**Symposium Title:** Examining Social Behavior and its Physiological Correlates in Boys and Girls with Intellectual and Developmental Disability

**Chair:** Jennifer Bruno¹

**Co-Chair:** Ellen Wilkinson²

**Discussant:** David Hessl³

**Overview:** Deficits in social behavior are a pervasive feature of individuals diagnosed with an intellectual and developmental disability (IDD) (Beadle-Brown et al., 2005). These deficits are particularly pervasive in individuals with fragile X syndrome (FXS), the most commonly inherited form of IDD (Crawford et al., 2015). Extensive work has determined that the development of appropriate social gaze behavior is a critical prerequisite to subsequent language development, emotion recognition, social engagement, and general learning through joint attention (Mirenda et al., 1983), however little research has sought to better understand the parameters under which deficits in social gaze behavior occur in FXS and the general IDD population. The development of appropriate social behavior can therefore be considered a critical target for behavioral interventions for children with IDD. In this symposium, we present three papers examining the breadth and specificity of social avoidance behavior and its physiological correlates in boys and girls with FXS in comparison to boys and girls with a general IDD diagnosis. The first presentation outlines data validating a parent-survey that can be used to quickly and easily quantify the severity of eye contact avoidance in children with IDD. The second presentation examines if a social gaze behavioral training intervention, proven to increase social gaze behavior in boys with FXS, can be applied to a general IDD population. The final presentation presents pilot functional near-infrared spectroscopy (fNIRS) data to examine the neural underpinnings of social behavior in girls with FXS. Together these presentations will emphasize the importance of increasing our understanding of the heterogeneity of aberrant social gaze and social avoidance behavior in children with IDD.

**References/Citations:**

**Paper 1 of 3**

**Paper Title:** Reliability and Validity of a Screening Tool for Measuring Social Gaze Avoidance in Boys with Developmental Disabilities

**Authors:** Ellen H. Wilkinson² & Scott S. Hall²

**Introduction:** Eye gaze avoidance is a significant and disabling problem for children with developmental disabilities (Wing et al., 2005). Few measures, however, are available to quickly and easily quantify the severity of eye contact avoidance in children with developmental disabilities. In this study, we examined the reliability and validity of the Eye Contact Avoidance Scale (ECAS), a 16-item informant-based questionnaire that has 5 subscales: speaking, listening, inability, difficulty, and anxiety (Venema & Hall, 2017).

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Methods: Caregivers completed the ECAS as part of a larger study investigating the effects of social gaze training in children with developmental disabilities. The sample consisted of 243 boys with a developmental disability aged 7-18 years (mean age = 11.79, SD = 3.07). Data were also obtained on the frequency and severity of the child’s problem behaviors (e.g., aggression, self-injury, stereotypy and elopement). 52 caregivers completed the ECAS again approximately 4 weeks later.

Results: The alpha coefficient of the total ECAS score was .95, and ranged from .86 to .88 for the subscales, indicating excellent internal consistency. The test-retest alpha coefficients for the total ECAS score was .77 and ranged from .73 to .80 for the subscale scores, indicating acceptable reliability. There was a significant effect of age (t = 2.13, p = .034) and communication ability (F = 4.08, p = .006) on the total ECAS score. Children with higher levels of social gaze avoidance exhibited more frequent problem behaviors such as aggression and elopement.

Discussion: Results suggest that this measure has acceptable to excellent internal consistency and reliability on a general developmental disability population. The effects of communication ability, age, and problem behavior further support the validity of the measure (Cohen et al., 1991). This survey could potentially be used in situations that require a quick and easy way to measure eye gaze behavior - for example, to measure changes following a pharmacological or behavioral intervention. Results will also be compared to a sample of 148 boys with fragile x syndrome.

References/Citations:

Paper 2 of 3

Paper Title: Examining the Specificity of a Behavioral Skills Training Package for the Treatment of Social Gaze Avoidance in Fragile X Syndrome

Authors: Tobias C. Britton, Caitlin E. Gannon, & Scott S. Hall

Introduction: Children diagnosed with fragile X syndrome (FXS), the most common known inherited form of intellectual disability, commonly exhibit significant impairments in social gaze behavior during interactions with others (Cohen et al., 1988; Hall et al., 2007). Few studies, however, have examined the extent to which social gaze behavior may be improved using behavioral training methods. In a previous study, we showed that discrete trial instruction (DTI) plus relaxation training could significantly improve social gaze behavior in boys with FXS (Gannon et al., 2018). In the present study, we examined whether this intervention could also be effective for children with intellectual and developmental disability (IDD).

