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Problem behaviour in adolescent boys with fragile X syndrome: relative prevalence, frequency and severity

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Abstract

Background—A large proportion of boys with fragile X syndrome (FXS), the most common known inherited form of intellectual disability (ID), exhibit problem behaviours (e.g. aggression, self-injury, property destruction and stereotypy) that can negatively impact the health and safety of others as well as the individual concerned. However, data are limited concerning the relative prevalence, frequency and severity of problem behaviours exhibited by boys with FXS compared with those by boys with mixed-aetiology ID who also exhibit problem behaviours.

Method—As part of a larger study on problem behaviour, we obtained survey data on 85 adolescent boys with FXS and 155 age-matched boys with mixed-aetiology ID who exhibited at least one form of problem behaviour.

Results—For boys with FXS, stereotypy was reported to be more prevalent ($\chi^2 = 4.52$, $P = 0.012$), self-injury was reported to more frequent ($U = 2525$, $P = 0.010$) and aggression was reported to be less severe ($U = 4176$, $P = 0.029$) than for boys with mixed-aetiology ID. Ratings of aggression and property destruction were highly correlated in each group in terms of both frequency and severity ($r = 0.60$ to 0.71). Examination of the data by age indicated that the relative frequency of self-injury decreased with age in boys with FXS ($\chi^2 = 8.29$, $P = 0.040$).

Conclusions—Taken together, these results refine and extend previous studies concerning the specificity of the behavioural phenotype in FXS and indicate that specific forms of problem behaviour shown by boys with FXS appear to differ from those exhibited by boys with mixed-aetiology ID in terms of prevalence, frequency and severity. Studies employing more objective measures of frequency and severity, including direct observations, are needed to confirm these findings.

Keywords

aggression; behavioural measurement methods; behavioural phenotypes; destructive behaviour; fragile X; self-injury; stereotypy

Introduction

A significant proportion of individuals diagnosed with fragile X syndrome (FXS), the most common known form of inherited ID, exhibit a variety of behavioural issues such as autistic

behaviours, sensory issues, anxiety and hyperarousal, as well as specific problem behaviours such as aggression, self-injury, property destruction and stereotypy (Symons *et al.* 2003; Hall *et al.* 2008; Hessler *et al.* 2008; Langthorne & McGill 2012). These behaviours can negatively impact the health and safety of the individual concerned and others (Wheeler *et al.* 2016). FXS is a genetic condition caused by mutations to the *FMR1* gene on the long arm of the X chromosome at Xq27.3, resulting in significantly reduced FMRP expression, the protein product of the gene (Verkerk *et al.* 1991). Given that problem behaviours in FXS occur at higher prevalence rates than would be expected for individuals with ID in general (Moss *et al.* 2009; Arron *et al.* 2011), it seems likely that genetic factors may confer increased risk for problem behaviours in this syndrome (Hessler *et al.* 2008; Hall 2009).

To date, the vast majority of previous studies involving individuals with FXS have employed questionnaire/informant ratings of problem behaviours (Hall *et al.* 2008; Hessler *et al.* 2008; Newman *et al.* 2015; Wheeler *et al.* 2016). These studies have indicated that a high proportion of individuals with FXS, particularly male participants, show multiple forms of problem behaviour. For example, in a sample of 47 children and adolescents with FXS who showed problem behaviour, aged 2 to 17 years, Newman *et al.* (2015) reported that 85% engaged in aggression, 80% engaged in self-injury, and 100% engaged in stereotypy, with 72% engaging in all three behaviours. Similarly, in a sample of 50 male participants with FXS, aged 8 to 24 years, Hessler *et al.* (2008) reported that 75% of the sample engaged in aggression, 79% engaged in self-injury and 98% showed stereotypic behaviour.

In terms of behaviour frequency and severity, several studies have documented the impact that problem behaviour can have on caregivers. In a sample of 570 male participants with FXS, aged 3 to 67 years, Wheeler *et al.* (2016) reported that aggression had caused injuries to caregivers in 30% of cases and had caused injuries to peers or friends in 22% of cases. In the majority of cases (70%), however, caregivers reported the behaviour to be mild, with 23% exhibiting moderate aggression and only 7% of cases exhibiting severe aggression. Similarly, in a sample of 489 male participants with FXS who were reported to show self-injury (mean age = 15.9 years), Symons *et al.* (2010) reported that the behaviour was rated as mild in 65% of cases, moderate in 29.9% and severe in only 4.6% of cases. In terms of behaviour frequency, however, 39% were reported to show self-injury on a daily basis, with 32.2% exhibiting the behaviour weekly, 9.6% monthly and 19.2% less than once a month. In the study conducted by Hessler *et al.* (2008), 33% were reported to exhibit aggressive behaviour on a daily basis, 33% on a weekly basis and 33% on a monthly basis. The average frequency of self-injury reported in the sample was weekly, while the modal frequency was daily. Finally, of those who showed stereotypy, the average frequency reported was daily, with 21% exhibiting hourly, 59% exhibiting daily, 14% exhibiting weekly and 6% exhibiting monthly stereotypic behaviour. Although these studies suggest that problem behaviour shown by boys with FXS can occur on a frequent basis with mild-to-moderate severity, it is unknown whether the relative prevalence, frequency and severity of problem behaviour in boys with FXS is equivalent to those displayed by boys with ID who also show problem behaviour but who do not have FXS, given that previous studies have not included a comparison group of individuals with mixed-aetiology ID.

In the present study, therefore, we recruited boys with FXS and boys with mixed-aetiology ID (but not FXS) who were reported to exhibit at least one form of problem behaviour and compared the data obtained on the relative prevalence, frequency and severity of problem behaviour in the two groups. To enhance the objectivity of the severity ratings, we included definitions of what constituted 'mild', 'moderate' and 'severe' and included an option to rate severity as 'not a problem'. Given the wide age range of participants included in previous studies, we limited the age range to boys aged 11 to 18 years in order to focus on this critical developmental period. We had three main questions: First, are particular forms of problem behaviour exhibited by boys with FXS more prevalent, frequent and/or severe than those exhibited by boys with mixed-aetiology ID? Second, to what extent do problem behaviours co-occur in each group? Finally, does the relative prevalence, frequency and/or severity of problem behaviour increase with age in either group?

Method

Participants

As part of a larger study investigating the determinants of problem behaviour in adolescents with FXS, we posted a series of advertisements via the National Fragile X Foundation, online parent support groups and social media inviting caregivers of children with FXS and intellectual and developmental disabilities who engaged in problem behaviour to participate in the study. The announcement stated:

We are conducting a study to evaluate how environmental and biological factors affect the display of severe problem behaviour (e.g., aggression, self-injury, etc.) in adolescents with fragile X syndrome. To determine your child's eligibility for the study, please complete the brief survey by clicking on the arrow at the bottom of the page.

An identical survey that stipulated adolescents with intellectual and developmental disabilities was also posted. Completed surveys were obtained on 116 male participants with FXS and 245 male participants with ID, aged 2 to 61 years, as well as 11 female participants with FXS and 49 female participants with ID. Given that the focus of the study concerned adolescent male participants, we excluded data on female participants as well as data obtained on male participants who were above or below the adolescent age range (defined here as 11 to 18 years) in each group. The final study sample therefore consisted of 85 boys with FXS and 155 boys with mixed-aetiology ID (but not FXS) who were aged 11 to 18 years and showed at least one form of problem behaviour.

