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Parenting of children with Down syndrome compared to fragile X syndrome

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Abstract

Children with Down syndrome (DS) and fragile X syndrome (FXS) struggle with language development. Parenting variables, such as responsiveness to children's communication attempts (Maternal Responsivity), and techniques used to support and teach appropriate behaviour (Behaviour Management) are known to have a significant impact on early child development. We examined these two aspects of parenting style via coded, videotaped parent-child interactions in two groups of participants matched on child age (2–5 years) and child expressive language level: mothers of children with DS and mothers of children with FXS. The mothers differed in their use of gestures and redirecting the child's attention. Overall, mothers in both groups of children appeared to adapt appropriately to their children's developmental needs.

Keywords

Intellectual disability; maternal responsivity; mother-child interactions; language development

The acquisition of language is an essential, remarkably complex part of child development. It can be disrupted by a number of factors, including the presence of a developmental disability. Developmental disabilities with known genetic origins, such as Down syndrome (DS) and fragile X syndrome (FXS), are of particular interest to researchers since they provide a unique opportunity to explore the interaction of different genetic disorders and a child's environment over time on language development. The influential work of Hart and Risley (1995) has shown the cumulative impact of parenting in the development of a child's early language skills such as vocabulary. A wide range of studies have demonstrated that highly responsive parenting throughout the early childhood period is associated with a number of benefits in terms of language, cognitive, social and emotional development [4, 12–13, 18, 22, 24]. Conversely, long-term exposure to a harsh and/or unresponsive and directive style of parenting is associated with suboptimal outcomes across the same developmental domains [3, 8]. This work is grounded in the transactional model of language

development which posits that a mother's ability to engage in responsive parenting is influenced by both maternal and child factors. Research has found maternal factors such as education level, mental health and overall stress, as well as child factors such as the presence of a developmental disability, intelligibility, ASD status and behavior problems, can have a significant impact on responsive parenting [24].

While many studies have examined the impact of parenting in children with developmental disabilities, no studies have directly compared parenting of young children with different known genetic disorders. This type of comparison can help pinpoint the child effects associated with a specific disorder on parenting behaviour. Therefore, the purpose of our study was to examine two general aspects of parenting style, maternal responsivity and behaviour management, in two groups of mother-child dyads in which the child has either FXS or DS.

DS and FXS are both characterized by intellectual disability and language impairments, although there are important differences in the behavioural phenotype of the two disorders that could influence a mother's ability to implement and maintain a highly responsive style of parenting. It has been estimated that 30% to 45% of boys with FXS have a co-diagnosis of an autism spectrum disorder (ASD) [2,11], although there are also heightened rates of ASD in children with DS. ADHD and behaviour problems are also reported in the literature for children with FXS. Research has demonstrated that the presence of an ASD diagnosis, ADHD and behaviour problems have a significant impact on parenting style [1, 9]. Children with FXS have significant language delays beginning with delayed first words and extending to more complex language such as grammar [4,25]. Children with DS also have significant and pervasive difficulties with language, but demonstrate a relative strength in their use and understanding of gestures [5]. Parents are often tuned in to their child's strengths and weaknesses and may already be capitalizing on this skill. However, children with DS have significant problems with intelligibility, which is known to influence maternal responsivity [24]. By directly comparing the effects of these genetic disorders we may be able to estimate the unique impact associated with each disorder. This knowledge could then be used to guide remediation efforts so as to optimize intervention outcomes with each group.

Research on maternal responsivity within developmental disabilities, including DS and FXS has generally indicated that while many parents are highly directive and often shoulder the burden of the interaction, they are typically responsive to their child's developmental level and needs, and often exhibit a warm style of parenting [4, 16, 19–20, 23, 25]. In fact, parents seem to be in tune with their child's developmental and language level and adjust their parenting accordingly in studies on children with DS and FXS. However, very few comparative studies have been completed in neurodevelopmental disorders, despite the fact that there could be key differences in parenting style that could be intervention targets, particularly in the realm of behaviour management techniques that parents often use for children with intellectual disability

The specific aim of this paper was to examine maternal responsivity and behaviour management for a group of mothers who have a young child with FXS compared to mothers who have a young child with DS. We matched the children in both groups on chronological

age and expressive language level. We matched on expressive language level because many maternal behaviours are tied to the child's expressive output [20, 25]. We focused on two general types of maternal behaviours, maternal responsivity and behaviour management. Together they encompass a large portion of the mother's behaviour to the child, and have been directly linked to child outcomes in the literature [24]. Maternal Responsivity includes behaviours known to promote language development. Behaviour Management consists of directive behaviours used by parents to manage the child's behaviour. We hypothesized the two groups would be similar in Maternal Responsivity behaviors given that we matched the groups on expressive language abilities, but that the mothers of children with FXS would employ proportionally more Behaviour Management techniques.

