative adjectives are words that describe a noun by comparing it to another noun (e.g., bigger, smaller). A superlative adjective is a word that describes a noun when it is being compared to two or more nouns (e.g., biggest, smallest). By age 3 years, children can make relative judgments based on size by understanding the concept of “big” and “little” (Ebeling & Gelman, 1988). Ehri and Ammon (1972) showed that the acquisition of “many” and “few” develops much later than “big” and “little” which most 4-year-olds in their study could distinguish. Other studies have explored the understanding of relational concepts in preschoolers such as a study of 4 year 10 month – 5 year 11 month-old participants who were reported scoring

poorly on a task assessing understanding of “less” (Holland & Palermo, 1975). Within adjective categories, there seems to be a bias from an early age for the positive adjective member as seen in a study of 50–61-month-olds who performed better on tasks involving the understanding of “more” and worse on tasks assessing understanding of “fewer” (Estes, 1976).

Throughout the past decade, researchers have begun to understand the pivotal role verbal and object counting play in the development of various skills (Torbeyns, Gilmore, & Verschaffel, 2015). Verbal counting is the ability to verbally speak the number word sequence in order, both forward and backward. Learning the number order sequence occurs in two distinct phases (Fuson, Richards, & Briars, 1982). First, children begin in the acquisition phase of counting in which they understand the number sequence as a connected whole such that the different numbers are not understood separately. For example, they can count to 10 but only in a sequence through rote memorization such that they do not understand each number word separately. The elaboration phase then follows, and children gradually begin to understand the sequence in terms of its separate number words and develop an understanding of the relations between the number words (Fuson, Secada, & Hall, 1983; Fuson, Richards, & Briars, 1982). For a child to understand the concept of cardinality, a child must understand the counting principles, rules that underly counting such as one-to-one correspondence, and have relative mastery of the verbal number word sequence (Clements & Sarama, 2007). However, verbal components of numerical processing and magnitude/quantity components of numerical processing engage different psychological and neural representations (Dehaene et al., 1999; Dehaene et al., 2003). Some components of mathematics are reliant on linguistic or verbal knowledge while others are reliant on magnitude representation (O’Hearn & Landau, 2007). Magnitude representation is the

approximate mental representation of size or number order while linguistic and verbal knowledge are language-based facts or skills.

Numerous studies have reported a strong relation between children's general language and math skills. Two meta-analyses demonstrated a significant relation between vocabulary knowledge and mathematical performance (Lin, Peng & Zeng, 2021; Peng et al., 2020). A longitudinal study used the Research-Based Early Mathematics Assessment-Short Form (REMA; Weiland et al., 2012) to assess the math knowledge of TD participants averaging 4 years 7 months old at the beginning of the school year and 6 months later (Rittle-Johnson, Zippert, & Boice, 2019). The REMA assesses comprehension of math skills such as numeracy knowledge and geometric knowledge. The researchers found that spatial abilities, such as form perception, spatial visualization, and visual-spatial working memory; and verbal abilities, including receptive vocabulary, predicted performance on math concepts tested in the study both concurrently and longitudinally (Rittle-Johnson, Zippert, & Boice, 2019). Due to the importance of mathematics in early learning, it is essential to investigate the development of these abilities in children with intellectual and developmental disorders.

Early Mathematical Abilities in Children with Williams Syndrome

Few studies have addressed the development of mathematical abilities in individuals with WS. Initial studies suggested a general deficit in mathematics without looking at individual mathematical abilities (Krajcsi et al., 2009). Understanding the early mathematical skills of children with WS and their predictors may help to reveal the specific relative strengths and weaknesses that exist within the subject as well as which cognitive abilities underlie the acquisition of these skills. Furthermore, understanding the cognitive predictors of mathematical ability in children with WS will provide a foundation for developing appropriate methods by

which to assess and teach mathematics to children with WS (O’Hearn & Landau, 2007; Simms et al., 2019). Overall, several studies have shown that almost all mathematical abilities are impaired in children with WS, but in line with overall cognitive abilities (Van Herwegen & Simms, 2020). However, researchers have found that some components of mathematics are more impaired than others in individuals with WS (O’Hearn & Landau, 2007). Additionally, individuals with WS, as well as TD individuals, seem to perform better on less advanced mathematical questions, regardless of question type, that usually appear closer to the beginning of a standardized assessments as seen in a study utilizing the Test of Early Mathematical Ability (TEMA-2; Ginsburg & Baroody, 1990; O’Hearn & Landau, 2007). These findings suggest that children with WS may acquire mathematical skills in approximately the same order as TD children.

Specifically, individuals with WS have difficulties with relational/conceptual language (Mervis & John, 2010). Cardinality also seems to be a difficulty for many children with WS as seen when Ansari and colleagues (2003) reported a group of fourteen 6–11-year-olds with WS to understand cardinality similar to TD children with matching mental age 3–4-years. Many studies examining mathematical abilities in individuals with WS revealed that individuals with WS often have difficulties with mathematical concepts that engage skills underlying spatial ability (Ansari et al., 2003; O’Hearn & Landau, 2007).