Measures

Caregivers in each group first completed basic demographic information about their child's sex and age and whether their child had an additional diagnosis of a genetic disorder, significant motor disability or neurological disorder (if known). Caregivers were then required to indicate whether their child displayed at least one or more of the following forms of problem behaviour: aggression (e.g. hitting, kicking, punching and throwing items at others), self-injury (e.g. head-hitting, self-scratching and self-biting), property destruction (e.g. breaking items, pushing over tables or chairs) or stereotypy (e.g. hand flapping, body

rocking and spinning objects repetitively). Caregivers in each group then rated the frequency and severity of each form of problem behaviour using questions derived from Part 1 of the Functional Analysis Screening Tool (FAST; Iwata *et al.* 2013). Response options for frequency of occurrence were the following: 'never', 'less than weekly', 'weekly', 'daily' or 'hourly'. Response options for severity were 'not a problem', 'minor (i.e. is disruptive, but results in little risk to property or health)', 'moderate (i.e. results in property damage or minor injury)' or 'severe (i.e. results in a significant threat to health or safety)'.

Data analysis—All data analyses were conducted using SPSS 20 (SPSS Inc., Chicago, IL, USA). To examine differences between the groups in terms of the relative prevalence, frequency and severity of problem behaviour in each group, Pearson chi-square difference tests were used for binary data and Mann–Whitney *U* tests for ordinal data. Spearman rank-order correlations were conducted to examine the extent to which the behaviours co-occurred in each group. Finally, to determine whether the frequency and/or severity of problem behaviour increased with age, we conducted Kruskal–Wallis tests for each form of problem behaviour within each group. In all analyses, the alpha value for statistical significance was set at 0.05.

Results

The mean age of boys in the FXS group was 14.02 years ($SD = 2.29$ years), and the mean age of boys in the control group was 13.73 ($SD = 2.07$ years), a non-significant difference between the groups [$t(238) = 1.01$, $P = 0.312$]. In boys with FXS, caregivers reported the following additional diagnoses: ADHD (four cases), ASD (15 cases), PDD-NOS (one case), Duarte Galactosemia (one case), epilepsy (two cases) and haemophilia (one case). In the control group, additional diagnoses reported included the following: Down syndrome (two cases), Dubowitz syndrome (one case), Prader–Willi syndrome (three cases), Sotos syndrome (one case), septo-optic dysplasia (one case), 9p deletion syndrome (one case), 1q21.1 deletion syndrome (one case), ADHD (six cases), ASD (34 cases), shaken baby syndrome (one case), Tourette syndrome (two cases), cerebral palsy (four cases), dysgenesis of the corpus callosum (one case), epilepsy (five cases) and OCD (one case).

Relative prevalence

Table 1 shows the percentage of cases reported to show each form of problem behaviour in each group.

In boys with FXS, the relative prevalence of problem behaviour was highest for stereotypy (90.6%) and lowest for property destruction (62.4%). Pearson chi-square tests indicated that the relative prevalence of stereotypy was significantly higher in boys with FXS than in controls. Conversely, the relative prevalence of property destruction and aggression was significantly lower in boys with FXS than in controls. In boys with FXS, 42.4% showed all four forms of problem behaviour, 29.4% showed three forms, 20% showed two forms and only 8.2% showed one form. In the controls, 50.3% showed all four forms of behaviour, 32.9% showed three forms, 11.6% showed two forms and 5.2% showed one form. There were no differences between the groups in terms of the number of forms of problem behaviour displayed, Mann–Whitney $U = 5780$, $P = 0.09$.

Relative frequency

Figure 1 (left panel) shows a breakdown of the frequency ratings for each form of problem behaviour by group.

For boys with FXS, the most frequent form of problem behaviour reported was stereotypy, with 20.8% exhibiting the behaviour hourly, 53.2% daily, 13% weekly and 13% less than weekly. The least frequent form of problem behaviour reported was property destruction, with 0% exhibiting the behaviour hourly, 18.9% daily, 35.8% weekly and 45.3% less than weekly. Overall, the median frequency rating for aggression, self-injury and property destruction was weekly, whereas the median frequency rating for stereotypy was daily. Statistical comparison of the groups indicated that the relative frequency of self-injury was higher for boys with FXS than for controls ($U = 2525$, $P = 0.010$). There were no differences between the groups in terms of the relative frequency of aggression, property destruction and stereotypy.

Relative severity

Figure 1 (right panel) shows a breakdown of the severity ratings for each form of problem behaviour by group. Again, the data show that the severity of each form of problem behaviour was relatively similar between the groups. For boys with FXS, the most severe form of problem behaviour reported was aggression, with 18.6% being rated as severe, 32.9% moderate, 41.4% mild and 7.1% not a problem. Property destruction was also rated as a severe form of problem behaviour, with 11.3% rated as severe, 43.4% moderate, 41.5% mild and 3.8% as not a problem. The least severe form of problem behaviour reported was stereotypy, with 0% rated as severe, 13% moderate, 62.3% mild and 24.7% as not a problem. Overall, the median severity rating for aggression and property destruction was moderate, indicating that these behaviours usually resulted in property damage or minor injury, whereas the median severity rating for self-injury and stereotypy was mild indicating that these behaviours were considered to be disruptive but resulted in little risk to property or health. Statistical comparison of the groups indicated that the relative severity of aggression was significantly lower in boys with FXS than in controls ($U = 4176$, $P = 0.029$). There were no differences between the groups in terms of the reported relative severity of self-injury, property destruction and stereotypy.

Co-occurrence of problem behaviour forms

Table 2 shows Spearman correlations obtained between each form of problem behaviour for each group.

For each group, the upper correlation matrix corresponds to the correlations obtained between the frequency ratings, and the lower correlation matrix corresponds to the correlations obtained between the severity ratings. In terms of behaviour frequency, the ratings between aggression and property destruction were significantly positively correlated (FXS group: $r(85) = 0.656$, $P < 0.001$; controls: $r(155) = 0.595$, $P < 0.001$). Smaller but significant positive correlations were also obtained for the frequency ratings between self-injury and stereotypy in each group (FXS group: $r(85) = 0.328$, $P < 0.001$; controls: $r(155) = 0.287$, $P < 0.001$). In terms of severity, the ratings between aggression and property

destruction were also highly positively correlated (FXS group: $r(85) = 0.709$, $P < 0.001$; controls: $r(155) = 0.639$, $P < 0.001$). However, the association between self-injury and stereotypy was significant only in the control group for the severity ratings ($r(155) = 0.316$, $P < 0.001$).

Association with age

Figure 2 shows a breakdown of the frequency ratings for each behaviour in each group across 2-year age bands.

Kruskal–Wallis tests were conducted to examine whether there was an effect of age on the frequency ratings within each group. These analyses indicated that for both groups, there was a significant effect of age on the frequency of self-injury (FXS group: $\chi^2(3) = 8.29$, $P = 0.040$; controls: $\chi^2(3) = 9.43$, $P = 0.024$). Post-hoc analyses indicated that the frequency ratings were significantly lower for boys with FXS aged 17 to 18 years than for boys with FXS aged 11 to 12 years ($P = 0.008$). Conversely, the frequency ratings were significantly higher for boys with mixed-aetiology ID aged 13 to 14 years than for boys with mixed-aetiology ID aged 11 to 12 years ($P = 0.016$) and boys with mixed-aetiology ID aged 15 to 16 years ($P = 0.01$). There were no other effects of age on the frequency ratings for the other forms of problem behaviour in either group.

Figure 3 shows the corresponding breakdown of the severity ratings for each behaviour in each group across 2-year age bands.

Kruskal–Wallis tests were conducted to examine whether there was an effect of age on the severity ratings within each group. No significant results were found in either group.

