Symposium Title: Speech Intelligibility in Fragile X Syndrome

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Overview: Moderate to severe delays have been reported in all aspects of language development in fragile X syndrome (FXS), including expressive and receptive vocabulary, syntax, and pragmatics (Fryns, Jacobs, Kleczkowska, & Van den Berghe, 1984; Newell, Sanborn, & Hagerman, 1983; Roberts, Mirrett, & Burchinal, 2001; Sudhalter, Scarborough, & Cohen, 1991). However, research on the speech production of individuals with FXS is more limited, consisting primarily of case reports (Madison, George, & Moeschler, 1986; Paul, Cohen, Breg, Watson, & Herman, 1984). Previously, the speech of children with FXS has been described as "odd" or "delayed" or showing features of cluttering, which may include rapid rate, abnormal prosody, or telescoping or condensing of words (Hanson, Jackson, Hagerman, Opitz, & Reynolds, 1986). Speech intelligibility is a critical aspect of expressive communication, defined as "the capability of the speaker to produce an understandable spoken message" (Kent, 1993, p. 224).

In studies of speech disorders, intelligibility is measured either objectively or subjectively. For subjective measures, listeners quantify their perception of a speaker's intelligibility by rating what they heard (Darley, Aronson, & Brown, 1969; Weismer & Laures, 2002). Objective measures involve transcription (typically orthographic) or forced-choice recognition of words by listeners, yielding a percentage of words or utterances identified correctly relative to the target that the speaker intended to produce (Yorkston & Beukelman, 1978). In one study that used an objective measure of connected speech intelligibility, the percentage of intelligible words, boys with FXS showed significantly reduced speech intelligibility compared to nonverbal mental-aged matched typically developing (TD) peers, even though these groups had similar scores on measures of phonological accuracy (Barnes et al., 2009). Despite these reports, there are currently no treatments shown to improve speech intelligibility in individuals with FXS.

The first presentation will explain how perceptual ratings of features such as speech rate and intonation distinguish the speech of individuals in FXS from individuals with Down syndrome and how these features contribute to reductions in intelligibility in FXS. The second presentation will investigate how speech intelligibility can change as a result of a parent-implemented language intervention. The third presentation will examine the effect of partner familiarity on a measure of speech intelligibility in narrative language samples. Collectively, these presentations highlight an important aspect of communication in FXS, with clinical implications for identifying speech-language intervention targets in this population.

References:


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Paper 1 of 3

Paper Title: Cross-Sectional Comparison of Speech Intelligibility and Features of Speech Motor Dysfunction in Boys with Fragile X Syndrome and Down Syndrome

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Introduction: Fragile X syndrome (FXS) and Down syndrome (DS) are the two most common genetic disorders that cause intellectual disability. Although the cognitive and linguistic phenotypes of these disorders have been described, speech production in FXS remains relatively unexplored. In one study by Barnes and colleagues (2009), boys with FXS did not differ from nonverbal mental age-matched typically developing (TD) boys on phonological accuracy and phonological processes but produced fewer intelligible words than TD boys. Finding of decreased intelligibility in FXS relative to nonverbal mental age-matched TD peers despite similar scores on all measures of phonological accuracy suggests that the reduced speech intelligibility in children with FXS may not be due to differences in phonological accuracy, but rather to factors such as prosody (rate, intonation) and fluency. Because phonological accuracy cannot fully explain the reduced intelligibility in FXS, it is important to assess other factors such as rate, fluency, and prosody in this population. In this study we hypothesized that males with FXS will present with a profile of speech features that are different from males with DS who are known to have poor intelligibility. By using perceptual ratings derived from the speech motor scale of the Predictive Cluttering Inventory (PCI) and comparing ratings between FXS and DS, we provide a preliminary description of unique features that characterize the speech of males with FXS and evaluate whether these speech features contribute to intelligibility.

