



Behavior in Angelman Syndrome

A Clinical Framework for Assessment and Management

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An Angelman Syndrome Foundation Clinical Guidance Document Based on Published Literature, Clinical Experience, and Expert Consensus

Overview

Behavioral challenges in Angelman syndrome (AS) are often multifactorial and can arise from medical, communicative, sensory, and environmental factors. Typical behavioral challenges arise from frustration related to inability to communicate. However, behavioral changes may also be the first sign of pain, illness, sleep disruption, anxiety, environmental stressors, or unmet needs. Before focusing on how to stop a behavior, it is important to understand what the behavior may be communicating and what factors may be contributing to it. Effective management requires systematic identification of root causes, including underlying medical, developmental, and environmental contributors, and sudden or worsening behavioral symptoms should generally be presumed to have an underlying cause.

This guide aims to provide a practical framework for evaluating and managing behavioral challenges in AS.



1 Recognize Syndrome-Specific Behavioral Phenotypes

Some behaviors may represent manifestations of the AS neurobehavioral phenotype rather than a separate psychological/emotional disorder. In AS attention seeking behaviors are common and these may become extremely challenging and even unsafe.

Characteristic AS behavior also include:

- Frequent laughter and excitability
- Hyperactivity
- Short attention span
- Sensory-seeking behaviors
- Mouthing behaviors
- Repetitive or ritualized behaviors
- Strong social interest and attention seeking behaviors

Appreciating the baseline phenotype helps to distinguish expected characteristics from new concerns.



2 Rule out medical causes first

When dealing with recent changes in behavior, an evaluation for a medical issue as an underlying cause of the behavior change should be performed.

When evaluating a new or worsening behavior, first ask: *"Why now?"*

Particular attention should be paid to:

- Gastroesophageal reflux disease
- Constipation
- Dental pain
- Otitis media (ear infection) or sinus disease
- Seasonal allergies
- Urinary tract infections
- Orthopedic pain (such as back pain related to scoliosis)
- Medication side effects
- Nutritional deficiencies, including iron deficiency, or weight loss
- Menstrual discomfort, hormonal changes (e.g. cyclical, 3-5 days prior to period/menses)

Neurologic Contributors

- Breakthrough seizures
- Postictal behavioral changes
- Non-epileptic myoclonus

Behavioral regression or changes in awareness should prompt consideration of EEG and neurological evaluation.

Review of nutritional status and physiological stress

Poor weight gain, feeding difficulties, or catabolic states (breaking down or losing overall mass) can significantly affect mood and behavior. Physiological stress may present primarily as behavioral dysregulation.

Questions to consider:

- Has there been recent weight loss?
- Is caloric intake adequate?
- Are feeding times stressful?
- Is the patient experiencing recurrent vomiting or feeding intolerance?

Pubertal and hormonal influences

- Menstrual discomfort
- Premenstrual mood changes
- Hormonal fluctuations

Behavioral manifestations of pain may include:

- Irritability
- Aggression
- Self-injury
- Sleep disruption
- Increased stereotypies
- School refusal
- Regression of skills
- Worsening or new onset of myoclonus

3 Assess sleep

Sleep disturbances are highly prevalent in AS. Sleep problems are associated with daytime behavioral dysregulation and can contribute to challenging behaviors.

Sleep issues include:

- Difficulty falling asleep
- Frequent awakenings
- Early morning awakening
- Sleep-disordered breathing
- Restless sleep
- Medication effects
- Nighttime seizures
- Enuresis (bed-wetting)

Sleep hygiene and behavioral approaches are essential and greatly benefit sleep patterns. Even with these approaches, still some individuals chronically struggle with sleep issues. Medication should be considered in more severe cases. Improvement in sleep alone may improve behavior.





4 Ask “What is this behavior communicating?”

Once medical contributors have been addressed or ruled out, the next step is to try to evaluate the function of the behavior; in other words, what this behavior is trying to communicate or accomplish?

Given the limited expressive communication abilities in AS, behavioral changes may represent an attempt to communicate anxiety, frustration, sensory overload, or unmet needs.

That is why the ABC (Antecedent-Behavior-Consequence) analysis is recommended as an essential step in the evaluation of behaviors.

Transitions, disruption of routine, separation from the caregiver, and preferred item removal are the most common precipitants of behavioral escalation in AS.

Inadvertent reinforcement (attention, escape from demands) can play a role in maintaining the challenging behavior, and consistent caregiver responses are essential.

Every challenging behavior should prompt clinicians to ask whether the individual has a reliable and efficient way to communicate the need being expressed by the behavior.

Analyze Behavioral Function

Common functions include:

- Acquire or maintain attention
- Access to preferred items or activities
- Sensory or self-stimulatory reinforcement
- Escape from demands
- Over-excitement, over-stimulation

When challenging behaviors occur in school, parents can request that the school conduct a Functional Behavioral Assessment to better understand the purpose of the behavior and guide development of an individualized behavior support plan. Behavioral data and functional assessment are invaluable in developing a consistent plan for parents, teachers and care staff, and help clinicians determine whether the plan needs to be adjusted.

Questions to consider:

- Is the behavior new or longstanding?
- Has there been a recent change in the routine, caregivers, school placement, or environment?
- Is the behavior occurring in specific settings or across multiple situations?
- What happens immediately before and after the behavior?

The ABC Model in Angelman Syndrome

Understanding behavior as communication

EXAMPLE 1: Individual seeking attention by grabbing and pulling a peer closer

A ANTECEDENT
What happens BEFORE the behavior?

The individual wants attention and social interaction with a peer. They grab the peer's arm and pull them closer. The peer does not want physical contact and moves away or says "no."

No, I don't want that.

I want attention and to play!

COMMON TRIGGERS

- Desire for attention or interaction
- Peer not engaging
- Change in activity or transition
- Loneliness or wanting to play

B BEHAVIOR
What does the individual DO?

The individual becomes upset and escalates to aggression toward the peer.



EXAMPLES OF BEHAVIOR

- Hitting, pinching, scratching
- Pushing, biting, or pulling hair
- Chasing or blocking the peer
- Loud screaming or yelling

C CONSEQUENCE
What happens AFTER the behavior?

The peer and adults give the individual attention and respond.

Let's take a break. I'm here.

OUTCOME

- The individual gains attention and interaction
- The peer stays or continues engaging
- The behavior may happen again

CLINICAL INTERPRETATION

Possible Function:
Access to attention and social interaction.

WHAT THE BEHAVIOR MAY BE COMMUNICATING

- "I want to play with you."
- "Don't leave."
- "Pay attention to me."
- "I don't know another way to keep this interaction going."

PREVENTION / REPLACEMENT SKILLS

- Teach and practice ways to gain attention (AAC, gestures, words)
- Reinforce appropriate requests
- Provide scheduled opportunities for social interaction
- Teach peers and staff how to respond consistently
- Use visual supports for expectations

EXAMPLE 2: Individual spitting or screaming while caregiver is engaged in conversation with others

A ANTECEDENT
What happens BEFORE the behavior?

The caregiver is talking with others (e.g., family members, staff). The individual is nearby and seeks attention.

I want attention! I'm not included.

COMMON TRIGGERS

- Caregiver focused on others
- Feeling left out or ignored
- Desire for connection
- Waiting or not being engaged

B BEHAVIOR
What does the individual DO?

The individual spits, screams, or both to get the caregiver's attention.



EXAMPLES OF BEHAVIOR

- Spitting
- Screaming or yelling
- Loud vocalizations
- Throwing items or dropping to the floor

C CONSEQUENCE
What happens AFTER the behavior?

The caregiver stops talking with others and gives the individual attention and responds.

I see you. I'm here.

OUTCOME

- The individual gains attention and engagement
- The interaction shifts to focus on them
- The behavior may happen again in the future

CLINICAL INTERPRETATION

Possible Function:
Access to attention and connection.

WHAT THE BEHAVIOR MAY BE COMMUNICATING

- "Look at me."
- "I want to be part of what is happening."
- "I need interaction."
- "Don't forget about me."

PREVENTION / REPLACEMENT SKILLS

- Provide frequent, planned attention
- Use visual schedules or timers for turn-taking
- Teach ways to request attention (AAC, gestures, tapping)
- Acknowledge calm, appropriate bids for attention
- Create inclusive opportunities for engagement

★ **REMEMBER:** Behavior is communication. When we understand the why, we can respond with empathy, reduce distress, and build connection. Individuals with Angelman syndrome thrive with understanding, structure, and supportive relationships.

The ABC Model

Understanding behavior in an individual with Angelman Syndrome



In Angelman syndrome, behavior is communication. This framework helps us understand what might be happening, what the behavior means, and how we can respond to support the individual's needs.



Remember:
Look for causes first. Behavior is rarely "just" behavior.

A ANTECEDENT
What happens BEFORE the behavior?

The trigger, event, or situation that happens right before the behavior.

EXAMPLE FOR ANGELMAN SYNDROME

It is time to transition from a preferred activity (music/video) to something non-preferred (therapy or getting dressed).

