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Prevalence, phenomenology, aetiology and
predictors of challenging behaviour in Smith-
Magenis syndrome

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Abstract

Background: The prevalence, phenomenology aetiology and correlates of four forms of challenging behaviour in 32 children and adults with Smith-Magenis syndrome (SMS) were investigated.

Method: Cognitive assessments, questionnaires and semi-structured interviews were used to gather data on intellectual disability, verbal and physical aggression, destructive behaviour and self-injury and on characteristics known to be associated with aggression.

Results: Aggression in SMS was more prevalent (87%), but not more severe, than aggression in contrast groups. Aggressive behaviour was more frequently associated with environmental contingencies (e.g. attention, escape and access to tangibles) than self-injury and destructive behaviours. Severity of challenging behaviours was associated with high impulsivity.

Conclusion: Aggression is seen in the majority of people with SMS. Results suggest that behavioural disinhibition and operant social reinforcement are associated with the manifestation of aggression.

Keywords: Smith Magenis syndrome, aggression, self-injurious behaviour, aggressive behaviour, impulsivity, behavioural phenotype, social reinforcement, Autism Spectrum Disorder.

Introduction

Smith-Magenis syndrome (SMS) is caused by an interstitial deletion or a heterozygous point mutation at 17p11.2p11.2 encompassing the retinoic acid induced 1(RAI1) gene (Smith *et al.*, 1986; Girirajan, Elsas, Devriendt & Elsea, 2005; Slager *et al.*, 2003). SMS is characterised by moderate to severe intellectual disability, speech and language delay (Greenberg *et al.*, 1991; Udwin, Webber & Horn, 2001) and a constellation of physical and cognitive characteristics, together with specific behaviours that, arguably, form part of the 'behavioural phenotype'; (Allanson, Greenberg & Smith, 1999; Dykens & Smith, 1998; Smith, Dykens & Greenberg, 1998; Arron *et al.*, In press, Oliver *et al.*, In press).

Prevalence figures for several forms of challenging behaviour in people with SMS are high. Estimates for physical aggression range from 38% to 93% (Madduri, Turcich & Lupski, 2002; Webber, 1999) with most reports citing figures above 70% (e.g. Colley, Leversha, Voullaire, & Rogers, 1990; Dykens *et al.*, 1993, 1997; Horn, 1999; Arron *et al.*, In press). Prevalence figures for self-injury are higher and range from 70% to 97% (Greenberg *et al.*, 1991; Greenberg *et al.*, 1993; Dykens and Smith, 1998; Finucane, Dirrigl & Simon, 2001; Arron *et al.*, In press). These figures contrast with those for groups with intellectual disabilities of mixed aetiology, where rates fall below 30% (e.g. Borthwick-Duffy, 1994; Deb *et al.*, 2001, Emerson *et al.*, 1997, 2001; Eyman & Call, 1977; Hill & Bruininks, 1984; Smith, Branford, Collacott, Cooper & McGrother, 1996). Given the high prevalence rates further research is warranted.

Case reports suggest that forms of aggressive behaviours in SMS are similar to those seen in mixed aetiology intellectual disability. These include hitting, punching, head banging, self-biting and destroying property (Colley *et al.*, 1990; Crumley, 1998; Finucane *et al.*, 1993; Finucane *et al.*, 1994; Finucane *et al.*, 2001; Greenberg *et al.*, 1991; Hagerman, 1999; Smith *et al.*, 1986; Stratton *et al.*, 1986). However, several case reports also describe aggressive behaviours that are unusual such as poking others' eyes (Finucane *et al.*, 1994), forceful hugging (Smith *et al.*, 1998) and punching fists through walls and windows (PRISMS, 2004), and rare forms of self injury such as onychotillomania (pulling finger and toe nails out) and polyembolokoilomania (insertion of foreign objects into bodily orifices).

The aetiology of aggression in SMS has yet to be systematically investigated. Evidence supporting the hypothesis that aggression in SMS has an exclusively biological cause is limited and comprises case reports of pharmacological interventions (Crumley, 1998; Hagerman, 1999; Smith *et al.*, 1998; Smith & Gropman, 2001). Similarly, there is limited anecdotal evidence to suggest that environmental contingencies such as photic stimuli, transitions and aversive stimuli shape and maintain aggression in people with SMS (Smith *et al.*, 1998; Haas-Givler & Finucane 2000; Smith *et al.*, 1986; Smith *et al.*, 1998). However, the observational study of Taylor and Oliver (2008) did provide evidence for self-injury and aggression in SMS being maintained by contingent attention and the authors suggest that this reward might be potent in this syndrome, and thus the function common, given the propensity for children and adults with SMS to seek adult contact (Moss *et al.*, 2009). This possibility warrants examination.

