

Evaluating Challenging Behaviors in Adults with Down Syndrome

Down Syndrome Medical Interest Group: DS-ASD Workgroup

Challenging behavior escalations (e.g., aggression towards self or others) in an adult with Down syndrome may indicate the need to:

1. Explore etiology of the behavior and any recent environmental changes or perceived stressors.
2. Rule out a medical cause.
3. Evaluate mental health.
4. Consider an autism or regression assessment as indicated.

Behavioral Assessment

Important considerations:

- In order to help reduce the risk of exacerbating anxiety and escalating behaviors, remember the need for a gentle tone and longer processing time when clinically assessing individuals with DS.
- Sensorimotor behavior (stimming) can be a stress reliever in individuals exhibiting challenging behaviors, necessitating tolerance of this behavior in the office.
- Be sensitive of your tone when working with the caregiver.

Explore etiology of challenging behaviors:

- Many behaviors do not have a medical or psychological cause; rather they are situational.

Review any recent environmental changes or perceived stressors:

- Consider abuse/neglect.
- Individuals with DS tend to remember events like a movie: the emotions, words, actions that were stressful may continue to replay in their minds as a relived experience.
- Timing of behavioral escalations may help identify the cause.



Think about the ABC's (antecedent – behavior – consequence/result):

- Seek to gain information on common antecedents that may be triggering the challenging behaviors including:
 - Sensory disturbances
 - Fear/anxiety/stress/frustration
 - Unmet physical needs (e.g., hunger)
 - Pain
 - Removal of preferred objects, people, or pets
 - Extreme frustration due to inability to communicate needs & being misunderstood

- Seek to understand functions or “goals” of the challenging behavior including:
 - Obtaining desired objects or privileges
 - Attention or reactions from others
 - Escape or avoidance from undesired stimuli or tasks
 - Self-stimulatory actions

Sometimes this exercise can clarify situational issues requiring attention and provide helpful guidance for implementing solutions. (See discussion of Addressing Challenging Behaviors below.)

A more extensive medical, behavioral and mental health assessment may be indicated if straightforward solutions are not apparent, or if additional conditions are suspected, and are discussed below.

New onset, subacute (over weeks to months) decline in functional skills previously attained, associated with challenging behaviors and a decline in communication/social skills can be a manifestation of Down Syndrome Regression Disorder. References containing recommendations for evaluation and work up:

- [Assessment and Diagnosis of Down Syndrome Regression Disorder: International Expert Consensus - PMC \(nih.gov\)](#) ¹
- [Down Syndrome Disintegrative Disorder: A Clinical Regression Syndrome of Increasing Importance | Pediatrics | American Academy of Pediatrics \(aap.org\)](#) ²
- https://link.springer.com/chapter/10.1007/978-3-319-90083-4_7 ³

¹ Santoro et al., “Assessment and Diagnosis of Down Syndrome Regression Disorder: International Expert Consensus.”

² Rosso et al., “Down Syndrome Disintegrative Disorder: A Clinical Regression Syndrome of Increasing Importance.”

³ Chicoine and Capone, “Regression in Adolescents and Adults with Down Syndrome.”



The Medical Work-Up

The following medical conditions can be responsible for changes in behavior.

Consider:

- Vision impairment
 - ophthalmology evaluation
 - consider functional vision evaluation
- Hearing impairment
 - audiology evaluation
 - may require sedated ABR
- Thyroid disorder
 - exam, labs
- Sleep disorder – chronic sleep apnea, and/or sleep stage disturbances
 - request a sleep study (may be able to substitute home sleep monitor; acquire an audio-visual recording of sleep for review)
- Seizure disorders (Consider neuro consult)
 - ask about history of Infantile Spasms*
 - check EEG if tonic clonic movements or change in consciousness
 - Note: tremors, tics, and involuntary movements, are unlikely to be seizure.
- Cardiac issues: (Consider cardiology consult, cardiac work up)
 - Congenital heart disease* - consider acquired valvular problems
 - Other cardiac issues – tendency for hypotension, bradycardia with syncope
- GI issues: (GI consult may be indicated)
 - Celiac disease – consider evaluation
 - Anatomical gastrointestinal (GI) tract anomalies/malformations
 - Constipation
 - consider x-ray to rule out fecal impaction. Consider bowel cleanout. Constipation can be painful and impact willingness to eat, or present as new behaviors.
 - GE Reflux
 - Inflammatory esophagitis can result in intermittent pain, vomiting, and not wanting to eat. Consider an empiric trial of an antacid and/or motility agent to see if apparent pain improves.
 - New onset or worsening swallowing difficulties & dysphagia
 - may result in food selectivity, refusal, or restricted diet. If there is choking, gagging, watery eyes, difficulty swallowing or history of aspiration, check a modified barium swallow (or clinical feeding evaluation if the individual cannot tolerate study). Refer to OT/SLP familiar with feeding problems if needed.
- Breathing conditions or illnesses (i.e., pneumonia)
- Painful skin conditions, such as folliculitis, hidradenitis, ingrown toenails
- Dental problems including infections/abscesses
 - dental x-ray may be helpful if exam is difficult to obtain
- Musculoskeletal concerns, for example, flat feet, early arthritis, joint hypermobility, ligamentous hyperlaxity
- Headache, migraine, head pain



