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.
Methods   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.

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Introduction

Smith-Magenis syndrome (SMS) is caused by an interstitial deletion or a heterozygous point mutation at 17p11.2 encompassing the retinoic acid induced 1 gene (Smith *et al.* 1986; Slager *et al.* 2003; Girirajan *et al.* 2006). SMS is characterised by moderate to severe intellectual disability (ID), speech and language delay (Greenberg *et al.* 1991; Udwin *et al.* 2001) and a constellation of physical and cognitive characteristics, together with specific behaviours that, arguably, form part of the 'behavioural phenotype' (Dykens & Smith 1998; Smith *et al.* 1998; Allanson *et al.* 1999; Arron *et al.* 2010; 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% (Webber 1999; Madduri *et al.* 2002) with most reports citing figures above 70% (e.g. Colley *et al.* 1990; Dykens *et al.* 1993, 1997; Horn 1999; Arron *et al.* 2010). Prevalence figures for self-injury are higher and range from 70 to 97% (Greenberg *et al.*

1991, 1993; Dykens & Smith 1998; Finucane *et al.* 2001; Arron *et al.* 2010). These figures contrast with those for groups with IDs of mixed aetiology, where rates fall below 30% (e.g. Eyman & Call 1977; Hill & Bruininks 1984; Borthwick-Duffy 1994; Smith *et al.* 1996; Emerson *et al.* 1997, 2001; Deb *et al.* 2001). 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 ID. These include hitting, punching, head banging, self-biting and destroying property (Smith *et al.* 1986; Stratton *et al.* 1986; Colley *et al.* 1990; Greenberg *et al.* 1991; Finucane *et al.* 1993, 1994, 2001; Crumley 1998; Hagerman 1999). 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 polyembolokoilamania (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; Smith *et al.* 1998; Hagerman 1999; 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.* 1986, 1998; Haas-Givler & Finucane 2000). However, the observational study of Taylor & 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 IDs of mixed aetiology. These include Autism Spectrum Disorder, impaired cognitive ability, communication impairment and impulsiveness (McClintock *et al.* 2003; Arron *et al.* 2010). Anecdotal reports, case

studies and large-scale systematic investigations indicate that over 80% of people with SMS evidence high rates of impulsivity (Dykens *et al.* 1993, 1997, 2000; Clarke & Boer 1998; Dykens & Smith 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 SMS 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 multisyndrome survey (see Oliver *et al.* in press). Information packs were sent to those families caring for individuals diagnosed with SMS aged over 6 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.

Measures

Primary carers completed a number of questionnaires and acted as informants for standardised interviews. Additionally, participants with SMS

Table 1 Demographic characteristics of participants

	Severity range	Score on assessment	Percentage of participants	Number of participants
Age	Mean = 15.09 years; range = 6 to 39 years; SD = 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			
Cognitive impairment*	Mild	55–69	12.5	4
FSIQ/VABS standard score	Mild–moderate	50–54	15.6	5
	Moderate	40–49	28.1	9
	Severe–profound	<40	43.8	14
Communication impairment	Mild	55–69	12.5	4
VABS standard score	Mild–moderate	50–54	9.4	3
	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

* 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 Vineland Adaptive Behaviour Scales (VABS) 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.] FSIQ, Full-Scale Intelligence Quotient; SD, standard deviation.

were directly assessed by a researcher using the Wechsler Intelligence Scales for Children (cognitive assessment) and the Childhood Autism Rating Scales (CARS) (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 *et al.* 1994) is a two-part questionnaire (Harris *et al.* 1994; Joyce *et al.* 2001) completed by carers to ascertain the frequency, management difficulty and severity of 14 topographies of physical aggression (e.g. 'pinching people', 'biting people') and 18 other challenging behaviours (e.g. 'eating inappropriate things' and 'spitting at people').

