

Accessible summary: Behaviour Across the Lifespan in Cornelia de Lange syndrome

Groves L, Oliver C & Moss, J. (2021). Behaviour across the lifespan in Cornelia de Lange syndrome. *Current Opinion in Psychiatry*, 34, 112–117

This paper reviews recent research studies of Cornelia de Lange syndrome (CdLS). The review suggests that there are key changes with age in aspects of the profile of people with CdLS.

There appears to be a ‘critical period’ between the ages of 15-22 years where there is much change. This is followed by a period of relative stability. The traits of CdLS which appear to change over time are:

- *Autism characteristics.* A preference for routine or ‘insistence on sameness’ appears to stay the same or strengthen with age. Social withdrawal or avoidance of social situations becomes more problematic with age.
- *Anxiety.* This appears to stay the same or increase with age.
- *Negative affect (mood, interest and pleasure).* It appears that mood remains constant over time, but interest and pleasure decrease with age.
- *Cognitive abilities.* General abilities appear to stay the same, whilst receptive language (ability to understand other’s speech) improves and verbal memory (memory for information presented audibly) decreases with age.

Researchers are not sure why these changes occur. However, they suggest it is likely to be due to a combination of factors. These include:

- *Biological factors.* Genetic changes which cause CdLS may have an impact on neural health.
- *Cognitive factors.* A decline in verbal memory may affect the ability to keep up with social interactions. Some people may cope by withdrawing and avoiding new situations. This can cause social anxiety, social withdrawal and a loss of interest and pleasure.
- *Environmental factors.* Many life changes occur during late adolescence and early adulthood, such as finishing school and moving to adult health services. Changes can lead to uncertainty and loss of routines. Those with unrecognised needs such as co-occurring autism can find it especially hard. These transitions require more cognitive effort which can cause anxiety. Some may cope by withdrawing into routine which can lead to a loss of interest and pleasure.

This paper indicates that there are very specific changes with age in people with CdLS. Researchers think this is due to the unique profile of CdLS combined with environmental changes that are typical of the transition to adulthood. This can lead to reduced coping and put people with CdLS at an increased risk of poor mental health during this critical period.



It is important to remember that not every person with CdLS may experience these changes and that there is no evidence that these are inevitable. With better understanding of these changes, we can improve the support available to individuals with CdLS as they become older and through periods of transition and disrupted routine. Specific recommendations to support individuals include:

- Ensuring that families and professionals are aware of possible changes with age to aid early recognition and implementation of strategies to support individuals with CdLS as they grow older. More information on identifying and managing behaviour in CdLS can be found in the international consensus guidelines (Lay Summary of the international guidelines can be found here: [Diagnosis and management of Cornelia de Lange Syndrome - CdLS World](#)).
- Approaching periods of transition or changes in the environment, which could be possible triggers for the behaviours, slowly and with support.
- Anxiety is prevalent in people with CdLS so understanding this and accessing resources to support individuals is critical (see our Cerebra Anxiety guide: <https://www.findresources.co.uk/anxiety-pain-and-challenging-behaviour>).

We want to continue to better understand the key changes with age of people with CdLS and the causes of these changes. We hope this will enable better, targeted support and lead to improved mental health outcomes.

Accessible summary written by Dr Laura Wilby, Dr Laura Groves, Prof Chris Oliver & Dr Jo Moss.

If you would like to read the full article please click on the link here: [\(PDF\) Behaviour across the lifespan in Cornelia de Lange syndrome \(researchgate.net\)](#)