Published in final edited form as:

J Intellect Disabil Res. 2012 October ; 56(10): 974–983. doi:10.1111/j.1365-2788.2012.01578.x.

Language development in school-age females with fragile X syndrome

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Abstract

Background—Females with fragile X syndrome (FXS) have a wide range of cognitive and language abilities. The range of language outcomes experienced by females with FXS, however, has been relatively unexplored. The purpose of this exploratory study was to examine receptive and expressive language, with a focus on vocabulary and syntax, in a group of school-age females with FXS.

Method—Twenty-one girls with FXS ages 7 to 15 participated in the study. The girls completed a receptive vocabulary test, nonverbal IQ test, and an expressive language sample.

Results—The mean IQ for this group of females was at the cutoff for intellectual disability. Vocabulary was an area of strength relative to nonverbal cognition. Age and nonverbal IQ were significant predictors of vocabulary performance.

Conclusions—The data suggest that a substantial portion of the sample would qualify for speech and language services. This study highlights the need for continued research in the area of language and cognitive development in girls with the full mutation of fragile X.

Fragile X syndrome (FXS) is the most common inherited cause of intellectual disability, affecting approximately 1 in every 3600 males and 1 in every 4000 to 6000 females although recent estimates suggest an even higher prevalence (Crawford et al., 2001; Fernandez-Carvajal, Walichiewicz, Xiaosen, Pan, Hagerman, & Tassone, 2009; Hagerman, 2008). The syndrome is caused by the excessive lengthening of a trinucleotide (CGG) repeat in the *FMR1* gene on the X chromosome (Verkerk et al., 1991), which in turn leads to a reduction or absence of the protein (FMRP) that is normally produced by that gene (Darnell, Warren, & Warren, 2004). Language and cognitive delays are virtually universal in males with FXS (Abbeduto, Brady, & Kover, 2007). Females with FXS are less affected on average and display a wider range of behavioral and neurocognitive outcomes than affected males, with some females functioning in the intellectually average range (Keysor & Mazzocco, 2002; Mazzocco, 2001). The range of language outcomes experienced by females with FXS, however, has been relatively unexplored (Abbeduto et al., 2007; Hagerman, 2002; Hatton, Wheeler, Sideris, Sullivan et al., 2009; Murphy & Abbeduto, 2003). The present study was designed to provide data on the range of language outcomes characteristic of females with FXS. Such data will be a useful foundation for further research designed to test specific hypotheses about the nature, correlates, and causes of language impairments in affected females.

There is evidence that, as a group, females with FXS have impairments in language. Mazzocco and colleagues (2006) found that in comparison to girls with typical development, girls with FXS asked fewer questions, including fewer questions that logically followed

from the previous utterance. In a study of adult women, Simon et al. (2001) found that females with the full mutation had difficulty in selecting appropriate humorous endings for stories that they read relative to IQ-matched women without FXS. Simon et al. concluded that the females with FXS failed the task because they were unable to follow and make connections between the elements and propositions of a discourse (i.e., establish coherence).

These studies, however, were limited by their reliance on a single measure of language. Language is not a unitary ability and thus, different aspects of language can be impaired to varying degrees across and even within neurodevelopmental disorders. In Down syndrome, for example, syntax, especially expressive syntax, is more severely impaired than is vocabulary (Chapman, 2003). In contrast, syntax, although impaired, is a relative strength in individuals with Williams syndrome (Mervis, 2009). In autism, subtypes with different profiles of relative strengths and weaknesses in language have been identified (Tager-Flusberg & Joseph, 2003). Such asynchronies across language have implications for clinical assessment and intervention and provide clues as to the nature of the learning mechanisms that are impaired in these disorders (Abbeduto, Evans, & Dolan, 2001). There is a need, therefore, to examine multiple dimensions of language within the same samples of females with FXS.

Preliminary data suggest that females with FXS also manifest a profile of linguistic strengths and weaknesses. In a study focused on verbal perseveration, Murphy and Abbeduto (2007) found that males with FXS produced conversational device repetitions (e.g., “that’s a wrap” and “that’s it”) significantly more often than did females with FXS, although gender differences did not emerge for other types of perseveration (e.g., repetitions at the utterance level or topic repetitions). Such findings suggest that females with FXS may display a more nuanced profile of relative strengths and weaknesses, with some aspects of language as impaired as in their male counterparts. The small sample of females in this study ($n = 8$), however, makes firm conclusions impossible. The present study was designed to examine multiple dimensions of language in females with FXs, using a larger sample.