Method

Participants

The participants in these analyses are drawn from two separate studies. The FXS sample ($n = 19$ mother-child dyads) was recruited from a larger longitudinal study of 55 mother-child dyads examining family adaptation to FXS (Sterling & Warren, 2014). We only included boys with FXS in the current analyses to control for gender differences. Eighteen of the mothers had the premutation and one had the full mutation. The DS sample ($n = 19$ dyads) was recruited from a separate study on responsivity in DS which included 22 mother-child dyads with DS (Warren, Brady, Sterling, Fleming, & Marquis, 2010; Brady, Warren, Keller, & Sterling, 2014). Participants with DS included 8 girls and 11 boys. The children in both groups had a mean age of 39.9 months (range: 22–60 months); they were matched at the group level on chronological age ($t(36) = .72, p = .48, d = -.13$) and expressive raw scores from the Mullen Scales of Early Learning (MSEL; $t(36) = .20, p = .84, d = .10$; DS: $M = 20.26, SD = 7.40$; FXS: $M = 19.53, SD = 7.30$) [15]. Given that many of our children were at floor on the subscales, we report raw scores. The groups were matched on maternal education ($t(36) = .49, p = .63, d = .15$; DS: $M = 15.8$ years, $SD = 1.77$; FXS: $M = 15.54, SD = 1.61$). The groups differed significantly on raw receptive language scores ($t(36) = 2.03, p = .050, d = .66$; DS: $M = 25.58, SD = 8.46$; FXS: $M = 20.63, SD = 6.46$), and approached significance in overall MSEL standard scores ($t(30) = 2.01, p = .053, d = .65$; DS: $M = 57.00, SD = 9.18$; FXS: $M = 52.00, SD = 5.73$).

Procedures

Data collection took place during a single visit by trained research assistants in the participants' home. All dyads completed videotaped mother-child interactions. The dyads were asked to read or look at books together, play together, and make and eat a snack together. A standard set of toys, books and food items were presented to each dyad. In addition, the FXS sample completed an unstructured interaction, consisting of daily activities. Each child was given the MSEL [15], a standardized assessment for children birth through 68 months. The standard score is based on a mean of 100, standard deviation of 15. The DS group had a mean standard score of 57 ($SD = 9.18$), and the FXS group had a mean standard score of 52 ($SD = 5.73$). For the visual reception subscale the children with DS had a raw mean score of 24 ($SD = 6.80$) and the children with FXS had a mean of 24.77 ($SD =$

6.17). On the fine motor subscale, the children with DS had a raw mean score of 22.75 ($SD = 4.59$), and the children with FXS had a mean of 22.70 ($SD = 4.26$).

For the FXS sample, 25 minutes of total mother-child interaction video were collected for each dyad (5 minutes of book, 5 minutes of free play, 5 minutes of snack, and two 5 minute segments from the unstructured interaction). For the DS sample a total of 21 minutes of mother-child interaction video were collected (7 minutes of book, 7 minutes of free play, and 7 minutes of snack). The coding system was adapted from previous studies, and has been used in studies of maternal responsivity in FXS and DS [12–13, 20, 25]. In order to account for the time discrepancies, all codes were converted into rates of behaviours per minute (see table 1 for definitions and examples). An utterance was coded if the mother paused her speaking for at least three seconds, or if the mother's behaviour (utterance and/or gesture) followed a child communication act. The codes were the dependent variables for this study, and were grouped by facilitating (maternal responsivity) or directive (behaviour management) behaviours.

1. Maternal Responsivity: Gestures, Recodes, Comments, Request for Verbal Complies
2. Behaviour Management: Redirects, Request for Behavioural Complies, Zaps

Research assistants were trained to identify and code the specific maternal behaviours to a training criterion of at least 80% agreement in three different practice samples before coding participant files. Once this criterion was met, two trained coders independently coded maternal behaviours for each file. We then compared transcripts and resolved disagreements through consensus. To determine the inter-judge reliability for the variables analyzed, we calculated intraclass correlation coefficients (ICCs), using the absolute agreement and single measure values for each score [17]. The ICCs were high across behaviours (mean ICC for DS group: .91, range = .73-.98; mean ICC for FXS group: .89, range = .79 - .97).

Results and Discussion

In order to compare the two groups, we completed *t*-tests on the specific codes. See table 2 for more information. Our analyses showed a significant difference in use of gestures and redirects between the two groups. Given our small sample size, we completed Cohen's *d* effect size calculations. The mothers of children with DS used gestures at a significantly higher rate compared to the mothers of children with FXS (mean rates: DS = 2.45, FXS = 1.90). In addition, the mothers of children with FXS used redirects at a significantly increased rate compared to the mothers of children with DS (mean rates: DS = .18, FXS = .33). While not significant there was a small effect size for differences between the two groups in mothers' use of comments (mean rates: DS = 2.84, FXS = 2.43), and mothers' use of zaps (mean rates: DS = .28, FXS = .41).