The Challenging Behaviour Interview (CBI) (Oliver *et al.* 2003) is a two-part interview which assesses the incidence and severity of challenging

behaviour. Interviewees identify whether a behaviour has been displayed in the past month. Fourteen questions then determine the severity of each behaviour, e.g. 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 IDs: (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 *et al.* 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 cogni-
tive functioning in participants. The lowest IQ
scores on the Wechsler scales fall within the range
of severe ID (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 chil-
dren and adults who were uncooperative with the
Wechsler Intelligence Scales ($n = 6$), the Vineland
Adaptive Behavior Scales – Interview edition,
Survey Form (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 VABS-SF (Sparrow *et al.* 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 ID.
The communication domain has 67 items and is
divided into three sub-domains (receptive, expres-
sive and written). Standard and age equivalent
scores may be calculated for each domain and the
composite score, whilst age-equivalents are calcu-
lated 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 10-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 (Sadeh 2004). The single item relating
to 'sleeping in carer's bed' was removed from the
scoring and carers of participants of all ages com-
pleted 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
(CARS; Schopler *et al.* 1986) is a brief rating scale
that was used to assess autistic type behaviour in
participants. It focuses on 14 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 dis-
played '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.
The three items relating to verbal behaviour were
removed and sub-scales were prorated for non-
verbal individuals (rated on the VABS expressive
communication domain as aged 30 months or
below).

Repetitive behaviour. The Repetitive Behaviour
Questionnaire (Moss *et al.* 2009) is a 19-item ques-
tionnaire designed for use with people with ID to
explore the frequency of repetitive behaviours. The
19 items comprise five sub-scales: stereotyped
behaviour, rule governed behaviour, insistence on
sameness, restricted interests and repetitive use of
language.

Impulsivity. The Dysexecutive Questionnaire
(DEX) (Wilson *et al.* 1996) and the Dysexecutive
Questionnaire for Children (DEX-C) (Emslie *et al.*
2003) are two versions of the same 20-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 Syn-
drome (Wilson *et al.* 1996; Emslie *et al.* 2003,
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 non-verbal
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 with previously published rates of aggressive behaviour shown by individuals with IDs 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 IDs of mixed aetiology. Twenty-two studies that recruited large samples ($n > 100$) were selected and reviewed (see Appendix 1). 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 ID 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 IDs, and Oliver *et al.* (2003) reported the use of the CBI in a sample of 40 adults (aged 17–58 years) with moderate–severe IDs and 47 children (aged 4–12) with severe IDs. 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 ID 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 sub-scale), 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 Smith-Magenis syndrome

Based on responses to the CBI 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 with the highest preva-

Table 2 Binomial tests comparing prevalence of topographies of aggression in adults aged 19 years or above with and without Smith-Magenis syndrome (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	0.003*
Grabbing	84	87.5	56	0.070
Kicking	59	75	26	0.005
Pinching	59	50	21	0.066
Biting	50	50	9	0.003*
Pulling hair	41	25	17	0.406
Using objects as weapons	38	37.5	13	0.074
Head butting	31	12.5	4	0.279
Choking or throttling	25	25	5	0.057
Throwing things at people	47	62.5	27	0.038
Scratching	28	25	26	0.653

* A Bonferroni correction was applied and effects associated with a *P*-value of <0.004 were considered significant. Effects marked with an asterisk are significant to this level.

lence figures found for challenging behaviour reported in the literature in people with mixed aetiological IDs (see *Methods*). Results showed that self-injurious behaviour, physical aggression and destructive behaviour were all significantly more prevalent in individuals with SMS compared with those with IDs of mixed aetiologies (*P* < 0.001). Although verbal aggression was more prevalent in individuals with SMS relative to those with IDs of mixed aetiologies, this effect did not reach significance following a Bonferroni correction (*P* = 0.022).

Topographies of physical aggression in Smith-Magenis syndrome

The mean number of topographies of physical aggression displayed by participants, as yielded by the CCB, was seven [range 1–13, standard deviation (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 subsample of individuals with SMS over the age of 19 (*n* = 8) and these previously published rates shown by a large group of individuals with IDs 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.