A number of 'risk markers' are associated with challenging behaviours in individuals with intellectual disabilities of mixed aetiology. These include Autism Spectrum Disorder,

impaired cognitive ability, communication impairment and impulsiveness (McClintock, Hall & Oliver, 2003; Arron et al., In press). Anecdotal reports, case studies, and large scale systematic investigations indicate that over 80% of people with SMS evidence high rates of impulsivity (Dykens, Finucane & Gayley, 1993, 1997; Dykens & Smith, 1998; Dykens, Hodapp & Finucane, 2000; Clarke & Boer, 1998; Oliver et al., In press). Although the mechanisms by which impulsivity might influence aggression are unclear, it seems likely that as the phenotype of Smith-Magenis syndrome encompasses this 'risk marker' then this might be predictive of the presence and severity of challenging behaviour.

In summary, the existing literature suggests that individuals with SMS have a heightened probability of exhibiting aggressive and impulsive behaviours. However, it is uncertain whether or not the presentation and aetiology of aggression in people with SMS is unusual. There is evidence that environmental contingencies, specifically positive operant reinforcement by contingent attention, might maintain aggression and self-injury but there has been no large scale evaluation of this hypothesis. Finally, the relationship between impulsivity and aggression in SMS warrants examination. The aims of this study are to investigate the prevalence and phenomenology of aggressive behaviour in SMS and their association with environmental events and examine the relationship between aggression and impulsivity.

Method

Recruitment and Participants

Families were contacted via the Smith-Magenis Syndrome Foundation (UK based support group) to recruit participants into a multi-syndrome survey (see Oliver et al., In

press). Information packs were sent to those families caring for individuals diagnosed with SMS aged over six who had consented to take part in further research (N = 40). Thirty-two of the families contacted participated in the research. All carers reported that participants had been diagnosed with SMS by medical professionals following genetic tests. Table 1 displays the characteristics of the participants.

+++ Insert Table 1 here +++

Measures

Primary carers completed a number of questionnaires and acted as informants for standardised interviews. Additionally, participants with SMS were directly assessed by a researcher using the Wechsler Intelligence Scales for Children (cognitive assessment) and the Childhood Autism Rating Scales (observation). Detailed information on the measures can be found below.

Demographic information A brief demographic questionnaire was used to gather information about the characteristics of informants and participants (such as age, gender, relationship to participant, age at diagnosis).

Challenging Behaviour

The Checklist for Challenging Behaviour (CCB; Harris, 1993; Harris Humphreys, & Thomson, 1994) is a two-part questionnaire (Harris *et al.*, 1994; Joyce, Ditchfield and Harris, 2001) completed by carers to ascertain the frequency, management difficulty and severity of fourteen topographies of physical aggression (e.g. ‘pinching people’, ‘biting people’) and

eighteen other challenging behaviours (e.g. 'eating inappropriate things' and 'spitting at people').

The Challenging Behaviour Interview (CBI) (Oliver, McClintock, Hall, Smith, Dagnan & Stenfert-Koese, 2003) is a two-part interview which assesses the incidence and severity of challenging behaviour. Interviewees identify whether a behaviours has been displayed in the past month. Fourteen questions then determine the severity of each behaviour, for example, questions cover frequency, damage caused and necessary restraint. Physical aggression, destructive behaviour and self-injury total scores were summed to ascertain a total severity score. Higher scores denote greater severity.

Function of Aggression

The Questions about Behavioural Function (QABF) (Matson & Vollmer, 1995) is a 25 question tool used to explore associations between challenging behaviour and five types of environmental events that have been associated with behaviour difficulties in people with intellectual disabilities: 1) self-stimulation 2) demand escape 3) access to tangibles 4) attention and 5) relief of pain or discomfort. A 'total function score' is obtained and mean total function scores for the five functions may be used to determine which functions are more prominent for which behaviours (Applegate, Matson & Cherry, 1999). The higher the score for a given function, the more likely it is that the challenging behaviour has that function.

