VERBAL PERSEVERATION IN BOYS WITH FRAGILE X SYNDROME WITH AND WITHOUT AUTISM COMPARED TO BOYS WITH DOWN SYNDROME

Gary Everett Martin

A dissertation submitted to the faculty of the University of North Carolina at Chapel Hill in partial fulfillment of the requirements for the degree of Doctor of Philosophy in the School of Medicine, Department of Allied Health Sciences, Division of Speech and Hearing Sciences

Chapel Hill
2008

Approved by:

Joanne Roberts, Ph.D.

Elizabeth Crais, Ph.D.

Nancy Helm-Estabrooks, Sc.D.

Stephen Hooper, Ph.D.

Linda Watson, Ed.D.

ABSTRACT

GARY EVERETT MARTIN: Verbal Perseveration in Boys with Fragile X Syndrome with and without Autism Compared to Boys with Down Syndrome
(Under the direction of Joanne Roberts)

This study compared boys with FXS with (n = 29) and without (n = 30) autism spectrum disorder (ASD) to boys with Down syndrome (DS; n = 29) and younger typically developing (TD) boys (n = 29) on measures of verbal perseveration (i.e., excessive self-repetition) during a play-based interaction, picture-supported story retelling, and verbal word fluency test (sample sizes varied according to context). Types of perseveration examined in interaction and narration included utterance-level (successive repetition of a word, phrase, or utterance) and ideational (excessive repetition of an idea, topic, or theme). Types of perseveration examined during the verbal fluency test included stuck-in-set (inappropriate continuation of a category), recurrent (inappropriate repetition of a previous response after an intervening response or stimulus), and continuous (inappropriate prolongation of a behavior without interruption). Conversational device repetition (repetition of rote sayings and phrases) in interaction and narration, and loss-of-set responses (responses inappropriate for a given category) during the verbal fluency test were also examined.

The results revealed that boys with FXS, regardless of autism status, produced significantly more ideational perseveration during play-based interaction than did boys with DS and younger TD boys after controlling for nonverbal mental age. Further, the boys with FXS with ASD displayed significantly more ideational perseveration during interaction than all other groups, and more utterance-level perseveration than boys with DS. In narration, the

boys with FXS without ASD and boys with DS produced significantly less utterance-level perseveration than did the TD boys. Further, in interaction as compared to narration, boys with FXS (with and without ASD) displayed significantly more ideational perseveration; boys with FXS with ASD, boys with DS, and TD boys produced significantly more conversational device repetition; and TD boys displayed significantly less utterance-level perseveration. On the verbal fluency test, boys with FXS (with and without autism) and boys with DS produced significantly more loss-of-set responses than TD boys. These findings suggest that boys with FXS display more perseveration than would be expected based on nonverbal mental age or intellectual disability in general, and that autism status as well as language sampling context affect perseveration in boys with FXS.

To my mother, Cheryl Martin, and my grandmother, Eva Colandrea, for their unwavering love, support, and prayers

ACKNOWLEDGEMENTS

The completion of this dissertation would not be possible if not for the efforts of many individuals. First, I acknowledge my mentor, Dr. Joanne Roberts, a tireless researcher, for her direction and time. Second, I greatly appreciate the assistance of colleagues of the Carolina Communication Project and the larger FPG community, especially Kristin Cooley, Anne Taylor, Jan Misenheimer, and John Sideris. Third, I especially thank the children and families who have taken part in the Carolina Communication Project. Fourth, I extend my gratitude to the members of my dissertation advisory committee, Drs. Nancy Helm-Estabrooks, Linda Watson, Betsy Crais, and Steve Hooper, for sharing their expertise and for making this a better dissertation. Fifth, I thank my research assistants, Jackie Assal and Jenna Mory, for their great work. Sixth, I acknowledge Lauren Moskowitz, Mike Underwood, Alan Roark, Chris Kocher, and Lindsay Moskowitz for their friendship. Lastly, I express my thanks to my family, particularly my brother, Dr. Scott Martin, who told me several years ago that the path to a Ph.D. was a marathon, not a sprint, and who has encouraged me throughout this process. In addition, I thank my parents, Cheryl Martin and Russell Martin, for their belief in me, and my grandparents, Russell Martin Sr. and Grace Martin, for their love and support early in my life. I especially thank my departed grandfather, John A. Colandrea, who has continued to support me over these last few years through my grandmother, Eva Colandrea, whom I also appreciate greatly.

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CHAPTER 1

INTRODUCTION

Fragile X syndrome (FXS) is the most common inherited cause of intellectual disability (ID; Dykens, Hodapp, & Finucane, 2000; Hagerman & Hagerman, 2002), affecting 1 in 4,000 males and 1 in 8,000 females (Crawford, Acuna, & Sherman, 2001; Turner, Webb, Wake, & Robinson, 1996). In FXS, an increase in Cytosine-Guanine-Guanine (CGG) repeats in the Fragile X Mental Retardation-1 (FMR1) gene on the X chromosome causes the gene to become methylated (shut down). Consequently, Fragile X Mental Retardation Protein (FMRP) is either reduced in quantity or not produced at all (Oostra & Willemsen, 2003). As FMRP is believed to be essential for normal brain development, this deficiency in FMRP is thought to account for the numerous physical and behavioral characteristics associated with the syndrome (Devys, Lutz, Rouyer, Bellocq, & Mandel, 1993; Jin & Warren, 2003). In females, the unaffected X chromosome likely moderates the effects of the affected X chromosome (Hagerman, 2002). Thus, males with FXS are more severely affected than females (Hagerman & Hagerman, 2002; Loesch et al., 2003; Reiss & Dant, 2003).

Verbal perseveration, or the excessive self-repetition of a spoken word, phrase, sentence, or topic, is a frequently reported behavioral characteristic of males with FXS (Abbeduto & Hagerman, 1997; Hanson, Jackson, & Hagerman, 1986; Madison, George, & Moeschler, 1986; Murphy & Abbeduto, 2007; Roberts, Martin, et al., 2007; Sudhalter, Cohen, Silverman, & Wolf-Schein, 1990; Wolf-Schein et al., 1987). In fact, many

researchers contend that verbal perseveration is a defining behavioral characteristic of FXS (Abbeduto, Brady, & Kover, 2007; Abbeduto & Hagerman, 1997; Bennetto & Pennington, 1996; Roberts, Chapman, Martin, & Moskowitz, 2008). This argument is reflective of a major goal in developmental disabilities research, which is to distinguish syndromes on the basis of language phenotypes (Rice, Warren, & Betz, 2005; Tager-Flusberg, 2005).

From a clinical perspective, the problem of verbal perseveration is significant for several reasons. Frequent perseveration may affect profoundly conversational skills, present a major obstacle to social interaction, and be frustrating for caregivers and other communication partners. Further, perseveration can affect test performance (e.g., scoring lower on a word list generation test due to repetitive responses) and thereby skew results by "blocking" underlying skills (Helm-Estabrooks & Albert, 2004).

The present study examined verbal perseveration in boys with FXS with and without autism spectrum disorder (ASD), boys with Down syndrome (DS) and younger typically developing (TD) boys of similar nonverbal mental age (MA) during play-based interaction, picture-supported story retelling, and a verbal word fluency (word list generation) test. In this chapter, I review the literature on verbal perseveration in FXS. I begin, however, by providing a brief overview of the behavioral phenotype of FXS apart from verbal perseveration, focusing on language as well as related cognitive and psychopathological characteristics. I also briefly describe the literature on verbal perseveration in adult neurological disorders, because, in many ways, this body of literature can serve as a model for fragile X researchers. The chapter concludes with a rationale for the current study. The term *perseveration* is used interchangeably with the more precise descriptor *verbal*

perseveration in the remainder of this chapter, although it is noted that perseveration is a behavior that can occur in multiple modalities.

Behavioral Phenotype of FXS

The majority of males with FXS have moderate or severe ID (Abbeduto & Chapman, 2005; Bennetto & Pennington, 1996) with specific impairments in working memory, sustained attention, and attention-switching (Hooper, Hatton, Baranek, Roberts, & Bailey, 2000; Munir, Cornish, & Wilding, 2000; Wilding, Cornish, & Munir, 2002). Most females with FXS have an IQ above 70, although impairments in executive function and attention have been reported (Bennetto, Pennington, Taylor, & Hagerman, 2001; Cornish, Sudhalter, & Turk, 2004). Moreover, about 70% of males with FXS meet diagnostic criteria for Attention Deficit Hyperactivity Disorder (ADHD; Baumgardner, Reiss, Freund, & Abrams, 1995; Borghgraef, Fryns, Dielkens, Dyck, & Van den Berghe, 1987; Bregman, Leckman, & Ort, 1988), and psychopathology in the form of social anxiety is frequently observed in both males and females (Bregman et al., 1988; Hagerman, 2002; Hessl et al., 2001; Mazzocco, Baumgardner, Freund, & Reiss, 1998).

Researchers generally agree that about 25% of males with FXS also have autism (Bailey et al., 1998; Dykens & Volkmer, 1997; Hagerman, 2002). Prevalence estimates closer to 50% have been reported, however, in studies that combined pervasive developmental disorder-not otherwise specified (PDD-NOS) with the more "classic" autistic disorder (Demark, Feldman, & Holden, 2003; Kaufmann et al., 2004; Philofsky, Hepburn, Hayes, Hagerman, & Rogers, 2004). Core features of autistic disorder according to the *Diagnostic and Statistical Manual of Mental Disorders-4th edition (DSM-IV-TR)* include qualitative impairments in social interaction and communication, as well as repetitive,

stereotyped, and restricted patterns of behavior, activities, and interests (American Psychiatric Association [APA], 2000). The DSM-IV diagnosis of autistic disorder is reserved for children who display a certain number of symptoms within each category of impairment, whereas the diagnosis of PDD-NOS is given for children who display similar behaviors at a "subthreshold" level.

The behavioral phenotype of FXS also includes notable speech and language impairments, although studies in this area of inquiry have focused almost exclusively on males. Despite considerable variability among individuals, most males appear to have delays relative to nonverbal MA in expressive syntax (Paul, Cohen, Breg, Watson, & Herman, 1984; Paul et al., 1987; Price et al., 2008; Roberts, Hennon, et al., 2007), as well as poor speech intelligibility (Madison et al., 1986; Paul et al., 1987; Barnes et al., in review). In males, receptive and expressive vocabulary are reported to be relative strengths (Abbeduto et al., 2003; Roberts, Price et al., 2007) or delayed relative to MA (Price, Roberts, Vandergrift, & Martin, 2007; Roberts, Hennon, et al., 2007; Sudhalter, Scarborough, & Cohen, 1991) depending on the sampling method. Findings for receptive syntax are also mixed. Several studies have found receptive syntax in males with FXS to be commensurate with expectations for nonverbal MA (Abbeduto et al., 2003; Paul et al., 1984; Paul et al., 1987;), whereas one recent study (Price et al., 2007) reported that boys with FXS scored lower than TD controls on a measure of receptive syntax ability.

Pragmatic aspects of language may be particularly impaired in individuals with FXS, even when compared with individuals with other forms of ID. The conversational discourse of males with FXS has been characterized by gaze aversion (Cohen et al., 1988; Cohen, Vietze, Sudhalter, Jenkins, & Brown, 1991), noncontingent (off-topic or tangential) language

(Roberts, Martin et al., 2007; Sudhalter & Belser, 2001; Wolf-Schein et al., 1987), and perseveration (Roberts, Martin, et al., 2007; Sudhalter et al., 1990; Wolf-Schein et al., 1987). Females with FXS are reportedly shy and may have difficulty initiating conversational interactions (Hagerman, Hills, Scharfenaker, & Lewis, 1999; Lesniak-Karpiak, Mazzocco, & Ross, 2003; Mazzocco, Kates, Baumgardner, Freund, & Reiss, 1997).

Several studies have investigated the effects of autism status on language and communication in individuals with FXS. Children with both FXS and autism have been found to perform more poorly on broad measures of expressive and receptive language (Bailey, Hatton, Skinner, & Mesibov, 2001; Philofsky et al., 2004; Rogers, Wehner, & Hagerman, 2001) and to produce more noncontingent language (Roberts, Martin, et al., 2007) than do children with FXS without autism. However, autism status in FXS does not appear to specifically affect expressive vocabulary (Roberts, Price et al., 2007) or receptive and expressive syntax (Price et al., 2008; Price et al., 2007). Findings for receptive vocabulary, however, have been mixed (Lewis et al., 2006; Price et al., 2007). The effects of autism status on verbal perseveration are discussed within the next section.

Verbal Perseveration in FXS

Non-Comparative Research

A few early studies reported perseveration to be common in males with FXS but did not include comparison samples of TD individuals or individuals with other forms of ID. Specifically, Fryns, Jacobs, Kleczkowska, and Van den Berghe (1984) reviewed the admission records of 21 institutionalized males with FXS, and discovered reports of perseveration in the records of 10 of these males. Madison and colleagues (1986) directly observed topic perseveration in the conversations of 4 adult males and one boy with FXS, all

of whom were from a single family. Similarly, all 10 boys studied by Hanson et al. (1986) were reported to produce perseveration during interactions with an examiner.

Only 3 of 15 young males, however, were reported to produce perseveration in an investigation by Prouty et al. (1988). Prouty and colleagues defined perseveration as "repetitive phrases which were interspersed with other phrases" (p. 132); 2 additional males demonstrated word repetitions. Neither Fryns et al. (1984) nor Hanson et al. (1986) provided operational definitions of perseveration; thus, it is not known which specific behavior or behaviors were measured in these two studies.

Comparative Research

Two recent studies have reported perseveration to be more common in boys with FXS than in younger TD boys. Levy, Gottesman, Borochowitz, Frydman, and Sagi (2006) found that boys with FXS produced more perseveration (defined as consecutive production of the same utterance three or more times) than MLU-matched TD boys during conversation with an examiner. Roberts, Martin, et al. (2007) also studied perseveration in examiner-child interactions and reported that boys with FXS produced more perseveration than TD boys, even after controlling for nonverbal MA. Roberts, Martin, et al. coded perseveration "when the child repeated certain words, phrases, or sentences (or very similar phrases or sentences) within or across turns, or when the child repeatedly spoke on a single topic or theme of topics" (p. 481). Similar to the criteria used by Levy et al. (2006), the coding scheme of Roberts, Martin, et al. required at least two repetitions for a behavior to be considered perseveration.