Frequency and severity of aggression in Smith-Magenis syndrome

Frequency, management difficulty and severity scores for 14 topographies of physical aggression were ascertained using the CCB. The mean item frequency score for the SMS group was 3.5 (range 2.29–5.00, SD 0.69) (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 0.63) (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 0.62) (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 with those shown by individuals with ID because of 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 with 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 ID because of 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 sub-scale 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.

A series of repeated measures ANOVAs with a single within-subjects factor (function sub-scale) was conducted, to test for differences between the functions of each form of behaviour. There were significant (to the adjusted level of $P < 0.01$) main effects of function in the data for physical aggression ($F_{4,108} = 13.74$, $P < 0.01$) and verbal aggression ($F_{4,52} = 9.14$, $P < 0.01$).

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: $t(27) = 6.46$, $P < 0.001$; VA: $t(13) = 5.67$, $P < 0.001$] or being related to pain and discomfort [PA: $t(27) = 3.92$, $P = 0.001$; VA: $t(13) = 4.52$, $P = 0.001$]. There was also significantly more physical and verbal aggression associated with an escape function than either a self-stimulatory function [PA: $t(27) = 6.30$, $P < 0.001$; VA: $t(13) = 3.19$, $P = 0.007$] or pain and discomfort [PA: $t(27) = 4.40$, $P < 0.001$; VA: $t(13) = 4.81$, $P < 0.001$]. Finally, there was significantly more physical aggression associated with access to tangibles than with either self stimulation [$t(27) = 6.30$, $P < 0.001$] or pain and discomfort [$t(27) = 6.30$, $P < 0.001$]. Thus, both physical and

Table 3 Mean total function scores derived for each form of aggression evaluated

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

Data are presented as mean (standard deviation).

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, Diagnostic and Statistical Manual of Mental Disorder-IV index of hyperactive impulsive behaviour [$r(31) = 0.77$, $P < 0.001$] and the global restless impulsive index [$r(31) = 0.72$, $P < 0.001$]. 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 IDs (see *Introduction*). These variables were age, sleep distur-

bance (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 & Koch's (1977) criteria] with hyperactivity (Conner's Attention Deficit/Hyperactivity Disorder index) and autistic type behaviours (CARS total score), relationships which both attained significance, and with degree of cognitive impairment (Wechsler Intelligence Scale for Children 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 SMS. 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

Variable	Pearson's *r* and *P*-values
Age	$r = -0.01, P = 0.931$
Sleep score (total ISQ score)	$r = 0.36, P = 0.044$
Intellectual ability (WISC IQ/VABS standard composite score)	$r = -0.12, P = 0.516$
Vineland receptive communication score	$r = -0.46, P = 0.008$
Vineland expressive communication score	$r = -0.46, P = 0.009$
Hyperactivity (Conners' ADHD index)	$r = 0.56, P < 0.005*$
Impulsivity (DEX-/DEX-C total score)	$r = 0.72, P < 0.001*$
CARS total score	$r = 0.53, P < 0.005*$
RBQ total score	$r = 0.24, P = 0.202$

Table 4 Correlations between total severity of aggression scores (sum of physical aggression, destruction and self-injurious behaviour severity scores as measured with the Challenging Behaviour Interview) and potential predictor variables

* Correlation coefficients are marked with an * that are significant to a corrected level of $P < 0.005$ (following a Bonferroni correction).
ADHD, Attention Deficit/Hyperactivity Disorder; CARS, Childhood Autism Rating Scale; DEX, Dysexecutive Questionnaire; ISQ, Infant Sleep Questionnaire; RBQ, Repetitive Behaviour Questionnaire; VABS, Vineland Adaptive Behaviour Scales; WISC IQ, Wechsler Intelligence Scales Full Scale IQ.

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; Webber 1999; Arron *et al.* 2010; Finucane *et al.* 2001). 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 IDs 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 ID 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 SMS.