Assessing characteristics associated with aggression

Cognitive functioning

The Wechsler Intelligence Scale for Children - third edition (Wechsler, 1991) and Wechsler Adult Intelligence Scale –third edition (Wechsler, 1997) were used to assess cognitive functioning in participants. The lowest IQ scores on the Wechsler scales fall within the range of severe intellectual disability (IQ 20 to 40), thus a proportion of participants with severe and profound impairment scored at the basal level. These participants were given a nominal score of 20. In a minority of children and adults who were uncooperative with the Wechsler Intelligence Scales (N=6), the VABS-SF was used as a measure of the level of ability (see below). Dykens *et al.* (1997) found a high correlation between IQ and Vineland composite standard scores in SMS.

Communication

The Vineland Adaptive Behavior Scales – Interview edition, Survey Form (VABS-SF) (Sparrow, Balla & Chicchetti, 1984) measures personal and social adaptive behaviour levels divided into four domains: daily living skills, communication, social ability and motor skills. It is suitable for use with carers of individuals with intellectual disability. The communication domain has 67 items and is divided into three sub domains (receptive, expressive and written). Standard and age equivalent scores may be calculated for each domain and the composite score, whilst age-equivalents are calculated for each sub-domain. High levels of reliability have been established (Sparrow *et al.*, 1984).

Sleep disturbance

The Infant Sleep Questionnaire (ISQ), Morrell, (1999) is a ten-item questionnaire that assesses sleep problems for clinical and research purposes. It is designed for use with young children, but has been used in research with older participants (Sadler *et al.* 2000). The single item relating to ‘sleeping in carer’s bed’ was removed from the scoring and carers of

participants of all ages completed the questionnaire. An overall sleep score may be obtained by summing the scores from questions in part one and higher scores denote increased disturbance.

Behaviours associated with Autism Spectrum Disorder

The Childhood Autism Rating Scale (Schopler, Reichler, DeVellis, & Daly, 1986) is a brief rating scale that was used to assess autistic type behaviour in participants. It focuses on fourteen dimensions regarding particular characteristics, abilities and behaviours and is completed by the investigator after a period of observation. A total score is computed by summing individual ratings (out of 60) and may be used to denote the degree to which individuals displayed 'autistic type behaviours'.

Hyperactivity

The Conners' Parent Rating Scale Revised; Long version (Conners, 1997) is an 80-item questionnaire commonly used in clinical settings to screen for Attention Deficit/Hyperactivity Disorder (ADHD). The three items relating to verbal behaviour were removed and subscales were prorated for nonverbal individuals (rated on the Vineland Adaptive Behaviour Scales expressive communication domain as aged 30 months or below).

Repetitive Behaviour

The Repetitive Behaviour Questionnaire (Moss et al., 2009) is a 19 item questionnaire designed for use with people with intellectual disability to explore the frequency of repetitive behaviours. The 19 items comprise five subscales: stereotyped behaviour, rule governed behaviour, insistence on sameness, restricted interests and repetitive use of language.

Impulsivity

The DEX (Wilson *et al.*, 1996) and the DEX-C (Emslie, Wilson, Burden, Nimmo-Smith & Wilson, 2003) are two versions of the same twenty-item informant-based questionnaire which sample a range of problems commonly associated with the Dysexecutive syndrome. It measures impulsivity in the areas of emotion and personality, motivation, behaviour and cognitive ability. The questionnaires form part of the child and adult versions of the Behavioural Assessment of the Dysexecutive Syndrome (Emslie *et al.*, 2003; Wilson, Alderman, Burgess, Emslie & Evans, 1996 respectively). Items on the DEX/DEX-C may be summed to provide an overall executive dysfunction score ranging from 0 to 80. The two items requiring individuals to speak in full sentences were removed and the total score was prorated for nonverbal individuals.

Procedure

Testing was carried out directly with participants in schools, day centres or homes prior to or following completion of carer interviews and questionnaire packs. Six participants (18.7%) refused to participate in assessment using the Wechsler scales. In these instances, the full Vineland was administered to the parents to obtain a level of functioning for the individual with SMS.

Data Analysis

In order to ascertain whether or not individuals with SMS are at increased risk of showing aggressive behaviour, the percentage of individuals showing different types of aggressive behaviour in the present sample of individuals with SMS was compared to previously published rates of aggressive behaviour shown by individuals with intellectual disabilities of mixed aetiologies. A hand and electronic search was undertaken to identify

research papers that investigated the prevalence and phenomenology of aggression in populations of individuals with intellectual disabilities of mixed aetiology. Twenty studies that recruited large samples ($N > 100$) were selected and reviewed (see Appendix A). These papers were chosen because they have been frequently cited, employed varied methodology and used samples of people with a range of cognitive abilities. In order to make a conservative estimate of risk, the highest of these published prevalence rates of aggression were used for comparison with the individuals with SMS. These figures were: 54% for physical aggression (Davidson *et al.*, 1996; 707 children, mild-profound disability, <22 years), 48.7% for self-injury (Kobe *et al.*, 1994; 203 non-ambulatory individuals with severe and profound disability, 6 months-73 years) and 25.9% and 39.3% for verbal aggression and destruction respectively (Eyman & Call, 1977; 1827 individuals with mild-profound disability living in a hospital environment).

