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Journal

Developmental Neurorehabilitation, 21(1)

ISSN

1751-8423

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Publication Date

2018-01-02

DOI

10.1080/17518423.2017.1369189

Peer reviewed

HHS Public Access

Author manuscript

Dev Neurorehabil. Author manuscript; available in PMC 2019 January 01.

Published in final edited form as:

Dev Neurorehabil. 2018 January ; 21(1): 48–63. doi:10.1080/17518423.2017.1369189.

Distance delivery of a spoken language intervention for school-aged and adolescent boys with fragile X syndrome

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Abstract

A small randomized group design ($N = 20$) was used to examine a parent-implemented intervention designed to improve the spoken language skills of school-aged and adolescent boys with FXS, the leading cause of inherited intellectual disability. The intervention was implemented by speech-language pathologists who used distance video-teleconferencing to deliver the intervention. The intervention taught mothers to use a set of language facilitation strategies while interacting with their children in the context of shared story-telling. Treatment group mothers significantly improved their use of the targeted intervention strategies. Children in the treatment group increased the duration of engagement in the shared story-telling activity as well as use of utterances that maintained the topic of the story. Children also showed increases in lexical diversity, but not in grammatical complexity.

Keywords

Distance teleconferencing; expressive language sampling; narrative storytelling; parent-implemented intervention

Language is learned best within the context of spoken interactions between children and their more skilled conversational partners,[1,2] especially when these partners are verbally responsive and provide high quality spoken language input.[3,4] Shared story-telling is especially well suited for providing children with such input.[5] In fact, there is considerable evidence that shared story-telling promotes language acquisition,[6] especially when the child and adult collaborate to jointly construct a book's meaning.[7] Over the course of repeated shared story-telling interactions, the adult partner can provide ongoing opportunities for reciprocal exchanges that move the child from being a passive listener to an active

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Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

No other authors have financial disclosures or declarations of interests to report.

participant in talking about the story (e.g., dialogic book reading).[8] The current study reports on a small-scale, randomized controlled trial designed to examine the efficacy of an interactive, shared story-telling intervention for boys with fragile X syndrome (FXS), ranging from 10- to 16-years of age.

Genotype and phenotype of FXS

FXS, the most common inherited cause of intellectual disability,[9] is caused by a mutation in a gene (*FMR1*) on the X chromosome.[10] In the full mutation, a repetitive CGG trinucleotide sequence expands to 200 or more repeats when inherited from a mother who is a carrier or who has the full mutation herself. This expansion leads to methylation and transcriptional silencing of the gene, reducing or eliminating the gene's protein product, FMRP.[11] FMRP is critical for the maturation and pruning of synapses, neural connectivity, and neurodevelopmental processes.[12] Males with the *FMR1* full mutation tend to be more affected than females given the protective presence of an unaffected X chromosome in females. Almost all males with the *FMR1* full mutation have an intellectual disability, with most having IQs between 40 and 55.[13] In addition to intellectual disability, boys with FXS display marked delays in the ability to use spoken language effectively for interpersonal communication, and delays often are more severe than expected for their levels of cognitive functioning.[14,15] These delays negatively impact the ability to function independently in academic, social, and vocational settings.[16] Thus, there is a need for interventions that improve spoken language for this population to help prevent a negative developmental cascade.

Other phenotypic characteristics of FXS include anxiety and social withdrawal,[17,18] hyperarousal and inattention,[19] tangential (i.e., off topic) and perseverative speech,[20,21] and symptoms of autism.[22–24] The syndrome also is associated with high rates of various challenging behaviors, including aggression and self-injury,[25,26] which often function to escape task and social demands, including those associated with conversation.[27] Collectively, these phenotypic characteristics are likely to limit engagement in the kinds of communicative interactions that support the acquisition of more advanced language skills. An intervention focused on improving the spoken language skills of boys with FXS should, therefore, include an emphasis on increasing engagement and active participation in intervention-related activities.

Expressive language in FXS

Studies using standardized measures of expressive language have revealed significant delays and slower rates of growth for boys with FXS relative to younger typically developing children matched on the nonverbal cognitive level.[28,29] In a within-group analysis of language profiles using standardized measures of language in different domains, Thurman and colleagues found that expressive syntax was delayed in boys with FXS relative to both vocabulary, receptive syntax, and nonverbal cognitive level.[30] However, the spoken language challenges experienced by males with FXS extend beyond what is captured by standardized assessments and include the ways in which spoken language is used to communicate in more

naturalistic interactions (e.g., during back-and-forth conversations with a conversational partner).

For a large group of boys with FXS, averaging between 8- and 9-years of age, Price and colleagues[31] derived measures of syntactic complexity and morphological development based on spoken language produced during administration of the Autism Diagnostic Observation Schedule (ADOS).[32] After controlling for the nonverbal cognitive developmental level and maternal education, boys with FXS displayed shorter mean length of utterance (MLU) and lower scores for grammatical complexity than did younger typically developing children. Additionally, boys with FXS used significantly fewer noun and verb phrases, as well as fewer different types of sentence constructions than typically developing boys. A similar study found that 6- to 16-year-old males with FXS produced fewer morphological endings on nouns and verbs during interactions with an examiner than did younger typically developing children, with between-group differences significant even after controlling for the nonverbal cognitive developmental level, maternal education, and speech intelligibility.[33]

Abbeduto and colleagues[34] have developed a standardized approach for evaluating spoken language ability using samples of expressive language collected in two different contexts: narration of a wordless picture book and an interview-style conversation with an examiner. These sampling procedures have been used in several studies characterizing the spoken language of individuals with FXS. In one study, the authors[35] found that 10- to 17-year-old boys with FXS produced shorter sentences and used fewer different vocabulary words in the narrative context than did younger typically developing participants, even after controlling for nonverbal mental age. In the narrative context, participants with FXS were also less fluent (i.e., produced a higher proportion mazes, dysfluencies, and repetitions) than the younger typically developing children.

Perseveration, the repetition of words, phrases, and topics, interferes with successful spoken communication for males with FXS.[36] The amount of perseverative speech produced by males with FXS is higher than that observed for typically developing children matched on the language level,[37] as well as children with other neurodevelopmental disorders.[38] Perseverative speech has been hypothesized to be related to other aspects of the FXS behavioral phenotype including hyperarousal, attentional impairments, and social anxiety, which may be exacerbated in situations that are perceived as socially demanding, such as dyadic topic-focused interactions. Mothers of children with FXS, almost without exception, anecdotally report that it is extremely difficult to engage their verbal sons with FXS in conversational interactions (e.g., describing what happened at school; discussing an upcoming trip) that last more than just a few reciprocal turns. Thus, in addition to supporting the growth of vocabulary and grammar, we reasoned that the visual supports and structured context provided by the storybooks might constrain the topic of conversation and decrease the amount of tangential language and off-topic responses that are characteristic of the spoken language of boys with FXS. We also reasoned that the sequential nature of the story events in the books and the use of targeted strategies by the mothers could be expected to help the boys to maintain the topic of conversation and result in more frequent initiations and conversational turns.

Despite the pressing need, few behavioral interventions have targeted improvements in spoken language for boys with FXS. In fact, several authors have noted the paucity of intervention research on FXS in general.[39–41] With specific regard to language interventions, after conducting a search of 34 journals, only five studies were identified as targeting communication skills in FXS,[42] each of which enrolled only a single participant with FXS. None of these studies focused on improving higher-level spoken language skills, instead focusing on single behavioral responses, such as signing “please” or using requesting to obtain adult attention.[43,44] In the current study, we situated a spoken language intervention within the naturalistic context of ongoing conversational interactions to provide frequent opportunities to target and ameliorate the key challenges to spoken language experienced by school-aged and adolescent males with FXS.

Shared story-telling

The current intervention was designed to improve spoken language in males with FXS by training mothers in strategies to support their child’s active participation in shared storytelling interactions. Shared story-telling provides a familiar setting for parent/child interactions beginning in early childhood[45] and continues to provide a vehicle for scaffolding children’s language skills and literacy development even as children are learning to read independently.[46,47] Both correlational and experimental studies have supported the positive relationship between shared picture book reading and children’s vocabulary, oral language complexity and narrative skills.[48] For example, the amount of time very young children spend listening to stories is correlated with their language skills at age 5 and their reading comprehension at age 7.[49] During shared story-telling, the adult verbally relates the story to the child, guides discussions about story content and, by adding information, asking questions, and prompting child responses, encourages the child to gradually assume more responsibility for telling the story independently.[50] Research studies implementing shared storytelling interventions, such as those based on dialogic reading, have suggested that it is not merely the exposure to an oral story that supports later language but the strategies the adult partner uses when engaged in the shared reading activity that are important for supporting language growth.[51] The core principle shared by these intervention approaches is to provide the child with ongoing opportunities to hear and practice using developmentally advanced language within the context of telling the story. For speech-language pathologists, shared story-telling represents a dynamic and naturalistic context for delivering a spoken language intervention.[52,53] Although the majority of spoken language interventions embedded in shared story telling have involved younger children,[48,51–53] we reasoned that this approach would be well matched to the developmental needs of older boys with FXS. Because the behaviors of the adult partner can be modified during shared story-telling to provide the child with a more optimal language learning experience,[54] interventions based on shared story-telling can result in improvements in structural aspects of spoken language, such as vocabulary and grammar.[55–57] As mentioned previously, we also expected the visual supports provided by the pictures in the storybooks and the sequential nature of the story events to aid in constraining the amount of off-topic and tangential language that was produced.