Frequency and severity were 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 & Bruininks (1984) and Kebbon & Windahl (1986) reported that self-injury and destructive behaviour in people with IDs 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 ID of mixed aetiology reported in the literature (Oliver *et al.* 2003) using the CBI, no differences were found. Therefore, although all aggressive behaviours are more prevalent in individuals with SMS when compared with 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 ID 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 ID 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 & 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 IDs. (Emerson & Bromley 1995; Applegate *et al.* 1999).

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 & Bromley (1995) and Applegate *et al.* (1999) found the same pattern in samples of people with mixed aetiological IDs. 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 IDs. 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 (e.g. 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 IDs of mixed aetiology and genetic syndromes (e.g. Emerson 1998; McClintock *et al.* 2003; Arron *et al.* 2010). 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; Swann & Hollander 2002; Rojahn *et al.* 2004; Petty & Oliver 2005).

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 with those with IDs because of 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 IDs because of 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 IDs because of 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 ID because of 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 and all included individuals with a range of degrees of ID. 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.

Acknowledgements

References

Allanson J. E., Greenberg F. & Smith A. C. M. (1999) The face of Smith-Magenis syndrome: a subjective and objective study. *Journal of Medical Genetics* **36,** 394–7.

Applegate H. R., Matson J. L. & Cherry K. E. (1999) An evaluation of functional variables affecting severe problem behaviours in adults with mental retardation by using the Questions About Behavioural Function Scale (QABF). *Research in Developmental Disabilities* **20,** 229–37.

Arron K., Oliver C., Berg K., Moss J. & Burbidge C. (2010) Prevalence and phenomenology of self-injurious and aggressive behaviour in genetic syndromes. *Journal of Intellectual Disability Research* DOI: 10.1111/j.1365-2788.2010.01337.x.

Borthwick-Duffy S. A. (1994) Prevalence of destructive behaviours: a study of aggression, self-injury and property destruction. In: *Destructive Behaviour in Developmental Disabilities; Diagnosis and Treatment* (eds T. Thomson & D. B. Gray), pp. 3–23. Sage Publications, London.

Clarke D. J. & Boer H. (1998) Problem behaviours associated with deletion Prader-Willi, Smith-Magenis and Cri-du-Chat syndromes. *American Journal on Mental Retardation* **103,** 246–71.

Colley A. F., Leversha M. A., Voullaire L. E. & Rogers J. G. (1990) Five cases demonstrating the distinctive behavioural features of chromosome deletion 17 (p11.2 p11.2) (Smith-Magenis syndrome). *Journal of Paediatrics and Child Health* **26,** 17–21.

Conners C. K. (1997) *Conners' Parent Rating Scale-Revised: Long Version.* Psychological Corp, New York.

Crumley F. E. (1998) Smith-Magenis syndrome. *Journal of the American Academy of Child and Adolescent Psychiatry* **37,** 1131–2.

Davidson P. W., Jacobsen J., Cain N. N., Palumbo D., Sloane-Reeves J. E., Quijano L. E. *et al.* (1996) Characteristics of children and adolescents with mental retardation and outwardly directed aggressive behaviour. *American Journal on Mental Retardation* **101,** 244–55.

Deb S., Thomas M. & Bright C. (2001) Mental Disorders in adults with intellectual disability 2; the rate of behaviour disorders among a community based population aged between 16 and 64 years. *Journal of Intellectual Disability Research* **45,** 506–14.

Dykens E. M. & Smith A. C. M. (1998) Distinctiveness and correlates of maladaptive behaviour in children and adolescents with Smith-Magenis syndrome. *Journal of Intellectual Disability Research* **42,** 481–9.

Dykens E. M., Finucane B. & Gayley C. (1993) Neuropsychological and behavioural profiles in individuals with Smith-Magenis syndrome. *American Journal of Human Genetics* **53,** 425.

Dykens E. M., Finucane B. & Gayley C. (1997) Brief report: cognitive and behavioural profiles in persons with Smith-Magenis syndrome. *Journal of Autism and Developmental Disorders* **27,** 203–11.

Dykens E. M., Hodapp R. M. & Finucane B. M. (2000) *Genetics and Mental Retardation Syndromes: A New Look at Behaviour and Interventions.* Paul H Brookes Publishing Co, Baltimore, MD.

Emerson E. (1998) Working with people with challenging behaviour. In: *Clinical Psychology and People with Intellectual Disabilities* (eds E. Emerson, C. Hatton, J. Bromley & A. Caine), pp. 127–53. John Wiley & Sons, Chichester.

Emerson E. & Bromley J. (1995) The form and function of challenging behaviours. *Journal of Intellectual Disability Research* **39,** 388–98.

Emerson E., Alborz A., Reeves D., Mason H., Swarbrick R., Kiernan C. *et al.* (1997) *The HARC Challenging Behaviour Project. Report 2: the Prevalence of Challenging Behaviour.* Hester Adrian Research Centre, University of Manchester, Manchester.

Emerson E., Kiernan C., Alborz A., Reeves D., Mason H., Swarbrick R. *et al.* (2001) The prevalence of challenging behaviours: a total population study. *Research in Developmental Disabilities* **22,** 77–93.

Emslie H., Wilson F. C., Burden V., Nimmo-Smith I. & Wilson B. A. (2003) *Behavioural Assessment of the Dysexecutive Syndrome for Children.* The Thames Valley Test Company, Bury St. Edmunds, England.

Eyman R. K. & Call T. (1977) Maladaptive behaviour and community placement of mentally retarded persons. *American Journal on Mental Deficiency* **82,** 137–44.

Finucane B. M., Jeager E. R., Kurtz M., Weinstein M. & Scott C. I. (1993) Eye abnormalities in the Smith-Magenis contiguous gene deletion syndrome. *American Journal of Medical Genetics* **45,** 443–6.

Finucane B. M., Konar D., Haas-Givler B., Kurtz M. & Scott C. I. (1994) The spasmodic upper body squeeze: a characteristic behaviour in Smith-Magenis syndrome. *Developmental Medicine and Child Neurology* **36,** 70–83.

Finucane B. M., Dirrigl K. & Simon E. W. (2001) Characterisation of self-injurious behaviours in children and adults with Smith-Magenis syndrome. *American Journal on Mental Retardation* **106,** 52–8.

Girirajan S., Vlangos C. N., Szomju B. B., Edelman E., Trevors C. D., Dupuis L. *et al.* (2006) Genotype-phenotype correlation in smith magenis syndrome; Evidence that multiple genes in 17p11.2 contribute to the clinical spectrum. *Genetics in Medicine* **8,** 417–27.

Greenberg F., Guzzetta V., de Oca-Luna R. M., Magenis R. E., Smith A. C. M., Richter S. F. *et al.* (1991) Molecular analysis of the Smith-Magenis syndrome: a possible contiguous gene syndrome associated with del (17)(p11.2). *American Journal of Human Genetics* **49,** 1207–18.

Greenberg F., Lewis R. A., Killian J., Glaze D. G., Williamson W. G., Patel Q. *et al.* (1993) Updated clinical findings in Smith-Magenis syndrome. *American Journal of Human Genetics* **53,** 110.

Haas-Givler B. & Finucane B. (2000) *Educational considerations and classroom strategies: children with SMS.* Parents and Researchers interested in Smith-Magenis syndrome, second National Conference on Smith-Magenis syndrome, September 21-24-2000, Arlington, Virginia.

Hagerman R. J. (1999) Psychopharmacological interventions in fragile X syndrome, fetal alcohol syndrome, Prader-Willi syndrome, Angelman syndrome, Smith-Magenis syndrome, and velocadiofacial syndrome. *Mental Retardation and Developmental Disabilities Research Reviews* **5,** 305–13.

Harris P. (1993) The nature and extent of aggressive behaviour amongst people with learning difficulties (mental handicap) in a single health district. *Journal of Intellectual Disability Research* **37,** 221–42.

Harris P., Humphreys J. & Thomson G. (1994) A checklist of challenging behaviour: the development of a survey instrument. *Mental Handicap Research* **7,** 118–33.

