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Reading in children with fragile X syndrome: Phonological awareness and feasibility of intervention

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**Reading in Children with Fragile X Syndrome: Phonological Awareness and Feasibility of  
Intervention**

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### **Abstract**

Individuals with FXS present with significant deficits in reading skills, but scant research exists to understand the characteristics of the reading delays or best practices for reading instruction with this population. Study 1 examined the relationship between phonological awareness and reading skills in individuals with FXS. Study 2 evaluated the feasibility of a web-based reading intervention, which incorporated phonological awareness and phonics instruction but was originally developed for mainstream students, for children with FXS. Results suggest that phonological awareness and reading skills are correlated in this population, and that instruction targeting phonological awareness and phonics should not be ruled out for individuals with FXS. Further studies are needed to examine their potential effects.

## Introduction

Fragile X syndrome (FXS) is a genetic disorder caused by an expansion of cytosine-guanine-guanine nucleotide sequence on the X-linked *Fragile X Mental Retardation 1 (FMR1)* gene (Santoro, Bray, & Warren, 2012) that affects approximately 1 in 2,500 to 1 in 6,000 males and 1 in 8,000 females (Fernandez-Carvajal et al., 2009). The *FMR1* gene produces fragile X mental retardation protein (FMRP), a protein crucial for brain development that is reduced in individuals with FXS, resulting in cognitive impairment (Darnell & Richter, 2012). FXS is the most common known cause of inherited intellectual disability, with approximately 90% of males and 50% of females with FXS presenting with these impairments (Hessl et al., 2009).

FXS occurs in both sexes, but males tend to show more severe impairments than females. Depending on the amount of FMRP produced, females may present as typical or similar to their male counterparts (Gallagher & Hallahan, 2012). The severity varies across individuals, but clinical characteristics of FXS often include developmental delays, learning challenges, social and behavioral problems, anxiety, executive function difficulties, and language and communication impairments (Gallagher & Hallahan, 2012). Two of the most common co-morbid conditions associated with FXS include intellectual disability (ID), which occurs in nearly all males with FXS (Bailey, Raspa, Holiday, Bishop, & Olmsted, 2009) and autism spectrum disorder (ASD) in about 60% of males with FXS (Harris et al., 2008; Klusek, Martin, & Losh, 2014).

### **Literacy Skills of Individuals with FXS**

Relatively little is known about literacy skills and reading trajectories in individuals with FXS. However, it is clear that the reading skills of males with FXS are delayed relative to chronological-age expectations. A large national survey of families of individuals with FXS

documented that less than half of adult males with FXS (44%) were able to read basic picture books and only 59% knew letter sounds (Bailey et al., 2009). Increased knowledge regarding reading and literacy skills in males with FXS is critical for a number of reasons. First, poor reading skills can negatively impact academic and social success (e.g., Chapman, Tunmer, & Prochnow, 2000; Miles & Stipek, 2006). Second, reading and literacy are clearly associated with improved adult outcomes in both the general population and in clinical groups such as ASD (Kutner et al., 2007; LeBlanc, Riley, & Goldsmith, 2008). Only 10% of adult males with FXS are able to live independently, and 95% need assistance with daily living tasks (Hartley et al., 2011). Increased literacy skills in FXS would likely contribute to greater independence and improved quality of life associated with greater educational attainment, better employment options, and increased social and leisure opportunities. Third, given the high association with ASD and the presence of ID in nearly all males with FXS, research on literacy skills in FXS can contribute to our understanding ASD and ID and to special education and early intervention fields more broadly.

Despite documented literacy impairments in FXS and the academic and functional significance of such delays, best practices for literacy intervention for individuals with FXS are unclear (e.g., Brazendale, Adlof, Klusek, & Roberts, 2015). A primary question is whether individuals with FXS benefit from the same methods of instruction as children with typical development, and specifically, whether or not they can learn to read from phonologically based reading instruction, which is currently considered to be best practice for teaching typically developing children to read (National Early Literacy Panel, 2009; Ryder, Tunmer, & Greaney, 2008).

Phonological awareness refers to the ability to focus on and manipulate the sounds in spoken language, whereas phonics refers to relationships between letters and sounds (Scarborough & Brady, 2002). In typically developing children, early phonological awareness skills predict later reading abilities (Melby-Lervåg, Lyster, & Hulme, 2012), and treatment studies indicate a causal relationship (National Early Literacy Panel, 2009; National Reading Panel, 2000). It is believed that having explicit awareness of the sounds in words makes it easier for beginning readers to learn to associate alphabet letters with corresponding sounds in reading and spelling. There is substantial evidence that explicit, systematic phonological awareness and phonics instruction is highly effective in improving decoding and comprehension in typically developing children, as well as in those with learning disabilities, such as dyslexia (National Early Literacy Panel, 2009; National Reading Panel, 2000; Ryder et al., 2008).

Due to the lack of research on reading instruction in FXS, clinical guidelines have been largely based on descriptive studies of the FXS profile and on expert opinions. Recent clinical guidelines recommend that a whole-word approach be used when teaching literacy and reading to individuals with FXS, as opposed to phonologically based methods (Braden, Riley, Zoladz, Howell, & Berry-Kravis, 2013; Braden, 2000, 2002; National Fragile X Foundation Education Project, 2012). These guidelines are based on a report that children with FXS display weakness in sequential processing skills relative to simultaneous processing (Hodapp et al., 1992). Given this cognitive profile, it has been assumed that individuals with FXS will be better able to process information as a whole rather than as separate parts (National Fragile X Foundation Education Project, 2012); however, a link between poor simultaneous processing and impaired phonetic decoding in FXS or other populations has not been directly studied. Moreover the original findings regarding the difficulty of simultaneous and sequential information in FXS is

not seen in all tasks and many not extend to the auditory domain (Dykens, Hidapp, & Leckman, 1994). Whole-word approaches teach children to read by recognizing and memorizing words as visual wholes, without taking into account the individual sounds of each letter (Lemons & Fuchs, 2010). Some studies have successfully implemented whole word instruction as an initial step in reading for individuals with Down syndrome (Byrne, MacDonald, & Buckley, 2002). Although whole-word approaches can be effective for teaching children to recognize a limited set of taught words, little evidence exists for generalization to untaught words or that whole-word approaches lead to improved reading comprehension (Connor, Alberto, Compton, & O'Connor, 2014).