McDuffie and colleagues used a single-case multiple baseline design across three mother-child dyads to test the initial promise of a parent-implemented spoken language intervention for school-aged and adolescent boys with FXS.[58] The intervention was embedded into the context of shared story-telling to provide a structured context for practicing sustained and on-topic conversational interactions between mothers and their children. Mothers were trained by a team of speech-language pathologists and served as their child's conversational partner during the book sharing activities. The intervention, which lasted 12 weeks, was completely delivered into participants' homes using a laptop computer and distance video-teleconferencing software. In addition to a laptop computer, each family received a tablet computer containing a library of digitized picture books. One book was used during each week of the intervention and mothers were provided with a script for each book.

A series of parent education sessions was provided at the beginning of the intervention to individually introduce the content of the intervention to each mother. Three types of intervention sessions were conducted each week: (1) a coaching session during which the clinician provided feedback to the mother while she interacted with her son around the shared topic of the wordless book; (2) a homework session during which the mother video-recorded a book sharing interaction with her child and sent it digitally to the clinician; and (3) a feedback session during which the clinician reviewed the homework session with the mother and set goals for the next week of the intervention.

Mothers were taught to use three empirically-based language support strategies while interacting with their sons: Recasts,[59] WH-questions,[60] and Fill-in the Blank prompts.[55] Recasts are adult responses that relate to the central meaning of the child's immediately preceding utterance and provide advanced lexical or grammatical information. WH-questions can prompt the child to answer specific questions about story content, thus supporting story comprehension as well as the ability to remain on topic and retell the story accurately. Fillin the blank prompts (e.g., "he's climbing up the _____") provide a sentence frame paired with rising intonation that helps a child to use a vocabulary word that he knows but may not be able to use independently.

McDuffie et al.[58] found that mothers learned to use the targeted language support strategies. For both mothers and children, participation in the intervention increased the number of spoken utterances that were related to the topic of each story. Because mothers and children produced more story-related utterances, the length of time that each dyad remained engaged in co-telling the story increased from an average of 4 min during baseline to an average of more than 14 min during the intervention. Relative to baseline levels, all three boys increased the number of unique vocabulary words they used during the book sharing interactions and two of the three boys increased the length of the utterances they produced while telling the stories. Mothers reported a high degree of satisfaction with their participation in the intervention and were particularly pleased to be able to participate in a sustained interaction with their children on a regular basis. In summary, the results suggested that a parent-implemented language intervention based on shared story telling has promise for improving the spoken language skills of school-aged boys and adolescent boys with FXS.

The current study

The pilot study conducted by McDuffie et al.[58] was clinically important for several reasons. First, the intervention was embedded in an activity that represents a naturalistic context for parent/child communication. Second, involving the mother in the delivery of the intervention increased the likelihood that the targeted strategies would continue to be used following the conclusion of the formal intervention sessions. Finally, delivery of the intervention by means of distance video-teleconferencing made it cost effective and with the potential to broaden access to the intervention by families regardless of geographic location. The study was limited, however, by the inclusion of only three dyads and the use of a single case design. The current study provided a more rigorous test of the efficacy of the intervention by using a randomized control design to address the following research questions:

1. Does participation in the intervention increase child engagement in shared story telling interactions with the mother as well as the frequency with which mothers and children produce story-related utterances? Do these gains in engagement and story-related talking generalize to clinic-based interactions with the mother? Do child gains in engagement and story-related talking generalize to clinic-based interactions with an unfamiliar examiner?
2. Does participation in the intervention increase maternal use of targeted intervention strategies (i.e., Recasts, WH-questions, Fill-in-the-blank prompts) during a shared storybook interaction in the home? Do maternal gains in the use of targeted intervention strategies generalize to a clinic-based interaction with the child?
3. Does participation in the intervention increase child use of more diverse vocabulary and more complex grammatical constructions in a shared story-telling interaction with the mother? Do child gains in lexical diversity and grammatical complexity generalize to a clinic-based interaction with the mother and with an unfamiliar examiner?
4. During a shared story-telling interaction with the mother, is there a significant association between pretreatment child characteristics and child gains in engagement, story-related talking, lexical diversity, and grammatical complexity at Post-treatment?

Method

Participants

A total of 34 boys with FXS were screened for participation in the current study. Three boys did not meet initial eligibility criteria and nine families decided not to participate. Dyads were recruited through postings on a listserv for families affected by FXS or through a database of previous research participants. Children met the following eligibility criteria: (a) confirmed diagnosis of full mutation FXS; (b) between 10 and 16 years of age; (c) daily use of at least 3-word utterances according to maternal report; (d) English was the primary language spoken in the home; and (e) no uncorrected sensory or motor impairments severe enough to preclude processing and responding to verbal language input according to

maternal report. An additional child eligibility criterion included spoken language appropriate for administration of modules 1 or 2 of the ADOS as determined during the Pretreatment assessment (described below). Informed consent was obtained and all intervention procedures were approved by the University's Institutional Review Board. A battery of standardized tests, informant report, and expressive language sampling measures were administered to 22 children during the pretreatment assessment at a university-based research center. For one child, testing could not be completed due to very severe challenging behaviors and this child was subsequently excluded from participation. One additional child was excluded because his spoken language was appropriate for an ADOS module 3, indicating that this child had achieved the ability to regularly use complex sentences. The 20 eligible boys and their mothers were randomly assigned after the pretreatment assessment to either the active treatment group or a treatment-as-usual comparison group using a list of random numbers. One child in the comparison group did not complete the Post-treatment language samples in the home and did not return for the Post-treatment assessment. The boys enrolled in the study generally communicated in 1–3 word phrases, used some grammatical markings (possessives, past tense, -ing), and were able to express their immediate needs and wants. Conversational speech was marked by tangential and off-topic utterances as well as repetitions of words and phrases. Nine out of ten boys in each of the groups met criteria for a classification of autism spectrum disorders according to the ADOS. Characteristics of each dyad at the Pre-treatment visit are presented in Table 1.

Expressive language sampling

Samples of spoken language during shared story-telling were collected in three different contexts at the Pre- and Post-intervention time points. Language samples were collected in the family home during the two weeks prior to, and following, the twelve intervention sessions and served as the primary outcome measure for the study. A language sample also was collected with each child's mother at the Pre- and Post-treatment assessment at the research clinic. A final language sample was collected at the research clinic with an unfamiliar examiner at the Pre- and Post-treatment assessment. Procedures for transcribing expressive language samples have been developed and tested in previous studies.[14,34,35] In the current study, language samples were transcribed and coded by trained research assistants to derive the variables of interest (see below: Transcription, Coding, and Interobserver Agreement)

Expressive language sample with mother at home—Each mother was asked to complete three language samples with her child at the Pre- and Post-treatment period. The samples involved a shared story-telling interaction and were collected over a period of approximately two weeks following the Pre-treatment clinic visit and prior to the Post-treatment clinic visit. Three pairs of books were used for the language samples in the home. In order to create pairs of books that maintained a comparable style, each pair of books had the same author and was selected from a series of books. The books were edited to have the similar page lengths (± one page) and were digitized and uploaded to the iPad provided to the family. One book from each pair was used for the Pre-treatment language sample and the other pair member was used for the Post-treatment sample. Within pairs, the books were counter-balanced across participants and time points. The book pairs were as follows: (1)

Pair A: *Suddenly* and *Oops* by Colin McNaughton; (2) Pair B: *If You Give a Pig a Party* and *If You Take a Mouse to School* by Laura Numeroff and Felicia Bond; (3) Pair C: *Just a Day at the Pond* and *Just a Little Music* by Mercer Mayer.

