



NATIONAL **FRAGILE X** FOUNDATION  
**FRAGILE X** CLINICAL & RESEARCH CONSORTIUM

Consensus of the Fragile X Clinical &  
Research Consortium

---

# BEHAVIORAL CHALLENGES IN FRAGILE X SYNDROME

First Published: June 2011  
Last Updated: December 2018  
[fragilex.org/consensus](http://fragilex.org/consensus)



## Introduction

Fragile X syndrome (FXS) is a genetic condition that causes developmental disabilities in both males and females. Though FXS occurs in both genders, males are more frequently affected than females, and often with more severity. FXS is the most common cause of inherited intellectual disability and the most common-known single gene cause of autism spectrum disorder (ASD) that accounts for about 2-3 percent of all ASD cases. At this time, there is no medical test, such as a blood test or brain scan, which can diagnose ASD; rather it is diagnosed by interview and clinical observation. Anxiety, especially social phobia and specific phobias, as well as generalized anxiety, is very common in FXS and often disabling.

FXS is a genetic disorder caused by a full mutation in the Fragile X Mental Retardation (FMR1) gene. Full mutations in FMR1 cause a deficit in Fragile X Mental Retardation Protein (FMRP). This deficit in FMRP results in abnormal communication among brain cells, which in turn affects learning and behavior. In many, though not all cases, FXS causes a spectrum of behavioral conditions in affected individuals, which can be challenging to their families, peers, and teachers, and can limit their interactions and opportunities within the wider society. At this time, there is no cure for the condition; therefore, management of FXS is largely focused on treatment of the challenging behaviors.

Behavior management tools may include medication(s) and/or other treatment modalities, but ideally involves the combined efforts of a transdisciplinary team that ideally includes a psychologist, a physician who can prescribe and manage medication treatment (often a psychiatrist, neurologist, or developmental pediatrician), occupational therapist, speech language therapist and educators, behavior analyst all working together with the family, school and community providers.

## Behavior and Fragile X

Individuals with FXS often present with many endearing personality qualities and relative adaptive strengths in addition to challenging behaviors. While there are commonalities in behavior challenges in FXS, the intensity, frequency, and duration vary greatly and are influenced by other factors, such as their environment and medical conditions. Behavior problems can worsen if they are reinforced in a variety of ways; a behavioral specialist can help to specify factors that may exacerbate behavioral problems. It is critical to address behavioral concerns with an individualized approach.

The most common behavioral challenges seen in FXS include behaviors associated with generalized anxiety, social interaction difficulties (this includes social anxiety/withdrawal and social aloofness), attention deficit hyperactivity disorder (ADHD), self-injury, and aggression. Addressing these challenges needs to be done with consideration to the person's level of intellectual functioning, speech/language abilities, and ASD symptoms, if applicable.

High states of arousal, or hyperarousal, a common feature of individuals with FXS, are thought to be biologically driven but triggered or increased by environmental and social stimulation.

Individuals experiencing overstimulation or hyperarousal may feel anxious or fearful. Some may become flushed, their ears and cheeks may become red, and their palms may begin to sweat. Their behavior may appear disorganized, and they may begin to engage in repetitive movements or speech. Some may become obstinate, refuse an activity or attempt to leave the stimulating environment. They may become unfocused, inattentive, impulsive, or aggressive. Avoidance/escape behaviors are more prominent in individuals with limited ability to communicate. Social withdrawal and phobias can evolve into full blown reclusion especially in adolescents and adults as the demands of schedules in later life do not necessitate going out of their homes on a regular schedule.

Many males, and some females with FXS, exhibit behavioral symptoms consistent with a diagnosis of ASD, a developmental disorder primarily characterized by a selective impairment in social interaction, stereotyped and repetitive behaviors, and communication deficits. Specifically, people with ASD have differences in how they understand and react to people and social situations, which result from differences in how their brains process socially-relevant information. Symptoms of ASD appear in early childhood. It is a lifelong disorder, though symptoms change over time. In some cases, the cause of ASD is known, such as FXS, a genetically-defined condition that can be diagnosed by a DNA blood test, unlike ASD that is a *behaviorally*-defined diagnosis. This does not mean that individuals with FXS have two separate, unrelated disorders; the FMR1 mutation is simply the underlying cause of their ASD symptoms.