To date, no empirical studies of literacy interventions in children with FXS have been published. However, a few recent studies have examined literacy profiles and associations between phonological awareness and reading skills in children with FXS. One report found that boys with FXS performed comparably or superior to younger typical boys matched on nonverbal mental age on measures of word reading and passage comprehension (Klusek et al., 2015). These findings suggest that basic reading skills may be a strength for males with FXS relative to cognitive expectations, consistent with prior work by Johnson-Glenberg (2008). In contrast, Klusek et al. (2015) found that boys with FXS showed low phonological awareness compared to younger, mental-aged matched typical boys, suggesting that cognitive factors alone could not account for their poor phonological awareness. Further, Klusek et al. (2015) found that ASD symptom severity was significantly correlated with performance on the phonological awareness tasks, but not with word reading skills (Klusek et al., 2015).

A related study followed the same sample of boys longitudinally as Klusek et al. (2015). Results showed that despite overall poorer phonological awareness skills, boys with FXS acquired phonological awareness skills at a rate that was similar to their younger, mental-age-

matched peers after differences in nonverbal cognitive ability were controlled, with a plateau detected at about 10 years of age (Adlof, Klusek, Shinkareva, Robinson, & Roberts, 2015). Roberts et al. (2005) reported similar findings; namely, a plateau in letter/word recognition acquisition in boys with FXS at the same age, consistent with parent reports of increasing literacy skills through age 10 years (Bailey et al., 2009). Importantly, concurrent and predictive relationships between phonological awareness and reading have been documented in boys with FXS, suggesting that the relationship between these skills in FXS is similar to that reported in typical development (Adlof et al., 2015; Klusek et al., 2015).

### **Approaches to Literacy Instruction in Children with Other Developmental Disabilities**

Given the lack of treatment studies examining reading instruction for children with FXS, it is useful to consider evidence from intervention studies involving populations with other developmental disabilities that share some cognitive and behavioral characteristics with FXS, such as DS and idiopathic ID. Similar to FXS, early assumptions regarding reading instruction in these populations suggested that these individuals were visual learners who could not learn from phonics approaches alone, but instead would benefit from whole-word or sight-word instruction (Buckley, 1985; Cossu, Rossini, & Marshall, 1993). However, studies examining the reading skills of youth with DS and ID have found that phonological awareness ability is highly correlated with reading achievement in these groups, despite literacy profiles that include weaknesses in phonological decoding relative to word identification skills (Bird, Cleave, & McConnell, 2000; Cupples & Iacono, 2002). Moreover, numerous intervention studies provide evidence that explicit instruction in phonological awareness and phonics promotes reading acquisition in individuals with DS and ID, resulting in gains in letter-sound knowledge, recognition of taught words, novel word decoding, and word reading fluency (Allor, Mathes,



Roberts, Cheatham, & Al Otaiba, 2014; Browder, Ahlgrim-Delzell, Courtade, Gibbs, & Flowers, 2008; Burgoyne et al., 2012; Cologon, Cupples, & Wyver, 2011; Fredrick, Davis, Alberto, & Waugh, 2013). Collectively, these intervention studies have emphasized the importance of using intensive, systematic, visually appealing, repetitive, and multi-modal reading programs with a phonological component to see clinical gains. Several studies indicated that some individuals with ID and DS need a significant amount of time (e.g., a year or more) to display gains in reading skills (Allor et al, 2014; Fredrick et al., 2013), and therefore educators need to be sufficiently trained in goal-setting and monitoring progress to effectively implement these interventions.

### **The Present Studies**

There is a strong need for empirical evidence to guide educational decision-making with regard to reading for individuals with FXS. In this paper, we present two studies of reading development in FXS with the goal of informing educational decision making for this group. Study 1 evaluates concurrent relationships between phonological awareness skills and reading level in males with FXS during adolescence. In Study 2 the feasibility of a web-based reading intervention developed for typically developing children is evaluated with a group of individuals with FXS.

#### **Study 1**

Study 1 builds on emerging evidence of a relationship between phonological awareness and reading in children with FXS (Adlof et al., 2015; Klusek et al., 2015) through replication in an independent sample. Whereas prior work focused on children, this study focused on adolescents. Additionally, it used assessment tools that were designed to capture variation in lower-level skills, and therefore, may be less subject to floor effects. Very little is known about

reading development and its predictors in groups with ID during adolescence, despite evidence that individuals with ID can continue to make literacy gains during this development period (Roberts, Leko, & Wilkerson, 2013). Previous research has produced mixed results regarding the relationship between ASD symptom severity and reading-related measures in children with FXS (Adlof et al., 2015; Klusek et al., 2015; Nation et al., 2006). Therefore, Study 1 addressed the following research question: are phonological awareness skills associated with concurrent reading skills in adolescent males with FXS, after accounting for nonverbal cognitive ability and ASD symptom severity?

## **Method**

**Participants.** Participants included 26 adolescent males with FXS who were enrolled in a larger, longitudinal study of language development. All participants were native speakers of English and able to speak in a minimum of 2-3 word phrases at study entry, per parent report. The full mutation of fragile X was confirmed via medical record review. Recruitment primarily took place in the [details omitted for blind review] regions of the United States, through social media, family support groups, and the assistance of the [details omitted for blind review]. Demographic and descriptive information for the males is presented in Table 1.

**Procedures.** Assessments were administered by a trained researcher who had experience working with individuals with developmental disabilities. Assessments took place within a broader research protocol that spanned two days, including time for breaks. The Institutional Review Board (IRB) of the [details omitted for blind review] approved all procedures.

### **Assessments.**

***Nonverbal Cognitive Ability.*** The Brief IQ Composite of the Leiter International Performance Scale-Revised (Leiter-R; Roid & Miller, 1997) measured nonverbal cognition. This

measure has been used extensively in studies of children with FXS and other developmental disabilities (Skinner et al., 2005; Tsatsanis et al., 2003) and shows good reliability with Cronbach's alphas for the Brief IQ subtests ranging from .75-.88 in the norming sample. Growth Scale Values were used in analysis, which provide an equal interval conversion of the raw score that corrects for variability in item difficulty (Roid & Miller, 1997).

***ASD Symptom Severity.*** The Autism Diagnostic Observation Schedule-2 (ADOS-2; Lord et al., 2012) was administered and scored by examiners who were research reliable. The ADOS-2 involves a semi-structured play-based interaction, from which behaviors consistent with ASD are directly observed. Comparison Scores were used in the present study to index the severity of ASD symptoms given the strong association of ASD in FXS. Comparison Scores range from 0-10, with scores equal to or greater than 3 consistent with a diagnosis of ASD. Because ASD symptoms in adolescents with FXS are relatively stable over time (Hernandez et al., 2009), the larger study measured ASD at the initial assessment only. Thus, ADOS-2 scores were drawn from an assessment that occurred approximately one year prior to the literacy measures.

