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Perseveration in the Connected Speech of Boys with Fragile X Syndrome with and Without Autism Spectrum Disorder

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Abstract

Verbal perseveration is a frequently reported language characteristic of males with Fragile X syndrome and may be a defining feature or hallmark of the syndrome. We compared the verbal perseveration of boys with Fragile X syndrome with (n = 29) and without (n = 30) autism spectrum disorder, boys with Down syndrome (n = 27), and typically developing boys (n = 25) at similar nonverbal mental ages. During a social interaction, boys with both Fragile X syndrome and autism spectrum disorder produced significantly more topic perseveration than all other groups. In social interaction as compared to narration, boys with Fragile X syndrome (regardless of autism status) produced significantly more topic perseveration. These findings suggest that autism status, as well as language sampling context, affect perseveration in boys with Fragile X syndrome.

Keywords

Fragile X syndrome; autism; Down syndrome; perseveration; X-linked

Fragile X syndrome is the most common, known inherited cause of intellectual disability (ID; Dykens, Hodapp, & Finucane, 2000; Hagerman & Hagerman, 2002), with the full mutation occurring in about 1 in 2,500 to 1 in 5,000 individuals (Coffee et al., 2009; Fernandez-Carvajal et al., 2009; Hagerman, 2008) and males more severely affected than females (Hagerman & Hagerman, 2002; Loesch et al., 2003; Reiss & Dant, 2003). Characteristics associated with Fragile X syndrome include anxiety (Bregman, Leckman, & Ort, 1988; Cordeiro, Ballinger, Hagerman, & Hessl, 2011; Hagerman, 2002), attention

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problems (Hooper, Hatton, Baranek, Roberts, & Bailey, 2000; Wilding, Cornish, & Munir, 2002), and autism (Hagerman & Hagerman, 2002). Fragile X syndrome is also associated with language impairment (Abbeduto, Brady, & Kover, 2007; Finestack, Richmond, & Abbeduto, 2009; Roberts, Chapman, Martin, & Moskowitz, 2008), with verbal perseveration, or the excessive self-repetition of a spoken word, phrase, sentence, or topic (Abbeduto & Hagerman, 1997), frequently reported among males with Fragile X syndrome (e.g., Murphy & Abbeduto, 2007; Roberts, Martin, et al., 2007; Sudhalter, Cohen, Silverman, & Wolf-Schein, 1990). Despite the widely cited claim that verbal perseveration is a hallmark of Fragile X syndrome (e.g., Sudhalter, et al., 1990), the role of autism status in the perseveration of individuals with Fragile X syndrome remains largely unexamined, different types of perseveration are generally not delineated, and the majority of studies to date have only examined perseveration in a single context (conversation samples). Thus, in the present investigation, we examined differing types of verbal perseveration in boys with Fragile X syndrome with and without autism spectrum disorder, boys with Down syndrome, and younger typically developing boys in two language sampling contexts: social interaction and narration.

Many researchers contend that verbal perseveration is a defining feature of the Fragile X syndrome behavioral phenotype (e.g., Abbeduto, et al., 2007; Bennetto & Pennington, 2002; Roberts, Chapman, Martin, et al., 2008). This argument is reflective of a major goal in developmental disabilities research, which is to distinguish syndromes on the basis of behavioral phenotypes, including language phenotypes (Rice, Warren, & Betz, 2005; Tager-Flusberg, 2005). Below we review the existing literature on perseveration in Fragile X syndrome and present our rationale for examining the role of autism and language sampling context.

Verbal Perseveration in Fragile X Syndrome

Although several early studies reported perseveration to be common in males with Fragile X syndrome, these studies did not include comparison samples of typically developing individuals or individuals with other forms of ID (Fryns, Jacobs, Kleczkowska, & van den Berghe, 1984; Hanson, Jackson, & Hagerman, 1986; Madison, George, & Moeschler, 1986). In a study by Prouty et al. (1988), however, only 3 of 15 young males displayed perseveration and two others demonstrated word repetitions. The rather low incidence of perseveration identified in this study may be accounted for by the definition of *perseveration* as “repetitive phrases which were interspersed with other phrases” (p. 132). Note that neither Fryns et al. nor Hanson et al. provided operational definitions of perseveration; thus, it is not known which specific behavior or behaviors were measured in the two studies.

Two recent studies reported perseveration (with varying definitions) to be more common in boys with Fragile X syndrome than in younger typically developing boys (Levy, Gottesman, Borochowitz, Frydman, & Sagi, 2006; Roberts, Martin, et al., 2007). Levy et al. (2006) found that boys with Fragile X syndrome produced more perseveration (defined as consecutive production of the same utterance) during conversation with an examiner than typically developing boys matched for mean length of utterance. Roberts, Martin, et al. (2007) also found that boys with Fragile X syndrome produced more perseveration (defined as the repetition of certain words, phrases, or sentences within or across turns, or when the child repeatedly spoke on a single topic or theme of topics) than typically developing boys in examiner-child interactions, after controlling for nonverbal mental age.

A few investigations have also found that males with Fragile X syndrome produced more perseveration (definition sometimes not provided) than either males with Down syndrome (Roberts, Martin, et al., 2007; Wolf-Schein et al., 1987) or autism (Sudhalter, et al., 1990).

Sudhalter et al. (1990) were the first investigators to propose a taxonomy that denoted specific types of perseveration in individuals with Fragile X syndrome. In a study of "deviant repetitive language" (comprised of perseveration, echolalia, jargoning, and "affirming by repetition") in boys and adult males with Fragile X syndrome, Down syndrome, and autism, Sudhalter et al. distinguished three types of perseveration: (a) *phrasal*, the sequential repetition of a phrase wherein the repetition is identical or almost identical to the original phrase; (b) *sentential*, the sequential repetition of a sentence, wherein the repetition is identical or almost identical to the original sentence; and (c) *topic*, the "incessant" talk on a topic that includes reintroductions considered to be tangential to the current topic. Despite the use of this taxonomy to analyze speech samples, all types of perseveration were combined for analysis; they were either combined with each other to form a single variable of "perseveration" or combined with other repetitive behaviors to form a single variable of "deviant repetitive language." This approach to analyzing the data was used again in other studies by Sudhalter and colleagues (Belser & Sudhalter, 1995; Sudhalter, Scarborough, & Cohen, 1991). Sudhalter et al. (1990) found that males with Fragile X syndrome produced more deviant repetitive language than did males with Down syndrome during interactions with both a familiar person and an unfamiliar person (the examiner). Moreover, a post-hoc analysis revealed that males with Fragile X syndrome produced specifically more perseveration than males with autism. Note, however, that males with Fragile X syndrome were not compared to males with Down syndrome specifically on the dimension of perseveration.

Perseveration and Autism Status

Fragile X syndrome is the leading single-gene disorder associated with a diagnosis of autism (Hagerman & Hagerman, 2002). Recent studies suggest that 18–52% of males with Fragile X syndrome also meet criteria for autism on gold standard instruments, and that almost half to nearly three quarters of males (43–74%) may be on the autism spectrum (Clifford et al., 2007; Hall, Lightbody, & Reiss, 2008; Kaufmann et al., 2004; Philofsky, Hepburn, Hayes, Hagerman, & Rogers, 2004; Rogers, Wehner, & Hagerman, 2001). There is some evidence that autism in boys with Fragile X syndrome is associated with greater language and communication difficulties (e.g., Bailey, Hatton, Skinner, & Mesibov, 2001; Estigarribia et al., 2011; Roberts, Martin, et al., 2007).