Methods: Participants were 31 males with DS (M age = 14.1, range = 7;7 - 23;8) and 31 males with FXS (M age = 15.3, range = 6;1 - 23;0). Consistent with previous research, males with FXS had a significantly higher mean percent of consonants correct on the Goldman Fristoe Test of Articulation (M = 87.33, SD = 14.1) than males with DS (M = 75.24, SD = 15.22; t = 3.25, df = 59.64, p = 0.002). Expressive language samples were collected from a narrative task in which an examiner followed a standardized script of limited prompts to guide the participant in re-telling a story from a wordless picture book. Narrative samples were transcribed and analyzed using SALT (Systematic Analysis of Language Transcripts; Miller & Iglesias, 2012). The average speech intelligibility in connected speech, calculated as the percent of utterances that were complete and intelligible for males with DS was 76.3% (SD = 19.6) and for males with FXS was 79.4% (SD = 20.1). Results of a t-test revealed that groups did not differ significantly on speech intelligibility (t = 0.61, df = 60, p = 0.54). The presence/absence of 10 speech motor features from the PCI were then rated from audio recordings of the narrative language sample. The speech motor features of the PCI were: 1) articulation errors, 2) irregular speech rate, 3) telescopes or condenses words, 4) rapid rate, 5) rate progressively increases, 6) irregular melody or

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Results: Overall, a higher number of males with FXS were rated as having features from the speech motor domain of PCI present than males with DS on 8 out of 10 of features (irregular speech rate, telescopes or condenses words, rapid rate, rate progressively increases, irregular melody or stress pattern, initial loud voice trailing off to unintelligible murmur, lack of pauses between words and phrases, repetition of multisyllabic words and phrases), suggesting a unique speech profile for FXS. As expected, given articulation scores on the GFTA, articulation errors were present in the majority of participants in both groups (DS = 87%, FXS = 84%). 10 of 31 (32%) males with DS presented with 3 or more PCI speech features, whereas 16 of 31 males with FXS had 3 or more PCI speech features (52%). Results of a t-test revealed that males with FXS were rated as having significantly more PCI speech features than males with DS ($t = 2.40$, $df = 60$, $p = 0.02$). Although speech intelligibility was not significantly different between the two groups, results of a linear regression model indicated that the total number of speech features present had a significantly negative impact on the percent of intelligible utterances for both groups ($F (1, 60) = 4.03$, $p = 0.049$).

Discussion: Results confirm with objective empirical data previous descriptions from case reports of the presence of abnormal speech features in males with FXS and provide evidence that, in addition to reduced articulatory accuracy, features such as abnormal rate and prosody contribute negatively to speech intelligibility (Hanson, Jackson, Hagerman, Opitz, & Reynolds, 1986; Madison, George, & Moeschler, 1986; Paul, Cohen, Breg, Watson, & Herman, 1984). Clinical implications of these results highlight the need for thorough speech assessment for males with FXS that includes evaluation of rate and prosodic features, in addition to articulation/phonology and speech intelligibility. Future research is needed to quantify the range of severity of abnormal features, beyond determining the presence or absence of particular features, to advance understanding of speech profiles in FXS and develop targeted speech interventions for this population.

References:
FXS have shown that the participating youth demonstrated improvements in their expressive skills, particularly expressive lexical diversity (McDuffie et al., 2016, 2018). We hypothesized that a positive change in expressive language as a result of participation in a PILI may also lead to improvements in speech intelligibility in older school-aged children and adolescents with FXS.

