

Behavioral challenges in children and adults with CdLS

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The term ‘behavioral phenotype’ is used to describe a strong association between specific behavior and a genetic syndrome. Understanding the associations between behavior and genetic syndromes provides very useful information for all people with an intellectual disability as well as those who have specific syndromes. As we continue to study behavior in Cornelia de Lange Syndrome (CdLS) we often learn about the causes of behavior that are shown across all people with an intellectual disability. This information helps to guide education, treatment, assessment and early intervention strategies for people with CdLS and others. This is one of the main reasons why we study behavioral phenotypes.

It is worth remembering that a number of medical and health problems are associated with CdLS that might be relevant to behavioral problems including gastrointestinal, eye, dental, ear and orthopedic problems. We will only briefly mention these problems as more detailed information is available elsewhere. It is important to know the health problems that are associated with CdLS and how they might be effectively treated. This is always the first step in trying to understand change in behavior.

Previous research into the behavioral characteristics of the syndrome has largely focused on the prevalence and nature of self-injurious behavior in CdLS and possible underlying causes. More recently, research has widened and has begun to consider other behavioral characteristics of the syndrome. Interest in these ‘new’ areas of research in CdLS have enabled a broader understanding of the syndrome and has provided a good foundation for future research, assessment and intervention. In this article we focus on three areas of behavior that we feel are key to understanding the problems faced by people with CdLS and their caregivers. These are self-injurious behavior, autistic-like characteristics and behavioral changes with age. In each section we outline the nature of the behavior, the possible causes of the behavior and, most importantly, we outline the strategies we think may help with these problems.

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Self-injurious behavior in Cornelia de Lange syndrome:

What is the nature of self-injurious behavior in CdLS?

Self injurious behavior occurs in about 60% of children and adults who have CdLS at a level which causes tissue damage (bruising or bleeding) at least once in a month. Severity and frequency vary significantly between people from an occasional scratch, bang or bump to



frequent self-injury resulting in open wounds in a small minority. The most important things to know about self-injury in CdLS are that it is not inevitable and that it can be prevented and reduced with the right kinds of assessments and interventions.

It is worth noting that whilst self-injury is common in CdLS it is no more common than we would expect by chance. If we compare the prevalence of self-injury in CdLS with that seen in people who are matched for degree of disability and mobility, age and sex then there is very little difference. What does differ between these groups are the more mild forms of self-injury (very mild skin picking and the occasional bump of the head for example) which we see much more commonly in CdLS than a matched group. The other difference is that in CdLS the self-injury is much more likely to take the form of biting and to be directed towards the hands\lower arms.

Self-injury has a significant impact on quality of life for everyone concerned and deserves the attention of researchers and clinicians alike. Here we outline the causes and treatments for self-injury in CdLS.

What are the causes of self-injurious behavior in CdLS?

There are three main causes of self-injurious behavior in CdLS and there are other factors that can make things worse when any of these causes is responsible. The most important thing to do is to keep an open mind about the possible causes of self-injury and then to systematically work through assessments to address each cause.

1. Pain and discomfort

The research of Angelo Selicorni and his colleagues in Milan has confirmed the clinical observation of many people that gastro-esophageal reflux is associated with self-injury. The evidence for this is mounting with the Milan group showing that self-injury decreases when reflux is treated and we have recently found that self-injury is significantly associated with reflux related behavior such as back-arching, excessive drinking in the morning, teeth-grinding, touching the throat and chest area and a number of other behavior. Another important finding is that the behavioral signs of reflux are not just associated with self-injury but also with aggression and damage to the environment. We have just developed a questionnaire that lists all the reflux associated behavior and we hope it will be ready for distribution soon. There should be no doubt in anyone's mind that reflux causes self-injury and must be properly investigated before anything else is done. These investigations should take place even if it is felt that reflux

has been successfully treated in the past because things can change over time.



It is important to remember that there are some other common causes of pain for people with CdLS such as middle ear infection, crowding of teeth, tooth decay, pain in the hips and discomfort in the sinuses (possibly related to reflux). These should always be investigated even if the self-injury may not be directed toward where you think the source of the pain might be. People can self-injure to block chronic pain and a common example is hair-pulling.

2. A learned behavior

Our most recent research shows that for some people with CdLS self-injury can be a learned behavior. In other words the behavior occurs because it is followed by a rewarding event or interaction. Most commonly this is contact with someone with whom the person would like to spend time, no matter how brief this time might be. The other common reason is that the person wants something to stop or go away. This is usually a task or chore but can sometimes be an overwhelming social situation (see below).