In the study of several neurodevelopmental disorders, it is the distinction between lexical and syntactic development that has proven to be useful in defining the linguistic phenotype (e.g., Down syndrome and Williams syndrome). Unfortunately, there have been virtually no studies focused on the lexical or syntactic capabilities of females with FXS. In a study of adolescents and young adults, Abbeduto et al. (2003) reported that females were not as delayed as males in terms of receptive vocabulary or receptive syntax. The females in the Abbeduto et al. study, however, were quite impaired in terms of nonverbal IQ (mean score of 55.5) and not representative of the full range of affectedness. Moreover, the sample of females was quite small ($n = 6$). In the present study, we recruited a larger sample of females who were representative of the full range of affectedness and focused on the dimensions of vocabulary and syntax.

It also is not clear what factors account for within-syndrome variation in linguistic outcomes among females with FXS (Murphy & Abbeduto, 2003). Research with boys with FXS has indicated that receptive and expressive vocabulary scores and syntax are strongly related to cognitive developmental level (Abbeduto et al., 2003; Roberts et al., 2007). This relationship also has been found for speech production in males with FXS (Roberts et al., 2005). Studies of this relationship in females, however, have yielded inconsistent findings, with some studies finding no relationship (Abbeduto et al., 2003; Murphy & Abbeduto, 2007). Interpreting these findings for females is difficult, however, because different dimensions of language have been measured across multiple studies instead of within the same group, and the studies have involved small samples comprised of only females with intellectual

disabilities. In the present study, we examined the relationship for both vocabulary and syntax and with a larger and more representative sample of females with FXS.

The purpose of this study was to examine receptive and expressive language, with a focus on receptive vocabulary and expressive syntax, in a group of school-age females with FXS. The first goal was to outline the range of impairments in receptive vocabulary and syntax that is typical for females with FXS. The second goal was to determine whether cognitive ability predicts receptive vocabulary and expressive syntax in this population.

Method

Participants

The participants were 21 females with the FMR1 full mutation. The diagnosis was confirmed by molecular genetic testing for all participants. The participants were drawn from two cohorts: the first author's dissertation study and a large, longitudinal study focused on the language development of children with FXS (McDuffie et al., 2010; McDuffie et al., in press). Although previous publications have emerged from these projects, none has focused exclusively on females and these measures. The girls were native English speakers according to parent report. Additionally, all were reported by parents as using multi-word phrase speech with some regularity. The girls ranged in age from 7 to 15 years, with a mean age of 11 years, 8 months. The participants for the two projects were recruited nationally via postings to Internet listservs and websites, mailings to professionals, attendance at national and/or regional parent meetings, advertisements on nationally syndicated radio shows and in newspapers in selected urban areas, and through a university registry of families with children with developmental disabilities.

Procedures

The assessments for most participants were completed in a university laboratory setting, but some participants were assessed in their homes. After obtaining written informed consent from the legal guardian, and oral assent from the child, a number of standardized tests and an expressive language sample were completed. The assessments reported on in this paper were part of more comprehensive protocols from each of the projects from which they were drawn. The assessment was videotaped using a video recorder. Participants were given breaks between testing as needed. The assessments were completed over a one to two day period, depending on the child's needs. See McDuffie et al. (2010; in press) for more information about the protocols.

Standardized Tests

Receptive vocabulary—The *Peabody Picture Vocabulary Test, Fourth Edition* (PPVT-IV; Dunn & Dunn, 2007) or the *Peabody Picture Vocabulary Test, Third Edition* (PPVT-III; Dunn & Dunn, 1997) was administered (Because the data were collected over several years, we began with the PPVT-III and switched to the PPVT-IV when it became available; reliability between two instruments reported at .84). In the standardized test, participants are asked to point to one drawing from among several alternatives that depicted the word spoken by the examiner. The mean for this test is 100, with a standard deviation of 15. Scores between 70 and 85 are considered borderline impairment, and scores below 70 are considered impaired. Although children seldom qualify for language-related services in schools or health care settings on the basis of a single behavioral measure or standardized test, scores under 85 (i.e., one standard deviation from the mean) would raise concerns and, if corroborated by other tests and sources of information, would likely lead to services.