Individuals with autism who do not have Fragile X syndrome have also been reported to use verbal perseveration (Baker, 2000; Koegel & Frea, 1993; Ross, 2002). Thus, it may be that individuals with both Fragile X syndrome and autism produce more perseveration than those with Fragile X syndrome only. This possibility, however, has been largely unexplored. Several studies reviewed earlier in this paper (i.e., Belser & Sudhalter, 1995; Fryns, et al., 1984; Hanson, et al., 1986; Madison, et al., 1986; Prouty, et al., 1988; Wolf-Schein, et al., 1987) did not report the autism status of participants with Fragile X syndrome. Other studies reviewed earlier (i.e., Ferrier, Bashir, Meryash, Johnston, & Wolff, 1991; Levy, et al., 2006; Murphy & Abbeduto, 2007; Sudhalter, et al., 1990; Sudhalter, et al., 1991) excluded only individuals with Fragile X syndrome who met full criteria for a diagnosis of autistic disorder. This suggests that the remaining sample included those who met criteria for the "subthreshold" diagnosis of pervasive developmental disorder–not otherwise specified (American Psychiatric Association, 2000) and therefore were on the autism spectrum.

In the only published study to date that investigated the role of autism status in perseveration in Fragile X syndrome (Roberts, Martin, et al., 2007), the frequency of perseveration during conversation did not significantly differentiate boys with Fragile X syndrome with autism spectrum disorder (classified as "autism" or "autism spectrum" on the Autism Diagnostic Observation Schedule [ADOS]; Lord, Rutter, DeLavore, & Risi, 2001) and boys with

Fragile X syndrome without autism spectrum disorder . However, a moderate effect size (*d* = .51) suggested that significant group differences, with perseveration more common in boys with Fragile X syndrome and autism spectrum disorder, may have been found with a larger sample. Note, too, that Roberts, Martin, and colleagues did not analyze separately different types of perseveration.

Perseveration and Language Sampling Context

With the exception of one study by Murphy and Abbeduto (2007), perseveration in Fragile X syndrome has only been examined in conversation samples. As noted by Abbeduto, Benson, Short, and Dolish (1995), a thorough examination of language production skills of individuals with ID (and also of typically developing individuals) involves both conversational and narrative samples, given the advantages of one context over another (e.g., increased standardization of narration, larger utterance corpus of conversation) and the effects of sampling context on various language skills. For adolescent males and females with Fragile X syndrome, Murphy and Abbeduto (2007) reported that repetition of topics occurred more often in a structured conversation than in narration from a wordless picture book. The topic of an utterance was considered repetitive if it recurred without the addition of new information, or if it was reintroduced in a way that was tangential or unrelated to the current focus of discourse. This finding provides additional support for examining context effects on perseveration in Fragile X syndrome in the present study.

Current Study

To date, no studies have compared individuals with Fragile X syndrome with and without autism spectrum disorder to individuals with other forms of ID and typically developing children vis-à-vis different types of perseveration in multiple language contexts. In the current study, types of perseveration were based on the taxonomy described by Murphy and Abbeduto (2007) and included utterance-level (successive repetition of a word, phrase, or utterance), topic (excessive repetition of a topic, theme, or idea), and conversational device (excessive repetition of rote sayings and phrases). The inclusion of younger typically developing boys of similar nonverbal mental age helps to determine whether any or all types of perseveration exhibited by boys with Fragile X syndrome may be attributed to developmental level. The comparison sample of boys with Down syndrome helped to determine whether certain or all types of perseveration are specific to Fragile X syndrome, or whether they can be attributed more generally to the presence of ID. The current study was designed to address the following research questions:

1. Do boys with Fragile X syndrome, boys with Down syndrome, and younger typically developing boys differ in the production of utterance level, topic, and conversational device perseveration during social interaction?
2. Do the boys differ in the production of the different types of perseveration during narration?
3. For boys with Fragile X syndrome, what is the role of autism status in perseveration?
4. For all boys, what is the role of language sampling context in perseveration?

We hypothesized that, after controlling for nonverbal mental age, boys with Fragile X syndrome (with and without autism spectrum disorder) would produce more perseveration during social interaction than boys with Down syndrome and typically developing boys, and that boys with both Fragile X syndrome and autism spectrum disorder would produce more perseveration during interaction than boys with Fragile X syndrome only. We hypothesized

further that topic perseveration for boys with Fragile X syndrome, regardless of autism status, would be more common in social interaction as compared with narration.

Method

Participants

The boys who participated in this study were drawn from a larger longitudinal investigation of the speech and language skills of boys with Fragile X syndrome (FXS) with autism spectrum disorder (FXS-ASD), Fragile X syndrome only (FXS-O), Down syndrome (DS), and typically developing (TD) boys (Roberts, Price, et al., 2007). The participants with FXS and DS were recruited from schools, genetic clinics, developmental clinics, and physicians' offices in the eastern United States. Additionally, boys with FXS were recruited from the Research Participant Registry Core of the Carolina Institute for Developmental Disabilities at the University of North Carolina at Chapel Hill. TD boys were recruited from schools, childcare centers, and physicians' offices in North Carolina. Because females with FXS generally show less severe impairments than males with FXS because females have a second, normally functioning X chromosome (Hagerman & Hagerman, 2002; Loesch et al., 2002), only boys were included in this study.

At study entry, all boys were producing at least 40 words expressively and combining at least two words (i.e., mean length of utterance [MLU] > 1.1) according to parent report, and all groups displayed similar distributions of nonverbal mental ages on the Brief IQ composite of the Leiter International Performance Scale—Revised (Leiter-R; Roid & Miller, 1997). Boys were excluded if their average hearing threshold was greater than 25 dB HL in the better ear, based on a screening across 500; 1,000; 2,000; and 4,000 Hz; using a Grason Stadler GSI 16 or 17, or MAICO MA 40 audiometer. Boys with DS and TD boys first were screened for Fragile X syndrome and autism and excluded if they showed signs of either disorder. In subsequent testing, boys with DS and TD boys were excluded if they scored in the autism or autism spectrum range on the ADOS (Lord, Rutter, DeLavore, & Risi, 2001). In the larger study, 1 TD boy and 6 boys with DS failed the hearing screening and 2 boys with DS scored in the autism spectrum range on the ADOS and therefore were not targeted for coding in the present study. TD boys included in this study did not have a developmental disability or speech and language difficulties. Spoken English was the primary language for all children. See Table 1 for background characteristics of all participant groups. Groups did not differ significantly in mental age, $F(3, 95) = .35$, $p = .7989$. Groups did, however, differ significantly in chronological age, $F(3, 95) = 43.22$, $p < .001$; the TD group was significantly younger than all other groups with no other significant group differences.

In the current investigation, boys with Fragile X syndrome were divided into two groups according to autism status: (a) FXS with ASD (FXS-ASD) and (b) FXS without ASD, or "FXS only" (FXS-O). The group of boys with FXS and ASD included those boys with FXS who were classified by the ADOS as having autism or autism spectrum. See the Assessments section for a description of the ADOS.

Fragile X syndrome without autism spectrum disorder (FXS-O)

Thirty boys with FXS only (FXS-O) participated in the study. These boys ranged in chronological age (CA) from 6.0 to 15.8 years ($M = 11.5$, $SD = 2.3$) and in nonverbal mental age (MA) from 4.0 to 7.7 years ($M = 5.5$, $SD = 0.7$). Eighty-three percent of the boys were Caucasian; 13% were African American; and 3% were of a different ethnicity. All boys had a diagnosis of full mutation FXS, which was confirmed by DNA analyses.