They are enjoying the music and don't want it to stop.

WHAT TO LOOK FOR

- Transition or routine change
- Removal of a preferred item/activity
- End of attention or interaction
- New demand or non-preferred task
- Sensory overload or environmental change

B BEHAVIOR
What does the individual DO?

The observable action or response.

EXAMPLE FOR ANGELMAN SYNDROME

The individual throws themselves to the floor, laughs loudly, flaps arms, and may cry or become more agitated.



WHAT TO LOOK FOR

- Laughter, squealing
- Body dropping or falling
- Arm flapping, spinning, pacing
- Crying, agitation, vocalizations
- Self-injury or aggression (if severe)

C CONSEQUENCE
What happens AFTER the behavior?

The response or outcome that follows the behavior.

EXAMPLE FOR ANGELMAN SYNDROME

The adult gives extra attention, lets the individual continue the activity, or removes the demand (e.g., skips therapy).

The behavior may have gotten what they wanted: attention or a break.

WHAT TO LOOK FOR

- Attention (talking, cuddling, eye contact)
- Escape from a demand or task
- Access to a preferred item/activity
- Delay or avoidance continues
- Sensory input (movement, noise, etc.)



WHY THIS MATTERS

In Angelman syndrome, challenging behavior often means the individual is trying to communicate a need, feeling, or wanting something. Understanding the ABCs helps us respond in a way that teaches, supports, and reduces future distress.



USE THIS TO SUPPORT

- ✓ Identify the reason behind the behavior
- ✓ Teach alternative ways to communicate
- ✓ Change the environment or routine
- ✓ Reinforce positive communication

GOALS:

- ✓ Reduce frustration
- ✓ Increase communication
- ✓ Build connection
- ✓ Improve quality of life

♥ Every behavior is communication. With understanding and support, we can help individuals with Angelman syndrome feel safe, connected, and successful. ★

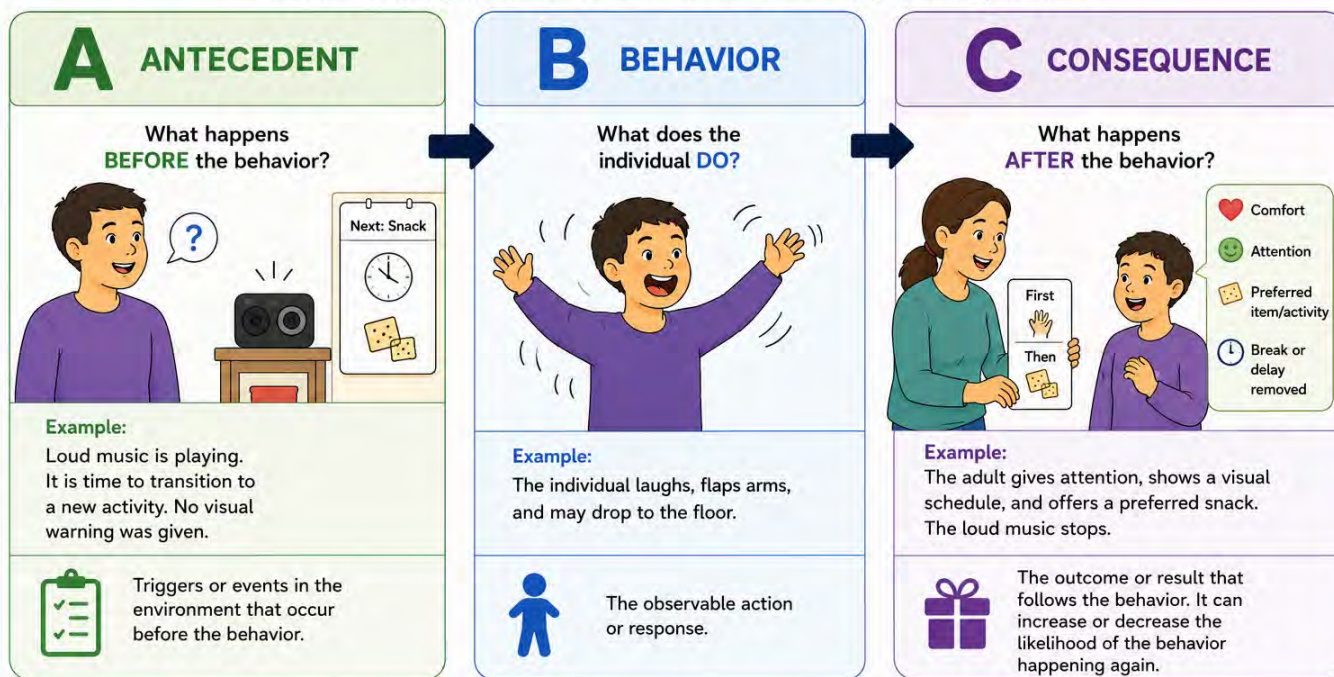




The ABC Model



Understanding behavior for an individual with Angelman Syndrome



Why this matters for Angelman Syndrome

Understanding the ABCs helps us see what the individual is trying to communicate. It helps us change the environment and teach skills in a supportive way.



Focus on strengths.
Build connection.
Celebrate progress.

These images were generated by AI after instructions based on real cases clinical experience. The images presented were generated using artificial intelligence based on prompts developed from real clinical cases and the authors' clinical experience. They are intended for educational purposes only and do not depict actual patients.

5 Evaluate for Anxiety

Anxiety is increasingly recognized as a major contributor to behavioral challenges in AS. Evaluating anxiety in AS requires a different approach than in verbally fluent individuals because anxiety manifests with behavioral, physiologic, and functional changes rather than verbal reports of worry.

Look for Physiologic Signs

- Avoidance behaviors
- Increased stereotypies or myoclonus
- Sweating, flushing, tachycardia, hyperventilation
- Separation difficulties
- Gagging, vomiting or retching
- Refusal to eat
- Distress in specific environments
- Fearful affect

Triggers may include:

- Changes in a long-term routine
- Novel environments
- Separation/fear of separation from caregivers
- Medical procedures
- Social demands
- Sensory triggers (e.g. loud noise, crowded environment)
- Performance demands

Many families report:

"The behavior started when we entered the building." or "She starts gagging before therapy."

These can be important clues to understanding the purpose of the behavior.

Obtain Information Across Settings

- Does it occur at home?
- Does it occur at school?
- Does it occur with specific caregivers?

Patterns often reveal anxiety triggers.

When evaluating anxiety, ask:

"Is the individual with AS trying to avoid something because they don't want to do it, or because it makes them anxious?"

6 Use a Multidisciplinary Approach

Complex behavioral concerns often require collaboration among:

- Primary care providers and specialists depending on the clinical presentation
- Psychologists or behavior specialists
- Therapists and school teams
- Encourage communication between specialists who may be helping with behavior (AAC specialist, special education teacher, behavior specialist)

Communication as Behavioral Intervention

- Functional Communication Training (FCT); teaching a communicative replacement for a challenging behavior has the strongest evidence base for behavior reduction in AS
- Improvement of communication strategies reduces frustration-driven aggression; consistent use across home, school, and clinic settings augments the likelihood of treatment success



6 Use a Multidisciplinary Approach



Medication selection should target a clearly defined symptom cluster (anxiety, hyperarousal, aggression, sleep disturbance, compulsive behaviors) rather than a diagnosis of "behavior problems."

Examples include:

- Anxiety: SSRIs, buspirone, selected benzodiazepines
- Hyperarousal or impulsivity: guanfacine or clonidine
- Sleep disturbances: melatonin, clonidine, trazodone, Benadryl, mirtazapine, other sleep-directed interventions
- Severe irritability or aggression: carefully selected atypical antipsychotics

Medications should be used in conjunction with medical evaluation and behavioral interventions.

Medication Management

Prescribing principles include:

- Start low, go slow. Individuals with AS are often sensitive to side effects, particularly constipation, sedation and behavioral activation
- The evidence base for psychopharmacology specifically in AS is limited; most prescribing is extrapolated from the broader IDD and ASD literature
- Antiepileptic drugs (AEDs), particularly valproate, lamotrigine, and clobazam, can have both positive and negative behavioral effects and should be reviewed as part of any behavioral assessment

Pharmacologic Options by Behavioral Target (Duis et al. 2022; Barry et al. 2005; Published IDD Literature)

Behavioral Target	Medication	Clinical Notes
Anxiety	Buspirone	Well tolerated; useful for chronic anxiety; slower onset of effect (1- 6 weeks)
Anxiety	Sertraline, Fluoxetine, Escitalopram (SSRIs)	Start low, titrate slowly; watch for constipation, appetite increase, headache, nausea and behavioral activation. Slow onset of effect (1 - 6 weeks)
Anxiety	Clonidine, lorazepam, clonazepam, propranolol, hydroxyzine	Can be used as needed for anxiety, especially with procedure, blood draws and maybe anticipated separation
Irritability / Aggression / Self-Injurious	Guanfacine, clonidine (alpha-2 agonist)	Targets hyperarousal, impulsivity, and anxiety; also clonidine supports sleep initiation. Monitor constipation
Irritability / Aggression / Self-Injurious	Risperidone, Olanzapine, Seroquel, Aripiprazole (atypical antipsychotics)	Low doses can improve mood regulation, may also help with hyperactivity; can increase weight, cholesterol and blood sugar, low risk for worsening tremor/muscle twitches. Watch for fatigue and constipation

