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Self-injurious behaviour in Cornelia de Lange syndrome: 2. Association with environmental events.

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Abstract

Background. Self-injurious behaviour is commonly seen in Cornelia de Lange syndrome (CdLS). However, there has been limited research into the aetiology of self-injury in CdLS and whether environmental factors influence the behaviour.

Methods. We observed the self-injury of 27 individuals with CdLS and 17 participants who did not have CdLS matched for age, gender, level of intellectual disability and mobility. Descriptive analyses were used to determine the extent to which environmental events were associated with self-injury.

Results. Lag sequential analysis of the association between self-injurious behaviour and environmental events revealed no differences between the two groups in terms of either the number or degree of environmental associations.

Conclusions. The results suggest that the associations between the environment and self-injury in CdLS do not differ from those seen in the broader population of people with intellectual disability. By implication the social reinforcement hypothesis is equally applicable to both groups.

Keywords: **Cornelia de Lange syndrome, self-injurious behaviour, descriptive analysis, social reinforcement, behavioural phenotype**

Introduction

Cornelia de Lange syndrome (CdLS) is a genetic disorder typically associated with moderate to severe intellectual disability and notable physical characteristics (Jackson, Barr, Kline and Koch, 1993; Kline *et al.*, 1993; Oliver *et al.*, 2008). In addition to the distinct physical phenotype, individuals with Cornelia de Lange syndrome are reported to show higher levels of autistic like behaviour, compulsive behaviour, expressive communication deficits (Oliver *et al.*, 2008, Moss *et al.*, 2008; Moss *et al.*, 2009) and a high prevalence of self-injurious behaviour (SIB) of between 50% and 60% (Hyman, Oliver & Hall, 2002; Oliver *et al.*, in press).

Self-injury was first reported in CdLS when Shear, Nyhan, Kirman and Stern (1971) described two children. Subsequently, Nyhan (1972) observed ‘self-mutilation’ in children with both Lesch-Nyhan and Cornelia de Lange syndromes and used the term ‘behavioural phenotype’ and suggested the self-injury in both groups was biologically determined. Since this report there has often been an implicit assumption when using the term ‘behavioural phenotype’ that an individual’s genotype is indirectly responsible for the manifestation of specific behaviours and it has also been generally accepted that self-injury is part of the behavioural phenotype of CdLS (e.g. Deb, 1997; Udwin & Dennis, 1995)

When data for individuals with intellectual disability of heterogeneous aetiology are considered, prevalence figures for self-injury in CdLS do appear to be comparatively high (Beck, 1987; Berney, Ireland & Burn, 1999; Gualtieri, 1990; Hawley, Jackson & Kurnit, 1985; Hyman, Oliver & Hall, 2002; Johnson, Ekman, Friesen, Nyhan & Shear, 1976). Thus, there is some evidence of an association between the CdLS and self-injury. However, recent work has suggested that this syndrome-behaviour relationship diminishes once key risk

markers for clinically significant self-injury are controlled (Oliver et al., in press).

There is evidence that suggests that environmental events can influence self-injurious behaviour in CdLS. For example, Berney *et al.* (1999) reported that the SIB in 76% of their sample of 49 people with CdLS, was exhibited in response to recognisable triggers including: “thwarting or frustration” (34%), “anxiety or fear” (18%), “boredom” (13%) and “demand avoidance” (5%). Similarly, Gualtieri (1990) reported that self-injury occurred when individuals were “angry”, “frustrated”, “sick”, “when unreasonable demands were made of them”, “for attention” or “in response to pain”. Dossetor, Couryer and Nicol (1991) reported that the frequency of SIB and aggression was related to the mood of the participant they observed. Finally, Johnson *et al.* (1976) found that the highest levels of self-injurious and ‘autistic acts’, in five out of seven participants with CdLS, were seen when a familiar person was talking to or touching them, perhaps indicating that this person acted as a discriminative stimulus or establishing operation for the behaviour. These reports provide preliminary evidence to suggest that SIB in CdLS can be affected by environmental factors. However, given that these findings are derived predominantly from anecdotal reports, or from the use of unevaluated, measures further investigation is required.

Studies that report behavioural interventions for self-injury provide additional support for the contention that environmental events might influence self-injurious behaviour. Singh and Pullman (1979) used punishment and differential reinforcement of other behaviour to successfully reduce self-injury in a child with CdLS. Menolascino, McGee and Swanson (1982) eliminated SIB in another child by ignoring and positively reinforcing engagement in other tasks. Dosseter *et al.* (1991) found that although a functional analysis in a young woman with CdLS was inconclusive, ignoring the self-injurious behaviour helped in its

reduction. Also, massage for 30 minutes twice a day resulted in steady improvements that were maintained 18 months later. The success of the intervention suggests that the self-injury may have served a stimulatory function or may have been reinforced by social attention, both components of massage. Bay, Mauk, Radcliffe and Kaplan (1993) devised a treatment for frequent self-biting in a child with CdLS comprising scheduled attention, extinction and reinforcement of appropriate play behaviour. Results showed a decrease in the frequency of the maladaptive behaviours in the attention-related conditions. Finally, Kern, Mauk, Marder and Mace (1995) demonstrated the efficacy of scheduled attention for self-injurious breath holding displayed by a 17-year old participant with CdLS. Although these interventions suggest that operant factors contribute to self-injury in at least some individuals with CdLS this support is limited as the descriptive nature of the reports detailing successful interventions does not provide robust evidence.

