

Hyperarousal in Fragile X Syndrome: Teenage and Adult Issues

NFXF International Conference, Miami
July 27, 2012

Richard Belser, Ph.D.
Vicki Sudhalter, Ph.D.



New York State Institute for Basic Research
in Developmental Disabilities
Staten Island, NY

Outline of the Presentation

1. Intro to arousal modulation and hyperarousal
2. Triggers considered within sensory environments
3. Triggers considered within behavioral contexts
4. General considerations for people who work with or care for individuals with Fragile X Syndrome

Arousal Modulation and Hyperarousal

Arousal Modulation / Hyperarousal

“Arousal” refers to a general state of nervous system activation that is reflected in behavior, physiological activity and emotional experience.

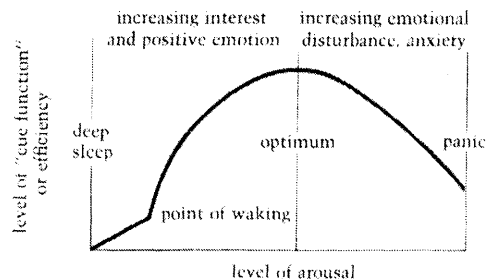
Arousal Modulation / Hyperarousal

There is an optimum level of arousal for the production of all well controlled behavior.

“Hypoarousal” refers to a state of low nervous system activation that interferes with the production of effective organized behavior.

“Hyperarousal” refers to a state of high nervous system activation that interferes with the production of effective organized behavior.

Inverted - U function of Arousal



Arousal Modulation / Hyperarousal

People with Fragile X Syndrome:

1. Have weak homeostatic arousal regulatory mechanisms, so they are less able than others to maintain an optimal arousal state.

Arousal Modulation / Hyperarousal

People with Fragile X Syndrome:

2. Are overly sensitive to many types of stimulation, which in the absence of effective regulation causes them to easily become hyperaroused.

Arousal Modulation / Hyperarousal

People with Fragile X Syndrome:

3. Remain affected by stimulation for a prolonged period after it has ended, which contributes to the duration and severity of their hyperaroused state.

Arousal Modulation / Hyperarousal

Hyperaroused individuals may experience:

- Difficulty focusing and sustaining attention on assigned tasks
- Difficulty performing tasks that require careful coordination and fine motor skill
- Impulsive behaviors
- Impaired speech and language

Hyperarousal Triggers Considered Within Specific Sensory Environments

Environmental Domains

- Physical environment
 - Visual, Auditory and Tactile stimulation
- Social environment
 - Eye contact and Personal space
- Temporal environment
 - Transitions and Unpredictability
- Personal environment
 - Pain, Anxiety and Sensory Integration Issues ->

The Physical Environment

Lighting

- Avoid fluorescent lighting when possible, in favor of incandescent lamps or natural sunlight.
- Insure that the lighting is not too bright, or directly in one's eyes.
- Avoid glare and reflections from shiny surfaces.

The Physical Environment

Acoustics

- Provide sound dampening over hard walls to reduce reverberation.
- Reduce the volume of radios or televisions.
- Insulate room from outside sounds such as hallway noises or loud machinery.
- Provide ear plugs or headphones.

The Social Environment

Eye Contact

- Avoid making direct eye contact
- Do not insist on mutual eye contact
- Do not confuse 'looking away' with 'not paying attention'
- Sit side-by-side, rather than facing one another
- Allow the wearing of sunglasses

The Social Environment

Other Stressors

- Crowded and busy environments
- Violations of personal space
- Teasing and bullying
- Rejection and isolation

The Temporal Environment

Transitions

Try to ease transitions by providing as much continuity as possible between activities:

- Avoid unnecessarily abrupt changes.
- Include familiar staff or peers.
- Walk the individual through a new environment.

The Temporal Environment

Transitions

Incorporate activities that include heavy work or exercise prior to transitions, especially transitions to challenging situations that involve unavoidable high stimulation.