However, children with WS show relatively good number familiarity, such as reciting number words (Steele, Brown, & Scerif, 2012), and small number recognition, the ability to recognize and manipulate small numbers and sets of items (Ansari et al., 2003; O’Hearn & Landau, 2007). Steele and colleagues (2012) reported that their participants, with an average age of 6.7 years, had good knowledge of the counting sequence. A study by Thomas et al. (2006) reported that their participants, mean age of 24 years and 7 months, with WS were accurate and

fast at naming numerals. Researchers suggest that some of these differences are related to the WS cognitive profile (Paterson et al, 2006). Some of the specific strengths in number familiarity and small number recognition may be due to relative strengths in verbal ability among individuals with WS. O’Hearn and Landua (2007) reported that performance on the TEMA-2 (Ginsburg & Baroody, 1990) for their participants with WS was higher than expected given their visuo-spatial abilities, and their score on the TEMA correlated with their verbal abilities. Another study showed superior performance on a verbal estimation task compared to a numerical comparison task in individuals with WS, further suggesting people with WS are better at number tasks that engage language systems (Libertus et al. 2014). Lastly, Ansari and colleagues (2003) found that verbal mental age (MA) correlated significantly with success on cardinality tasks.

Overall, while most studies have shown that the mathematical abilities of individuals with WS do not exceed those of a TD child of age 8–9 years (Van Herwegen & Simms, 2020), there seems to be variation in their abilities depending on the type of mathematical question. While several studies have suggested an atypical development of mathematical abilities in WS, there is a lack of studies examining multiple aspects of mathematical development and the abilities of preschoolers with WS (Van Herwegen & Simms, 2020). Likewise, previous studies used small sample sizes and/or they included participants across a wide age range.

Present Study

The present study aimed to provide a better understanding of the early mathematical abilities of 48-month-old children with WS and to determine the relation between early mathematical abilities and other cognitive abilities for these children. Two research questions concerning early mathematical abilities in 48-month-old children with WS were addressed:

1. What are the characteristics of early mathematical abilities of 48-month-old children with WS as measured by the Early Number Concepts subtest of the Differential Ability Scales–second edition (DAS-II; Elliott, 2007)?
2. What are the concurrent cognitive predictors of early mathematical abilities and how much of the variability in early mathematical abilities can be accounted for by individual differences in concurrent verbal, nonverbal reasoning, and spatial abilities?

Method

Participants

The participants were seventy-two 48-month-olds (Mean = 48.47 months, $SD = 0.28$, Range: 48.00–48.95, 43 boys) with genetically confirmed classic deletions of the WS region. All participants were native English speakers. All United States census regions were represented (22% Midwest, 19% Northeast 53% South, 6% West). The racial/ethnic distribution of the sample was 86% White non-Hispanic, 7% White Hispanic, 4% Multiracial non-Hispanic, 1% Asian non-Hispanic, and 1% African American non-Hispanic. Data collection began in December 2006 and ended in June 2021.

Measures

Differential Ability Scales–Second Edition (DAS-II)

The DAS-II Early Years Upper-Level Battery (DAS-II; Elliott, 2007) is a standardized assessment of cognitive and school readiness abilities, which is normed for children aged 3;6–8;11. The DAS-II, for 48-month-old children, is composed of 6 core subtests making up the three core clusters (Verbal, Nonverbal Reasoning, Spatial). The DAS-II also includes a diagnostic subtest measuring Early Number Concepts.

Verbal Ability. Verbal ability was measured by the DAS-II Verbal Cluster. This cluster is composed of two subtests: Verbal Comprehension and Naming Vocabulary. In the Verbal Comprehension subtest, the participants are required to follow instructions provided verbally by the examiner. This test measures understanding of syntax, knowledge of prepositional and relational concepts, and vocabulary knowledge. The Naming Vocabulary subtest measures concepts such as expressive language skills and vocabulary knowledge of nouns. A T-score for the Naming Vocabulary subtest and Verbal Comprehension subtest was obtained. The T-scores from each subtest are then summed to obtain a cluster SS which has a mean of 100 and a standard deviation of 15. The Verbal SS was used to measure verbal ability.

Nonverbal Reasoning Ability Nonverbal reasoning ability was measured by the DAS-II Nonverbal Reasoning Cluster, which includes two subtests: Pictures Similarities and Matrices. In the Pictures Similarities subtest, participants are required to match a card to one of four pictures presented in a stimulus book based on similarities. In the Matrices subtest, participants must find which alternative completes a pattern presented in a stimulus book. Performance was measured by the Nonverbal Reasoning SS.