***Phonological Awareness.*** The Oral Language (Phonological Awareness) subtest of the Gates-MacGinitie Reading Tests- 4<sup>th</sup> Edition, Pre-Reading Level (MacGinitie, MacGinitie, Maria, & Dreyer, 2000) was used to measure phonological awareness. This assessment was chosen for the present study because it is developmentally appropriate for individuals with severe intellectual disabilities. The Phonological Awareness subtest includes 20 items testing the knowledge of the basic structure of spoken words, including knowledge of phoneme sequencing, matching, and segmentation. The Phonological Awareness subtest at the Pre-Reading Level has high internal consistency in the norming sample, with a Kuder-Richardson Formula 20 reported at .83 (MacGinitie et al., 2000). The range of scores observed in our sample of participants (see

Table 2) suggests that the Pre-Reading level was appropriately calibrated for our sample of participants. Raw scores were used in analyses. Given that the Gates-MacGinitie is designed for classroom-based testing, the administration was modified to apply to a one-on-one assessment setting: standardized prompts were addressed to the individual rather than the class as a whole and participants were instructed to point to responses rather than mark their responses in pencil. The Total Pre-Reading Extended Scale Score, representing each individual's position in relation to the entire range of achievement in grades K-12, was also computed as a group descriptive. This score includes four subtests: Literacy Concepts, Oral Language (Phonological Awareness), Letters and Letter-Sound Correspondences, and Listening Comprehension.

**Reading.** The Word Identification and Word Attack subtests of the Woodcock Reading Mastery Tests-III (WRMT-III; Woodcock, 2011) were administered to assess the ability to read aloud single words (Word Identification) and pronounceable pseudowords of increasing complexity (Word Attack). Analyses used Growth Scale Value scores, which estimate absolute performance on an equal-interval scale. The WRMT-III have been used in a number of prior studies of reading in individuals with other disabilities (e.g., Chen, Cordier, & Brown, 2013; Cullen, Alber-Morgan, Schnell, & Wheaton, 2014). Split-half reliability coefficients for the Word Attack and Word Identification subtests range from 0.71-0.93 and test-retest reliability ranged from 0.85-0.88 in the norming sample for the same age range of the present sample.

**Data analysis.** Analyses were conducted in SAS 9.4 (SAS Institute, 2013). The data were first examined for normality. Extreme floor effects were observed for the Word Attack subtest of the WRMT-III; 21 of the 26 participants obtained a raw score of "0". Given the limited variability in the scores, these data were only examined descriptively. Some floor effects were also observed for the Word Identification subtest, but to a lesser degree: 7 participants obtained a

raw score of “0”. No participants scored at the floor for the Phonological Awareness subtest of the Gates-MacGinitie (see Table 1 for descriptive statistics). To correct for positive skew, the Box Cox transformation (Box & Cox, 1964) was applied to the WRMT-III Word Identification scores ( $\lambda=.125$ ), Phonological Awareness scores of the Gates-MacGinitie ( $\lambda =0$ ), and the Leiter-R Brief IQ Growth Scale Value ( $\lambda =5$ ). The ADOS Comparison Scores were normally distributed.

Linear regression models were used to test phonological awareness as a predictor of word identification skill. First, maternal education level and household income (proxies for socioeconomic status) were examined in relation to the phonological and reading variables of interest. These variables were not correlated with the phonological awareness or reading measures in the present sample (all  $r$ 's < .16, all  $p$ 's > .423) and therefore, to maximize degrees of freedom, were not included as covariates in the final model. In the final model, nonverbal cognitive ability and ASD severity (indexed with the comparison score of the ADOS -2), were first entered in level 1, followed by the full model including the Phonological Awareness subtest. Examination of Cook's  $D$  (Cook, 1977) regression diagnostics revealed a single observation with a high influence on the regression results ( $D = 0.31$ ). Analyses were repeated excluding this point, with no change in the sign or significance of the coefficients for the predictor of interest. Notably, excluding this observation improved model fit ( $R^2$  increased from .46 to .62). Given that the observation was believed to represent a valid observation from the larger population, it was included in the presented analyses.

## **Results**

Descriptive statistics for the phonological awareness and reading measures are presented in Table 2. The relationship between phonological awareness performance and word attack skills

was examined descriptively: the mean phonological awareness score for the participants who obtained a raw score of “0” on the WRMT-II Word Attack subtest was 5.3 ( $n = 21$ ,  $SD = 1.85$ , range = 2-10). In contrast, the participants who scored above the floor on the Word Attack subtest obtained an average phonological awareness score of 11.4 ( $n = 5$ ,  $SD = 5.03$ , range = 5-19).

The model testing phonological awareness as a predictor of word identification skills was significant,  $F(3,22) = 6.33$ ,  $p = .003$ ,  $R^2 = .46$ . Phonological awareness scores explained significant variance in word identification scores, after controlling for nonverbal cognition and ASD symptom severity ( $R^2\Delta = 0.19$ ,  $p = .011$ ). Regression coefficients are provided in Table 3.

## **Discussion**

The results of Study 1 indicate a strong positive relationship between phonological awareness skills and concurrent oral word reading skills in adolescent males with FXS. This replicates the findings of previous studies that used different assessment tools with younger participants. Results indicated that, even in late adolescence, phonological awareness explained significant unique variance (19%) in word reading skills, beyond the effects of nonverbal cognition and ASD severity. These findings suggest that phonological awareness is an important component of reading in adolescents with FXS, as it is for typically developing children.

The pattern of generally low scores obtained by the adolescent participants on the various reading measures in this study also illustrates an important point with regard to the assessment of reading skills in this population. There is a shortage of standardized assessments of literacy-related skills in children with severe developmental disabilities. Our selection of measures was informed by previous research and by clinical experience, but we were unable to measure individual differences in pseudoword decoding skills due to extreme floor effects on the WRMT-

III Word Attack subtest. Descriptively, those participants who obtained a score above floor also had higher phonological awareness scores on the Gates-MacGinitie than those who scored at floor. However, without a measure that is sensitive to individual differences in decoding skills at the lowest end of the distribution, we are unable to evaluate whether phonological awareness uniquely predicts pseudoword decoding. This problem is not unique to the current study, and the need for valid measures of literacy skills that are sensitive to individual differences in children with severe developmental disabilities remains important to both research and clinical practice.