Two recent studies have advanced research in this area. Arron *et al.* (2006) used experimental functional analytic methodology to investigate self-injurious behaviours in nine children with CdLS and found that self-injurious behaviour varied with levels of social contact in three children. Moss *et al.* (2005) used descriptive analysis to examine the variability of self-injurious behaviour across environmental setting events in children with Cornelia de Lange syndrome and found that seven out of eight of the participants showed at least one form of self-injurious behaviour that was associated with a setting event. The study also demonstrated that the relationship between setting events and environmental events was extremely variable across individuals and concluded that the precise nature of the association warrants clarification.

To date, no large-scale descriptive or functional analytic studies have been conducted to

systematically assess the influence of the environment on self-injurious behaviour in people with CdLS, or other syndromes, nor have studies compared the influence of the environment on SIB to the influence seen in those without the syndrome. In this study we use systematic observational methods and analyses to identify the presence and degree of association between self-injurious behaviour and naturally occurring environmental events in people with CdLS and make comparisons with a control group.

As this is the first large-scale study to investigate the effect of the environment on self-injury in a syndrome group, it seems appropriate to determine both whether or not environmental events are associated with self-injury in individuals with CdLS of all ages and also to establish whether SIB is associated with environmental events *to a greater or lesser degree* in CdLS than a non-syndrome group by comparing the strength of the association. In this study we will compare the groups in terms of 1) the *proportion of individuals* who demonstrate an association between SIB and the environment and 2) the *degree of association* that is found in these individuals for both self-injury as a whole and for each specific form and target of self-injury.

Method

Participants

Twenty-seven individuals with CdLS and seventeen people who were comparable in terms of age, gender, mobility and level of ability took part in this study which was part of a larger project delineating the behavioural phenotype of CdLS (Oliver *et al.*, 2008; Oliver *et al.*, In press). The full recruitment procedure is reported in Oliver *et al.* (2008). The recruitment procedure yielded a group of 54 people with CdLS. Of the 54 participants, 27 displayed self-injurious behaviour, as defined in part one of the Challenging Behaviour Interview (Oliver, McClintock, Hall, Smith, Dagnan, *et al.*, 2003) and displayed sufficient instances of self-

injury to analyse the environmental associations with the behaviour. These 27 participants were included in this study. Of the 46 comparison group participants seventeen engaged in self-injurious behaviour as defined by the part one of Challenging Behaviour Interview (Oliver *et al.*, 2003) and displayed sufficient instances of SIB for analysis. The characteristics of the CdLS and comparison participants are summarised in table 1.

+++++++ Insert table 1 here ++++++

Instruments

Sony TRV-48E video camera recorders were used to record observational data and LCD fold out screens used to minimise observer reactivity. Two observers coded observational data using Obswin 32 software (version 3.0) (Martin, Oliver & Hall, 2000). Obswin 32, uses real time data collection and enables behaviours to be recorded both as ‘discrete events’ (with nominated 1 second duration) and ‘durations’ in which onset and offset times are recorded.

Procedure

Observers visited each participant in their usual day-care environment (e.g. school, day centre or home) and collected observational data of participants over the course of a typical day. Activities observed were characteristic of such settings (e.g. meal times, group activities, individual activities and leisure time). In order to observe a naturalistic environment, observers endeavoured to remain in the background of the day-care setting. Following data collection, one observer coded participant self-injurious behaviours whilst the other coded social interactions using Obswin 32 software (version 3.0) (Martin, Oliver & Hall, 2000) After each coding session, the data files from the two observers were merged to form a single data file, containing both self-injury and social interaction codes.

Observational Behavioural Definitions and Coding

Operational definitions of self-injurious behaviours were developed from informal observations and preliminary analysis of videotapes. Each individual topography of SIB was operationally defined yielding twenty types of self-injury that were all coded separately (see Oliver *et al.*, in review). In order to reduce data, the topographies of self-injury were collapsed into twelve categories: six forms (e.g. picking, poking, biting etc) and six locations (e.g. hand, face, head directed) as shown in table 2.

+++++++ Insert table 2 here ++++++++

These categories were selected in order to reduce data, whilst still allowing topographically related comparisons to be made. These categories were used because the behavioural phenotype literature suggests that syndrome groups may be more likely to exhibit particular self-injurious forms (e.g. picking in Prader-Willi syndrome (Greenswag, 1987; Whitman & Accardo, 1987)), or may target particular locations ((e. g. lips and fingers in Lesch Nyhan syndrome (Anderson & Ernst, 1994)) and the applied behaviour analytic literature indicate the possibility of different forms of behaviour being differentially effected by social environmental events (Oliver, Murphy and Corbett, 1993).

