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Review article

The prevalence of aggression in genetic syndromes: A review

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ABSTRACT

Research into behavioural phenotypes identifies both environmental and organic factors as influencing aggression in children and adults with genetic disorders associated with intellectual disability. However, in contrast to self-injury there is a paucity of research that compares aggression across relevant syndromes. The primary aim of this review is to examine the association between aggression and genetic syndromes by analysis of prevalence studies. The review also examines the literature on the form of the behaviour and influence of environmental factors.

Results imply that certain syndrome groups (Cri du Chat, Smith-Magenis, Prader-Willi, Angelman, Cornelia de Lange, and Fragile X syndromes; estimates over 70%) evidence a stronger association with aggression than others (e.g. Williams and Down syndromes; estimates below 15%). However, the strength of association is difficult to quantify due to methodological differences between studies. The results from examining form and environmental influences highlight the importance of phenotype–environment interactions. Research employing group comparison designs is warranted and future work on the assessment and intervention of aggression in genetic syndromes should consider the importance of phenotype–environment interactions.

Contents

1. Method . . . 1052
 1.1. Selecting syndromes for inclusion . . . 1052
  1.1.1. Search strategy . . . 1052
 1.2. Syndromes included for review . . . 1053
  1.2.1. Cri du Chat syndrome . . . 1053
  1.2.2. Smith-Magenis syndrome . . . 1053
  1.2.3. Fragile X syndrome . . . 1054
  1.2.4. Angelman syndrome . . . 1055
  1.2.5. Cornelia de Lange syndrome . . . 1055
  1.2.6. Prader-Willi syndrome . . . 1055
  1.2.7. Williams syndrome . . . 1055
  1.2.8. Down syndrome . . . 1055
2. Results . . . 1055
 2.1. Prevalence of aggression in selected syndromes . . . 1055
  2.1.1. Comparison of prevalence across genetic syndromes . . . 1055

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2.1.2. Methodological influences on prevalence estimates . . . 1062
2.2. Comparison of prevalence across syndrome groups . . . 1063
2.3. Interim summary . . . 1064
2.4. Form of aggression and the influence of environmental factors . . . 1064
2.4.1. Form . . . 1064
2.4.2. Summary and considerations . . . 1064
2.4.3. Environmental influences . . . 1064
2.4.4. Summary and considerations . . . 1066
3. Discussion . . . 1066
Acknowledgements . . . 1068
References . . . 1068

Aggression in individuals with intellectual disability (ID) impinges on quality of life, carer well being, and contributes to the breakdown of residential placements (Hastings, 2002; Tausig, 1985). Prevalence estimates for aggression in ID vary widely, in part because of methodological differences (Borthwick-Duffy, 1994; Harris & Russell, 1989; Quine, 1986; Sigafoos, Elkins, Kerr & Atwood, 1994). A recent review of prevalence studies of aggression in ID that limited inclusion to those reporting 'physical aggression', suggested rates of physical aggression in ID are likely to lie at the upper end of the widely cited 2–20% estimate (Davies & Oliver, 2013). For example, Tyrer et al. (2006) and Crocker et al. (2006) examined prevalence in over 3000 individuals with ID and reported overall rates of 14% and 24.4% respectively. Similarly, Smith, Branfield, Collacot, Cooper, and McGrother (1996) reported a prevalence of 22% in 2202 adults with ID. Relative risk analyses of studies reporting prevalence across age groups has indicated that aggression increased with age until mid-adulthood (Davies & Oliver, 2013).

A robust and diverse research literature highlights the importance of environmental factors generally, and operant theory specifically, in the development and maintenance of aggression. It has been demonstrated repeatedly in experimental studies that such behaviours can be sensitive to, and maintained by socially, and non-socially mediated forms of reinforcement such as attention or the presentation of tangible items from carers (Carr & Durand, 1985), and removal of task demands (Iwata, Pace, Kalsher, Cowdery, & Cataldo, 1990). In a review of functional analytic studies, Hanley, Iwata, and McCord (2003) demonstrated that in 50 of 52 studies, aggression was mediated by an operant reinforcement process.

Although operant theory has significant empirical support, there is a broad consensus that biological factors also play a role in behaviours such as self-injury and aggression (e.g. Arron, Oliver, Moss, Berg, & Burbridge, 2011; Langthorne & McGill, 2012; May et al., 2009). Certain syndrome groups evidence a comparatively higher prevalence of self-injury, aggression, and destructive behaviour than others, and forms of behaviour also differ across genetic syndromes (Arron et al., 2011).

In addition to syndrome related associations, certain person characteristics are known to be associated with aggression. McClintock, Hall, and Oliver (2003) found that Autism Spectrum Disorder (ASD), Attention-Deficit/Hyperactivity Disorder (ADHD), being male, and reduced communication skills were all associated with aggression. Within genetic syndromes, Arron et al. (2011) found that impulsivity and over-activity were significantly higher in participants showing aggression than in those who did not. Furthermore, in studies examining risk factors for aggression in ASD, lower IQ, poorer expressive and receptive language, and repetitive behaviours have been identified as associated with aggression (Dominick, Davis, Lainhart, Tager-Flusberg, & Folstein, 2007; Kanne & Mazurek, 2011).

Evidence from the operant and behavioural phenotype literatures suggest that both environmental and organic factors may play a role in the manifestation of aggression in genetic syndromes (see Oliver et al., 2013; Tunnicliffe & Oliver, 2011). However, there is a paucity of research that directly compares aggression across syndromes, and this is in contrast to the literature on self-injury. This lack of research is surprising given that many of the risk factors known to be associated with aggression (i.e. impulsivity, over-activity, repetitive behaviours, ASD, and reduced communication abilities) are frequently described in certain genetic syndromes (e.g. Clarke & Boer, 1998; Finucane, Konar, Haas-Givler, Kurtz, & Scott, 1994; Hagerman, 2002; Oliver, Berg, Burbidge, Arron, & Moss, 2011). Identification of increased risk for aggression would facilitate the implementation of early intervention strategies to reduce or replace behaviours before they become established. Furthermore, as it has been suggested that successful interventions require knowledge of underlying operant influences (Harvey, Boer, Meyer, & Evans, 2009) it is necessary to investigate the role of environmental influences on aggression across syndromes.

This review will examine the extent to which aggression is associated with specific genetic syndromes by analysis of studies that report the prevalence of aggression in these groups. This will ascertain whether certain syndromes show a heightened association with aggression in comparison to others. The review will then examine the literature outlining the form of the behaviour in these groups, and literature that examines the influence of environmental factors on aggression.

1. Method

1.1. Selecting syndromes for inclusion

1.1.1. Search strategy

Due to the number of syndromes that might potentially be investigated, screening was undertaken to identify which syndrome groups were reported in research papers relevant to the review. A search using Ovid PsychInfo was conducted on