Overall, these results converge with existing work regarding the association between phonological awareness and reading acquisition in males with FXS. We note that the current sample was limited to males who had intellectual disability; additional research is needed to address whether results generalize to higher-functioning individuals or females with FXS. Furthermore, conclusions about the directionality of the relationship from this study are not possible given its concurrent correlational design, as well as a lack of information about participants' prior educational experiences, specifically regarding reading instruction and any prior interventions, including potentially pharmacological treatments, that may have been tried. In typical development, the relationship between phonological awareness and word reading skills is reciprocal, with phonological awareness initially predicting later word reading skills in younger children and a developmental shift for reading skills to then predict later phonological awareness skills at older ages (Hogan, Catts, & Little, 2005). Taken together, these findings point to the need for studies of causality in FXS to determine whether phonologically based instructional approaches lead to improvements in reading skills.

## **Study 2**

Despite known reading deficits in individuals with FXS, no published studies have evaluated methods of teaching reading to this population. Thus, the purpose of Study 2 was to test the feasibility of a web-based reading instruction program, originally developed for mainstream students, with a small group of individuals with FXS. We aimed to: (1) determine whether individuals with FXS can successfully complete a mainstream reading intervention that explicitly targets phonological awareness and phonics as well as other literacy skills, (2) examine factors related to successful program completion, and (3) determine whether the program showed evidence of potential effectiveness. Addressing these questions with a small sample would provide important data to guide future research on reading interventions with this population.

## **Method**

**Participants.** Participants included 8 individuals with FXS (7 males and 1 female) ranging in age from 7 to 23 years. All participants were able to communicate verbally with a minimum phrase length of 2-3 words. A diagnosis of the full mutation of fragile X was confirmed through medical records. Four participants were recruited through [details omitted for blind review], and four were recruited through [details omitted for blind review]. Recruitment was done primarily through the [details omitted for blind review]. This study was part of a larger study designed to evaluate the effectiveness of multiple reading interventions for participants with differing skill levels. Table 4 provides participants' demographic and descriptive information.

**Procedures.** Baseline and six-month assessments were administered by two trained speech-language pathologists with extensive experience working with individuals with developmental disabilities generally and FXS specifically. Assessments were completed in two to four hours, typically divided in two sessions. Following the documentation of baseline levels,



participants were enrolled in the web-based reading intervention and instructed to spend 15-20 minutes on the program four times per week at home or at school. Procedures were approved by the IRBs of [details omitted for blind review].

### **Assessments.**

***Cognitive Ability.*** The Stanford-Binet, Fifth Edition (SB5; (Roid, 2003) was used to document the participants' overall level of cognitive functioning. The SB5 is an individually administered assessment of intelligence and cognitive abilities, normed for individuals aged 2 through 85+ years, and it has been used frequently with the FXS population (Berry-Kravis, Sumis, Hervey, & Mathur, 2012; Leigh et al., 2013). The full battery provides a Full Scale IQ (consisting of all 10 subtests) as well as a Nonverbal IQ and a Verbal IQ, which have internal reliabilities ranging from .95 to .98 (Roid, 2003). Studies indicate that the broad abilities measured by the SB5, are predictive of academic achievement (Evans, Floyd, McGrew, & Leforgee, 2002; McGrew & Hessler, 1995).

***ASD Symptom Severity.*** Descriptive data on ASD symptom severity were available for seven participants. Scores from the ADOS-2 (described in Study 1) were available for six of the eight participants. Autism symptomology for one participant was assessed using the Social Responsivity Scale, Second Edition (SRS-2; Constantino & Gruber, 2012). This 65-item parental report instrument has strong psychometrics, with an internal reliability coefficient of .95. This scale examines social skills across five sub-areas including Social Cognition, Social Awareness, Social Communication, Social Motivation, and Restricted and Repetitive Behaviors. These combine to give an overall indicator of risk of ASD, ranging from within normal limits to severe. Overall, 7/7 (100%) of the participants for whom we had a measure of autism symptoms were above the threshold criteria suggesting an ASD diagnosis.

**Early Reading Skills.** The Test of Early Abilities-Third Edition (TERA-3; Reid, Hresko, & Hammill, 2000) was used to assess participants' reading skills. This assessment provides scaled scores in three areas, as well as an overall standard reading composite. The Alphabet subtest examines the participant's knowledge of general alphabet principles, including letter recognition, letter sounds, and syllables. The Conventions subtest measures overall print conventions, including spelling, capitalization, punctuation, and general familiarity with printed materials. The final subtest, Meaning, assesses the participant's ability to infer meaning from printed materials, including common logos as well as letters, sentences, and paragraphs. The TERA-3 has norms for children 3.5-8.5 years, and shows adequate reliability across those ages, with Cronbach's alpha ranging from .83-.91 across subtests. This test has been used in previous studies of children with ASD, where it was found to be positively correlated with nonverbal IQ, expressive language, and receptive language (Davidson & Weismer, 2014). It has also been found to be sensitive to change following an intensive reading intervention (Huffstetter, King, Onwuegbuzie, Schneider, & Powell-Smith, 2010).

**Reading Comprehension.** The Woodcock-Johnson-III Passage Comprehension subtest (WJ-III; Woodcock, McGrew, & Mather, 2001) was used to assess reading comprehension. This subtest required participants to silently read brief texts (i.e., sentences or paragraphs) of increasing difficulty and supply a missing word for each. The WJ-III is highly correlated with several other established reading measures, and normative data is available for ages 2 to 90 years. Internal consistency reliability for this subtest ranged from .78 to .96 across the ages.

**Intervention.** Participants were enrolled in the HeadSprout Early Reading Program (HERP). Several factors led the investigators to believe the HERP might be useful for individuals with FXS. First, the program explicitly targets five essential reading skills identified

by the National Reading Panel (2000), including phonemic awareness and phonics, as well as fluency, vocabulary, and comprehension. Second, the use of a computerized program capitalizes on the natural motivation that computers provide for individuals with FXS (Hall, Hustyi, Hammond, Hirt, & Reiss, 2014). Third, several studies of the HERP have provided evidence of effectiveness for at-risk students, including those with special needs such as autism and ADHD (Clarfield & Stoner, 2005; Grindle, Hughes, Saville, Huxley, & Hastings, 2013; Huffstetter et al., 2010; Pindiprolu & Forbush, 2009; Whitcomb, Bass & Luiselli, 2011). Furthermore, the HERP provides built-in analytics to measure usage and progress from a distance. In addition, the web-based platform ensured student access from any computer with Internet access, such that the intervention could be delivered either at school or in the home.