In addition to the previous studies selected for the large mixed intellectual disability samples they employed, two previous studies administered measures that were used in the present study, providing direct points of comparison. Joyce *et al.* (2001) reported the use of the CCB in a sample of 448 adults over the age of 19 with intellectual disabilities, and Oliver *et al.* (2003) reported the use of the CBI in a sample of 40 adults (aged 17-58 years) with moderate-severe intellectual disabilities and 47 children (aged 4-12) with severe intellectual disabilities. Comparison of the present results on the CCB and CBI with these retrospective data (using binomial tests and one sample t-tests respectively) allowed comparisons of the prevalence rates of different topographies of aggressive behaviour and of the and severity of aggressive behaviours between individuals with SMS and those with intellectual disability of mixed aetiologies.

The functions of aggressive behaviour in SMS were investigated using the QABF. Results from this measure were analysed using a series of repeated measures ANOVAs with a single within-subjects factor (function subscale), to assess possible differences in the proportions of each form of aggressive behaviour being associated with the five different functions assessed.

Possible associations between the different measures of impulsivity employed were assessed using Pearson's correlations. The relationships between scores for the overall severity of aggressive behaviour as measured by the CBI (sum of physical aggression, destruction and self-injurious behaviour CBI severity scores; see *Measures*) in these individuals with SMS and age, sleep disturbance, cognitive ability, receptive and expressive communication, hyperactivity, impulsivity, autistic type behaviour and repetitive behaviour (known risk factors for challenging behaviour) were examined using Pearson's and Spearman's correlations. Throughout the analysis alpha levels were adjusted by application of the Bonferroni correction in order to reduce the chances of type-one errors.

Results

Phenomenology of Aggressive behaviour in Smith-Magenis Syndrome

Prevalence of aggression in SMS

Based on responses to the Challenging Behaviour Interview 96.9% (31) of participants displayed self-injurious behaviour, 87.5% (28) exhibited physical aggression, 81.3% (26) destructive behaviour and 43.8% (14) were verbally aggressive. Using Binomial tests the prevalence figures of different forms of aggression seen in the SMS group were compared to the highest prevalence figures found for challenging behaviour reported in the literature in

people with mixed aetiological intellectual disabilities (see methods section). Results showed that self-injurious behaviour, physical aggression and destructive behaviour were all significantly more prevalent in individuals with SMS compared to those with intellectual disabilities of mixed aetiologies ($p < .001$). Although verbal aggression was more prevalent in individuals with SMS relative to those with intellectual disabilities of mixed aetiologies, this effect did not reach significance following a Bonferroni correction ($p = .022$).

Topographies of physical aggression in SMS

The mean number of topographies of physical aggression displayed by participants, as yielded by the Checklist for Challenging Behaviour, was seven (range 1-13, SD 3.56). Across the whole sample of individuals with SMS hitting and grabbing were the most prevalent topographies of aggression (shown by >80% of individuals), with biting, kicking and pinching also shown in more than half of the individuals (see Table 2). A comparison of the prevalence rates of the aggressive behaviours measured by the CCB in individuals over the age of 19 with and without SMS was possible using data published previously by Joyce et al. (2001; see *Methods*). Binomial tests were used to compare the prevalence rates of aggressive behaviours in a sub sample of individuals with SMS over the age of 19 ($n=8$) and these previously published rates shown by a large group of individuals with intellectual disabilities of mixed aetiologies (see Table 2). These analyses revealed that in individuals over the age of 19 there was a significantly higher prevalence rate of hitting and biting in those with SMS than in those without the syndrome.

+++ Insert table 2 here+++

Frequency and severity of aggression in SMS

Frequency, management difficulty and severity scores for fourteen topographies of physical aggression were ascertained using the Checklist of Challenging Behaviour. The mean item frequency score for the SMS group was 3.5 (range 2.29 – 5.00, SD .69) (where 1-never, 2-rarely, 3-occasionally, 4-often, 5-very often). The mean item management difficulty score was 2.63 (range 1.14-4.00, SD .63) (where 1-no problem, 2 -slight problem, 3-moderate problem, 4-considerable problem, 5-extreme problem) and the mean item severity score was 1.93 (range 1.00 – 3.00, SD .62) (1-no injury, 2 -minor injury, 3-moderate injury, 4-serious injury, 5-very serious injury).