**Method:** The current study included 28 parent-child dyads who participated in a double-blind placebo-controlled study of lovastatin and PILI in which all participating families dyads received PILI. Youth participants were 26 males and two females ($M$ age at visit $1 = 13.53, SD = 2.40$, range = $10.09 - 17.77$). In this 12-week PILI, parents were taught to use four verbally responsive language support strategies (i.e., models of story related talking, recasts of child utterances, open-ended WH-questions, and intonation prompts) while interacting with their son or daughter in the context of shared storytelling using wordless picture books. At the pre-intervention assessment visit, youth participants completed multiple standardized assessments, including the Leiter-R ($M$ nonverbal IQ = 44, $SD = 7.70$), PPVT ($M$ age equivalent = 5.94, $SD = 2.29$), EVT ($M$ age equivalent = 5.93, $SD = 2.10$), and CASL-Syntax Construction subtest ($M$ age equivalent = 3.99, $SD = 1.77$). Furthermore, following the pre-intervention assessment visit and prior to their participation in the intervention, parent-child dyads completed three shared storytelling interactions using wordless picture books that were uploaded to an iPad. Parent-child dyads completed three additional shared storytelling interactions with different picture books after completing their participation in the intervention and prior to their post-intervention assessment visit to the clinic. These samples were recorded in the home via Skype and transcribed by highly trained research staff using SALT (Systematic Analysis of Language Transcripts; Miller & Iglesias, 2012). The dependent variable in the current study, percentage of intelligible words, was derived from these parent-child narrative language samples. Wilcoxon Signed-ranks tests were used to examine whether there was a significant difference in the percentage of intelligible words in the parent-child language samples from pre-intervention to post-intervention.

**Results:** First, we examined if there was a significant difference in intelligibility in words from pre-intervention to post-intervention in the entire sample of participants ($n = 28$). Descriptive summaries revealed that the median percentage of intelligible words produced by the child at pre-intervention was 84.0% (min = 50%, max = 97%). At post-intervention, the median percentage of intelligible words produced was 88.5% (min = 56%, max = 98%). Results of a Wilcoxon Signed-ranks test indicated a significant improvement in percentage of intelligible words ($Z = -2.385, p = 0.017, r = 0.45$) from pre- to post-intervention. Given the variability in mean length of utterance in morphemes (MLUm) within the sample, we examined whether there was a difference in percentage of intelligible words as a function of MLUm. We found a significant improvement in percentage of intelligible words for children who had an MLUm < 2 at baseline ($Z = -2.357, p = 0.018, r = 0.59$), but no significant improvement in percentage of intelligible words for children who had an MLUm ≥ 2 at baseline ($Z = -0.714, p = 0.475, r = 0.21$).

**Discussion:** This study is the first to demonstrate that participation in a PILI that targets improvements in spoken language also leads to increases in speech intelligibility for the participating youth with FXS. This improvement was only observed in the youth who had more limited syntactic complexity (i.e., MLUm < 2) at the pre-intervention assessment visit. We suspect that the observed increases in speech intelligibility may be due to the fact that the participating youth were spending a significant amount of time each week interacting with their parents in shared storytelling activities. During these activities, parents were taught to use clear story-related language models and to prompt their children to talk about the story (using wh-questions and intonation prompts), thereby providing the children with multiple opportunities to practice their speech and language skills in a semi-structured and supportive activity. These results suggest that future studies of language interventions in FXS, or other neurodevelopmental disabilities that involve co-occurring speech impairments, should investigate whether gains in language skills correspond with improvements in speech production.

**References:**


**Paper 3 of 3**

**Paper Title:** Measurement of Speech Intelligibility: Variation Depending on Partner Familiarity

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**Introduction:** Language sampling is often used as an assessment measure to describe expressive language skills, including speech intelligibility, of children and adolescents. However, the language produced by the participant during the language sampling assessment may be influenced by variations in the context in which the sample was taken, such as setting, materials, and partner. Previous studies have reported on the effects of language sampling task (conversation or narration) on speech intelligibility in males with fragile X syndrome (FXS; Kover, McDuffie, Abbeduto, & Brown, 2012); however, no study has described the effects of partner familiarity on speech intelligibility in this population. Previous studies have measured child speech intelligibility in children with delayed/impaired speech in a variety of contexts, including parents as raters of speech intelligibility (Flipsen, 1995), intelligibility scores derived from parent-child language samples (Hustad, Allison, McFadd, & Riehle, 2014), and intelligibility scores derived from examiner-child interactions (Binger, Ragsdale, & Bustos, 2016). However, little is known about the effects of partner familiarity on speech intelligibility within populations of children with delayed/impaired speech, and as we have highlighted previously, speech intelligibility in FXS has received limited attention. To address these gaps, the current study examined whether there are differences in youth speech intelligibility in parent-child and examiner-child narrative language samples. Given that parents are their child’s most familiar communication partner, we anticipate that parent behavior during the interaction, such as glossing or recasting the child’s utterances, would provide richer context within the language sample for interpreting the child’s speech productions. Therefore, we hypothesize that the youth’s speech will be more intelligible in the parent-child language samples compared to the examiner-child language sample.