The HERP is an Internet-based beginning reading program developed with a goal of bringing kindergarten students up to a second grade reading level through the completion of 80 “episodes” or lessons (Layng, Twyman, & Stikeleather, 2003). The HERP is designed to adapt to each user’s individual learning pace, and each instructional segment is taught to mastery before progressing to the next segment. As students interact with the program, their number of response opportunities and the accuracy of their responses are tracked. If the mastery criterion for a particular segment is not met on the first try, the student is provided a corrective teaching sequence followed by new opportunities to demonstrate mastery; this routine is repeated until the mastery criterion is met. Thus, the instruction is individualized for each student by adapting to their pattern of correct versus incorrect responses.

The first 23 episodes in the HERP focus on the basic understanding that letters represent sounds and that letters and sounds can be combined to form words. Foundational instruction in phonological awareness and phonics is provided with segmenting and blending activities, and a

corpus of common vocabulary words is established. These episodes proceed at a slower pace to allow the participants to get comfortable with the mechanics (i.e., manipulating the mouse, following the directions given by the computer) without putting undue pressure on them to do so quickly. In episodes 24-40, the pace of instruction is accelerated, and students begin reading sentences and stories of increasing length and complexity. By the end of this section, a reading vocabulary of 500 words has been established. In episodes 41-56, multiple sounds are introduced in each episode, and participants are expected to be able to segment and decode more advanced real and nonsense words. The focus on comprehension is also increased. The final episodes, 57-80, target expanding reading vocabulary, fluency, and comprehension.

Each participant was assigned to begin with the first episode. The HERP documentation suggests that an average episode should take an average student 15-20 minutes to complete and recommends that students consistently use the program three to five times per week for optimal benefits. Thus, parents and/or teachers were instructed to implement the program a minimum of four times per week for 15-20 minutes per session. Following the recommended schedule would have enabled participants to complete all 80 episodes, as long as their pace of episode completion was relatively similar to that of average students. Investigators assessed frequency of use and progress through the training program with weekly remote monitoring through the HERP website allowed the investigators to assess frequency of use and episode progress. Parents and teachers were also contacted approximately weekly via e-mail, telephone, or Skype/Facetime to discuss student progress, parent/teacher concerns, and ways to facilitate participation with the program.

## **Results**

The first research question asked whether individuals with FXS could successfully complete a mainstream reading intervention that explicitly targets phonological awareness and phonics as well as other literacy skills. To answer this question, we examined students' average weekly usage of the site, as well as cumulative progress through the instructional program at the conclusion of the 6-month intervention period. These data are displayed for each subject in the last five columns of Table 5. All of the participants were able to use the program, but participants varied in their average usage each week, and therefore, their cumulative usage across the 6-month intervention period. Whereas the recommended usage was at least four, 15-20 minute sessions per week (60-80 minutes total), the average number of days that participants logged into the site ranged from 1.33 to 3.40 per week, and the average number of minutes logged on the program ranged from 30.2 to 68.9 per week (i.e., 50-100% of the recommended weekly minutes). Participants also varied in the average amount of time required for them to master an episode, from 15.8 minutes to 40.9 minutes. After six months, all participants had logged more than ten hours of cumulative time in the HERP program (ranging from 10.2 hours to 30.37 hours). In this time, all but one participant (88%) had progressed beyond the introductory episodes (i.e., beyond episode 24), five participants (62.5%) had progressed to the final section of episodes, and two of those had successfully completed the entire program (another participant completed the program a few days after the study ended). Overall, these results suggest that the participants with FXS were able to access and use the intervention developed for mainstream students, and most made substantial progress toward program completion.

Note that, because progress through the HERP episodes was contingent on demonstrating mastery of the episode content, participants took different amounts of time to reach the same episode. For example, Participants A and H both completed the final episode, but their

cumulative time in HERP differed by 6.1 hours. Also, Participants E and G completed 22 and 25 episodes, respectively, but their cumulative time in HERP differed by 6.9 hours. Accordingly, our second research question involved examining factors that might explain the degree to which children would successfully progress through the intervention. To address this question, we considered both quantitative data and qualitative feedback from parents and participants. We used Spearman correlations to examine the relationship between participants' age and pretest measures of cognitive and reading abilities at pretest, their average number of weekly logins, their average weekly usage in minutes, and their pace of episode mastery, as measured by the average number of active minutes required to pass an episode. The two usage measures, weekly logins and weekly minutes, were positively and significantly correlated ( $r = .86, p < .01$ ). The correlations between usage measures and episode mastery pace were nonsignificant ( $r = -.55$  and  $-.24$ , respectively), although the direction of the trends suggests that individuals who logged in more often took less time to progress through the episodes. As shown in Table 5, usage was significantly positively correlated with scores on two of the TERA pretests, indicating that individuals with better reading skills at pretest tended to log in more often and spend more time on the program each week than individuals with lower reading skills.

In addition to literacy skills, parent and teacher feedback suggested that other factors affected program usage. For example, one of the two participants who completed the full program was the only participant not currently enrolled in school, having aged out the previous year. Therefore, he did not need to fit in episode completion around a full school schedule. The other participant who completed the entire program was the only female, had the highest level of literacy skills at the onset of the program, and was also able to complete episodes at a faster rate than the other participants. This may have made use of the HERP more appealing, as the time on

the computer was always relatively short. Although the majority of parents/teachers reported that the program was easy to access and seemed appropriate for the developmental level of the participant, compliance seemed to decrease as the study progressed. Per the parent of Participant G, initially her son “love[d] the program. He is the one who is reminding me that he needed to complete reading episodes.” After one month, she reported that he “needs to be asked several times to start his episodes” and at the five-month mark she would “often find [herself] begging him to complete required activities.” These remarks were echoed at varying levels by most of the parents/teachers participating in the study. Furthermore, subject participation was impacted by a multitude of factors, such as the family relocating, lapses in home Internet connection, holidays, challenging student behavior/resistance to using the program, and prolonged student illness including hospitalization. For example, Participant E became seriously ill with pneumonia during the study and was hospitalized and missed several weeks of school. His mother reported being extremely stressed by the illness and that getting him well took precedence over any academic work. In contrast, Participant F, who completed the study during his school day, was fairly consistent in his use of the program. His failure to complete the 80 episodes in six months was primarily due to breaks in the academic calendar. Collectively, these reports underscore the range of factors that may influence fidelity to a treatment schedule, especially for a population with high special needs.