Understanding the behavioral challenges and common triggers in FXS are the key to effective behavior management. Developmental differences, such as delayed language and sensory sensitivities, often contribute to behavioral challenges, especially in younger individuals. A loss of capacity for self-regulation, manifesting in aggressive outbursts or self-injurious behaviors is often observed in individuals with FXS, often based on a



specific trigger, which may or may not be easy to identify. Difficulty handling transitions, new environments and changes in routine often present behavioral challenges, potentially including aggressive outbursts.

## Initial step - Comprehensive Evaluations

Addressing behavioral challenges in an individual with FXS should start with a comprehensive evaluation. A transdisciplinary team approach ideally includes: educators, school administrator, psychologist, psychiatrist or other physician if medication is being considered, behavior analyst, occupational therapist, speech/language pathologist and caregivers. If the child has previously had a comprehensive evaluation, the evaluation should be reviewed and updated. There should be documentation of relative strengths and weaknesses across all developmental domains. Comprehensive evaluations should be used whenever developing behavioral support plans, behavior intervention plans, educational plans, specially designed instruction, vocational plans, etc.

The following are important components of a transdisciplinary comprehensive evaluation.

- A thorough psychological and adaptive behavior evaluation should be conducted and/or reviewed regularly. If school-aged, academic and achievement assessments are important aspects to include. Adults with FXS benefit from vocational and adaptive assessments as part of an annual service plan review. Input from family, independent providers, and school personnel should be solicited. This information is crucial to deciding if the goals and strategies are appropriate and realistic for the person with FXS.
- Many children with FXS should be formally evaluated for autism spectrum disorder (ASD). A diagnosis of autism is typically made by a specialist (psychologist, developmental pediatrician, neurologist, or psychiatrist). A comprehensive evaluation for ASD should include an in-depth parent interview and formal assessments of cognitive skills, adaptive skills, speech and language skills, social development, and behavioral differences.
- An occupational therapy (OT) evaluation should be conducted to evaluate and create a plan to address fine motor skills, motor planning, sensory issues, self-regulation skills, adaptive functioning and daily life skills. An OT evaluation can identify sensory sensitivities in a person with FXS and the profile of dysregulation

that often is associated with these difficulties. Challenging behaviors can be the result of the ways the child acts to try to eliminate, avoid or cope with the aversive sensory input. Examples include taking off clothes, throwing oneself on the ground to avoid entering a loud space, or covering ears. Additionally, the child may seek or need more sensory input (leaning on or pushing into people or stuffing too much food in the mouth) as a way of coping or helping to deal with the challenges they are struggling to manage. A child can be both sensory seeking and sensory avoidant, which can prove challenging for caregivers. The OT evaluation helps to describe the way the sensory or motor foundations in the child might be best addressed, through direct intervention or environmental and interactional changes in the world around them. Identifying daily strategies to address the sensory and motor challenges and to support the acquisition of self-regulation and coping skills is central to the OT's role on the team and important for a comprehensive behavior intervention plan. Additionally, difficulties with coordination and skill development, and with fine motor skills and handwriting skills compounded by dislike for hand over hand instruction may be triggers for challenging behaviors. These elements should be included in the OT evaluation.