A few investigations have found that males with FXS produced more perseveration than either males with DS (Roberts, Martin et al., 2007; Wolf-Schein et al., 1987) or autism

(Sudhalter et al., 1990), whereas one study (Paul et al., 1987) did not find that institutionalized males with FXS differed from males with autism or nonspecific forms of ID on a measure of topic perseveration. Specifically, Wolf-Schein et al. (1987) reported that boys and adult males with FXS produced more perseveration (definition not provided) than males with DS during conversations with a familiar and unfamiliar person. Roberts, Martin, et al. (2007) reported similar findings for younger samples of boys with FXS and DS.

Sudhalter and colleagues (1990) were the first researchers studying perseveration in individuals with FXS to propose a taxonomy that denoted specific types of perseveration. In an investigation of "deviant repetitive language" (comprised of perseveration, echolalia, jargoning, and "affirming by repetition") in boys and adult males with FXS, DS, and autism, Sudhalter et al. (1990) 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 implementation of this taxonomy, different types of perseveration were collapsed for analysis (i.e., 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") in this and other studies by Sudhalter and colleagues (Belser & Sudhalter, 1995; Sudhalter, Scarborough, & Cohen, 1991).

Nevertheless, Sudhalter et al. (1990) found that males with FXS produced more deviant repetitive language than males with DS during interactions with both a familiar person and an unfamiliar person (the examiner). Moreover, a post-hoc analysis revealed that

males with FXS produced specifically more perseveration than males with autism. Unfortunately, Sudhalter et al. (1990) did not compare males with FXS and DS specifically on the dimension of perseveration.

Two studies have examined behaviors that are consistent with definitions of perseveration, although researchers did not use the term *perseveration* to describe these behaviors. In a study by Baumgardner et al. (1995), parents and teachers rated boys with FXS higher than boys with other forms of ID on a checklist item related to using repetitive words and phrases. Ferrier, Bashir, Meryash, Johnston, and Wolff (1991) reported that children and adults with FXS produced more partial repetitions of their own previous utterances than individuals with DS and autism.

Effects of Autism Status

Because individuals with autism who do not have FXS have also been reported to display verbal perseveration (Baker, 2000; Koegel & Frea, 1993; Ross, 2002), individuals with both FXS and autism may produce more perseveration than those with FXS only. However, the effects of autism status on perseveration in FXS have been largely unexplored. Several studies (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 FXS, whereas other studies (Ferrier et al., 1991; Levy et al., 2006; Murphy & Abbeduto, 2007; Sudhalter et al., 1990, 1991) excluded individuals with FXS on the basis of their autism status.

Specifically, individuals with FXS were excluded from previous studies for meeting DSM-IV (APA, 2000) diagnostic criteria for autistic disorder (Murphy & Abbeduto, 2007), DSM-III (APA, 1980) criteria for infantile autism or childhood onset pervasive

developmental disorder (Sudhalter et al., 1990), or DSM criteria for "autism" (Ferrier et al., 1991; Levy et al., 2006; Sudhalter et al., 1991). "Autism," however, has never been a DSM diagnostic category, although a number of researchers, clinicians, and members of the general public use "autism" as a synonym for "autistic disorder" (whereas others use "autism" as the umbrella term for all autism spectrum disorders). Thus, it is likely that individuals who met criteria for the "subthreshold" DSM-IV diagnosis of PDD-NOS remained in all of these previous investigations.

Three studies (Baumgardner et al., 1995; Paul et al., 1987; Roberts, Martin et al., 2007) reported inclusion of individuals with FXS and co-occurring autism in addition to those with FXS only. However, Baumgardner et al. and Paul et al. did not investigate the effects of autism status on perseveration, and data for individual participants were not reported. In the only published study to date that investigated the effects of autism status on perseveration in FXS (Roberts, Martin et al., 2007), the frequency of perseveration during conversation did not significantly differentiate boys with FXS with ASD (classified as "autism" or "autism spectrum" on the *Autism Diagnostic Observation Schedule [ADOS]*, Lord, Rutter, DiLavore, & Risi, 2001) and boys with FXS without ASD. However, a moderate effect size indicated to the researchers that actual group differences may have gone undetected. The direction of the effect size would suggest that perseveration is more common in boys with both FXS and ASD.

Effects of Language Sampling Context

Except for one recent study, perseveration in FXS has been examined in conversation only. Murphy and Abbeduto (2007) reported that *topic repetition* occurred more often in conversation (10-minute interaction with an examiner wherein they discussed predetermined

topics such as school, pets, and hobbies) than in narration (storytelling from a wordless picture book) for adolescent males and females with FXS. 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.

Murphy and Abbeduto (2007) coded two other categories of repetition for which there were no context effects: *utterance-level repetition* and *conversational device repetition*. The definition of utterance-level repetition incorporated phrasal and sentential perseveration as described by Sudhalter et al. (1990), but it also included successive word repetitions. Conversational device coding captured repetition of rote phrases and expressions (e.g., "right on" or "I don't know") that were considered to be "fillers" or "placeholders" in that they performed the mechanics of conversation without necessarily adding semantic information.

Murphy and Abbeduto (2007) proposed two explanations for their finding that topic repetition happened more frequently in conversation than in narration. First, the interpersonal demands of conversation (e.g., eye contact) may have caused the individual with FXS to become "hyperaroused." Hyperarousal, or the physiological state of increased and unregulated nervous system excitation (Sudhalter et al., 1990), is commonly regarded as a possible underlying mechanism for perseveration in individuals with FXS (see next section). The second explanation presented by Murphy and Abbeduto presumes a relationship between perseveration and executive functions. The added structure of the narrative format, they reasoned, may have lessened the effects of impairments in executive functioning. Clearly, context effects were thought to reflect the mechanism or mechanisms that account for perseveration. A review of all previously proposed causal theories for perseveration in FXS is provided next.

Explanatory Theories

A number of mechanisms have been theorized to explain the verbal perseveration exhibited by individuals with FXS, although most theories have attempted to account for all types of perseveration with a single mechanism. The majority of arguments have come from Sudhalter and colleagues (Belser & Sudhalter, 1995; Cornish et al., 2004; Sudhalter et al., 1990, 1991). According to the word-retrieval account (Sudhalter et al., 1990), the individual with FXS produces perseveration while searching for the correct word to say, or to provide him or herself with the time necessary to process the message of a communication partner. Sudhalter et al. (1991) later hypothesized that perseveration may be related to deficits in expressive syntax. The individual with FXS, they argued, may repeat unmastered syntactic structures as a means of practicing such structures. Syntactic measures, however, were not found to correlate significantly with perseveration in the study by Sudhalter et al. (1991), and the word-retrieval hypothesis has not been tested.

The hyperarousal theory (Belser & Sudhalter, 1995; Cornish et al., 2004; Sudhalter et al., 1990) is the most commonly cited explanatory theory for perseveration in FXS. According to this account, the individual with FXS becomes hyperaroused and anxious during social interactions, particularly because the social gaze of a communication partner makes him or her "uncomfortable," and produces verbal perseveration in response. Belser and Sudhalter (1995) specified that the discomfort associated with social gaze *is* anxiety, which directly reflects a heightened state of arousal. The precise mechanism for how hyperarousal may cause perseveration was also presented by Belser and Sudhalter. They proposed that a "sentence template" normally precedes the production of every sentence, and

that this template does not decay quickly enough in the hyperaroused individual. As a result, characteristics of the sentence (words, phrases, and topics) are repeated in the next sentence.

Cornish and colleagues (2004) expanded upon the hyperarousal theory. They proposed that hyperarousal, through the activation of extraneous neuronal connections, may interfere with a person's ability to inhibit or switch attention. This deficit in inhibitory control, they reasoned, may act to free impulsive tendencies of the individual with FXS to talk continually about his or her favorite topic (which may also be well-rehearsed and therefore less anxiety-producing than unfamiliar topics). Abbeduto and Hagerman (1997) also suggested that perseveration may result from problems inhibiting highly salient or prior responses, although they implicated frontal lobe abnormalities independent of hyperarousal.

The hyperarousal hypothesis has been tested only once. Belser and Sudhalter (1995) found that the skin conductance levels (a measure of autonomic arousal) of 2 males with FXS were greater when an experimenter faced them in conversation than when the examiner looked away, whereas no effects of eye gaze were found for one male with DS and another male with ADHD. Moreover, males with FXS produced more "deviant language" (perseveration and tangential language) in the eye contact condition compared with the non-eye contact condition, whereas the other two participants did not produce any deviant language in either condition. The taxonomy of perseverative behaviors used in this study was largely consistent with the taxonomy applied previously by Sudhalter and colleagues (Sudhalter et al., 1990, 1991), except that Belser and Sudhalter also coded perseveration at the word level and expanded upon the definition of topic perseveration to characterize "the inability to be diverted from the topic of choice" (p. 276).

Recently, explanatory theories of perseveration in FXS have begun to recognize that different types of perseveration may be related to different underlying problems. Citing the work of Miller (1991), Murphy and Abbeduto (2007) suggested that utterance-level repetition may reflect false starts or word-finding difficulties. Moreover, they proposed that topic repetition may be related to hyperarousal or executive dysfunction, and that conversational device repetition may represent an individual's way of staying engaged in discourse in the face of working memory deficits.

Summary and Limitations of Previous Research

In summary, verbal perseveration in individuals with FXS has been a subject of published studies for over 20 years. In early investigations without comparison samples, perseveration was reported to be a common behavioral characteristic of males with FXS. Since that time, a small number of studies have found that males with FXS produce more perseveration during conversation than males with DS, males with autism, and younger TD boys. Within individuals with FXS, findings from one study suggest that topic perseveration is more common in conversation than in picture-supported storytelling. Researchers have proposed several mechanisms to account for perseveration in persons with FXS, although the most widely cited explanatory theory presumes a primary role of hyperarousal.

Previous research into perseveration in individuals with FXS has been limited in several respects. A few investigators (Fryns et al., 1984; Hanson et al., 1986; Wolf-Schein et al., 1987) did not provide a definition of perseveration. Other researchers have defined and even delineated different types of perseveration, but either did not analyze their data according to type (Belser & Sudhalter, 1995; Sudhalter et al., 1990) or did not include a non-FXS comparison sample (Murphy & Abbeduto, 2007). Therefore, no studies have compared

individuals with FXS to individuals with autism or other forms of ID on different types of perseveration. Further, the consequence of autism status on perseveration in FXS has been largely unexplored. Finally, except for a single study of perseveration in both narration and conversation (Murphy & Abbeduto, 2007), perseveration in individuals with FXS has been examined in conversation only; in fact, perseveration has never been studied in a context other than connected speech (conversation and narration).

Perseveration in Adult Neurological Disorders

The study of perseveration in adults with neurological disorders has a long and rich history, and studies in this area of inquiry can serve as a model for fragile X researchers. Interestingly, although this body of literature has been in existence for more than 100 years, no previous studies of perseveration in FXS have cited the work of researchers in this field. Neisser (1895) introduced the term *perseveration* to indicate the inappropriate recurrence or continuation of a behavior after a switch in task requirements. In a more recent publication, Lezak, Howieson, Loring, Hannay, and Fischer (2004) defined perseveration as the "repetitive prolongation or continuation of an act or activity sequence, or repetition of the same or a similar response to various questions, tasks, or situations" (p. 84).

Perseveration has been examined in persons with neurological disorders such as aphasia (Helm-Estabrooks, Ramage, Bayles, & Cruz, 1998; Sandson & Albert, 1987; Shindler, Caplan, & Hier, 1984), Alzheimer's disease (Bayles, Tomoeda, Kaszniak, Stern, & Eagans, 1985; Bayles, Tomoeda, McKnight, Helm-Estabrooks, & Hawley, 2004; Shindler et al., 1984), Parkinson's disease (Bayles et al., 1985; Ebersbach, Hättig, Schelosky, Wissel, & Poewe, 1994; Sandson & Albert, 1987; Stoffers, Berendse, Deijen, & Wolters, 2001), and closed-head injury (Hotz & Helm-Estabrooks, 1995a; Lombardi et al., 1999). See Hotz and

Helm-Estabrooks (1995b) for a comprehensive review of the literature on perseveration in adult neurological disorders.

Types of Perseveration

Researchers of perseveration in adults with neurological disorders agree that "perseveration is not a unitary phenomenon but a collection of different forms of behaviour" (Hotz & Helm-Estabrooks, 1995b, p. 152). Sandson and Albert (1984) proposed a taxonomy that included the following categories of perseveration: (a) *stuck-in-set*, the inappropriate continuation of a framework or category; (b) *recurrent*, the inappropriate repetition of a previous response to a later stimulus or after an intervening response; and (c) *continuous*, the inappropriate prolongation of a behavior without interruption. Soon after, Bayles et al. (1985) used the term *ideational perseveration*, modifying a description introduced by Freeman and Gathercole (1966), to indicate the repetition of an idea during an object description task (e.g., describing a nail as "very small" after describing it as "quite small," or describing a marble as belonging to a dress after giving a similar description for a button). These types of perseveration have been examined on various structured tasks, including verbal fluency (word list generation) tests, concept definition tests, and tasks requiring individuals to produce repeated graphomotor sequences.

Proposed Neurobehavioral Mechanisms for Perseveration

Three neurobehavioral mechanisms have been argued most often to account for perseveration in persons with neurological disorders (Hotz & Helm-Estabrooks, 1995b). These mechanisms include (a) impaired attention, the failure to disengage one's attention from a stimulus (Sandson & Albert, 1987); (b) impaired memory, the failure to inhibit a response that remains active in short-term memory while searching for a target response in

long-term memory (Hudson, 1968; Sandson & Albert, 1987; Shindler et al., 1984); and (c) disordered motor patterns, the inertia of movement (Luria, 1965).

Several researchers have attempted to relate the different types of perseveration proposed by Sandson and Albert (1984) to specific neurobehavioral mechanisms. Stuck-in-set perseveration has been argued to reflect impairments in executive function (Possin et al., 2005; Sandson & Albert, 1984), including problems in switching attention (Christman, Boutsen, & Buckingham, 2004). Similarly, continuous perseveration may be associated with executive dysfunction (Possin et al., 2005) and reflect difficulties in disengaging attention (Sandson & Albert, 1987). Continuous perseveration in some modalities may be due to motor impairment or persistent motor memory traces (Luria, 1965), such as a person adding additional loops to the grapheme 'm'. Finally, a number of researchers contend that recurrent perseveration results from the reactivation of memory traces and a person's inability to inhibit a previous response that remains active (Hudson, 1968; Pietro & Rigrodsky, 1986; Sandson & Albert, 1987; Shindler et al., 1984).

Loss-of-Set Behavior

In addition to problems in switching cognitive set (stuck-in-set perseveration), persons with some neurological disorders may have difficulty maintaining the correct response set (Lezak et al., 2004). A behavior that is possibly related to perseveration, "loss-of-set" is demonstrated by the individual who begins to respond appropriately given a particular cognitive set, and then slips into another category of response. Errors of this type were examined in the present study because individuals with FXS have been reported to produce many noncontingent contributions to conversational discourse (Roberts, Martin et al., 2007; Sudhalter & Belser, 2001; Wolf-Schein et al., 1987). Therefore, I reasoned that

boys with FXS may produce responses on a verbal fluency test that are unrelated or tangential to the intended semantic category.

Current Study

In the present investigation, I examined different types of verbal perseveration in boys with FXS with and without ASD, boys with DS, and younger TD boys during play-based interaction, picture-supported story retelling, and a verbal word fluency test. Types of perseveration measured in interaction and narration included utterance-level perseveration and ideational perseveration, whereas types of perseveration measured during the verbal fluency test included stuck-in-set, recurrent, and continuous perseveration. Two related verbal behaviors were also examined: conversational device repetition in interaction and narration, and loss-of-set responses during the verbal fluency test.

Motivations for the Present Investigation

There were several specific motivations for the current study. First, perseveration has oftentimes been defined as a single behavior in the fragile X literature. However, perseveration can take many forms in connected speech alone, such as the successive repetition of words, phrases, and entire utterances, or the excessive repetition of ideas and topics. Completely lacking are studies comparing boys with FXS, boys with DS, and TD boys on different types of perseveration. Second, the effects of autism status on perseveration in individuals with FXS have been largely unexplored, and boys with FXS with and without ASD have not been compared previously on different types of perseveration. Third, conversation or play-based interaction has served as the language sampling context for all previous investigations of perseveration in FXS except for one. Moreover, perseveration in FXS has not been examined in a context other than connected speech.

Rationale for Comparisons with Younger TD Boys and Boys with DS

Study participants included boys with FXS with and without ASD, boys with DS, and younger TD boys of similar nonverbal MA. The inclusion of younger TD boys of similar nonverbal MA helps to determine whether any or all types of perseveration and related behaviors exhibited by boys with FXS are appropriate for their developmental level. DS, the most common genetic cause of ID (Centers for Disease Control and Prevention, 2006), is an ideal comparison sample because boys with FXS and boys with DS have similar levels of ID and somewhat similar speech and language profiles. Children with DS have impaired speech, vocabulary, and grammar (Chapman, Seung, Schwartz, & Kay-Raining Bird, 1998; Roberts, Long, et al., 2005; Sigman & Ruskin, 1999), although pragmatics is considered to be a relative strength for individuals with DS (Beeghly, Weiss-Perry, & Cicchetti, 1990; Leifer & Lewis, 1984; Tager-Flusberg & Anderson, 1991). Whereas boys with FXS and boys with DS have been compared previously on overall measures of perseveration (Roberts, Martin et al., 2007; Wolf-Schein et al., 1987), they have not yet been compared on specific types of perseveration. Thus, the comparison sample of boys with DS helps to determine whether certain or all types of perseveration are specific to FXS, or whether perseveration can be attributed more generally to the presence of ID.

Rationale for Comparisons Based on Autism Status

Autism status could affect perseveration in boys with FXS. Individuals with autism without FXS have been reported to produce verbal perseveration (Baker, 2000; Koegel & Frea, 1993; Ross, 2002), and so it is possible that boys with both FXS and ASD produce more perseveration than those with FXS only. Moreover, researchers who have attempted to exclude individuals with FXS on the basis of autism status (Ferrier et al., 1991; Levy et al.,

2006; Murphy & Abbeduto, 2007; Sudhalter et al., 1990, 1991) likely only succeeded in excluding those with an autism profile consistent with a DSM-IV (APA, 2000) diagnosis of autistic disorder. However, nearly 50% of individuals with FXS may qualify for a diagnosis of either autistic disorder or PDD-NOS (Demark et al., 2003; Kaufmann et al., 2004; Philofsky et al., 2004). Therefore, it is likely that a number of the participants with FXS included in these previous studies were on the autism spectrum. In the present study, the boys in the "FXS Only" sample were not categorized as having either autism or autism spectrum on the *ADOS* (Lord et al., 2001); either of these categorizations placed boys with FXS in the "FXS with ASD" group.

Rationale for Play-Based Interaction and Narration Coding Scheme

A detailed description of all measures will be presented in the next chapter. However, inconsistencies in previous approaches to the study of perseveration in FXS necessitate some discussion of the present coding scheme prior to Chapter 2. In the current study, types of self-repetitive language examined in play-based interaction and narration were utterance-level perseveration, ideational perseveration, and conversational device repetition. This taxonomy is most similar to the one described by Murphy and Abbeduto (2007).

Utterance-level perseveration. The repetition involved in perseveration is by definition "excessive" (Abbeduto & Hagerman, 1997; Abbeduto, Brady, & Kover, 2007). The fact that researchers (except for Murphy & Abbeduto, 2007) used the term *perseveration* to describe successive repetitions of words, phrases, and entire utterances indicates that these investigators considered such repetitions to be excessive. However, Sudhalter and colleagues (Belser & Sudhalter, 1995; Sudhalter et al., 1990, 1991) coded perseveration contingent upon just one repetition (i.e., two or more successive productions of the same linguistic unit),

whereas Levy (2006) and Roberts, Martin, et al. (2007) required two or more repetitions (i.e., three or more successive productions of the same linguistic unit). In fact, Murphy and Abbeduto (2007) did not apply the label *perseveration* to describe repetitions of this type. Instead, they coded utterance-level *repetition* given one or more successive repetitions of a word, phrase, or entire utterance. Because of this discrepancy in coding criteria, two studies of utterance-level perseveration could potentially report very different findings.

In the present investigation, I defined utterance-level perseveration as one or more successive repetitions of a word, phrase, or entire utterance. I considered the close proximity of even the first repetition to make it excessive and therefore perseverative. In fact, some support of this criterion comes from the adult neurological literature on perseveration. Bayles et al. (1985) provided the examples "that's-that's" and "it was-it was" for continuous verbal perseveration in their study of adults with neurological disorders. Nevertheless, I applied a subcode for instances of two or more repetitions.

Ideational perseveration. The category of ideational perseveration was utilized to capture the excessive repetition of topics, themes, and ideas. The naming of this category should not indicate that I examined a behavior different from the behavior identified as "topic perseveration" in previous investigations. In fact, Murphy and Abbeduto (2007) described topic repetition as the repetition of topics, themes, or ideas. I regarded "ideational" to be the more inclusive – and, thus, the more accurate – descriptor.

In contrast to the case for utterance-level perseveration, several previous studies have defined topic perseveration with words and criteria that clearly indicate the "excessive" nature of such repetitions. For example, researchers have described repetitions of this type as "incessant" and "tangential" (Belser & Sudhalter, 1995; Sudhalter et al., 1990). Even Murphy

and Abbeduto (2007) described topic *repetition* as the repetition of topics, themes, or ideas in a way that (a) does not add new information, or (b) is tangential or unrelated to the current focus of discourse.

Clearly, the identification of ideational perseveration during connected speech is challenging. Unlike the procedure for identifying utterance-level perseveration, the repetition of a topic, theme, or idea cannot simply be called perseveration given a certain number of repetitions. Roberts, Martin et al. (2007) required *at least* 2 repetitions for a topic or theme to be considered perseverative. In the current study, ideational perseveration was coded with similar criteria. The decision to require at least 2 repetitions was a conservative one.

Conversational device repetition. Repetition of conversational devices (e.g., "right on" or "I don't know") was not considered perseveration because I did not assess the excessiveness of these repetitions. In fact, Bayles et al. (1985) included a non-perseveration category of *carrier phrases* which included filler words and phrases such as "let's see," "oh," and "well." They considered members of this category to be "devices" that individuals with dementia use to fill pauses during periods of disorganized thinking and uncertainty.

I examined conversational device repetition because such repetitions may have figured into reports of perseveration found in previous studies that did not define perseveration (Fryns et al., 1984; Hanson et al., 1986; Wolf-Schein et al., 1987). These investigations either measured behaviors using a rating scale (Wolf-Schein et al., 1987) or simply described participants as displaying or not displaying perseveration (Fryns et al., 1984; Hanson et al., 1986). It is unlikely that an observer in these previous studies would have even realized that a device was used repetitively if it recurred just once during an

interaction. As such, I coded conversational device repetition only when a particular device was repeated two or more times.

Theoretical Perspective

Studies of language and communication in individuals with FXS have generally presumed a relationship between language and nonlinguistic aspects of the behavioral phenotype such as cognitive ability and psychopathology (Abbeduto & Chapman, 2005). This perspective is consistent with an interactionist theory of language development, which stresses a bidirectional relationship between language and nonlinguistic development as well as the critical role of experience gained through social interaction (Chapman, 2000).

Of course, the problem of perseveration does not fit intuitively into any theoretical perspective of language development. Such theories attempt to understand how children acquire language skills. Perseveration, however, is simply not a language "skill." Perhaps perseveration is a verbal behavior that *affects* language development. For example, a child's ability to maintain a topic of conversation that is introduced by a communication partner would clearly be affected if the child tended to stick to just one or two topics of choice. Moreover, a communication partner might adopt a more directive interaction style with a child who produces perseveration, and this style could affect language learning. Such a scenario is also consistent with an interactionist perspective of language development.

Perseveration in individuals with FXS may also be conceptualized from a capacity and processing perspective, wherein anxiety and task demands influence the production of perseveration by decreasing the attention available for inhibiting previous responses. The attention and information processing capacity model, introduced by Kahneman (1973), assumes a limited amount of attention capacity. Interestingly, according to attentional control

theory (Eysenck, Derakshan, Santos, & Calvo, 2007), anxiety causes attention to be allocated to threat-based stimuli. Thus, according to this account, attention in the anxious individual is not available for other processes for which it is normally allocated, such as inhibiting task-irrelevant responses. In fact, individuals with FXS have been reported to have deficits in attention and inhibition (Munir, Cornish, & Wilding, 2000; Wilding, Cornish, & Munir, 2002), although the effects of anxiety on these impairments have not been explored in individuals with FXS.

Moreover, a limited capacity framework proposes that performance decreases as tasks become more difficult (Alt & Plante, 2006). For example, in play-based interaction as compared with picture-supported story retelling, multiple sources of information must be encoded all at once. These demands could further tax the attention system and prevent it from effectively inhibiting previous responses.

Research Questions and Hypotheses

The current study was designed to address the following research questions:

1. Do boys with FXS, boys with DS, and younger TD boys differ in the production of self-repetitive language (utterance-level perseveration, ideational perseveration, and conversational device repetition) during either play-based interaction or narration?
2. Do boys with FXS, boys with DS, and TD boys differ in the production of stuck-in-set, recurrent, and continuous perseveration, as well as loss-of-set behavior, on a verbal fluency test?
3. For boys with FXS, what are the effects of autism status on verbal perseveration and related verbal behaviors?

4. For all boys, what are the effects of language sampling context on verbal perseveration and related verbal behaviors?
5. For all boys, what are the relationships between perseveration during play-based interaction and narration and perseveration on a verbal fluency test?
6. Do findings for utterance-level perseveration differ when using a coding criterion of one or more repetitions versus a coding criterion of two or more repetitions?

I hypothesized that, after controlling for nonverbal MA, boys with FXS (with and without ASD) would produce more utterance-level and ideational perseveration during play-based interaction than boys with DS and TD boys; boys with FXS-ASD would produce more utterance-level and ideational perseveration during interaction than boys with FXS-O; and boys with FXS (with and without ASD) would produce more loss-of-set errors than boys with DS and TD boys on the verbal fluency test. I hypothesized further that ideational perseveration for boys with FXS, regardless of autism status, and conversational device repetition for all groups would be more common in interaction as compared with narration.

CHAPTER 2

METHOD

Participants

Four groups of boys (boys with FXS without ASD, boys with FXS with ASD, boys with DS, and TD boys) participated in this study. The boys were drawn from a larger longitudinal investigation of the speech and language skills of boys with FXS, boys with DS, and TD boys (Roberts, Hennon, 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. TD boys were recruited from schools, childcare centers, and physicians' offices in North Carolina. As females with FXS generally show less severe impairments than males with FXS, due to females having a second normally functioning X chromosome (Hagerman & Hagerman, 2002; Loesch et al., 2002), only boys were included in this study.

To be enrolled in the larger study, boys with FXS and boys with DS had to be 16 years of age or younger, whereas TD boys were between 2 and 6 years of age. At study entry, all boys were producing at least 40 words expressively and combining at least two words (i.e., MLU > 1.1), and all groups displayed similar distributions of nonverbal MAs on the Brief IQ composite of the *Leiter International Peformance Scale – Revised* (*Leiter-R*; Roid & Miller, 1997). Boys with FXS and DS were excluded when their average hearing threshold was greater than 30 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. TD boys were excluded when their average hearing threshold was greater than 25 dB HL in the better ear. Boys with DS and TD boys were screened for FXS and autism, and excluded accordingly. 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 et al., 2001), as described later. TD boys also could not have a developmental disability or speech and language difficulties. Spoken English was the primary language for all children.

In the current investigation, boys with FXS were divided into two groups according to autism status: (1) FXS with ASD and (2) FXS without ASD (FXS-Only). The group of boys with both FXS and ASD included those boys with FXS who were classified by the *ADOS* (Lord et al., 2001) as having "autism" or "autism spectrum." See the *Assessments* section for a description of the *ADOS*. The following sections, as well as Table 2.1, present background characteristics of participants in the present study.

Fragile X Syndrome without Autism Spectrum Disorder (FXS-O)

Thirty boys with FXS who did not have ASD (FXS-O) participated in the current study. These boys ranged in chronological age from 6.0 to 15.8 years (M = 11.5, SD = 2.3) and in nonverbal 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. Maternal education ranged from 11 to 20 years (M = 14.0, SD = 2.2).

Fragile X Syndrome with Autism Spectrum Disorder (FXS-ASD)

Twenty-nine boys with FXS who also had ASD (FXS-ASD) were participants in the current study. These boys ranged in chronological age from 6.4 to 15.5 years (M = 10.6, SD

Table 2.1

Chronological Age and Leiter Developmental Age of Boys with Fragile X Syndrome without Autism Spectrum Disorder (FXS-O), Boys with Fragile X Syndrome with Autism Spectrum Disorder (FXS-ASD), Boys with Down Syndrome (DS), and Typically Developing (TD) Boys

	FXS-O (N=30)	FXS-ASD (N=29)	DS (N=29)	TD (N=29)
Chronological age (in years)				
M	11.5	10.6	10.0	5.1
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. Samples varied somewhat according to language sampling context. Conversation = 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. Verbal fluency test = 19 boys with FXS-O, 15 boys with FXS-ASD, 15 boys with DS, and 18 TD boys.

= 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. Maternal education ranged from 11 to 20 years (*M* = 14.5, *SD* = 2.3).

Down Syndrome (DS)

Twenty-nine boys with DS participated in the present study. These boys ranged in chronological age from 6.3 to 16.0 years (*M* = 10.0, *SD* = 2.8) and in nonverbal MA from 4.1 to 8.2 years (*M* = 5.2, *SD* = 0.9). Ninety-three percent of the boys were Caucasian, and 7% were African American. Parents reported that trisomy 21 was the source of DS for all boys. Maternal education ranged from 12 to 20 years (*M* = 15.8, *SD* = 2.3).

Typically Developing (TD) Boys

Twenty-nine TD boys were study participants. These boys ranged in chronological age from 3.9 to 6.5 years (*M* = 5.1, *SD* = 0.8), and in nonverbal MA from 3.6 to 8.2 years (*M* = 5.2, *SD* = 1.1). Sixty-six percent of the boys were Caucasian, 24% were African American, and 10% were of a different ethnicity. Maternal education ranged from 12 to 20 years (*M* = 16.5, *SD* = 2.3).

Assessments

Each boy was assessed at his school, in his home, or at a university research center. Testing occurred in a quiet area and lasted for approximately 6 hours (frequently over two days) 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* (Roid & Miller, 1997) 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 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 FXS

Autism classification of the boys with FXS 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." Examiners, who completed *ADOS* training and were reliable with an *ADOS* trainer, scored videotapes of *ADOS* interactions. Reliability computed on 16% of the boys was .93 on diagnosis (range of .83-.96) and .89 for individual items (range of .83-.96). Eight of the boys with FXS 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 "spectrum."

Language Sampling and Transcription Procedures

Play-Based Interaction

Play-based interactions that occurred during administration of the *ADOS* (Lord et al., 2001) were examined in this study. During the *ADOS*, the examiner elicited language using developmentally appropriate games and toys. The *ADOS* includes a variety of structured and semi-structured activities, such as a block activity, free-play, bubble and balloon blowing, a demonstration task, storybook reading, and a pretend birthday party.

Trained research assistants transcribed the language samples from videotapes using the *Child Language Data Exchange System* (*CHILDES*; MacWhinney, 1995), and a second researcher verified and corrected all transcripts via audiotapes. Transcription procedures, the verifier, and a number of the transcribers were the same as those described by Price et al. (2008). In addition, 30 of the transcripts utilized in the current study were also analyzed for syntactic complexity by Price and colleagues. Morpheme-to-morpheme agreement reported by Price et al. was 85% for boys with FXS-O, 85% for boys with FXS-ASD, 90% for boys with DS, and 95% for TD boys.

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 (14 sentences in length) while showing the pictures to the child. Next, the examiner asked the child to tell the story while looking through the book.

A trained research assistant transcribed the narratives using *CHILDES* (MacWhinney, 1995), and a second researcher verified and corrected all transcripts via audio recordings. All of the transcripts utilized for the present investigation were also analyzed for story grammar elements by Estigarribia et al. (in preparation). Morpheme-to-morpheme agreement in the study by Estigarribia and colleagues was 85% for boys with FXS-O, 84% for boys with FXS-ASD, 81% for boys with DS, and 88% for TD boys.

NEPSY Verbal Fluency

The Verbal Fluency subtest of the *NEPSY, A Developmental Neuropsychological Assessment* (Korkman, Kirk, & Kemp, 1998) was designed to assess a child's ability to quickly generate words given semantic and phonemic categories. For the assessment of semantic fluency, the examiner asked the child to name as many different animals as he could (as quickly as possible) and then as many food or drink items. According to administration guidelines, the examiner gave the child two examples for each category. A time limit of 60 seconds was imposed for each category. Phonemic categories were not examined in the current study because too few of the boys were able to complete these tasks.

A trained research assistant transcribed all of the *NEPSY* samples from audio recordings, and the author transcribed about 12% of the samples (two from each participant group) for reliability. Morpheme-to-morpheme agreement between the two transcribers was 89% for the FXS-O group, 84% for the FXS-ASD group, 90% for the DS group, and 97% for the TD group.

Measures of Perseveration and Related Verbal Behaviors

Types of perseveration and related verbal behaviors were coded during play-based interaction, narration, and the verbal fluency test. Coding procedures and descriptions of the coding categories are provided next.

Play-based Interaction Measures

Coding of self-repetitive language produced during play-based interaction was done initially from the language transcripts, and verified later from the video. To obtain a language sample that was most reflective of genuine conversation, only certain activities of the *ADOS* (Lord et al., 2001) were coded. These activities included free-play, balloon and bubble blowing, a demonstration task, and/or a pretend birthday activity. 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. Criteria used for determining the occurrence of perseveration was similar to that described by Murphy and Abbeduto (2007). Utterances were coded for the following: (a) utterance-level perseveration; (b) ideational perseveration; and (c) conversational device repetition, as described below. See Appendix A for further details about the coding system.

Utterance-level perseveration. Utterance-level perseveration was coded when a child repeated a word (e.g., "hold <u>hold</u> on."), phrase (e.g., "I'm gonna <u>I'm gonna</u> take a break for now"), or entire utterance (e.g., "We need to put these away. <u>We need to put these away</u>.") one or more times. To count as utterance-level perseveration, the linguistic units had to be said 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). Utterance-level perseveration was not coded if a child repeated himself for emphasis or to repair a communicative breakdown (e.g., repeating a question or statement directed at the examiner because the examiner did not

respond the first time). Sub-coding for one (e.g., "hold hold on") versus two or more (e.g., "hold hold hold on") repetitions was also conducted.

Ideational perseveration. Ideational perseveration was coded when a child repeated ideas, topics, and themes excessively (at least two repetitions). Repetitions of this type did not need to be produced in immediate succession; ideas, topics, and themes could be reintroduced throughout the interaction. To determine if repetitions of this type were excessive, coders considered aspects of the interaction such as whether (a) the examiner could not divert the child from a topic, (b) the child offered the same information that he had added previously, and (c) the reintroductions were noncontingent (tangential or unrelated) with respect to the current focus of discourse. The following is an example of ideational perseveration from a study participant with FXS-ASD:

> 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 repetition. Conversational devices were defined as rote sayings and phrases (e.g., "Oh man," "Okay," "I don't know," "cool," and "hmm"). Conversational device repetition was coded when a conversational device was repeated two or more times throughout an interaction.

Narration Measures

Coding of self-repetitive language produced during picture-supported story retelling was done from the language transcripts. In order for a narrative language sample to be

eligible for coding, the child needed to produce at least nine intelligible utterances. Coding categories were the same as those used for the play-based interaction context.

Computation of Variables for Play-Based Interaction and Narrative Contexts

Four variables each were computed for the play-based interaction and narrative contexts: the proportion of utterance-level perseveration defined as one or more repetitions, the proportion of utterance-level perseveration defined as two or more repetitions, the proportion of ideational perseveration, and the proportion of conversational device repetition. For example, the proportion of ideational perseveration was computed by dividing the number of utterances coded as ideational perseveration by the total number of utterances.

NEPSY Verbal Fluency Measures

Coding of responses on the verbal fluency test was done from language transcripts. In order for a transcript to be eligible for coding, the child needed to produce at least five intelligible responses in each semantic category. Responses were coded for (a) stuck-in-set perseveration; (b) recurrent perseveration; (c) continuous perseveration; and (d) loss-of-set, as described below. Perseveration categories were those described by Sandson and Albert (1984). See Appendix B for further details about the coding system.

Perseveration. Stuck-in-set perseveration was coded when the child's first response for the category of food and drinks was an animal name (appropriate for the preceding task). If the child continued to name animals, these responses were also coded for this category of perseveration. A child's response was classified as *recurrent perseveration* if the response had already been given for that particular semantic category (e.g., "cow, tiger, monkey, cow, frog"). *Continuous perseveration* was coded when the child gave the same response twice or more in a row within the same task (e.g., "cow, tiger, tiger, monkey, frog").

Three perseveration variables were computed for the verbal fluency test. For example, the variable for recurrent perseveration was the proportion of responses categorized as this type of perseveration, computed by dividing the number of recurrent perseverations by the total number of responses.

Loss-of-set. Three types of loss-of-set behavior were coded. A child's response was coded as *proactive interference loss-of-set* if the child began to name food/drink items and then returned to naming animals (e.g., "hamburger, apple juice, camel"). *Tangential loss-of-set* was coded when the child's response was related to the current semantic category but not appropriate for the category (e.g., "great white shark, shark tooth"). *Unrelated loss-of-set* was coded when the child gave a response that did not appear to be related, even loosely, to the semantic category (e.g., "dog, hot air balloon").

Four loss-of-set variables were computed: the proportion of all loss-of-set responses (total loss-of-set), the proportion of proactive interference loss-of-set, the proportion of tangential loss-of-set, and the proportion of unrelated loss-of-set. For example, the variable for tangential loss-of-set was the proportion of responses classified as this type of loss-of-set, computed by dividing the number of responses coded as tangential loss-of-set by the total number of responses. The proportion of all loss-of-set responses was computed by dividing the number of utterances coded as either proactive-interference, tangential, or unrelated loss-of-set by the total number of utterances.

Coding Reliability

The author and two research assistants coded the language samples. The first research assistant, who coded the play-based interaction and narration samples, was an undergraduate student taking courses in speech and hearing sciences. The second research assistant, who

coded the verbal fluency context, was a Master's student in Linguistics. The coders were trained extensively by reviewing the coding systems and participating in consensus coding activities.

For each language sampling context, reliability was computed on a random subset (12%) of the samples using the kappa statistic (Cohen, 1960). Intercoder agreements using kappa statistics are generally lower than those computed by other methods since the kappa takes chance agreement into account (Fleiss, 1981); levels between .61 and .80 are judged to be "substantial" and levels between .81 and 1.0 are considered "almost perfect" (Landis & Koch, 1977). For play-based interaction, mean intercoder agreement computed on three boys from each group was .81 for utterance-level perseveration, .69 for ideational perseveration, and .77 for conversational device repetition. For narration, overall reliability computed on 3 boys from each group was .93 for utterance-level perseveration, .77 for ideational perseveration, and 1.0 for conversational device repetition. For the verbal fluency test, mean intercoder agreements computed on 2 boys from each group were 1.0 for all of the following codes: recurrent and continuous perseveration, and proactive interference, tangential, and unrelated loss-of-set. The kappa for stuck-in-set perseveration could not be computed because neither of the coders identified stuck-in-set in any of the randomly selected samples.

Data Analysis Strategy

A series of statistical models, all controlling for nonverbal MA, tested the effects of group (FXS-O, FXS-ASD, DS, and TD) on the measures of perseveration (utterance-level, ideational, stuck-in-set, recurrent, and continuous) and related behaviors (conversational device repetition and loss-of-set). An additional model tested the effects of context (narration and play-based interaction) on utterance-level perseveration, ideational perseveration, and

conversational device repetition. Each multivariate model was run as a hierarchical linear regression (HLM), also referred to as a multilevel model, whereas one analysis of covariance (ANCOVA) tested for group differences on the total measure of loss-of-set.

Sample sizes varied according to language sampling context. For play-based interaction, participants included 25 boys in each group. For narration, data were available for 26 boys with FXS-O, 23 boys with FXS-ASD, 25 boys with DS, and 25 TD boys. For the verbal fluency test, data were available for 19 boys with FXS-O, 15 boys with FXS-ASD, 15 boys with DS, and 18 TD boys.

The outcomes examined in the first HLM included the proportion of play-based interaction utterances classified as utterance-level perseveration (defined as one or more successive repetitions of a word, phrase, or entire utterance), ideational perseveration, or conversational device repetition. A second HLM, which controlled for the number of utterances produced in addition to nonverbal MA, tested whether groups differed on the different types of self-repetitive language (utterance-level perseveration again defined as one or more repetitions) during narration. The first and second models were repeated defining utterance-level perseveration as two or more successive repetitions of a word, phrase, or entire utterance. Another HLM tested whether the frequency of each type of self-repetitive language differed between interaction and narration for each group of boys.

For the next HLM, the outcomes examined included the proportion of responses on the verbal fluency test classified as stuck-in-set, recurrent, or continuous perseveration. Whereas an ANCOVA tested whether the groups differed on the total measure of loss-of-set, another HLM tested for groups differences on the different types of loss-of-set responses (proactive interference, tangential, and unrelated).