The language samples collected in the home represented the primary outcome measure in the current study and can be considered a test of near generalization of intervention effects as the mother was the conversational partner and the book was digitized and loaded onto an iPad as was the case for the intervention sessions. This sampling context differed from the intervention, however, in that no feedback was provided to the mother by the clinician during the sample and the mother was not provided with a script for the book. The following outcome variables were derived from the language samples in the home: child engagement, maternal and child story-related talking, child number of different words, child mean length of utterance (MLU) in morphemes, and maternal use of targeted intervention strategies. After transcription and coding, the outcome variables were averaged across the three language samples to derive a composite measure for each variable of interest. Administration was untimed but typically lasted less than 15 min.

Expressive language sample with mother at clinic—Each mother was asked to complete a language sample with her child at both the Pre- and Post-treatment visits to the research clinic. The mother was instructed to look through the book with her child and to encourage her child to tell the story. Two books by Mercer Mayer were used: *Frog, Where Are You* and *One Frog Too Many*. Books were counterbalanced, and each participant received a different book at Pre-and Post-intervention. Each mother was kept blind to (i.e., did not observe) the language sampling procedure with the unfamiliar examiner. The expressive language sample with the mother in the clinic represented a generalization context as it involved a different setting and materials than did the intervention sessions. In addition to being at the clinic, the context differed from the intervention sessions in that the mother used a hard cover book for the sample (instead of a digitized book on the iPad) and did not have a script. The following outcome variables were derived from the language sample with the mother in the clinic: child engagement, maternal and child story-related talking, child number of different words, child mean length of utterance (MLU) in morphemes, maternal use of targeted intervention strategies. Administration was untimed but typically lasted less than 15 min.

Expressive language sample with an unfamiliar examiner at clinic—In this language sampling context, the examiner guided the participant as they looked at the pages of a wordless picture book to get the gist of the story. After viewing the book, the participant was asked to tell the story page-by-page. The examiner followed a script designed to maximize the participant's contribution and avoid scaffolding the participant's talk. Two books by Mercer Mayer were used: *Frog Goes to Dinner* and *Frog on his Own*. Books were counterbalanced, and each participant received a different book at Pre- and Post-intervention. This language sampling context represented a test of far generalization as the setting, conversational partner, materials, and interaction style differed from the intervention sessions. The following child outcome variables were derived from the language sample in the clinic with an unfamiliar examiner: engagement, number of different words, and mean

length of utterance (MLU) in morphemes. Administration was untimed but typically lasted less than 15 min.

Structure of the intervention program

The intervention program for each mother/child dyad consisted of six types of activities: (a) behavior support sessions, (b) parent education sessions, (c) weekly coaching sessions, (d) weekly homework sessions, (e) weekly clinician feedback sessions, and (f) weekly observation sessions. The sequence of sessions (c) through (f) (i.e., coaching, homework, clinician feedback, and observation sessions) was maintained during each week of the intervention. The primary interventionist was a licensed speech-language pathologist (i.e., SLP). The SLP was assisted by two speech-language clinicians (i.e., Clinicians) both of whom had a master's degree in communication disorders and who were completing a clinical fellowship year under the supervision of the SLP. The SLP delivered the parent education and weekly coaching sessions. The Clinicians reviewed the video-recordings of the homework sessions with the SLP and provided the weekly feedback sessions to the mothers. They also emailed a written summary of the weekly feedback session to the mother. The SLP and Clinicians watched and discussed the observation sessions. Each type of intervention session is explained in more detail below.

Video-teleconferencing equipment—Each family was loaned an "11 MacBook Air™ laptop computer, a 16GB 9.7" iPad Air, and a Plantronics M165 Marque 2 Ultralight Bluetooth Earpiece. The Bluetooth earpiece was used during coaching sessions so that the child would not hear the SLP's comments to the mother. A digitized version of the 12 books selected by each mother were loaded onto an iPad using the Apple iBooks™ application. The clinical team used a 13.3 in MacBook Pro™ laptop computer and initiated video calls using the built-in iSight™ web camera and Skype™ software. Coaching and observation sessions were captured using eCamm™ Call Recording Software. A wireless broadband internet connection was used and secured through 128-bit advanced encryption. Homework sessions were independently recorded by the mother using the iSight™ camera and Photobooth™ software application on the MacBook. These sessions were uploaded to the clinical team using Dropbox™, a cloud-based storage system.

Book selection—Following the Pre-intervention assessment at the research clinic, each mother selected 12 wordless books for use during the intervention sessions (see Table 2). Digitized versions of the selected books, which were edited for length and to remove all text, were uploaded to the iPad and assigned to a random order for use during the 12 weeks of intervention sessions. Prior to each week's coaching session, the child selected one book to read from a choice of the next three books on the randomized list.

Antecedent behavior supports—Prior to initiation of treatment, a Board Certified Behavior Analyst (BCBA) completed the Functional Assessment Interview (FAI)[61] with each mother via a 60-min interview using distance video-teleconferencing (i.e., Skype™). This interview was used to develop hypotheses regarding establishing operations and discriminative stimuli associated with the occurrence of and consequences maintaining challenging behavior during book-sharing. Hypotheses statements were then confirmed by

observation of three video-recorded baseline sessions of mother-child book-sharing activities. The BCBA then developed and implemented a behavior support plan to be used by the mother during all intervention sessions. The behavior support plan included antecedent and consequence-based strategies to (a) decrease the likelihood of challenging behaviors and (b) increase child engagement during the book-sharing activities.[62] Each behavior support plan included: (a) two antecedent strategies (e.g., First_____Then_____ visual card with a highly preferred activity available contingent on completion of the book sharing activity); and (b) three consequence-based strategies including a token economy to differentially reinforce engagement (e.g., token delivery paired with social praise contingent on engagement in the book-sharing activity).

Prior to the introduction of the intervention, each mother in each of the two conditions (treatment and comparison) participated in 2 hours of distance training focusing on the behavioral support strategies including Powerpoint introduction of the strategies, the BCBA modeling strategy use, and maternal practice with child. Mothers were provided with a laminated icon representing each book, a choice board, a visual schedule, and a token tower. Mothers were encouraged to use the behavior support strategies during all intervention activities and the use of these strategies was prompted and reinforced by the BCBA during coaching and feedback sessions.

Parent education sessions—Following the behavior support sessions, the SLP used Skype™ to deliver an individualized PowerPoint presentation containing information about the format and content of the language intervention to each mother. As a precaution against fatigue, the presentation was divided into two one-hour sessions that were delivered during the same week, at the mother's convenience. Mothers also had access to an electronic copy of the PowerPoint during the course of the intervention. Following the parent education sessions, each mother received an electronic copy of a written script for each of the 12 books selected for shared story-telling.

Coaching sessions—A coaching session, delivered by the SLP, was the first step in the weekly sequence of intervention-related activities. Each coaching session was conducted via Skype™, digitally captured, and coded to measure fidelity of intervention delivery. During coaching, the mother wore the Bluetooth earpiece so that the SLP could interact with her without distracting the child. During the coaching session, the mother and child looked at, and talked about, the book selected for that week of the intervention. The SLP guided the mother's responses by modeling story-related vocabulary and sentence structure, by suggesting or modeling the use of the targeted intervention strategies, and by reinforcing the mother's independent use of the strategies.

Fidelity of clinician coaching: Each coaching session was coded from videotape by one member of a team of four trained observers to evaluate fidelity of intervention delivery (i.e., the frequency of coaching behaviors used by the SLP). Twenty percent of sessions, which were randomly selected, were recoded by a second trained observer to evaluate interobserver reliability. Agreement was calculated using the formula: number of agreements/(number of agreements + disagreements) multiplied by 100. For each dyad, agreement was averaged within the following 5 categories of coaching behavior: (1) General coaching behaviors

(e.g., providing a verbal model of a story-related utterance, providing a verbal model reinforcing a child utterance; providing the mother with reinforcement for an utterance, providing the mother with a prompt to transition to the next page of the book); (2) recasts (verbal prompt for mother to use a recast; verbal model of a recast; verbal reinforcement of mother for using a recast); (3) WH-questions (verbal prompt for mother to use a WH-question; verbal model of a WH-question; verbal reinforcement of mother for using a WH-question); (4) Fill-in-the-blank prompts (verbal prompt for mother to use a Fill-in-the-blank prompt; verbal model of a Fill-in-the-blank prompt; verbal reinforcement for using a Fill-in-the-blank prompt); (5) Behavior management (e.g., prompt for mother to provide child with verbal praise and token; verbal praise for mother's use of praise/token). The number of times the mother used an intervention strategy spontaneously or when prompted by the clinician was also coded. See Table 3 for means and ranges for each category of coaching behavior, mean length of coaching sessions, and interobserver agreement.