Spatial Ability. Spatial ability was measured by the DAS-II Spatial Cluster. This cluster includes two subtests: Pattern Construction and Copying. For 48-month-olds of below average spatial ability (including all participants in this study), the Pattern Construction subtest involves the replication of three-dimensional patterns of increasing complexity that are first described or modeled by the examiner. In the Copying subtest, participants are required to copy on a blank sheet of paper a shape or pattern either drawn by the examiner or presented in a stimulus book. Performance was measured by the Spatial SS.

Early Mathematical Ability. Early mathematical ability was measured by the DAS-II Early Number Concepts (ENC) subtest. This subtest measures the child's development of quantitative concepts. Some of the number concepts and skills tested in this subtest include reciting by rote numbers up to 10, counting up to 10 objects using one-to-one correspondence, and making comparisons of set sizes. Other items include comprehension of relational concepts (e.g., quantitative or magnitude terms such as "few," "biggest"). Performance is measured by a T-score (mean = 50, $SD = 15$).

Procedure

The DAS-II was administered as part of the 48-month-old assessment for children with WS at the Neurodevelopment Sciences Laboratory at the University of Louisville. The DAS-II was administered and scored according to the standardized procedures by trained doctoral students or research assistants.

Data Analysis

Data were analyzed using IBM SPSS–Version 27. To characterize the early mathematical abilities of 48-month-old children with WS, descriptive statistics for each measure were obtained. An item frequency analysis for each item in the ENC subtest was performed. To investigate concurrent cognitive predictors of early mathematical ability, a multivariate linear regression was performed with ENC T-score as the dependent variable. The independent variables were the SSs for Verbal, Nonverbal Reasoning, and Spatial Clusters.

Results

Early Mathematical Skills as Measured by the DAS-II ENC Subtest

Item analysis was performed to delineate the mathematical abilities of the participants in this study. The items were split into three categories: items that assessed counting ability, number recognition and manipulation, and relational language.

Counting Ability

Table 1 shows the responses to item 3 on the DAS-II ENC. This item measures both the ability to recite by rote numbers up to 10 as well as one-to-one correspondence with pointing up to 10. As indicated in the table, half of the participants were able to count by rote to 10. However, only 8.3% of participants demonstrated understanding of one-to-one correspondence with pointing up to 10.

Table 1

Performance of Participants on Rote Counting and Counting Using One-to-One Correspondence While Pointing

Item	Highest Number Counted Correctly			
	0-2	3-5	6-9	10
Recite in correct order	18.1	13.9	18.1	50
Count with one-to-one correspondence	62.5	20.8	8.3	8.3

Note. $N = 72$.

Further analyses of performance on item 3 from the DAS-II ENC are presented in Figures 1 and 2. From Figure 1, it is evident that half of the participants were able to recite by rote up to the number 10. Figure 2 shows that only six participants were able to count objects in one-to-one correspondence with pointing up to 10. Twenty participants were unable to count with one-to-one correspondence while pointing, resulting in a score of zero. Therefore, almost 28% of the participants have no understanding of the principle of one-to-one correspondence. A large decline in the number of participants able to count with one-to-one correspondence is noted as numerals increase along the x-axis.