**Methods:** Nineteen mother-child dyads who participated in a parent-implemented language intervention (PILI) described in McDuffie et al. (2018) were included in this study. Youth participants were 19 males with FXS between the ages of 10 and 17 years. The dependent variable in this study, percentage of intelligible words, was collected prior to and following each dyad’s participation in a 12-week, distance-delivered PILI described in the previous symposium study. Parent-child dyads completed a shared storytelling task using an unfamiliar wordless picture book (‘Frog, Where Are You?’ or ‘One Frog Too Many’) at each visit to the clinic. Parents were able to review the story before engaging with their son but received minimal instructions on how to go through the book. In addition, each child completed a narrative task with an unfamiliar examiner at these visits. Examiners were trained to providing minimal prompting and scaffolding for the youth. During this narrative task, the youth paged through an unfamiliar wordless picture book (‘Frog Goes to Dinner’ or ‘Frog on His Own’) two times: once to quietly review the story, and again to talk about the story with minimal prompting from the examiner. Each parent-child and examiner-child interaction was audio-recorded and transcribed using SALT (Systematic Analysis of Language Transcripts; Miller & Iglesias, 2012). Wilcoxon Signed-ranks tests were used to examine whether there was a significant difference in the percentage of intelligible words in the parent-child and examiner-child language samples at the pre- and post-intervention assessment visits.

**Results:** First, we examined whether there were differences in the percentage of intelligible words produced by the child in the parent-child and examiner-child narrative language samples obtained at the pre-intervention assessment visit. Descriptive results for the parent-child narrative language samples revealed the median percentage of intelligible words produced by the child was 95.44% (min = 86.75, max = 99.03) at pre-intervention and 93.85% (min = 67.14, max = 98.93) at post-intervention. Descriptive results for the examiner-child narrative language samples revealed the median percentage of intelligible words produced by the child was 95.92% (min = 74.80, max = 99.57) at pre-intervention and 93.53% (min = 72.17, max = 99.47) at post-intervention. Results of a Wilcoxon Signed-ranks test indicated that there was no difference in the percentage of intelligible words based on the partner ($Z = -0.322, p = 0.748, r = 0.07$). Then, we examined whether there were differences in the percentage of intelligible words produced by the child in the parent-child and examiner-child narrative language samples obtained at the post-intervention assessment visit. Results of a Wilcoxon Signed-ranks again indicated that there was no difference in the percentage of intelligible words based on the partner ($Z = -0.644, p = 0.52, r = 0.15$).
Discussion: We found that there were no differences in child speech intelligibility based on partner familiarity at either of the assessment visits. Given the small sample size and the relatively high speech intelligibility observed in the sample, it is possible that partner-dependent differences would be observed in a sample that includes children with poorer speech intelligibility. However, the results of this study provide preliminary evidence that partner familiarity does not affect the measurement of speech intelligibility in narrative language samples with individuals with FXS, which supports the growing body of work looking at the feasibility and validity of parent-administered assessments and conducting assessments in the home using distance technology. Being able to participate in research or receive services from home via distance technology is particularly important for low-incidence populations such as FXS who may not live close to research facilities and clinics. Future studies should examine differences between the use parent-facilitated language samples obtained in the home using video-teleconferencing technology and those obtained in the clinic using trained examiners.

References: