

A Behavioural Approach to the Assessment and Treatment of Severe Self-Injury in a Woman with Smith-Magenis Syndrome: A Single Case Study

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Abstract. A behavioural approach was utilized in the assessment and treatment of severe self-injurious behaviour (SIB) in a woman with Smith-Magenis Syndrome (SMS). The results showed a marked reduction in the frequency of SIB following the introduction of functional assessment driven behavioural interventions (differential reinforcement of other behaviour, self-monitoring and social reinforcement, and extinction). There were also considerable reductions in the frequencies of aggressive behaviour towards others, the use of rapid tranquilization and the use of control and restraint. The results of this study suggest environmental factors may play a significant role in maintaining some self-injurious behaviour shown by people with Smith Magenis Syndrome. The results are discussed in relation to emerging models of SIB in SMS.

Keywords: Smith-Magenis Syndrome, self-injurious behaviour, differential reinforcement of other behaviour, self-monitoring and social reinforcement, extinction, functional assessment.

Introduction

Smith-Magenis Syndrome (SMS) is a genetic disorder resulting from a chromosomal deletion of a proportion of 17p11.2., (Smith et al., 1986; Greenberg et al., 1991). The syndrome is estimated to occur in 1 in 25,000 live births (Finucane, Hains-Dirrigl and Elliot, 2001). People with SMS have a range of characteristic physical features (Smith et al., 1986; Chen, Potocki and Lupski, 1996). The majority of older children and adults with SMS function within the moderate intellectual disability range (Dykens, Finucane and Gayley, 1997). Children and adults with SMS show very high levels of behaviour problems, including self-injury, impulsivity, aggression, and stereotypies (Finucane et al., 2001; Dykens and Smith, 1998; Smith, Dykens and Greenburg, 1998a). Self-injurious behaviour (SIB) is found in over 90% of people with SMS. The most frequently occurring SIBs are self-biting, (77–93%), head banging (55%), skin picking (52%), self-hitting (62–71%), and removal of nails (29–55%) (Dykens and Smith, 1998; Finucane et al., 2001). Sleep disturbance is very common in people with SMS (Smith, Dykens and Greenburg, 1998b) and is a very strong predictor of behavioural problems (Dykens and Smith, 1998).

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Finucane et al. (2001) have begun to articulate a theoretical model that may be helpful in understanding the genesis and maintenance of SIB in people with SMS. They suggest that SIB may first occur as a result of gene expression, a possible mechanism for this being unusual sensations arising in fingers and toes due to peripheral neuropathy. This SIB may then be reinforced by social consequences.

The literature on treatment for behavioural problems in SMS is sparse. Expert opinion suggests that total communication approaches (e.g. sign language), early intervention therapies (e.g. speech, physical and occupational therapies, and sensory integration), special education and medication approaches have been found to be useful in reducing some of the negative developmental and behavioural characteristics of SMS in children (PRISMS, 2003). However, medication appears to be the dominant approach for treating self-injurious and aggressive behaviours displayed by people with SMS, despite only partially successful effects being reported (The Smith-Magenis Foundation, 1994).

The role of environmental reinforcement in the maintenance of SIB in the Finucane et al. (2001) hypothesis would suggest that a behavioural approach might be useful in assessment and treatment. This single case study therefore investigated the effectiveness of a behavioural approach to the assessment and treatment of severe SIB displayed by an adult woman diagnosed with Smith-Magenis Syndrome and associated learning disabilities.

Method

Participant and setting

Jane is a 36-year-old woman with a diagnosis of Smiths Magenis Syndrome (evidenced by genetic testing). Jane has a mild learning disability and attended a school for children with learning disabilities. She can speak and write in simple sentences and is able to read at a basic level (primary school level). Jane has a long history of challenging behaviour, including SIB, which began in her pre-school years. She has experienced major losses of caregivers due to death or placement change and is a survivor of sexual abuse.

At the start of treatment Jane was an inpatient in a specialist NHS acute assessment and treatment unit for people with learning disabilities and additional mental health/behavioural problems. She was self-injuring by inserting objects into a wound on her left forearm. The wound was the result of earlier self-injury. Jane was also pulling out her nails and assaulting others; neither of these behaviours were targeted by this intervention.

Previous treatments had consisted of pharmacotherapy and attempted individual psychotherapy; there had been three attempts during the previous 2 years to engage Jane in individual psychotherapy by three different therapists. No therapist had been able to reliably access Jane's internal cognitive or emotional states. The severity of the injury had resulted in visits to the accident and emergency department and a plaster cast being applied in an unsuccessful attempt to prevent further self-injury to the area. Intervention for the SIB to the forearm was prioritized because the wound had become infected and medical opinion indicated that there was a significant risk of gangrene.