Figure 1

Histogram of Highest Number Recited in Correct Sequence by Rote

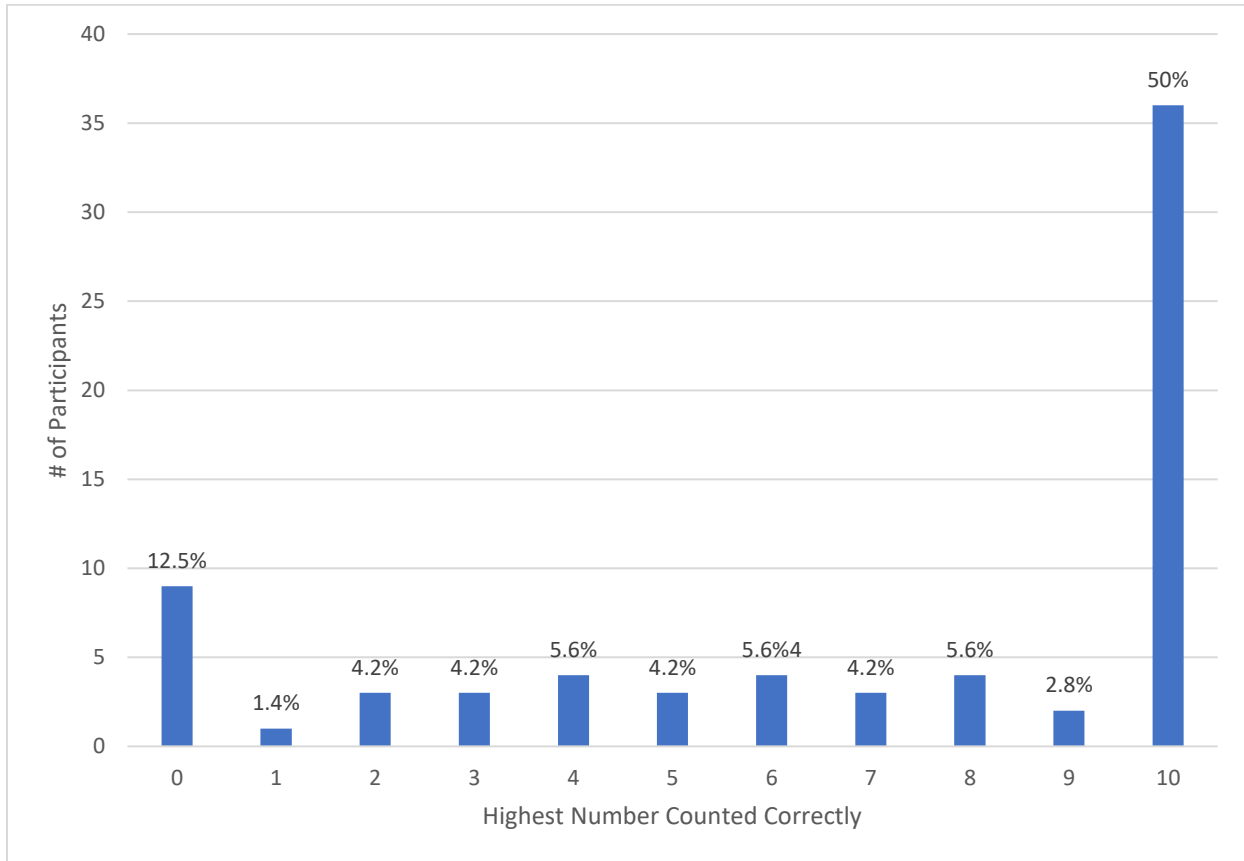
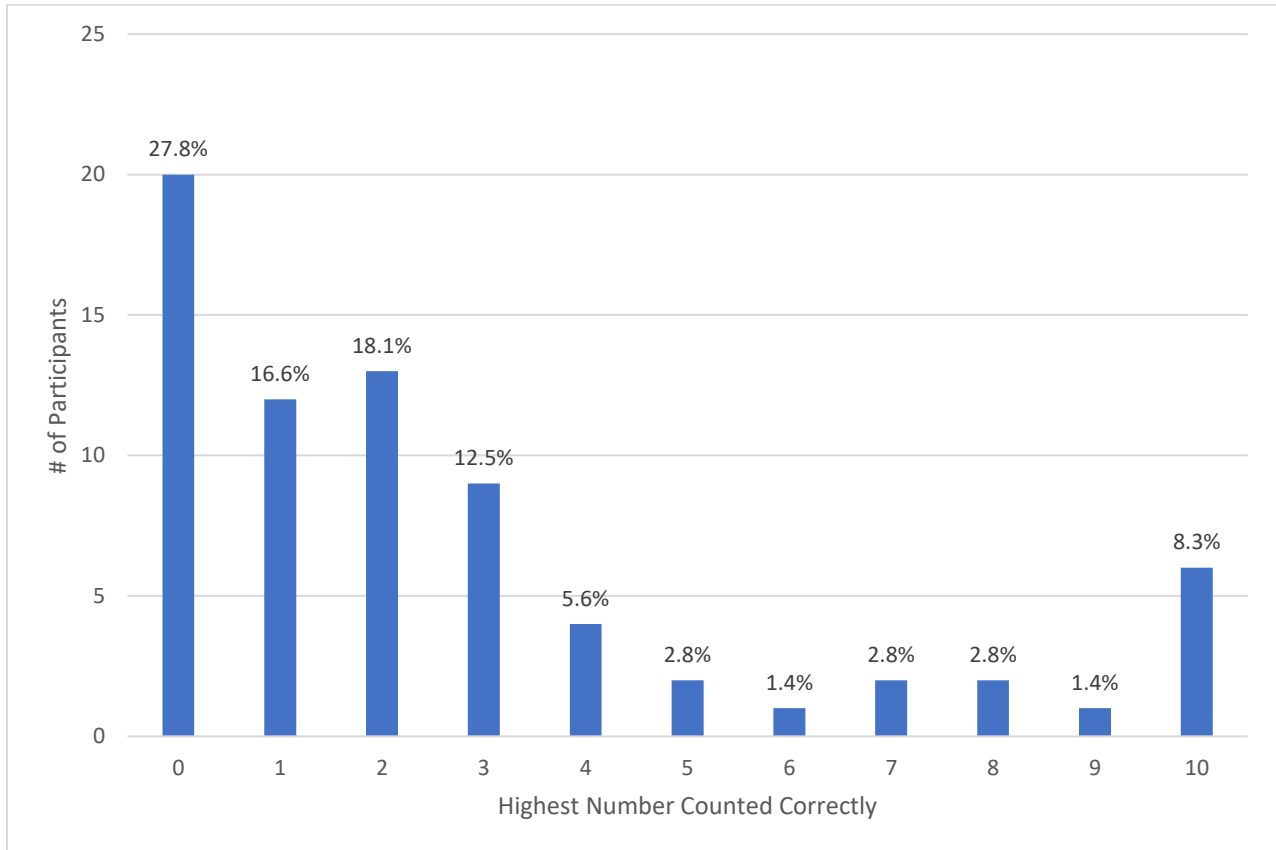