Fragile X syndrome with autism spectrum disorder (FXS-ASD)

Twenty-nine boys with FXS who also had ASD (FXS-ASD) were study participants. These boys ranged in CA from 6.4 to 15.5 years (M = 10.6, SD = 2.8) and in nonverbal MA from 4.7 to 6.6 years (M = 5.3, SD = 0.4). Ninety percent of the boys were Caucasian, and 10% were African American. All boys had a diagnosis of full mutation FXS, which was confirmed by DNA analyses.

Down syndrome (DS)

Twenty-seven boys with DS participated in this study. These boys ranged in CA from 6.3 to 16.0 years (M = 10.1, SD = 2.8) and in nonverbal MA from 4.1 to 8.2 years (M = 5.2, SD = 0.9). Ninety-six percent of the boys were Caucasian; and 4% were African American. Parents reported that trisomy 21 was the source of DS for all boys.

Typically developing (TD)

Twenty-five TD boys were study participants. These boys ranged in CA from 3.9 to 6.5 years (M = 5.2, SD = 0.8) and in nonverbal MA from 3.6 to 8.2 years (M = 5.2, SD = 1.1). Sixty-eight percent of the boys were Caucasian; 24% were African American; and 8% were of a different ethnicity.

Assessments—Each boy was assessed either at his school, in his home, or at a university research center. Testing occurred in a quiet area and lasted for approximately 6 hr and included several breaks. All sessions were videotaped with a Sony Digital8 video camera (Model DCR-TVR27) and were audiotaped with a portable Digital Auditory Tape TASCAM (DA-P1).

Nonverbal cognition

The Brief IQ composite of the Leiter-R was used to assess nonverbal cognition. The Brief IQ composite is based on the results of four subtests: Figure Ground, Form Completion, Sequential Order, and Repeated Patterns. Participants were asked to locate an item in a picture, arrange items according to a pattern, or select the next item in a sequence. The Leiter-R was standardized on 1,719 individuals aged from 2 years to 20 years. High levels of reliability have been reported for the Brief IQ composite, with alpha reliability coefficients for the subtests ranging from .75 to .88 and a test–retest coefficient of .96. The Leiter-R also correlates strongly (.85 to .86) with other regularly used IQ tests. Age equivalent scores were computed for all children in the present study according to published norms.

Autism spectrum disorder in Fragile X syndrome

Autism classification of the boys with Fragile X syndrome was determined through administration of the ADOS (Lord et al., 2001), a standard observation of communicative and social behaviors that distinguishes autism from other developmental disorders and from typical behavior. During the ADOS, an examiner engages the child in a series of structured and semi-structured activities that are designed to provide the child with opportunities to exhibit communicative and social behaviors that are either typical or are indicative of ASD. The ADOS yields categorical scores of autism, spectrum, and no autism. Trained examiners scored videotapes of the ADOS, and reliability computed on 16% of the boys was .93 on diagnosis (range = .81–1.00) and .89 for individual items (range = .83–.96). Eight of the boys with Fragile X syndrome in the current study were classified as having autism; 21 were classified as having autism spectrum; and 30 were classified as having neither autism nor autism spectrum. The FXS-ASD group included the boys classified as either autism or autism spectrum.

Language Sampling and Coding Procedures

Social interaction

Social interactions that occurred during administration of the ADOS were examined in this study. A nationally assembled group of experts in language in ASD recently recommended the ADOS as a context for collecting a natural language sample (Tager-Flusberg et al., 2009). To obtain a language sample that was most reflective of genuine conversation, only certain activities of the ADOS were coded. These activities included free play, balloon and bubble blowing, a demonstration task, and/or a pretend birthday activity. Perseveration produced during social interaction was coded initially from language transcripts. Next, coders viewed a videotape of the interaction in order to determine (a) referents that were unclear in the transcript and (b) pause time between repetitive utterances. The first 100 intelligible utterances from the selected contexts were coded. For one boy with FXS-ASD and one TD boy, only 96 and 97 utterances, respectively, were available for coding.

Narration

The Renfrew Bus Story (Crowley & Glasgow, 1994) was administered to obtain samples of picture-supported story retelling. In this story, a bus runs away from its driver and goes through a series of events before ultimately being saved by the driver. The story booklet is four pages in length with three pictures to a page. Following the test protocol, the examiner read the story to the child from a script (15 sentences in length) while showing the 12 pictures to the child that went along with the story. Next, the examiner asked the child to tell the story while looking through the book (i.e., “Now you tell me the story. Once upon a time there was a …”). Perseveration was coded from the language transcripts. For a narrative language sample to be eligible for coding, the child needed to produce at least 9 intelligible utterances. On average, boys with FXS-O produced 16.3 (SD = 5.9; range = 9–33) intelligible utterances; boys with FXS-ASD produced 16.7 (SD = 5.2; range = 9–27); boys with DS produced 17.6 (SD = 4.7; range = 11–27); and TD boys produced 19.1 (SD = 7.3; range = 12–42) intelligible utterances during narration.

Transcription

Trained research assistants transcribed the social interaction and narrative language samples from videotapes using the Child Language Data Exchange System (CHILDES; MacWhinney, 1995). Utterance boundaries were determined by a change in speakers, intonation contour, and an obvious pause. In cases of more than two conjoined independent clauses, utterances were separated at the second conjunction following Systematic Analysis of Language Transcripts (SALT; Miller & Chapman, 2008) guidelines. A second trained researcher verified and corrected all original transcripts using high-quality audio recordings. For reliability purposes, a random subset of original transcripts (12% for social interaction and 14% for narration) was independently verified and corrected by a third researcher via the audio recordings and morpheme-by-morpheme agreement calculated by comparing transcripts of the second and third researchers. For the social interaction context, agreement was 91% overall, 94% for boys with FXS-O (n = 3), 89% for boys with FXS-ASD (n = 3), 87% for boys with DS (n = 3), and 93% for TD boys (n = 3). For the narrative context, agreement was 95% overall, 98% for boys with FXS-O (n = 3), 90% for boys with FXS-ASD (n = 3), 91% for boys with DS (n = 4), and 98% for TD boys (n = 4).

Perseveration coding categories

Criteria used for determining the occurrence of perseveration was based on that described by Murphy and Abbeduto (2007). Utterances were coded for the following: (a) utterance-level perseveration, (b) topic perseveration, and (c) conversational device perseveration, as described below.

Utterance-level perseveration

Utterance-level perseveration was coded when a child repeated a word (e.g., "He he he he he goes."), phrase (e.g., "Put it back in the bag in the bag."), or entire utterance (e.g., "We need to put these away. We need to put these away."). To be coded as utterance-level perseveration, the linguistic units had to be spoken in immediate succession, and the repetition had to be identical or almost identical to the original unit (the two could differ by only one morpheme). If a child repeated himself for emphasis (e.g., "I really *really* like ice cream") or to repair a communicative breakdown (e.g., repeating himself because the examiner did not respond), these utterances were not coded as perseverations.

