
Anxiety in people with Cornelia de Lange syndrome

People with Cornelia de Lange syndrome (CdLS) may have a strong preference for sameness and routine. This means that when those routines are disrupted, for example with changes at home or in day services (e.g. going on holidays), this can be quite anxiety provoking. People with CdLS can also find demanding social situations very anxiety provoking.

The signs that someone is feeling anxious can vary, and may not be immediately obvious, especially in people who may not be able to easily tell you how they are feeling. However, these signs might include restlessness, pacing, forms of self-injury (mainly picking and scratching the arms and legs), irritability, loss of appetite or reporting feeling physically unwell.

Communication

When someone feels anxious, difficulties communicating this to others and making themselves understood may worsen the anxiety. People with CdLS vary widely in their ability to use speech to communicate their needs, and sometimes other communication aids might be useful. Visual aids can be a fantastic way to improve communication and the confidence to communicate. The National Autistic Society has lots of tips for helping people who have difficulties coping with certain situations and how to make the best use of visual aids; these strategies may also be relevant for people with CdLS.

Improving communication can also help clarify the causes of anxiety. It can be very easy to assume that an individual is anxious is about one thing, when in fact it is about another. Clearing up any misunderstandings can reduce or even remove the source of anxiety.

Avoidance

People (with and without intellectual disability) learn very quickly to avoid situations which make them feel anxious. It can be difficult for parents of people with CdLS to negotiate when to respect people's decisions to avoid anxiety-provoking situations, and when to persuade them otherwise. Whilst removing tasks that cause anxiety can be useful in the short term, this may maintain the anxiety or worsen this over time (because you never learn that you can cope with the anxiety, or that the situation/object is not really that frightening).

Sometimes it is possible to gently encourage a person to start with relatively easy situations or only experience these for a short period of time, before working up to situations which cause more anxiety. Periods of anxiety are generally self-limiting, and sometimes it is a question of helping a person to tolerate some anxiety at the beginning in order to learn that the anxiety passes and the situation becomes enjoyable.

Decision Making

People with CdLS have also been reported to have difficulties with making decisions. Recent research from the Cerebra Centre for Neurodevelopmental Disorders has shown that difficulties with some cognitive processes called “executive functions”, specifically attention switching and working memory, may partly explain these difficulties. For example, when faced with a number of choices you need to remember your options (working memory) and be able to move back and forth between them (attention switching) in order to make your decision. Our research showed that decisions with high demands on switching attention and working memory led to longer decision making times, and also provoked anxious responses from individuals with CdLS. Using picture cards or visual aids (reducing memory demand) and only giving two options at once (reducing attention switching demands) may help people make decisions and lessen the anxiety associated with this.

Other Strategies

Other strategies include using social stories and visual timetables to help to explain to people what activities they will be doing on a certain day. If anxiety is triggered by changes in routine or by fear of new and unfamiliar situations, then this may be helpful to help people prepare for this.

If there are unexpected changes to plans or routines, using a cue to indicate that change is coming can often be effective at reducing the anxiety associated with this. For example, by using a particular picture card. Sometimes families report that telling a person about a change a long time before it happens can heighten anxiety and lead to worrying until the event comes to pass. Therefore, cuing a change closer to the time (5-10 minutes before) may be a helpful middle position as opposed to ‘springing’ changes on the person or warning them far in advance.

Getting help

If the difficulties are severe your GP might suggest a referral to a psychiatrist or psychologist.

Resources

National Autistic Society has some good general advice for navigating difficult/anxiety provoking situations and tips for how best to use visual aids and social stories - <http://www.autism.org.uk/>

The Cerebra anxiety guide has some more information on recognising anxiety and suggestions for strategies to overcome this - <http://w3.cerebra.org.uk/help-and-information/guides-for-parents/cerebra-anxiety-guide-guide-parents/>