Nonverbal intelligence—The Brief IQ subtests of the *Leiter Test of Nonverbal Intelligence-Revised* (Leiter-R Roid & Miller, 1997) served as the measure of nonverbal cognition. The four subtests comprising the Brief IQ composite are: Figure Ground, Form Completion, Sequential Order, and Repeated Patterns. In these subtests, individuals are asked to find an item in a picture, choose the next item within a sequence, or arrange items in a pattern, respectively. The mean for this test is 100, with a standard deviation of 15. Scores of 70 and below (i.e., two standard deviations below the mean) are considered in the range of intellectual disability, and, if corroborated by information about concomitant impairments in adaptive functioning, would likely lead to a diagnosis and qualifications for special education services (American Association on Intellectual and Developmental Disabilities, 2010). One participant refused to complete all four subtests.

Expressive syntax—An expressive language sample was completed with each participant in order to evaluate expressive syntactic abilities. The language sample involved an interview-style conversation designed to elicit spontaneous speech. Each participant talked with a female examiner for 10 to 15 minutes. In general, the examiner asked open-ended questions and avoided yes-no questions, while trying to elicit as much talk from the participant as possible and keeping her own talk to a minimum. Standardization was ensured across language samples by the use of a script of topic introductions and follow-up questions that were likely to be of interest to all participants. Language samples were audiotaped. Mean length of utterance (MLU) was computed in morphemes as described in Brown (1973). Based on Brown's stages (1973) we would expect all participants to have a minimum MLU of 4.5. By the age of 5, children with typical development have complex expressive syntax at an adult-like level.

Coding and reliability procedures—The standardized tests were scored on-line, with accuracy of administration and scoring double checked independently by a second examiner who reviewed the originally scored protocol. Data entry was also double-checked. The audiotaped language samples were transcribed using Systematic Analysis of Language Transcripts (SALT; Miller & Chapman, 2000) according to typical child language research conventions. For each language sample, a second trained transcriber listened to the tape and marked changes or perceived discrepancies on the typed transcript. The original transcriber then verified or updated the transcript before the data were analyzed. Inter-transcriber agreement was good across contexts and groups with overall agreement of 90%.

Results

Range of Impairments

The first research question focused on the range of impairment in school-age females with full mutation FXS. We addressed this question through a number of descriptive statistics designed to reflect group tendencies and variability. We found a broad range of outcomes in terms of both cognitive and language variables. See Table 1 for more information. Nine of the girls in this sample (45%) had scores below 70 on the Leiter-R Brief IQ, which would be consistent with a diagnosis of intellectual disability (see Figure 1). Four girls scored 3 standard deviations or more below the mean. Eight girls (40%) had scores between 70 and 83, which is more than one standard deviation below the mean. Thus, only 15% of the sample scored better than 85 (i.e., less than 1 SD below the mean).

As a group, the girls achieved significantly higher scores on the measure of receptive vocabulary compared to the Leiter-R findings based on a matched pairs t-test (see Figure 2, and Table 1). Nevertheless, there was considerable variability in PPVT scores. Three of the girls (14%) had scores below 70 (2 standard deviations below the mean), and another 3 girls

were in the borderline range for language impairment (1 to 2 standard deviations below the mean).

In terms of expressive syntactic complexity, only 2 girls scored lower than age expectations (Figure 3). MLUs ranged from 4 to 8.75, indicating that many girls were using syntactically complex language in conversation. Some girls, however, were quite delayed as evidenced by the fact that by 4 years of age, typically developing children achieve an MLU of 4.5 (Brown, 1973). Based on age norms available from the SALT software (Miller & Chapman, 2000), mean MLU for children with typical development in the age range of our study is 6.07. There were two girls with MLUs lower than 4.5; one girl had an MLU of 4.0 (age 13 years 3 months) scored 1.33 standard deviations below the mean for her age. The second participant (age 11 years 4 months) had a mean MLU of 4.2, which was 1.3 standard deviations below the mean.