In addition to self-injurious behaviours, Social Environmental States were also coded. The applied behaviour analytic literature has identified social events that act as establishing operations that are commonly related to self-injurious behaviour in individuals with intellectual disabilities (Iwata *et al.*, 1982; Oliver, 1995). Positive reinforcement has most often been discussed in terms of the presentation of attention or tangibles contingent on the occurrence of behaviour; negative reinforcement, by the contingent removal of instructional demands or removal of social attention; and finally automatic reinforcement by sensory or

perceptual consequences. These events were used as the basis of environmental codes for this study. *Attention* and *demands* were coded as Social Environmental States and *denials* as a third Social Environmental State (as denials may be interpreted as the removal or denial of tangibles, which potentially evoke SIB). Finally, *no social contact* was coded (when the individual was not subject to any verbal or physical interaction, demands, denials or reprimands from others). It was hypothesised that if an individual's behaviour was maintained by sensory consequences, and s/he received no form of social stimulation their self-injury may be associated to this condition. Thus, in order to ascertain the extent to which self-injurious behaviours were associated with social events, social interactions were coded under the four exhaustive Social Environmental State categories shown in table 3.

+++++++ Insert table 3 here ++++++

Inter-observer reliability

Inter-observer reliability was calculated for 26.33% of observations. Two raters exchanged target codes and each independently coded a minimum of 15% of the observations for each participant. Kappa values were calculated for each behavioural code on a 10s interval-by-interval basis. Kappa values derived for both social interaction codes and single self-injurious behaviour codes were all 'good' (.60 to .75) or 'excellent' (above .75) (Fleiss, 1981 in Bakeman & Gottman, 1997) with a range of .67 to 1.00.

Data Analysis

Data files for each individual were appended, and converted into 10s intervals using Obswin 32 software (version 3.0) (Martin *et al.*, 2000). The GSEQ statistical analysis package (Bakeman & Quera, 1995) was employed for analysis of observational data in order to

determine the presence of co-occurring of self-injurious behaviours and Social Environmental States. Such associations may be defined as the temporal relation of two types of behaviours or events within an observation session (Yoder & Feurer, 2000).

Yule's Q is an index of association that can be used to assess the association between two events. Scores range from negative to positive one, and a score of zero indicates 'no association'. Yule's Q scores are unaffected by the number of tallies in the data and consequently allow resultant scores to be compared across participants and groups regardless of whether observation files are of equal length. Additionally, the calculation of Yule's Q is advantageous because it is based on conditional probabilities. Yule's Q scores were derived for each behaviour category displayed by each participant in each of the four Social Environmental States. A sufficiently large behaviour sample is required for Yule's Q to be interpretable. Values were calculated if the behaviour and Social Environmental State each occurred in at least ten, ten second-intervals. Positive rather than negative associations were used to compare the co-occurrence of SIB and environment across CdLS and comparison groups. Positive associations reflect 'real associations' i.e. only when the two co-occur is it likely that either positive or negative reinforcement processes are contributing to the exhibition of the behaviour.

Higher Yule's Q scores (either positive or negative) reflect stronger associations and the degree to which self-injurious behaviours are associated with Social Environmental States which may be variable. As such, comparisons between the CdLS and Comparison groups can be made at this level (using independent samples t-tests and Mann-Whitney U tests) to determine whether specific forms of behaviour had stronger associations to Social Environmental States in one group than the other. A second approach is to determine a cut-

off whereby associations are deemed ‘significant’ in order to make comparisons between the groups at a categorical level. A cut-off score of .6 or above was employed. At .6 the behaviour and environment are five times more likely to occur in the presence of each other than by chance. For all comparisons Mann-Whitney U and Fishers Exact tests were employed only when data were drawn from five or more individuals in each group.

Results

CdLS and Comparison Group Demographic Information

Independent samples t-tests and χ^2 tests revealed no significant differences between the comparison group and CdLS group in terms of gender, wheelchair use, place of residence, and Vineland Daily Living Skills domain standard score (see table 1 for descriptive data).

Thus, the groups are comparable on these variables.

Time Spent in Social Environment States

There were no differences between the two groups in either the percentage of time that demands (CdLS - 4.06%, comparison - 4.13%, $t(42) = .06$, $p = .95$) or denials (CdLS - .41%, comparison - .42%, $t(42) = .07$, $p = .95$) were observed. However, there was a difference in the percentage time in which attention (which excluded demands and denials) occurred (CdLS - 21.72%, comparison - 11.47%, $t(\text{unequal variances } (40)) = 4.27$, $p < .01$), and thus also a difference in the time in which no social interaction occurred (CdLS - 73.84%, comparison - 83.94%, $t(42) = 2.89$, $p < .01$). Individuals diagnosed with CdLS spent more time receiving attention than the comparison group and thus less in the no social contact state. Although this means that the observational periods were different between groups, the measurement of environmental association remains valid because Yule’s Q is unaffected by the base rate of each variable.

Co-occurrence of Social Environmental States and Self-Injurious Behaviour

A key objective was to investigate the associations between self-injurious behaviour and Social Environmental States. For this analysis, self-injury was investigated at a global level as well as at the level of injurious ‘forms’ and ‘locations targeted’. Yule’s Q scores were calculated for each type of self-injury displayed in each of four Social Environmental States (demand, denial, attention and no social contact). Table 4 shows the number of self-injurious forms and locations that were displayed at a sufficient level for analysis in each group (i.e. in at least ten, ten second intervals).

+++++++ Insert table 4 here ++++++

Figures 1 and 2 present *all* Yule’s Q scores yielded for participants in both CdLS and comparison groups for each form and location of self-injury. It should be noted that for each behaviour displayed by an individual, a Yule’s Q score is presented for up to four Social Environmental States. Consequently, individuals may have a single behaviour significantly associated with more than one Social Environmental State i.e. where a number of states were associated with a behaviour.

+++++++ Insert figures 1 & 2 here ++++++

The data in figures 1 and 2 show that the degree of association between a Social Environmental State and self-injury is variable in both the CdLS and Comparison groups. Figures 1 and 2 demonstrate that there is a spread in the Yule’s Q scores for all forms and locations of self-injury relating to all Social Environmental State for both groups. Additionally, relatively few Yule’s Q scores fall above the arbitrary cut-off (.6) that has been

used to denote a significant relationship between the behaviours and specific Social Environmental States.

The number of significant associations (i.e. Yule's Q scores above .6) observed in both the CdLS and Comparison groups are summarised in Table 5, which also shows the Social Environmental States for which associations were found. In summary, the data in table 5 show that a minority of self-injurious behaviours were found to be associated with a Social Environmental State in both the CdLS and comparison groups. In those with CdLS, the majority of behaviours that were significantly associated were related to the demand and attention conditions. The behaviours significantly associated in the comparison group were related to all environmental conditions evenly. In order to reduce these data for further analysis, each behaviour was associated with a maximum of one Social Environmental State environment. Thus, when a single behaviour had a Yule's Q score above .6 for more than one Social Environmental State, the environment in which the highest value was found is reported.

+++++++ Insert table 5 here ++++++

Comparing co-occurrence of self-injury and Social Environmental States across groups

Comparison at the participant level

The research aimed to compare the CdLS and Comparison groups in terms of proportion of individuals who demonstrate an association between any form of self-injurious behaviour and environmental event. Each individual was classified as either demonstrating an association between SIB and a Social Environmental State or not. An association was deemed present if *any* Yule's Q score calculated (i.e. any form, any location or for global self-injurious behaviour) was above .6 for that individual. In the CdLS group, 29.6% yielded an

association between self injury and a Social Environmental State, whilst in the comparison group 52.9% of participants showed an association but this difference was not statistically significant ($\chi^2(1) = 2.39, p = .12$; odds ratio of 2.67, 95% confidence interval .76 to 9.43)

In order to determine whether Social Environmental States would be associated with self-injurious behaviour to a *lesser degree* in the CdLS group than the comparison group, an independent samples t-test was conducted. The t-test compared the single highest Yule's Q score calculated for each participant within the CdLS (mean .51, SD .26) and Comparison groups (mean .60, SD .21) for any given form, location or for global self-injury across all four Social Environmental States. No significant difference was found for the mean Yule's Q values ($t(42) = 1.16, p = .25$).

Comparison at a behavioural level

In order to determine the association between specific forms or locations of self-injury and Social Environmental States the proportion of participants engaging in each form/location/global self-injury for whom the behaviour co-occurred with any Social Environmental State (i.e. with a Yule's Q scores of 0.6 or above) was calculated. When sufficient numbers of people in each group displayed the behaviour (i.e. when data were drawn from five or more individuals in each group), these proportions were compared across the two groups using Fishers exact test. Twenty seven people with CdLS and seventeen people in the comparison group showed global self injury. Of these, an association between the behaviour and a Social Environmental State was found in 7.4% of people with CdLS and 11.8% of participants in the comparison group. For individual forms of injury, fourteen people with CdLS and six people in the Comparison group displayed biting (of whom, 21.4% and 0% showed associations respectively) nineteen with CdLS and eleven

individuals in the comparison group exhibited picking (10.5% and 18.2% showed associations) whilst five people with CdLS and seven people in the comparison group showed striking (40% and 28.6% yielded associations). For locations of injury, nine people with CdLS and five people in the comparison group directed injury towards their body (of whom, 33.3% and 80% showed associations respectively) ten with CdLS and seven individuals in the comparison group exhibited injury towards their face (10% and 14.3% showed associations) whilst nineteen people with CdLS and nine people in the comparison group showed striking (15.3% and 0% yielded associations). Fishers exact tests found no significant differences in the proportion of people demonstrating an association between a Social Environmental State and either global self-injury or for any form or location of injury between the two groups.

Finally, an analysis was undertaken to determine whether individual forms or locations of self-injury were associated with a Social Environmental State to a lesser degree in the CdLS than the Comparison group. For every participant, the *highest* of the four Yule's Q scores was ascertained for each behaviour. When sufficient numbers of people in each group displayed the behaviour, Mann-Whitney-U tests were employed to compare the highest Yule's Q scores between the CdLS and comparison groups. The median Yule's Q scores for global self-injury in the CdLS group and comparison group was .26, and .24 respectively (Mann-Whitney U = 203.5, p = .53). In terms of biting the median scores were .36, and .25 (Mann-Whitney U = 19, p = .06), for picking .25, and .33 respectively (Mann-Whitney U = 72, p = .16) and for striking .33, and .45 respectively (Mann-Whitney U = 17, p = .94). For locations of self injury, median Yule's Q scores for body directed injury were .51 and .76 for CdLS and comparison groups respectively (Mann-Whitney U = 15, p = .32), for face directed injury .28 and .33 respectively (Mann-Whitney U = 29, p = .56) and finally for

hand directed injury were .28 and .27 respectively (Mann-Whitney $U = 82$, $p = .86$). Again no differences were found between groups in terms of median Yule's Q scores.

In summary, no differences were found in terms of the proportion of individuals in each group who demonstrate an association between SIB and Social Environmental States.

Additionally, the degree of association that is found for either self-injury as a whole or for each specific form of self-injury did not differ between the two groups.

Discussion

In this study we employed naturalistic observation and descriptive analyses to investigate the presence and degree of association between SIB and environmental events in a cohort of individuals with Cornelia de Lange syndrome and a comparison group selected for their similarities for self-injury risk markers. The aim was to compare the presence and degree of association between self-injury and high risk environmental events in individuals with and without the syndrome. We also investigated whether the presence and degree of association between environmental events and self injury differed between the CdLS and comparison group for specific forms of self-injury. Comparison of the groups demonstrated no differences in risk markers for self-injury, thus suggesting the groups were comparable on key variables, and assessment of inter-observer reliability showed good agreement between observers.

Four analyses revealed no significant differences between individuals with CdLS and the comparison group in terms of associations between SIB and environmental events. The two groups were compared in terms of the number of individuals demonstrating an association

between high risk environmental events and *any* form of self-injurious behaviour. 29.6% of those with CdLS and 52.9% of those in the comparison group demonstrated at least one significant association. The odds ratio showed that the comparison group were 2.7 times more likely than those with CdLS to exhibit a behaviour that was associated to one of the four environmental states. However, the difference between the two groups was not significant suggesting that individuals with CdLS are neither more nor less likely to have behaviours associated with the environment than those without the syndrome. The proportion of individuals with CdLS, just under a third, who show SIB that is related to an environmental event is similar to that reported by Arron et al. (2006) for their experimental functional analytic study, although the methods used in the present study are more likely to identify an association.

Further analyses compared the proportion of individuals manifesting significant environmental associations at a finer grained level i.e. at the level of forms and locations as well as for global self-injury. Analyses were carried out at this level in order to determine whether or not there were particular forms of self-injury that were more likely not to be associated with environmental events in Cornelia de Lange syndrome i.e. due to gastrointestinal pain (Jackson, Kline, Barr & Koch 1993, Luzzani *et al.*, 2003; Berg et al., 2007; Collis et al., 2008), or directed towards the hands (Kline, Krantz, Goldstein, Koo & Jackson, 2001; Oliver et al., in press). If this were the case, it would be expected that the proportion of significant environmental associations in the CdLS group for specific locations or forms, would be lower than for the comparison group. Again no differences were apparent. However, because analyses were only performed if at least five participants formed the derived proportion, only six of the behavioural forms and locations were compared.

Two further analyses were conducted which, rather than comparing the proportion of significant associations across the two groups, examined Yule's Q scores and thus the degree of association. There were no differences between the two groups when comparing the single highest Yule's Q score for each participant, nor were any differences found when comparing the highest Yule's Q scores for each behavioural form and location across the two groups. Therefore, it seems that although a trend was apparent in the first analysis, together, the four comparisons reveal no significant differences across the groups. Thus, for the self-injurious behaviours there is no difference between the groups in the degree to which they are associated with the environmental states used in this study.

These results suggest that the self-injury exhibited by those with CdLS is neither less nor more likely to be governed by social environmental factors than those without the syndrome. It may therefore be suggested that self-injury is not related more specifically to a nonenvironmental aetiology than those without the syndrome and this weakens the argument that this behaviour at this level forms part of the behavioural phenotype of the syndrome in the conventional sense of this term. This finding has implications for the interventions used for those displaying the behaviour. The present study suggests that because SIB in some individuals with CdLS can be associated with social environmental states, thus assessment strategies should include those currently employed by applied behaviour analysts (see Iwata et al., 1982; Hall and Oliver, 1992). The findings also extend the list of syndromes in which operant processes can be shown to contribute to behaviour within a syndrome (e.g. Oliver, Murphy and Corbett, 1993; Taylor and Oliver, 2008, Strachan et al., 2009; Oliver et al., 2007; Woodcock, Oliver, & Humphreys, 2009; Hall, Oliver, & Murphy, 2001; Millichap et al., 2003).