All models included nonverbal MA (as measured by the *Leiter-R*) to adjust the estimated means. For all analyses, pairwise tests comparing the participant groups were investigated when the *F* statistic indicated a significant overall effect of group. As a precaution against Type I errors, the *p* value for all pairwise tests was adjusted for multiple comparisons using Tukey's HSD (Honestly Significant Differences) method. Spearman correlations were also calculated among specific variables within and across language sampling contexts.

CHAPTER 3

RESULTS

Descriptive Analyses

In the following paragraphs, patterns of perseveration and related behaviors as displayed during the three language sampling contexts (play-based interaction, narration, and the verbal fluency test) within and across groups are described. The means and standard deviations that are reported in this section have not been adjusted for nonverbal MA.

Play-Based Interaction and Narration

Means and standard deviations for the proportion of utterances categorized as utterance-level perseveration, ideational perseveration, and conversational device repetition for the contexts of play-based interaction and narration are reported in Table 3.1. Here, utterance-level perseveration was defined as one or more successive repetitions of a word, phrase, or entire utterance. The number of utterances produced during narration varied by child. The mean numbers of utterances produced by boys with FXS-O, FXS-ASD, DS, and TD boys during narration were 16.3 (*SD* = 5.9), 16.7 (*SD* = 5.2), 17.6 (*SD* = 4.7), and 19.1 (*SD* = 7.3), respectively.

On average, utterance-level perseveration (defined as one or more repetitions) occurred in 10% of utterances during play-based interaction for boys with FXS-O, 13% for boys with FXS-ASD, 7% for boys with DS, and 10% for TD boys. Ideational perseveration occurred in 9% of utterances for boys with FXS-O, 14% for boys with FXS-ASD, and 5% for

Table 3.1

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

	Play-Based 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)
Ideational perseveration	.09 (.05)	.14 (.09)	.05 (.05)	.05 (.04)	.03 (.09)	.04 (.08)	.03 (.07)	.04 (.07)
Conversational device repetition	.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. Utterance-level perseveration = one or more repetitions.

both boys with DS and TD boys. Conversational device repetition 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.

Spearman partial correlations (partialing nonverbal MA) among the conversational outcome variables for each participant group were also calculated. Spearman correlations were appropriate due to the skewed distributions of some variables. None of the correlations were significant.

In narration, utterance-level perseveration (defined as one or more repetitions) 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. Ideational perseveration (3% to 4%) and conversational device repetition (0% to 1%) occurred infrequently for all groups. Despite the use of Spearman correlations, some of the correlations among the outcome variables were still not estimable due to the lack of variance for some variables (ideational perseveration for boys with DS, and conversational device repetition for the boys with FXS-ASD and DS). For TD boys, ideational perseveration significantly correlated with conversational device repetition ($r = .76$, $p = .0025$). No other correlations were significant.

When utterance-level perseveration was defined as two or more successive repetitions, it occurred infrequently in both play-based interaction and narration for all groups. Given this stricter criteria, utterance-level perseveration occurred in 1% of utterances during interaction for boys with FXS-O, 3% for boys with FXS-ASD, 1% for boys with DS, and 2% for TD boys on average. In narration, it occurred in 1% of utterances for boys with FXS-O, 2% for boys with FXS-ASD, 2% for boys with DS, and 3% for TD boys on average. Correlations were not calculated for this variable in order to limit the number of correlations.

NEPSY Verbal Fluency Subtest

Samples for a number of children who were administered the *NEPSY* could not be coded for this study. Reasons for exclusion were the following: (a) administration issues (divergence from standard directions, starting over after the child had begun giving responses, or giving a child a piece of candy after every response), (b) a child having fewer than 5 responses per semantic category, (c) a child repeating example items only (possible misunderstanding of task requirements), and (d) incomplete audio. This information is provided for each group in Table 3.2.

For those boys whose data were included in the current study, the number of responses produced during the *NEPSY* varied by child. The mean numbers of responses produced by boys with FXS-O, FXS-ASD, DS, and TD boys were 29.3 (*SD* = 15.5), 31.3 (*SD* = 10.5), 28.7 (*SD* = 16.9), and 22.8 (*SD* = 6.2), respectively. Means and standard deviations for stuck-in-set, recurrent, and continuous perseveration, and proactive interference, tangential, unrelated, and total loss-of-set responses are shown in Table 3.3. On average, stuck-in-set perseveration occurred infrequently for all groups (0% to 1% of responses categorized as stuck-in-set). Recurrent perseveration constituted 17% of responses for boys with FXS-O, 22% for both boys with FXS-ASD and boys with DS, and 10% for TD boys. Continuous perseveration made up 6% of responses for boys with FXS-O, 4% for boys with FXS-ASD, and 2% for both boys with DS and TD boys.

Loss-of-set constituted 10% of responses on the verbal fluency test for boys with FXS-O, 13% for boys with FXS-ASD, 9% for boys with DS, and 2% for TD boys on average. Proactive interference loss-of-set occurred infrequently for all groups (0% to 2%). Tangential loss-of-set made up 6% of responses for boys with FXS-O, 4% for both boys with

Table 3.2

Children Excluded from NEPSY Coding and Reasons for Exclusion

		No. (%) not coded due to:				
Group	Total administered	Administration	Fewer than 5 responses in either category	Repetition of example items only	Incomplete audio	Total no. (%) of samples not coded
FXS-O	26	2 (8%)	5 (19%)	0 (0%)	0 (0%)	7 (27%)
FXS-ASD	26	1 (4%)	7 (27%)	1 (4%)	2 (8%)	11 (42%)
DS	23	3 (13%)	1 (4%)	2 (9%)	2 (9%)	8 (35%)
TD	28	4 (14%)	5 (18%)	0 (0%)	1 (4%)	10 (36%)

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

Table 3.3

Means and Standard Deviations for Proportions of Responses on the NEPSY Verbal Fluency Subtest Categorized as Perseveration or Loss-of-Set

Outcome	FXS-O (N=19)	FXS-ASD (N=15)	DS (N=15)	TD (N=18)
	M (*SD*)	*M* (*SD*)	*M* (*SD*)	*M* (*SD*)
Perseveration				
Stuck-in-set	.01 (.02)	.01 (.02)	.00 (.02)	.00 (.00)
Recurrent	.17 (.17)	.22 (.11)	.22 (.10)	.10 (.17)
Continuous	.06 (.09)	.04 (.04)	.02 (.02)	.02 (.03)
Loss-of-Set				
Proactive Interference	.01 (.04)	.01 (.03)	.02 (.04)	.00 (.01)
Tangential	.06 (.09)	.04 (.04)	.04 (.04)	.00 (.01)
Unrelated	.03 (.07)	.08 (.12)	.03 (.04)	.02 (.04)
Total Loss-of-Set	.10 (.11)	13 (.16)	.09 (.08)	.02 (.04)

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

FXS-ASD and boys with DS, and 0% for TD boys. Unrelated loss-of-set constituted 3% of responses for boys with FXS-O, 8% for boys with FXS-ASD, 3% for boys with DS, and 2% for TD boys.

Spearman partial correlations among the outcome variables for each group were also examined. To minimize the number of correlations, only the total loss-of-set variables were used. For TD boys, correlations with stuck-in-set perseveration were not estimable due to the lack of variance on this variable. For boys with FXS-ASD, loss-of-set significantly correlated with both stuck-in-set ($r = .58$, $p = .0463$) and continuous perseveration ($r = .61$, $p = .0365$). For TD boys, loss-of-set significantly correlated with recurrent perseveration ($r = .57$, $p = .0399$). No other correlations were significant.

Group Comparisons

In this section, analysis results comparing the groups on the outcome measures for each of the language sampling contexts are reported. The means reported here have been adjusted for nonverbal MA.

Play-Based Interaction

The first model compared types of self-repetitive language (utterance-level perseveration, ideational perseveration, conversational device repetition) across participant groups (FXS-O, FXS-ASD, DS, TD) for the play-based interaction context, controlling for nonverbal MA. The model was run as an HLM via the computer software program SAS PROC MIXED (SAS Institute, 2002). In this model, utterance-level perseveration was defined as one or more successive repetitions of a word, phrase, or entire utterance.

The F statistic, $F(2, 287) = 8.71$, $p < .0001$, indicated a significant effect of group on the production of self-repetitive language. In addition, a significant interaction between

diagnosis and type of self-repetitive language, $F(6, 287) = 4.71$, $p = .0001$, indicated that between-group differences varied by type of self-repetitive language. The between-group differences and adjusted means (adjusted for nonverbal MA) are presented in Table 3.4. The boys with FXS-ASD produced significantly more utterance-level perseveration ($M = 0.13$) than did the boys with DS ($M = 0.07$). The boys with FXS-ASD also produced significantly more ideational perseveration ($M = 0.14$) than boys with FXS-O ($M = 0.09$), boys with DS ($M = 0.05$), and TD boys ($M = 0.05$). Additionally, the boys with FXS-O produced significantly more ideational perseveration than boys with DS and TD boys. The groups did not differ significantly on the measure of conversational device repetition.

The model was repeated defining utterance-level perseveration as two or more successive repetitions of a word, phrase, or entire utterance. The change in the model did not impact the findings for ideational perseveration and conversational device repetition. However, the previous significant difference found for utterance-level perseveration when the criterion was one or more repetitions were not replicated with a criterion of two or more repetitions.

Narration

Types of self-repetitive language were also compared across participant groups for the narrative context, controlling for nonverbal MA and number of utterances produced. The model was run as an HLM via SAS PROC MIXED (SAS Institute, 2002). In this model, utterance-level perseveration was defined as one or more successive repetitions of a word, phrase, or entire utterance.

The F statistic, $F(3, 283) = 2.50$, $p = .0599$, did not indicate a significant effect of group on the production of self-repetitive language. However, a significant interaction

Table 3.4

Adjusted Means (adjusted for Leiter-R mental age) and Between-Group Differences for the Types of Self-Repetitive Language in Play-Based Interaction

Outcome	FXS-O (N=25)	FXS-ASD (N=25)	DS (N=25)	TD (N=25)
Utterance-level perseveration	0.10[a,b]	0.13[a]	0.07[b]	0.10[a,b]
Ideational perseveration	0.09[b]	0.14[a]	0.05[c]	0.05[c]
Conversational device repetition	0.04[a]	0.04[a]	0.05[a]	0.05[a]

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

between diagnosis and type of self-repetitive language, $F(6, 283) = 2.34$, $p = .0321$, indicated that between group differences varied by type of self-repetitive language. The between-group differences and adjusted means are presented in Table 3.5. The TD boys produced significantly more utterance-level perseveration ($M = 0.20$) than boys with FXS-O ($M = 0.12$) and DS ($M = 0.11$). The groups did not differ significantly on either ideational perseveration or conversational device repetition.

The model was repeated defining utterance-level perseveration as two or more successive repetitions. The change in the model did not impact the findings for ideational perseveration and conversational device repetition. However, the previous significant differences found for utterance-level perseveration were not replicated using the new criterion.

NEPSY Verbal Fluency Subtest

Perseveration. The next analysis compared types of perseveration (stuck-in-set, recurrent, continuous) produced during the verbal fluency test across participant groups, controlling for nonverbal MA. The model was run as an HLM via SAS PROC MIXED (SAS Institute, 2002). The F statistic, $F(3, 188) = 2.39$, $p = .0699$, did not indicate a significant effect of group on the production of perseveration. In addition, the interaction between diagnosis and type of perseveration, $F(6, 188) = 7.74$, $p = .0659$, was not significant. Pairwise tests were not conducted due to the nonsignificant results, although adjusted means are presented in Table 3.6.

Loss-of-Set. The groups were next compared on the total measure of loss-of-set errors, controlling for nonverbal MA. The model was run as an ANCOVA, with nonverbal MA as the covariate, via the software program SAS PROC GLM (SAS Institute, 2002). The

Table 3.5

Adjusted Means (adjusted for Leiter-R mental age and number of utterances) and Between-Group Differences on the Measures of Self-Repetitive Language in Narration

Outcome	FXS-O (N=26)	FXS-ASD (N=23)	DS (N=25)	TD (N=25)
Utterance-level perseveration	0.12[b]	0.14[a,b]	0.11[b]	0.20[a]
Ideational perseveration	0.03[a]	0.04[a]	0.03[a]	0.04[a]
Conversational device repetition	0.00[a]	0.00[a]	0.00[a]	0.01[a]

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

Table 3.6

Adjusted Means (adjusted for Leiter-R mental age) by Group on the Measures of Perseveration on the NEPSY Verbal Fluency Subtest

Outcome	FXS-O (N=19)	FXS-ASD (N=15)	DS (N=15)	TD (N=18)
Stuck-in-set perseveration	0.01	0.00	0.00	0.00
Recurrent perseveration	0.17	0.22	0.21	0.11
Continuous perseveration	0.06	0.04	0.02	0.02

Note. FXS-O = fragile X syndrome only; FXS-ASD = fragile X syndrome with autism spectrum disorder; DS = Down syndrome; TD = typically developing. Significance tests of these means were not conducted due to the nonsignificant effect of group and the nonsignificant effect of the interaction between group and type of perseveration.

F statistic, $F(3, 64) = 3.03$, $p = .0416$, indicated a significant effect of group. The boys with FXS-O ($M = 0.10$), FXS-ASD ($M = 0.13$), and DS ($M = 0.09$) produced significantly more loss-of-set errors than did the TD boys ($M = 0.02$). The boys with FXS-O, FXS-ASD, and DS did not significantly differ from each other. The between-group differences and adjusted means are presented in Table 3.7.

The fifth analysis compared specific types of loss-of-set responses (proactive interference, tangential, unrelated) across participant groups, controlling for nonverbal MA. The model was run as an HLM via SAS PROC MIXED (SAS Institute, 2002). The *F* statistic, $F(3, 62) = 2.91$, $p = .0416$, indicated a significant effect of group on the production of loss-of-set errors (consistent with the results of the ANCOVA). However, the interaction between diagnosis and type of loss-of-set, $F(6, 126) = 1.96$, $p = .0769$, was not significant. Pairwise tests were not conducted due to the nonsignificant results, although adjusted means are presented in Table 3.7.