Predictors of Vocabulary and Syntax

The second research question concerned the relationship between language and cognition. Multiple linear regression analysis was used to examine the relationship between age and nonverbal IQ and the measures of receptive vocabulary and expressive syntax. The dependent variables were PPVT raw scores (receptive vocabulary) and MLU (expressive syntactic complexity), with each examined in separate regression equations. We selected raw scores over standardized scores for the PPVT as our dependent variable; this was due to the fact that raw scores are more sensitive to improvement in abilities than standard scores, which have tend to have a relative high “floor” for the population persons with intellectual disabilities. In terms of receptive vocabulary, chronological age and IQ were significant predictors of PPVT raw scores (see Table 2). Together, these predictors accounted for 29% of the variance in PPVT raw scores. Both chronological age and nonverbal IQ were significant and unique predictors in the final model, and positively related to PPVT scores.

The same regression model was run for expressive syntactic complexity. In this analysis, however, the independent variables of age and nonverbal IQ did not account for a significant amount of variance in MLU (see Table 3).

Discussion

The girls with FXS in this study displayed a broad range of language and cognitive abilities. The mean nonverbal IQ for this group of females was at the cutoff for intellectual disability, with almost half achieving nonverbal IQs consistent with an intellectual disability and a few even scoring more than three standard deviations below the mean for the test (i.e., in the range of moderate intellectual disability). At the same time, however, three females achieved nonverbal IQs in the typical range. Thus, the broad net we cast in terms of recruitment sources appears to have been successful in sampling the full range of affectedness in females with FXS.

The first goal of the study was to provide data on the range of impairment characteristic for females with FXS in the domains of receptive vocabulary and expressive syntax. It was found that receptive vocabulary was an area of strength relative to nonverbal cognition for females with FXS on average. Nevertheless, there was a wide range of affectedness in receptive vocabulary, with 40% of females scoring well below the cutoff for language impairment (i.e., more than one standard deviation below the mean, mean of 100). Thus, speech-language clinicians should carefully assess receptive vocabulary in any female with FXS to determine the need for intervention.

In terms of expressive syntactic abilities, again, a wide range of abilities characterized the sample. In general, MLU was near expectations based on chronological age for many females in our sample, suggesting that expressive syntax is not an area of great challenge for them. At the same time, however, 10% of the females in our sample achieved MLUs more typical for a preschooler despite a mean chronological age near 12 and lower bound of 7 years. Again, speech-language clinicians should be cognizant of the possibility of expressive syntactic delays in some girls with FXS on their caseloads, although for girls with FXS expressive syntax is not an area requiring thorough assessment or focused intervention.

The second goal of the study was to examine the relationship between nonverbal cognitive ability and language, which we did by including both chronological age and nonverbal IQ as predictors of vocabulary score and MLU. We found that chronological age and nonverbal IQ were significant predictors and together accounted for 29% of the variance in receptive vocabulary scores. This finding is most parsimoniously interpreted as suggesting that not only FXS status, but improved cognitive skills and experience in the world facilitate vocabulary learning. An important next step will be to determine more precisely the specific cognitive achievements and experiences that facilitate that learning. It will also be important to determine the factors that contribute to vocabulary learning in females with FXS beyond what is expected relative to their age and cognitive skills. Such factors are likely to include biological variables, such as FMRP levels and background genes. It is also likely, however, that more proximal organismic variables, such as level of social anxiety will be important, as will a range of experiences, from home to school. Identifying such factors will be important for determining potential routes to intervene in order to optimize receptive vocabulary in this population. In the interim, our results suggest that targeting cognitive skills in addition to vocabulary would be wise.

In the case of our measure of syntactic complexity, neither chronological age nor nonverbal IQ was a significant predictor of MLU. This null finding highlights the pressing need for additional research to identify that factors that are predictors of this dimension of language ability. Such data are critical for developing effective interventions targeting expressive syntactic outcomes. Our results do not provide reason to focus on cognitive precursors of syntactic learning for our population.

