



Expert Perspective Interview Behavior and Medication

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1. Can you provide an overview of CdLS and how it affects behavior?

Children and adults with CdLS will manifest a higher risk for behavioral and emotional disturbances. The risk for individuals with intellectual disability is at least 50% for a diagnosable psychiatric condition, while in the general population, it is about 10% for a treatable condition. The risk for individuals with CdLS is at least 50% or higher.

2. What are some common behavioral challenges associated with CdLS?

A common behavioral challenge is the manifestation of excessive anxiety. It can take the form of selective mutism, social avoidance, and dysregulation with fear triggers. Management of the environment is important in order to decrease anxiety-related behaviors.

3. How does CdLS impact emotional regulation and social interactions?

For those individuals with CdLS and emotional dysregulation and social difficulties, it is mostly understood as an anxiety/hyperarousal-driven condition rather than a truly autistic manifestation. This is because autism is characterized by a lack of social awareness and no social interest, which is not the case in most individuals with CdLS.

4. What are the most frequent behavioral issues in individuals with CdLS?

Repetitive behaviors, the need for routine, and disruption if an individual is unable to engage in these repetitive behaviors are frequent concerns in individuals with CdLS. A more extreme form of repetitive behaviors that can occur is self-injurious behaviors (SIB), which can cause physical harm to specific body areas.

5. Are there specific triggers that tend to cause behavioral challenges in individuals with CdLS?

As in most individuals with ID, common triggers for CdLS are overstimulating environments, sensory overstimulation, and lack of attention. Controlling the environment can help manage these behaviors.

6. How do these behaviors compare to individuals with other genetic conditions?

The profile of individuals with CdLS is characterized by high levels of repetitive behaviors and extreme anxiety. Some individuals with CdLS show excessive hyperactivity as well, and other SIB is of concern. Like most other genetic disorders, repetitive behaviors are common, and social reciprocity (the ability to connect with others) is sometimes deficient but relatively less affected compared to 'classic autism'.





7. What early signs indicate a potential behavior issue in someone with CdLS?

There are no particular early signs, as these behaviors can occur at any age or in the early years. In others, the behaviors are not apparent till later in life.

8. How can parents/caregivers differentiate between typical behaviors and those requiring intervention?

An impairment in parents' quality of life is a strong indicator that intervention is warranted. This threshold may be different among families, but seeking early assistance is encouraged, as treating maladaptive behaviors early improves prognosis.

9. Are there co-occurring conditions (such as anxiety, sensory issues, or self-injury) that parents should watch for?

The common symptoms noted above may be considered co-occurring conditions (anxiety, etc.), but may also be regarded as an integral part of CdLS' behavioral phenotype. For example, repetitive behaviors and SIB are hallmark behavioral manifestations in CdLS.

10. What strategies can parents/caregivers use to manage behavior issues at home?

If problem behaviors arise, consultation with an applied behavioral analyst is encouraged. The indicated approach is usually the ABA model (Applied Behavioral Analysis), and the Functional Analysis (FA) of behaviors can guide treatment. The behaviorist may also guide parents in managing behaviors. Generally, positive reinforcement of good behaviors is the most effective strategy.

11. When is a medication typically considered for behavioral challenges in individuals with CdLS?

Medication consultation may be warranted when problem behaviors are identified, and a substantial trial of ABA therapy is unsuccessful. There may be acute cases of SIB or disorganized behaviors (psychosis) where medication treatment may be started earlier.

12. What medications are commonly prescribed for behavioral issues in CdLS, and what symptoms do they target?

There are four categories of medications: stimulants/alpha agonists for hyperactivity/hyperarousal; SSRIs for anxiety/depression; mood-stabilizers for bipolar disorder or mood swings; neuroleptics for SIB, aggression.

13. How do you determine the proper medication and dosage for someone with CdLS?

Consultation with a psychiatrist with experience in developmental disability and ID is a necessary step to obtain the correct medication and dosing.





14. Are there any medications that should be avoided or used cautiously in individuals with CdLS?

In general, individuals with CdLS will have more sensitivity to medication side effects and may even experience paradoxical effects with psychiatric medications. *Consultation with an expert and a cautious approach are warranted when psychotropics are prescribed.*

15. How do medications for behavior interact with other common medical treatments that individuals with CdLS may receive?

There is no special interaction with other medications applied for medical conditions in CdLS, but a check for medication interactions is necessary in those cases where multiple medications are used.

16. What signs indicate that medication works effectively for behavior management?

To monitor medication effects, a rating scale such as the Aberrant Behavior Checklist (ABC-C) is encouraged. Qualitative reports from caregivers are usually the main form of feedback on medication effects.

17. How long does it typically take to see results from a new medication?

It depends on the medication, wherein for stimulants and ADHD it can be 1-2 days, and for SSRIs and repetitive behaviors it can be 4-6 weeks.

18. What are the most common side effects of behavior-related medications in CdLS individuals?

There is no particular pattern of side effects of medications. It is individual to each medication and its profile.

19. How can caregivers and healthcare providers monitor and assess the impact of medication over time?

The follow-up of medication use should be with visits every 3-6 months for an increase or decrease in dosing depending on symptoms.

20. When should caregivers consider adjusting or discontinuing medication?

If the medication is not as effective as initially experienced or during the child's growth, it might need to be re-evaluated.

21. Should medication always be paired with behavioral therapy or other interventions?

The first approach to the management of maladaptive behaviors is the application of a behavioral management plan. When it is not effective or sufficiently therapeutic, medication may be considered.





22. Can non-medication approaches be tried before or alongside medication?

Non-medication approaches should be tried before medication intervention, except in select cases.

23. How do sleep, diet, and sensory factors influence the need for medication in individuals with CdLS?

For particular patients, diet, exercise, and sensory factors may play a role in maladaptive behaviors. A thorough assessment of these areas of function is warranted when developing a behavioral plan.

24. What role does anxiety play in behavioral challenges, and can it be managed without medication?

Anxiety may respond to behavioral interventions, before medication is tried, or used along with medication.

25. How can schools and care providers best support individuals with CdLS?

IEP or individualized educational plans are necessary for youth with CdLS attending school.

26. How do CdLS behavior challenges vary across individuals and ages?

A key time in the development of behavioral challenges in individuals with CdLS is adolescence. In several instances, adolescence is accompanied by mood swings and disruptive behaviors.

27. Can caregivers and professionals use any universal strategies to improve quality of life?

A behavioral plan with positive reinforcements (rewards) for good behaviors is a universal intervention that is suggested.

28. How can the broader community (schools, healthcare professionals, employers) become more aware and supportive of individuals with CdLS?

The CdLS Foundation's advocacy can help families and individuals with CdLS receive more support from schools and communities.