The third research question asked whether the program demonstrated evidence of potential effectiveness. Without a control group or control condition (e.g., a baseline period with multiple assessments but no intervention), it is impossible to test the effectiveness of the intervention, as positive changes observed between pre- and post-test could be attributed to practice effects or normal maturation. However, evidence of statistically significant gains

between pre- and post-test would provide evidence of potential effectiveness. Figure 1 displays the change scores for each participant on each test. Wilcoxon signed-rank tests were conducted to examine changes in raw scores on all standardized reading measures, and Cohen's  $r$  was used as a measure of effect size. As this was a feasibility study involving a small sample, no adjustments for multiple comparisons were made. No statistically significant differences in pre- and post-test scores were detected on the TERA-3 subtests. The effect size for the Alphabetic subtest was medium ( $z = 1.90, p = .06, r = .67$ ), whereas small effect sizes were observed for the Conventions ( $z = 1.34, p = .18, r = .47$ ) and Meaning subtests ( $z = -0.41, p = .69, r = -.15$ ). We note that while most participants increased their scores, two participants had negative change scores on each of the three TERA-3 subtests (affecting a total of 3 participants across all subtests). However, large, statistically significant gains ( $t = 2.39, p < .02, r = .85$ ) were observed on the WJ Passage Comprehension subtest, where seven of the participants showed improved scores, and one participant maintained the same score. Overall, these results provide mixed evidence of potential effectiveness.

## **Discussion**

Study 2 was designed to test the feasibility of a web-based reading intervention targeting phonemic awareness and phonics for individuals with FXS. Results indicated that individuals with FXS were able to use the HERP intervention, which was originally developed for a mainstream population. Because participants had to demonstrate mastery of the instructional targets to progress from one instructional segment to another, the final episode completed represents their intervention progress. Such progress varied, and while all but one participant progressed past the introductory section, only two (25%) of the participants successfully completed the program in the six-month period, while a third completed it shortly after the study



ended. Most previous studies have implemented the HERP for a shorter duration—ranging from a few days (Plavnick et al., 2014) to eight weeks (Huffstetter et al., 2010; Pindiprolou & Forbush, 2009) to 60 school days (Clarfield & Stoner, 2005). However, the participants in this study showed progress is comparable to that observed Grindle et al. (2013) who found that their participants with ASD required 22-43 weeks (5-10 months) to complete all 80 sessions.

Results indicated that usage and progress were related to a number of factors, including initial literacy skills, school and family issues which affected scheduling, and participant interest. The correlational analyses demonstrated that individuals with stronger reading skills at pretest were more likely to use the HERP program and showed greater progress through the HERP episodes. Such findings have important practical implications, as they suggest a need for consideration of initial skills before prescribing the intervention program. Future studies could examine precursor skills that need to be in place prior to implementing a program such as HERP, and whether pre-teaching (perhaps face-to-face) some prerequisite skills increases usage and effectiveness. We note that Grindle et al. (2013) implemented pre-teaching as needed for their participants with ASD, and all participants completed the full HERP program, albeit at differing rates (22-43 weeks).

In addition, feedback from parents in our study in highlighted the effort required to maintain consistent usage of the HERP. This may reflect traits often associated with FXS such as increased anxiety or poor attention, participant frustration with the increasing difficulty of the program, fatigue at being asked to complete additional tasks outside of the school day, or other unknown factors. It does highlight the burden faced by parents when attempting to implement an at-home intervention program for an extended period. We note that most published studies of HERP have involved usage during the school day (Clarfield & Stoner, 2005; Grindle et al., 2013;

Plavnick et al., 2014). In one study that tested parent implementation of computerized interventions (contrasting parent implementation of HERP vs. another computerized intervention), 5/30 children (17%) did not complete the assigned 40 sessions; however, those who did complete the sessions, showed significant gains on standardized reading assessments (Pindiproulou & Forbush, 2009). Fidelity is a challenge with almost any study performed outside of a lab. However, it is important for future studies to consider the potential effects of interventions employed in more natural environments. Future studies could examine whether forming cooperative relationships with schools and incorporating the program as part of the school day can achieve better fidelity to the treatment schedule. Additionally, the use of specific behavior intervention strategies to supplement the HERP may help increase fidelity whether used in school or at home. For example, Plavnick et al. (2014) found that students with autism showed increased engagement and reduced off-task behaviors during use of HERP when a behavior intervention package was applied. Future studies might consider specific behavior training for parents when implementing an in-home intervention such as the HERP.

This study involved a small sample and no control group, and the participant sample was heterogeneous, with variability in age and intellectual abilities, as well as other factors. Also, we were not able to consider important variables, such as the use of medications for attention, anxiety, or the type of reading instruction the participants had or were receiving in school. As this study's focus was on the feasibility of a reading intervention in a population with a wide spectrum of abilities, and there is no published data on reading intervention in FXS, it was decided to include all participants. These limitations restrict the evaluation of efficacy, but we examined changes in pre- and post-test scores for evidence of potential effectiveness. Overall, the results were mixed, with large, statistically significant gains observed on the WJ Passage

Comprehension subtest, and non-significant gains on three subtests from the TERA. Obviously, more statistical power is needed to adequately assess treatment efficacy and effectiveness. In addition, the ability to detect treatment effects may also be affected by measurement issues. In this case, the selection of the TERA was based on its use in previous studies of children with autism (Davidson & Weismer, 2014) and its ability to detect significant treatment effects related to preschoolers' usage of the HERP (Huffstetter et al., 2010). However, considering the performance of the students in this study, we questioned whether the progression of item difficulty on the TERA was sufficiently calibrated to detect gains across the wide range of ability levels represented in our participant sample. For example, the last few items on the TERA Alphabetic subtest (items 26-29) require students indicate the number of syllables or phonemes in printed multisyllabic words. Successful completion of such items requires the student to read the word, hold its pronunciation in memory, and count the number of syllables or phonemes to provide the answer. We note that no participant earned a score higher than 26 at either test time. It is possible that participants who initially scored in this range actually made reliable gains in literacy skills, but item difficulty in this portion of the TERA progressed too rapidly to detect it. In contrast, significant gains on the WRMT-III Passage Comprehension may have been detected because the progression of item difficulty was more consistent within the range of ability exhibited by the participants in this study. Future studies should consider the progression of item difficulty across the range of ability (or potential ability) of the participant sample. However, we acknowledge that this is a particularly difficult challenge when working with the FXS population, where there is such heterogeneity in cognitive and academic abilities.

