

Consensus of the Fragile X Clinical & Research Consortium on Clinical Practices

Hyperarousal in Fragile X Syndrome



First Issued: June 2011

Updated: October 2012

Introduction

Arousal refers to a general state of nervous system activation that is reflected in behavior, physiological activity and emotional experience. There is an optimum level of arousal that is necessary for controlled behavior, and this level is usually maintained by internal homeostatic mechanisms. One of the characteristics of fragile X syndrome (FXS) is an impairment of this homeostatic control of arousal. Another is a heightened sensitivity to environmental and social stimulation. Together, these factors combine to cause boys and girls with FXS to become aroused more easily, and to a greater extent, than others, and to remain in such a “hyperaroused” state for prolonged periods of time. While hyperaroused, such individuals are less able to control their behavior. They may act impulsively, have difficulty focusing and sustaining attention to tasks, exhibit speech and language impairments, and emit other “problem” behaviors.

Diagnosis & Recognition

The hyperarousal discussed above is believed to underlie many of the phenotypical behaviors commonly associated with fragile X syndrome. Individuals experiencing hyperarousal may become flushed, their ears and cheeks may become red, and their palms may begin to sweat. Their behavior becomes disorganized and they may begin to engage in repetitive movements. Some may become obstinate and not wish to continue an activity or to remain in the stimulating environment. They may become unfocused, inattentive, impulsive, aggressive, and display symptoms that mimic anxiety. Their behavior may become disorganized, repetitive and less efficient. Their speech may also become repetitive and perseverative.

Current Treatment Recommendations

When problem behaviors occur, it is important to recognize that they may be caused by hyperarousal, and to first try to identify any sources that may be overstimulating the person. The initial intervention should always be to try to reduce environmental stimulation. When that is not possible, it may be advisable to remove the person from the overstimulating environment.

A non-stimulating, quiet and “safe” place should be designated, so that when hyperarousal occurs, the individual can have a place to quickly retreat to where he will be able to de-arouse, regain his composure, and re-establish a sense of well-being before returning to his previous activity.

Hyperarousal in Fragile X Syndrome

Proactive strategies to prevent hyperarousal include modifying the physical environments of individuals with FXS (e.g., their homes, classrooms, and workplaces) to reduce their stimulating quality. Examples might include providing natural or incandescent lighting instead of fluorescent lighting, and introducing sound-dampening material such as curtains or acoustic tiles in order to minimize sound reverberation and echo.

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.

Unfamiliarity and unpredictability can be arousing, and attempts to reduce them 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 her 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 its arousing quality, and can also recommend ways to provide an appropriate sensory diet to help her maintain control and feel calm and focused throughout the day.

Teaching individuals with FXS personal stress-reducing strategies such as "square breathing," muscle relaxation techniques, and visualization of tranquil places, can also help them cope with unavoidable exposures to stimulation.

Medication may also be helpful in some cases, by reducing the person's threshold for becoming hyperaroused when exposed to stimulating environments.

Frequently Asked Questions

What are the types of conditions that will arouse individuals with fragile X syndrome?

Hyperarousing conditions vary according to the particular sensitivities of the individual. This is why an evaluation of the environment should be made to determine what might be causing the problem. Generally, auditory and visual stimulation, environmental complexity,

Hyperarousal in Fragile X Syndrome

unpredictability and eye contact are all stimuli that can trigger hyperarousal in many people with fragile X syndrome. Other triggers can include confrontational encounters, being made the center of attention, being asked to produce novel answers to new questions, and being asked to perform personally challenging tasks.

Will my child always experience hyperarousal or will he outgrow it?

Individuals with FXS are always susceptible to hyperarousal. Their environments may change, and their responses to hyperarousal may change as they age, but it remains an important consideration. It is useful to periodically re-evaluate the potential sources of stimulation in their environment, especially following changes in the home, school or vocational setting.

What is the role of eye contact in hyperarousal?

Making eye contact is a social behavior that signals interest in, and attention to, another person. Individuals with FXS tend to have social anxiety, and asking them to make eye contact can be very arousing. It is preferable to let them initiate eye contact to the degree that they are comfortable with it, and to avoid demanding it. The avoidance of eye contact is one way that individuals are able to exert some self-control over their own state of arousal, and when eye contact is forced on them it deprives them of this useful coping strategy.

Can medication help?

Medications may help an individual with FXS avoid becoming aroused too easily. The use of medications must be coupled with the prescribing physician's understanding of the environment and an awareness of the particular stimuli that cause an individual to become aroused. Because some environmental stimulation is unavoidable, individuals should be provided with strategies and tools to help them cope with it. In some cases appropriate medication may be considered one such tool.

What can I as a parent or caregiver do to help an individual with FXS de-arouse?

The most important thing is to eliminate or reduce the source of stimulation. When that is not possible, the individual must be removed from the stimulating environment. A quiet, non-arousing place should be available, where he can feel comfortable and safe, and can have time to regain his composure and self-control. Occupational therapy interventions, such as brushing or joint compression, may help him calm down. Hyperaroused individuals often recognize that they have lost control and may feel scared and guilty about their reaction. It is therefore important to express an understanding that they are not responsible for their emotional state, to offer support, and to praise them for trying to manage their behavior.

Hyperarousal in Fragile X Syndrome

Even non-verbal children will respond to such expressions of love, support and understanding, and be calmed as a result.

Additional Resources

The National Fragile X Foundation (www.fragilex.org) has articles and other materials on sensory integration and hyperarousal that can provide additional insights and suggestions regarding the effects of hyperarousal and strategies for preventing and coping with it.

Author note: This *recommendation* was authored by Vicki Sudhalter, PhD, and was reviewed and edited by consortium members both within and external to its Clinical Practices Committee. It has been approved by and represents the current consensus of the members of the Fragile X Clinical & Research Consortium.

Funding: This project was made possible by Cooperative Agreement U01DD000231 from the Centers for Disease Control and Prevention to the Association of University Centers on Disabilities (AUCD) and RTOI 2008-999-03 from AUCD to W.T. Brown in support of the National Fragile X Clinical and Research Consortium. The findings and conclusions in this report are those of the authors and do not necessarily represent the official position of the Centers for Disease Control and Prevention.

*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 www.fragilex.org)*