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Health and sleep problems in Cornelia de Lange Syndrome: A case control study

Running Head: Health and sleep in Cornelia de Lange Syndrome

Abstract

Background. Self-injury, sleep problems and health problems are commonly reported in Cornelia de Lange syndrome but there are no comparisons with appropriately matched participants. The relationship between these areas and comparison to a control group is warranted.

Method. 54 individuals with CdLS were compared to 46 participants with intellectual disability of mixed aetiology who were similar in terms of degree of intellectual disability, mobility, age and gender using informant based measures of health problems, sleep and self-injury.

Results. Participants with CdLS experienced significantly more current and lifetime health problems with eye problems and gastrointestinal disorders prominent. Although 55% of those with CdLS experienced sleep problems this prevalence was not different from the comparison group. Sleep disorder was not associated with self-injury in the CdLS group, skin problems were associated with severity of self-injury but not presence.

Discussion. People with CdLS experience a range of significant health problems and have more health concerns than others with the same degree of intellectual disability. Although this has been well documented in the past, the current high prevalence of health concerns indicates the need for regular health assessment and appropriate intervention.

Introduction

Cornelia de Lange Syndrome (CdLS) is a rare genetic syndrome characterised by facial anomalies, small stature, limb abnormalities, low birth weight and excessive hair growth (Ireland, Donnai & Burn, 1993; Jackson, Kline, Barr & Koch, 1993; Kline, Stanley, Belevich, Bidsky, Barr & Jackson, 1993). Individuals with CdLS are commonly reported to have a severe to moderate degree of intellectual disability, sensory abnormalities, poor communication, autism spectrum features and compromised mobility (Berney, Ireland & Burn, 1999; Kline *et al.*, 1993, Oliver *et al.*, In review, Moss *et al.*, In press). The genetic abnormality causing CdLS has been mapped for 20 to 50% of affected individuals to the NIPBL gene, located at chromosome 5p13.1 (Krantz *et al.*, 2004; Tonkin, Wang, Lisgo, Bamshad & Strachan, 2004) with two further gene loci on chromosome 10 and X being described recently (Musio *et al.*, 2006; Deardorff *et al.*, 2007). Prevalence is estimated at 1 in 50,000 live births (Beck, 1976; Beck & Fenger, 1985) although this is thought to be an underestimate with more mildly affected individuals increasingly being identified.

The NIPBL gene is expressed in the developing skeleton and soft tissue of the limbs, hands, spinal column, face and head including the ear canal, the atrial and ventricular areas of the heart, oesophagus, trachea and lungs (Tonkin *et al.*, 2004). The compromised development in these areas may lead to the specific physical and sensory abnormalities that are often reported in CdLS involving the limbs and other bone structures, central nervous system, gastrointestinal system, peripheral senses, ears and organs such as the heart (Kline, Krantz, Goldstein, Koo & Jackson, 2001; Luzzani, Macchini, Valade, Milani & Selicorni, 2003). Health problems may develop from these abnormalities and are commonly described in individuals with CdLS (Jackson *et al.*, 1993; Luzzani *et al.*, 2003, Berg *et al.*, 2007).

Results from published research examining health problems in CdLS are presented in table 1 and indicate that individuals with CdLS experience a number of diverse health problems with gastro-intestinal problems prominent. However, many of the studies are based on clinical examination or a review of case notes of referred samples and have not always employed reliable and valid measures and comparison groups. Communication difficulties often experienced by individuals with severe intellectual disability, mean that reporting the presence and nature of health difficulties is

problematic (Beange, McElduff & Baker, 1995; Lennox & Kerr, 1997). Studies utilising carer reports to measure health problems, such as Berney *et al.* (1999), Gualtieri, (1990) and Hawley *et al.* (1985), may facilitate identification of problems and provide insight into health issues that are of immediate concern and likely to be having impact on the individual's quality of life.

+++++ Insert Table 1 here ++++++

Previous research has indicated that individuals with intellectual disabilities are more likely to experience health problems than the general adult population (Jansen *et al.*, 2004; Sutherland, Couch, Iacono, 2002) and the prevalence of specific health problems in people with CdLS appears to be further elevated. However, the prevalence of problems in CdLS may be comparable to those in intellectual disabilities populations e.g. the prevalence of skin conditions is 6 to 17% and epilepsy is 5 to 24% in people with intellectual disabilities (Jansen *et al.*, 2004). As studies of the health status in CdLS have not included comparison groups, further investigation is needed in order to identify the specific health concerns associated with CdLS.

In addition to the effects of health problems on quality of life, illness may influence sleep which in turn may impact on well-being (Brylewski & Wiggs, 1998). Empirical and anecdotal evidence suggests that sleep problems are more common in people with intellectual disabilities with 32 to 39% of people experience sleeping problems, 27 to 29% reporting settling problems and 44 to 56% waking at night (Brylewski & Wiggs, 1998, 1999; Stores, Stores & Buckley, 1996). Studies of sleep in CdLS have indicated that 55 to 72% of individuals experience problems, with settling difficulties described in 26% and waking problems in 31% (Berney *et al.*, 1999; Gualtieri, 1990; Sarimski, 1997). However, as studies have not incorporated comparison groups it is unclear whether the prevalence of sleep problems in CdLS is higher than would be expected when compared to people with a similar degree of intellectual disability.

Individuals with CdLS are also thought to be at risk for the development of self-injurious behaviour, with prevalence rates for self-injury being approximately 55 to 64% in people with CdLS compared to 3 to 17% in community intellectual disability populations (Berney *et al.*, 1999; Collacott, Cooper, Branford & McGrother, 1998;

Hyman, Oliver & Hall, 2002; Oliver, Murphy & Corbett, 1987; Sloneem et al., In review). When shown, self-injury is often reported to be severe in CdLS, causing significant damage and harm (Bryson, Sakati, Nyhan & Fish, 1971; Dossetor, Couryer & Nicol, 1991). Environmental factors can be related to self-injury in CdLS as is the case for others with an intellectual disability (Moss, et al., 2005; Arron et al., 2006). Recent research has shown that clinically significant self-injury is not more prevalent in CdLS than a matched comparison group. However, the high prevalence is cause for concern (Sloneem et al., In review).

It has been hypothesised that self-injury may develop as a response to pain from illness or trauma as a way of blocking pain sensation (Oliver, 1993) and that experiencing health problems and sleep deprivation may influence operant processes and increase self-injury (Carr et al., 1995). A number of studies have indicated that children and adults showing self-injury are significantly more likely to have difficulties with sleeping than individuals without self-injury (Brylewski & Wiggs, 1999; Piazza, Fisher, & Shorer, 1997; Symons, Davis, & Thompson, 2000). Given the reported association of CdLS with health problems, sleep difficulties and self-injurious behaviour, the relationship between these factors in people with CdLS warrants further investigation.

To identify the health needs and sleep problems of people with CdLS, we conducted a survey to examine the presence and severity of health and sleep problems. A group of individuals with intellectual disability of mixed aetiology matched for age, gender, ability and mobility was recruited for comparison. A measure of the presence and severity of self-injurious behaviour was incorporated to examine the relationship between self-injury, health and sleep problems in both groups.

Method

Participants

75 families, who had been recruited via the CdLS Foundation parent group were contacted directly. Another 112 families were contacted via the CdLS Foundation parent group. From the 144 families who consented to the study, 54 participants with CdLS were selected based on diagnostic information, age (over two years) and distance to the research base (for full details of inclusion criteria, see Oliver *et al.*, In

review a). These 54 participants were visited in their usual day care setting as part of the larger study and parents and carers were sent a questionnaire pack.

Two methods of recruitment were adopted to identify a matched comparison group. Teachers and key workers of participants with CdLS identified up to two individuals who were similar to the index participant in terms of age, gender, mobility and ability. Secondly, schools and day centres that had already been visited were approached. Information packs were sent to parents and carers of the 876 individuals attending these establishments. 153 (17.5%) consent forms and questionnaires were returned. Participants were included in the current study by matching to the CdLS participants. Matching criteria were age (+/- 2 years), gender, wheelchair use ('never', 'sometimes' or 'often') and self-help skills as determined by the Wessex scores (transformed into 'able', 'partially able' and 'not able'; Kuschlick, Blunden & Cox, 1973). 46 participants were identified for the comparison group.

Table 2 shows participant characteristics in terms of the Wessex self help scale, mobility score, age and gender for the groups. 21 individuals in the comparison group had syndrome diagnoses including: Down Syndrome (n=8), Autism (n=3), Cerebral Palsy (n=2), Congenital Rubella (n=1), Fragile X Syndrome (n=1), Ito Syndrome (n=1), Landau Kleffner Syndrome (n=1), Miller Dieker Syndrome (n=1), Prader-Willi Syndrome (n=1), Soto Syndrome (n=1) and intellectual disability resultant from Reye Syndrome (n=1)

+++++++ Insert Table 2 here ++++++

10.9% of the comparison group participants were on anti-psychotic medication at the time of the study, 23.9% were taking anti-epileptic medication and 31.3% other forms of medication. 3.7% of the CdLS group were taking anti-psychotic medication at the time of testing, 11.1% were taking anti-epileptic medication and 42.6% other types of non-psychoactive medication.

Measures

A measure of background information regarding age, gender and diagnosis was included.

Self-injurious Behaviour

The Challenging behaviour Interview (CBI) (Oliver *et al.*, 2003) is a two-part interview used to assess the prevalence and severity of challenging behaviors displayed by people with intellectual disabilities. In part one, five forms of challenging behaviours (including self-injurious behaviour) are presented. Self-injury is defined as ‘Non-accidental behaviors which produce temporary marks or reddening of the skin or cause bruising, bleeding or other temporary or permanent tissue damage [Examples - Self-biting, head banging, head punching or slapping, removing hair, self scratching, body hitting, eye poking or pressing]’. Informants state whether each of the behaviours have been displayed in the past month. In part two, for each behaviour identified, fourteen questions are asked to determine the severity of the behaviour. Each question has a four or five-point Likert scale, lower scores reflecting less severe behaviour. The scores for each of these questions are summed to provide an overall severity score for each behavior. Psychometric properties of the interview have been calculated, i.e. test–retest reliability, inter-rater agreement, concurrent validity and content validity were generally reported to be good. (Oliver *et al.* 2003).

Health Questionnaire

A health questionnaire, measuring the presence and severity of 15 health problems, was developed for use in the current study. The Tenth Revision of the International Statistical Classification of Diseases and Related Health Problems or ICD-10 (World Health Organization, 1992) was used to classify health problems. Informants are asked to rate the presence and severity (0 = never occurred to 3 = severe problem) of problems occurring ever in the person’s life and over the last month. Scores were summed to produce an Overall Health Score indicating severity of health problems for the previous month and during the person’s life. The total numbers of health problems during the person’s life and the previous month were also calculated

Inter-rater reliability was collected on a sample of 24 individuals by asking two parents or carers to complete the measure independently. Mean item level reliability Kappa co-efficient for health problems ever occurring was .72 (range .32 to 1.00). Item level reliability Kappa co-efficients for the occurrence of health problems over the last month ranged from .32 to 1.00 (mean .76). Intra-class correlation co-efficient scores for the overall health problem score and total number of health problems

occurring over the last month were .65 and .73 respectively. Intra-class correlation co-efficient scores for the overall health problem score and total number of health problems occurring ever during the person's life were .71 and .68 respectively.

The Infant Sleep Questionnaire (ISQ; Morrell, 1999)

The Infant Sleep Questionnaire is a ten item, informant based measure of sleep in infants. The original version was adapted by substituting the phrase 'your baby' to 'the person you care for'. Informants respond to questions relating to the person's sleeping habits over the previous month. Items are collated to give an overall sleep score and the subscales of 'waking at night' and 'going to sleep', with higher scores indicating more difficulties. Comparisons of the ISQ to Richman's sleep diaries have demonstrated good concurrent validity and the test retest reliability correlation co-efficient of the ISQ score is reported to be .92 (Morrell, 1999).

Data Analysis

Due to the high number of statistical tests being conducted an alpha value of less than .01 was used to indicate significance except for those tests used to evaluate comparability of the groups.