- A comprehensive speech and language evaluation provides a great deal of information and can shed light on the function of challenging behaviors. Delayed speech and language development is noted in many boys with FXS. Oral motor difficulties may significantly impact sound production skills, making expressive language difficult to understand. Some individuals may be minimally verbal due to delayed or deficient oral motor planning skills. Difficulty with expressive speech and language and/or limited means of functional communication is a cause frustration for many boys with FXS. Also, attentional deficits and severe autistic features are also associated with relatively greater impairment in receptive language. In a state of hyperarousal, language demand can increase the arousal further, compounding an already compromised situation. Language frustration often results in challenging behaviors including: crying, grabbing items, hand biting or aggression. Refusal to follow directions may not be oppositional behavior but rather due to receptive language deficits (inability to understand the directions), expressive language deficits (inability to refuse or respond to the direction) or delayed auditory processing. Some boys and men may say "NO" initially in response to any direction. The transdisciplinary team should assess and parse out if this is a learned response to accommodate for poor auditory processing, poor receptive language skills or heightened state of arousal rather than noncompliance.



- A physical therapy (PT) evaluation should be completed to assess the level of joint laxity (looseness) and generalized mild hypotonia. Additionally, general gross motor skills are often included in the PT evaluation. These conditions are noted with varying degrees in most individuals with FXS. Mild hypotonia is often associated with fatigue following physical exercise. Fatigue may be a reason for refusal to continue an activity, particularly if it involves prolonged exertion. Frustration with motor skills can lead to resistance and hesitant participation and the PT evaluation can assist the team in best ways to alleviate these challenges
- Evaluation for ADHD can provide an understanding of how inattentiveness and/or poor impulse control may contribute to behavioral issues, such as lack of focus and poor response to redirection. Destroying materials or swiping at stimuli may be symptomatic of poor impulse control. Hyperactivity may also affect the person's ability to stay engaged long enough to finish an activity or comply with a demand. An evaluation of ADHD symptoms assists in planning activities/goals that are appropriate and in alignment with the person's attention span, (e.g., time on task, waiting time).
- A sleep study should be conducted if persistent sleep issues are noted by caregivers or daytime drowsiness is noted.
- Approximately 10-12% of individuals with FXS have a co-occurring seizure disorder, with a higher prevalence in males (13-15%) and in those with a diagnosis of ASD. If the person with FXS has a seizure disorder, an evaluation by a neurologist may provide additional information for planning and interventions. Some anti-convulsants used to treat seizures can aggravate behavior. It is important for the team to help the neurologist monitor behavior when medications are changing, so that adjustments can be made if behavior specifically worsens after medication changes. Likewise, some anti-convulsants have positive behavioral effects and thus careful weaning of medication for seizures is warranted.
- A functional behavioral assessment (FBA) should be conducted to develop a hypothesis of the function(s) of the challenging behavior and the maintaining consequences. To start, the team, often led by a behavior analyst or psychologist, should carefully describe the challenging behavior of concern in an observable and measurable way. This description provides consistency among the family and those working with the person with FXS so that data collection can be used to determine whether progress is being made once an intervention is started.

- Steps in the FBA process include identification of the following: medical concerns, history of the behavior and treatment approaches, possible triggers (activities, people, places, specific demands, etc.), preferences and possible reinforcers, and consequences that will not maintain the challenging behavior. Outcomes of the FBA include strategies for positive behavior support plans, and behavior intervention plans including strategies to prevent the behavior from occurring (antecedent strategies) and strategies to teach alternative /replacement behaviors. Ongoing collection and analysis of data is recommended to test the hypothesis of the FBA and identify triggers, trends in behavior, and multiple functions and maintaining consequences of challenging behaviors.

The culmination of the multidisciplinary evaluation process is best utilized when the information is integrated into a plan that is consistently utilized across providers and environments and maintained with consistent communication and collaboration. It is important to collect baseline and post intervention assessments. Regularly scheduled team meetings should be convened to review progress, update goals, strategies, reinforcers, etc. in a continuous and concerted plan.

## Behavior Management and Current Treatment Guidelines

Central to an effective plan for addressing behavioral concerns is a transdisciplinary approach across home, school and community settings. Effective management of physical and medical issues such as seizures should be addressed prior to a behavior intervention plan. Additionally, the team should ensure that there are appropriate accommodations to address functional communication skills and that the educational, vocational, or community program is appropriate for the individual's needs.

Behavior problems serve a purpose (or a function) and are often a form of communication. The functions of behavior are sometimes reduced to four basic categories: 1) to gain sensory input, 2) to receive attention from others, 3) to access preferred objects or materials, and 4) to escape or avoid something undesirable (e.g., activity, person, place, demand). Thoughts and feelings are private behaviors but may contribute to observable behavior. For example, if a person with FXS is feeling anxious, observable behaviors may include: flushed face, red ears or increased hand flapping, verbalizing the same statement repeatedly (perseveration) or mouthing of clothing. These behaviors often occur just prior to elopement or dropping to the ground and may function as escape from uncomfortable feelings, loud noises, fearful settings or activities, etc.



The purpose or function of the behavior is generally appropriate (individual is thirsty and wants a drink). It is the behavior the person utilizes that is not acceptable (grabbing someone's water). In this example, it is recommended to teach alternative appropriate behaviors. For example, functional communication training can be used to teach the appropriate way to request and access a desired item. Reinforcement for engaging in the appropriate strategy is access to the desired item as well as additional reinforcement such as social praise, although praise may not be effective for everyone with FXS. It is also important to be mindful of the need for supports and instruction in appropriate ways to express frustration, fear or protest (e.g., crowded setting, change in schedule) rather than engaging in challenging behaviors. Anxious individuals may become hyper-aroused causing already compromised communication skills to worsen and result in more significant behavioral challenges. An FBA conducted by the transdisciplinary team who are familiar with FXS will contribute to a better understanding of the multiple functions and maintaining consequences of challenging behaviors seen in individuals with FXS.

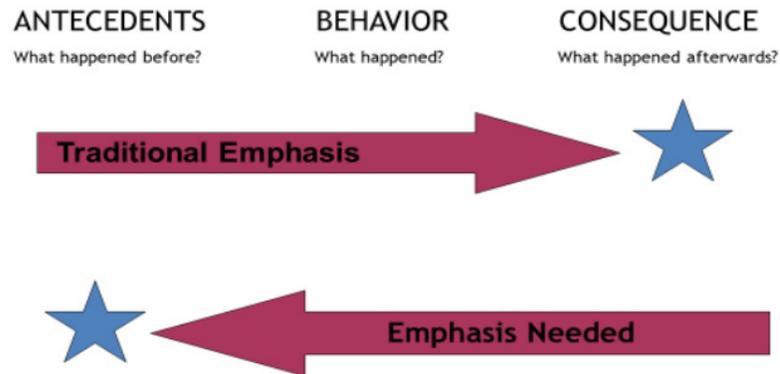
Common triggers for challenging behaviors in children and adults with FXS include:

- Forced eye contact
- Unclear expectations
- Verbal instructions that are unclear, too rapidly spoken or not at developmental level
- Lack of consistency and routine
- Environmental noises, crowds, and close proximity to others
- Transition, even when from preferred to preferred activities
- Having to wait without clear understanding of the time requirement
- Heightened emotion or excitement

Traditional strategies for addressing behavioral issues in children with intellectual and neurodevelopmental disorders include the use of positive reinforcement for appropriate behavior, shaping, reward systems, time-out, loss of privileges, and ignoring negative behavior. Such strategies are widely used in educational and residential settings. These approaches are “consequence-based” in that they are implemented after a behavior has occurred. While useful in shaping, reinforcing, increasing, or maintaining positive behaviors, these approaches are less effective for decreasing inappropriate behaviors with individuals with FXS. Preventative or antecedent strategies facilitate the learning of new skills, rather than addressing inappropriate behaviors that have already occurred.