Figure 2

Histogram of Highest Number Correctly Counted to with One-to-One Correspondence



To determine the relation between counting by rote and counting with one-to-one correspondence while pointing, a bivariate Pearson correlation was calculated. This correlation was statistically significant ($r = 0.37, p < .01$).

Items Addressing Numerical Abilities

Table 2 provides the percentage of correct answers for items 1, 2, 11, 14, and 17–19 of the DAS-II ENC subtest. These items assess numerical abilities such as those that involve

manipulation of small numbers of objects, word problems assessing cardinality and addition, and recognizing the smallest and largest numbers in a set.

According to the table, 69.1% of participants demonstrated an understanding of the concept of “one” while only 41.7% indicated an understanding of the concept of “two.” A quarter of participants were able to pick out the image that showed no (or zero) items. Only one participant was able to answer a word problem that demonstrated understanding of addition and cardinality. No participants were able to pick out the smallest number in a set of 6 numbers and only one participant picked out the largest number in a set of 6 numbers.

Table 2

Percentage of correct answers on items assessing numerical abilities in the DAS-II Early

Number Concepts Subtest

Item Number and Ability Measured	Percent of participants who answered correctly
1. Understanding the concept of “one”	69.4
2. Understanding concept of “two”	41.7
11. Understanding concept of “no” items	25.0
14. Solving simple word problem requiring addition of small numbers and understanding of cardinality	0
17. Solving simple word problems requiring addition of small numbers and understanding of cardinality	1.4
18. Able to pick out the number that represents the smallest quantity in a set of 6 numbers	0
19. Able to pick out the number that represents the largest quantity of 6 numbers	1.4

Note. N=72

Items Addressing Relational Language

Table 3 provides the percent of correct answers for items 4–10, 12, 13, 15, and 16 of the DAS-II ENC subtest. These items assess understanding of relational language such as knowing which object is the largest in a set of objects.

More than half of the participants demonstrated an understanding of “big” and “little” indicating some comprehension of simple relational concepts. Most of the remaining relational concepts were identified correctly by fewer than a quarter of the participants.

Table 3

Percentage of correct answers on items assessing relational language in the DAS-II Early Number Concepts Subtest

Item Number and Ability Measured	Percent correct
4. Understanding concept of “big”	61.1
5. Understanding concept of “little”	56.9
6. Understanding concept of “largest”	22.2
7. Understanding concept of “smallest”	40.3
8. Understanding concept of “full”	22.2
9. Understanding concept of “empty”	44.4
10. Understanding concept of “many”	20.8
12. Understanding concept of “whole”	6.9
13. Understanding concept of “most”	4.2
15. Understanding concept of “few”	1.4
16. Understand concept of “least”	2.8