To assess whether the self-injury is learned you can keep charts that will show you whether there is a pattern to something in the environment triggering the behavior. These are useful for behavior that occurs between around five and ten times a day. If the behavior is more frequent then a more structured assessment is required. The most commonly used method is called experimental functional analysis (also known as analogue conditions). This can be conducted by an applied behavior analyst or clinical psychologist and involves systematically manipulating levels of social contact and tasks to see if the self-injury is associated with a lower or higher level of attention or the presence of a task. When we used this method in our recent research, we found that seven out of ten people with CdLS showed evidence that the behavior was learned.

3. Anxiety and low mood

From interviews with parents and caregivers, particularly of more able young adults with CdLS, we think that some forms of self-injury are associated with anxiety (see below). At present we only have anecdotal evidence for this and we need to do more research. The most common form of self-injury in this case is picking at the arms and legs and this may or may not break the skin. This behavior can occur when the person feels under stress and this is often related to an upset in routine or a change in the person's broad social environment, for example moving to a new school. If health problems are ruled out then this mild picking of arms or legs might be related to increased anxiety (that can sometimes look like low mood) and environmental causes of this must be assessed.

What parents and caregivers can do to reduce self-injurious behavior

For self-injury that is related to health conditions the best way to intervene in an effective way is to resolve any pain or discomfort. This means seeking the right assessment of a health condition and, if necessary, doing this assertively. Theodore Roosevelt gave good advice on



how to approach this when he said “Speak softly and carry a big stick”. The first assessment should be for reflux, especially if there are other signs that this might be present. After that the other well known health problems associated with CdLS should be investigated and treated.

It is said that prevention is better than cure and this is certainly true for learned behavior. Even if the self-injury is initially occurring in response to pain, it is possible for the person to learn to show self-injury if the behavior is followed by a reward. When you first see self-injurious behavior, be careful how you respond. Try to delay your natural protective response by saying to yourself “One, two what should I do?” This will give you time to check whether you are about to reward the behavior. You can then work out a way of responding that keeps the child safe but is not rewarding.

There are two main components to any effective strategy for learned behavior and applied behavior analysts and clinical psychologists should help with these. The first is finding a way of responding to the behavior that is not rewarding, whilst protecting the child from harm, and the second is teaching the child an effective way of communicating what they have learned to say with their self-injury. This is called Functional Communication Training and can be very effective in the long term. These interventions need planning and everyone needs to be on board. This is not an easy task but it is important. People learn quickly when the same outcome is always achieved. For self-injury that appears to be related to anxiety then the strategies described below can be helpful.

Three things tend to make self-injurious behavior worse in CdLS. The first is the problem with inhibiting behavior that we see in many people with CdLS (see below). This means that when a behavior is rewarded it is very difficult for the person to stop showing the behavior in the future and this is one reason why prevention is better than cure. The second is the likely impaired pain perception that we think many people with CdLS have in their arms. This may mean that if the behavior is learned the natural pain that would normally be experienced, when biting for example, does not check the behavior and so the bite can be harder than would otherwise be the case. The third is the poor expressive communication that children and adults with CdLS experience. This means that effective communication that can access rewards from others is hampered and self-injury is a behavior that can replace poor communication in a very effective way.

It is certainly not inevitable that anyone who has CdLS will show self-injury. The work of the Foundation in raising awareness of painful medical conditions such as reflux has without doubt contributed to a decrease in the overall number of people with CdLS who start to show self-injury. This raised awareness of health conditions is important and we must do more to try to understand how people show pain when they cannot communicate. We are also working on early detection and intervention for self-injury. This is a large scale project involving other syndrome groups who face similar problems. Finally, the most important



thing to remember when you are confronted with self-injurious behavior is that it doesn't have to be like this.

Autistic Spectrum Disorder in Cornelia de Lange Syndrome:

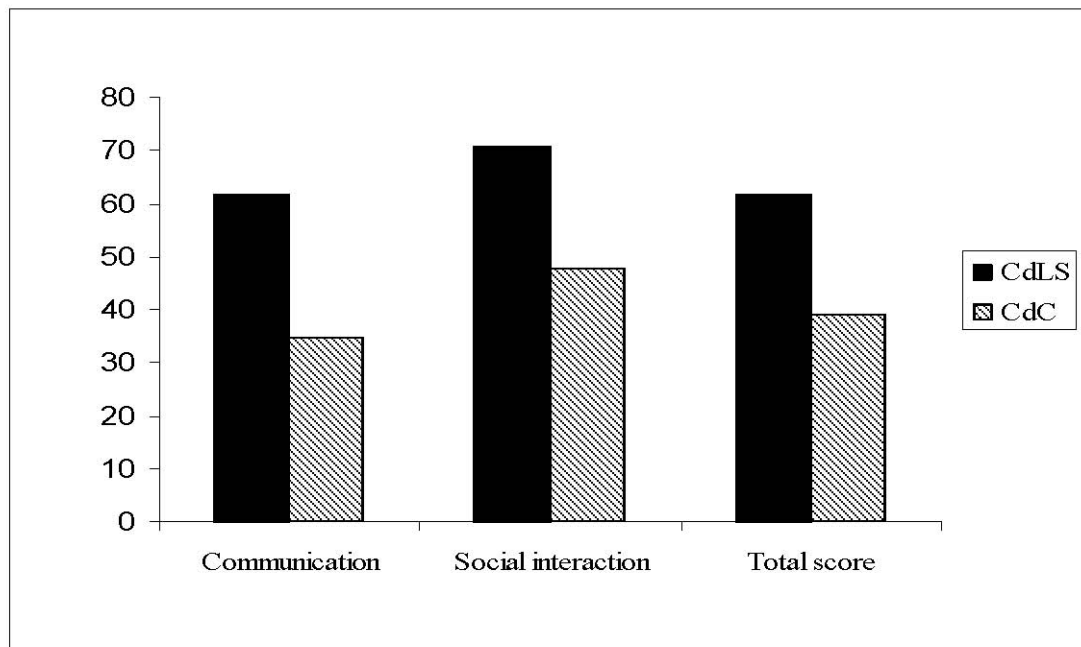
What is the nature of autistic spectrum disorder in CdLS?

Several recent studies of individuals with CdLS have indicated that autistic-like characteristics are associated with CdLS. The three core characteristics of autistic spectrum disorder are:

1. Poor or unusual social interaction skills.
2. Delayed development or difficulties in both verbal and non-verbal (e.g. gestures, pointing and showing) communication.
3. The presence of repetitive behavior, an insistence on sameness, restricted interests and behavior such as lining objects up.

In trying to understand autistic spectrum disorder in CdLS it is important to compare children and adults with the syndrome with other children with the same degree of intellectual disability. This means we can be sure that any differences are associated with the syndrome rather than just being caused by an intellectual disability. When we do this our research shows that people with CdLS are more likely to reach the diagnostic cut-off scores for autism on an observational assessment of autistic characteristics than people with Cri du Chat syndrome (CdCS) (who have a comparable level of ability and receptive language skills). A diagnostic cut-off score is the minimum score which an individual with a confirmed diagnosis of autism would achieve. Figure 1 shows the percentage of individuals with CdLS and CdCS who score above the diagnostic cut-off for autism on the communication scale, the social interaction scale and total score of the Autism Diagnostic Observation Schedule. The figure shows that approximately 60% of people with CdLS score on this assessment at a level that would be expected to be observed in individuals with autism. This does not necessarily mean that all individuals with CdLS should receive a diagnosis of autism. Rather it suggests that autistic-like characteristics are common in individuals with CdLS and that some people with CdLS might be considered to fall within the autistic spectrum.

Figure 1: Percentage of individuals with CdLS and Cri du Chat syndrome (CdCS) who score above the diagnostic cut-off for autism on the Autism Diagnostic Observation Schedule.



The graph shows that more individuals with CdLS than individuals with CdCS score above the diagnostic cut off for autistic spectrum disorder. The diagnostic cut off score indicates the minimum level that an individual with confirmed diagnosis of autistic spectrum disorder would achieve.

When we look at the nature of the autistic-like characteristics in people with CdLS in more detail, our findings suggest that impairments in communication are the most prominent autistic-like characteristic associated with the syndrome. Although we have yet to identify the nature of communication impairments in CdLS more precisely, our results and observations, and those of past research, suggest that both verbal and nonverbal communication skills are compromised. Additionally, in those individuals who develop verbal communication, reluctance to use speech in particular environments, social situations or with unfamiliar people seems to be a very common problem. This is called selective mutism and we think the prevalence may be as high as 10% in people with CdLS who are able to speak.