Topic perseveration

Topic perseveration was coded when a child repeated a topic, theme, or idea two or more times in an excessive manner. Repetitions of this type did not need to be produced in immediate succession; topics, themes, and ideas could be reintroduced at any point throughout the language sample. If a child was offering obligatory information to a question or responding to a request for clarification, topic perseveration was not coded. To determine if repetitions were excessive, coders considered criteria such as whether (a) the examiner could not redirect the child away from a topic; (b) the child offered the same information that he had provided previously, without adding any new information; and (c) the reintroductions were noncontingent (tangential or unrelated) with respect to the current focus of discourse. The following is an example of topic perseveration from a study participant with FXS-ASD during social interaction:

> The examiner tells the child, "When we get to the bottom of the bag, we'll see if we found any masks," and the child immediately says, "And with blood?" Throughout the remainder of the interaction, the child produces the following utterances: "Where the blood? …Where the blood? … the blood? You have blood somewhere? … Do you have a blood mask with blood? … Do you have fake blood? … Do you have blood? … It was blood. … Screen blood. … Do you have blood? Do you have screen blood? … Where's the blood at? Where is it? … Do you have a blood mask?"

Conversational device perseveration

Conversational device perseveration was defined as the excessive repetition of rote or conventional words, phrases, or sayings (e.g., "Oh man," "Okay," "I don't know," "cool," and "hmm"). Repetition of a conversational device was considered perseverative if the device occurred three or more times. Expressions that were obligatory (e.g., "uh huh," "mhm," or "yeah" in response to a yes/no question) were not coded as conversational device perseverations.

Coding reliability

Two coders independently coded the language samples. For each language sampling context, reliability was computed on a random subset (12%) of the samples using the kappa statistic (Cohen, 1960). For social interaction, mean intercoder agreement computed on 12 boys (3 boys from each group) was .81 for utterance-level, .69 for topic and .77 for conversational device perseveration. For narration, overall reliability computed on 3 boys from each group was .93 for utterance-level, .77 for topic, and 1.0 for conversational device perseveration. Intercoder agreements using kappa statistics are generally lower than those computed by other methods since the kappa takes chance agreement into account (Fleiss, 1981); kappa statistics ranging from .61 to .80 are considered *substantial* and those between .81 and 1.0 are considered *almost perfect* (Landis & Koch, 1977).

Computation of variables

Three variables each were computed for the social interaction and narrative contexts: (a) the proportion of utterance-level perseveration, (b) the proportion of topic perseveration, and (c) the proportion of conversational device perseveration. For example, the proportion of topic perseveration was computed by dividing the number of utterances coded as topic perseveration by the total number of intelligible utterances.

Data Analysis Strategy

Group (FXS-O, FXS-ASD, DS, and TD) differences on each of the measures of perseveration (proportion of utterances classified as utterance-level, topic, and conversational device) were tested using one multivariate model, run as a hierarchical linear model (HLM) controlling for nonverbal MA. A context variable indicated whether the proportion of perseverative utterances was from social interaction or narration. Interactions with context were included to test for differential group differences and type effects. Nonverbal MA (as measured by the Leiter-R) was included in the model to adjust the estimated means. The repeated measurement within subject across contexts results in dependence across those subjects. HLM manages that dependence through the estimation of random effects, essentially estimating and controlling for effects for each subject (see Bryk & Raudenbush, 1992; Burchinal, Nelson, & Poe, 2006, for complete discussions). Random intercepts were included in these models.

Results

Descriptive Analyses

Means and standard deviations (not adjusted for nonverbal MA) for the proportion of utterances categorized as utterance-level, topic, and conversational device perseveration for the contexts of social interaction and narration are reported in Table 2. On average, utterance-level perseveration occurred in 10% of utterances during social interaction for boys with FXS-O, 13% for boys with FXS-ASD, 7% for boys with DS, and 10% for TD boys. Topic perseveration occurred in 9% of utterances for boys with FXS-O, 14% for boys with FXS-ASD, and 5% for both boys with DS and TD boys. Conversational device perseveration occurred in 4% of utterances for boys with FXS-O, 3% for boys with FXS-ASD, and 5% for both boys with DS and TD boys. In narration, utterance-level perseveration occurred in 11% of utterances for boys with FXS-O, 14% for boys with FXS-ASD, 11% for boys with DS, and 21% for TD boys on average. Topic (3% to 4%) and conversational device perseveration (0% to 1%) occurred infrequently during narration for all groups.

Group Comparisons

The HLM was run via the computer software program SAS Proc Mixed (SAS Institute, 2002). Results indicated a significant effect of group on the production of perseveration, $F(3, 101) = 4.28$, $p = .0069$. In addition, a significant 3-way interaction between diagnosis, type of perseveration, and context, $F(6, 470) = 2.28$, $p = .0349$, indicated that between-group differences varied by type of perseveration, and that these differences varied also by context. The between-group differences, adjusted means (adjusted for nonverbal MA), and standard errors for the social interaction context are presented in Table 3. The boys with FXS-ASD produced significantly more utterance-level perseveration ($M = 0.13$) than did the boys with DS ($M = 0.07$, $p = .0023$) and significantly more topic perseveration ($M = 0.14$) than boys with FXS-O ($M = 0.09$, $p = .0097$), boys with DS ($M = 0.05$, $p < .0001$), and TD boys ($M = 0.05$, $p < .0001$). The groups did not differ significantly on the measure of conversational device perseveration.

The between-group differences, adjusted means, and standard errors for the narrative context are presented in Table 4. The TD boys produced significantly more utterance-level perseveration ($M = 0.21$) than boys with FXS-O ($M = 0.11$, $p < .0001$), FXS-ASD ($M = 0.14$, $p = .0016$), and DS ($M = 0.11$, $p < .001$). The groups did not differ significantly on either topic or conversational device perseveration during narration.

Comparison of Social Interaction and Narration

The HLM also addressed the effects of language sampling context on perseveration in each of the four groups, controlling for nonverbal MA. The TD boys produced significantly more utterance-level perseveration in narration than in social interaction ($p < .001$). The boys with FXS-O and FXS-ASD produced significantly more topic perseveration in interaction than in narration ($p = .006$ and $p < .001$, respectively). The boys with FXS-ASD, boys with DS, and TD boys produced significantly more conversational device perseveration in interaction than in narration ($p < .001$, $p = .012$, and $p = .035$, respectively).

Discussion

Verbal perseveration is a frequently reported characteristic of the language profile of males with Fragile X syndrome. Moreover, many researchers have argued that verbal perseveration is a hallmark or defining feature of the behavioral phenotype of Fragile X syndrome (e.g., Abbeduto, et al., 2007; Roberts, Chapman, Martin, et al., 2008; Sudhalter, et al., 1990). In contrast to previous studies (Levy, et al., 2006; Roberts, Martin, et al., 2007), we did not find statistically significant differences in verbal perseveration during social interaction between boys with Fragile X syndrome without ASD (FXS-O) and boys with Down syndrome or typically developing boys. In the earlier studies, however, researchers did not compare groups on different types of perseveration; thus, the current findings are not directly comparable with previous findings. In the present investigation, boys with FXS-O produced topic perseveration in 9% of utterances during interaction, compared with 5% of utterances for typically developing boys and 5% of utterances for boys with Down syndrome. Although these differences were not statistically significant, the effect sizes were medium to large ($d = .54$ and .71, respectively). This suggests that we might find significant differences with a larger sample size.