### **Conclusions**

Evidence-based practice with regard to reading instruction in children with FXS is limited by a paucity of research on reading instruction for this population. A primary question is whether individuals with FXS can benefit from phonologically based reading instruction, such as that recommended for typically developing children. The two studies presented in this paper addressed this issue by examining the predictive relationship between phonological awareness and reading in adolescent males with FXS, and by testing the feasibility of a web-based, comprehensive reading intervention package that stressed phonological awareness and phonics. The results of Study 1 converged with those of previous studies (Adlof et al., 2015; Klusek et al., 2015) to show that phonological awareness skills are uniquely predictive of reading abilities, after controlling for nonverbal cognitive abilities and ASD severity. This relationship parallels that seen in typical development. While causality was not tested in Study 1, the results are in line with the hypothesis that improvement in phonological awareness may be associated with improvement in overall reading skills, as is the case for typically developing individuals and those with reading and learning difficulties.

The results of Study 2 demonstrated that individuals with FXS are able to engage in a web-based reading intervention program designed for mainstream beginning readers. However, progress within the program was variable and significantly associated with baseline skills. Overall, the results of Study 2 provided positive evidence of feasibility but mixed evidence of potential effectiveness. Additional studies with a larger sample and a control group or condition are needed to determine whether this intervention or other comprehensive interventions that incorporate specific instruction in phonological awareness can be effective for improving reading skills in individuals with FXS. Descriptive data from both studies highlighted the need for careful consideration with regard to test selection for this population. Identifying valid and

reliable measures of literacy skills is particularly difficult for individuals with FXS, who often present with severe intellectual disabilities (cf. Baker, Spooner, Ahlgrim-Dezell, Flowers, & Browder, 2008). Future studies may need to utilize a combination of researcher-designed and published assessments.

Overall, our findings indicate that phonological awareness skills predict reading skills in individuals with FXS, and that individuals with FXS are able to participate in a mainstream reading intervention program that includes specific instruction in phonological awareness and decoding. Taken together with findings that typically developing children and children with other developmental disabilities benefit from phonologically based reading instruction, these findings suggest that phonologically based reading instruction approaches should *not* be ruled out for individuals with FXS. Given the strong evidence for explicit, code-focused instruction across multiple populations, including individuals with severe developmental disabilities (Allor et al., 2014), we recommend that educators consider similarly explicit instruction when working with individuals with FXS, unless empirical evidence that contraindicates such practices is gathered. Ultimately, there is an important need for studies that examine best practices for improving reading and reading-related skills in this population.

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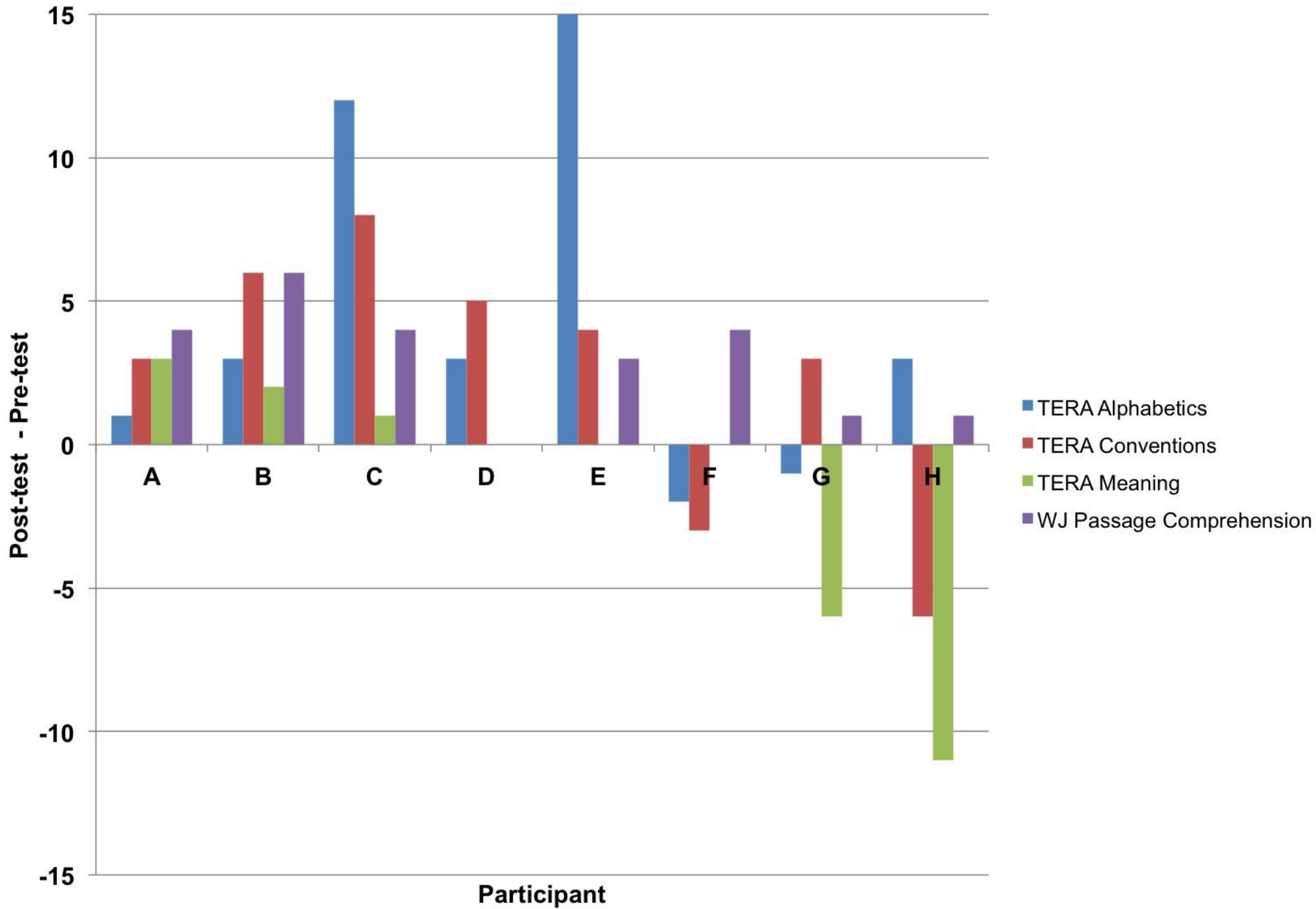