Hill B. K. & Bruininks R. H. (1984) Maladaptive behaviour of mentally retarded individuals in residential facilities. *American Journal of Mental Deficiency* **88,** 380–7.

Horn I. A. (1999) *The cognitive and behavioural phenotype of Smith Magenis syndrome.* Unpublished doctoral thesis. Institute of Psychiatry, Kings College, London.

Joyce T., Ditchfield H. & Harris P. (2001) Challenging Behaviour in Community Services. *Journal of Intellectual Disability Research* **45,** 130–8.

Kebbon L. & Windahl S. I. (1986) Self injurious behaviour results of a nationwide survey among mentally retarded persons in Sweden. In: *Science and Service in Mental Retardation* (eds J. Berg & J. M. Dejong), pp. 142–8. Methuen, London.

King B. H. (1993) Self-injury by people with mental retardation: a compulsive behaviour hypothesis. *American Journal on Mental Retardation* **98,** 93–112.

Kobe F. H., Mulick J. A., Rash T. A. & Martin J. (1994) Nonambulatory persons with profound mental retardation: physical developmental, and behavioural characteristics. *Research in Developmental Disabilities* **15,** 413–23.

Landis J. R. & Koch G. G. (1977) The measurement of observer agreement for categorical data. *Biometrics* **33,** 159–74.

McClintock K., Hall S. & Oliver C. (2003) Risk markers associated with challenging behaviours in people with intellectual disabilities: a meta-analytic study. *Journal of Intellectual Disability Research* **47,** 405–16.

Madduri N. S., Turcich M. & Lupski J. R. (2002) Low adaptive behaviour and cognitive functioning in patients with Smith-Magenis Syndrome[del(17)(p11.2p11.2)]. *American Journal of Human Genetics* **17,** 109.

Matson J. L. & Vollmer T. R. (1995) *Users Guide: Questions About Behavioural Function (QABF).* Scientific Publishers, Inc, Baton Rouge, LA.

Morrell J. M. B. (1999) The Infant Sleep Questionnaire: a new tool to assess infant sleep problems for clinical and research purposes. *Child Psychology and Psychiatry* **4,** 20–6.

Moss J., Oliver C., Arron K., Burbidge C. & Berg K. (2009) The prevalence and phenomenology of repetitive behavior in genetic syndromes. *Journal of Autism and Developmental Disorders.* **39,** 572–88.

Oliver C., McClintock K., Hall S., Smith M., Dagnan D. & Stenfert-Kroese B. (2003) Assessing the severity of challenging behaviour: psychometric properties of the Challenging Behaviour Interview (CBI). *Journal of Applied Research into Intellectual Disabilities* **16,** 53–61.

Oliver C., Berg K., Burbidge C., Arron K. & Moss J. (in press) Delineation of behavioral phenotypes in genetic syndromes. Comparison of autism spectrum disorder, affect and hyperactivity. *Journal of Autism and Developmental Disorders* DOI: 10.1007/s10803-010-1125-5.

Petty J. & Oliver C. (2005) Self-injurious behaviour in individuals with intellectual disabilities. *Current Opinion in Psychiatry* **18,** 484–9.

PRISMS (2004) *PRISMS Parents and Researchers Interested in Smith-Magenis Overview page*. Available at. http://www.smithmagenis.org/overview.htm (retrieved 24 September 2004).

Rojahn J., Matson J. L., Naglieri J. A. & Mayville E. (2004) Relationships between psychiatric conditions and behaviour problems among adults with mental retardation. *American Journal on Mental Retardation* **109,** 21–33.

Sadeh A. (2004) A brief screening questionnaire for infant sleep problems: Validation and findings for an Internet sample. *Pediatrics* **113,** E570–E577.

Schopler E., Reichler R. J., DeVellis R. F. & Daly K. (1986) Toward objective classification of childhood autism: Childhood Autism Rating Scale (CARS). *Journal of Autism and Developmental Disorders* **10,** 92–103.

Slager R. E., Lynn T., Newton T. L., Vlangos C. N., Finucane B. & Elsea S. H. (2003) Mutations in RAI1 associated with Smith-Magenis syndrome. *Nature Genetics* **33,** 466–8.

Smith A. C. M. & Gropman A. (2001) Smith-Magenis Syndrome. In: *Management of Genetic Syndromes* (eds S. B. Cassidy & J. E. Allanson), pp. 363–87. Wiley-Liss, Inc., Oxford.

Smith A. C. M., McGavran L., Robinson J., Waldstein G., Macfarlane J., Zonona J. *et al.* (1986) Interstitial deletion of (17)(p11.2p11.2) in nine participants. *American Journal of Medical Genetics* **24,** 393–414.

Smith S., Branford D., Collacott R. A., Cooper S.-A. & McGrother C. (1996) Prevalence and cluster typology of maladaptive behaviours in a geographically defined population of adults with learning disabilities. *British Journal of Psychiatry* **169,** 219–27.

Smith A. C. M., Dykens E. & Greenberg F. (1998) Behavioural Phenotype of Smith-Magenis syndrome (del 17 p11.2). *American Journal of Medical Genetics* **81,** 179–85.

Sparrow S. S., Balla D. A. & Chiccetti D. V. (1984) *Vineland Adaptive Behaviour Scales: Interview Edition. Survey Form Manual*. Circle Pines: American Guidance Survey.

Stratton R. F., Dobyns W. B., Greenberg F., DeSana J. B., Moore C., Fidone G. *et al.* (1986) Interstitial Deletion of (17) (p11.2p11.2): report of six additional patients with a new chromosome deletion syndrome. *American Journal of Medical Genetics* **24,** 421–32.

Swann A. C. & Hollander E. (2002) *Impulsivity and Aggression: Diagnostic Challenges for the Clinician: A Monograph for Continuing Education Credit*. ACCESS Medical Group, Arlington Heights, IL.

Taylor L. & Oliver C. (2008) The behavioural phenotype of Smith-Magenis syndrome: evidence for a gene-environment interaction. *Journal of Intellectual Disability Research* **52,** 830–41.

Udwin O., Webber C. & Horn I. (2001) Abilities and attainment in Smith-Magenis syndrome. *Developmental Medicine and Child Neurology* **43,** 819–22.

Webber C. (1999) *Cognitive and behavioural characteristics of children with Smith-Magenis Syndrome*. Unpublished Doctoral Thesis. University of Leicester, UK.

Wechsler D. (1991) *Wechsler Intelligence Scale for Children*, 3rd edn. The Psychological Corporation, San Antonio, TX.

Wechsler D. (1997) *Wechsler Adult Intelligence Scale*, 3rd edn. The Psychological Corporation, San Antonio, TX.

Whitman T. L. (1990) Self regulation and mental retardation. *American Journal of Mental Retardation* **94,** 347–62.

Wilson B. A., Alderman N., Burgess P. W., Emslie H. & Evans J. J. (1996) *Behavioural Assessment of the Dysexecutive Syndrome*. Thames Valley Test Company, Bury St Edmunds, England.

Appendix 1

Bihm E. M., Poindexter A. R. & Warren E. R. (1998) Aggression and psychopathology in persons with severe or profound mental retardation. *Research in Developmental Disabilities* **19,** 423–38.

Borthwick-Duffy S. A. (1994) Prevalence of destructive behaviors: a study of aggression, self-injury and property destruction. In: *Destructive Behavior in Developmental Disabilities: Diagnosis and Treatment* (eds T. Thomson & D. B. Gray), pp. 3–23. Sage Publications, London.

Bruininks R. H., Olson K. M., Larson S. A. & Lakin K. C. (1994) Challenging behaviors among persons with mental retardation in residential settings; implications for policy, research and practice. In: *Destructive Behavior in Developmental Disabilities: Diagnosis and Treatment* (eds T. Thompson & D. B. Gray), pp. 24–48. Sage Publications, Thousand Oaks, CA.