In addition to impairments in communication, our results suggest that social interaction skills might also be compromised in CdLS. However, we think that the nature of these difficulties might be somewhat different to those observed in individuals with a diagnosis of autism. From our observations and consultations with parents, social difficulties in people with CdLS

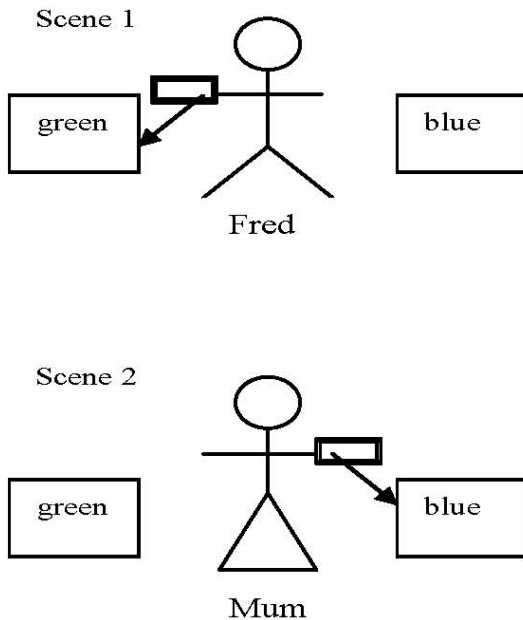
are likely to take the form of social anxiety. This means that people with CdLS might find social situations highly anxiety provoking; this in turn might lead to social withdrawal or reluctance to take part in group activities. Others might appear extremely shy when around unfamiliar people and take time to ‘warm up’ in unfamiliar environments. Physical signs of anxiety include becoming pale, starting to shake, avoiding eye contact and looking tense and uncomfortable.

With regard to repetitive behavior our recent research suggests that “compulsive-like” behavior is very common in CdLS, particularly tidying up and lining up behavior. These types of repetitive behavior are also commonly observed in children and adults with autistic spectrum disorders.

What is the cause of autistic like characteristics in CdLS?

There are several explanations for the social, communicative and behavioral difficulties experienced by people with autistic spectrum disorders. One of these, which might help us to understand the cause of autistic-like characteristics in people with CdLS, such as difficulties with social interactions, is the ‘Theory of Mind’ account. This suggests that people with autistic spectrum disorders do not develop the ability to understand that their own thoughts and beliefs about the world may be different to those of other people. This means that when faced with a social situation people may find it difficult to understand or ‘read’ the social situation and thus social interactions become anxiety provoking (see Box 1).

Box 1. The ‘Theory of Mind’ account of autism, autistic spectrum disorder and autistic-like characteristics



The false belief task:

Fred puts his bar of chocolate in the green box and then leaves the room. While he is away, his Mum moves his chocolate bar to the blue box.

Where will Fred look for his chocolate bar when he comes back?

Children with autistic spectrum disorders find it very difficult to understand that Fred has different knowledge about the situation to themselves and will be more likely to answer that Fred will look in the blue box for his chocolate as this is where *they* know the chocolate is. Children with good theory of mind skills understand that Fred will look in the green box, as this is the last place *he* saw the chocolate.

A second theory, the ‘Executive Dysfunction’ account, might be useful in explaining some of the repetitive behaviors observed in the syndrome. This theory suggests that people with autistic spectrum disorders have difficulty in regulating their behavior. This includes stopping an ongoing response or behavior, generating new responses and behaviors when they are needed and not starting inappropriate responses or behaviors. This means that people might find it difficult to move from one activity or from one behavior to another (and so keep repeating the same behavior or activity) and find it difficult to not start a behavior or response when that behavior is not appropriate (even though they “know” they should not show a behavior; see Box 2).

Box 2. The Executive Dysfunction account of autism, autistic spectrum disorder and autistic-like characteristics

RED BLUE YELLOW

RED BLUE YELLOW

RED BLUE YELLOW

The Stroop Task:

In this task, the individual is asked to read the colour of the ink the word is printed in and not the word itself. In doing so, the individual must inhibit the typical response to read the word and focus on the colour of the print. People with autistic spectrum disorders find it difficult to inhibit the typical response and might be slow to complete this task. Try to do this quickly and you will see what we mean!

What parents and caregivers can do to help with autistic characteristics in CdLS:

The nature of autistic-like behaviors and characteristics in people with CdLS is extremely variable. Consequently, the way in which these behavior and characteristics are managed will vary according to the level of ability of the individual and the precise nature of the difficulties. The following section provides a brief overview of some of the types of interventions that have been demonstrated to be useful in helping individuals with autistic spectrum disorders.

Communication skills:

For those individuals who have not developed verbal communication, encouraging nonverbal communication skills (such as gesturing, pointing and showing), the use of objects of reference or formal sign or picture exchange system, such as Makaton sign language or PECS, may be important. However, input from speech and language therapists and educational specialists are critical to make sure the method is matched to the child. Rewarding the use of nonverbal communication skills by responding to the individual in some way (this might be responding to a request or simply engaging with the person) may help to encourage the individual to use these techniques more consistently and increase motivation to use their skills. The aim in this situation is to ensure that formal nonverbal



strategies are more effective or efficient for the individual than other less appropriate communication strategies that the individual might have developed.