Functional assessment of self-injurious behaviour to arm

A functional assessment of the targeted SIB was carried out by use of:

- Antecedent-behaviour-consequence (ABC) charts
- Motivational Assessment Scale (MAS) (Durand and Crimmins, 1988) completed by 20 members of staff
- Semi-structured interviews with nursing staff regarding setting events, antecedents and consequences of the targeted SIB

Setting events. The majority (65% of ABC charts) of episodes of the targeted SIB occurred during the night. A significant minority of episodes (20% of ABCs) occurred in the daytime when Jane was in a room on her own on the Unit (of her own volition). The behaviour did not occur when Jane was in the presence of staff she valued highly (those to whom Jane made repeated requests to spend time with) or when she was engaged in preferred community-based activities (activities Jane requested and engaged in when presented).

Antecedents. The behaviour occasionally occurred in response to demands, but in the majority of cases there was no recorded social antecedent (because the behaviour usually occurred when Jane was alone).

Reinforcers. Eleven of the 20 MAS scales identified positive social reinforcement and 9 of the 20 MAS identified negative social reinforcement as the primary reinforcer. On 60% of ABC charts nurses had recorded “visit to A&E department” as a consequence of the SIB. “Positive attention” (social interaction including empathy, sympathy, medical treatment) was recorded as a consequence of the SIB on 85% of ABCs. “Negative attention” (criticism, reprimand, control and restraint) was recorded as a consequence of the SIB on 15% of ABCs. “Escaping tasks or situation” was recorded as a consequence of the SIB on 15% of ABCs. Multiple consequences were recorded for some episodes of SIB, hence the percentages above add up to over 100%.

Treatment hypotheses

On the basis of the functional assessment we generated the following treatment hypotheses:

1. Jane’s SIB was reinforced by a number of internal and social consequences.
2. The reinforcer that was most active in maintaining Jane’s SIB was positive social attention received outside of the treatment unit (e.g. in accident and emergency).
3. SIB had a low response cost for Jane because, typical of many people with SMS, she had decreased sensitivity to pain (Greenburg et al., 1991, 1996).
4. The delivery of positive social attention away from the treatment unit contingent on non-occurrence of SIB would result in an increase in the rate of behaviours other than SIB.
5. Ensuring that positive social attention away from the treatment unit was not a consequence of SIB would reduce the frequency of SIB through a process of extinction.
6. The response cost of SIB would be increased if Jane engaged in written self-monitoring.

Treatment

Consent to treatment. The authors assessed Jane’s capacity to consent to behavioural treatment. Jane was able to understand and retain information regarding the costs and benefits of taking part in this specific treatment package, and what the treatment entailed. We therefore

felt she had the capacity to consent to the treatment. Jane consented to the treatment package and her consent was regularly reviewed.

Differential Reinforcement of Other Behaviour (DRO) for non-occurrence of SIB. This intervention consisted of Jane being offered the opportunity to go to town to buy a cassette with a member of staff (positive social attention away from the treatment unit) at the end of every 24-hour period during which Jane had not self-injured. This frequency of reinforcement was twice the frequency of the SIB, ensuring a higher rate of reinforcement for other behaviours than for SIB.

For each half week in which Jane did not self-injure she was offered the opportunity to go out for a meal with a chosen member of nursing staff. This provided a more powerful version of the same reinforcer (positive social attention away from the unit, but with chosen staff and for a longer time) for longer periods of displaying behaviours other than SIB.

Two weeks after the DRO was implemented there had been no occurrence of SIB. The daily outings to buy a cassette stopped after Jane had said that she did not want to go out to get any more cassettes. The twice-weekly meal outings became the sole reinforcer for non-occurrence of SIB.

Both levels of reinforcement in the DRO schedule were available in addition to Jane's regular programmed social and vocational outings into the community. In addition, Jane had access to positive social attention on the treatment unit. The fact that Jane declined the opportunity to go out and buy cassettes with staff provided some evidence that Jane was not deprived of social attention or community activities.