Davidson P. W., Jacobsen J., Cain N. N., Palumbo D., Sloane-Reeves J. E., Quijano L. E. *et al.* (1996) Characteristics of children and adolescents with mental retardation and outwardly directed aggressive behavior. *American Journal on Mental Retardation* **101,** 244–55.

Deb S., Thomas M. & Bright C. (2001) Mental Disorders in adults with intellectual disability 2; the rate of behaviour disorders among a community based population aged between 16 and 64 years. *Journal of Intellectual Disability Research* **45,** 506–14.

Emerson E. & Bromley J. (1995) The form and function of challenging behaviours. *Journal of Intellectual Disability Research* **39,** 388–98.

Emerson E., Alborz A., Reeves D., Mason H., Swarbrick R., Kiernan C. *et al.* (1997) *The HARC Challenging Behaviour Project. Report 2: the Prevalence of Challenging Behaviour.* Hester Adrian Research Centre, University of Manchester, Manchester.

Emerson E., Kiernan C., Alborz A., Reeves D., Mason H., Swarbrick R. *et al.* (2001) The prevalence of challenging behaviors: a total population study. *Research in Developmental Disabilities* **22,** 77–93.

Eyman R. K. & Call T. (1977) Maladaptive behaviour and community placement of retarded persons. *American Journal of Mental Deficiency* **82,** 137–44.

Fleming I. & Stenfert-Kroese B. (eds) (1993) *People with Learning Disability and Severe Challenging Behaviour; Advances in Research, Service Delivery and Interventions.* Manchester University Press, Manchester.

Fovel J. T., Lash P. S. & Barron D. A. (1989) A survey of self-restraint, self-injury, and other maladaptive behaviors in an institutionalized retarded population. *Research in Developmental Disabilities* **10,** 377–82.

Hardan A. & Sahl R. (1997) Psychopathology in children and adolescents with developmental disorders. *Research in Developmental Disabilities* **18,** 369–82.

Harris P. (1993) The nature and extent of aggressive behaviour amongst people with learning difficulties (mental handicap) in a single health district. *Journal of Intellectual Disability Research* **37,** 221–42.

Hill B. K. & Bruininks R. H. (1984) Maladaptive behaviour of mentally retarded individuals in residential facilities. *American Journal of Mental Deficiency* **88,** 380–7.

Jacobson J. W. (1982) Problem behaviour and psychiatric impairment within a developmentally disabled population 1: behaviour frequency. *Applied Research in Mental Retardation* **3,** 121–39.

Kiernan C. & Kiernan D. (1994) Challenging behaviour in schools for pupils with severe learning difficulties. *Mental Handicap Research* **7,** 177–201.

Kobe F. H., Mulick J. A., Rash T. A. & Martin J. (1994) Nonambulatory persons with profound mental retardation: physical developmental, and behavioral characteristics. *Research in Developmental Disabilities* **15,** 413–23.

Maclean W. E., Stone W. L. & Brown W. H. (1994) Developmental psychopathology of destructive behavior. In: *Destructive Behavior in Developmental Disabilities* (eds T. Thompson & D. B. Gray), pp. 68–79. Sage, Thousand Oaks, CA.

Quine L. (1986) Behaviour problems in severely mentally handicapped children. *Psychological Medicine* **16,** 895–907.

Qureshi H. (1993) Prevalence of challenging behaviour in adults. In: *People with Learning Disability and Severe Challenging Behaviour* (eds I. Fleming & B. Stenford-Kroese), pp. 11–42. Manchester University Press, Manchester.

Qureshi H. & Alborz A. (1992) Epidemiology of challenging behaviour. *Mental Handicap Research* **5,** 130–45.

Smith S., Branford D., Collacott R. A., Cooper S.-A. & McGrother C. (1996) Prevalence and cluster typology of maladaptive behaviours in a geographically defined population of adults with learning disabilities. *British Journal of Psychiatry* **169,** 219–27.