Research in Developmental Disabilities 43–44 (2015) 150–166

Contents lists available at ScienceDirect

Research in Developmental Disabilities

Functional analysis and treatment of problem behavior exhibited by children with fragile X syndrome

Patricia F. Kurtz [a,b,*], Michelle D. Chin [a], Ashley N. Robinson [a,b], Julia T. O'Connor [a,b], Louis P. Hagopian [a,b]

[a] Kennedy Krieger Institute, Baltimore, MD, United States
[b] Johns Hopkins University School of Medicine, Baltimore, MD, United States

ARTICLE INFO

Article history:
Received 9 December 2014
Received in revised form 16 June 2015
Accepted 25 June 2015
Available online 14 July 2015

Keywords:
Fragile X syndrome
Problem behavior
Self-injurious behavior
Aggression
Functional analysis
Treatment
Generalization

ABSTRACT

The efficacy of function-based interventions for the treatment of severe problem behavior exhibited by individuals with intellectual and developmental disabilities (IDD) is well established. However, few studies have reported on behavioral interventions in fragile X syndrome (FXS) specifically. The present study is a consecutive case-series analysis that reports on functional analysis and treatment of problem behavior of nine children with FXS. Assessment findings were consistent with previous research indicating that among individuals with FXS, problem behavior is more commonly maintained by escape from demands and access to tangible items, relative to the broader population of individuals with IDD. Functional analysis-based behavioral interventions resulted in a mean reduction in problem behavior of 95.2% across the nine participants. Additionally, generalization of treatment effects from controlled clinical settings to home, school, and community was demonstrated. The current findings suggest that function-based behavioral interventions shown to be effective with the broader population of individuals with IDD are also effective for individuals with FXS. Our results in combination with those of previous studies describing functional analysis outcomes provide additional evidence for a unique functional behavioral phenotype for severe problem behavior in individuals with FXS. Implications of study findings for early intervention and prevention of problem behavior in children with FXS are discussed.

1. Introduction

Fragile X syndrome (FXS) results from an expansion of the cytosine-guanine-guanine (CGG) sequence within the fragile X mental retardation 1 (FMR1) gene and is the leading known genetic cause of heritable intellectual disability. Affecting approximately 1 in 4000 males and 1 in 8000 females (Crawford, Acuña, & Sherman, 2001), males with the disorder typically express more severe deficits and behavior problems than females due to the expansion occurring on the X chromosome (Bailey, Raspa, Olmsted, & Holiday, 2008). In addition to intellectual deficits, FXS is commonly associated with autism spectrum disorder (ASD), with prevalence estimates suggesting between 21 and 60% of individuals with FXS meet ASD

* Corresponding author at: Neurobehavioral Unit, Kennedy Krieger Institute, 707 N. Broadway, Baltimore, MD 21205, United States. Tel.: +1 443 923 2840.
E-mail address: kurtz@kennedykrieger.org (P.F. Kurtz).

http://dx.doi.org/10.1016/j.ridd.2015.06.010
0891-4222/

diagnostic criteria (Bailey et al., 2008; Hall, Lightbody, Hirt, Rezvani, & Reiss, 2010; Harris et al., 2008; Hatton et al., 2006; Kaufmann et al., 2004). Beyond cognitive impairments and the occurrence of ASD, FXS is associated with delayed language development, speech abnormalities, deficits in adaptive skills, and social abnormalities (Abbeduto, Brady, & Kover, 2007). Behavior problems are another key feature of the FXS phenotype, encompassing attention difficulties, hyperactivity/impulsivity, hyperarousal, social anxiety, stereotypic/repetitive/perseverative behaviors, poor eye contact, and self-injurious behavior (SIB) and aggression (Bailey et al., 2008; Hagerman, 2002; Hatton et al., 2002; Tranfaglia, 2011).

Parents of children with FXS report that SIB and aggression are among the most significant behavioral challenges (Hatton et al., 2000), as they often result in injury to self or others. Characteristic forms of SIB in individuals with FXS include finger/hand biting, head hitting, picking/pulling skin/hair, self-hitting, and self-scratching (Hall, Lightbody, & Reiss, 2008; Richards, Oliver, Nelson, & Moss, 2012; Symons, Clark, Hatton, Skinner, & Bailey, 2003; Symons, Byiers, Raspa, Bishop, & Bailey, 2010). Recent prevalence estimates for SIB in males with FXS range from 39 to 79% (Bailey et al., 2008; Hall et al., 2008; Hessl et al., 2008; Richards et al., 2012; Symons et al., 2010), with many individuals displaying two or more forms (Symons et al., 2003). The relative risk of SIB in FXS is also as much as 2.91 times more than those of other genetic syndromes, specifically Down syndrome (Richards et al., 2012). Further, the prevalence of aggression in males with FXS ranges from 38 to 75% (Powis & Oliver, 2014). Relative to those with FXS only, the significance and severity of behavior problems increases for individuals with co-morbid FXS and ASD (Hernandez et al., 2009; Kaufmann et al., 2004; Symons et al., 2010; Wolff et al., 2012). For the purposes of this paper, the term "problem behavior" will refer to SIB, aggression, and destructive behaviors.