Results

Statistical analyses of the matching criteria showed no significant difference between the groups for age ($t(97) = -.08, p=.94$), gender ($\chi(1) = .14, p=.71$), ability ($\chi(1) = .67, p=.72$) and mobility ($\chi(1) = .33, p=.85$).

Health and sleep problems between the CdLS and comparison groups

Comparisons of the type of health difficulties shown by the two groups indicated that significantly more individuals in the CdLS group had experienced eye problems, gastrointestinal problems, problems with genitalia and a hernia during their lives, see table 3. Significantly more individuals with CdLS had also experienced gastrointestinal problems and were affected by their limb abnormalities during the previous month.

+++++++ Insert Table 3 here ++++++

T-test comparisons of the number of health difficulties and the Overall Health score indicated that individuals with CdLS had experienced significantly more health problems and more severe problems during their lives and the previous month than the comparison group (see table 4). No significant differences in participant's sleep scores were found between individuals in the CdLS and comparison groups.

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

Health problems and self-injurious behaviour

Examination of the presence of self-injurious behaviour in individuals with and without specific health problems in the CdLS group revealed that participants who had not experienced eye problems in the previous month were more likely to show self-injury, see table 5. In the comparison group, participants with skin problems were more likely to show self-injury than those without skin problems.

+++++++ Insert Table 5 here ++++++

The results of comparisons of the severity of self-injury between participants who had experienced specific health problems and those who had not experienced health problems are displayed in table 6. The results for the CdLS group indicate that participants with skin problems displayed significantly more severe self-injury than those without skin problems. No significant differences were observed in the comparison group for severity of self-injury between participants with and without health problems.

+++++++ Insert Table 6 here ++++++

Sleep and self-injurious behaviour

Table 7 displays the mean sleep scores and T-test comparisons for participants showing and not showing self-injury in the CdLS and comparison group. Results reveal no significant differences in sleep scores in either group. Correlation analysis also indicated that severity of self-injury was not associated with severity of sleep

problems in the CdLS and comparison groups (CdLS group: $r = -.03$; $p = .87$; Comparison group: $r = .20$; $p = .44$).

+++++++ Insert table 7 here ++++++

Discussion

The results of this study indicate that individuals with CdLS are likely to experience more frequent and severe health difficulties than matched individuals. Specific health problems found to be associated with CdLS include gastro-intestinal problems, eye problems and genitalia and limb abnormalities. In line with previous research, the results of the current study indicate that 71% of individuals with CdLS experience gastro-intestinal problems during their lives, with 44% of participants having been affected by gastro-intestinal problems during the previous month. The proportion of participants with CdLS displaying eye problems (67% during their lives and 31% during the previous month) was also similar to previous findings. The percentages of individuals with abnormalities of the limbs and genitalia in the present study were lower than results found in previous research. This may be due to the definitions employed. However, the results indicate that the prevalence of limb and genital problems is significantly higher in people with CdLS than individuals with mixed aetiology intellectual disabilities and results for the prevalence of genital abnormalities for males only in the current study (58%) is similar to the results of previous research. Overall, the similarity of the data on health conditions in CdLS in this study to previous studies and similarity of distribution of degree intellectual disability in this sample (see Oliver et al., In review) to previously described samples suggest that this sample is representative of the population of those who have CdLS

In accordance with previous findings the current study suggests that around 75% of people with CdLS experience dental problems and 73% have problems with their ears. The prevalence rates in the CdLS group were not significantly higher than those found for individuals with mixed aetiology intellectual disabilities although it is notable that differences approached significance. Previous studies have also suggested that heart defects and epilepsy are associated with CdLS. The prevalence of these difficulties in

CdLS in the current study appears to be high (39% heart defects and 33% epilepsy). However, these figures were not significantly different to prevalence rates found for people with mixed aetiology intellectual disabilities.