An unexpected finding was that typically developing boys produced more utterance-level perseveration during narration (21% of utterances) than did boys with FXS-O (11%), FXS-ASD (14%), and Down syndrome (11%). At present, the factors underlying these group differences are unclear. According to Murphy and Abbeduto (2007), utterance-level perseveration may reflect word-finding difficulties, consistent with the word-retrieval account of perseveration in Fragile X syndrome introduced by Sudhalter and colleagues (1990). For example, a child might say, “I want the want the want the ladder,” repeating the linguistic unit “want the” while searching for the word “ladder.” Perhaps the typically developing boys were expending more effort than the other boys in their attempts to retrieve story elements from memory. If this was the case, then the pronounced occurrence of utterance-level perseveration in the typically developing group might reflect better story-retelling skills or a greater attempt to retell the story accurately and completely. For example, one typically developing boy produced the following statement that was coded for utterance-level perseveration: “When when the driver found out he was in the water, he called he called a tow truck to pull him out of the water.” In fact, these narrative samples were previously examined for recall of story grammar elements (see Estigarribia, et al., 2011), and typically developing boys recalled more story actions than boys with Fragile X syndrome with and without ASD.

Perseveration and Autism Status

In the present investigation, the boys with FXS-ASD produced significantly more topic perseveration (14% of utterances) during social interaction than all other groups. This finding adds to a growing body of research indicating differences in language characteristics between individuals with Fragile X syndrome depending on their autism status (e.g., Bailey et al., 2001; Roberts, Martin, et al., 2007; Rogers et al., 2001). Currently, the factors underlying the observed group differences for topic perseveration during social interaction and the apparent effect of autism status are unclear. Many researchers (e.g., Belser & Sudhalter, 1995; Cornish, Sudhalter, & Turk, 2004; Murphy & Abbeduto, 2007) have attributed perseveration in individuals with Fragile X syndrome to the excessive arousal ("hyperarousal") and/or anxiety that are well documented features of the behavioral phenotype (Cordeiro, et al., 2011; Hagerman, 2002; Hessl et al., 2001). According to this account, an individual becomes hyperaroused and/or anxious during social interactions because the social gaze of a communication partner, in particular, makes him or her "uncomfortable," and produces verbal perseveration in response (Belser & Sudhalter, 1995; Cornish, et al., 2004; Sudhalter, et al., 1990).

Findings from one recent study suggest that social anxiety occurs more frequently in individuals with both Fragile X syndrome and autism than in those with Fragile X syndrome alone (Cordeiro, et al., 2011). Thus, it is possible that increased anxiety in individuals with both Fragile X syndrome and autism leads to increased perseveration in these individuals. Alternatively, if perseveration is a hallmark of Fragile X syndrome, it is possible that some individuals with Fragile X syndrome may receive a classification of autism or autism spectrum *because* of their increased topic perseveration, which could reduce the amount of reciprocal conversation and affect the overall quality of rapport between the individual and his or her communication partner.

The boys with FXS-ASD in this study also produced significantly more utterance-level perseveration (13% of utterances) during social interaction than did boys with Down syndrome (7%). If utterance-level perseveration is, in some instances, reflective of the word retrieval process (as mentioned previously), perhaps word-finding skills during interaction are more impaired in boys with FXS-ASD than they are in boys with Down syndrome.

Perseveration and Language Sampling Context

The present study was the first to compare boys with Fragile X syndrome (with and without ASD), boys with Down syndrome, and typically developing boys on measures of perseveration during both a social interaction and a narration context. Our findings indicate that the context of language sampling plays a role in perseveration. Boys with Fragile X syndrome, both with and without ASD, produced significantly more topic perseveration during social interaction with the examiner than when narrating a story. This is consistent with the work of Murphy and Abbeduto (2007), who found that adolescent males with Fragile X syndrome (without autistic disorder) repeated topics more often in conversation than in picture-supported storytelling. Murphy and Abbeduto offered two explanations for this finding. First, they proposed that the interpersonal demands of conversation such as maintaining eye contact (versus narration from a picture book) may have caused the individual with Fragile X syndrome to become hyperaroused, consistent with the hyperarousal hypothesis of perseveration discussed previously. Second, they reasoned that the added structure of the narrative format may have lessened the effects of impairment in executive functioning. Indeed, providing picture support and two presentations of the book in both studies may have reduced some of the demands of self-formulated storytelling and resulted in reduced perseverations arising from linguistic-executive dysfunction. In the current study, children were additionally provided with a structured script, which may have

further reduced some narrative discourse demands notwithstanding placing demands on short-term memory. Findings from the two studies together suggest that perseveration is relatively infrequent for boys with Fragile X syndrome during picture-supported storytelling whether or not a structured script is provided.

As previously discussed, the surprising finding that typically developing boys produced significantly more utterance-level perseveration in narration than in social interaction may have resulted from their efforts to recall the story elements precisely. Additionally, narratives have been found to elicit longer and more complex utterances from typically developing individuals than does conversation (Dollaghan, Campbell, & Tomlin, 1990; Thordadottir, 2008), and complex and longer sentences are more likely to produce dysfluencies in the typically developing population than are shorter and less complex sentences (Rispoli & Hadley, 2001). Results of the present study suggest that at least some of these dysfluencies may manifest themselves as perseverations. Perhaps perseveration at the utterance level is reflective of typically developing children's attempts to produce more complex syntax.

Study Strengths and Limitations

The strengths of the study reported here are at least threefold. First, sample sizes of boys with Fragile X syndrome, boys with Down syndrome, and typically developing boys were large compared with sample sizes of many previous studies. Second, the mental age–matched comparison groups of boys with Down syndromeand typically developing boys helps to determine whether perseveration exhibited by boys with Fragile X syndrome is a function of developmental level or ID in general. Third, boys with Fragile X syndrome both with and without ASD were included so we could investigate the relationship between autism status and perseveration in Fragile X syndrome. Finally, we examined three different types of perseveration in two different language sampling contexts.

The present study also has several limitations that have implications for future research. First, we used a picture-supported story retelling task for our narrative sample, and such a task is not necessarily representative of a child's performance on either story-retelling tasks that lack picture supports or story generation tasks with or without picture supports. This narrative task also elicited a small number of utterances compared with the social interaction context. Future studies should examine perseveration in different types of narrative tasks and with longer language samples, including personal narratives. Second, the FXS-ASD group consisted of boys who were classified as autism or autism spectrum on the ADOS (Lord et al., 2001), but the other current gold standard measure of autism, the Autism Diagnostic Interview—Revised (ADI-R; Lord, Rutter, & Le Couteur, 1994), should be used to classify children in addition to the ADOS in forthcoming studies. Unlike the ADOS, which assesses only current functioning with an unfamiliar person (the examiner) in one setting, the ADI-R gathers information about the individual's functioning across a variety of contexts from a parent. Thus, use of both instruments to assess individuals with Fragile X syndrome might well improve valid classifications of autism. Third, adding a comparison group of children with ASD who do not have Fragile X syndrome would further our understanding of the contribution of autism status to perseveration in boys with both Fragile X syndrome and ASD. Fourth, while the observed group and language context differences and within-group variability prompt discussion of underlying mechanisms of perseveration in boys with Fragile X syndrome, these mechanisms were not directly examined in the current study. Future studies should investigate predictors of individual differences in perseveration in boys with Fragile X syndrome, such as anxiety/hyperarousal, executive functions, and autism severity. Findings from predictor studies may inform interventions that target perseveration, such as addressing anxiety to manage perseveration if anxiety is indeed found to be a significant predictor of perseveration.

Clinical Implications

From a clinical perspective, the problem of verbal perseveration is significant for several reasons. Frequent perseveration may have a profound negative impact on conversational skills, presenting a major obstacle to social interaction and frustrating caregivers and other communication partners. As pointed out by Helm-Estabrooks and Albert (2004), perseveration may also affect performance during assessments of various cognitive linguistic domains and skew test results by blocking underlying skills.

