

The CdLS Be-Well Checklist

Behaviour and WellBeing in Cornelia de Lange Syndrome

This syndrome sensitive version of the Be-Well Checklist was compiled by The CdLS Foundation UK & Ireland Scientific and Clinical Advisory Team with additional input from Dr. Paul Mulder and Dr. Sylvia Huisman

(To be used alongside the Be-Well checklist. Download available at: www.cerebra.org.uk/download/be-well-checklist)

- For each item, CdLS related problems or influences are noted along with information on interactions with other checklist items, CdLS related signs and age. For each item the most common signs to look for can be found in the main Be-Well checklist. The CdLS Care Card should also be used alongside the checklist. The CdLS Care Card is available at: www.cdls.org.uk/media/files/cdls_carecard_2018.pdf

Checklist item	CdLS related problems or influences	Interaction with other checklist items, CdLS related signs and age information
Pain and discomfort. (See also the CdLS Care Card)	Gastro-oesophageal reflux and cough/ aspiration. Constipation. Volvulus (can be caused by malrotation). Tooth decay (can be caused by reflux). Middle ear infections (can be caused by reflux). Respiratory infections/sinusitis (can be caused by nasal polyps). Hip displacement. Blepharitis (not specifically painful but can be uncomfortable). Retinal detachment (increased risk by myopia)	Can directly cause self-injury. Can cause the behaviour to start in the first place and then later becomes learned. Can make learned behaviours more likely to occur, especially escape/ avoidance behaviours. Can affect sleep.
Sensory sensitivity	Over sensitivity (mainly hearing, vision and touch) and under sensitivity are reported.	Can affect sleep. Can cause phobias to develop. Can be a trigger for learned behaviours and anxiety. Can affect mood (getting bored/depressed by under stimulation). Can lead to avoidance of social situations.

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Anxiety or low mood	<p>Anxiety: can increase in late childhood through to early adulthood. Mainly seen in social settings (especially with strangers or groups), busy environments (e.g. supermarkets) and when upcoming events are new or unpredictable. Significant change (e.g. new class, change of established routines) can trigger anxiety.</p> <p>Low mood: Mood tends to be stable with age but interest and pleasure can decline in teenage and early adulthood years.</p>	<p>Anxiety: can affect sleep, and pain and discomfort, and cause skin picking and other forms of self-injury. Can be a response to sensory sensitivity, or unpredictable events or situations. Can be a trigger for learned behaviours. Can cause low mood.</p> <p>Low mood: can be caused by pain and discomfort, anxiety, and lack of choice and opportunity.</p>
Sleep difficulties	(As appropriate for age). Can be caused by generalised anxiety and anxiety around settling, and pain (especially from reflux when lying down, and aspiration).	Poor sleep can make impulsivity and learned behaviours more likely.
Emotional outbursts	(As appropriate for age). Emotional outbursts are uncommon but can be triggered by anxiety or pain.	Can be related to changes in routine or sensory over/under sensitivity as well as pain and anxiety.
Impulsive or insists on sameness	<p>Impulsivity can increase in teenage years and control of behaviour may be difficult at times.</p> <p>Insistence on sameness can increase from late childhood to early adulthood.</p>	<p>Impulsivity: can make anxiety, learned behaviours and emotional outbursts more likely. Anxiety can increase impulsivity.</p> <p>Insistence on sameness: might be a response to anxiety that is caused by unpredictability and can lead to avoiding events or places.</p>

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Social differences	Mixed picture. Social anxiety can increase from childhood into early adulthood and lead to greater social isolation and reluctance to speak to others (selective mutism), especially strangers, or when having to 'lead' the conversation. However, motivation for social contact and eye contact remain at the same level.	Can be related to insistence on sameness as a coping strategy particularly during periods of unpredictability or changes in routine.. So, only speaking to the same people when in the same place, or only speaking about the same thing.
Learned behaviour	Behaviour is triggered by something, such as being asked to do something, go somewhere, stop doing something, someone moving away from the person, being refused or unable to get something, going into a specific place. Behaviour tends to stop in the short term when the trigger is removed or stops.	Probably the most common cause of behaviours and is related to nearly all other checklist items. Pain, sensory sensitivity and anxiety can be triggers for behaviours such as self-injury, aggression, destroying the environment etc. with the behaviour rewarded by the removal of these triggers.
Communication	<p>Early hearing impairment can delay speech. Gestures used more than speech.</p> <p>As an absolute minimum, the person should be able to reliably and effectively communicate these nine things: that they want or need something to start, stop or pause, that they want or need to leave a situation or event, and that they want or need social contact, to go somewhere, food or drink, activities or help.</p>	Being able to communicate these things decreases the negative impact of most other checklist items on behaviour and wellbeing.

The Be-Well Checklist was developed by Oliver, C., Adams, D., Allen, D., Crawford, H., Heald, M., Moss, J., Pearson, E., Richards, C., Waite, J., Welham, A., Wilde, L., and Woodcock, K. The Universities of Aston, Birmingham, Leicester, Surrey, Warwick and Wolverhampton, UK and Griffiths University, Australia.