Behavioral Target	Medication	Clinical Notes
Irritability / Aggression / Self-Injurious	Valproate, clobazam, topiramate, lamotrigine	Dual role as mood stabilizer and antiepileptic; useful when behavior change co-occurs with seizure activity or EEG changes. Topiramate can reduce appetite. Valproate can worsen motor function
Hyperactivity / Impulsivity	Guanfacine and clonidine (short and long acting), atomoxetine and viloxazine	Monitor constipation, fatigue, nausea and appetite decrease; slow onset effect (2 - 4 weeks). Consider a long-acting liquid clonidine in patients sensitive to fatigue
Hyperactivity / Impulsivity	Stimulants	Stimulants generally used cautiously in AS; may exacerbate seizures, hyperactivity or irritability; low-dose trials with close monitoring only, maybe better tolerated in adults

Monitoring & Safety Considerations

- Atypical antipsychotics (risperidone, aripiprazole): baseline and periodic weight, Body Mass Index, fasting blood sugar, cholesterol, and movement disorder assessment
- SSRIs: monitor closely for behavioral activation (increased agitation, irritability, hyperactivity) particularly in the first 2–4 weeks; more common in AS than in neurotypical populations
- Alpha agonists (clonidine, guanfacine): monitor blood pressure and heart rate; avoid abrupt discontinuation
- Valproate: can cause worsening motor skills and should not be considered in individuals in AS with major motor deficits; LFTs, CBC, ammonia levels; drug interactions with other AEDs are common and clinically relevant





Frequently Asked Questions

This approach transforms this FAQ section from a collection of answers into a practical demonstration of the clinical reasoning process this guide is aiming to provide

My child is struggling with transitions. How can I best support them?

1. This is fairly common in AS
2. Can visual support (visual schedules or social stories) or AAC help?
3. Are we accidentally reinforcing avoidance?
4. What proactive strategies (reward systems) can be implemented?
5. Could anxiety, particularly separation anxiety, be contributing?

My child's behavior has suddenly changed. Could there be a medical reason?

1. Why now?
2. Evaluate for medical issues; pain, constipation, reflux, seizures/EEG
3. Assess sleep
4. Review medications and potential side effects
5. Assess nutritional status
6. Evaluate anxiety
7. Perform functional behavioral assessment (FBA) to determine what drives behavior

How do I know if a behavior is related to communication frustration?

1. Is the child able to communicate this need another way?
2. Does the behavior occur when communication demands increase?
3. Does the behavior decrease when AAC is available?
4. What is the behavior accomplishing?
5. Can a replacement communication skill be taught? For instance, a communication strategy to get attention

What should I do when a behavior is happening at school but not at home?

1. Ask for a Functional Behavioral Assessment to determine antecedents and consequences.
Is it possible to get a behavior consultant involved to support the school with the evaluation?
2. Why does it happen only at school?
3. Is the social school environment over stimulating?
4. Is the individual seeking attention from peers?
5. Is the child or teen struggling with a different teacher or aide?
6. Are communication supports consistent?
7. Could anxiety be school-specific?

When should a family seek help from a behavioral specialist?

After:

1. Medical causes have been considered
2. Communication supports have been optimized

Or immediately if:

- Safety is a concern
- Self-injury is severe
- Aggression is escalating
- School participation or day program attendance is affected
- Improvement in ability to separate at night for bedtime
- Developing a toilet training plan

How do sleep, pain, anxiety, or seizures affect behavior?

- Pain often causes irritability, aggression, and self-injury. Pain can also worsen tremors and lead to new onset of myoclonus.
- Sleep deprivation and anxiety reduce emotional regulation.
- Seizures may present as sudden behavioral change or regression.
- Anxiety may present as recurrent vomiting or anxiety about walking and fear of falling

Sources: ASF Behavior Training Series (angelman.org); Duis et al. 2022 AS Standards of Care, Mol Genet Genomic Med; Barry et al. 2005 Behavioral Aspects of AS; GeneReviews AS Chapter (Dagli et al., updated 2025); Keary CJ & Ochoa-Lubinoff C. ASF Educational Webinar Series.