The literature reports a considerable amount of variability concerning the effectiveness of differential reinforcement procedures. Some studies have found only marginal reductions in CB, whereas others have achieved more complete suppression (O'Brien and Repp, 1990; Luiselli, Myles, Evans and Boyce, 1985; Russo, Cataldo, and Cushing, 1981). More specifically some studies suggest DRO schedules may be ineffective with low frequency behaviours occurring less than once or twice a day (Homer and Peterson, 1980; Whitaker, 1996). However, other studies have shown significant reductions in target behaviours with a frequency of less than two episodes per month (Whitaker, 1992; Redman, 1987).

Self-monitoring linked with social reinforcement. Twice per day Jane was prompted by staff to record on a chart whether she had self-injured or not by placing a tick or a cross in the appropriate box. Self-monitoring alone has shown considerable promise as an intervention for challenging behaviour (Reese, Sherman and Sheldon, 1984; Rudrud, Ziarnik and Coleman, 1984). Staff were also instructed to praise Jane when she marked a tick for a period of non-SIB and to remain neutral if she marked a cross for self-injury. Staff kept a separate record of SIB and the delivery of reinforcement on the DRO was not contingent on Jane completing the self-monitoring.

Extinction. Staff were instructed not to take Jane to accident and emergency for wound dressing following SIB unless absolutely necessary on medical grounds. Instead, they were to dress the wound on the treatment unit whilst maintaining a neutral rather than empathic interpersonal style. The aim was to prevent post SIB access to the reinforcer that we hypothesized was maintaining the SIB (social attention away from the treatment unit). No treatment at accident and emergency was required during the intervention period.

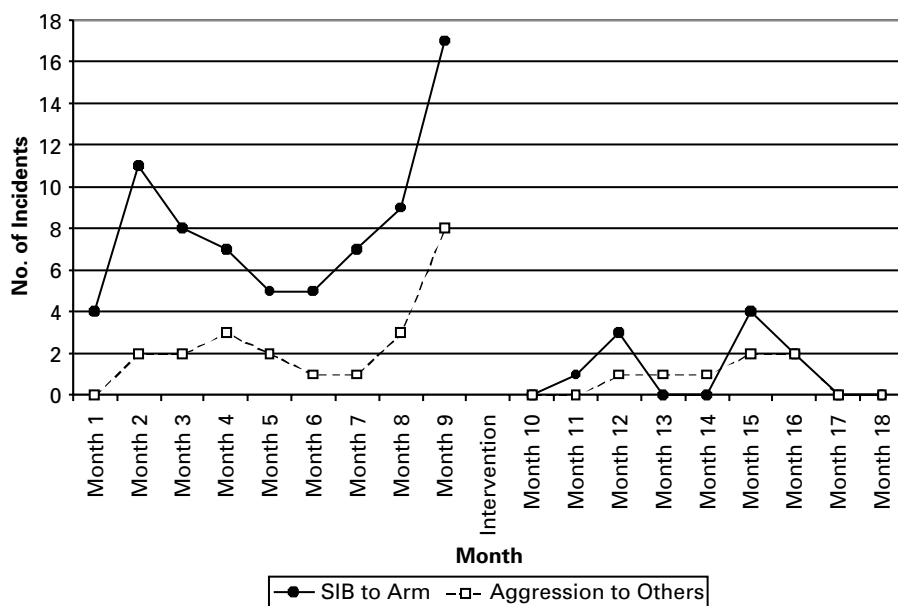


Figure 1. Frequencies per month for SIB to arm and aggression to others pre and post intervention

Pharmacotherapy. Pharmacotherapy consisted of two types of antipsychotic (depot zuclopenthixol decanoate and oral chlorpromazine) medication and an antidepressant (fluoxetine) and was kept constant throughout the baseline, treatment and follow-up periods. Rapid tranquilization medication (PRN) was a combination of Lorazepam and/or Haloperidol.

Results

Incidents of SIB and aggressive behaviours towards others were recorded by nursing staff on ABC charts. Incidents of the use of control and restraint and administration of rapid tranquilization were recorded in the nursing records. There was a marked reduction in the frequency of SIB and in aggressive behaviour towards others post intervention (Figure 1). There was also a considerable reduction in the frequency of use of control and restraint techniques and the administration of rapid tranquilization (Figure 2).

Discussion

Behavioural interventions (DRO, extinction, and self monitoring with social reinforcement) appear to have been effective in producing a clinically significant and sustained reduction in Jane's self-injury. The interventions were designed from hypotheses generated about the function of the SIB as a result of functional assessment. These hypotheses were based on the role of environmental reinforcement in maintaining the SIB. The apparent success of the interventions provides some support for these environmental reinforcement hypotheses.