Studies examining sleeping difficulties in people with CdLS have found higher rates for sleeping problems than those described in studies of people with mixed aetiology intellectual disabilities. However, this study demonstrates that when compared to matched participants with intellectual disabilities, people with CdLS are not significantly more likely to experience difficulties with sleep. 52% of carers of people with CdLS reported that the person they care for experiences mild to severe problems compared to 44% of carers of individuals in the comparison group. These findings are comparable to previous findings indicating that 55 to 72% of people with CdLS and 32 to 39% of people with intellectual disabilities have sleep problems (Berney *et al.*, 1999; Brylewski & Wiggs, 1998, 1999; Gualtieri, 1990; Sarimski, 1997; Stores *et al.*, 1996).

Examinations of the relationship between the presence of self-injurious behaviour and health problems in the participants with CdLS revealed an unexpected finding. Participants who had not experienced eye problems in the previous month were more likely to show self-injury than those who had experienced eye problems. This is in contrast to predictions that the presence not the absence of health problems might be associated with self-injurious behaviour and is difficult to interpret. The presence of sight impairment might be expected to be associated with the presence of both eye problems and self-injury. Although some research has indicated that visual impairments may be associated with self-injurious behaviour (Collacott *et al.*, 1998; Nøttestad & Linekar, 2001), more rigorous studies have found no association between self-injury and visual problems (Chadwick, Piroth, Walker, Bernard & Taylor, 2000; Murphy, Hall, Oliver & Kissi-Debra, 1999).

In CdLS Health problems appear to be associated with the severity of self-injurious behaviour rather than simply its presence. Individuals with CdLS experiencing skin problems displayed more severe self-injury than those without skin problems and a similar relationship between self-injury and dental problems approached significance. However, in both instances causal direction was not investigated. This is a novel

finding that has not previously been reported and indicates that the presence of health problems might be associated with the severe self-injurious behaviour often reported in CdLS. Previous research indicating an association between ear infections and gastroesophageal -reflux and self-injury, and the findings reported by Luzzani *et al.* (2003) suggesting that gastro-intestinal difficulties might be associated with self-injurious behaviour in CdLS, were not replicated in this study. Luzzani *et al.* utilised ph and endoscopy examinations to measure gastro-intestinal problems and due to the difficulties communicating the symptoms of health problems for people with severe intellectual disabilities the current study may not have identified all individuals experiencing gastro-intestinal problems. The observation that Luzzani *et al.* identified 65% of individuals to be currently experiencing gastro-reflux compared to 44% in this study supports this possibility. In summary, it appears that the presence of health problems in CdLS may be associated with the severity of self-injurious behaviour but not the presence.

The findings of the study indicate that individuals with CdLS do not show a different pattern of sleep problems when compared to matched individuals with mixed aetiology intellectual disabilities and sleep problems are not related to self-injury. Difficulties with sleep were not associated with presence or severity of self-injury in the CdLS or the comparison group. This is in contrast to previous research on sleep and self-injury in people with intellectual disabilities showing that children and adults showing self-injury are significantly more likely to have difficulties with sleeping than individuals without self-injury (Brylewski & Wiggs, 1999; Piazza, Fisher, & Shorer, 1997; Symons, Davis, & Thompson, 2000). Differences in these findings may be explained by the use of questionnaire measurement. Informant based questionnaires rely on an awareness of the participant's sleep and carers may not always be aware of the participant's sleep disturbances. More detailed observational examinations of sleep are warranted.

Due to the limited instruments available for the measurement of the health status and sleeping behaviour of people with intellectual disabilities, in this study we employed new and adapted measures in order to examine these domains. An adapted version of Infant Sleep Questionnaire was incorporated, with demonstrated reliability and validity. However, the psychometric properties of this measure have not been

examined for use with adults with intellectual disabilities. Although utilising carer reports of health issues provides information regarding immediate and relevant health difficulties, and can help to limit the potential threat of biased samples associated with methodologies such as clinical examination and reviews of case notes, the construct validity of carer reports remains problematic. Efforts were made to ensure the face validity and reliability of the health measure included in this study. However, further examination of the construct validity of the health questionnaire is warranted. A related methodological issue for this study was the use of a sample that is in contact with the syndrome support group and thus exposed to information about health conditions and this might not have been the case for the comparison group.

