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

Sensory Processing and Integration Issues in Fragile X Syndrome

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Sensory Processing and Integration Issues in Individuals with Fragile X Syndrome

Introduction

Individuals with fragile X syndrome (FXS) frequently have sensory processing and sensory integration problems (Hagerman, 2002; Riley, 2011). Sensory based hyperarousal has been noted to be the most troubling and prevalent feature of FXS (Reiss, 2006). These problems are frequently described by parents and professionals, and recent research supports their inclusion within the phenotypic presentation of FXS. For example, children with FXS have enhanced sympathetic nervous system reactivity to social-emotional stimuli (Cohen, in press). Individuals with FXS have profound deficits in sensori-motor gating as measured in the auditory domain, which implies greater difficulty filtering auditory stimuli (Yuhas, 2010; Hessl, 2009; Frankland, 2004). Individuals with FXS have reduced habituation to sensory input (Miller, McIntosh, et al., 1999), and overall sensory based hyperarousal has long been reported in individuals with FXS (Belser and Sudhalter, 1995). Further, recent research advances suggest that sensory based processing issues characterized in animal models of FXS will increase understanding of the biology underlying FXS. For instance, Rotschafer and Razak reported that auditory hyper-excitability is a robust and reliable biomarker in Fmr1 KO mice (2014).

Sensory integration (SI) is a specific frame of reference used in occupational therapy and is based on the work of A. Jean Ayres., Ph.D., OTR (Ayres, 2005). Within the field of occupational therapy, the terms sensory integration and sensory processing are used to refer to the theory that describes a range of difficulties seen in many neurodevelopmental disorders, as well as the treatment approach that is suggested for use to remediate the difficulties. There are various therapists and theorists who may suggest somewhat different terminology to describe the difficulties and accompanying intervention methods, and use of terminology can vary by geographic location. Many professionals and parents may be familiar with the work of Ayres, who founded the field, or with important contributors such as Pat Wilbarger, Winnie Dunn and Lucy Miller. Thanks to the efforts of these women, among many others, the field is evolving in its descriptions of the sensory processing difficulties and the sensory integrative treatment approach. For the purposes of this consensus document, an overview which is most consistent with basic neuroscience, rather than with a particular theorist or method is provided. This is done to provide an overarching review of sensory-based issues that are seen in FXS specifically. Therefore, the terms sensory processing and sensory integration will be used interchangeably. Additionally, as SI is used within the context of occupational therapy practice, the term OT/SI will be used to refer to the intervention under consideration for this consensus document.

Sensory Integration: A natural occurrence in everyone

Sensory processing involves detecting sensory stimuli in the environment, processing this information, and integrating it into meaningful information, action and adaptation (Baranak et al. 2008). For most people, sensory integration occurs unconsciously. SI involves organizing

sensations from all of our senses, giving us the ability to hear, see, smell, touch and taste and also includes the hidden senses of movement and body awareness. The brain attempts to process these sensations in a way that will allow the person's nervous system to maintain a sense of order which is essential for daily functioning. The ability to do this appropriately is necessary for normal development. In fact, the idea of sensory integration is consistent with theories of the role that experience plays in brain development (Greenough, Black and Wallace, 1987). Think about how you maintain your balance when walking, especially if the surface is unfamiliar or uneven – your basic sense of body in space and in relation to the ground and to movement works with your balance (vestibular) system, eyes and spatial awareness to give you coordinated and responsive balance skill. This is one example of sensory integration. If you worked on your balance every day, this experience would quickly translate into a higher level of balance skill. But, more importantly, experiences as a young toddler learning to cruise, fall and get back up are essential for your sense of balance to develop so that you can take your first steps and venture into the world upright. Sensory integration through experience drives this learning and brain development.

Issues Surrounding Sensory Integration

In June 2012, the American Academy of Pediatrics published a policy statement on the use of Sensory Integration Treatment for children with developmental and behavioral disorders, which would apply to those with FXS (AAP, 2012). The policy statement stated that because there is no universally accepted diagnostic schema, sensory processing disorder should not be diagnosed. However, both the DSM5 and Zero to Three's *Diagnostic Classification of Mental Health and Developmental Disorders of Infancy and Early Childhood Revised* recognize that there exists sensory processing and regulatory differences and/or disorders. In fact, under DSM5's definition of autism, the presence of either sensory hypo- or hypersensititivites to the environment are included as a part of the disorder, so there is increasing understanding of the pivotal nature of sensory based difficulties in neurodevelopmental disorders. While currently there is not a recognized stand-alone "sensory processing disorder", children with Fragile X may be recognized to have sensory based difficulties that often impair functioning in school, with peers, at work, and with the family.

As with all AAP policies, the academy was conservative in endorsing occupational therapy as an acceptable form of treatment for sensory integrative difficulties within the context of a comprehensive treatment plan. They encouraged physicians to guide families to know of the limitations of research as well as the potential benefits of the treatment. As with any intervention, it is important to monitor progress toward goals and to evaluate the effectiveness of the intervention for each individual child (AAP, 2012). As the term "sensory" can be attached to many techniques or tools, it can be a source of confusion. This confusion we will try to clarify later in this document. Further, the American Occupational Therapy Association was in support of the AAP statement on Sensory Integration, though they did respond with concern for the lack of inclusion of current research on the efficacy of sensory integration. See response

statement from AOTA then-President, Florence Clark in a letter to the editor as well as a formal AOTA statement (Clark, 2012).

Before further discussion on OT/SI intervention for individuals with FXS, it is important to clarify in general what is included as authentic OT/SI and what is not. There are many strategies that may have a label of "sensory", and unfortunately, these masquerade as authentic therapy, though they do not meet the neural-based evidence for therapy as defined within the profession of OT, nor do they meet the criteria established by the recently devised Ayres Sensory Integrations (ASI) fidelity tool (Parham LD, Roley SS, May-Benson TA, Koomar J, Brett-Green B, Burke JP, Cohn ES, Mailloux Z, Miller LJ, Schaaf RC. , 2011). Many of these inauthentic therapies are passive in nature, non-individualized, and not evidence informed, such as, passive exposure to lights, sounds, motion, touch including the Snoezelen; attempts to reactively use a sensory "trick", such as a weighted vest or a fidget toy, to address complex behavior; use of lavender aromatherapy in a classroom for setting a calm environment, to name a few. These so-called "sensory strategies" in and of themselves will not create change for a person with FXS and are not endorsed within the scope of recommendations outlined in this consensus document. Rather, OT/SI is a relationship based intervention that employs specifically tailored and enhanced sensory and motor experiences, together with environmental supports and scaffolded, developmentally appropriate tasks that are targeted at a specified goal or outcome and facilitated by the therapist to evince an adaptive response. As Dr. Ayres stated, “It is the events between the sensation and the response wherein the story of sensory integration lies” (Ayres, 1972, p.11). This is what makes the intervention powerful and unique. The evidence base of neuroplasticity parallels this process, wherein change is a product of experience that produces adaptation (Stackhouse, 2014). This is more complex in humans than in animals, and requires a therapeutic process to obtain when neurodevelopment is not happening neurotypically.

For example, if a child with FXS is struggling with hyperarousal, an OT/SI approach would include identifying the specific target of adaptation, which would be a more calm, organized arousal state, and then, selecting from the core elements of treatment (sensory, task, environment, predictability, self-regulation and relational interaction) to promote the changes required to meet the goal, or, the highest level of adaptive response possible. The therapist then scaffolds the elements into both direct treatment as well as carry-over programming for school and home, so that the adaptation is extended into daily life. Perhaps the intervention could include a weighted back pack, a balance practice program, or a chewable oral support, but, these would not be used indiscriminately, rather, incorporated within the framework of the core elements of treatment as noted above. Importantly OT/SI is aimed to promote optimal performance and outcomes for children and families with FXS and is often a primary referral from physicians and source of ongoing support for these families.