Table 1

*Participant Demographic and Descriptive Statistics*

Variable	
Chronological Age	
Mean ( <i>SD</i> )	18.50 (2.02)
Range	16.04-23.88
Maternal Education Level (%)	
High school or lower	56.53
Bachelor's degree	17.38
Master's degree	26.09
Household Income (%)	
<20k	4.35
21-40k	13.05
41-60k	13.05
61-80k	17.40
81-100k	17.40
>100k	34.79
Race (%)	
Caucasian	85.46
African American	11.54
Other	4.00
Nonverbal IQ <sup>1</sup>	
Mean ( <i>SD</i> )	38.54 (6.00)
Range	36.00-56.00
Nonverbal Mental Age <sup>1</sup>	
Mean ( <i>SD</i> )	7.32 (1.37)
Range	5.78-11.37
ASD Symptom Severity <sup>2</sup>	
Mean ( <i>SD</i> )	5.46 (2.39)
Range	1.00-10.00

*Note.* <sup>1</sup>Measured with the Brief IQ Scale of the Leiter-R. <sup>2</sup>Indexed with the Comparison Score of the ADOS-2. Scores  $\geq 3$  are consistent with a diagnosis of ASD (scores within the range of 3-4 indicate low level of ASD-related symptoms, 5-7 a moderate level, and 8-10 a high level).

Table 2

*Descriptive Statistics for Performance on Phonological and Reading Measures*

Measure	Mean (SD) Range
Gates-MacGinitie Pre-Reading Total, Extended Scale Score <sup>1</sup>	272.15 (27.56) 241.00-333.00
Gates-MacGinitie Phonological Awareness subtest, Raw Score <sup>2</sup>	6.50 (3.60) 2.00-19.00
Transformed Gates-MacGinitie Phonological Awareness subtest, Raw Score <sup>2</sup>	1.75 (0.49) 0.69-2.94
WRMT-III Word Identification subtest, Raw Score <sup>3</sup>	8.23 (8.24) 0-31.00
WRMT-III Word Identification subtest, Growth Scale Value Score <sup>4</sup>	394.69 (51.81) 336.00-521.00
Transformed WRMT-III Word Identification subtest, Growth Scale Value Score <sup>4</sup>	8.87 (0.27) 8.56-9.50
WRMT-III Word Attack subtest, Raw Score <sup>5</sup>	1.46 (4.14) 0-19.00
WRMT-III Word Attack subtest, Growth Scale Value Score <sup>6</sup>	428.08 (21.81) 418.00-508.00

*Note.* <sup>1</sup>Possible Extended Scale Scores range from 74-454; scores within the range of 74-330 are consistent with achievement at the kindergarten level. <sup>2</sup>Possible raw scores on the Oral Language subtest range from 0-20. <sup>3</sup>Possible raw scores on the Word Identification subtest range from 0-46. <sup>4</sup>Possible Growth Scale Value scores on the Word Identification subtest range from 336-599; a Growth Scale Value of 500 corresponds with the achievement of an average student finishing third grade. <sup>5</sup>Possible raw scores on the Word Attack subtest range from 0-26. <sup>6</sup>Possible Growth Scale Value scores range from 418-550; a Growth Scale Value of 500 corresponds with the achievement of an average student finishing third grade.



Table 3

*Regression Coefficients Depicting Phonological Awareness as a Predictor of Word Identification*

<i>Word Identification</i>				
Effect	Standardized Beta	<i>T</i>	<i>p</i>	<i>R</i> <sup>2</sup>
Step 1				
Intercept	0	40.51	.001*	
Nonverbal Cognition	0.29	0.04	.020*	.28
ASD Severity	-0.28	-1.57	.130	
Step 2				
Intercept	0	39.97	.001*	
Nonverbal Cognition	0.23	1.34	.192	.46
ASD Severity	-0.37	-2.27	.033*	
Phonological Awareness	0.46	2.78	.011*	

\**p* < .05

Table 4

*Participant Demographic Information, Pretest Reading Scores, and HERP Usage Data*

Subj.	Sex	Age (Yrs)	VIQ	NVIQ	FSIQ	AUT Status	TERA Alph. Pre	TERA Conv. Pre	TERA Mean. Pre	WJPC Pre	HERP Mean Login/ Week	HERP Mean Min/ Weeks	HERP Mastery Pace (Min/ Episode)	HERP Total Min.	HERP Last Episode Comp- leted
A	F	8.83	68	59	62	AUT <sup>1</sup>	25	16	16	13	3.40	62.39	15.38	1230	80
B	M	8.92	53	53	51	AUT <sup>1</sup>	20	11	13	11	2.10	42.97	16.74	1105	66
C	M	10.08	44	43	41	AUT <sup>1</sup>	6	5	7	5	1.33	33.94	40.92	1023	25
D	M	10.33	----	---	---	AUT <sup>1</sup>	3	3	7	4	2.13	30.38	15.83	855	54
E	M	12.58	49	43	44	AUT <sup>1</sup>	5	4	8	6	1.48	30.17	27.82	612	22
F	M	14.08	43	42	40	AUT <sup>2</sup>	22	7	13	8	3.25	68.94	23.06	1822	79
G	M	14.5	53	43	46	----	24	10	15	14	2.15	52.72	17.97	1348	75
H	M	23.75	47	43	42	AUT <sup>1</sup>	21	9	14	8	2.76	57.89	19.95	1596	80

*Note.* Participants listed in order of chronological age. VIQ, NVIQ, and FSIQ were measured with the Stanford-Binet Fifth Edition (Roid, 2003). IQ data for one participant were not available. <sup>1</sup>Measured with the ADOS/ADOS-2 (Lord et al., 2012). <sup>2</sup>Measured with the Social Responsivity Scale, Second Edition (Constantino & Gruber, 2012). Autism status for one participant was not available. HERP mean logins/week was calculated as (total logins during study period)/(number of weeks between first login and posttest). HERP mean minutes/week was calculated as (total minutes during study period)/(number of weeks between first login and posttest).

Table 5

*Spearman Rank Correlations Between Usage and Pretest Reading and Cognitive Skills*

	Usage: Logins/Week	Usage: Minutes/Week	Mastery Pace: Days/Episode
Chronological Age	0.14	0.14	0.38
TERA Alpha	0.76*	0.86*	-0.31
TERA Conv	0.52	0.64+	-0.48
TERA Mean	0.76*	0.71*	-0.47
WJ Passage	0.54	0.60	-0.40
Verbal IQ	0.23	-0.05	-0.81*
Nonverbal IQ	0.12	-0.12	-0.71+
Mastery Pace	-0.55	-0.24	1.00

\*  $p < .05$ , +  $p < .1$