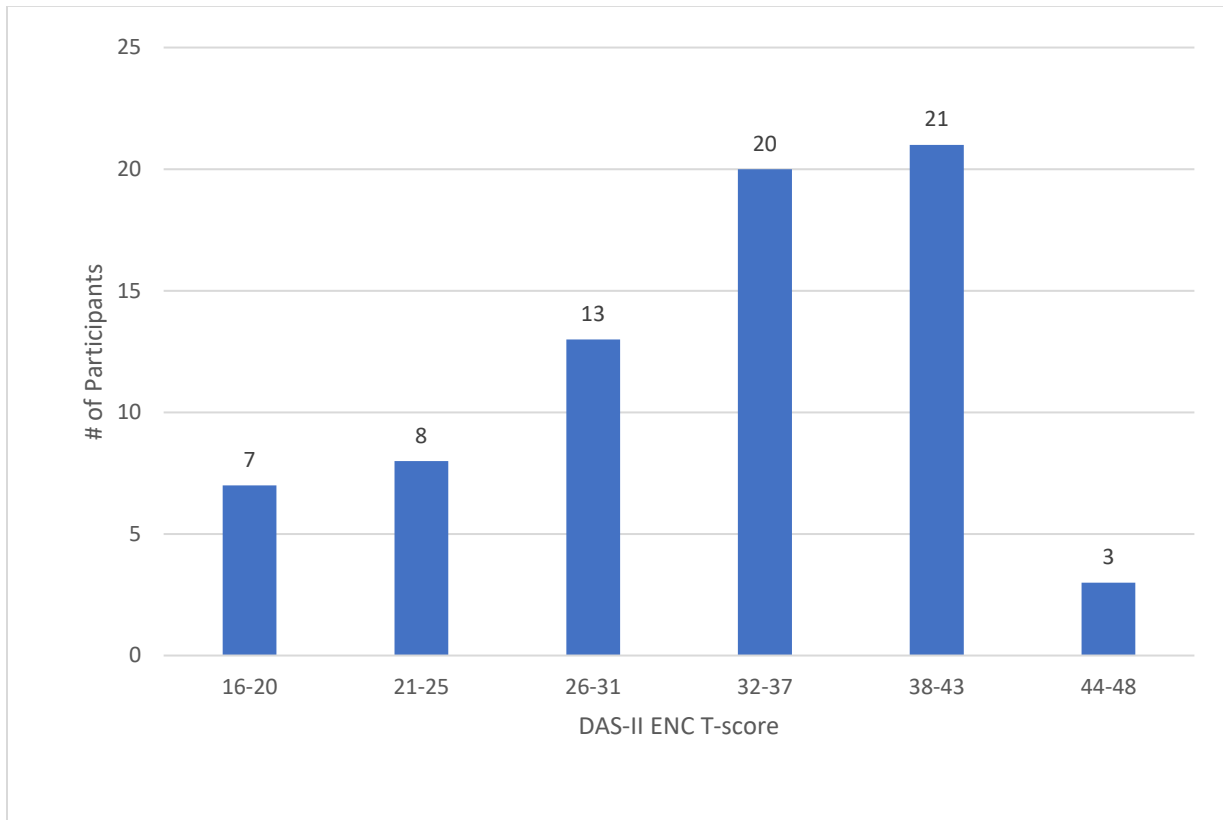
Note. N = 72

Concurrent Cognitive Predictors of Mathematical Abilities for 48-Month-Old Children with WS

A histogram of the ENC T-scores is shown in Figure 3. This histogram demonstrates that there was considerable variability in mathematical ability among the participants.

Figure 3

Histogram of DAS-II ENC T-Scores



Note. DAS-II = Differential Ability Scales, second edition; ENC = Early Number Concepts

A multiple regression analysis was conducted to determine the concurrent cognitive predictors of ENC T-scores. Descriptive statistics for the dependent and independent variables included in the analysis are presented in Table 4. A wide range of mathematical abilities, as

determined from the ENC T-scores, was noted. The DAS-II ENC T-score for seven individuals in the sample was 16, the lowest possible score for 48-month-olds, indicating that these children were unable to answer any item correctly. In contrast, one participant earned a T-score of 48 on the DAS-II ENC indicating that her mathematical abilities were close to the average for TD 48-month-olds ($T = 50$). However, no child was able to score above the average of TD 48-month-olds. The mean DAS-II ENC T-score for this sample of 48-month-olds with WS was 32.97 ($SD = 8.22$), which demonstrated that the average level of early number concepts ability for this sample was almost 2 SDs below the average for TD children in the same age group.

A wide range of DAS-II Verbal SSs was also noted, with values from 30 – 102 for the participants. One participant obtained a Verbal SS of 30, the lowest possible score for 48-month-olds. Two participants scored above the mean SS of 100 for the general population. These scores show the great variability of verbal abilities in children with WS. While still indicating delay compared to typically developing children, the mean score of DAS Verbal SS of the sample being 74.14 ($SD=16.60$) shows a relative strength in verbal ability compared to that of spatial ability; the mean Spatial SS was 55.96 ($SD=9.60$).

Table 4

Descriptive Statistics for Variables Included in the Multiple Regression Analysis

Measure	Mean	Median	SD	Range
DAS-II ENC T	32.97	34.00	8.22	16* – 48
DAS-II Verbal SS	74.14	77.00	16.60	30* – 102
DAS-II Nonverbal Reasoning SS	75.82	76.50	14.96	41 – 106
DAS-II Spatial SS	55.96	55.00	9.60	41 – 85

Note. $N = 72$; *lowest possible score for 48-month-olds; DAS-II = Differential Ability Scales, second edition; ENC = Early Number Concepts; SS = standard score; T = T-score.

Bivariate Pearson correlations among the measures included in the multiple regression were computed (see Table 5). All measures were significantly correlated ($p < .001$). The ENC T-score and the DAS-II Verbal SS showed the strongest significant correlation among the variables tested ($r = .83$).