For more able individuals who develop verbal communication skills, role-play techniques can be used to teach people to increase the range of conversational topics and encourage more sophisticated conversational skills. This technique can be used to practice how an individual might introduce himself in a social situation or respond to common social questions. Encouraging initiation of conversation can be more difficult to develop.

Social interaction skills:

For younger children, or those who have not yet developed communication skills, a technique called Structured Interference has been used in individuals with autistic spectrum disorder to encourage and facilitate social interaction. Structured Interference helps the individual to understand the value of social interaction with others. The technique involves parents or caregivers engaging with the individual during what might typically be solitary play and withholding an object that might be required to complete the activity such as a piece of a jigsaw puzzle or a block for building. The individual would need to engage in some way with the adult in order to obtain the object and finish the activity. This technique can be extended to encourage individuals to engage with siblings.

For older children or those who have developed verbal communication skills, teaching simple conversational skills and appropriate responses to social questions can be helpful. Role-play techniques similar to those described above can be a useful way for the individual to 'practice' social interaction skills and be better equipped for social situations when they arise.

Repetitive behaviors:

For some children and families, repetitive behaviors, rituals or insistence on sameness behaviors are a source of great difficulty and disruption to family life. As the individual gets older, repetitive and restricted interests might become increasingly marked and restrict the individual and their family from engaging in other activities. It is important to note that repetitive behaviors may be difficult to stop completely and even if this is achieved, there is a good chance that the individual will begin to engage in a different type of repetitive behavior in its place. Consequently, the aim is to manage repetitive behaviors by reducing the severity or frequency of the behavior, establishing rules regarding when and where the individual is allowed to engage in the behavior or adapting the focus of the behavior to something that is more appropriate. Using these management techniques early on in younger children or when the behaviors first start to appear might make these behaviors easier to manage in the future.

A popular technique used to reduce or replace repetitive behavior in individuals with autistic spectrum disorder is the 'Graded Change Technique'. This approach works to gradually change the repetitive behavior by slowly, but progressively, changing the nature, severity or



frequency of the behavior. For example, slowly limiting the number of objects that the individual is allowed to line up or restricting the number of rooms in the house in which the individual is allowed to engage in the behavior, limiting the number of 'special objects' the individual is allowed to carry with them or establishing rules regarding when and where the individual is allowed to carry the objects with them. For many families, insistence on sameness and adherence to routine can be very problematic. In such situations, making sure that changes are predictable can help. Giving the individual plenty of warning about any changes to the environment or routine by talking about what will happen, when it will happen and developing a timetable for the change might help to make the changes less anxiety provoking for the individual.

These are just some of the strategies that might help. Given that there is some overlap between the problems experienced by children and adults with CdLS and those with an autistic spectrum disorder, it is always useful to look at the strategies recommended for children and adults with autism.

Behavior change in adolescents and young adults with CdLS

There is some research that shows that some adolescents and young adults with CdLS show a change in their behavior and may experience low mood and/or anxiety as they become older. This research is at an early stage and we have yet to answer several questions such as: what is the true nature of this change? (is it really low mood and social anxiety or something else?), how many people show this change? (it may be that we have seen more people with these problems because parents and carers know we are interested in this area) and what might be the cause of this change?

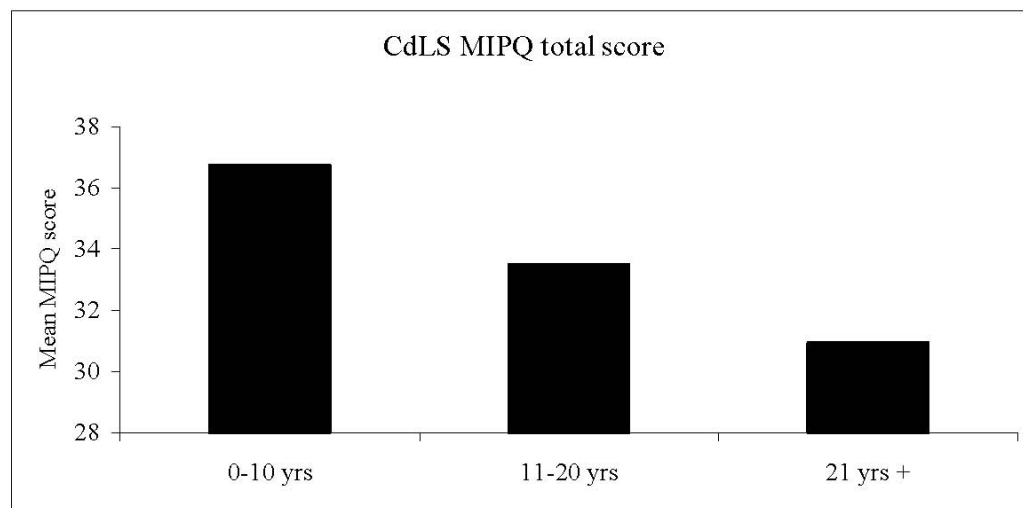
What is the nature of this change?