Context Effects

In the following paragraphs, analysis results are reported for the within-group comparisons that addressed the effects of language sampling context on the outcome variables. Also reported are the significant results for the across-context correlations. Given the previous nonsignificant findings for utterance-level perseveration when it was defined as two or more repetitions, all analyses below refer to utterance-level perseveration defined with the criterion of one or more repetitions.

Relationships Between Outcome Variables in Play-Based Interaction and Narration

For each group, Spearman partial correlations were calculated for each type of self-repetitive language (utterance-level perseveration, ideational perseveration, conversational

Table 3.7

Adjusted Means (adjusted for Leiter-R mental age) by Group on the Measures of Loss-of-Set on the NEPSY Verbal Fluency Subtest

Outcome	FXS-O (N=19)	FXS-ASD (N=15)	DS (N=15)	TD (N=18)
Total Loss-of-Set	0.10[a]	0.13[a]	0.09[a]	0.02[b]
Subtypes				
Proactive Interference	0.02	0.01	0.02	0.00
Tangential	0.06	0.04	0.02	0.00
Unrelated	0.03	0.08	0.03	0.02

Note. FXS-O = fragile X syndrome only; FXS-ASD = fragile X syndrome with autism spectrum disorder; DS = Down syndrome; TD = typically developing. Significance tests of subtype means were not conducted due to the nonsignificant effects of the interaction between group and type of perseveration.

device repetition) produced during play-based interaction and its narrative counterpart. Despite the use of Spearman correlations, some of the correlations among the outcome variables were still not estimable due to the lack of variance for some variables (conversational device repetition during narration for the boys with FXS-ASD, and ideational perseveration and conversational device repetition during narration for the boys with DS). For boys with FXS-O, utterance-level perseveration (defined as one or more repetitions) in interaction and narration significantly correlated ($r = .60$, $p = .0379$). No other correlations were significant.

Comparison of Play-Based Interaction and Narration

The final model addressed the effects of language sampling context (play-based interaction and narration) on self-repetitive language in each of the four groups, controlling for nonverbal MA. The model was run as an HLM via SAS PROC MIXED (SAS Institute, 2002). A significant 3-way interaction between diagnosis, type of self-repetitive language, and context, $F(6, 470) = 2.28$, $p = .0349$, indicated that between-group and scale differences varied by context.

The TD boys produced significantly more utterance-level perseveration (defined as one or more repetitions) in narration than in play-based interaction ($p < .001$). The boys with FXS-O and FXS-ASD produced significantly more ideational 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 repetition in interaction than in narration ($p < .001$, $p = .012$, and $p = .035$, respectively).

Relationships Between Perseveration on the NEPSY Verbal Fluency Subtest and Perseveration in either Play-Based Interaction or Narration

Group comparisons of perseveration on the verbal fluency test and perseveration during play-based interaction and narration were not conducted due to the differing coding schemes. However, correlations were examined between perseveration on the verbal fluency test (stuck-in-set, recurrent, continuous) and perseveration in interaction and narration (utterance-level and ideational). Correlation analyses included only the perseveration variables (and not conversational device repetition and loss-of-set) to minimize the number of correlations. Several correlations were not estimable due to the lack of variance for two variables (ideational perseveration in narration for the boys with DS, and stuck-in-set perseveration for the TD boys).

The sample sizes available for these correlation analyses typically dropped in comparison with the previous analyses. Fifteen boys with FXS-O had both play-based interaction and verbal fluency data, and 16 had both narration and verbal fluency data. Fifteen boys with FXS-ASD had both interaction and verbal fluency data, and 13 had both narration and verbal fluency data. Twelve boys with DS had both interaction and verbal fluency data, and 13 had both narration and verbal fluency data. Fourteen TD boys had both interaction and verbal fluency data, and 14 had both narration and verbal fluency data. Significant correlations were found only for the boys with FXS-ASD. Utterance-level perseveration (defined as one or more repetitions) in interaction significantly correlated with both recurrent perseveration ($r = .60$, $p = .0401$) and continuous perseveration on the *NEPSY* ($r = .63$, $p = .0275$).

CHAPTER 4

DISCUSSION

Verbal perseveration is a frequently reported characteristic of males with FXS (Madison et al., 1986; Murphy & Abbeduto, 2007; Roberts, Martin, et al., 2007; Sudhalter et al., 1990), and many researchers have argued that it is a defining feature of the behavioral phenotype of FXS (Abbeduto et al., 2007; Abbeduto & Hagerman, 1997; Bennetto & Pennington, 1996; Roberts, Chapman, et al., 2008). The study of perseveration is vital because perseveration can be exceedingly frustrating for caregivers and other communication partners, present a major obstacle to social interaction, and also affect an individual's ability to demonstrate underlying skills during assessments in various domains. However, previous studies of perseveration in individuals with FXS have been limited in several respects. Perseveration has often been defined as if it were a single behavior, and conversation has served almost exclusively as the language sampling context. The present study was the first to (a) compare boys with FXS, boys with DS, and TD boys of similar nonverbal MA on different types of perseveration; (b) examine the effects of autism status on the production of different types of perseveration by including boys with FXS with and without ASD; (c) examine perseveration in a context other than connected speech (a verbal fluency test, in addition to play-based interaction and narration); and (d) apply a taxonomy of perseveration from the literature on adult neurological disorders.

The results revealed that boys with FXS, regardless of autism status, produced significantly more ideational perseveration (excessive repetition of ideas, topics, and themes) during play-based interaction after controlling for nonverbal MA than did boys with DS and younger TD boys. Further, the boys with FXS-ASD produced significantly more ideational perseveration than did boys with FXS-O, and significantly more utterance-level perseveration (successive repetition of a word, phrase, or entire utterance one or more times) during interaction than did boys with DS. These differences were not found for the narrative context. However, in narration, the boys with FXS-O and the boys with DS displayed significantly less utterance-level perseveration than did the TD boys after controlling for nonverbal MA and number of utterances produced. Further, in interaction as compared to narration, boys with FXS, regardless of autism status, produced significantly more ideational perseveration; boys with FXS-ASD, boys with DS, and TD boys produced significantly more conversational device repetition; and TD boys displayed significantly less utterance-level perseveration. On the verbal fluency test, the three disability groups (FXS-O, FXS-ASD, and DS) produced significantly more loss-of-set responses than did the TD boys. Implications of these findings are discussed in the following sections.

Boys with FXS

FXS with and without ASD

For boys with FXS, regardless of autism status, findings of the present investigation suggest that perseveration occurs more frequently in play-based interaction than would be expected based on nonverbal MA. Further, findings suggest that perseveration in boys with FXS cannot simply be explained by the presence of ID. These findings are consistent with those of other researchers (Roberts, Martin, et al., 2007; Wolf-Schein et al., 1987). However,

previous studies did not compare groups on different types of perseveration, and so the present findings are not specifically comparable with previous findings. In the present investigation, both the boys with FXS-O and the boys with FXS-ASD produced significantly more ideational perseveration (9% and 14% of utterances, respectively) during interaction than did boys with DS (5%) and TD boys (5%).

Currently, the factors underlying the observed group differences for ideational perseveration during play-based interaction are unclear. Researchers (Belser & Sudhalter, 1995; Cornish et al., 2004; Murphy & Abbeduto, 2007; Sudhalter et al., 1990) have commonly attributed perseveration in individuals with FXS to the excessive arousal and anxiety that are well documented features of the behavioral phenotype (Hagerman, 2002; Hessl et al., 2001; Mazzocco et al., 1998). Specifically, Belser and Sudhalter (1995) proposed that hyperarousal causes the "sentence template" that precedes the production of an utterance to decay too slowly, and that characteristics of a previous sentence (e.g., the topic) are unintentionally repeated. The more recent of these arguments (Cornish et al., 2004; Murphy & Abbeduto, 2007) implicated hyperarousal in tandem with deficits in inhibitory control and attention-switching as a cause for topic perseveration (similar in description to ideational perseveration). For example, Cornish and colleagues (2004) suggested that hyperarousal activates extraneous neuronal connections, and thereby interferes with one's ability to inhibit or switch attention.

It is also possible that anxiety influences the production of ideational perseveration by decreasing the attention available for inhibiting previous responses. According to attentional control theory (Eysenck et al., 2007), anxiety causes attention to be allocated to threat-based stimuli, finding its source, and deciding how to respond. Thus, according to this account,

attention in the anxious individual is not available for other processes, such as inhibiting task-irrelevant responses. This argument is consistent with Kahneman's (1973) attention and information processing capacity model, which assumes a limited amount of attention capacity. In the anxious child with FXS, perhaps attention is spent avoiding eye contact or dealing with internal feelings of "worry" rather than inhibiting prior responses. Furthermore, attentional control theory proposes that the executive function of cognitive shifting is affected when attention is allocated to threat-based stimuli. Thus, the child with FXS may not be able to follow topic changes made by a communication partner, instead remaining "stuck" on a single topic. Although potential mechanisms were not examined in the current study, there is evidence that boys with FXS have greater difficulty switching attention and inhibiting repetition than do boys with DS and TD boys (Wilding et al., 2002), which could help to explain the present findings.

The observed group differences for ideational perseveration during play-based interaction could also be due to the boys with FXS lacking an understanding of what Grice (1975) termed the Cooperative Principle, and the conversational maxim of "Do not make your contribution more informative than is required" in particular. Clearly, a speaker who continues to reintroduce the same information is not adhering to this rule. Deficits in theory of mind could support this hypothesis. That is, someone who cannot attribute knowledge to a communication partner would be unaware that previous contributions to conversation have already been stored by that partner.

Boys with FXS did not significantly differ from either of the other two groups on any type of perseveration during the verbal fluency test. However, the sample sizes for this context (15 to 19 in each group) were considerably smaller than for conversation (25 in each

group) and narration (23 to 26 in each group), and this may have affected the ability to detect group differences in the samples that may be indicative of actual differences in the population. Indeed, recurrent perseveration constituted 17% of responses for boys with FXS-O, 22% for both boys with FXS-ASD and boys with DS, and only 10% for TD boys. Additionally, continuous perseveration made up 6% of responses for boys with FXS-O, 4% for boys with FXS-ASD, and only 2% for both boys with DS and TD boys. With more participants, boys with FXS with and without ASD (along with boys with DS) may have been found to produce significantly more recurrent perseveration than the younger TD boys, and boys with FXS with and without autism may have been found to produce significantly more continuous perseveration than either boys with DS and TD boys. Stuck-in-set perseveration, on the other hand, occurred infrequently for all groups (0% to 1% of responses). However, the use of only two semantic categories limited the opportunity for any of the boys to produce this type of perseveration. Moreover, providing examples may have given all boys an advantage for "getting into set" even if they had a natural tendency to evidence problems in set-switching.

The groups did differ significantly on the overall measure of loss-of-set behavior on the verbal fluency test. Boys with FXS-O (10% of responses) and boys with FXS-ASD (13%) produced significantly more loss-of-set errors than did TD boys (2%), but did not significantly differ from boys with DS (9%). Loss-of-set behavior may reflect difficulties in sustained attention (Kaplan, Sengor, Gurvit, Genc, & Guzelis, 2006). Therefore, boys with FXS (as well as boys with DS) may not have been able to attend long enough to the category at hand. In fact, boys with FXS show greater impairments in sustained attention than do boys with DS (Munir et al., 2000), who themselves evidence greater impairments in sustained

attention than do TD children (Brown et al., 2003). In addition, the group differences for loss-of-set responses may reflect richer semantic knowledge of the TD boys. Specifically, the TD boys may have possessed better categorical knowledge than did the children with intellectual disability in the present investigation.

The boys with FXS did not significantly differ from the other groups on the specific types of loss-of-set behavior. However, tangential loss-of-set made up 6% of responses for boys with FXS-O, 4% for both boys with FXS-ASD and boys with DS, and 0% for TD boys. With more participants, boys with FXS with and without ASD (along with boys with DS) may have been found to produce significantly more tangential loss-of-set than the younger TD boys.

Effects of Autism Status

In the present investigation, the boys with FXS-ASD produced significantly more ideational perseveration (14% of utterances) during play-based interaction than did the boys with FXS-O (9%). Consistent with the argument that deficits in attention-switching may underlie ideational perseveration in boys with FXS to some extent (see previous section), attention-switching in children with ASD without FXS has been found to be impaired compared with both TD children (Landry & Bryson, 2004; Rinehart, Bradshaw, Moss, Brereton, & Tonge, 2001) and children with DS (Landry & Bryson, 2004). Perhaps the added effect of autism on attention-switching resulted in boys with FXS-ASD producing more ideational perseveration than boys with FXS-O. Moreover, if ideational perseveration, in part, reflects lack of understanding of Grice's (1975) conversational maxim of "Do not make your contribution more informative than is required," and if this deficit can be explained by

theory of mind ability (see previous section), then the well-documented theory of mind deficits in autism (see Tager-Flusberg, 1999, for a review) could help to explain this result.

Additionally, the boys with FXS-ASD produced significantly more utterance-level perseveration (13% of utterances) during play-based interaction than did boys with DS (7%). Murphy and Abbeduto (2007) proposed that utterance-level repetition may reflect word-finding difficulties. For example, a child could say "I want the want the want the ball," repeating the linguistic unit "want the" while searching for the word "ball." Perhaps word-finding skills during interaction are more impaired in boys with FXS-ASD than they are in boys with DS. In addition, utterance-level perseveration as described in the fragile X literature is consistent with descriptions of continuous perseveration found in the adult neurological disorders literature. Sandson and Albert (1987) proposed that continuous perseveration reflects difficulties in disengaging attention. For example, a child could say "I want the ball. I want the ball. I want the ball," because he or she cannot disengage attention from the "ball" (or, more specifically, the desire to obtain it) long enough for the communication partner to respond. In fact, utterance-level perseveration in interaction significantly correlated with continuous perseveration on the verbal fluency test for the boys with FXS-ASD in the present investigation. As previously discussed, attention-switching is a problem for children with ASD without FXS (Landry & Bryson, 2004; Rinehart et al., 2001), and this may help to explain this result. Of course, these two possible explanations for this research finding also highlight the potentially important differences in *types* of utterance-level perseveration (i.e., repetition of phrase or repetition of entire utterance). This distinction was not addressed in the present investigation.

