Background
Adults with Fragile X Syndrome (FXS) have shown benefit in receptive vocabulary from in cognition, verbal reasoning, and overall language scores. 7 Males with FXS perform well below chronological age on measures of receptive and expressive vocabulary. 8 The original language and behavior test was administered to all subjects (language: SEK) throughout the study.

Methods
Subjects
- The original sample consisted of 42 adults (33 males, 9 females) with FXS recruited through the FXS Clinic, Rush University Medical Center, Chicago, and was completed only with the cooperation of the subject's legal guardian.

Measures
- All measures were consistently administered by the same experienced examiner to ensure reliability.

Randomization and Dosing
- After baseline testing (week 1), subjects underwent a 4-week placebo lead-in period, and then were randomized to CX516 or placebo per computer-generated randomization list maintained in the pharmacy. All examiners, investigators, subjects, and families were blinded to treatment status throughout the study.

Results
- The ICC of .88 indicates very good test-retest reliability. The PLS-4 was used as an outcome measure for intervention efficacy studies with adults with cognitive impairment, suggesting that the PLS-4 may be a useful tool for monitoring progress in adults with cognitive impairment.

Discussion
- The PLS-4 provides a reliable benchmark over repeated trials in adults with FXS for clinical trials research.

Implications for Future Research
- There is a wide range of language and cognitive function represented in adults with FXS and a need for additional language skills in these adults. Therefore, the identification of reliable measures that may be used in clinical trials research over a time frame of less than 6 months is critical.

Conclusions
- The PLS-4 represents one such reliable measure for adults with a mental age of less than 6 years, 0 months, despite the fact that it was developed for use in young children.

References