A behavior plan that is “front-loaded” with antecedent strategies is recommended so that the person with FXS is supported and set up to engage in appropriate behaviors which will then be reinforced by positive consequences. (see figure below)

## THE ABCs OF BEHAVIOR



While no two individuals with FXS present with the same repertoire and/or severity of behavioral issues, there can be striking similarities. Knowledge of behavioral and learning characteristics associated with FXS provides a foundation to develop appropriate supports and interventions.

Evidence-based behavioral interventions grounded in Applied Behavior Analysis (ABA) are often effective. ABA is not a specific program but rather a behavioral framework from which specific therapeutic interventions (e.g. Lovaas therapy, verbal behavior, discrete trial, etc.) have been developed. As such, there may be wide variability from one ABA program to another. It is important to keep in mind that systematic instruction, careful ongoing assessment, positive reinforcement, and attention to the ABC’s (antecedents, behaviors, and consequences) of behavior are essential aspects of any successful therapeutic program, regardless of whether or not it is designated as “ABA.” Decisions about the appropriateness of ABA services for a child with FXS should be made after careful evaluation of the individual. Discrete trial training (DTT) and intensive table teaching (ITT) procedures may need to be less direct and may include another student to utilize peer modeling or turn taking. Staff working with these students should continue to glean ideas and potential strategies from well-established programs while keeping in mind the syndrome-specific characteristics associated with FXS.

The following general treatment guidelines may be useful to customize the environment and set the stage for success for the person with FXS.

## **General treatment guidelines:**

**Speech and Language:** For many young males and some adolescents and adults with FXS, verbal speech and communication is not functional across settings. Rapid rate of speech or poor intelligibility and anxiety interfere with effective communication. Augmentative and Alternative communication systems (AAC) may be suggested for individuals with FXS. AAC refers to all tools that are used to supplement or replace speech when it is not sufficient to meet the individual's needs. AAC includes high-tech tools, such as tablet technology, as well as low-tech strategies, such as a picture communication and sign language. Access to language may increase functional communication and reduce frustration which in turn may decrease challenging behaviors.

**Feeding:** Though not always a behavior problem, feeding time may be fraught with frustration due to sensory, motor, or medical reasons. The child may overstuff his mouth, be aversive to specific textures and have hypotonic oral muscles. Feeding can require patient and gradual introduction of new foods. Speech and/or occupational therapy can help develop appropriate use of the mouth and oral cavity, normalize sensory and motor functions and build healthy routines around meals and food. Nutritionist/Dietician can also play a role to help with picky eating, and weight issues.

**Anxiety:** When people experience anxiety or stress, they often have both physical and emotional reactions. Physical symptoms may include increased heart rate, redness of face or neck, sweating, stomach ache, trouble breathing, muscle tension, frequent urination, gagging or trouble swallowing. Emotional reactions may include feelings of fear, worry, or apprehension. During times of stress, transition, or frustration the sympathetic nervous system is activated by the brain, and the body responds with a "fight or flight" response. Commonly reported anxiety triggers in FXS include: being rushed, changes in routine, new plans or routines, making decisions, going from one place to another, transitioning from one activity to another, items out of place, loud noises or crying children.

Social anxiety is present to some extent in the vast majority of individuals with FXS. This can range from shyness to a clinical diagnosis of social anxiety disorder. When evaluating social responsiveness, individuals with FXS are often motivated to be social, but become overwhelmed by the intensity of the social interaction and expectations causing a negative or avoidant reaction. Some triggers for social anxiety include: being put on the spot, forced eye contact, standing up in front of people, making phone calls, attending a party or large gathering and meeting new people.

Many people with FXS exhibit the following behaviors in response to anxiety: laughing, fidgeting, pacing, crying, biting their hands, running out of the room, crawling under a table, shutting down (immobilized, standing still, remaining silent), hitting or throwing objects. Some individuals may develop selective mutism, which is when the person does not speak in unfamiliar or uncomfortable situations but has better communication in a familiar setting. Some children and adults with FXS exhibit behaviors that appear to be oppositional and defiant but may have the function to escape the anxiety of entering a new environment or a transitioning even when it is to a preferred activity. The seeming compliant, lovable child may become reluctant to follow directions such as cross a threshold, go out of the house, or disengage in a preferred activity. It can be helpful to give the child a “job” to perform or a transitional object to carry for another person while crossing the threshold. In this case, the child becomes focused on the task at hand rather than the transition. Try to provide the activity before the actual transition (e.g., before the bell rings for a change of class) so the child is complying with one activity and not having to cope with environmental stimuli and fear of transition at the same time. Providing a picture schedule helps define the activity and may reduce the anticipatory anxiety associated with it.

Anxiety triggers and behavioral responses vary from person to person, by age and/or developmental level as well as by the consequences to the behavior. Treatment for anxiety includes: limit known triggers while teaching self-regulatory behaviors, gradual introduction of triggers, and proactive strategies such as visual supports, video modeling, role play and rehearsal. Some older and higher functioning individuals with FXS benefit from Cognitive Behavior Therapy (CBT) and therapeutic interventions that involve the acquisition of coping skills. Medication choices are like those for the general population and should be viewed as adjunctive therapy that supports implementation of behavioral treatments. Input from a transdisciplinary team should be solicited when developing an intervention protocol for a person with FXS.

Gaze aversion, thought of as an emotional coping mechanism, is also a distinctive feature of FXS, related to a hyper-reactive sympathetic response to faces and other social stimuli. Family and staff working with people with FXS should be aware that gaze aversion (actively avoiding eye contact) could be due to a sensory overstimulating effect of eye gaze and social initiation by others that is uncomfortable for the person with FXS. Anyone working with an individual with FXS should not demand eye contact, due to the aggravation of hyperarousal and anxiety provoked by this in FXS. It should be noted that many people with FXS will make eye contact once they become more comfortable with the person.



**Hyperarousal:** When problem behaviors occur, it is important to recognize that they may be caused by overstimulation or hyperarousal. If a source of overstimulation is identified, environmental accommodations may be helpful. When accommodations are not possible, it may be advisable to remove the person from the over-stimulating environment. It is beneficial to teach children and adults with FXS an appropriate way to request a break when overstimulated. A quiet, “safe” place could be designated as a place to retreat and regain composure. Some individuals with FXS may take an extended amount of time to de-escalate.

It is important to recognize that the sources of stimulation vary within environments such as classrooms, and to situate individuals away from arousing stimuli. For example, seat the child away from the bell that signals class changes, or away from open doors that admit distracting sounds from adjunct rooms or hallways.

When exposure to intense stimulation is unavoidable, such as during assemblies, fire drills or other busy events, a familiar person should accompany the individual to help him remain calm and provide continuity and support. Preparing the person with FXS for intense stimulation should be included in programming (desensitization procedures, etc.) or be a part of the behavior support plan. It may also be prudent to provide noise cancelling headsets, I-pod with music and ear buds, etc., to help “ground” the individual when the stimulation is unavoidable.

Unfamiliarity and unpredictability can be arousing and attempts to reduce these for the person with FXS are highly advisable. Strategies along these lines may include engaging in role play to help an individual anticipate social situations or providing picture schedules to prepare for changes in activities or routines.

An occupational therapist can provide valuable insight and advice regarding accommodations to improve an individual’s physical and social environment to reduce overstimulation as well as how to help the individual with FXS acquire the self-regulation skills needed to manage their hyperarousal.

**Sensory:** Intervention for sensory sensitivities typically involves a program developed by an occupational therapist (OT). For some individuals with FXS, a treatment that is focused on desensitization and/or amelioration of uncomfortable sensations is applicable (e.g., loud noises, crowded settings, texture sensitivities). For some individuals engaging in undesired sensory-seeking behaviors, the OT can help develop a plan to substitute a more appropriate behavior serving the same sensory-seeking function,

or function or can work on a plan to extinguish the behavior. In some situations, accommodations are encouraged, e.g. when a parent is mowing the yard, noise cancelling headphones can be worn.