Independent homework sessions—The homework session was the second step in the sequence of weekly intervention activities and provided a context from which to evaluate the mother's independent use of the strategies targeted during coaching sessions. Within four days of each coaching session, the mother was expected to share that week's story with her child, record the interaction using the Photobooth application on the iPad, and subsequently, upload that recording to the clinical team using Dropbox™. The mother was asked to talk about the entire book with her child, just as the dyad had practiced during the coaching session, and she was encouraged to use the written script as needed.

Clinician feedback sessions—The Clinician reviewed the homework submitted by the mother and used this recording to create clips illustrating parent use of the targeted intervention strategies. The clinician then participated in a SKYPE™ call with the mother and, using the video-clips of the homework session, provided the mother with feedback about her performance and the child's responses, answered any questions posed by the mother, and engaged in joint problem solving relative to the child's behavior and spoken language performance. Clinician comments were also summarized in writing and sent to the mother electronically following each feedback session.

Observation sessions—The final step in the sequence of weekly intervention activities consisted of the Clinician observing, via SKYPE™, as the mother-child dyad engaged in the shared book reading activity. The goal of this session was to provide the child with an additional opportunity to practice each book. Additionally, this session provided the clinician with an opportunity to observe the mother's independent use of targeted intervention strategies as well as the child's participation in the book sharing activity. No feedback or coaching was provided to the mother during this session.

Transcription, coding, and interobserver agreement

Transcription and coding of language variables—Digitized video-recordings of the expressive language samples in three contexts (home with mother, clinic with mother, clinic with unfamiliar examiner) were transcribed using Systematic Analysis of Language Transcripts (SALT),[63] a software program that enables the systematic transcription of

samples of spoken language. Transcription was completed by highly trained research assistants following standardized procedures.[34] Transcription involved a first draft by a primary transcriber, doublechecking by a second transcriber, and final editing by the primary transcriber. Use of this process averages >90% inter-observer agreement.[35] Finalized SALT transcripts were additionally coded by another group of trained observers to generate the following maternal and child variables:

Maternal story-related talking: Maternal utterances that directly related to the semantic or conceptual content of the story included: (a) models of story-related vocabulary and grammar (e.g., "The duck is thinking about getting a flashlight."), (b) general story prompts (e.g., "How does the story start?"), (c) utterances that signaled transitions (e.g., "Let's turn the page and see where the penguin goes next."), and d) use of a targeted intervention strategy (i.e., Recasts, WH-questions, Fill-in-the-blank prompts). The following types of utterances were excluded from the count of maternal story-related utterances: (a) utterances judged to be unrelated to the story; (b) requests for labels and sound effects; (c) yes/no questions; (d) questions without interrogative reversals (e.g.," The duck did what?"); (e) choice questions (e.g., "Who was driving – the farmer or the goat?"); and, (f) utterances related to behavior management.

Child story-related talking: Child utterances directly related to the semantic or conceptual content of the story were coded from the finalized SALT transcripts. Repetitions and completely unintelligible utterances were not included. The resultant transcripts were used to generate the variables for lexical diversity and mean length of utterance for the three language sampling contexts.

Maternal use of recasts: Maternal story-related utterances that were contingent upon and related to the meaning of the immediately preceding child utterance and which provided additional or corrective semantic or grammatical content were coded as Recasts.

Maternal use of WH-questions: Maternal story-related utterances that were in the form of a WH-question were coded to yield the frequency of WH-questions. WH-questions that were simple requests for labels were not included in the count of maternal WH-questions.

Maternal use of fill-in-the-blank prompts: Maternal story-related utterances that were partial utterances ending with rising intonation and an expectant pause and provided children with the opportunity to "fill-in the blank" were coded.

Coding of child engagement—Child engagement was coded directly from the session videotapes at the pre- and post-treatment with a digital video coding software package using a 5-sec partial interval coding system. The child was judged to be engaged in the shared story-telling interaction if he was looking at the pages of the book (i.e., the iPad screen), saying something related to the story content, looking at the mother, or answering the mother's question about the story. The child was required to show engagement for three cumulative seconds within the interval in order for that interval to be coded as engaged. Coding began at the first 5-sec interval in which the mother introduced the book and ended with the final 5-sec interval in which the mother or child was talking about the book.

Intervals were coded as either engaged or unengaged. Intervals in which the mother was not involved in the book sharing activity (i.e., answering her phone, using a behavior support strategy) were coded as not-applicable.

Interobserver agreement for variables derived from transcripts: Four trained observers completed all primary coding using the finalized SALT transcripts from all three language sampling contexts. Twenty percent of these transcripts were randomly selected and independently recoded for interobserver agreement. Intraclass correlations were then computed for the following summary level variables used in the analyses: maternal Recasts, WH-questions, and Fill-in-the-blank prompts, child engagement, and maternal and child story-related utterances. Intraclass correlation coefficients were above .950 for all variables in each language sampling context.

Interobserver agreement for engagement coding: One trained observer completed all primary coding of digitized videos from the pre- and post-treatment sessions with the mother at home, the mother at the clinic, and the examiner at the clinic. A second trained observer independently recoded 20% of these sessions which were randomly selected. Intraclass correlation coefficients were above .950 for all engagement variables.

Analysis plan

Independent samples t-tests were used to test for group equivalence on the maternal and child characteristics that were measured at the Pre-treatment. For the first research question, two-way mixed ANOVAs with Time (Pre-treatment/ Post-treatment) as the repeated measure and Group (Treatment/Comparison) as the between-participants factor were used to determine whether changes in (a) duration of child engagement and (b) frequency of maternal and child story-related talking in the home could be attributed to participation in the shared story-telling intervention. The dependent measures in these analyses were composite variables derived by averaging across the three book sharing interactions conducted in the home at the Pre- and Post-treatment. An additional set of ANOVAs were used to examine generalized changes in child engagement and maternal and child story-related talking in the clinic-based interaction with the mother. A final set of ANOVAs were used to examine generalized changes in these dependent variables in the clinic-based interaction with an unfamiliar examiner.

For the second research question, two-way mixed ANOVAs with Time (Pre-treatment/Post-treatment) as the repeated measure and Group (Treatment/Comparison) as the between-participants factor were used to determine whether changes in maternal use of the targeted intervention strategies in the home could be attributed to participation in the shared story-telling intervention. In these analyses, each dependent measure was a composite variable representing the frequency of each category of parent strategy use averaged across the three shared story-telling interactions conducted in the home at the Pre- and Post-treatment. A separate set of ANOVAs were used to evaluate generalized changes in the dependent variables during a shared story-telling interaction with the mother in the clinic.

For the third research question, two-way mixed ANOVAs with Time (Pre-treatment/Post-treatment) as the repeated measure and Group (Treatment/Comparison) as the between-

participants factor were used to determine whether changes in child lexical diversity (i.e., number of different words) and grammatical complexity (mean length of utterance in morphemes) could be attributed to participation in the intervention. For the interactions conducted in the home, composite variables representing number of different words and mean length of utterance in morphemes were derived by averaging across the three story-telling interactions at the Pre-and Post-treatment. An additional set of ANOVAs were used to evaluate generalized changes in lexical diversity and grammatical complexity in a clinic-based interaction with the mother. A final set of ANOVAs were used to examine generalized changes in these dependent measures in a clinic-based interaction with an unfamiliar examiner.

For the fourth research question, bivariate correlations were used to examine potential associations between Pretreatment characteristics of children in the Treatment group and gains in child engagement, story-related talking, lexical diversity, and grammatical complexity from Pre- to Post-treatment. Correlational analyses were conducted for the shared story-telling interaction with the mother in the home, the shared story-telling interaction with the mother in the clinic, and the narrative language sample with an unfamiliar examiner in the clinic. For each language sampling context, gain scores were computed by subtracting the value of the dependent variables of interest at the Post-treatment from the value of the corresponding variable at the Pretreatment.

Results

Preliminary analyses

Characteristics of each dyad at the Pre-treatment visit are presented in Table 1. Child chronological age showed a trend for differing between the groups, $t(18) = -1.79$, $p < .09$, two-tailed, with the mean age of the treatment group ($\bar{X} = 13.92$, $SD = 2.26$) exceeding the mean age of the comparison group ($\bar{X} = 12.46$, $SD = 1.23$). Thus, we examined the bivariate correlations between child chronological age and each of the child and maternal dependent measures at the Pre- and Post-treatment. None of these correlations reached significance, with all p-values exceeding .23. In light of the nonsignificant associations between child chronological age and the relevant outcome variables, we did not control for child age in the remainder of the analyses. The groups did not differ on any of the other child or maternal variables at the Pre-treatment, with all p's >.50.