This is the first report of the effectiveness of behavioural interventions for reducing SIB in a person with SMS. The finding that SIB in someone with SMS can be affected

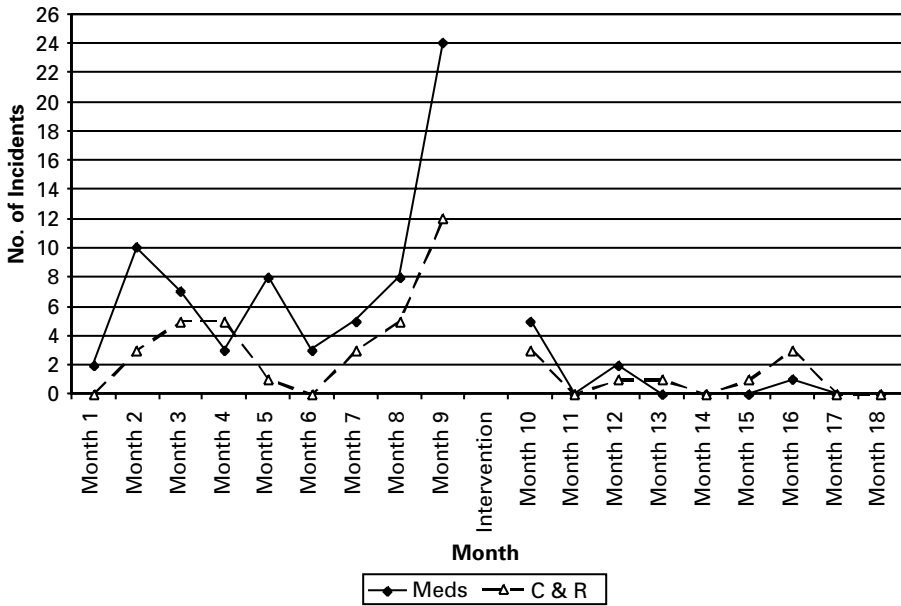


Figure 2. Frequencies per month for the use of control and restraint (C&R) and rapid tranquilization (PRN – Meds) pre and post intervention

by altering environmental reinforcement contingencies is consistent with Finucane et al.'s (2001) emerging model of the genesis (gene expression) and maintenance (environmental reinforcement) of SIB in people with SMS.

The design of the study (AB) was not truly experimental; no attempt was made to control variables systematically (with the exception of medication – the treatment unit's psychiatrist kept Jane's medication regime consistent throughout the study period at our request). As such, it is possible that it was another factor rather than the introduction of behavioural interventions that led to the reductions in SIB. However, the lengths of the pre intervention baseline and post intervention follow-up (9 months in each case) strengthen the case that the intervention package was the most salient influence on the SIB. An ABAB withdrawal design would have provided a more powerful demonstration of the influence of the behavioural treatment package but would have posed ethical problems related to the strengthening the SIB.

The results also show a reduction in aggressive behaviour towards others following introduction of the interventions. Although a functional assessment of aggressive behaviour was not carried out, the reduction suggests that the aggressive behaviour may have been maintained by the same contingencies as the SIB. Nail pulling continued at a constant frequency throughout baseline and intervention phases suggesting it was maintained by different contingencies to the targeted SIB. This highlights the need for a separate functional assessment to be carried out for each topography of SIB displayed by a person with SMS; nail pulling is a very frequent behaviour (85% of people) in older cohorts (mean age 25) with SMS (Finucane et al., 2001).

The reduction in SIB achieved by Jane led to significant improvements in her lifestyle. She was able to move out from the treatment unit where she had been for 4 years into a dispersed housing scheme run by a social care agency (where she still lives). SIB and aggression had

previously led to the break down of community placements. Post intervention Jane was exposed to much lower frequencies of control and restraint and rapid tranquilization (both of which pose significant health risks to people who receive them).

This study demonstrates that behavioural approaches show considerable promise for reducing the nearly universal SIB in people with Smith-Magenis Syndrome. We believe that the primacy of an overly deterministic genetic model have discouraged behavioural therapists and researchers from using behavioural approaches to understand and treat SIB in people with SMS. We hope that this study will encourage more behavioural therapists to become involved in this area and report their findings. Particularly useful will be reports of functional assessment and treatment of other topographies of SIB, especially nail pulling.

Studies of the relationship between age and the extent to which SIB has a social function may elucidate the developmental history of SIB in SMS. Investigations of behavioural interventions in early childhood may shed light on whether SIB emerging through gene expression can be prevented from acquiring social functions – a possible route for the prevention of SIB in people with SMS.

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