The Temporal Environment

Unpredictability

Try to increase predictability by reviewing any upcoming changes in routine, e.g.:

- Scheduling of special events.
- Substitute or replacement staff.
- Holidays or vacations.

The Temporal Environment

Transitions & Unpredictability

Use photos and picture schedules to prepare individuals for upcoming activities and changes in routine.

The Personal Environment

- Pain due to injury or illness, such as a toothache, indigestion, etc
 - Consult a physician for treatment
- Anxiety due to fear, excitement or worry about a past or impending event
 - Consult a psychologist or psychiatrist
- Sensory Integration Dysfunction
 - Consult an occupational therapist

Sensory Diet

- One's sensory diet refers to type of sensory stimulation (visual, auditory, tactile, vestibular, oral-motor, etc.) that an individual experiences throughout the day.
- An appropriate sensory diet should be designed for each individual in consultation with an occupational therapist who is knowledgeable about Fragile X syndrome.

Sensory Diet

Exercise should be included in the life of all teens and adults with Fragile X syndrome.

- Walking, biking, playing sports, weight training
- The most calm and self-controlled adults with Fragile X have the most toned bodies because of their regular engagement in exercise.
- Exercise is also important for weight control

Hyperarousal Triggers Considered Within Specific Behavioral Contexts

Contexts for Hyperarousal

Hyperarousal can occur in various contexts:

- Residential Life
- Transportation
- Social Relationships
- Employment

Contexts for Hyperarousal Residence

Insure that the physical and sensory characteristics of his room are appropriate and not too stimulating.

Ask him if it is a comfortable space for him, and try to accommodate any reasonable changes.

Insure that his clothes & blankets are comfortable, and do not trigger any tactile sensitivities.

Contexts for Hyperarousal Residence

Try to find compatible roommates

- Someone with a calm personality

Try to accommodate food preferences

- Consider texture as well as taste

Assign appropriate chores

- Outdoor chores such as gardening
- Incorporate physical activity

Contexts for Hyperarousal Transportation

Transition to/from car or bus

Issues when riding in a car

- Seatbelts may be uncomfortable or aversive
- Back seat may be preferable

Issues when riding a bus

- Rules and regulation
- Co-passengers

Contexts for Hyperarousal Social Relationships

Individuals with Fragile X syndrome experience normal sexual development, but have difficulty demonstrating socially acceptable behaviors.

Close personal contact with a member of the opposite sex may trigger social anxiety, which can directly interfere with one's behavior, and cause embarrassment which further increases arousal.

Contexts for Hyperarousal Social Relationships

Hyperarousal may cause impulse control problems, which lead to inappropriate sexual contact.

- Use video and/or personal modeling instruction to teach appropriate social interactional behaviors.

Contexts for Hyperarousal Social Relationships

Hyperarousal may increase tactile sensitivity causing an aversion to touch, that interferes with appropriate social and sexual contact.

- Desensitize tactile defenses using occupational therapy strategies and calming procedures.

Contexts for Hyperarousal Social Relationships

Individuals with Fragile X syndrome have difficulty maintaining adult relationships, because they often become overwhelmed by the social demands and expectations.

- Provide support groups, and opportunities for supervised social interaction.
- Model appropriate social routines.

Contexts for Hyperarousal Employment

Consider jobs that appropriately match the characteristics of Fragile X Syndrome.

Avoid jobs that require:

- Fine motor skills
- Improvising on the spot
- Overstimulating environments

Contexts for Hyperarousal Employment

Examples of good matches include:

- Tasks that involve exercise, such as materials handling, pushing carts, stocking shelves.
- Outside work, such as farming, working with animals, landscaping.
- Pushing people in wheelchairs in a nursing home.

Contexts for Hyperarousal Employment

- Consider how timing may change an employment experience, for example, working in a store or stocking shelves during off hours.
- Consider the relationship between an individual and his boss and co-workers, and choose co-workers carefully.

General Considerations

for people who work with or care for individuals with fragile X syndrome

Behavioral Management Issues

Employers and caretakers should plan ahead to identify a quiet non-stimulating sheltered area where an aroused individual can go to calm down if he becomes hyperaroused.