These findings support and extend previous research into SIB in CdLS. Oliver *et al.* (in press) found that once risk markers for clinically significant self-injury had been controlled, self-injury was no more prevalent in people with CdLS, nor was it different in its presentation from the self-injury displayed by individuals without the syndrome. The present research adds to this finding by suggesting that the extent to which environmental factors affect people with and without the syndrome is not different.

There are several points arising from the present study that are worth noting. Firstly, the level of environmental associations in *both* groups appears to be relatively low in comparison with the levels seen in applied behaviour analytic literature. For example, Iwata *et al.* (1994) found differential or uniformly high responding was observed in 95.4% of 154 participants. One explanation for the discrepancy between the results of this study and others in the applied behaviour analytic field is that although such analyses informed the present study of the environments to be identified as observational variables, the intermittent schedules of reinforcement and uncontrolled environmental variables (particularly the salience of both establishing operations and discriminative stimuli) seen in natural observations allows a number of factors to influence behaviour simultaneously. However, the controlled nature of experimental analysis eliminates such confounds and thus amplifies the effects of environmental events, thereby strengthening the associations found. Additionally, for experimental analyses, higher numbers of associations may be reported because the literature is prone to increased reporting of positive results with, for example, the contemporary Applied Behaviour Analytic literature focusing on demonstrating interventions when behaviour has an identified function. However, the two groups in the present study are recruited from comparable sources and have undergone the same analyses thus ensuring that comparisons drawn between the two are valid.

The relatively low numbers of associations between SIB and the environment also suggest that caution needs to be exercised when making inferences about the environmental conditions that are associated with SIB across groups. Although associations are indicative of functionally maintained behaviour, given the number of behaviours analysed, a proportion may be expected to be found at above .6, i.e. the dispersion of Yule's Q scores in figures 1

and 2 may be seen to resemble those that would be predicted by chance (analogous of normal distribution curves).

The present analyses found no differences between participants in the CdLS and comparison groups in terms of environmental factors that associate with the behaviour. However, the environments analysed in this study were limited to those that are believed to be influential in individuals with intellectual disability of mixed aetiology, i.e. without syndromes. It is therefore possible that differences *do* exist between individuals with and without CdLS, but that they were not revealed in the present study because the analysis only focused on four specific types of environmental conditions. Finally, for those with CdLS who do not evidence an association between environmental events there are a number of physical characteristics specifically related to CdLS that may impact on self-injurious behaviour and warrant further examination. Conditions that should be considered for further investigation in relation to self-injury in people with CdLS include, pain associated with gastro-intestinal disorder (Luzanni *et al.* 2003; Hall *et al.*, 2008; Collis *et al.*, 2008), peripheral sensory neuropathy (Kline, 2001) and behavioural dysregulation i.e. hyperactivity, stereotypy, compulsions (Oliver *et al.*, in press).

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	CdLS group (N = 27)	Comparison group (N = 17)
Mean age	14.44 years (SD 7.31)	15.38 years (SD 9.55)
Male	14 (52%)	10 (59%)
Place of residence (Living in parental home)	26 (96%)	14 82%
<u>Wheelchair use</u>		
Never	14 (52%)	11 (65%)
Occasionally	11 (41%)	5 (29%)
Always	2 (7%)	1 (6%)
<u>Vineland Adaptive Behavior Scale: Daily Living Skills domain</u>	27.48	24.24
Mean standard score	(14.74SD)	(SD 8.84)

Table 1. Demographics of Cornelia de Lange syndrome and comparison participants

VARIABLE	TOPOGRAPHIES INCLUDED
Global self-injurious behavior	Body pick, face pick, hand pick, head pick, neck pick, body poke, ear poke, eye poke, face poke, finger insertion, body hit, face hit, head hit, hand bite, lip bite, body bang, body throw, elbow flick, head bang, mouth flick
BEHAVIOR BY FORM	
Picking self	Body pick, face pick, hand pick, head pick, neck pick
Poking self	Body poke, ear poke, eye poke, face poke
Insertion	Finger insertion
Striking self	Body hit, face hit, head hit
Biting self	Hand bite, lip bite
Body to object	Body bang, body throw, elbow flick, head bang
BEHAVIOR BY LOCATION	
Body SIB	Body bang, body hit, body pick, body poke, body throw
Face SIB	Face hit, face pick, face poke
Sensory SIB	Ear poke, eye poke
Head SIB	Head bang, head hit, head pick, neck pick
Hand SIB	Hand bite, hand pick, finger insertion
Mouth SIB	Lip bite, mouth flick

Table 2. Self-injurious variables created by combining individual topographies of behavior.

ENVIRONMENTAL STATE	TOPOGRAPHIES INCLUDED
Demand	Any verbal or physical request from the caregiver to do something, requiring a response from the child.
Verbal / Physical Denial	Any verbal or physical refusal by the caregiver / other adult of something that the child wants or is attempting to do.
Attention (excluding demands and denials)	All social contact ('verbal interaction', 'verbal reprimand', 'physical interaction' and 'physical reprimand') excluding demands and denials
No social contact	The absence of 'demand', 'denial' and 'attention excluding demands and denials'

Table 3. Social Environmental States derived by combining social interaction codes.

	CdLS group (N = 27)	Comparison group (N = 17)
	No. analysed	No. analysed
<u>SIB forms</u>	19	11
Picking	3	3
Poking	14	6
Biting	5	7
Striking	4	1
Body to object	45	28
<u>Total</u>		
<u>SIB locations</u>		
Body	9	5
Face	10	7
Head	6	9
Hand	19	2
Mouth	1	0
Sense organ	4	3
<u>Total</u>	49	26

Table 4. Number of self-injurious forms and locations analysed for Cornelia de Lange syndrome and comparison groups.

	CdLS (N = 27)			Comparison (N = 17)		
	No. analysed	Presence of environmental association	Environment	No. analysed	Presence of environmental association	Environment
SIB forms	45	8 (17.78%) (n = 7)	Demand - 2 Denial - 1 Attention - 4 No contact - 1	28	5 (17.86%) (n = 5)	Demand - 1 Denial - 1 Attention - 1 No contact - 2
SIB locations	49	8 (16.33%) (n = 6)	Demand - 5 Denial - 0 Attention - 2 No contact - 1	26	7 (26.92%) (n = 6)	Demand - 0 Denial - 2 Attention - 3 No contact - 2
Global self-injury	27	2 (7.4%) (n = 2)	Demand - 2 Denial - 0 Attention - 0 No contact - 0	17	2 (11.76%) (n = 2)	Demand - 0 Denial - 0 Attention - 0 No contact - 2

Table 5. Summary of environmental associations found in Cornelia de Lange syndrome and comparison groups for self-injurious forms, locations and global self-injury. (“n =” in cells refers to total number of people for whom an association was identified at that level of analysis).