Findings of the current investigation have several important implications for assessment and intervention. Given that boys with FXS-ASD produced more topic perseveration than did boys with FXS-O, the diagnosis of ASD should be a consideration for assessment. That being said, considerable individual differences occurred in our sample. For example, one 8-year-old boy with FXS-ASD produced topic perseveration in 27% of utterances during social interaction, whereas another 8-year-old boy with FXS-ASD used topic perseveration in just 4% of utterances. Similarly, for one 14-year-old boy with FXS-O, 22% of utterances during social interaction included topic perseveration. In contrast, another 14-year-old boy with FXS-O produced topic perseveration in only 2% of utterances. Whereas perseveration was overall very infrequent in the Down syndrome group, one 7-year-old boy with Down syndrome produced topic perseveration in 25% of utterances during social interaction. Thus, despite statistically significant differences across diagnostic groups, high individual variability in our data indicates that perseveration should be a focus of comprehensive language assessments regardless of diagnosis. The context for assessing language also should be considered, given our finding that boys with Fragile X syndrome, regardless of autism status, perseverated on topics more often during play-based interaction than when retelling a story.

Given the negative effects of perseveration on communication and social interaction, speech and language interventions for boys with Fragile X syndrome who produce verbal perseveration should include goals for decreasing these behaviors. Roberts, Chapman, Martin, and Moskowitz (2008) advised that clinicians first try to identify possible causes of perseveration (e.g., anxiety) and possible functions of a particular child's perseveration in a given context (e.g., escaping social interaction, maintaining social interaction, gaining reassurance or a desired item/activity) and select intervention strategies based on the identified functions. Specific strategies recommended by Scharfenaker, O'Connor, Stackhouse, Braden, and Gray (2002) for managing verbal perseveration in individuals with Fragile X syndrome included monitoring anxiety levels, utilizing verbal redirection, and allowing additional processing time. Our finding that topic perseveration was frequent for boys with Fragile X syndrome during social interaction but not during picture-supported narration is consistent with results from the Murphy and Abbeduto (2007) study. Based on this finding, McDuffie, Chapman, and Abbeduto (2008) suggested that clinicians use pictures to support conversational performance and decrease perseveration. Indeed, incorporating visual supports into social interaction and conversation may help to increase predictability, lessen anxiety, and reduce perseveration.

A method suggested for managing the perseverative behaviors of individuals with aphasia, which is to raise such behaviors to a "level of awareness" for conscious control of perseverative tendencies (Helm-Estabrooks & Albert, 2004, p. 100), may also be successful with some children with Fragile X syndrome. Finally, as previously mentioned, reliable identification of the underlying mechanisms of types of perseveration in children with Fragile X syndrome would inform intervention approaches.

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References

Abbeduto L, Benson G, Short K, Dolish J. Effects of sampling context on the expressive language of children and adolescents with mental retardation. Mental Retardation. 1995; 33(5):279–288. [PubMed: 7476250]

Abbeduto L, Brady N, Kover ST. Language development and Fragile X syndrome: Profiles, syndrome-specificity, and within-syndrome differences. Mental Retardation and Developmental Disabilities Research Reviews. 2007; 13(1):36–46. [PubMed: 17326110]

Abbeduto L, Hagerman RJ. Language and communication in Fragile X syndrome. Mental Retardation and Developmental Disabilities Research Reviews. 1997; 3(4):313–322.

American Psychiatric Association. Diagnostic and statistical manual of mental disorders. Washington, DC: Author; 2000. (Text rev.)

Bailey DB, Hatton DD, Skinner M, Mesibov G. Autistic behavior, FMR1 protein, and developmental trajectories in young males with Fragile X syndrome. Journal of Autism and Developmental Disorders. 2001; 31(2):165–174. [PubMed: 11450815]

Baker MJ. Incorporating the thematic ritualistic behaviors of children with autism into games: Increasing social play interactions with siblings. Journal of Positive Behavior Interventions. 2000; 2(2):66–84.

Belser RC, Sudhalter V. Arousal difficulties in males with Fragile X syndrome: A preliminary report. Developmental Brain Dysfunction. 1995; 8(4):270–279.

Bennetto, L.; Pennington, BF. Neuropsychology. In: Hagerman, RJ.; Hagerman, PJ., editors. Fragile X syndrome: Diagnosis, treatment, and research. Vol. 3. Baltimore, MD: Johns Hopkins University Press; 2002. p. 206-248.

Bregman JD, Leckman JF, Ort SI. Fragile X syndrome: Genetic predisposition of psychopathy. Journal of Autism and Developmental Disorders. 1988; 18:343–354. [PubMed: 3170453]

Bryk, AS.; Raudenbush, SW. Hierarchical linear models: Applications and data analysis methods. Newbury Park, CA: Sage; 1992.

Burchinal M, Nelson L, Poe M. Growth curve analysis: An introduction to various methods for analyzing longitudinal data. Monographs of the Society for Research in Child Development. 2006; 71:65–87.

Clifford S, Dissanayake C, Bui QM, Huggins R, Taylor AK, Loesch DZ. Autism spectrum phenotype in males and females with Fragile X full mutation and premutation. Journal of Autism and Developmental Disorders. 2007; 37(4):738–747. [PubMed: 17031449]

Coffee B, Keith K, Albizua I, Malone T, Mowrey J, Sherman SL, Warren ST. Incidence of Fragile X syndrome by newborn screening for methylated FMR1 DNA. American Journal of Human Genetics. 2009; 85(4):503–514. [PubMed: 19804849]

Cohen J. A coefficient of agreement for nominal scales. Educational and psychological Measurement. 1960; 20(1):37–46.

Cordeiro L, Ballinger E, Hagerman R, Hessl D. Clinical assessment of DSM-IV anxiety disorders in Fragile X syndrome: Prevalence and characterization. Journal of Neurodevelopmental Disorders. 2011; 3:57–67. [PubMed: 21475730]

Cornish K, Sudhalter V, Turk J. Attention and language in Fragile X. Mental Retardation and Developmental Disabilities. 2004; 10:11–16.

Crowley, J.; Glasgow, C. The Renfrew bus story. Centreville, DE: Centreville School; 1994. (American ed.)

Dollaghan CA, Campbell TF, Tomlin R. Video narration as a language sampling context. Journal of Speech and Hearing Disorders. 1990; 55:582–590. [PubMed: 2381199]

Dykens, E.; Hodapp, RM.; Finucane, BM. Genetics and mental retardation syndromes: A new look at behavior and interventions. Baltimore, MD: Paul H. Brookes; 2000.

Estigarribia B, Martin GE, Roberts JE, Spencer A, Gucwa A, Sideris J. Narrative skill in boys with Fragile X syndrome with and without autism spectrum disorder. Applied Psycholinguistics. 2011; 32(2):359–388. [PubMed: 21516264]

Fernandez-Carvajal I, Walichiewicz P, Xiaosen X, Pan R, Hagerman PJ, Tassone F. Screening for expanded alleles of the FMR1 gene in blood spots from newborn males in a Spanish population. Journal of Molecular Diagnostics. 2009; 11(4):324–329. [PubMed: 19460941]

Ferrier LJ, Bashir AS, Meryash DL, Johnston J, Wolff P. Conversational skills of individuals with Fragile-X syndrome: a comparison with autism and Down syndrome. Developmental Medicine and Child Neurology. 1991; 33(9):776–788. [PubMed: 1834506]

Finestack LH, Richmond EK, Abbeduto L. Language development in individuals with Fragile X syndrome. Topics in Language Disorders. 2009; 29(2):133–148. [PubMed: 20396595]

Fleiss JL. Balanced incomplete block designs for inter-rater reliability studies. Applied Psychological Measurement. 1981; 5:105–112.

