Title: An Improved Descriptive Profile of IQ in Fragile X Syndrome Using Deviation Scoring On the Stanford Binet 5 across Multiple Study Sites

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Introduction: This study aims to characterize the cognitive profile in the fragile X syndrome (FXS) full mutation in a more sensitive manner and in a broader sample than has been done before to our knowledge, by presenting a large collection of deviation IQ scores generated from assessment of the Stanford Binet Intelligence Scales, Fifth Edition (SB5). We combine data from several sites in order to provide descriptive IQ information in FXS. Compared to standardized IQ scoring, this deviation IQ scoring has implications for better understanding of individuals’ cognitive profiles as well as valuable implications for detecting changes, as is critical for measuring improvement in clinical trials. These data were collected across multiple sites as part of several NIH grants to validate the NIH Toolbox and expressive language sampling as outcome measures for FXS, a CDC-funded natural history study of FXS, and baseline data from several clinical trials of new medications in FXS, all projects in which the SB5 was administered.

Method: We performed deviation score calculations according to the method described in Hessl et al (2008) and Sansone et al (2014) on SB5 data from individuals with FXS, ages 3-51 years. Although data is still being analyzed and we are in the process of identifying and removing duplicate participants from the total sample of approximately 400, preliminary analyses are presented here from a subsample of these individuals. We used raw scores from the SB5 to perform deviation score calculations and generate deviation IQ scores. We used the Vineland Adaptive Behavior Scales, 2nd or 3rd edition (VABS-2 or VABS-3) depending on the study, to correlate Adaptive Behavior Composite (ABC) with IQ scores.

In final analyses we will present results from individual subtests of the SB5 as well as provide additional analyses on the individuals at the floor of the SB5, to provide detailed description of the cognitive profile of FXS with individual variation on subtests below the typical floor. We plan to test our method by examining the subset of the entire sample with a standardized FSIQ of 40 and present the correlations between the deviation IQ and Vineland composite scores, in order to demonstrate the validity of IQ scores below the normal floor. We will also present SB5 change sensitive scores, as this scoring approach is another method of value in measuring improvement and may have relevance in the clinical trial setting. Additionally, we plan to conduct analyses to provide information about what signifies clinically meaningful improvement.

Results: Although data are still being analyzed, in our preliminary results from 226 individuals ages 3-51 years (47 females, mean age = 17.03, SD = 9.11; 177 males, mean age = 14.99, SD = 6.33; 2 unreported gender), the standardized full scale IQ (FSIQ) ranged from 40 to 95, with a mean of 47.09 and a SD of 10.84. There were 93 individuals, or 41.15% of the sample, with a standardized FSIQ of 40 and present the correlations between the deviation IQ and Vineland composite scores, in order to demonstrate the validity of IQ scores below the normal floor. We will also present SB5 change sensitive scores, as this scoring approach is another method of value in measuring improvement and may have relevance in the clinical trial setting. Additionally, we plan to conduct analyses to provide information about what signifies clinically meaningful improvement.

Discussion: This data is important in regards to accurately characterizing the cognitive profile of fragile X syndrome, especially due to the variability that is present across individuals with floored scores on the standardized IQ. The method allows for the portrayal of individuals’ strengths and weaknesses across domains versus a flat floored profile across domains. The deviation scoring method is useful for any individual with one or more floored subscale scores. Sensitive measures are critical in order to detect changes over time, such as in clinical trials, which require sensitive measures in order to detect small but meaningful improvement. These results will be relevant for future longer term treatment trials aiming to shift IQ. Additionally, having properly matched groups in a trial depends on accurate and sensitive measurement of functioning in each group.
References/Citations:
