Research in Developmental Disabilities xxx (xxxx) xxx–xxx

Contents lists available at ScienceDirect

Research in Developmental Disabilities

journal homepage: www.elsevier.com/locate/redevdis

Review article

How common are challenging behaviours amongst individuals with Fragile X Syndrome? A systematic review

Rebecca Lyndsey Hardiman*, Peter McGill

Tizard Centre, University of Kent, United Kingdom

ARTICLE INFO

Number of completed reviews is 2

Keywords:
Fragile X Syndrome
Self injurious behaviour
Aggression
Property destruction
Problem behaviour
Challenging behaviour
Intellectual disability
Learning disability
Genetic syndromes
Behavioural phenotypes

ABSTRACT

Fragile X Syndrome (FXS) appears to be associated with an increased risk for engaging in challenging behaviour, particularly self-injury, relative to those with mixed aetiology learning disabilities. Such behavioural issues are reported to be of high concern for those providing support. As such, this systematic review aimed to gain further epidemiological data regarding challenging behaviours in individuals with FXS, including: self-injurious behaviour (SIB), hand-biting as a specific topography of SIB, aggression and property destruction. Twenty eight manuscripts were identified which reported the prevalence of a relevant topography of behaviour, with widely varying prevalence estimates. Weighted averages of the prevalence of behaviours were calculated across studies. Comparison of proportions revealed significant gender differences and differences in the prevalence of types of behaviour. It is hoped that this comprehensive overview of data on this clinically significant topic will help to inform and drive future investigation to understand and provide effective intervention for the benefit of those with FXS.

What this paper adds?

This paper adds a systematic and comprehensive overview of the prevalence of challenging behaviours in individuals with Fragile X Syndrome. These behaviours are likely to have a negative impact upon the individuals themselves and those who support them. As such a deeper knowledge of the frequency of these behaviours is required in order to assist with the assessment of risk at the population level, to help to facilitate planning of service provision for these individuals and to contribute towards understanding of these behaviours, with an aim of developing effective support and intervention.

1. Introduction

Some individuals with intellectual disabilities engage in behaviour which challenges those around them. Between 10–20% of individuals with intellectual disabilities have been described to engage in such behaviours (Jacobson, 1982; Kiernan & Kiernan, 1994), which most commonly include self-injurious behaviour, aggression and property destruction (Emerson et al., 2001). A number of risk markers have been identified for engagement in challenging behaviours, including: expressive communication deficits (McClintock, Hall, & Oliver, 2003), as well as co-morbid conditions, such as autism and epilepsy (Smith & Matson, 2010). In addition, an increasing body of evidence demonstrates that the likelihood of engaging in challenging behaviour relates to the genetic aetiology of an individual's intellectual disability (for instance: Arron, Oliver, Moss, Berg, & Burbidge, 2011).

Fragile X Syndrome (FXS) is the most common inherited cause of intellectual disability (Mazzocco, 2000) and a genetic condition

* Corresponding author at: Hardiman Tizard Centre, Cornwallis North East, University of Kent, Cornwallis North East, Canterbury CT2 7NF, United Kingdom.
E-mail address: Rh432@kent.ac.uk (R.L. Hardiman).

https://doi.org/10.1016/j.ridd.2018.02.020
Received 3 February 2017; Received in revised form 28 December 2017; Accepted 26 February 2018

in which challenging behaviours are frequently reported (for instance: Hessl et al., 2008). The condition is caused by a CGG triplet expansion on the FMR1 gene, located on the long arm of the X chromosome. As a result of this expansion, the gene typically becomes methylated, resulting in cessation or suppression of its protein product FMRP, which is important in many aspects of development and brain function (Santoro, Bray, & Warren, 2012; Verkerk et al., 1991). As a result of the X-linked nature of the condition, females show more variable effects and, on average, show less clear effects. The widespread effects of the genetic mutation are associated with a phenotype including varying degrees of intellectual disability, anxiety, attention deficits and autistic-like behaviour (Bailey, Raspa, Olmsted, & Holiday, 2008; Cordeiro, Ballinger, Hagerman, & Hessl, 2010).

Self-injurious behaviour, aggression and destructive behaviour have all been described in individuals with this condition (for example: Hessl et al., 2008). Indeed, hand-biting has been described as part of the behavioural phenotype (Hagerman et al., 1992). It is reported by clinicians and parents that SIB and aggressive outbursts in FXS are often associated with sensory stimulation or unexpected change, which leads to the individual feeling overwhelmed and, in turn, hyperaroused (Miller et al., 1999) and stressed (Hessl et al., 2008). This has led to a suggestion that changes to the physiology of the stress response may be associated with the operant conditioning of challenging behaviours with an escape function, within this group (Hardiman & McGill, 2017; Langthorne & McGill, 2012; Langthorne et al., 2011). In addition, a number of characteristics commonly associated with FXS have been identified as risk factors for engagement in challenging behaviour, for people with intellectual disabilities (McClintock et al., 2003). These characteristics include: autism (Oliver, Berg, Moss, Arron, & Burbidge, 2011), over-activity and impulsivity (Baumgardner, Reiss, & Freund, 1995). The heightened presence of these risk factors in FXS, as well as the syndrome-specific factors discussed, highlights several possible associations between FXS and challenging behaviours.

Parents and carers report that the problem behaviour of their children with FXS (such as, aggressive behaviours) create a significant caregiving burden: physically, emotionally and financially (Bailey et al., 2012). Accordingly, in interviews with parents, behavioural problems are rated as being of greater concern than cognitive delays (Hatton et al., 2000). Furthermore, for adults with FXS, the presence of mental health problems, including SIB and aggressiveness, is associated with lower independence, poorer employment outcomes, fewer friendships (for women) and greater assistance required in everyday life (Hartley et al., 2011). Given the significance of such behaviours, for both individuals with FXS and their families, it is important to gain an in-depth understanding regarding these behaviours, including epidemiology, risk factors and interventions. Although there have been a number of studies investigating the prevalence of challenging behaviours in FXS, the results of individual studies vary widely, making them difficult to interpret. Understanding the prevalence of challenging behaviours across individuals with FXS aids with understanding the needs of, and planning services for, these individuals. As such, a systematic review of the literature was warranted in order to collate the findings on this important topic, allowing better description of the prevalence of such challenges in FXS, and to inform future discussion and research. Therefore, the aims of this review were to address the following questions:

1. What proportion of individuals with FXS engage in self-injurious behaviour, physical aggression or destructive behaviours?
2. Are there gender differences in the prevalence of each of these types of behaviours?

2. Methods

2.1. Inclusion criteria

For the purposes of this review, the challenging behaviour types were defined broadly as follows:

- Self-injurious behaviour (SIB): behaviour towards the self that has caused, or may cause, physical harm. Hand-biting was included as a specific topography where assessed in isolation in a study, given its association with FXS. This was defined as pressure being applied between the teeth to any part of the individual's own hands or fingers.
- Aggression: physically aggressive behaviour directed towards another person.
- Destructive behaviour: aggressive or damaging behaviour directed towards an individual's environment, such as objects or furniture.

Published data with sample sizes of 10 or more individuals reported to have FXS were included; where more detailed information about genetic status was provided, both full-mutation and mosaic cases were included. In addition, in order to be included, the studies were required to have sufficient data to calculate a percentage prevalence of either SIB (including hand-biting), aggression or property destruction. The presence of challenging behaviour was rated according to the data available in the article: both ratings on single items of questionnaires or interviews, and borderline or clinically significant scores on relevant subscales were included (see Table 1 for assessment measures). If the use of a measure which may have generated data relevant for this review was listed, but the results not provided in sufficient detail for inclusion, authors were contacted to request further details (where data were obtained in this fashion, this is noted in Table 1). There were no limits on the dates of publication for inclusion.

2.2. Literature search

An electronic search of four databases (PsychINFO, PubMed, Web of Science, and SCOPUS) using a string including the terms "Fragile X Syndrome" (including variants, plus Medical Subject Headings (MeSH): Martin Bell or Escalante) and "Challenging Behaviour" (including variants, plus MeSH: problem behaviour, behaviour problems, maladaptive behaviour, aberrant behaviour,