Behavioral Management Issues

Employers and caretakers should:

- Be aware of the environmental and social factors that can lead to hyperarousal.
- Recognize the signs of hyperarousal.
- React appropriately when hyperarousal occurs.

Signs of Hyperarousal

- Hyperactive, impulsive, distractible
- Rocking, flapping, stereotypic behaviors
- Covering eyes or ears
- Flush ears or neck

Signs of Hyperarousal

- Rapid or repetitive speech
- Perseverative or tangential language
- Biting of hand or clothing
- Aggression towards self or others

Response to Hyperarousal

- Identify the stimulating trigger, and try to reduce the stimulation at its source.
- If necessary, remove the individual from the over-stimulating environment.

Response to Hyperarousal

- Apply Occupational Therapy procedures to promote calming and self-regulation.
- These include:
 - Deep pressure
 - Body brushing
 - Joint compression

Self-Response to Hyperarousal

Individuals with Fragile X syndrome should be taught to recognize the signs of hyperarousal in themselves, and how to respond appropriately when they become aware of their hyperaroused state, so that they are better able to cope with stimulating situations on their own.

Self-Response to Hyperarousal

Coping strategies an individual can use include:

- Asking to be excused from an arousing situation.
- Using headphones or sunglasses as needed.
- Invoking calming imagery.
- Adopting slow (square) breathing.
- Providing himself with muscular or oral-motor stimuli.

Self-Response to Hyperarousal

While it is appropriate for someone with Fragile X syndrome to take these measures to effectively avoid or mitigate the effects of hyperarousal, it is important that his supervisor and co-workers understand why he is acting that way, so that they do not misinterpret his behavior as lazy, arrogant or goofing off.

Consider Individual Differences

- Although there are many situations that commonly lead to hyperarousal, it is important to recognize personal idiosyncratic triggers.
- Such personal triggers may not be apparent, especially if they represent conditioned reminders of previous arousing experiences.

Functional Behavioral Assessment

In such cases, a Functional Behavioral Assessment (FBA) can help explain the factors that trigger hyperarousal reactions under certain conditions, and can guide your efforts to deal with context-specific behavior problems.

Functional Behavioral Assessment

An FBA is based upon three types of data, which may referred to as the ABC's:

- Antecedent -- the stimulus or stimuli to which the individual responds.
- Behavior -- the behavior that we see exhibited by the individual.
- Consequence -- the stimulus or stimuli that the individual receives (or that he is stopped being subjected to) as a result of his behavior.

Functional Behavioral Assessment

It is very important that the FBA be performed by a psychologist who understands Fragile X syndrome, and the causes and effects of hyperarousal, so that potentially arousing social and environmental stimuli are included in the behavioral analyses.

Skills Assessments Issues

- Skills assessments may be necessary in order to make decisions about vocational placement.
- Formal testing procedures include many triggers for hyperarousal.
- Such hyperarousal will interfere with one's performance, and almost certainly result in scores that underestimate his true capabilities.

Skills Assessments Issues

Every effort must be made to reduce arousal during skills assessments, for example by:

- Reducing unnecessary environmental stimulation
- Desensitizing to the assessment procedures
- Using occupational therapy calming techniques
- Employing covert assessment methods

Skills Assessments Issues

Covert methods are those that allow one to assess an individual without his knowledge.

- Observe his performance while he is working
- Consider peer or parent reports of his activities
- Incorporate the test into a game or activity

Individualized Service Plan (ISP)

One's ISP Should include:

- Relevant information about the causes and effects of hyperarousal.
- Any agreements regarding arousal related accommodations.
- A mandate that program officials and staff receive training on the relationship between hyperarousal and behavior.

In Conclusion

- In order to effectively work with and help individuals with Fragile X syndrome, the causes and effects of hyperarousal must be recognized and understood.
- Relatively simple accommodations to residence and workplace design and management can help to prevent or reduce hyperarousal, and the behavior problems that may be associated with it.