Research question 1

Means and standard deviations for maternal story-related talking are presented in Table 4. Means and standard deviations for child story-related talking and child engagement are presented in Table 5.

Shared book reading activity with the mother in the home—For the expressive language sample with the mother in the home, a two-way mixed ANOVA examining child engagement yielded a significant main effect of Time, $F(1,17) = 31.17$, $p = .001$, $\eta^2_{partial} = .647$, and a significant Time X Group interaction, $F(1,17) = 31.23$, $p = .001$, $\eta^2_{partial} = .648$. Post-hoc probing of the Time X Group interaction using an independent samples t-test

indicated that, at the Post-treatment time point, children in the Treatment group spent a significantly longer duration of time jointly engaged in the shared story-telling activity relative to children in the Comparison group, $t(17) = 2.70$, $p < .015$, $d = 1.24$.

For child story-related utterances produced during the shared story-telling activity in the home, a two-way mixed ANOVA yielded a significant effect of Time, $F(1,17) = 19.73$, $p = .001$, $\eta^2_{partial} = .537$, and a significant Time X Group interaction, $F(1,17) = 13.87$, $p < .002$, $\eta^2_{partial} = .449$. Post-hoc probing of the interaction with an independent samples t-test failed to yield a significant difference between the groups at the Post-treatment, $t(17) = -1.09$, $p < .291$, $d = -0.53$. However, paired samples t-tests revealed that, relative to the Pre-treatment time point, children in the Treatment group produced significantly more story-related utterances at Post-treatment, $t(9) = -8.21$, $p = .001$, $d = -3.12$, whereas children in the Comparison group did not change, $t(8) = -.40$, $p < .699$, $d = -0.33$.

Finally, for maternal story-related utterances produced during the shared story-telling activity in the home, a two-way mixed ANOVA yielded a significant main effect of Time, $F(1,17) = 52.21$, $p = .001$, $\eta^2_{partial} = .754$, and a significant Time X Group interaction, $F(1,17) = 56.98$, $p = .001$, $\eta^2_{partial} = .770$. Mothers in the Treatment group produced significantly more story-related utterances at the Post-treatment, $t(17) = -4.22$, $p < .001$, $d = -2.11$, than mothers in the Comparison group.

Shared story-telling activity with the mother in the clinic—For the shared story telling activity with the mother in the clinic, a two-way mixed ANOVA examining child engagement yielded a significant main effect of Time, $F(1,17) = 28,27$, $p = .001$, $\eta^2_{partial} = .624$, and a significant Time X Group interaction, $F(1,17) = 16.07$, $p = .001$, $\eta^2_{partial} = .486$. At the Post-treatment assessment in the clinic, children in the Treatment group spent a significantly longer duration of time jointly engaged with their mothers in the shared story-telling activity relative to dyads in the Comparison group, $t(17) = -2.09$, $p < .050$, $d = -0.97$.

For child story-related utterances produced with the mother during the shared story-telling activity in the clinic, a two-way mixed ANOVA yielded a significant main effect of Time, $F(1,17) = 15.54$, $p < .001$, $\eta^2_{partial} = .478$, and a significant Time X Group interaction, $F(1,17) = 4.36$, $p < .052$, $\eta^2_{partial} = .204$. An independent samples t-test failed to yield a significant difference between the groups at the Post-treatment, $t(17) = -.457$, $p < .653$, $d = -0.22$. However, paired samples t-tests revealed that, relative to the Pre-treatment time point, children the Treatment group produced significantly more story-related utterances at the Post-treatment, $t(9) = -5.36$, $p = .001$, $d = -2.79$, whereas children in the Comparison group showed no difference, $t(8) = -1.09$, $p < .31$, $d = -0.61$.

The analysis of maternal story-related utterances produced during the shared story-telling activity in the clinic yielded a significant main effect of Time, $F(1,17) = 24.74$, $p = .001$, $\eta^2_{partial} = .593$, and a significant Time X Group interaction, $F(1,17) = 33.58$, $p = .001$, $\eta^2_{partial} = .664$. Mothers in the Treatment group produced significantly more story-related utterances at the Post-treatment than mothers in the Comparison group, $t(17) = -3.63$, $p < .002$, $d = -1.78$.

Language sample with an unfamiliar examiner in the clinic—For child engagement, a two-way mixed ANOVA failed to yield a significant main effect of Time, $F(1,17) = 3.104$, $p = .098$, $\eta^2_{partial} = .154$, or a significant Time X Group interaction, $F(1,17) = .64$, $p = .435$, $\eta^2_{partial} = .036$. For child story-related talking, neither the effects of Time, $F(1,17) = 1.28$, $p = .274$, $\eta^2_{partial} = .070$, nor the Time X Group interaction, $F(1,17) = 2.10$, $p = .165$, $\eta^2_{partial} = .110$, reached significance.

Research question 2

Means and standard deviations for maternal use of targeted intervention strategies during shared story-telling in the home and in the clinic are presented in Table 4.

Maternal strategy use for shared story-telling in the home—For WH-questions, a two-way mixed ANOVA yielded a significant main effect of Group, $F(1,17) = 24.76$, $p = .001$, $\eta^2_{partial} = .593$ and a significant Group X Time interaction, $F(1, 17) = 22.85$, $p = .001$, $\eta^2_{partial} = .573$. Post-hoc probing of the interaction using an independent samples t-test confirmed a significant between group difference at the Post-treatment, $t(17) = -2.84$, $p < .011$, $d = -1.34$. Similarly, for use of expansions, the two-way mixed ANOVA yielded a significant main effect of Group, $F(1,17) = 58.42$, $p = .001$, $\eta^2_{partial} = .775$, and a significant Group X Time interaction, $F(1,17) = 56.39$, $p = .001$, $\eta^2_{partial} = .768$. Post-hoc tests confirmed a significant between-group difference at the Post-treatment, $t(17) = -6.46$, $p = .001$, $d = -3.38$, indicating that mothers in the Treatment group produced significantly more expansions following the intervention, relative to mothers in the Comparison group. Finally, for use of intonation prompts, the analysis yielded a significant main effect of Group, $F(1,17) = 21.46$, $p = .001$, $\eta^2_{partial} = .558$, and a significant Group X Time interaction, $F(1,17) = 21.46$, $p = .001$, $\eta^2_{partial} = .558$. Post-hoc probing confirmed a significant between group difference at the Post-treatment, $t(17) = -4.87$, $p = .001$, $d = -2.57$, indicating that mothers in the Treatment group produced more intonation prompts relative to mothers in the Comparison group.

Maternal strategy use for shared story-telling in the clinic—For WH-questions, a two-way mixed ANOVA yielded a significant main effect of Group, $F(1,17) = 8.62$, $p < .009$, $\eta^2_{partial} = .336$ and a significant Group X Time interaction, $F(1, 17) = 9.12$, $p < .008$, $\eta^2_{partial} = .349$. Post-hoc testing using an independent samples t-test revealed a marginally significant between-group difference at the Post-treatment, $t(17) = -2.04$, $p < .057$, $d = -0.94$, favoring mothers in the Treatment group. Additional follow-up testing using paired samples t-tests demonstrated a significant difference between Pre- and Post-treatment in the use of WH-questions for mothers in the Treatment group, $t(9) = -3.73$, $p < .005$, $d = -1.38$, but not for mothers in the Comparison group, $t(8) = .07$, $p < .943$, $d = 0.43$.

For use of expansions, a two-way mixed ANOVA yielded a significant main effect of Group, $F(1,17) = 23.21$, $p = .001$, $\eta^2_{partial} = .577$, and a significant Group by Time interaction, $F(1,17) = 22.578$, $p = .001$, $\eta^2_{partial} = .570$. There was a significant between group difference at the Post-treatment, $t(17) = -4.37$, $p = .001$, $d = -2.46$, indicating that mothers in the Treatment group produced significantly more expansions following the intervention relative to mothers in the Comparison group. Finally, for use of intonation prompts, the two-way

mixed ANOVA yielded a significant main effect of Group, $F(1,17) = 14.51$, $p < .001$, $\eta^2_{partial} = .460$, and a significant Group by Time interaction, $F(1,17) = 18.33$, $p < .001$, $\eta^2_{partial} = .519$. Post-hoc testing confirmed a significant between group difference at the Post-treatment favoring mothers in the Treatment group, $t(17) = -4.56$, $p = .001$, $d = -2.36$.

Research question 3

Means and standard deviations for lexical diversity and grammatical complexity are presented in Table 5.