Our findings indicate differences between the groups in several important parenting behaviours including gestures and redirects, and small effect sizes for comments and zaps. The mothers of children with DS were using gestures more often compared to the mothers of children with FXS. Gestures are a strength for many children with DS [5]. Perhaps the mothers were cued into this strength, and using gestures to help support their child's

communication. While not statistically significant the effect size estimate indicated that there was a different trend in the use of comments between the two groups, with the mothers of children with DS using more comments compared to the mothers of children with FXS. The children with DS had better receptive language raw scores, which could also be a potential explanation for these findings.

The groups were different on their use of redirects, with higher rates in the FXS group. Past research has demonstrated several phenotypic differences between these two groups, including higher rates of ASD and higher rates of ADHD in children with FXS as well as more behaviour problems [2, 11, 21]. Many males with FXS either meet criteria for ASD or display autistic-like symptoms, including poor eye contact, perseverative behaviours, etc. [6]. While we did not control for ASD symptoms in this study, future work should address the co-morbidity of ASD and the potential impact on the dyadic interaction. Hyperactivity is a common aspect of the behavioural phenotype in FXS particularly in males [7]. The use of redirects could be a strategy parents are using to manage behaviour. McCathren et al. (1995) noted that these types of directives, although useful for managing behaviour problems, are not optimal for language facilitation and are commonly discouraged in interventions targeting increased maternal responsivity. Therefore, in the course of intervention it may be useful to teach parents alternate strategies in order to maintain a highly responsive interaction style with their young child.

Our findings are constrained by a relatively small sample size, as well as the cross-sectional approach to our work. Investigating these issues with larger samples would allow for a more in-depth examination of these specific variables, utilizing a bidirectional approach to understanding the unique maternal and child variables influencing parenting in these two populations. Maternal variables such as IQ, age, stress and mental health would be important to consider in future work. The focus of the current study was on child variables. However, maternal variables play a significant role. For example, the mothers of the children with FXS are either premutation carriers or could have FXS. Premutation carriers are at an increased risk for mental health issues, including depression, anxiety, and executive functioning problems [2,25]. Understanding the unique impact of the premutation on parenting style is an important future step. Child variables such as the presence of an ASD co-diagnosis, joint attention abilities, intellectual disability and nonverbal abilities, in addition to speech intelligibility are important variables to consider. The presence of an ASD co-diagnosis could be particularly informative, given the deficits in social reciprocity and communication in ASD. Additionally, tracking these behaviours over time in a longitudinal design would allow us to investigate how the unique phenotypes might cumulatively shape these important parent-child interactions.

With these limitations in mind, our findings support previous literature indicating that mothers of children with developmental disabilities are able to adapt their parenting style. Overall the two groups of mothers were quite similar, and demonstrated a responsive style of parenting, including the use of language facilitating techniques such as recodes and comments. There were some group differences, including the mothers of children with DS using gestures at a higher rate, which may reflect the fact that children with DS tend to demonstrate a strength in communicating with gestures [5]. Future research could expand on

this study, by systematically examining the influence of key maternal and child variables, such as maternal mental health and stress, and child ASD symptoms, joint attention skills and nonverbal abilities. Given how important the early environment is for language development, future work focused on parent-child interventions designed to help facilitate language development might build more effectively on what the parents are already doing well and teach additional facilitative strategies that can be incorporated in daily living tasks.

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Table 1.

Definition of codes

Behavior:	Definition:	Example:
<i>Maternal Responsivity</i>		
Gestures	Sign language, gestures (“come here”, “stop”, “no”), tapping, clapping, or knocking, etc.	Mom points to the book and says “Do you want to read this?”
Request for Verbal Comply	Question/statement aimed at getting a verbal response	Mom says, “say ____” with the intent of eliciting a verbal response.
Comment	All comments; praise or phrases in reaction to something the child has done	Mom says “good job”, or “the ball is red”.
Recode	Verbal interpretation/expansion of child’s communication act	Child says “ba” and mom says “do you want your ball?”
<i>Behaviour Management</i>		
Request for Behavioural Comply	Directives to which the child can comply behaviourally	Mom says, “push this one”, or “I want you to do it”
Redirect	Mom references new object when child is actively attending to another object	Child is playing with a toy and mom says, “What book should we read?”
Zap	Restricting child’s behaviour in some way – not always negative	Mom says, “no stop that”, “don’t touch that”

Note. Codes adapted from Landry et al. (1998, 2001), and Warren et al. (2010)

Table 2.

Group comparisons for mothers of children with DS compared to mothers of children with FXS on the two factors

Behavior:	<i>t (df)</i>	<i>p</i>	<i>d</i>
<i>Maternal Responsivity</i>			
Gestures	-2.251(36) *	.031	-.73
Request for Verbal Comply	.257(36)	.799	.08
Comment	-1.229(26)	.227	-.40
Recode	-.371(36)	.713	-.12
<i>Behavior Management</i>			
Request for Behavioral Comply	-.371(36)	.71	-.12
Redirect	2.012(29) *	.054	.65
Zaps	1.087(36)	.284	.35

Note. Effect size estimates calculated using Cohen's *d*. Estimates interpreted as .2 = small, .5 = medium, .8 = large.

*
p < .05