Frequency and severity scores were also obtained using the CBI. For the present sample of individuals with SMS, the median scores for the CBI items relating to frequency of physical aggression, verbal aggression, destruction and self injury were all 3.0, indicating that, on average, the informant reported that they would definitely next see the behaviour by ‘this time tomorrow’, (but not as often as in the next hour). The CBI severity scores in the present sample of individuals with SMS were compared to those shown by individuals with intellectual disability due to mixed aetiologies using the data from the Oliver et al. (2003) study (see Methods). Using the age bands described by Oliver et al. (children: 4-12 years; adults: 17-58 years), the present sample was divided into the same child (n=15) and adult (n=17) groups. A series of one sample t-tests was conducted to compare severity scores yielded in child and adult participant groups in the present study to the scores reported by Oliver *et al.* (2003). This analysis revealed no significant differences in the severity of aggressive behaviour shown by individuals with SMS and those with intellectual disability due to mixed aetiologies.

Function of Aggressive Behaviours in Smith-Magenis Syndrome

Each form of aggressive behaviour was explored in relation to the five functions of challenging behaviour that the QABF assesses: self-stimulation, demand escape, access to tangibles, attention and relief of pain or discomfort. Results are shown in Table 3. For both physical aggression and verbal aggression, the attention subscale received the highest total score, followed by the escape tangible, then pain and discomfort and finally self-stimulation. In contrast, for both self-injury and destructive behaviour, self-stimulation yielded the highest totals followed by attention, then escape. For self injury this was followed by tangible and finally pain and discomfort, whilst in the case of destructive behaviour pain and discomfort yielded higher totals than the tangible function.

+++Insert Table 3 here+++

A series of repeated measures ANOVAs with a single within-subjects factor (function subscale) was conducted, to test for differences between the functions of each form of behaviour. There were significant (to the adjusted level of $p < .01$) main effects of function in the data for physical aggression ($F(4,108)=13.74, p < .01$) and verbal aggression ($F(4,52)=9.14, p < .01$).

Post hoc Bonferroni adjusted pairwise comparisons revealed that significantly more physical and verbal aggression was related to an attention function than either to a self stimulatory function (PA: $t(27)=6.46, p < .001$; VA: $t(13)=5.67, p < .001$) or being related to pain and discomfort (PA: $t(27)=3.92, p = .001$; VA: $t(13)=4.52, p = .001$). There was also significantly more physical and verbal aggression associated with an escape function than either a self stimulatory function (PA: $t(27)=6.30, p < .001$; VA: $t(13)=3.19, p = .007$) or pain

and discomfort (PA: $t(27)=4.40$, $p<.001$; VA: $t(13)=4.81$, $p<.001$). Finally, there was significantly more physical aggression associated with access to tangibles than with either self stimulation ($t(27)=6.30$, $p<.001$) or pain and discomfort ($t(27)=6.30$, $p<.001$). Thus, both physical and verbal aggression were more frequently associated with social communicative functions (attention, escape from demands, access to tangibles) than with non-communicative functions.

Phenomenology of Impulsive Behaviour in Smith-Magenis Syndrome

Pearson's correlations were undertaken to examine the association between the DEX/DEX-C and the Conners' Parent Rating Scale. The DEX/DEX-C total scores correlated strongly with two of the Conners' Parent Rating Scale indices, DSM-IV index of hyperactive impulsive behaviour ($r(31) = .77$, $P<.001$) and the global restless impulsive index ($r(31) = .72$, $p<.001$). The mean DEX/DEX-C score was 53.17 (Range 17-75, SD – 15.57).

Correlates and predictors of aggressive behaviour

Pearson's and Spearman's correlations were undertaken to investigate whether or not the severity of aggressive behaviour (sum of severity scores for physical aggression, destruction and self-injurious behaviour on the CBI; see *Methods*) in SMS was correlated with the nine variables that have been associated with challenging behaviour the previous literature including in individuals with intellectual disabilities (see *Introduction*). These variables were age, sleep disturbance (ISQ overall score), cognitive ability, receptive and expressive communication, hyperactivity, impulsivity, autistic type behaviour and repetitive behaviour (see Table 4). The severity of aggressive behaviour showed moderate strength relationships (according to Landis and Koch's (1977) criteria) with hyperactivity (Conner's

ADHD index) and autistic type behaviours (CARS total score), relationships which both attained significance, and with degree of cognitive impairment (WISC IQ/VABS SS), although this later relationship did not attain statistical significance. In addition to these moderate strength relationships, there was a substantial positive association between the severity of aggressive behaviour and impulsivity (DEX/DEX-C total score).