Child lexical diversity and grammatical complexity with the mother in the home—For lexical diversity, a two-way mixed ANOVA yielded a significant main effect of Time, $F(1,17) = 23.76$, $p = .001$, $\eta^2_{partial} = .583$, and a significant Group X Time interaction, $F(1,17) = 23.24$, $p = .001$, $\eta^2_{partial} = .578$. The effect of Group failed to reach significance. Post-hoc testing with an independent samples t-test failed to yield a significant difference between the two groups at the Post-treatment time point, $t(17) = -1.59$, $p < .13$, $d = -0.73$. However, paired samples t-tests revealed a significant difference at the Post-treatment, relative to Pre-treatment, in the number of different words produced by participants in the Treatment group, $t(9) = -7.45$, $p = .001$, $d = -2.57$, but not by participants in the Comparison group, $t(8) = -0.03$, $p < .975$, $d = -0.01$. For grammatical complexity, neither the main effects nor interaction were significant.

Child lexical diversity and grammatical complexity with the mother in the clinic—For lexical diversity, a two-way mixed ANOVA yielded a significant main effect of Time, $F(1,17) = 8.70$, $p < .009$, $\eta^2_{partial} = .338$, and a significant Group X Time interaction, $F(1,17) = 5.63$, $p = .030$, $\eta^2_{partial} = .249$. The effect of Group failed to reach significance. Post-hoc testing with an independent samples t-test failed to yield a significant difference between the two groups at the Post-treatment, $t(17) = -0.96$, $p < .348$, $d = -0.44$. However, paired samples t-tests revealed a significant difference at the Post-treatment, relative to the Pre-treatment, in the number of different words produced by participants in the Treatment group, $t(9) = -4.53$, $p < .001$, $d = -1.85$, but not by participants in the Comparison group, $t(8) = -0.35$, $p < .737$, $d = -0.14$. For grammatical complexity, neither the main effects nor interaction were significant.

Child lexical diversity and grammatical complexity in the clinic with an unfamiliar examiner—None of the main or interaction effects were significant.

Research question 4

Note that change during treatment for each dependent measure was calculated by subtracting the Post-treatment score from the pretreatment score. Thus, improvement was reflected by a negative number.

Bivariate correlations with child pre-treatment characteristics in the shared story-telling interaction with the mother in the home—There were significant bivariate associations between receptive vocabulary standard scores and gains in the frequency of child story-related utterances, $r(10) = -.739$, $p < .015$, as well as gains in the

number of different words used by the children, $r(10) = -.714$, $p < .020$, in the home. There was also a marginally significant association between receptive vocabulary standard scores and gains in the duration of child engagement, $r(10) = -.583$, $p < .077$. There was a significant bivariate association between expressive vocabulary standard scores and gains in the number of different words used by the children, $r(10) = -.666$, $p < .035$, in the home. Finally, there was a marginally significant association between expressive vocabulary standard scores and gains in the frequency of child story-related utterances, $r(10) = -.611$, $p < .060$. There were no significant associations for child chronological age, nonverbal IQ, autism severity, or standard scores for expressive grammar.

Bivariate correlations with child pre-treatment characteristics in the shared story-telling interaction with the mother in the clinic—There were significant bivariate associations between receptive vocabulary standard scores and gains in child engagement, $r(10) = -.838$, $p < .002$, frequency of child story-related talking, $r(10) = -.774$, $p < .009$, and the number of different words children used, $r(10) = -.659$, $p < .038$, in the clinic-based interaction with the mother. There were significant bivariate associations between expressive vocabulary standard scores and gains in child engagement, $r(10) = -.774$, $p < .009$, frequency of child story-related talking, $r(10) = -.672$, $p < .033$, and the number of different words, $r(10) = -.679$, $p < .031$, produced during the clinic-based interaction with the mother. There were no significant associations for child chronological age, nonverbal IQ, autism severity, or expressive grammar standard scores.

Bivariate correlations with child pre-treatment characteristics in the narrative language sample with an unfamiliar examiner in the clinic—None of the correlations was significant.

Maternal satisfaction

All mothers completed a satisfaction survey at the Post-intervention to reflect their impressions of the intervention program. In general, mothers reported a high degree of satisfaction with their participation. The results of the parent satisfaction survey are presented in Table 6.

Discussion

Boys with FXS are likely to speak in short phrases that are adequate for conveying their immediate needs and wants. Vocabulary and grammatical skills are, however, delayed relative to cognitive-level expectations and conversational interactions are negatively impacted by other co-occurring problems. Social anxiety and inattention may limit sustained engagement in conversational interactions, whereas repetitive, poorly articulated, and tangential speech may interrupt the sequential flow of information exchange between conversational partners. The goal of the current project was to improve the spoken language of boys with FXS by teaching their mothers to provide models of developmentally advanced vocabulary and grammar as well as to provide prompts to elicit child responses that continued the topic of the shared conversation. Interactions were situated within the context

of shared story-telling using wordless picture books, which provided both a visual structure and a linear sequence for maintaining a sustained and on-topic conversation.

Our analyses indicated that mothers in the Treatment group learned the three targeted intervention strategies (i.e., Recasting, WH-questions, and Fill-in-the-blank prompts) and, at the Post-treatment, used these strategies significantly more often than mothers of children in the Comparison group in both the home and in the clinic. At the Post-treatment, boys with FXS in the Treatment group spent a significantly longer duration of time engaged in the shared storytelling interaction with their mothers than did the boys in the Comparison group. Engagement in the shared story-telling experience sets the occasion for the child to be exposed to models of more advanced language from the adult. When the child is engaged, the adult can use WH-questions or Fill-in-the-blanks to prompt the child to take an additional turn in the shared story-telling interaction. When the child takes a spontaneous or prompted turn in the interaction, the adult can respond contingently by recasting the child's utterance, thus providing additional enriched language input. Thus, child engagement in the interactive context enables many of the transactional aspects of language learning to take place.

The results for story-related talking also indicated that children in the Treatment group were actively engaged in the shared story-telling interaction. Relative to the Pre-treatment time point, we found that children in the Treatment group produced more story-related utterances when interacting with their mothers in the home at the Post-treatment. The significant increase in mothers' use of story-related talking in the home completes the picture indicating that both mothers and children were more active participants in the shared story-telling activities because of the treatment. These results were mirrored for the shared story-telling interaction with the mother in the clinic; again, with enhanced engagement on the parts of both partners. However, we did not observe generalization of child engagement or story-related talking to the language sample with the unfamiliar examiner, perhaps indicating that the children were reticent to engage with the unfamiliar examiner and/or continued to need the scaffolding provided by the mother in order to maintain their engagement and participation in the story-telling activity. Put differently, the shared story-telling skills of participating boys may have been emerging and could not be applied independently with an examiner who an interactive style without extensive supports.

Children in the Treatment group used a significantly more diverse vocabulary at the Post-treatment than they did at the Pre-treatment while children in the Comparison group did not. This effect was observed with the mother in both the home and in the clinic. Once again, changes in lexical diversity at the Post-treatment were not observed in the language sample with the unfamiliar examiner. Here too, it appears that the lexical skills of the boys with FXS were not sufficiently developed to be applied without the considerable support provided by an active and responsive conversational partner.

Growth in grammatical complexity was not observed in any of the three language sampling contexts. One possible explanation for the lack of significant changes in mean length of utterance may be attributed to the mother's use of the language facilitation strategies. The use of WH-questions often resulted in one or two word responses (rather than a complete

utterance) from the child as did the use of Fill-in-the-blank prompts. Even though mothers were regularly recasting these and other short utterances by adding semantic and grammatical information, it does not appear that this was sufficient to result in increases in children's sentence length. It is also possible that the 12 week time frame of the intervention was not sufficient to observe changes in the use of more complex grammar.

Examination of bivariate correlations revealed a strong association between intervention gains and standard scores for receptive and expressive vocabulary. In general, children with higher levels of vocabulary ability showed bigger gains in duration of engagement and frequency of story-related talking and used more diverse vocabulary during the shared storytelling interactions with the mother. Unexpectedly, significant correlations were not observed between child treatment gains and pretreatment chronological age, nonverbal IQ, autism severity, or grammatical ability. It may be that children with higher levels of receptive vocabulary were able to follow the story content more effectively, leading to higher levels of engagement in the shared story-telling interaction. Similarly, children with higher initial levels of vocabulary competence may have been able to more efficiently learn to use a greater variety of different words as the result of the shared storytelling interaction. Such findings suggest the need to adapt our treatment for children with less well-developed vocabularies, perhaps through increasing the duration of the intervention, limiting the rate at which parents introduce new words, or increasing the number of opportunities parents provide to practice emergent vocabulary.