Our initial research shows that the change in behavior can take three main forms and these may occur in any combination. These include low mood and loss of interest and pleasure in activities once enjoyed, increased social anxiety or generalized anxiety, and an increase in or first appearance of aggressive outbursts or self-injurious behavior.

1. Low mood and a loss of interest and pleasure in activities once enjoyed.

Our recent research has indicated that some individuals with CdLS seem to show lower levels of mood and interest and pleasure in activities with age (see Figure 1). This may appear as: lack of motivation, loss of appetite, loss of self help skills previously learned including washing and dressing, appearing more tearful or tired and feeling more unwell than usual.

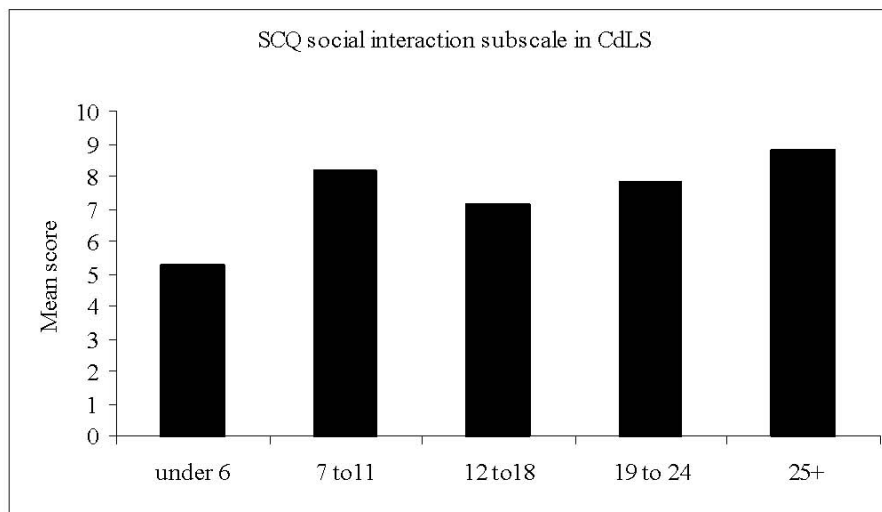
Figure 2: Lowering of mood and interest and pleasure in activities with age in CdLS



We used the Mood Interest and Pleasure Questionnaire (MIPQ), which is a measure of mood suitable for individuals with a range of intellectual abilities, in children and adults with CdLS. A lower score on this measure indicates lower levels of mood. The graph demonstrates the way in which scores on the MIPQ become significantly lower with age suggesting that changes in mood may occur as individuals become older.

2. **Anxiety / Social anxiety.** In the previous section, we described the presence of social interaction difficulties in individuals with CdLS that were associated with the presence of autistic spectrum disorders or autistic-like characteristics. We described the way in which people with CdLS might find social interactions highly anxiety provoking and how this might appear in people with CdLS. Our recent research shows that social interaction difficulties might become more pronounced as people with CdLS become older (See Figure 3). This might mean that social situations become more difficult for individuals to cope with or more anxiety provoking as they become older because of the increasing social demands that are made on people. See box 3 for an account of a social outing for young adults with CdLS which was organised by Lisa Collis, working with the CdLS Foundation (UK and Ireland). In some individuals, anxiety may not be restricted to social situations but might be more generalised, for example a person may feel anxious if there is a change in their routine (see below).

Figure 3: Changes in social interaction skills with age in CdLS



We used the Social Communication Questionnaire (SCQ), which is a measure of autistic like characteristics. In this measure a higher score indicates the presence of greater difficulties. The graph describes scores on the social interaction domain of the SCQ only. The graph demonstrates the way in which scores on the SCQ become significantly higher with age suggesting that changes in social interaction abilities might occur with age.

3. **An increase or the first appearance of aggression towards others, outbursts and self-injury (usually in the form of skin picking).** As discussed above, self-injurious behavior is reported to occur in around 60% of individuals with CdLS, although this is not considered to be significantly higher than expected given the degree of intellectual disability associated with the syndrome. In contrast to this, our research suggests that people with CdLS are significantly less likely to demonstrate aggression in comparison to other individuals with a similar level of ability who do not have CdLS. However, during adolescence and early adulthood some parents have reported an increase in the frequency and intensity of aggressive behavior (and self-injurious behavior). In some people, parents report that these behaviors occur for the first time during this period of change. Box 4 describes a young man who demonstrated an increase in aggressive outbursts during the period of early adulthood. For this young man, it is possible that changes at school triggered the aggressive behavior and when he moved to a day provision with less demands and more structure he appeared happier and the aggression decreased.