Fryns JP, Jacobs J, Kleczkowska A, van den Berghe H. The psychological profile of the Fragile X syndrome. Clinical Genetics. 1984; 25(2):131–134. [PubMed: 6705249]

Hagerman P. The Fragile X prevalence paradox. Journal of Medical Genetics. 2008; 45(8):498–499. [PubMed: 18413371]

Hagerman, R. The physical and behavioral phenotype. In: Hagerman, RJ.; Hagerman, PJ., editors. Fragile X syndrome: Diagnosis, treatment, and research. 3. Baltimore, MD: Johns Hopkins University Press; 2002. p. 3-87.

Hagerman, R.; Hagerman, P., editors. Fragile X syndrome: Diagnosis, treatment, and research. 3. Baltimore, MD: Johns Hopkins University Press; 2002.

Hall SS, Lightbody AA, Reiss AL. Compulsive, self-injurious, and autistic behavior in children and adolescents with Fragile X syndrome. American Journal of Mental Retardation. 2008; 113(1):44–53. [PubMed: 18173299]

Hanson DM, Jackson AW III, Hagerman RJ. Speech disturbances (cluttering) in mildly impaired males with the Martin-Bell/Fragile X syndrome. American Journal of Medical Genetics. 1986; 23(1–2):195–206. [PubMed: 3953648]

Helm-Estabrooks, N.; Albert, ML. Manual of Aphasia and Aphasia therapy. 2. Austin, TX: Pro-Ed; 2004.

Hessl D, Dyer-Friedman J, Glasrer B, Wisbek J, Barajas RG, Taylor A, Reiss A. The influence of environmental and genetic factors on behavior problems and autistic symptoms in boys and girls with Fragile X syndrome. Pediatrics [Evanston]. 2001; 108(5):88–104.

Hooper SR, Hatton DD, Baranek GT, Roberts JP, Bailey DB. Nonverbal assessment of IQ, attention, and memory abilities in children with Fragile-X syndrome using the Leiter-R. Journal of Psychoeducational Assessment. 2000; 18(3):255–267.

Kaufmann WE, Cortell R, Kau AS, Bukelis I, Tierney E, Gray RM, Cox C, Capone GT, Stanard P. Autism spectrum disorder in Fragile X syndrome: Communication, social interaction, and specific behaviors. American Journal of Medical Genetics. 2004; 129(3):225–234. [PubMed: 15326621]

Koegel RI, Frea WD. Treatment of social behavior in autism through the modification of pivotal social skills. Journal of Applied Behavior Analysis. 1993; 26(3):369–377. [PubMed: 8407685]

Landis JR, Koch GG. The measurement of observer agreement for categorical data. Biometrics. 1977; 33:159–174. [PubMed: 843571]

Levy Y, Gottesman R, Borochowitz Z, Frydman M, Sagi M. Language in boys with Fragile X syndrome. Journal of Child Language. 2006; 33:125–144. [PubMed: 16566323]

Loesch DZ, Bui QM, Grigsby J, Butler E, Epstein JL, Huggins RM, Taylor AK, Hagerman RJ. Effect of the Fragile X status categories and the Fragile X mental retardation protein levels on executive functioning in males and females with Fragile X. Neuropsychology. 2003; 17:646–657. [PubMed: 14599277]

Loesch DZ, Huggins RM, Bui QM, Epstein JL, Taylor AK, Hagerman RJ. Effect of the deficits of Fragile X mental retardation protein on cognitive status of Fragile X males and females assessed by robust pedigree analysis. Journal of Developmental and Behavioral Pediatrics. 2002; 23(6): 416–423. [PubMed: 12476071]

Lord, C.; Rutter, M.; DeLavore, PC.; Risi, S. Autism Diagnostic Observation Schedule. Los Angeles, CA: Western Psychological Services; 2001.

Lord C, Rutter M, Le Couteur A. Autism Diagnostic Interview—revised: A revised version of a diagnostic interview for caregivers of individuals with possible pervasive developmental disorders. Journal of Autism and Developmental Disorders. 1994; 24(5):659–685. [PubMed: 7814313]

MacWhinney, B. The CHILDES project: Tools for analyzing talk. Hillsdale, NJ: Erlbaum; 1995.

Madison LS, George C, Moeschler JB. Cognitive functioning in the Fragile-X syndrome: A study of intellectual, memory and communication skills. Journal of Mental Deficiency Research. 1986; 30(2):129–148. [PubMed: 3735410]

McDuffie, A.; Chapman, R.; Abbeduto, L. Language profiles of adolescents and young adults with Down syndrome and Fragile X syndrome. In: Roberts, JE.; Chapman, RS.; Warren, SF., editors. Speech and language development and intervention in down syndrome and Fragile X syndrome. Baltimore, MD: Paul H. Brookes; 2008. p. 117-141.

Miller, JF.; Chapman, RS. Systematic Analysis of Language Transcripts (SALT) [Computer Software]. Madison, WI: Language Analysis Lab; 2008.

Murphy M, Abbeduto L. Gender differences in repetitive language in Fragile X syndrome. Journal of Intellectual Disability Research. 2007; 51(5):387–400. [PubMed: 17391255]

Philofsky A, Hepburn SL, Hayes A, Hagerman R, Rogers SJ. Linguistic and cognitive functioning and autism symptoms in young children with Fragile X syndrome. American Journal of Mental Retardation. 2004; 109(3):208–218. [PubMed: 15072521]

Prouty LA, Rogers RC, Stevenson RE, Dean JH, Palmer KK, Simensen RJ, Coston GN, Schwartz CE. Fragile X syndrome: growth, development, and intellectual function. American Journal of Medical Genetics. 1988; 30(1–2):123–142. [PubMed: 3177438]

Reiss AL, Dant CC. The behavioral neurogenetics of Fragile X syndrome: Analyzing gene-brain-behavior relationships in child developmental psychopathologies. Development and Psychopathology. 2003; 15:927–968. [PubMed: 14984133]

Rice ML, Warren SF, Betz SK. Language symptoms of developmental language disorders: An overview of autism, Down syndrome, Fragile X, specific language impairment, and Williams syndrome. Applied psycholinguistics. 2005; 26(1):7–27.

Rispoli M, Hadley P. The leading-edge: The significance of sentence disruptions in the development of grammer. Journal of Speech, Language, and Hearing Research. 2001; 44:1131–1143.

Roberts, JE.; Chapman, R.; Martin, GE.; Moskowitz, L. Language of preschool and school-age children with Down syndrome and Fragile X syndrome. In: Roberts, JE.; Chapman, RS.; Warren, S., editors. Speech and language development and intervention in Down syndrome and Fragile X syndrome. Baltimore, MD: Paul H. Brookes; 2008. p. 77-115.

Roberts, JE.; Chapman, R.; Warren, S., editors. Speech and language development and intervention in Down syndrome and Fragile X syndrome. Baltimore: Paul H. Brookes; 2008.

Roberts JE, Martin GE, Moskowitz L, Harris AA, Foreman J, Nelson L. Discourse skills of boys with Fragile X syndrome in comparison to boys with Down syndrome. Journal of Speech, Language, and Hearing Research. 2007; 50:475–492.