Methods: Training data were obtained from 20 boys with IDD aged 8 to 18 years, and compared to the data obtained from 20 age- and developmental age matched boys with FXS who had received similar amounts of behavioral skills training as reported by Gannon et al. (2018). Both groups of boys evidenced significant deficits in social gaze behavior at baseline. Improvements in social gaze behavior were evaluated by conducting Theil-Sen regression analyses across the blocks of training trials for each participant.

Results: Results showed that 9 (45%) boys with FXS and only 2 (9%) boys with IDD evidenced significant improvements in social gaze behavior across the training blocks, a significant difference between the groups ($X^2(1) = 6.14, p = 0.013$). There were no effects of age or developmental age on treatment response across groups.

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Discussion: The results indicate that the behavioral skills training package was significantly more effective for boys with FXS compared to boys with IDD. Given that FXS has a distinct neurogenetic profile that includes significant symptoms of hyperarousal, these data suggest that the intervention effects were syndrome-specific. Our results further highlight the need for interventions specifically tailored to the underlying phenotype of FXS. Potential biomarkers of treatment response will be discussed.

References/Citations:

Paper Title: Social Behavior and Brain Activation Patterns in Girls with Fragile X Syndrome

Authors: Jennifer L. Bruno¹, Matthew Marzelli², Amy Lightbody¹, Cindy Lee¹, Kristi Bartholomay¹, & Allan L. Reiss¹

Background: Fragile X syndrome (FXS), the most common inherited cause of intellectual disability, occurs in ~1 in 4,000 males and ~1 in 8000 females (Crawford et al, 2015). The broader range of symptoms and overall higher IQ relative to boys allows females to play a particularly important role in understanding the complexities of the FXS phenotype. Social avoidance and social withdrawal are among the most significant behavioral challenges for females with FXS (Hagerman et al, 2017). We present a pilot study examining the neural underpinnings of social behavior in females with FXS using functional near infrared spectroscopy (fNIRS). fNIRS is a non-invasive optical imaging technique that can be used to quantify brain activation in naturalistic settings.

Methods: Participants included 17 girls with FXS (ages 6.71-14.36) and a comparison group of 14 girls without FXS who had a history of learning, behavior and/or developmental challenges (ages 7.65-14.21). Cognitive functioning was assessed via the Differential Abilities Scale (DAS). Parental report of behavior was acquired via the Vineland Adaptive Behavior Scale, Third Edition and the Social Responsiveness Scale (SRS-2). fNIRS was performed with a NIRScout (NIRx) and measurement channels were positioned over bilateral frontal, temporal and parietal regions. During fNIRS imaging, participants completed a naturalistic social interaction task. The task consisted of a conversation with a novel researcher using the structured procedures outlined previously (Murphy & Abbeduto, 2007; Kover & Abbeduto, 2010) and structured rest periods.

Results: Cognitive functioning was lower in the FXS group as indicated by the DAS general conceptual ability and verbal composite scores (p’s<0.001). The two groups did not differ on the Vineland adaptive behavior composite or on any individual subscale of the Vineland (p’s >0.10). Total scores on the SRS indicated a trend for greater symptom severity in the FXS group (p=0.085) but this difference was not significant after accounting for DAS verbal composite scores (p>0.10). However, higher scores for the FXS group on the Social Cognition subscale of the SRS did remain significant after accounting for DAS verbal composite scores (p=0.035). fNIRS data indicated unique patterns of brain activation for conversation vs. rest periods as signified by changes in concentration of oxygenated hemoglobin. In particular, we found less activation for the FXS group relative to the comparison group in right temporal regions including the superior temporal sulcus (STS, p=0.017).

Conclusions: To the best of our knowledge, this study represents the first investigation of functional brain activation patterns in association with naturalistic social interaction in girls with FXS. If confirmed in larger sample sizes, unique patterns of brain activation will advance a mechanistic understanding of social avoidance and social withdraw behaviors in females with FXS. Brain activation patterns could further be used to inform interventions and as a progress or outcome measure.
References/Citations