Box 3: An account of the social outing for young adults with Cornelia de Lange Syndrome

Background: Parents of some mildly affected individuals with CdLS have reported that their young adults sometimes feel they find it difficult to make friends. To find out more about this, a social outing to the theatre and a meal at a restaurant with five adolescents/young adults with CdLS was arranged, to encourage development of social interaction skills and friendships among individuals. This is an account of the outing.

Interaction: During the outing, four of the five individuals with CdLS didn't appear to start conversations with each other or with the volunteers who had come on the trip. This may have been because some individuals with CdLS experience social anxiety and so find interactions with unfamiliar people extremely difficult. Also, some people with CdLS may have a general difficulty with initiating conversations. For example, one parent commented that their child would never start a conversation even with a family member, which may mean that some people with CdLS take a very passive role in conversations. This would mean that they may respond if a person spoke to them but they are less likely to start talking to someone.

Social anxiety: During the outing, two individuals with CdLS seemed to experience high levels of anxiety that resulted in the individuals feeling unwell and having to remove themselves from the situation. Both individuals experienced this anxiety at the theatre and at the restaurant. The anxiety took the form of breathlessness, feeling faint and feeling sick. The anxiety experienced in the theatre may have been in response to the crowded unfamiliar setting and/or the fact that they did not know the volunteers or their peers. The anxiety experienced at the restaurant may have also been because of the high social expectations of having meal, such as having to interact with other people.

Coping strategy: During the performance at the theatre, one person with CdLS felt that she needed to make several phone calls to various family members despite missing some of the show. She may have done this to help re-establish order and predictability in this new situation. After making the phone calls, the person appeared happy and relaxed so it seemed that this was an effective coping strategy for dealing with this new situation.

Despite the ups and downs during the trip, the general feedback from parents and individuals showed that most people had enjoyed their afternoon and everyone learned a good deal about the problems that people were experiencing.

Box 4: Example of an individual with CdLS demonstrating an increase in aggressive outbursts during early adulthood

Over a period of five years Darren started to show a deterioration in his behaviour, particularly at school. This mainly took the form of experiencing obsessions with particular people and being aggressive towards them. Darren would repeat the particular person's name for several hours during the day and when the chance became available he would pinch them or push them. In addition, if the person that Darren was preoccupied with was in the same room, he would often show behavior indicative of anxiety; Darren would appear white and clammy, he would shake, bite his hand and pick his fingers. At this point Darren would be more likely to show aggression towards that person. The obsessive-like behaviour occurred approximately once a week initially and over time it gradually increased to every day.

By Christmas 2005, Darren left school because it caused him too much distress. Since leaving school, Darren seems much happier and is showing much less aggressive behaviour. Darren has recently started to attend a day unit one day a week, which caters for people with autism. The unit is very small and highly structured. Darren currently seems to enjoy attending the unit and when he arrives home afterwards, he appears happy and relaxed. In addition, the reduction in Darren's aggressive behaviour has remained consistent since starting the new placement.



What is the cause of this change?

There are a number of possible causes for these changes. The two main reasons for this change are:

1. **Pain and discomfort.** Pain and discomfort can lead to people experiencing low mood. Possible causes of pain and discomfort should always be investigated as a first step. The common causes of pain and discomfort in children and adults with CdLS are: reflux, middle ear infection, tooth infection, sinus disorders and hip problems. Other health problems can be experienced by children and adults with CdLS such as, kidney problems and bowel problems that can also cause people to feel low or lead to behavior change. We cannot emphasize enough the importance of investigating physical health conditions when there is behavior change.
2. **Life events and significant change:** From our observations, we have noted that the period of low mood or behavioral change in people with CdLS often seems to arise around the time of a life event or significant change (such as illness, bereavement in the family, moving house, changing from a school to a college or change of teacher). Many people with CdLS appear to show 'autistic-like' characteristics (see above), particularly a strong preference for routine and insistence on the sameness, which means that they can find such changes extremely difficult to cope with, due to the unpredictability and possible increase in social demands that these changes bring. This change may increase the person's level of anxiety. If the situation persists and the person repeatedly finds the situation overwhelming they may feel unable to cope with the related anxiety. This persistent heightened anxiety might, in turn, lead to the person experiencing low mood. For some people, aggression or behavior outbursts may also occur and these may lead to the person being excluded or removed from the new situation. This removal can act as a reward for this behavior because it has enabled the person to change their environment and re-establish predictability with a return to how things were before. See box 5 for an example of some of the difficulties a person with CdLS experienced during the transition from school to college.