- **Note: These are among medical conditions that are more common in comorbid DS and autism. For complete list see reference.⁴*

Laboratory evaluations that may warrant consideration include:

- TSH/FT4, thyroid antibodies
- CBC
- B12, Folate
- Ferritin/Iron studies
- Vitamin D
- Lead
- Celiac titers (even when no GI symptoms)

Helpful resources for medical work up:

- American Academy of Pediatrics guidance for care of children, adolescents with Down syndrome, April 5, 2023⁵
 - Aimed at pediatric age groups up to early twenties.
- Medical Care Guidelines for Adults with Down Syndrome (2020)⁶

Mental Health Assessment

Mental Health Conditions that may develop in adults with Down syndrome:

- Adjustment or grief reaction:
 - Difficulty coping with significant changes including loss of a loved one related to death or a parent/caregiver/sibling/ roommate leaving home, or relocation/move or change in living situation
 - Change in educational or work setting/ day-to-day life
 - Discuss potential relational stress, such as at work or day program
- Depression
- Mood disorder with psychotic features, disorganization
- Post Traumatic Stress Disorder (PTSD)
- Anxiety

⁴ Spinazzi et al., "Co-Occurring Conditions in Children with Down Syndrome and Autism: A Retrospective Study."
<https://pubmed.ncbi.nlm.nih.gov/36864370/>

⁵ Black, "Clinical Report Updates Recommendations for Care of Children With Down Syndrome: American Academy of Pediatrics."
<https://www.aap.org/en/news-room/news-releases/aap/2022/clinical-report-updates-recommendations-for-care-of-children-with-down-syndrome-american-academy-of-pediatrics/>

⁶ for Adults with Down Syndrome Workgroup, "Global Down Syndrome Foundation Medical Care Guidelines for Adults With Down Syndrome."
<https://www.globaldownsyndrome.org/wp-content/uploads/2020/10/Global-Down-Syndrome-Foundation-Medical-Care-Guidelines-for-Adults-with-Down-Syndrome-v.1-10-20-2020.pdf>



- Autism (previously undiagnosed)
 - Note:
 - Autism is not a mental illness. However, it may contribute to significant behavioral challenges, and adults with DS-ASD may be more vulnerable to additional mental health conditions, particularly with life changes and transitions that occur with growing older. See discussion below.
- Severe decline in functional skills: in the absence of prior autistic symptomatology, consider Adult DS Regression, which can have psychiatric manifestations – references noted above.

Helpful resource for mental health work up:

- Mental Wellness in Adults with Down Syndrome, 2nd Edition, d. McGuire, Ph.D., B. Chicoine, M.D., 2006, 2021,
https://adscresources.advocatehealth.com/assets/1/6/Mental_Wellness_full_book_final_2023.02.24.pdf?mibextid=Zxz2cZ⁷

Why consider Autism in an Adult with Down syndrome?

- Autism is more prevalent in DS than in the general population.⁸
- ASD may be the basis for an escalation of challenging behaviors in an adult with DS, possibly triggered by perceived stressors, environmental changes, or medical conditions.
- These behaviors may include:
 - Frequent or accelerated tantrums
 - “Drop and flops” (e.g., cries or falls to the ground and refuses to move)
 - Self-injurious behaviors (e.g., banging head against the wall)
 - Extreme difficulty with even minor changes in routine
 - Increased social isolation
 - Engagement in self-stimulatory behaviors
 - Aggression
- People with ASD may respond to a variety of behavioral interventions, and co-morbid mental health issues related to autism may respond to pharmacological interventions.
- A formal diagnosis of autism:
 - Can help an individual qualify for additional services such as behavioral support in the home, work, or day program setting
 - May help a family/caregiver more fully understand the individual
- Understanding a full diagnostic picture will allow caregivers to develop an environment that will promote independence and skill acquisition by that individual even in adulthood.

⁷ Dennis McGuire, Ph.D. & Brian Chicoine, M.D, “Mental Wellness in Adults with Down Syndrome A Guide to Emotional and Behavioral Strengths and Challenges.”

⁸ Richards et al., “Prevalence of Autism Spectrum Disorder Phenomenology in Genetic Disorders: A Systematic Review and Meta-Analysis.” <https://pubmed.ncbi.nlm.nih.gov/26341300/>



Autism in Down Syndrome: What does it look like?