One strength of the current study was the use of expressive language sampling as an outcome measure. Samples of expressive language have been recommended for use as outcome measures in studies of language and cognition in FXS,[64] as these measures are hypothesized to be sensitive to clinically meaningful changes in behavior, can be used to derive variables that represent many of the component skills of language, and have applicability over a wide range of developmental levels. The outcome measures used in the current study were derived from samples of expressive language collected in three contexts that differed in varying degrees from the context in which parent training occurred, thereby providing tests of the generalization of intervention effects. The language samples that were collected with the mother in the home closely resembled the intervention context, although the mother did not receive any feedback from the clinician or use a script during the collection of these samples. The language samples that were collected with the mother in the clinic differed in setting (i.e., location) from the intervention sessions. Additionally, in contrast to the coaching and homework sessions, a hard-cover book was used rather than an iPad and the mother did not have access to a script. Finally, the language samples that were collected during shared story-telling with an unfamiliar examiner differed from the intervention sessions in conversational partner, setting, and interaction style, in that the examiner provided only a minimal amount of scaffolding to the child.

A disappointing feature of our findings is the lack of generalization of intervention effects to the language sample with the unfamiliar examiner in the clinic. The demands of interacting with an unfamiliar examiner could potentially have elicited high levels of anxiety and social avoidance in the child, making an assessment of optimal levels of spoken language competence unlikely. Additionally, this sampling context differed in other potentially

important ways from the shared story-telling interaction with the mother. Whereas the mother interacted with her son in ways that were designed to scaffold the back and forth nature of the book sharing conversation, the language sample with the examiner utilized the standardized narrative language sampling protocol of Abbeduto and colleagues[35] and was designed to limit the amount of talk and extent of scaffolding provided by the examiner. Indeed, the examiner's comments were largely limited (e.g., "what's happening on this page?" or "here's the next page") so as to determine what the child could do independently. Achieving the level of generalization required in such a non-scaffolded interactional context may well require adaptation of our intervention, perhaps by increasing the duration of treatment, increasing the diversity of contexts in which parent coaching occurs (e.g., adding interactions that do not involve books or stories), or adding an additional conversational partner in some treatment sessions. It is also possible that the mother's scaffolding, which can be considered as a verbal prompt, should be faded gradually in order to facilitate generalization by lessening prompt dependence. Further, it is also possible that the schedule of reinforcement used during consequence-based strategies (i.e., token economy) would also need to be thinned in order to facilitate generalization. Testing such alternatives awaits future studies.

The present study has several limitations which should be mentioned. First, the length of the parent education sessions, coaching sessions, feedback sessions, and observation sessions was not held constant across dyads. Thus, some children may have received a more intensive dose of the intervention than other children. We did not measure fidelity of implementation for the parent education sessions and the sessions used to instruct the parents in the behavioral support strategies. This will be an important factor to include in future studies. We also failed to control for multiple significance testing in our analysis which would have been important given the large number of ANOVAs which were conducted. We further acknowledge that our sample size was fairly small although it represents a good sized sample for studies of children with FXS within a circumscribed age range. Finally, we did not measure maintenance or durability of the intervention effects, which would be important to assess in future studies.

In the present study, we have documented the promise of a parent-implemented shared story-telling intervention for improving engagement and spoken vocabulary in adolescents with FXS. We also found that effects are more limited in boys who begin treatment with more restricted vocabularies and that generalization is limited to situations in which a high degree of scaffolding continues to be provided. Such variability suggests the need for testing further adaptations of the intervention. Not surprisingly, there was variability among mothers in the rate at which they acquired the targeted strategies, which means that there was variability in effective "dose" of the intervention experienced by the boys with FXS. In subsequent analyses, we are working to characterize such parental variability and relate it to child outcomes. Those analyses will be used to develop adaptations of the intervention.

Targeted treatments that aim to normalize the molecular and cellular pathways underlying the FXS behavioral phenotype have shown promise in pre-clinical trials, most commonly using a knock-out mouse model of FXS.[61] The promise of these trials, however, has not been realized in clinical trials with human participants and many uncertainties remain about

how best to demonstrate treatments effects in clinical trials of FXS.[62] As enumerated by Berry-Kravis and colleagues,[65] major trial design issues include narrow dosing windows, restriction of treatment to older individuals, limited duration of treatment, large placebo effects, difficulty in selecting specific, psychometrically sound, and sensitive outcome measures, and lack of validated biomarkers to measure treatment effects at the neurobiological level.[66] One additional important issue is that individuals with FXS have experienced less that optimal learning opportunities over the course of the lifespan, resulting in a cascade of negative developmental effects.

Although a targeted treatment may correct impaired neuronal signaling pathways in FXS, it is unlikely that such a treatment can ameliorate deficits in experience-dependent learning that have accumulated over time. Two ways to address this problem are to treat individuals during a chronologically younger developmental time frame to take advantage of critical periods for learning and to combine a targeted treatment with an intensive behavioral intervention that, concurrently with the targeted treatment, creates an optimally enriched learning environment to support developmental outcomes such as language. An intensive learning intervention should facilitate changes in drug-related synaptic plasticity such that these changes are more likely to be detected within the time frame of the clinical trial.

Unfortunately, there are few behavioral interventions that have been empirically tested with individuals with FXS. The language intervention described in the present study represents a line of research that could be used in combination with a targeted treatment to assess the efficacy of that treatment on spoken language outcomes. At the same time, a medication may improve responsiveness to the behavioral treatment, thereby overcoming some of the limitations we have noted among boys with more limited starting vocabularies or the limited generalization observed with unfamiliar communicative partners.

Acknowledgments

The authors wish to thank the families who participated in this research. We also thank Dr. Wendy Machalicek for her assistance with videoteleconferencing procedures and formative contributions to conceptualizing the role of the BCBA in this program of intervention research.

Funding

This research was supported by grant U54 HD079125 from the National Institute of Child Health and Human Development to Dr. Leonard Abbeduto. Leonard Abbeduto has received financial support to develop and implement outcome measures for clinical trials from F. Hoffman-LaRoche, Ltd., Roche TCRC, Inc. and Neuren Pharmaceuticals Limited. Randi J. Hagerman has received funding from Marinus, Novartis, Alcobra, Neuren, and Curemark to carry out treatment studies in fragile X syndrome and ASD. She has also consulted with Roche/Genetech, Zynerba, Orid, and Novartis regarding treatment studies in fragile X syndrome.

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Table 1

Characteristics of participating dyads at pre-treatment assessment: Means and standard deviations.

Child and Maternal Characteristics	Treatment Group	Comparison Group	Test	*p*
Child Characteristics				
Chronological age (years)	13.92 (2.26)	12.46 (1.23)	$t_{(18)} = -1.79$	.09
Nonverbal IQ[1]	41.80 (8.72)	41.20 (8.23)	$t_{(18)} = -0.16$	.88
Autism severity[2]	6.00 (2.31)	5.50 (1.72)	$t_{(18)} = -0.55$	.59
Receptive vocabulary[3]				
Age equivalent	6.51 (2.55)	5.98 (1.31)	$t_{(18)} = -0.59$	.56
Standard score	52.90 (21.55)	55.80 (14.56)	$t_{(18)} = 0.35$	.73
Expressive vocabulary[4]				
Age equivalent	6.02 (1.69)	5.73 (1.16)	$t_{(18)} = -0.45$	.66
Standard score	56.20 (15.64)	58.5 (15.47)	$t_{(18)} = 0.33$	.75
Expressive syntax[5]				
Age equivalent	3.93 (2.10)	3.63 (1.12)	$t_{(18)} = -0.41$	.69
Standard score	43.50 (8.89)	40.60 (1.35)	$t_{(18)} = -1.02$	.32
Maternal characteristics				
Chronological age	44.20 (6.00)	44.00 (6.13)	$t_{(18)} = -0.07$	.94
Years of education	15.30 (1.77)	15.40 (2.46)	$t_{(18)} = 0.10$	.92
IQ[6]	109.30 (12.25)	104.40 (16.08)	$t_{(18)} = -0.77$	.45

Note.

[1] Leiter International Performance Scales-Revised (Roid & Miller, 1997);

[2] Autism Diagnostic Observation Schedule (Lord, Rutter, DiLavore, Risi, Gotham, & Bishop, 2012;

[3] Peabody Picture Vocabulary Test − 4th Edition (Dunn & Dunn, 2007);

[4] Expressive Vocabulary Test-2 (Williams, 2007);

[5] Comprehensive Assessment of Spoken Language: Sentence Formulation (Carrow-Woolfolk, 1999);

[6] Kaufman Brief Intelligence Test −2nd Edition (Kaufman & Kaufman, 2004).

Table 2

Wordless picture books used during intervention sessions.