Table 5

Bivariate Correlations among the Measures Included in the Multiple Regression Analysis

Measure	2	3	4
1. DAS-II Verbal SS	.70	.55	.83
2. DAS-II Nonverbal Reasoning SS		.53	.62
3. DAS-II Spatial SS			.48
4. DAS-II ENC T			–

Note. N = 72; DAS-II = Differential Ability Scales, second edition; ENC = Early Number Concepts; SS = standard score; T = T-score. All *ps* < .001

The multiple regression analysis is shown in Table 6. The DAS-II Verbal SS, DAS-II Nonverbal Reasoning SS, and DAS-II Spatial SS were used as predictors of mathematical ability, as measured by the DAS-II ENC T-score. A preliminary analysis was performed to confirm there was no violation of the assumptions of normality. The results of the multiple linear regression revealed that nonverbal reasoning ability and spatial ability were not statistically significant predictors in the model. However, the analysis revealed verbal ability to be a significant predictor of mathematical ability. A significant regression equation was found ($F(3,68) = 49.82, p < .001$), with an R^2 of .69 and adjusted R^2 of .67. The regression coefficient ($b=0.38, 95\% \text{ CI } [.28, .48] p < .001$) indicated that with each additional 1 SS-point increase in DAS-II Verbal SS, there is an increase of approximately 0.38 T-score point in DAS-II ENC, after controlling for Nonverbal Reasoning SS and Spatial SS. DAS-II Verbal SS uniquely accounted for 27% of the variability on DAS-II ENC. Verbal ability had a large effect with a Cohen’s f^2 value of 2.19.

Table 6

Multiple Regression Analysis Predicting DAS-II Early Number Concepts T-score

	<i>B</i>	<i>t</i>	<i>p</i>	95% CI for <i>B</i>	<i>semi-partial r</i>	Cohen's <i>f</i> ²
Constant	33.97	59.58	<.001	[31.87, 34.08]		2.22
DAS-II Verbal SS	0.38	7.71	<.001	[.28, .48]	.52	2.19
DAS-II Nonverbal Reasoning SS	0.04	0.68	.50	[-.07, .14]	.05	<0.01
DAS-II Spatial SS	0.03	0.37	.72	[-.12, .17]	.03	<0.01

Note. *N* = 72. All continuous independent variables were centered on their respective sample means. DAS-II = Differential Ability Scales, second edition; SS = standard score; CI = confidence interval

Discussion

The purpose of this study was to describe the mathematical abilities of 48-month-old children with WS and to determine the concurrent cognitive predictors of early mathematical abilities for these children. The Discussion will focus on addressing both aims of the study, comparing the current results to results of prior studies, discussing the implications of these findings, explaining limitations of the present study, and offering direction for future research.

Mathematical Abilities of 48-month-olds with Williams Syndrome

The first aim of the study was to determine the characteristics of early mathematical abilities of 48-month-old children with WS. Throughout the study, no child gave non-numerical word answers when asked questions requiring numerical answers. This is similar to findings for TD children, as seen in Fuson (1988), and signifies that the participants understand that number

words are different from other words in the English language and are used to answer questions pertaining to counting and numerosity. The data showed that half of the participants were able to count by rote to 10, a skill learned through rote memorization. These findings are in agreement with previous research by Ansari and colleagues (2003), who reported the ability of children with WS to recite and count small numbers with little error. However, while half of the participants were able to count to 10 using rote memorization, the majority of participants was not able to count objects in one-to-one correspondence with pointing over the number two, a skill most TD children have mastered before the age of 4 years (Clements & Sarama, 2010; Frydman & Bryant, 1988).

The data showed a large percentage of the participants were able to understand and manipulate the quantity of one, but there was a notable decrease in the number of participants able to understand and manipulate quantities of two. Two previous studies reported their participants with WS, who were several years older than the participants in the current study, to perform well on questions dealing with small number recognition and manipulation using objects (Ansari et al., 2003; O’Hearn & Landau, 2007). This suggests that development of this ability may begin around 48-months-old but continues to develop for years after. No participant was able to pick out the smallest number in a set and only one was able to pick out the largest number in a set.

Greater than half of the participants were able to compare two objects in terms of size when asked which one was the little or big item. However, fewer than half were able to compare more than two items in terms of size. This suggests that 4-year-olds with WS have difficulty with comparison when more than two objects are available but perform relatively well on questions regarding only two objects of varying size. This is consistent with the order of acquisition seen in

TD children in which understanding of simple terms (e.g., big, little) develops earlier than comparative and superlative forms of the same terms (Edmonston & Thane, 1999). However, because the participants completing items that compare only two objects have a 50% chance of guessing the correct answer and items that compare more than two items have less of a chance that the child will guess the correct answer, the difference seen in the results between these items may be due in part because of the difference in chance and not because of true understanding of the words. The participants also struggled with questions involving relational words such as “few”, “least” and “many”. These findings suggest difficulty comparing quantities and size of items, which is consistent with findings by Mervis and John (2010) where they indicated great difficulty with relational/conceptual vocabulary in 5–7-year-olds with WS. Therefore, it is reasonable to conclude that the problem with relational language persists even in older children with WS.