The examination of health and sleep in this study revealed that individuals with CdLS are likely to experience frequent and severe health problems. In particular, problems with the gastro-intestinal system, eyes, limbs and genitalia appear to be associated with CdLS. This high level of health problems might account for the higher ratio of facial expression of negative to positive affect observed in the syndrome (Collis et al., In press). Individuals with skin problems and possibly dental problems are likely to show more severe self-injurious behaviour although causal direction needs to be established. Given the limited communication skills of some people with CdLS, and therefore the difficulty indicating the symptoms and existence of health problems, these findings highlight the importance of an awareness of the potential health problems for professionals working with people with CdLS. Furthermore, as nearly three quarters of individuals with CdLS are likely to experience gastro-intestinal problems, it is reasonable to suggest that robust screening processes are recommended for all people diagnosed with CdLS.

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Health and sleep in Cornelia de Lange syndrome

CdLS Studies	N	Percentage of individuals experiencing health problems											
		limb abnorm- alities	gastro- intestinal	heart	dental	genit- alia (males)	genito- urinary	eye	ear	respira- tory	epile- psy	skin	cleft palate
Hawley <i>et al.</i> , 1985	64	33%	71%	28%	93%	94%	-	-	-	-	14%	-	-
Gualtieri 1990	138	-	41%	15%	-	-	8%	-	-	18%	25%	-	-
Sataloff <i>et al.</i> , 1990	45	-	-	-	-	-	-	-	-	-	-	-	59%
Ireland <i>et al.</i> , 1993	20	80%	10%	15%	-	-	-	-	-	-	5%	-	10%
Jackson <i>et al.</i> , 1993	310	-	48%	25%	-	73%	12%	50%	60%	25%	23%	16%	-
Sommer 1993	17	-	76%	-	-	-	-	-	-	-	-	-	-
Kousseff <i>et al.</i> , 1994	37	56%	49%	14%	-	-	33%	38%	-	-	38%	-	21%
Berney <i>et al.</i> , 1999	49	-	67%	-	-	-	-	-	-	-	29%	-	-
Tsukahara <i>et al.</i> , 1998	50	-	-	26%	-	-	-	-	-	-	-	-	-
Luzzani <i>et al.</i> , 2003	43	-	65%	-	-	-	-	-	-	-	-	-	-

Table 1: Percentage of individuals with CdLS showing specific health problems in published studies.

		CdLS Group	Comparison Group
Age (in years)	Mean	13.88	13.74
	(SD)	(8.58)	(7.99)
Gender	% Male	46	50
Wessex self-Help	% Not able	46	42
	% Partly able	41	49
	% Able	13	9
Wheelchair Use	% Always	11	11
	% Occasionally	33	28
	% Never	56	61

Table 2: Mean age and standard deviation and percentage of males, levels of ability and levels of mobility for CdLS and comparison groups

	Percentage of individuals showing health problem		Chi-statistic	p-value
	CdLS (N=52)	Comparison (N=44)		
Health Problems – ever during the person’s life				
Eye problems	66.7	36.4	8.70	<0.01*
Ear problems	73.1	50.0	5.42	0.02
Dental problems	75.0	51.2	5.82	0.02
Cleft Palate	18.0	4.5	4.10	0.04
Gastrointestinal problems	71.2	31.8	14.81	<0.01*
Bowel problems	26.9	29.5	0.08	0.78
Heart problems	38.5	22.7	2.75	0.10
Genitalia problems	28.8	4.5	9.66	<0.01*
Hernia	25.0	2.3	9.88	<0.01*
Limb abnormalities	44.2	18.2	7.40	0.01
Epilepsy	33.3	36.4	0.10	0.76
Lung problems	33.3	31.8	0.03	0.88
Liver problems	6.0	6.8	0.03	0.87
Diabetes	2.0	0.0	0.89	0.35
Skin problems	52.9	36.4	2.62	0.11
Health Problems – in the previous month				
Eye problems	30.8	13.6	3.96	0.05
Ear problems	30.8	11.4	5.25	0.02
Dental problems	30.8	13.6	3.96	0.05
Cleft Palate	3.8	2.3	0.20	0.66
Gastrointestinal problems	44.2	15.9	8.90	<0.01*
Bowel problems	23.1	13.6	1.39	0.24
Heart problems	5.8	2.3	0.73	0.40
Genitalia problems	5.8	0.0	2.62	0.11
Hernia	3.8	0.0	1.73	0.20
Limb abnormalities	34.6	4.5	13.07	<0.01*
Epilepsy	7.7	9.1	0.06	0.81
Lung problems	15.4	9.1	0.86	0.35
Liver problems	0.0	0.0	-	-
Diabetes	1.9	0.0	0.86	0.36
Skin problems	44.2	27.3	2.96	0.09