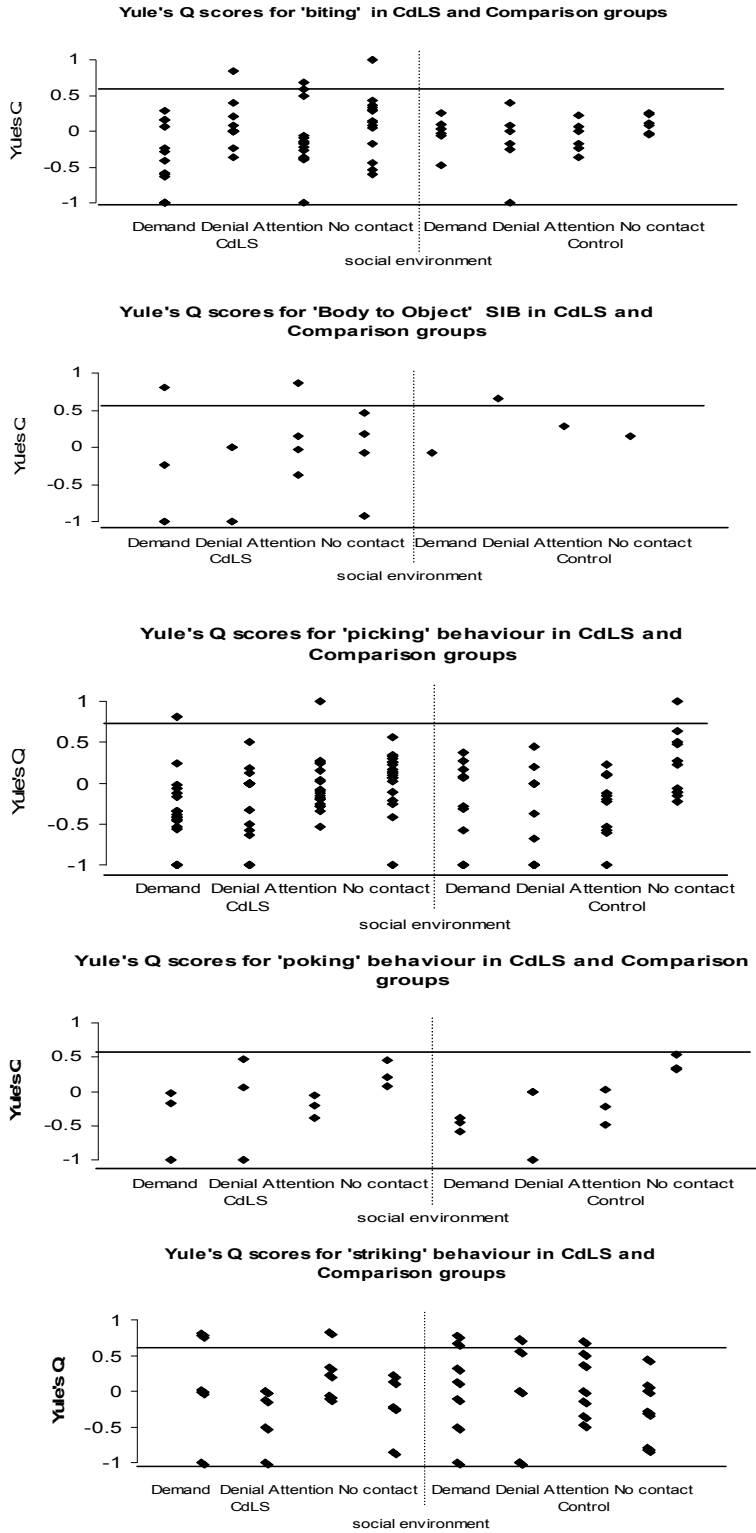


Figure 1. Yule's Q scores for each form of self-injury and each Social Environmental State for the Cornelia de Lange syndrome and comparison groups (black horizontal line denotes level of significant associations)

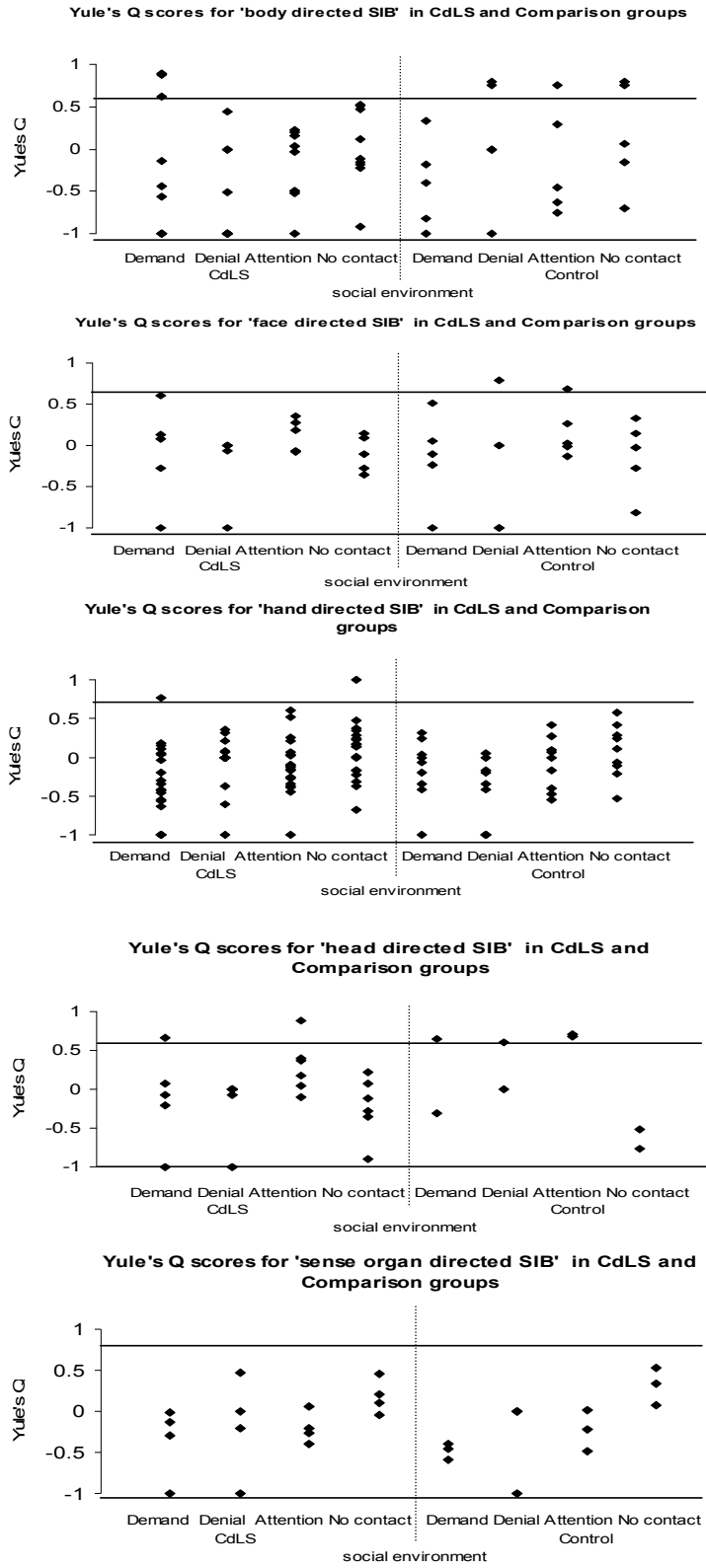


Figure 2. Yule's Q scores of each location of self-injury for each Social Environmental State for the Cornelia de Lange syndrome and comparison groups (black horizontal line denotes level of significant association)