Discussion

This study is the first to report a systematic investigation of the phenomenology and operant functions of challenging behaviour and the relationship between challenging and impulsive behaviours in individuals with Smith-Magenis syndrome. The results support and extend the findings of previous studies that describe increased prevalence of challenging behaviours and impulsivity in SMS and a relationship between challenging behaviours and environmental events, more specially contingent attention. The present study also found that impulsive behaviours (as measured by the DEX/DEX-C) are strongly related to challenging behaviour.

The prevalence data for self-injurious behaviour (96.9%) and physical aggression (87.5%) demonstrate that these two forms of behaviour are displayed by the vast majority of people with SMS. These findings are consistent with the high prevalence reported in previous studies (Colley *et al.*, 1990; Finucane *et al.*, 2001; Webber, 1999; Arron *et al.*, In press). High proportions of people also showed destructive behaviour and verbal aggression in SMS, (81.3% and 43.8% respectively). The prevalence rates of self-injurious behaviour, physical aggression and destructive behaviour were found to be significantly higher in participants

with SMS than in groups of people with intellectual disabilities of mixed aetiology described in the previous literature. These previously described groups were selected for comparison to the present SMS group because of particularly high prevalence rates of corresponding aggressive behaviours, which were higher than the published rates on other samples of individuals with intellectual disability of mixed aetiologies. The commonly accepted definition of phenotypic behaviours suggests that behaviours should be more prevalent in individuals with a specific genetic syndrome (i.e. SMS) than in individuals without that syndrome. Thus, these prevalence data and comparisons with carefully selected previous rates strengthen the assertion that these aggressive behaviours form part of the behavioural phenotype of Smith-Magenis syndrome.

Frequency and severity was investigated in order to assess the clinical relevance of the aggressive behaviour shown in people with SMS. In terms of frequency, all four forms of behaviour were found to occur, on average, on a daily basis. There is no doubt that this poses difficulties for those caring for individuals with the syndrome. However, Hill and Bruininks (1984) and Kebbon and Windahl (1986) reported that self-injury and destructive behaviour in people with intellectual disabilities of mixed aetiologies occurred on average at a daily to weekly basis, suggesting that while SMS appears to be associated with particularly high prevalence rates of aggressive behaviour, the frequency of this behaviour in each individual may be similar in individuals with SMS to in those without the syndrome.

When comparing severity of aggression between the participants with SMS and individuals with intellectual disability of mixed aetiology reported in the literature (Oliver *et al.*, 2003) using the Challenging Behaviour Interview no differences were found. Therefore, although all aggressive behaviours are more prevalent in individuals with SMS when

compared to people without the syndrome, there is no clear evidence that aggression, once manifested, is more severe in people with SMS.

The present study sought to describe the topographies of physical aggression shown by individuals with SMS. Across children and adults, most common topographies of aggression were hitting and grabbing (more than three quarters of the present sample) and biting, kicking and pinching were also very common (more than half of the sample). In individuals over the age of 19 comparison was possible with previously published data and individuals with SMS showed significantly more hitting and biting than individuals with intellectual disability of mixed aetiologies. Hitting and grabbing were the most common topographies of physical aggression in people with and without SMS, however biting was one of the least common aggressive behaviours in the mixed intellectual disability sample. Thus it appears that elevated rates of common forms of aggressive behaviours form part of the SMS behavioural phenotype, alongside high rates of aggressive behaviours not prevalent among individuals without the syndrome (such as biting). It may be that people with SMS display a wider repertoire of physically aggressive behaviours than people without the disorder.

Informants completed questionnaires to establish the specific functions that behaviours serve for individuals with SMS. Findings suggest that the aetiology of aggression in SMS is comparable to that seen in groups of people without the syndrome, supporting the hypothesis that operant factors are likely to play a role in the manifestation of the behaviour in people with SMS. For a relatively large proportion of participants, physical and verbal aggression were reported to be related to positive reinforcement through attention and this replicates and extends the findings of Taylor and Oliver (2008). It has been reported elsewhere that preference for being with adults is a notable feature of SMS (Moss et al., 2009) and this

would be consistent with this operant function. In a number of people, these behaviours were also related to escape from aversive situations and access to tangible items. Previous studies have shown similar functions in physical aggression in those with mixed aetiological intellectual disabilities. (Applegate *et al.*, 1999; Emerson and Bromley, 1995).