Table 3: Chi-square test results and percentage of individuals showing topographies of health problems ever and in the previous month (* indicates significant results).

Health and sleep in Cornelia de Lange syndrome

		CdLS	Comparison	T	p-value
Health Questionnaire					
<u>Health problems in the previous month</u>					
Overall health score	Mean	4.67	2.00	4.69	<0.01*
	<i>standard deviation</i>	3.53	1.93		
No. of health problems	Mean	2.82	1.23	5.13	<0.01*
	<i>standard deviation</i>	1.91	1.20		
<u>Health problems during the person's life</u>					
Overall health score	Mean	11.81	5.95	5.55	<0.01*
	<i>standard deviation</i>	6.19	4.06		
No. of health problems	Mean	5.90	3.61	4.88	<0.01*
	<i>standard deviation</i>	2.38	2.18		
Infant Sleep Questionnaire					
Overall sleep score	Mean	12.03	11.67	0.20	0.85
	<i>standard deviation</i>	9.15	8.54		
Going to bed score	Mean	5.58	5.14	0.45	0.65
	<i>standard deviation</i>	4.73	4.51		
Waking at night score	Mean	6.08	5.95	0.12	0.90
	<i>standard deviation</i>	5.10	4.82		

Table 4: T-test analyses and mean and standard deviation scores for CdLS and comparison groups for health and sleep problems.

Health and sleep in Cornelia de Lange syndrome

Health problem in last month		% of CdLS group showing self-injury			% of comparison group showing self-injury		
			Chi Square	P-value		Chi Square	P-value
Eye	Present	25.0	8.87	<0.01*	50.0	0.24	0.626
	Absent	69.4			39.5		
Ear	Present	56.3	0.00	0.963	40.0	0.00	0.965
	Absent	55.6			41.0		
Dental	Present	62.5	0.42	0.515	22.2	1.91	0.167
	Absent	52.8			36.8		
Gastro	Present	60.9	0.44	0.510	11.1	0.52	0.469
	Absent	51.7			43.2		
Hernia	Present	50.0	0.03	0.867	-	-	-
	Absent	56.0			40.9		
Skin	Present	56.5	0.01	0.922	75.0	7.93	<0.01*
	Absent	55.2			28.1		

Table 5: Percentage of individuals showing self-injurious behaviour with and without specific health problems in the CdLS and comparison groups.

Health and sleep in Cornelia de Lange syndrome

Health problem in last month		CdLS group			Comparison group		
		mean self-injury severity score	T	p-value	mean self-injury severity score	T	p-value
Eye	Present	11.50	1.62	0.118	9.67	1.15	0.269
	Absent	16.56			13.60		
Ear	Present	14.44	0.85	0.402	9.50	0.94	0.362
	Absent	16.50			13.37		
Dental	Present	18.90	-2.10	0.045	11.75	0.48	0.636
	Absent	14.26			13.29		
Gastro	Present	16.21	-0.30	0.765	11.50	0.39	0.705
	Absent	15.53			13.13		
Hernia	Present	17.00	-0.19	0.851	-	-	-
	Absent	15.82			12.94		
Skin	Present	19.00	-2.85	<0.01*	11.33	1.27	0.223
	Absent	13.31			14.56		

Table 6: Self-injury severity scores for participants with and without health problems in the CdLS and comparison groups

Presence of self-injury	CdLS group mean sleep score	T	p-value	Comparison group mean sleep score	T	p-value
Present	12.66	-0.53	0.600	10.23	0.89	0.377
Absent	11.30			12.64		

Table 7: Mean sleep scores for participants showing and not showing self-injurious behaviour in each group.