

# STRATEGIES FOR MANAGING DAY-TO DAY LIFE WITH FRAGILE X SYNDROME

Fragile X Syndrome (FXS) is the leading known inherited cause of Autism Spectrum Disorder (ASD) and Intellectual Disability (ID). It is recommended that all children with ASD or unexplained developmental delays should have genetic testing for FXS as part of their evaluation. <sup>\*2</sup>

Males with FXS inherit the syndrome from their mother, while females can inherit from their mother or father. Males tend to be more severely affected by this syndrome when compared to females. However, both males and females can present with deficits in cognition/learning, language, adaptive skills (self-help), motor skills, and social-emotional skills. <sup>\*2</sup>

Individuals with FXS have a unique cognitive-behavioral phenotype, or clinical presentation. They exhibit characteristics that differ from classic non-syndromic Autism Spectrum Disorder (ASD without FXS). Higher levels of anxiety in children with FXS correlates with increased social avoidance. This relationship between anxiety and social avoidance is actually stronger in children with FXS when compared to children with non-syndromic ASD. <sup>\*4,5</sup>

In addition to very high levels of anxiety, individuals with FXS have behavioral challenges most often related to hyper-arousal. When a child with FXS experiences hyper-arousal, they exhibit a “fight-or-flight” response and much of their behavior is no longer within their control. <sup>\*3</sup> For example, when a child with FXS is going to see a preferred person, their body may become overwhelmed with excitement when the individual arrives. This can trigger an episode of hyper-arousal where the child may act out aggressively despite being very excited to see their familiar friend.

Despite the above challenges, individuals with FXS also have several strengths. Individuals with FXS show strengths with social awareness (demonstrated by their

performance on facial perception assessments)! <sup>\*1,2</sup> This is atypical for children with more classic features of ASD (non-syndromic ASD). Children with FXS are very much aware of the emotions and mood of those around them. It’s important to understand that a child with FXS may appear to be in their own world, but they are in fact always listening and aware of what those around them are doing. However, due to hyper-arousal these individuals are often affected by the mood and behavior of those near them, which could have a positive or negative impact on the individual with FXS and their own behaviors. Another strength is in long-term memory and verbal-visual associations. Individuals with FXS will remember details over time. Objects, locations, or even people will prompt them to recall details about past events. <sup>\*1,2</sup>

Here are five tips and strategies for parents and caregivers to integrate throughout the day to help manage anxiety and hyper-arousal in order to prevent challenging behaviors and facilitate learning:

**1. Be aware of your own attitude and mood** – Your child is very much aware of your mood. Use this to your advantage. When you are calm and relaxed you are modeling the appropriate behavior for your child. Children with FXS learn so much by observing others and this can help them to relax when they are feeling anxious or distressed. In contrast, if a child with FXS sees you are feeling overwhelmed or upset, this may trigger them to experience hyper-arousal leading to challenging behaviors.

**2. Incorporate high interest materials when learning new concepts** – We know children with FXS will remember things overtime but this skill can be enhanced when high interest material is incorporated into a task that may otherwise not interest the child. When preferred interests are involved the child with FXS is more engaged and may have less anxiety allowing for learning to take place. This can also improve self-confidence when completing something difficult or new. <sup>\*1</sup>



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**3. Transitions are hard. Try side conversations to preview** – Transitions can trigger anxiety and hyper-arousal which leads to non-compliance and challenging behaviors. It is well known amongst experts working with children with FXS that side conversations are a great way to prepare them for a transition. <sup>\*1</sup> Directing the conversation towards another person results in less anxiety when compared to a direct instruction. It also allows the child to hear what will be coming next and prepare for the transition to the next activity. Don't have a 3rd person to give the instruction to? No problem! Just start talking to yourself out loud. I would also use this as an opportunity for your child to be a “helper”. For example, “I could really use your help carrying my car keys to the car so we can go for a ride”.

**4. Take advantage of peer models or even video modeling of a new skill** – Children with FXS are paying attention to your actions and learn well when they have had opportunities to observe others before they are expected to perform a task. <sup>\*1</sup> This can be applied to many scenarios. For example, watching a video of a child getting a haircut or watching a sibling get a haircut. This helps the child with FXS know what to expect before the event occurs. Then talk about all the steps as they are happening and even acting out the scenario at home.

**5. Know when to take a break** – When you know your child is already experiencing hyper-arousal, this is not the time to insist on completing a demand. Remember, your child is no longer in control of their body so before they can follow instructions, they need to calm down. This is a good time to decrease stimulation. For example, turn down the lights, lower the tone of your voice, or go to a more quiet space. Then provide support and label the child's feelings. For example, “I know you are mad because making your bed is hard. Let's just sit and relax and listen to some quiet music.” You may even offer things that are known to relax your child, such as a blanket, favorite toy to hold, or even just rubbing their back. When your child

is more relaxed, this may be a good time to show a video of how to complete the difficult task. Offer to help them and decrease the total demand to just one part that you know they can do successfully. Then offer lots of praise for their effort and success to boost confidence when you try the task again. <sup>\*1</sup>

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