**ADHD:** ADHD is common in FXS. Males with FXS typically present with the combined hyperactive-impulsive and inattentive subtype. Females are more likely to present with the inattentive subtype. When making a diagnosis of ADHD, it is essential to consider that other underlying issues, or simply young age, may present as ADHD. Toddlers and young children with FXS and ADHD symptoms often present with significant behavioral challenges, particularly hyperactivity. These behaviors can manifest as pacing, jumping up/down, excessive talking or constant moving. It is critical to address safety concerns by setting clear boundaries and rules for unsafe behaviors. Hyperactivity often lessens over time, replaced by impulsivity. Impulsive behaviors, noted in later childhood include difficulty waiting, rushing ahead of the group, and grabbing items. Hyperactivity, impulsivity and limited attention span are often barriers to learning. Inattentive behaviors can be addressed with strategies such as structured highly motivating learning opportunities with visual supports as well as specific positive reinforcement for attention to task. It is equally important to make environmental accommodations such as minimizing distractions.

Initial diagnosis of ADHD symptoms may simply involve observation. Once a conclusive diagnosis of ADHD is made, medication is often prescribed, and should be used in tandem with behavioral interventions. It is also important to consider if the individual with FXS has a co-occurring anxiety disorder, as some ADHD medications may exacerbate anxiety symptoms.

**Aggression:** Many significant behavioral problems in boys with FXS are secondary reactions to anxiety and accompanied by hyperarousal. These reactions often follow a predictable pattern of escalation or behavioral chain. Early signs typically include hand flapping, hand or finger biting, redness of face or ears, verbal perseveration (repetition of short phrases, noises, sounds, etc.), and chewing on clothes (collars, sleeves). If unabated, hyperarousal may then result in loud verbalizations which sometimes include cursing, threats, screaming, and making threatening gestures. These reactions can escalate to property destruction (throwing objects, slamming doors, kicking tables, etc.) and ultimately lead to physical aggression toward others. The key to effective behavior support for individuals with FXS, lies in recognizing and minimizing triggers such as reducing environmental distractions which lead to hyperarousal, and establishing a consistent behavioral approach across all settings. With the knowledge of known



triggers and early signs of escalation, appropriate interventions can be put in a behavior intervention plan to interrupt the behavioral chain and replace challenging behaviors with appropriate prosocial responses. Additionally, offering specific praise for utilization of coping strategies and refraining from aggressive behaviors is an important component of behavioral support.

Individuals with FXS will commonly lash out, either at an offending individual or a trusted person. Lashing out at a trusted person often happens when the individual with FXS wants to avoid the conflict with a person offending them. This is common among females and higher functioning males who are better able to formulate a more regulated response. Many times, the person with FXS exhibits signs of confusion or expresses sadness after the incident and once calm, will usually apologize. A protocol should be in place that clearly outlines how and when the person with FXS returns to regularly scheduled activities.

If aggression is severe or the individual who is aggressive does not respond to management strategies, medication may be necessary. Atypical antipsychotics are often utilized and helpful in individuals with irritability and temper tantrums.

**Self-Injurious Behaviors:** Many individuals with FXS engage in self-injurious behaviors, the most common of which is hand biting. They often explore objects with their mouths long into their development. It is common to see older children putting many things in their mouths (including fingers and pieces of clothing). Hands-to-mouth may have started as a form of self-soothing but may be inadvertently reinforced by attention given to the behavior by caregivers. What started as a self-soothing behavior, may now have multiple maintaining consequences. The transdisciplinary team should conduct an FBA and develop appropriate intervention strategies that may include teaching appropriate self-soothing activities, appropriate alternative activities, as well as desensitization techniques, if applicable. The team may note a behavioral chain such as biting on clothing, perseverative speech or verbal comments that are exhibited prior to hand-biting or finger chewing. Behavior strategies should include interruption and engagement in alternative and appropriate self-soothing activities. Often, an IEP includes behavioral goals which can be effective in strategizing and teaching replacement behaviors, such as using words or pictures to communicate a protest rather than self-injury. Goals should include encouraging these skills at a more independent level.