In contrast, it was found that for both self-injury and destruction, self-stimulation yielded the highest total function score. This suggests that these two behaviours may be maintained by sensory reinforcement or that operant factors are not influential. In addition to sensory stimulation, in a significant number of people, self-injury and destructive behaviours were also associated with the functions of attention, access to tangibles and escape from aversive situations. It is therefore possible that these behaviours additionally serve a communicative function in people with SMS. Both Emerson and Bromley (1995) and Applegate *et al.* (1999) found the same pattern in samples of people with mixed aetiological intellectual disabilities. It is important to note that the QABF employed here is restricted in the possible functions of behaviours that can be identified. The measure was used in the present study in line with previous research with people with intellectual disabilities. However, it is possible that some behaviours shown by individuals with SMS can be associated with different functions, possibly some which are idiosyncratic to individuals with SMS (for example a preference for being with adults as discussed above).

The current research aimed to investigate factors that may be associated with aggression in SMS. There was a near universal occurrence of the aggressive behaviour in the present sample, thus correlations between the *severity* of aggressive behaviours and risk marker characteristics were investigated. In contrast to what may be expected from the existing literature, results suggested that age, gender, sleep disturbance, level of

communication and repetitive behaviours were *not* significantly associated with the severity of any forms of aggression in people with SMS. The relationship between the severity of aggressive behaviour and the degree of cognitive impairment was associated with a medium effect size but this relationship did not attain significance in the present sample.

However, the overall severity of aggression in participants with SMS was significantly related to: hyperactivity, autistic type behaviours (moderate strength relationships) and impulsivity (substantial relationship). These findings are consistent with other prevalence studies and research investigating risk markers of aggression in groups of people with intellectual disabilities of mixed aetiology and genetic syndromes (e.g. Emerson, 1998; McClintock, Hall and Oliver, 2003; Arron *et al.*, In press). These results also suggest that impulsivity and aggression may be associated in SMS, as they have been thought to be in people without the syndrome (King, 1993; Petty & Oliver, 2005; Rojahn *et al.*, 2004; Swann, 2003; Swann & Hollander, 2002).

It is notable that the association between impulsivity and aggression in SMS was substantial and stronger than the associations between aggression and hyperactivity or autistic type behaviours. This supports the idea that impulsivity is an important factor in the manifestation of aggression in SMS and has implications when considering what intervention may be useful for people within this population. Intervention may include the use of medication to address impulsivity (although efficacy of medication to reduce impulsivity in SMS has yet to be established) and/or the development of self-regulation through the use of behavioural techniques and linguistic tools (Whitman, 1990).

An important limiting factor in the present study is the use of data from published studies retrospectively in order to compare aggression in individuals with SMS to those with intellectual disabilities due to mixed aetiologies. Including a comparison group in the present study would have allowed exact matching of method, measures and demographic characteristics across the samples, which would have made each comparison more informative. However, the focus in the present study was on highlighting that individuals with SMS are at very high risk for showing aggressive behaviour. Groups of individuals with intellectual disabilities due to mixed aetiologies are inherently heterogeneous, thus it would be difficult to control for all potentially confounding variables with a single comparison group. By reviewing a sample of good quality studies reporting on large sample sizes with a range of cognitive abilities and other demographic characteristics, we were able to select the highest of these published prevalence rates of aggressive behaviour in individuals with intellectual disabilities due to mixed aetiologies. In this way the present comparison provides a stringent test of the hypothesis that SMS will be associated with a greater prevalence of aggression than intellectual disability due to mixed aetiology. The support that was demonstrated for this hypothesis therefore emphasises the particular clinical relevance of understanding aggression in SMS.

It must also be noted that the comparisons reported with data from the Oliver et al. (2003) and Joyce et al. (2001) papers relating to the severity and phenomenology of aggressive behaviours in SMS, whilst benefiting from shared measures with the present study, could potentially be confounded by differences in demographic variables between the samples. However, the samples were matched for age all included individuals with a range of degrees of intellectual disability. Given the inherent difficulties associated with matching

it is unlikely that a comparison group recruited specifically for the present study would have provided a better match for the SMS sample.