In view of the genetic origin of the disorder, the presence of problem behavior in FXS is often thought to be linked to biological mechanisms (Hall, 2009; Langthorne et al., 2011). For example, research suggests that pain amplification may be reduced in FXS and that this impairment could contribute to the development of SIB (Peebles & Price, 2012). As a result of the focus on biological origins of SIB, the majority of treatment studies have emphasized the use of pharmacological interventions (cf. Hagerman et al., 2009; Hall, 2009). Unfortunately, outcomes vary widely across studies (Erickson, Wink, Schaefer, & Shaffer, 2014; Hall, 2009; Rueda, Ballesteros, & Tejada, 2009; Valdovinos, Parsa, & Alexander, 2009). In a survey of medication utilization for individuals with FXS, more than one third of parents reported that medication targeting SIB had little or no effect (Bailey et al., 2012).

Research findings have demonstrated that that environmental variables play a role in the maintenance of problem behavior in FXS (Bailey et al., 2008; Hall, DeBernardis, & Reiss, 2006; Hall et al., 2008; Hessl et al., 2001; Symons et al., 2010; Langthorne & McGill, 2012), autism spectrum disorders (ASD), and a range of other intellectual and developmental disabilities (IDD) that are both idiopathic and genetically caused. Applied behavior analytic-based approaches to treatment rely on functional analysis (Iwata, Dorsey, Slifer, Bauman, & Richman, 1994a/1982) to identify the environmental variables (antecedents and consequences) that maintain problem behavior (cf. Beavers, Iwata, & Lerman, 2013; Hanley, Iwata, & McCord, 2003). Commonly identified consequences that reinforce and maintain problem behavior in persons with IDD include provision of adult attention, access to tangible items, and escape from non-preferred tasks (Iwata et al., 1994a,b); however, a number of other functions for problem behavior have been identified (Lancioni, Singh, O'Reilly, Sigafoos, & Didden, 2012; Schlichenmeyer, Roscoe, Rooker, Wheeler, & Dube, 2013). Studies reporting on the functional analysis of problem behavior have included sub-populations of IDD such as individuals with ASD (Love, Carr, & LeBlanc, 2009; O'Reilly et al., 2010); prenatal drug exposure (Kurtz, Chin, Rush, & Dixon, 2008); and genetic disorders such as Angelman (Strachan et al., 2009), Cornelia de Lange (Bay, Mauk, Radcliffe, & Kaplan, 1993), Prader-Willi (Hall, Hustyi, Chui, & Hammond, 2014), Rett (Roane, Piazza, Sgro, Volkert, & Anderson, 2001), Soto (Harding et al., 2001), and Williams syndromes (O'Reilly, Lacey, & Lancioni, 2000).

In its consensus document on clinical practices regarding behavior problems in FXS, the Fragile X Clinical and Research Consortium recommended using functional analysis to identify potential causes for problem behavior (Picker & Sudhalter, 2012). Two recent studies have reported functional analysis outcomes for children recruited for participation in FXS research. Langthorne et al. (2011) conducted functional analyses in home or school settings with eight children (ages 8 to 15 years) with FXS who engaged in SIB, aggression, and destructive behaviors. Each participant's assessment included 3–5, 5-min sessions of each of the following conditions: attention, academic demand, social avoidance, tangible, no interaction, and unstructured play (control condition). Problem behavior was maintained by escape and/or access to tangible items for all participants; none exhibited problem behavior maintained by access to attention. Machalicek et al. (2014) replicated these procedures and findings—reporting outcomes of functional analyses with 12 young boys with FXS. Eight participants exhibited problem behavior maintained by escape from demands and/or escape from social interaction, nine children exhibited tangible-maintained problem behavior, and only three displayed attention-maintained problem behavior. These initial studies are important in that they highlight the presence of SIB, aggression, and destructive behavior in FXS, and provide support for the use of functional analysis in determining the environmental variables that maintain problem behavior. Furthermore, their consistent findings regarding escape and tangible functions for problem behavior suggest that there may be patterns of responding that are more prevalent among individuals with FXS, relative to individuals with IDD. Finally, they identify a gap in the literature with regard to behavioral treatment of problem behavior in individuals with FXS.

There are three decades of research supporting the efficacy of function-based interventions for severe problem behavior (Heyvaert, Maes, Van den Noortgate, Kuppens, & Onghena, 2012; Kahng, Iwata, & Lewin, 2002), including documented empirically supported treatments such as functional communication (FC) training (Kurtz, Boelter, Jarmolowicz, Chin, & Hagopian, 2011; Rooker, Jessel, Kurtz, & Hagopian, 2013) and noncontingent reinforcement (NCR; Carr, Severtson, & Lepper, 2009; Richman, Barnard-Brak, Grubb, Bosch, & Abby, 2015). Despite overwhelming empirical support for the use of applied behavior analysis in assessment and treatment of problem behavior in persons with IDD, there is a paucity of research