Roberts JE, Price J, Barnes E, Nelson L, Burchinal M, Hennon E, Moskowitz L, Edwards A, Malkin C, Anderson K, Misenheimer J, Hooper SR. Receptive vocabulary, expressive vocabulary, and speech production of boys with Fragile X syndrome in comparison to boys with Down syndrome. American Journal on Mental Retardation. 2007; 112:177–193. [PubMed: 17542655]

Rogers SJ, Wehner DE, Hagerman R. The behavioral phenotype in Fragile X: Symptoms of autism in very young children with Fragile X syndrome, idiopathic autism, and other developmental disorders. Journal of Developmental Behavioral Pediatrics. 2001; 22(6):409–417.

Roid, GH.; Miller, LJ. Leiter International Performance Scale—Revised. Wood Dale, IL: Stoelting; 1997.

Ross DE. Replacing faulty conversational exchanges for children with autism by establishing a functionally equivalent alternative response. Education and Training in Mental Retardation and Developmental Disabilities. 2002; 37(4):343–362.

Scharfenaker, S.; O'Connor, R.; Stackhouse, T.; Braden, ML.; Gray, K. An integrated approach to intervention. In: Hagerman, RJ.; Hagerman, PJ., editors. Fragile X syndrome: Diagnosis, treatment, and research. Baltimore, MD: Johns Hopkins University Press; 2002. p. 363-427.

Sudhalter V, Cohen IL, Silverman W, Wolf-Schein EG. Conversational analyses of males with Fragile X, Down syndrome, and autism: Comparison of the emergence of deviant language. American Journal on Mental Retardation. 1990; 94(4):431–441. [PubMed: 2137003]

Sudhalter V, Scarborough HS, Cohen IL. Syntactic delay and pragmatic deviance in the language of Fragile X males. American Journal of Medical Genetics. 1991; 38(2–3):493–497. [PubMed: 2018092]

Tager-Flusberg H. Designing studies to investigate the relationships between genes, environments, and developmental language disorders. Applied Psycholinguistics. 2005; 26:29–39.

Tager-Flusberg H, Rogers S, Cooper J, Landa R, Lord C, Paul R, Rice M, Stoel-Gammon C, Weatherby A, Yoder P. Defining spoken language benchmarks and selecting measures of expressive language development for young children with autism spectrum disorders. Journal of Speech, Language, and Hearing Research. 2009; 52:643–652.

Thordadottir ET. Language-specific effects of task demands on the manifestation of specific language impairment: A comparison of English and Icelandic. Journal of Speech and Hearing Research. 2008; 51:922–937.

Wilding J, Cornish K, Munir F. Further delineation of the executive deficit in males with Fragile-X syndrome. Neurospsychologia. 2002; 40:1343–1349.

Wolf-Schein EG, Sudhalter V, Cohen IL, Fisch GS, Hanson D, Pfadt AG, Hagerman R, Jenkins E, Brown WT. Speech-language and the Fragile X syndrome: Initial findings. ASHA. 1987; 29(7): 35–38. [PubMed: 2956959]

Table 1

Chronological Age and Leiter Developmental Age of Boys with Fragile X Syndrome Without Autism Spectrum Disorder, Boys with Fragile X Syndrome with Autism Spectrum Disorder, Boys with Down Syndrome, and Typically Developing Boys

	FXS-O (*n* = 30)	FXS-ASD (*n* = 29)	DS (*n* = 27)	TD (*n* = 25)
Chronological age (in years)				
M	11.5	10.6	10.1	5.2
SD	2.3	2.8	2.8	0.8
Range	6.0–15.8	6.4–15.5	6.3–16.0	3.9–6.5
Leiter-R developmental age (in years)				
M	5.5	5.3	5.2	5.2
SD	0.7	0.4	0.9	1.1
Range	4.0–7.7	4.7–6.6	4.1–8.2	3.6–8.2

Note. FXS-O = Fragile X syndrome only; FXS-ASD = Fragile X syndrome with autism spectrum disorder; DS = Down syndrome; TD = typically developing. Sample sizes varied according to language sampling context. Social interaction = 25 boys with FXS-O, 25 boys with FXS-ASD, 25 boys with DS, and 25 TD boys. Narration = 26 boys with FXS-O, 23 boys with FXS-ASD, 25 boys with DS, and 25 TD boys.

Table 2

Means and Standard Deviations for Proportion of Utterances Categorized as Utterance-Level, Topic, and Conversational Device Perseveration by Group and Context

	Social interaction				Narration			
	FXS-O (*n* = 25)	**FXS-ASD (*n* = 25)**	**DS (*n* = 25)**	**TD (*n* = 25)**	**FXS-O (*n* = 26)**	**FXS-ASD (*n* = 23)**	**DS (*n* = 25)**	**TD (*n* = 25)**
	***M* (*SD*)**	***M* (*SD*)**	***M* (*SD*)**	***M* (*SD*)**	***M* (*SD*)**	***M* (*SD*)**	***M* (*SD*)**	***M* (*SD*)**
Utterance-level perseveration	.10 (.07)	.13 (.09)	.07 (.05)	.10 (.05)	.11 (.08)	.14 (.12)	.11 (.10)	.21 (.15)
Topic perseveration	.09 (.05)	.14 (.09)	.05 (.05)	.05 (.04)	.03 (.09)	.04 (.08)	.03 (.07)	.04 (.07)
Conversational device perseveration	.04 (.04)	.03 (.03)	.05 (.04)	.05 (.04)	.00 (.01)	.00 (.02)	.00 (.00)	.01 (.02)

Note. FXS-O = Fragile X syndrome only; FXS-ASD = Fragile X syndrome with autism spectrum disorder; DS = Down syndrome; TD = typically developing.

Table 3

Adjusted Means (Adjusted for Leiter-R Mental Age), Standard Errors, and Between-Group Differences for the Types of Perseveration in Social Interaction

Outcome	FXS-O (*n* = 25)	FXS-ASD (*n* = 25)	DS (*n* = 25)	TD (*n* = 25)
Utterance-level perseveration	0.10(.01)[a,b]	0.13(.01)[a]	0.07(.01)[b]	0.10(.01)[a,b]
Topic perseveration	0.09(.01)[a]	0.14(.01)[b]	0.05(.01)[a]	0.05(.01)[a]
Conversational device perseveration	0.04(.01)[a]	0.04(.01)[a]	0.05(.01)[a]	0.05(.01)[a]

Note. FXS-O = Fragile X syndrome only; FXS-ASD = Fragile X syndrome with autism spectrum disorder; DS = Down syndrome; TD = typically developing. Different superscripts within a row indicate significant differences. If groups share the same letter, differences were not significant.

Table 4

Adjusted Means (Adjusted for Leiter-R Mental Age) and Between-Group Differences for the Types of Perseveration in Narration

Outcome	FXS-O (*n* = 26)	FXS-ASD (*n* = 23)	DS (*n* = 25)	TD (*n* = 25)
Utterance-level perseveration	0.11(.01)[a]	0.14(.01)[a]	0.11(.01)[a]	0.21(.01)[b]
Topic perseveration	0.03(.01)[a]	0.04(.01)[a]	0.03(.01)[a]	0.04(.01)[a]
Conversational device perseveration	0.00(.01)[a]	0.00(.01)[a]	0.00(.01)[a]	0.01(.01)[a]

Note. FXS-O = Fragile X syndrome only; FXS-ASD = Fragile X syndrome with autism spectrum disorder; DS = Down syndrome; TD = typically developing. Different superscripts within a row indicate significant differences. If groups share the same letter, differences were not significant.