The findings generate a number of further research questions. At present performance tests for impulsivity have not been standardised for use in the severely intellectually disabled population consequently there is reliance on impulsivity scores from informant based questionnaires. Use of direct behavioural tests of impulsivity and the link between these and aggression may be worth investigating further. It is important to note that the association between aggression and impulsivity in SMS does not imply causality. Although the association falls in line with the literature on risk markers for challenging behaviour (discussed above), it does not rule out the possibility that aggression in SMS may cause impulsivity or that a third variable is underpinning this association. The link between aggression and impulsivity needs to be explored to help to further understanding of the underlying aetiology of aggression in SMS.

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Table 1

Demographic characteristics of participants

Age	Mean= 15.09 years; range= 6 to 39 years; <i>SD</i> = 8.79			
Place of residence	84.4% (n=27) lived at home with parents 15.6% (n=5) lived in residential care			
Gender	43.8% (n=14) male			
	Severity Range	Score on Assessment	Percentage of Participants	Number of Participants
Cognitive impairment¹	mild	55- 69	12.5	4
	mild-moderate	50-54	15.6	5
FSIQ/	moderate	40-49	28.1	9
VABS standard score	severe- profound	< 40	43.8	14
Communication impairment	mild	55- 69	12.5	4
	mild-moderate	50-54	9.4	3
VABS standard score	moderate	40-49	15.6	5
	severe	35-39	3.1	1
	moderate- severe	24-34	31.3	10
	severe- profound	20-24	3.1	1
	profound	<20	25	8
	not verbal	-	46.9	15

¹ Where possible, cognitive impairment was ascertained using the Wechsler Intelligence Scales Full Scale IQ scores (WISC-III^{UK} and WAIS-III). For participants who were uncooperative, the The Vineland Adaptive Behaviour Scales interview edition, survey form was used instead. [Dykens *et al.* (1997) found a high correlation between IQ and Vineland composite standard scores in individuals with SMS.]

Table 2

Binomial tests comparing prevalence of topographies of aggression in adults aged 19 years or above with and without SMS.

	Percentage of SMS sample displaying the behaviour (n=32)	Percentage of SMS sample > 19 years of age displaying the behaviour (n = 8)	Percentage of mixed ID group (Joyce et al. 2001) displaying behaviour (n=448)	P value for comparison between individuals > 19 years old with and without SMS
Hitting	84	100	49	.003*
Grabbing	84	87.5	56	.070
Kicking	59	75	26	.005
Pinching	59	50	21	.066
Biting	50	50	9	.003*
Pulling hair	41	25	17	.406
Using objects as weapons	38	37.5	13	.074
Head butting	31	12.5	4	.279
Choking or throttling	25	25	5	.057
Throwing things at people	47	62.5	27	.038
Scratching	28	25	26	.653

* A Bonferroni correction was applied and effects associated with a *p* value of < .004 were considered significant. Effects marked with an asterisk are significant to this level.

Table 3

Mean total function scores derived for each form of aggression evaluated

Subscale	Physical aggression (N = 28)	Verbal aggression (N = 14)	Self-injury (N 31)	Destruction (N = 25)
Attention				
M (SD)	7.04 (5.04)	9.07 (4.01)	5.74 (5.48)	6.64 (5.3)
Escape				
M (SD)	6.96 (4.32)	7.14 (3.94)	4.29 (4.38)	8.56 (3.95)
Self-stimulation				
M (SD)	1.93 (2.73)	3.36 (4.67)	6.97 (5.26)	6.84 (5.09)
Pain and discomfort				
M (SD)	3.07 (3.95)	2.64 (4.47)	3.16 (4.43)	2.64 (3.94)
Tangible				
M (SD)	6.57 (4.42)	5.21 (5.01)	4.87 (4.70)	4.24 (4.68)

Table 4

Correlations between total severity of aggression scores (sum of physical aggression, destruction and self-injurious behaviour severity scores as measured with the CBI) and potential predictor variables

Variable	Pearson's <i>r</i> and <i>p</i> values
Age	$r = -.01, p = .931$
Sleep score (total ISQ score)	$r = .36, p = .044$
Intellectual ability (WISC IQ/VABS standard composite score)	$r = -.12, p = .516$
Vineland receptive communication score	$r = -.46, p = .008$
Vineland expressive communication score	$r = -.46, p = .009$
Hyperactivity (Conners' ADHD index)	$r = .56, p < .005 *$
Impulsivity (DEX-/DEX-C total score)	$r = .72, p < .001 *$
CARS total score	$r = .53, p < .005 *$
RBQ total score	$r = .24, p = .202$

* Correlation coefficients are marked with an * that are significant to a corrected level of $p < .005$ (following a Bonferroni correction).