Book	Author	Original Page Length	Edited Page Length	# of Dyads Selected
Badger's Fancy Meal	Keiko Kasza	32	19	7
Carl's Birthday	Alexandra Day	32	21	1
Chalk	Bill Thomson	40	20	7
Charlie the Ranch Dog	Ree Drummond	40	19	2
Cow Can't Sleep	Ken Baker	24	21	8
Duck on a Bike	David Shannon	40	19	10
Harry the Dirty Dog	Gene Zion	32	20	8
I Just Forgot	Mercer Mayer	24	21	9
I Took My Frog to the Library	Eric A. Kimmel	32	20	9
If You Give a Dog a Donut	Laura Numeroff	32	21	7
Lost and Found	Oliver Jeffers	32	21	6
Marley: Messy Dog	John Grogan	32	21	1
Mooncake	Frank Asch	32	21	5
Mr. Gumpy's Motor Car	John Burningham	32	19	5
Octopus Soup	Mercer Mayer	24	21	6
Pancakes for Breakfast	Tomie dePaola	32	21	8
Pete at the Beach	James Dean	32	20	2
Ready for Anything!	Keiko Kasza	32	21	7
That's Good! That's Bad!	Margery Cuyler	32	21	7
The Invisible Boy	Trudy Ludwig	40	21	1
The Perfect Pet	Margie Palatini and Bruce Whatley	32	19	4

Table 3

Frequency and percent agreement for clinician coaching behaviors.

Categories of Clinician Coaching	Weekly Intervention Sessions												Percent Agreement
	1	2	3	4	5	6	7	8	9	10	11	12	
WH-Questions													
Coached	19	21	20	23	22	17	22	20	22	20	17	19	94%
Range	7–28	8–28	5–46	8–36	9–35	6–28	3–36	7–49	6–34	9–27	7–25	11–35	75 – 100%
Parent Prompted[1]	16	19	19	22	21	16	21	19	20	18	16	18	94%
Range	6–24	7–29	5–46	7–35	9–33	6–29	3–34	7–46	5–34	8–26	6–22	9–33	75 – 100%
Parent Spontaneous[a]	20	21	19	23	23	22	25	26	26	25	22	30	87%
Range	4–38	1–38	5–41	6–42	6–46	11–38	5–44	7–58	8–54	4–52	6–39	19–43	33 – 100%
Expansions													
Coached	9	8	7	6	8	5	5	6	6	5	6	5	89%
Range	1–18	0–16	0–17	1–13	0–18	2–7	1–13	3–16	1–11	2–10	1–18	0–12	50 – 100%
Parent Prompted	5	5	5	4	5	3	4	5	4	3	5	4	82%
Range	1–13	0–12	0–13	1–13	0–11	0–6	0–11	1–14	1–9	1–7	1–16	0–10	40 – 100%
Parent Spontaneous	21	23	30	35	32	30	36	41	45	38	36	42	80%
Range	9–36	8–35	14–65	19–55	8–56	21–42	15–62	18–67	26–62	22–63	15–49	27–66	48 – 100%
Intonation Prompts													
Coached	6	8	6	6	7	6	6	6	6	5	5	6	92%
Range	0–15	2–16	1–13	1–16	0–15	1–14	0–14	0–17	0–13	1–12	0–9	0–14	67 – 100%
Parent Prompted	4	7	5	5	6	5	5	4	5	5	4	4	88%
Range	0–7	1–16	1–11	1–10	0–12	1–11	0–12	0–13	0–12	1–10	0–6	0–12	50 – 100%
Parent Spontaneous	11	11	12	18	13	14	16	19	20	15	14	19	91%
Range	0–24	1–19	0–44	1–65	5–31	3–33	5–32	4–42	5–36	7–28	1–41	7–32	60 – 100%
General													
Coached	60	56	45	48	54	50	46	52	47	53	50	44	90%
Range	45–75	38–73	28–60	23–64	37–69	34–64	21–62	28–91	28–65	29–72	31–71	31–60	77 – 100%
Behavior Management													
Coached	7	3	3	2	6	5	4	4	3	1	4	1	76%
Range	1–16	0–7	0–9	0–5	0–19	0–13	0–18	0–17	0–9	0–5	0–16	0–4	0– 100%

[1] Parent response to clinician prompt for strategy use

[a] Parent spontaneous use of targeted strategy

Table 4

Parent strategy use during narrative language sampling in two contexts: home and clinic.

	Treatment Group				Comparison Group			
	Pre-Treatment		Post-Treatment		Pre-Treatment		Post-Treatment	
Dependent Measure	***Mean***	***(SD)***	***Mean***	***(SD)***	***Mean***	***SD***	***Mean***	***(SD)***
Language Sample in Home								
Story-Related Talking	50.03	(33.78)	163.50	(66.53)	59.03	(29.66)	61.44	(30.27)
Wh-Questions	12.47	(9.96)	47.43	(25.26)	16.60	(14.48)	18.85	(17.40)
Expansions	3.63	(2.75)	37.20	(13.87)	5.00	(4.20)	5.74	(4.70)
Fill-in the Blanks	3.20	(4.77)	22.30	(11.62)	2.30	(2.50)	2.48	(3.86)
Language Sample in Clinic								
Story-Related Talking	69.20	(36.72)	153.80	(62.79)	70.00	(36.74)	69.33	(32.11)
Wh-Questions	20.60	(12.83)	44.40	(24.59)	21.10	(16.07)	22.11	(22.77)
Expansions	8.60	(7.46)	40.60	(22.07)	6.90	(7.43)	7.67	(4.69)
Fill-in the Blanks	4.30	(7.17)	21.40	(11.01)	4.40	(4.67)	3.67	(4.00)

Table 5

Child outcome variables in three language sampling contexts: home with mother, clinic with mother, clinic with unfamiliar examiner.

	Treatment Group				Comparison Group			
	Pre-Treatment		Post-Treatment		Pre-Treatment		Post-Treatment	
Dependent Measure	***Mean***	***(SD)***	***Mean***	***(SD)***	***Mean***	***(SD)***	***Mean***	***(SD)***
Language Sample in Home								
Engagement (secs)	285.43	(135.68)	779.77	(315.80)	350.20	(207.05)	363.48	(297.50)
Story-Related Talking	41.80	(25.70)	129.00	(45.90)	74.10	(69.00)	88.67	(106.86)
Number of Different Words	51.70	(33.51)	116.30	(45.16)	72.20	(55.73)	78.78	(57.37)
Mean Length of Utterance	2.41	(1.01)	2.42	(0.62)	2.35	(0.64)	2.42	(0.62)
Language Sample in Clinic with Mother								
Engagement (secs)	406.70	(201.76)	737.00	(329.31)	373.30	(220.72)	448.33	(268.08)
Story-Related Talking	71.30	(39.12)	128.80	(66.66)	83.60	(82.06)	108.33	(123.23)
Number of Different Words	68.80	(31.66)	103.70	(47.56)	71.10	(41.84)	80.00	(59.45)
Mean Length of Utterance	2.66	(1.05)	2.50	(0.80)	2.59	(0.87)	2.42	(0.62)
Language Sample in Clinic with Examiner								
Engagement (secs)	385.70	(86.44)	375.80	(80.67)	496.20	(147.64)	434.67	(150.39)
Story-Related Talking	57.50	(19.56)	60.60	(31.77)	90.50	(62.67)	71.11	(37.44)
Number of Different Words	73.20	(26.63)	76.40	(37.17)	98.90	(59.10)	87.74	(30.03)
Mean Length of Utterance	3.98	(1.32)	4.09	(1.52)	3.99	(1.20)	3.95	(1.38)

Table 6

Results of parent satisfaction survey.

Survey item	Mean	Range
Participation in the project was beneficial to me	6.9	6–7
Participation in the project was beneficial to my child	6.8	5–7
I found the clinicians easy to work with	7.0	7
The intervention materials were well organized	6.9	6–7
The information presented was new to me	6.3	4–7
I was able to use the laptop and tablet computers	7.0	7
I was able to use the Bluetooth	6.3	6–7
I enjoyed the coaching sessions	6.6	5–7
I enjoyed the homework sessions	6.7	6–7
I enjoyed the feedback sessions	7.0	7
I found the written feedback helpful	6.8	5–7
The intervention strategies were easier to use over time	6.9	6–7
My child’s spoken language has improved	6.6	5–7
My child’s understanding has improved during the book sharing activity	6.2	5–7
My child’s attention has improved during the book sharing activity	6.5	5–7
My child’s participation has improved during the book sharing activity	6.7	6–7
I have noticed improvements in spoken language outside of intervention	6.5	5–7
I am able to use the intervention strategies during everyday activities	6.3	5–7
I will continue to use intervention strategies	6.8	6–7
I would recommend this intervention to other families	7.0	7