- Autism spectrum disorder (ASD) is a lifelong, neurodevelopmental condition with symptoms presenting in early childhood. In an adult with DS, symptoms of autism should be evident during early childhood, even if they were not previously identified with a diagnosis of DS-ASD.
- It is important to understand DS developmental & behavioral norms:
 - See discussion of behavioral norms in Mental Wellness in Adults with DS book (above).
- Every individual with autism experiences a spectrum of symptoms across multiple domains:
 - Differences with social communication & social interaction/social initiation
 - Restricted interests or repetitive behaviors
 - Sensory differences
 - Behavioral challenges
- Autism may be accompanied by certain medical issues⁹ as well as mental health challenges with anxiety, depression, and attention.
- Compared with individuals with autism alone, individuals with DS-ASD may:
 - Show more social interest in peers
 - Have more significant cognitive challenges
 - Have slower processing speeds that may be misinterpreted as stubbornness
 - Display less severe repetitive behaviors
- See chart below for examples.
 (+/- indicates inconsistent, ↓ indicates less than expected, ↑ indicates more than expected)

Domain	DS	DS-ASD	ASD
Social Communication			
Eye contact*	+/-improvement by 2 years	↓	↓
Social smile*	↑	+/-	↓
Joint attention*	+/- but improves	↓	↓
Gestures	↑	↓	↓
Behaviors			
Insistence on sameness	+/-	↑	↑↑
Repetitive behaviors	+/-	↑	↑↑

*Consider evaluating vision and/or hearing impairments if these domains are affected.

⁹ Spinazzi et al., “Co-Occurring Conditions in Children with Down Syndrome and Autism: A Retrospective Study.”

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Helpful information to gather if suspecting DS-ASD:

- Behavior: inquire specifically about the areas of social communication/interaction, restricted interests, repetitive behaviors, and sensory sensitivity. If there are multiple domains impacted concurrently, this would point you towards seeking a formal autism evaluation.
- Potential screeners for ASD include: SCQ – Social Communication Questionnaire and SRS-2 – Social Responsiveness Scale.
- Videos from the individual's caregiver in different settings, particularly in how they respond to opportunities to engage with others, if possible, for the purpose of sending to the evaluator.
- School records, therapy records, medical records.

Where to refer individual for further autism evaluation:

- Local autism center of excellence
- Association of University Centers on Disabilities, www.aucd.org
- U.S. Down syndrome medical clinics listing, <https://www.globaldownsyndrome.org/research-medical-care/medical-care-providers/>
- Hospital (outpatient clinical psychology evaluation)

Addressing Challenging Behaviors

Immediate Considerations

- Simplify and monitor the environment - mitigate overstimulating settings, ensuring predictable routines are followed.
- It may be necessary to reduce expectations on the individual in order to help get their behavior more regulated.
- Address sleep disturbance which may exacerbate daytime behavior problems.
- Consider medications if appropriate:
 - If a mental health condition is present, then medication treatment may be beneficial.
 - It is not necessary to pursue a formal evaluation for autism in order to identify and treat existing medical conditions or to initiate medications for behavioral and mental health symptoms if indicated.
 - Thoughtful use of psychotropic medications can help in both short term (acute) situations and longer term (maintenance) situations.
 - In the above setting it is important to give the medication time to work and not push behavioral therapy or lifestyle changes until the individual has stabilized and is simply feeling better.
 - It is important to regularly review medications, to avoid confusing polypharmacy, and to adjust doses to the lowest effective level. It may be worth considering whether to wean and discontinue some medications once symptom stability has been achieved for an extended period of time (>9-12mo).
 - For pharmacotherapy treatment guidelines for adults with Down Syndrome, see Pharmacotherapy of Down syndrome. *Expert opinion on pharmacotherapy* <https://doi.org/10.1080/14656566.2018.1529167>¹⁰

¹⁰ Palumbo and McDougle, "Pharmacotherapy of Down Syndrome."



- Try to obtain in-home behavioral or personal support from a qualified adult professional.

Ongoing Behavioral Supports

- Interventions to address the above functions of behavior include:
 - Teaching appropriate means to obtain desired outcomes
 - Reinforce/pay attention to other, non-challenging behaviors
 - Make tasks/requirements smaller or more manageable
 - Attempt to replace, not stop, self-stimulatory behaviors
- Design interventions (such as 1:1 support for life skills and socialization, waiver to cover behavioral therapy).
- Ensuring individualized appropriate behavioral plans/therapy and supports are in place are essential to promote a healthy life for each individual. Research shows there is a benefit to behavioral therapy, to help build structure, routines, lower anxiety, and increase predictability of “what’s next”.
- If desired, a referral for behavioral interventions can be initiated without a formal diagnosis of ASD, possibly through waiver resources (consider referral while waiting for ASD assessment).
- Consider sensory assessments and interventions - usually provided by an occupational therapist.
- Consider communication assessments and interventions like augmentative and alternative communication - usually provided by a speech pathologist.
- Prepare documentation that the family can share with the employer, job coach, therapists, and Medicaid or other insurance provider in the case of a new diagnosis (importantly, an additional dx of ASD can establish eligibility for additional school and community-based services). Helpful resources:
 - The Arc, <https://thearc.org/>
 - Developmental Disability Administration (DDA), <https://www.dshs.wa.gov/dda>

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