## Proactive or Antecedent Strategies to support behavior and learning

### Effective Strategies to support children and adults with FXS include:

- Clear, concise verbal instructions; always remember to reduce verbiage
- Consistent expectations, routine and structure
- Visual supports and strategies, e.g., individual schedule
- Allow for possible delay of initial response to direction or instruction
- Ample personal space
- No demand for eye contact
- Limit known individual triggers, e.g., noises, crowds, specific environments
- Provide transition activities, e.g., job and/or transitional items
- Allow the child or adult closure before changing activities
- Consistent responses to challenging behaviors among caregivers and across settings
- Interrupt behavior chains; distract and engage in alternative behavior and/or venue

**Crisis Intervention Plan:** For school aged children, a Crisis Intervention Plan (CIP) should be included as part of the Individualized Education Program (IEP). For adults with FXS, it is essential to create a plan or protocol to be utilized in the day program, work and/or community settings. This should be developed and reviewed/updated annually. This plan will be critical to prepare staff to be proactive rather than reactive. The specific response to the crisis depends upon the individual situation. When there is a history of unsafe behaviors, it is crucial that the family and team develop a written crisis plan that focuses on safety. If crisis related behaviors are likely to occur across settings, separate crisis plans may be necessary.

This proactive crisis intervention plan should emphasize de-escalation interventions e.g., provide a quiet, safe, familiar place for the child/adult which removes the audience and possible triggers. The plan should delineate the behaviors that may be seen in a crisis as well as the different phases of escalation. It should include specific instructions of the interventions that will be implemented, who will be contacted with a list of current contact information. Contacts may include family members or a friend who would be helpful in a crisis, such as a psychologist or behavioral health provider who knows the child. It may also be necessary to include the local crisis line number, and walk-in centers or emergency rooms. In an emergency, as with anyone, you should call 911. If 911 should be called, emergency responders should be informed about the individual's unique needs.



## Conclusion

Interventions designed by a team who is knowledgeable of the behavioral, learning, medical, and physiological characteristics associated with FXS are usually helpful in reducing challenging behaviors. A proactive approach with appropriate supports and accommodations will likely foster positive outcomes and set up the child or adult with FXS to succeed in their home, school or community.

## Resources

There are many resources available on the National Fragile X website [www.fragilex.org](http://www.fragilex.org) including quarterly articles, webinars, medical guidelines, etc.

Current treatment guidelines and recommendations for a wide range of topics are available <https://fragilex.org/our-research/treatment-recommendations/>

Topics related to this guideline include: Educational Guidelines, Medications for Individuals with FXS, Assessment in FXS, Sensory Integration Issues, Physical Problems in FXS, Seizures in FXS, Hyperarousal, and Autism Spectrum Disorder in FXS.

Author note: This guideline was revised by Barbara Haas-Givler, MEd, BCBA, Cora M Taylor, PhD, Karen Riley, PhD, Marcia Braden, PhD, Dejan Budimirovic, MD, Jean Frazier, MD, Matthew Kinney, MA, BCBA, Tracy Stackhouse, MA,OTR, Sarah Scharfenaker, MA,CCC-SLP and Walter Kauffman, MD. This document was originally authored by Jonathan Picker MD, PhD, and Vicki Sudhalter, PhD. It has been reviewed and edited by members of the Fragile X Clinical & Research Consortium and represents the current consensus of its members.

*The **Fragile X Clinical & Research Consortium** was founded in 2006 and exists to improve the delivery of clinical services to families impacted by any Fragile X-associated Disorder and to develop a research infrastructure for advancing the development and implementation of new and improved treatments. Please contact the **National Fragile X Foundation** for more information. (800-688-8765 or [fragilex.org](http://fragilex.org))*