Box 5: Example of difficulties experienced by a person with CdLS during the transition from school to college

Abby visited the college that she was going to attend twice before she went there on a full-time basis. During the second week, Abby said that she no longer wanted to attend the college. Some evenings before college, Abby would appear quiet and withdrawn. On the following morning before college, Abby would complain of feeling unwell or tired. After this, Abby would often become angry and tearful because she did not want to attend college.

The college was able to identify that one activity Abby particularly did not enjoy was drama. Abby is relatively shy and drama may have caused her to become anxious because it involves high social demands, such as having to work with a group of people and perform in front of others. The college changed this activity to cookery, which Abby is much happier taking part in. This may be because cookery does not involve the same social expectations as drama and is consequently less stressful.

It is important to note that Abby did not say she felt “anxious”. This may be because she found it difficult to label this emotion correctly. Instead she said that she felt “tired” and “unwell” and her mood changed.

What can parents and caregivers do to manage this possible change in young adults with CdLS?

Given these changes there are a number of things that should be investigated and we would suggest:

1. Ensuring that all possible medical conditions are being appropriately treated and regularly reviewed. This also means being sure that all professionals and carers are aware of these conditions and the signs that something may be wrong.
2. If a change in situation or life event is going to happen and is unavoidable then the person should be prepared for the change as much as possible. This can be done by gradual introduction to the new situation using planned timetables of preferred, low demand and familiar activities in the new setting for short periods of time as the initial step. New activities and longer periods of activities can then be gradually introduced. See box 6 for an example of someone that is successfully integrating into the work place after leaving school using this method.



Box 6: Example of a successful transition from school to work

After visiting two mainstream colleges for typically developing young adults, Craig felt that he did not want to attend these colleges when he left school. Consequently, his father secured him a part-time job at his work place, a Steel works company. Craig was gradually integrated into the workplace over a long period of time. Initially Craig worked two hours per week at the Steel works and this has gradually increased to three days a week, working four hours on each day. Over time this will increase until Craig works there full time.

During his time at the Steel works company, Craig's responsibilities have also increased. Initially, Craig's job mainly involved sweeping the shop floor. Craig now has a wide range of responsibilities such as, shredding paper, processing timesheets of other employees and tidying up. Craig's work colleagues feel that he has increased in confidence since starting to work there. Although Craig was quite quiet when he started work, he has become much more sociable over time, greeting other members of staff when he sees them around the workplace and also enjoying their company at work. Craig often says that he really enjoys working there.

The key to this successful move for Craig was listening to Craig's concerns, gradual change and making sure there was predictability and routine.

3. If you are beyond the period of change then it is important to establish a highly structured routine within the new situation with preferred activities and reduced social demands. In the context of a school, college or day centre this means being sure that the professionals and carers working with the person are aware of these difficulties in order that they can help with implementing the routine. Again, new activities and social settings can be gradually introduced over time.
4. If the low mood persists then you should seek referral to Psychiatry or Clinical Psychology for an assessment and intervention.

Conclusion

It is clear that there are some behavioral challenges experienced by people with CdLS that have an impact on their quality of life and those involved in their care and support. It is also clear that although some behavior might be associated with CdLS, this does not mean that the behavior cannot be changed or that nothing can be done. We are seeing an increasing number of families who tell us about positive behavioral change when they were able to find



the cause of the problem. It is still surprising to us how frequently the cause is found to be a health condition and how frequently the health condition is found to be reflux. Even if you think this has been dealt with, it is always worth considering this as a possible cause.

We are grateful to the CdLS Foundation for sponsoring Jo Moss and Lisa Collis for their PhD work and we hope you think the results are worthwhile. We will do more to try and understand the behavioral challenges we see in CdLS and we will keep you updated on progress. In the meantime, remember: it doesn't have to be like this.

Suggested Reading:

Howlin, P. & Rutter, M. with Berger, M., Hemsley, R., Hersov, L. & Yule, W. (1989). Treatment of Autistic Children. Chichester: John Wiley & Sons.

Oliver, C., Moss, J., Petty, J., Arron, K., Sloneem, J. & Hall, S. (2003). Self-injurious behavior in Cornelia de Lange syndrome: A guide for parents and carers. Trident: London. Available free if you e-mail C.Oliver@Bham.ac.uk.