Although groups did not differ significantly, unrelated loss-of-set on the verbal fluency test constituted 8% of responses for boys with FXS-ASD, compared with 3% of responses for boys with FXS-O, 3% for boys with DS, and 2% for TD boys. With more participants, boys with FXS-ASD may have been found to produce significantly more unrelated loss-of-set than all other groups.

Boys with DS

In the present study, the boys with DS were not found to produce any type of perseveration more often than any other group in any of the three language sampling contexts. However, recurrent perseveration on the verbal fluency test constituted 22% of responses for boys with DS, compared with only 10% for TD boys. With more participants, boys with DS may have been found to produce significantly more recurrent perseveration than the younger TD boys.

The boys with DS did produce significantly more loss-of-set responses overall (9% of responses) than did TD boys (2%) on the verbal fluency test. As previously discussed, loss-of-set behavior may reflect difficulties in sustained attention (Kaplan et al., 2006), and children with DS have been found to evidence greater impairments in sustained attention than do TD children (Brown et al., 2003). Moreover, this finding might be explained by the richer semantic knowledge of the TD boys (see previous section). While the groups did not differ significantly on the specific types of loss-of-set behavior, tangential loss-of-set made up 4% of responses for boys with DS, and only 0% for TD boys. With more participants, boys with DS may have been found to produce significantly more tangential loss-of-set than TD boys.

TD Boys

In the present investigation, TD boys surprisingly produced more utterance-level perseveration (21% of utterances) during narration than did boys with FXS-O (11%) and boys with DS (11%). If Murphy and Abbeduto (2007) were correct when they posited that utterance-level perseveration in some instances may be reflective of the word retrieval process, perhaps the TD boys were expending more effort than the other boys during their attempts to retrieve story elements from memory. It is also possible that utterance-level perseveration in the TD boys reflected their use of repair strategies (minor revisions of their own statements), and thus their best attempts to be most accurate during the story retelling. Thus, the pronounced occurrence of perseveration in the TD group might actually relate to better storytelling skills.

Effects of Language Sampling Context

The present study was the first to compare boys with FXS, boys with DS, and TD boys on measures of perseveration during narration, and findings indicate that language sampling context plays a role in perseveration. None of the significant group differences found for the play-based interaction context were found for narration. Further, TD boys produced significantly more utterance-level perseveration in narration than they did in interaction, possibly due to word-retrieval or repair processes (discussed previously). Additionally, boys with FXS with and without ASD produced significantly more ideational perseveration in interaction than in narration. This result is consistent with the finding of Murphy and Abbeduto (2007), who reported that adolescent males with FXS without autistic disorder produced more topic repetition in interaction than in narration. Murphy and Abbeduto (2007) proposed two explanations for their finding. First, the interpersonal demands of conversation (e.g., eye contact) may have led to a state of heightened arousal and

anxiety in the males with FXS. Second, the added structure of the narrative format may have lessened the effects of any impairments in executive function. A more parsimonious explanation for this finding is that hyperarousal leads to inhibitory control and attention-switching problems through the activation of extraneous neuronal connections (Cornish et al., 2004).

The finding that ideational perseveration occurred more frequently during play-based interaction than in narration for boys with FXS can also be discussed within a limited capacity model. A limited capacity framework proposes that performance decreases as tasks become more difficult (Alt & Plante, 2006). In the current study, the narration task, being of the picture-supported story retelling format, arguably required less cognitive resources than the play-based interaction. During play-based interaction, multiple sources of information needed to be encoded simultaneously. Thus, the demands of play-based interaction could have further taxed the attention system and prevented it from effectively inhibiting previous responses or switching attention from one topic to another.

Finally, boys with FXS-ASD, boys with DS, and TD boys produced significantly more conversational device repetition in play-based interaction as compared with narration. This finding is not surprising given the nature (and even the name) of this category of self-repetition. Many conversational devices are acknowledgements of a communication partner (e.g., “mhm,” “okay”), and such devices are simply not necessary during narration. In general, conversational device repetition occurred infrequently across groups and across contexts compared with the two types of perseveration.

Within-group comparisons of perseveration on the verbal fluency test and perseveration during play-based interaction and narration were not conducted due to the

differing taxonomies. However, correlations were examined between perseveration on the verbal fluency test (stuck-in-set, recurrent, continuous) and perseveration in interaction and narration (utterance-level and ideational). The one significant finding was that the utterance-level perseveration of boys with FXS-ASD in interaction correlated with both recurrent and continuous perseveration on the verbal fluency test. It is possible that a common mechanism (maybe attention, as previously discussed) underlies all three types of perseveration in boys with FXS-ASD, although this was not examined in the present study.

Discrepancy in Findings for Utterance-Level Perseveration

One aim of the present study was to determine whether findings for utterance-level perseveration would differ depending on the coding criterion (one versus two or more repetitions). When utterance-level perseveration was defined as one or more successive repetitions of a word, phrase, or entire utterance, the boys with FXS-ASD were found to produce significantly more of this type of perseveration during play-based interaction than boys with DS, and the TD boys were found to produce significantly more utterance-level perseveration during narration than boys with FXS-O and boys with DS. Interestingly, these two significant findings were not replicated using a stricter criterion of two or more repetitions. This finding should generate discussion among researchers as to whether or not just one repetition of a word, phrase, or utterance constitutes "excessive self-repetition." It is argued here that even one immediate repetition is excessive (and, therefore, perseverative) due to the close proximity of the repetition to the original linguistic unit.

Study Strengths and Limitations

This study has several strengths. First, sample sizes of boys with FXS, boys with DS, and TD boys for play-based interaction and narrative contexts were large compared with

sample sizes of previous studies. Second, the MA-matched comparison groups of boys with DS and boys who were developing typically helps to determine whether perseveration and related behaviors exhibited by boys with FXS are a function of developmental level or ID in general. Third, boys with FXS with and without ASD were included in order to investigate the effects of autism status on perseveration in FXS. Finally, different types of perseveration were examined and multiple language sampling contexts were utilized.

The present study also had several limitations, which have implications for future research. First, sample sizes available for the examination of perseveration on the verbal fluency test were small compared with those available for play-based interaction and narration. Thus, the ability to detect group differences may have been limited. Future studies should utilize verbal fluency tests, but include a larger number of participants. Second, the FXS-ASD group consisted of boys who were classified as "autism" or "autism spectrum" on the *ADOS* (Lord et al., 2001). Added measures of autism should be used in forthcoming studies to classify children. Third, a comparison group of children with ASD without FXS would further our understanding of the contribution of autism status to perseveration in boys with both FXS and ASD. Future studies should make such comparisons. Fourth, the current study included only *boys* with FXS, and examination of perseveration among girls should be conducted in the future.

Fifth, examiner-child interactions that occurred during administration of the *ADOS* (Lord et al., 2001) served as the samples of play-based interaction, but the *ADOS* was not specifically designed to be a language sample. However, care was taken to code only those activities, such as free-play, that were considered to be most representative of natural conversation. Nevertheless, future investigations should examine perseveration during more

naturalistic contexts and with both familiar and unfamiliar conversation partners. Sixth, the narrative sample was from a picture-supported story retelling task, 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. Future studies should examine perseveration in different types of narrative tasks, including personal narratives. Seventh, all analyses in the present study included nonverbal MA as a covariate. Future investigations could also control for a verbal measure of intelligence and determine if the significant findings remain. Eighth, while the observed group differences and the observed effects of language sampling context certainly prompt discussion of underlying mechanisms of perseveration in boys with FXS, these mechanisms were not directly examined in the current study. Future studies should investigate predictors of perseveration in boys with FXS, such as anxiety, attention, inhibitory control, and theory of mind. Findings from predictor studies would likely have great implications for the management of perseveration. Finally, the current study examined perseveration at only one point in time. Future investigations should study perseveration in the same groups of children longitudinally and examine the factors that affect perseveration over time.

Clinical Implications

Findings of the current investigation have several important assessment and intervention implications. Because boys with FXS-ASD were found to produce more ideational perseveration than boys with FXS-O, whether a child with FXS also has a diagnosis of ASD should be considered during assessment. In addition, perseveration was found to be pronounced in boys with FXS compared with boys with DS and TD boys during play-based interaction only. Therefore, the evaluation of perseveration during play-based

interaction and conversation, as well as the examination of different types of perseveration, is recommended for clinicians.

Speech and language interventions for boys with FXS who produce verbal perseveration should include goals for decreasing these behaviors. The finding that ideational perseveration (which appears to be a problem for boys with FXS regardless of autism status) occurred more frequently in play-based interaction than in picture-supported story retelling suggests that adding structure to conversational interactions (e.g., using picture supports to maximize topic maintenance) might decrease perseveration. Scharfenaker, O'Conner, Stackhouse, Braden, and Gray (2002) suggested several other strategies for managing verbal perseveration in individuals with FXS, such as monitoring of anxiety levels and utilizing verbal redirection. Another method suggested for managing the perseverative behaviors of individuals with aphasia, which is to raise such behaviors to a "level of awareness" (Helm-Estabrooks & Albert, 2004), may also be successful with some children with FXS. For the child who perseverates on just a few topics, interventions might focus on increasing the number of topics on which he or she can converse. Of course, identification of the underlying mechanisms of perseveration in children with FXS and whether they differ according to type of perseveration and autism status would certainly inform intervention approaches.

Summary

In summary, boys with FXS with and without ASD were found to produce more perseveration during play-based interaction than boys with DS and younger TD boys. In particular, ideational perseveration was observed significantly more frequently during interaction among boys with FXS, regardless of autism status, than among boys with DS and TD boys after controlling for nonverbal MA. Further, during interaction, the boys with FXS-

ASD produced significantly more ideational perseveration than all other groups, and more utterance-level perseveration than boys with DS. These between-group differences were not found for the narrative context, and boys with FXS with and without ASD were found to produce significantly more ideational perseveration in interaction than in narration. On a verbal fluency test, boys with FXS-O, boys with FXS-ASD, and boys with DS produced significantly more loss-of-set responses than the younger TD boys. The groups did not differ on measures of perseveration on the verbal fluency test, although the ability to detect group differences was likely affected by sample size. These findings suggest that boys with FXS, regardless of autism status, produce more perseveration than would be expected based on nonverbal MA or ID in general, and that autism status as well as language sampling context affect perseveration in boys with FXS.

Appendix A

Coding Manual for Self-Repetitive Language (Play-Based Interaction Context)

Contents

Coding Outline

1. Utterance-level perseveration:

    a. **U2**

    b. **U3**

    c. In the "extra" column: **Q** (as appropriate)

2. Ideational perseveration: **(I)**

    a. Ideational perseveration questionable: **(IQ)**

    b. In the "extra" column: **Q** (as appropriate)

3. Conversational device repetition:

    a. **CVO or CVB**

    b. **CWO or CWB**

    c. In the "extra" column: **Q** (as appropriate)

Suggested Coding Steps

1. Add your initials to the end of the filename. Enter your initials in the "coder" cell of the excel spreadsheet.

2. Start with the beginning of the *ADOS* transcript. Do not code the "blocks" activity (which typically comes first). Code everything after "blocks" except for the "imitation" and "book reading" activities.

3. Code the first 100 intelligible utterances (do not code singing if a popular song).

4. Look through each of the 100 numbered utterances and determine if there are any utterance perseverations (coded as **U**; see section on utterance perseveration).

5. While performing step 4, look at each utterance to determine and record the major utterance elements (in most cases, the actual words). In the Excel document, there will be columns for subject, verb, object, and adjective (adj. column can also be used for adverbs). Enter the significant words in their respective categories on the same row as the utterance itself. Remember to pay attention to pronouns and their referents while filling out the columns.

6. After going through all of the 100 usable utterances once, go through them again. Here you are just concerned with: 1) child utterances that follow examiner questions and 2) child questions. This time, pay attention to the words entered in your "sentence element" columns, and highlight in red any words that will not be considered when coding ideational perseveration (**I**; see section on ideational perseveration for rules to follow)**:**

7. Looking only at the words in the subject, verb, object, and adjective columns that are *not* red, scan through the columns to see which words come up multiple times. To facilitate this process, take the first word that you are considering and highlight it in yellow. Scan through each of the four elemental columns. If the word occurs in the columns three times, highlight the word in blue and code **I**. For any time that the word occurs after three times, code another **I**. If the word does not re-occur, or only occurs twice, leave the word highlighted in yellow. Repeat the process until all words are highlighted .

8. Go back through each of the 100 utterances, looking for conversational device repetition (to be defined later). To do this, look within the text and highlight in pink any conversational expressions.

9. Go back through the pink conversational expressions and highlight in green any that are used three times. Once the expression has been used three times, code **C**. For any re-occurrences after that, code an additional **C**.

10. Look at your coded transcript again, focusing again on your elemental columns. This time, consider synonyms (e.g., does "bad," "awful," and "terrible" continue to come up?; or "pampers" and "diaper"?); categories (e.g., do you see "T-Rex," "Velociraptor," and "Dinosaur"; "wrench," "screwdriver," and "pliars"; "doctor," "hospital," "nurse"?); and themes (e.g., "smooshing" may be one element in the theme/idea of harming, destroying, damaging, or ruining; or a theme may consist of "quitting," "stopping," or otherwise taking a "break"). Modify your codes as appropriate (treating every synonym or category member as the same "element"). You do not always need to follow this rule: Category members may come up quite naturally (and represent quite good topic maintenance) and would not be coded as **I**.

    Note: "baby", "brother", "dad", etc. would not count as members of a category (they are figures commonly used together during administration of the ADOS).

    Note: synonyms in a row would not count towards **U**.

11. After coding **I** in this very objective way, go back through the utterances coded as **I**. Change all **I** codes to **IQ** (ideational perseveration – questionable) if consideration of the interaction as a whole makes it fairly obvious that these repetitions were not excessive or otherwise odd (see ideational perseveration section for more explicit criteria).