Despite the generally high means for our language variables, it is important to recognize that more than half of the participants in our study would qualify for services in a school setting based on their IQs alone. Moreover, our data suggest that a substantial portion of the sample would also qualify for speech and language services. In short, this study highlights the need for continued research in the area of language and cognitive development in girls with the full mutation. It also suggests, however, that language intervention may be less of a need for girls with FXS, on average, than are specific cognitive skills.

An important limitation of the present study is the lack of data on the mutation status of our participants (beyond that of the status of full mutation). Including information of FMRP levels and activation ratios may have helped to explain additional variance in our language measures. It is important to note, however, that the link between these biomarkers and cognitive abilities has not been especially strong. Some studies have noted relationships between FMRP and IQ, attention, and processing speed in females (Loesch et al., 2002), whereas others have not found predictive relationships or found these biomarkers to explain less variance than other more proximal individual-psychological variables (e.g., Bailey, Hatton, Tassone, Skinner, & Taylor, 2001). It will be important in the future to examine not only direct but also indirect relationships between FMRP and other biomarkers and behavioral outcomes. It may be, for example, that the relationship between FMRP and language is mediated by IQ. Additionally, the inclusion of other groups of females (e.g.,

girls with Autism spectrum disorders) matched on nonverbal IQ would allow for a more in-depth analysis of the unique impact of the FMR1 gene on cognitive and linguistic development. It would also provide much needed cross-syndrome comparisons which are particularly useful in creating clinical interventions and assessments.

Although the sample size is relatively large for a behavioral study of females with FXS, our power to detect small but potentially important differences, was limited by our sample size and the inherent behavioral variability among females. Nevertheless, ours is one of the larger samples of females with FXS in which language has been examined in any detail. Second, although we improved on previous studies by examining two distinct domains of language – vocabulary and syntax -- there is a need for attention to additional areas of the linguistic system, such as phonology and pragmatics, so as to gain a complete look at the language phenotype of females.

Acknowledgments

This research is supported in part by grants R01 HD024356, P30 HD03352, P30 HD002528-39, and T32 HD07489 from NICHD, as well as T32 DC000052 from NIDCD and a Rosen Summer Student Fellowship from the National Fragile X Foundation.

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Figure 1.
Range of scores for nonverbal IQ; based on the brief composite of the Brief IQ subtests from the Leiter Test of Nonverbal Intelligence-Revised (Leiter-R; Roid & Miller, 1997).

Figure 2.
Range of standardized scores for receptive vocabulary, as measured by the Peabody Picture Vocabulary Test (PPVT; Dunn & Dunn, 1997, 2007).

Figure 3.

Range of syntactic complexity based on a language sample involving an interview-style conversation designed to elicit spontaneous speech. Samples were transcribed using Systematic Analysis of Language Transcripts (SALT; Miller & Chapman, 2000) and MLU was calculated from SALT.

Table 1

Participant standardized test and expressive language sample information

	Chronological Age	PPVT¹	MLU²	Leiter Brief IQ¹
Mean	11;8	89.33 *	6.28	70.40
SD	1.82	13.75	1.23	16.23
Range	7–15;5	59–106	4.00–8.75	46–100

Note.

¹ Standard scores.² Measured in morphemes.

* p<.05PPVT standard scores significantly different from Leiter brief IQ scores based on a matched-pairs t-test.

Table 2

Predictors of PPVT raw scores

	<i>B</i>	<i>SE B</i>	β	95% CI
Constant	-16.24	48.63		[-118.85, 86.36]
Age	8.52	3.42	.48 [*]	[1.31, 15.73]
Nonverbal IQ	.67	.32	.41 [*]	[0, 1.34]
Adjusted R^2	.29			
<i>F</i>	4.92 [*]			
<i>df</i>	2, 19			

Note.

^{*}
 $p < .05$.^{**}
 $p < .01$.

Table 3

Predictors of syntactic complexity

	<i>B</i>	<i>SE B</i>	β	95% CI
Constant	7.03	2.48		[1.81, 12.25]
Age	-.20	.17	-.26	[-.57, .16]
Nonverbal IQ	.03	.02	.34	[-.01, .06]
Adjusted R^2	.10			
<i>F</i>	2.08			
<i>df</i>	2, 19			

Note.

*
 $p < .05$.

**
 $p < .01$.