Lastly, it is evident from the data that the participants were better at answering the earlier questions in the subtest and had more difficulty with the later questions. Because standardized assessments present items in order of difficulty, these findings indicate that the order of difficulty of the concepts included in this subtest is similar for children with WS and children in the general population, findings consistent with O’Hearn and Landau (2007).

Concurrent Cognitive Predictors of Early Mathematical Abilities of 48-Month-Old Children with WS

The second aim of the present study was to determine if concurrent verbal, nonverbal reasoning, or spatial abilities predicted mathematical ability at 48 months for children with WS. Verbal, Nonverbal Reasoning, and Spatial SSs were significantly positively correlated with each other as well as with Early Number Concepts T-scores. These significant correlations were

expected, based on Meyer-Lindenberg, Mervis, and Berman's (2006) review of prior studies that emphasized the interdependence of multiple cognitive abilities for individuals with WS. These findings argue against the modularity of mind hypothesis, which suggests that the mind is made up of neural modules that have distinct neural functions. The results of the multiple regression analysis indicated that of the three cognitive abilities measured, only verbal ability accounted for a significant amount of unique variance in mathematical skills. The significance of verbal ability in predicting mathematic ability is consistent with previous findings that suggest that some mathematical abilities are related to verbal ability in individuals with WS (O'Hearn & Landua, 2007; Ansari, 2003).

Ansari and colleagues (2003) suggest that the visuospatial impairments characteristic of the WS cognitive profile prevent normal understanding of cardinality such that the relative strength in language did not fully compensate. The findings of the current study may further support this claim in that relative weakness in spatial ability may hinder the development of mathematical concepts causing an overall impairment of the subject. The findings in the present study show that nonverbal reasoning ability is not a significant predictor of unique variance in mathematical ability in 48-month-olds with WS. However, nonverbal reasoning ability was significantly correlated with early mathematical ability so may contribute unique variance to more advanced mathematical abilities in children with WS.

Implications

The present study provides a valuable description of the mathematical abilities of young children with WS. These findings are beneficial to determine interventional goals for preschool education for children with WS. This study determined that 48-month-old children with WS performed well on questions involving rote counting up to 10, understanding the quantity of one,

and comparisons of two objects based on size. However, 48-month-olds with WS performed poorly on questions involving one-to-one correspondence and comparisons of more than two objects regarding size or quantity. These findings suggest that children with WS may need additional support with some components of mathematics, such as counting with one-to-one correspondence, cardinality, and relational concepts related to quantity.

This study determined that verbal ability is a significant concurrent predictor of mathematical ability in 48-month-old children with WS. Educational professionals may want to employ this information to tailor individualized approaches to teaching mathematics that include verbal strategies so children with WS can better learn and further develop mathematical skills. While spatial and nonverbal reasoning ability were not significant predictors of unique variance in mathematical ability, they did correlate significantly with mathematical ability in the bivariate correlation analysis. Therefore, it may also be important to allocate support in the development of spatial and nonverbal abilities as this may benefit the development of various mathematical skills.

Limitations and Directions for Future Study

As with all research, the design of the current study is subject to limitations. The rareness in occurrence for WS limited the diversity of the participant sample. Of the 72 participants in the study, 93% were White non-Hispanic, and only 6% of the participants came from the western region of the United States.

Additional research is necessary to characterize the mathematical abilities as well as to determine the influences of verbal ability on mathematical abilities in preschool-aged children with WS. Furthermore, a lack of available data on the external predictors, such as math language usage in the household, of mathematical abilities in children with WS limited the scope of this

study. Investigations into external predictors of mathematical abilities would contribute to the understanding of the development of mathematical skills in individuals with WS. Further research into the significant predictors of each individual mathematical skill may also help to promote understanding of which cognitive abilities predict ability in which individual component of mathematics. Therefore, further research that analyzes early mathematical skills in children with WS is necessary. In addition, research is needed to compare the overall mathematical abilities and their development in children with WS to other groups, including children with other neurodevelopmental disorders.

Lastly, it is important to note that significant correlations do not imply causal relations between variables in a correlation analysis. An experimental study design would be necessary to determine if verbal ability and certain mathematical abilities rely on similar processes. This type of study is critical if we are to begin to understand the causal relations in the development of mathematical abilities in children with WS.

Conclusion

Considerable variability is present in mathematical abilities among 48-month-old children with WS, ranging from very limited mathematical abilities to average abilities seen in TD populations of the same age group. Certain mathematical abilities were more advanced in children with WS than others, but the general pattern of development was consistent with TD populations. Furthermore, verbal ability and mathematical ability are very highly correlated and verbal ability is a significant unique predictor of mathematical ability in the population studied. The findings of this study may contribute to the understanding of the development of mathematical abilities in people with WS and support the development of interventional techniques that utilize verbal abilities when teaching mathematical concepts.

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