12. Go back through the transcript again, subdividing **U** and **C** codes into their respective sub-level codes

13. Finally, verify your codes while watching the video. Pay particular attention to 1) pronouns whose referents are clear while watching the video, and 2) whether or not **U** was coded because the child was repeating himself to repair a communicative breakdown (i.e., the examiner wasn't answering him). *Since this is the first time you are aware of the child's diagnosis, do not change any codes unless you are certain that watching the video was instrumental in your coding changes (this keeps you from being biased by a child's diagnosis).*

Utterance-Level Perseveration

General Definition:

Utterance perseveration is present when words, phrases, dependent clauses, and entire utterances are said more than once (immediately after the first time they are uttered), are basically the same in structure, and do not add novel semantic content. For example, the child says, "After the movie, after the movie we went for ice cream" or "They must have fainted too. They must have fainted." (Example from Murphy & Abbeduto, 2007)

Specific Rules:

1. The child must say the word, phrase, dependent clause, or entire utterance in immediate succession without interruption by the examiner for it to be **U.**

2. The child only needs to say the word, phrase, dependent clause, or entire utterance twice for it to be **U**. **U** can be coded given just one utterance (e.g., "I don't I don't know" or given two utterances (e.g., "I don't know. I don't know that."). In the latter case the code **U** would be entered next to the second utterance.

3. Do not count the repetition of only a sound/phoneme as **U**.

4. Allow for minor alterations in an utterance. Whatever two units you are considering as a repetition can differ by only one morpheme (word or bound morpheme). For the example, "I don't that I don't know anything," the 2 units you are considering are underlined and differ only by the word *that.*

5. Repetitive utterances are not coded as **U** if the child repeats himself to repair a communicative breakdown (e.g., if the examiner does not respond to the child within 3 seconds of the first utterance).

6. Sound effects said in succession do not count as **U** (but will be entered once into an element column).

7. **U** is not coded if the child repeats himself for emphasis or in any otherwise appropriate fashion (e.g., "I really really like ice cream.").

8. Do not code **U** if the child is clarifying something he said. For example, the child might say "Can I pull this thingy? This string thingy?" Also note that the second "thingy" would be highlighted red in the element column, so not to be considered when coding ideational perseveration.

Two levels of **U** based on number of repetitions

1. If a unit of language is repeated once (i.e., 2 successive productions), code **U2**

2. If a unit of language is repeated two or more times (i.e., 3 or more successive productions), code **U3**

***** If an entire utterance is repeated two or more times, use both codes. For example:

I want the ball
I want the ball (code U2)
I want the ball (code U3)

Additional Examples:

CHILD: I usually go I usually leave
…
CHILD: and and and that's it

…
CHILD: no no no the the pool's closed
…
CHILD: I usually go I usually go go home

Ideational Perseveration

General Definition:

Ideational perseveration occurs when ideas are repeated excessively (for example, The tester says, 'You can't think of any time you went on vacation?' and the participant responds, 'Out of North Carolina, no…I never went out of North Carolina…I have never gone out of home, out of North Carolina…Everything's in Carolina…' – Example from Murphy & Abbeduto (2007)). Ideational perseveration does not have to be produced in immediate succession; perseverative ideas can be reintroduced later in the interaction.

"Topic" perseveration not just about topics:

Although "topic" perseveration is a traditional code of fragile X researchers, "topic" is an elusive concept. Instead, we will examine "conceptual elements" of utterances. These are the words of a sentence once they are separated into: subject, verb, object, and adjective/adverb (or pronouns that refer to them).

Basic Rules:

1. Code an utterance with a **I** on the third time that a word in your element columns is repeated (if the word is *not* highlighted in red) and code an additional **I** for each occurrence after that.

2. If you see variations of a word that have the same root or meaning, consider these variations to be repetitions of that word (e.g., "mom" and "momma"; "ride" and "riding")

3. Synonyms and near-synonyms will count as the same element (e.g., "wash" and "clean"; see the previous section on "coding steps" for further instructions).

4. If you detect utterance perseveration, remember to include the word/s that contribute to the code of utterance perseveration only one time in the elemental columns (e.g., for "blue, blue" *blue* would be entered once in the columns).

When and when not to assign words to an elemental column:

1. If the child uses pronouns but you can determine what the pronoun is modifying, include a word.

2. If a word is said more than once in the same utterance but the utterance does not qualify as utterance perseveration, you can place the element twice in the columns (and these 2 occurrences of the word can count towards a code of **I**).

3. When separating the sentences into their elements, do not include "insignificant" words (i.e., words that are not "meaty" enough, too common, auxiliary verbs, or in other words not "conceptual" elements). Examples:

   you, I, me, down, who, what, where, when, why, how, thing, been, so, gonna, too, do, that, this, it, goes, go, get, up, wanna, come, on, all, did, got, something, don't, put, have, right, here, there, off, "look" or "watch" (if used as a directive), can, know, now, try, could, should, would, please, thank you, mmm, thanks, want, some

   Note: If any of the pronouns above refer to something, you can put their referent in the elemental columns (this *does not* include *you, I,* or *me*)

   Note: A child can potentially perseverate on *anything*, even on ideas represented with words like those above (the rule above is simply meant to save us time, because *typically* repetitive use of these words wouldn't lead to a code of perseveration). Be aware of this while coding.

<u>Excluding elements from consideration by highlighting in red:</u>

1. In general, after considering context, highlight in red (therefore eliminating from consideration) any words that are present because the child was offering completely obligatory information to a question, answering a request for clarification, requesting clarification, or using repetition for emphasis.

2. Scripted language *that is appropriate to the context of the ADOS* (e.g., "ready, set, go" or "one, two, three, go!" during balloon activity or when blowing out candles) should be highlighted in red.

3. Obligatory Elements:

   a. Completely obligatory example:

      Examiner: Who do you have?
      Child: Baby

      Because the response is completely obligatory, eliminate the word "baby" from consideration.

   b. Example of an answer that has both an obligatory and nonobligatory element:

      Examiner: What happened to the dog?
      Child: He flew away.

      He (refers to the dog) is obligatory because this same element was present in the examiner's question (so this would be highlighted in red in your

element column and not considered when coding **I**). However, you may still enter the verb "flew" in your element column.

4. Examples of when the child is asking for clarification:

    Examiner: My dog's name is Bailey.
    Child: Bailey?

    Examiner: It's crush?
    Child: Crush?
    Examiner: Yeah
    Child: Crush the soda?

    Anything the child says in response to the clarification requests, *what?, huh?, what did you say?, etc.* would be highlighted red and not counted when considering **I**. Answers to a *who* question would also be highlighted red *if* the question is a request for clarification.

5. Examples of other communicative breakdowns:

    Child: How you spin this?
    Examiner: Albert, I have –
    Child: How you spin this?

    Elements from the second child utterance become highlighted in red because the examiner didn't respond to his question the first time.

    Child: Love them.
    Examiner: Lava? (she didn't understand him)
    Child: I love them more and more.

    "Love" and "them [it's referent]" would be highlighted in red in the element columns. More would be entered in the element columns and *not* highlighted. Note: more and more would not be considered **U** because more was repeated for emphasis.

    Examiner: (singing)
    Child: Stop!
    Examiner: (singing)
    Child: Stop!
    Examiner: (singing)
    Child: Stop!

    The second two stops will be highlighted in red. The examiner does not acknowledge the child. However, if examiner first acknowledged the child

and continued singing (e.g., "Hold on"), additional "stops" would not be highlighted in red.

5. The element "baby" is always highlighted in red *when spoken during the "Birthday Party" activity*. Unlike in free play where there are a variety of things to talk about, the baby is central to this whole section of the ADOS and will keep coming up very appropriately.

Changing a Code of **I** to a code of **IQ** (Ideational perseveration – Questionable):

* In this step, we identify those repetitions already coded as **I** which are *really* perseverative (the codes for those that aren't considered excessive from a more subjective perspective are changed to **IQ**)

Subjective Criteria::

1. If the examiner seems to want to move on to something else, keep as **I**.

2. If elements are spread apart and it wouldn't feel to the communication partner that it was excessive, change to **IQ.**

3. If elements are spread apart but it is a very odd topic (e.g., "Ziki butt,") keep as **I.**

4. If the elements are in very close proximity, and doesn't feel like good topic maintenance, keep as **I.**

5. When the child makes the same exact point or offers the same exact information, keep as **I.**

6. If the idea comes up abruptly (i.e., tangential or noncontingent), keep as **I**.

Default Rules:

1. If the child appears to be perseverating on one of the family figures, or other ADOS toys, and you are "on the fence," change to **IQ**.

2. If you're on the fence about the first **I** given for an element, code as **IQ**.

3. If you've already coded something as **I** and you're on the fence again, code as **I** again.

4. If you already changed something to **IQ** for that element previously, and you're on the fence again, keep as **I**.

Additional Examples:

CHILD: I wanna go play with that car right now.
…
EXAMINER: Well, while these are out I have a couple of questions for you.
CHILD: How about play with the car now?
…
CHILD: I'll go get the car.
CHILD: When are you gonna get the car?
CHILD: We need to go get it.

Conversational Device Repetition

General Definition:

Conversational devices are rote sayings and phrases (e.g., "right on," "That's about it") that serve to control the interactional flow but do not add content to the topic (Murphy & Abbeduto, 2007).

Specific Rules:

1. We will NOT consider "umm" or "uh" as a conversational device.

2. If a child repeats a conversational device three times or more within the interaction, consider it conversational device repetition. Code with a **C** on the third time and code an additional **C** for each occurrence after that.

3. Some phrases that you may code as conversational devices include: "cool," "good deal," "uh huh," "mhm," "oh no," "OK," "OK good", "yeah," "wow, " "mmm," "well," "uh," "c'mon," (see below for exceptions)

4. Some words and phrases that won't count as **C**: "thank you," "please,"

5. There are exceptions to coding some conversational phrases as **C**. These include:

    a) If a phrase like "uh huh," "mhm," or "yeah" is in response to a yes/no question, it is an obligatory response and should not be considered.

    b) Do not consider "OK" as a conversational device if it is in response to the question "OK?" or something similarly obligatory.

    c) Do not consider "OK" as a conversational device if it is describing a condition or state, as in "I'm OK."

    d) Do not count phrases as conversational devices if the child is using them to request clarification, as in "huh?".

6. Conversational devices do not have to be in isolation. Sometimes conversational devices will begin a longer utterance. For example, a child might begin multiple utterances with "mmm…" - "Mmm okay…" "Mmm I don't know…" etc. In these

instances, the utterances *themselves* may actually be obligatory but we could consider them for **C** because the conversational devices were not obligatory.

Two levels of conversational device repetition: "vocal" vs. "real word" devices:

1. Vocal examples (code as **CV**): "hmm;" "mhm;" "ahh"

2. Real word examples (code as **CW**): "good deal;" "right on;" "cool" (may be a "habitual perseverative response")

Even more sub-levels of conversational device repetition: "stand-alone" vs. devices that begin longer utterances:

1. In the "conversational device" column, *go back* and code:

   a. **CVO** or **CWO**: The conversational device stands alone, meaning that it constitutes an entire utterance by itself (e.g., "OK")

   b. **CVB** or **CWB**: The conversational device begins a longer utterance (e.g., "OK that's what I meant")

Q Coding

Use this code for self-repetitive utterances (utterance-level, ideational, or conversational device) that occur after an examiner's "conceptual question" (conceptual questions are basically "difficult," put the child "on the spot," and require – for example – the child to explain something, give an opinion, or otherwise give important information).

1. In the "X" column, code **Q** next to an utterance coded as perseverative if the immediately preceding utterance includes the examiner asking a conceptual question.

Appendix B

Coding Manual for Perseveration and Loss-of-Set (Verbal Fluency Context)

Code Definitions

* If a slash (/) is used, this means that codes are entered in different columns in the excel document

Perseveration

Stuck-in-set perseveration **(S)**: the child's first response on a task is appropriate for a preceding task (e.g., saying "cow" when instructed to generate food or drink items because the previous task requested the child to name animals). If after the first response, the child continues to name items appropriate for the preceding task, this would also be coded for this category of perseveration. If the child names an animal *after* naming a food/drink item, this would be coded as *loss-of-set* as described below.

Recurrent perseveration **(R)**: the child offers a response within a task that was already given for that task, but not as the immediately preceding response (e.g., "*cow*, dog, monkey, *cow*, cat").

Continuous perseveration **(C)**: the child gives the same response twice or more in a row within the same task (e.g., "cow, *dog*, *dog*, *dog*, monkey, cat").

* If examiner happens to prompt the child (e.g., "what else?") and the child repeats his last response, this is coded as **R**, not **C**

Loss-of-set

Proactive Interference loss-of-set **(L/PI** *in the X column*)*:* the child starts to name food/drink items and then returns to animal names.

Tangential loss-of-set **(L/TG** *in the X column*)*:* the child gives a response or responses that would not be appropriate even for the preceding category. These responses will relate loosely to the semantic category but will not be appropriate for the category (e.g., when asked to name animals, the child responds "Mrs. Harris. Teacher. Bus driver. **Bus.")** (note that only the bolded response counts as "loss-of-set" because people are considered animals).

Unrelated loss-of-set **(L/N** *in the X column*): the child says something that is not at all related, even loosely, to either of the verbal fluency tasks (e.g., "computer" or "White House!").

If loss-of-set response also counts as recurrent perseveration code as **PIR, TGR, or NR**

Example coding

Add "**X**" to code column if response coded as perseveration or loss-of-set is an example item (i.e., cat, dog, pizza, milk)

Connected Speech

(CS): Child produces an utterance (i.e., does not simply name an item which is what the task actually requires)

Examiner Prompts

Code **E** if examiner gives a specific prompt (to count as *specific* must refer to the particular "set")

In X column, also code **P** if the examiner's prompt has to do with the child's perseveration or loss-of-set (don't enter anything if not about perseveration or loss-of-set)

Other Rules

- Always begin coding AFTER what seems to be the examiner's last bit of "direction-giving"
- Repairs: Do not code perseveration if child repeats himself as a response to a request for clarification
- "Cat Dog Cat Dog" (Not understanding the task)
    - If the child begins by simply repeating cat and dog and the examiner prompts for something "different" (or otherwise makes clear that the child needs to name *different* animals), act like those responses of cat and dog never happened if the child then appears to understand the task

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