Discussion

The primary aim of the present study was to determine whether problem behaviours shown by adolescent boys with FXS occurred with higher relative prevalence, frequency and/or severity than those of age-matched boys with mixed-aetiology ID who also showed problem behaviours. To accomplish this aim, we invited caregivers of boys in each group who showed at least one form of problem behaviour to rate the occurrence, frequency and severity of their child's behaviour using an online survey tool. Therefore, only adolescent boys from both groups who showed problem behaviour were included in the present study. Overall, the results for boys with FXS broadly supported previous findings indicating that some forms of problem behaviour (e.g. stereotypy and self-injury) occurred at a relatively high frequency in FXS but that in the majority of cases, these behaviours were considered to be relatively 'mild' (i.e. were disruptive, but resulted in little risk to property or health). Conversely, other forms of problem behaviour (e.g. aggression and property destruction) were reported to occur less frequently but were considered to be 'moderate' (i.e. resulted in property damage or minor injury). These data support the need to measure the frequency and severity of multiple forms of problem behaviour. Indeed, approximately 42% of boys with FXS included in the present sample showed all four forms of problem behaviour and approximately 30% of participants showed three forms of problem behaviour. When we examined the extent to which these behaviours were associated, high positive correlations

were obtained between aggression and property destruction in both frequency and severity domains. These data suggest that aggression and property destruction most likely form a response class that may serve the same purpose or function. Smaller but significant positive correlations were also obtained between the frequency of self-injury and stereotypy in each group, suggesting that self-injury and stereotypy may also be linked together in the child's repertoire (Guess & Carr 1991).

Comparison of the data across groups indicated that the relative frequency of self-injury was significantly higher in boys with FXS than in controls but that the relative severity of aggression was lower in boys with FXS than in controls. Given that the relative severity of self-injury was found not to differ between the groups, these data indicate that when self-injury occurs in boys with FXS, it does so more frequently, but at the same level of severity as controls. Similarly, given that the relative frequency of aggression was found not to differ between the groups, these data indicate that when aggression occurs in boys with FXS, it occurs at the same frequency but at a lower severity than in controls.

There are several strengths of the study that should be mentioned. First, to our knowledge, this is the first study to examine the relative prevalence, frequency and severity of problem behaviours shown by boys with FXS compared with those shown by boys with mixed-aetiology ID. Given that both groups of individuals showed at least one form of problem behaviour, comparison to a control group of boys with boys with mixed-aetiology ID allowed us to examine the specificity of the behavioural phenotype in FXS. As discussed by Hodapp and Dykens (2001), inclusion of a mixed-aetiology control group is preferable to an 'idiopathic' control group of individuals with ID because an idiopathic ID control group includes only those with unknown causes of ID, whereas a mixed-aetiology control group includes individuals with unknown causes of ID as well as known causes of ID. Thus, a mixed-aetiology control group is more representative of the general population of individuals with ID (Hodapp & Dykens 2001). Second, we focused our analyses on the adolescent period because anecdotal reports from caregivers indicated that adolescence may be a particularly problematic period for behavioural management, given the increasing height and weight of their child. In boys with mixed-aetiology ID, for example, we found that the frequency of self-injury was increased for individuals aged 13 to 14 years compared with the other age groups. We did not, however, find any increase in the frequency and/or severity of problem behaviour across age bands in either group for the forms of problem behaviour and in fact, there was a significant decrease in the frequency of self-injury with age in children with FXS. However, this finding should be tempered by the fact that this was a cross-sectional study, not a longitudinal study. In a previous longitudinal study using the Aberrant Behaviour Checklist (Aman & Singh 1994), we found that severity ratings of problem behaviour shown by boys with FXS also decreased over time (Hustyi *et al.* 2014). Whether this represents a true decrease or whether caregivers become inured or desensitised over time to behaviour problems is open to question.

There were several limitations of the study that also should be mentioned. First, the data were collected using a screening tool that relied upon caregiver reports of problem behaviour occurrence, frequency and severity. Although the majority of other studies have collected data in this way, caregiver reports are subject to recall bias, and it would be important to

validate the data by obtaining objective ratings of these behaviours in the child's natural setting. Second, we did not obtain estimates of the child's developmental level that could help explain the frequency and/or severity of problem behaviours in the two groups. Thus, although the groups were matched on chronological age, it is possible that the two groups may not have been matched on developmental age. Third, the sample was drawn from caregivers of individuals with ID or FXS who showed problem behaviour via online parent support groups, social media and the National Fragile X Foundation. Thus, the sample was self-selected, and caregivers of individuals who typically do not engage with social media and/or are not involved in support groups may have been less likely to participate in the study. It is also possible that the presence/absence of autism spectrum disorder could have impacted the frequency and/or severity of problem behaviours in the two groups.

In the present study, we resisted the temptation to generate a total problem behaviour score. Many investigators, for example, employ instruments that routinely assign numbers to the ordinal categories (e.g. 1 = less than weekly, 2 = weekly, 3 = daily and 4 = hourly) and then generate a total score by summing the numbers across forms of problem behaviour. As discussed by Rojahn *et al.* (2012), this implies that a total score is interpretable on an ordinal scale, which may not necessarily be the case. For example, a child who engages in a single form of problem behaviour on an hourly basis would not necessarily be equivalent to a child who engages in four forms of problem behaviour that each occurs on a less-than-weekly basis, yet both children would receive a total problem behaviour score of 4. Clearly, this is a significant issue in the measurement of problem behaviours that needs to be resolved.

In summary, stereotypy was reported to be more prevalent, self-injury was reported to be more frequent and aggression was reported to be less severe in boys with FXS than in boys with mixed-aetiology ID. Given that we attempted to enhance the objectivity of the severity ratings by including definitions of what constituted a mild, moderate or severe problem, we were somewhat surprised by the latter finding. One explanation for this finding concerns whether caregivers of boys with FXS may view aggression simply as part of the behavioural phenotype of FXS and therefore have a less severe perception of their child's behaviour than caregivers of boys with ID in general. Future research should involve direct observations of these behaviours and attempts to measure the impact of these behaviours more objectively (e.g. whether the behaviours actually result in broken furniture, cuts and bruises on the caregivers or on the individual concerned). In a previous study examining the impact of self-injury shown by individuals with Prader-Willi syndrome, for example, we employed the Self-Injury Trauma Scale (Iwata *et al.* 1990) to directly quantify the extent to which their behaviour produced tissue damage to the individual concerned (Hustyi *et al.* 2013). Specifically, we asked caregivers to note the number of wounds on the child's body and to provide severity ratings for the wounds they had observed using specific objective criteria. Similar measures could therefore be employed for individuals with FXS to provide more objective ratings of the frequency and severity of problem behaviour in the future.

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Figure 1.

Relative frequency and severity of problem behaviours shown by boys with fragile X syndrome (FXS) and controls.

Figure 2.
Relative frequency of problem behaviours across age for boys with fragile X syndrome (FXS) and controls.

Figure 3. Relative severity of problem behaviours across age for boys with fragile X syndrome (FXS) and controls.

Table 1

Relative prevalence of problem behaviours reported in each group

Form of problem behaviour	FXS (<i>N</i> = 85) (%)	Controls (<i>N</i> = 155) (%)	$\chi^2(1)$	<i>P</i>
Aggression	82.4	92.9	6.33	0.012
Self-injury	70.6	70.3	0.002	NS
Property destruction	62.4	85.2	16.17	<0.001
Stereotypy	90.6	80.0	4.52	0.033

Table 2

Spearman correlations obtained between forms of problem behaviour in each group

	Aggression	Self-injury	Property destruction	Stereotypy
FXS group ($N = 85$)				
Aggression	1.00	0.059	0.656**	0.114
Self-injury	0.295**	1.00	0.144	0.328**
Property	0.709**	0.411**	1.00	0.043
Stereotypy	0.039	0.188	0.106	1.00
Controls ($N = 155$)				
Aggression	1.00	0.252**	0.595**	0.138
Self-injury	0.226**	1.00	0.164	0.287**
Property	0.639**	0.148	1.00	0.127
Stereotypy	0.111	0.316**	0.047	1.00

For each group, the upper correlation matrix corresponds to correlations between the frequency ratings and the lower correlation matrix corresponds to correlations between the severity ratings.

**
 $P < 0.01$.