Review of the Evidence on Sensory Integration Interventions

There have been no controlled studies of the effects of any sensory integration (SI) interventions in FXS. Even in autism spectrum disorders (ASD), the lack of research on therapies to address this issue is notable. Wong et al in 2013 published a manual on therapies used in ASD, and determined that there was insufficient evidence for sensory diet or sensory integration modalities, in general, to include in their review of the literature of what are evidence-based therapies for ASD (Wong, 2013). Despite this, Autism Speaks includes OT/SI as one of the recommended interventions for individuals with ASD. Baranek performed a systematic review of sensory and motor intervention studies for children with autism and concluded that most categories of these interventions had mixed results (Baranek, 2002). For OT/SI-classical and other sensory-based approaches, the studies were very small, usually single-subject designs, with varying age ranges. In FXS specifically, Stackhouse and Scharfenaker (2006) presented single-subject case studies from two toddlers with FXS, suggesting positive goal attainment outcomes in a therapy intensive model that utilized an OT/SI approach which was customized for diagnostic specificity incorporating key features of the FXS phenotype. Below, we review the controlled studies that have examined the delivery of OT/SI as the intervention. Although there have been studies on single techniques, such as auditory integration training and ambient prism lenses, the use of a comprehensive SI approach based on solid principles in a manualized protocol and randomized controlled trial design are preferred and only possible now that a fidelity model has been established.

Perhaps the first controlled study to examine an OT/SI program intervention was a study conducted in Turkey using the Sensory Evaluation Form for Children with Autism to evaluate the differences in scores on this checklist from baseline to end of study (Fazlioğlu, 2008). Children with autism were separated into two groups each comprising 15 children, with the authors reporting significant differences in favor of the sensory integration therapy program intervention compared to a control group (abstract only).

In the first randomized controlled trial using an approach of modeling Ayres principles of sensory integration techniques, researchers enrolled children with ASD (autism and PDD-NOS) – 20 in the OT/SI intervention group and 17 in a comparison group that received a fine motor (FM) intervention (Pfeiffer, 2011). Both the evaluators and the parents were blinded to intervention status, and goal attainment scales (GAS; these scales tailor measurement of a procedure(s) to goals identified by the parent) were used as the primary outcome measures. Both groups demonstrated significant improvement in the GAS, although the OT/SI group did better than the FM group in terms of sensory processing, motor skills, and social functioning goals rated by parents ($p<0.05$) and teachers ($p<0.01$).

A randomized controlled trial was published following the Wong review that may give the best evidence to date for a manualized intervention adhering to the Ayres principles of sensory integration techniques and using GAS as the primary outcome (Schaaf, 2013). This trial included 17 children in the treatment group and 15 children in the usual care comparison group that were between 4 and 8 years old with ASD by ADIR (Autism Diagnostic Interview-Revised) and

ADOS (Autism Diagnostic Observation Schedule). The treatment group achieved significantly higher scores on the GAS (p=0.003), and on Self-Care Caregiver Assistance (p=0.008) and Social Function Caregiver Assistance (p=0.04) subtests, compared to the usual care group. Although evaluators were blind to the intervention status, parents were not, which may bias the reporting of outcomes by parents.

Neither of the trials used the standard intention-to-treat design, which necessitates including measurements on children who did not complete the programs as specified. One of the major challenges in research of the efficacy of SI procedures, are the myriad therapies that don't lend themselves well to treatment fidelity and/or replication. Using a manualized approach as above will be essential to assure that what is being measured is what has been defined as the intervention. Using a homogeneous group of children with SI problems will be important if small studies continue to be conducted. Due to the small size of the study, Schaaf et al could not use their stratification schema designed to remove the effects of cognitive function and severity (Schaaf, 2013). Study samples should be homogeneous in terms of their FXS phenotype. Co-factors that may introduce heterogeneity are autism status, severity of the FXS, cognitive or intellectual function, and the presenting SI problem(s), to name a few. Much more rigorous research is needed to demonstrate efficacy of SI therapies, that is, whether they truly work in the ideal situation, before effectiveness can be established in the real world and having a demonstrable impact on the burden of SI disorders.

Of note, there is little direct intervention research for individuals with FXS in a number of fields. For example, there is no more evidence-based research supporting treatment with SSRIs for FXS than OT/SI but we know this is quite accepted as a reasonable treatment. The limitation of studies in OT/SI only verifies that much research in many areas is needed.

Types of Sensory Integration Problems

Virtually all of the sensory systems can be impacted by sensory integration problems (Martin, 2012). There are two primary categories of sensory-based problems associated with the anatomical division of the streams of processing that take sensory information from the environment and body-based receptors into the brain. These pathways include two primary systems 1) sensory discrimination and 2) sensory modulation

- Sensory discrimination problems lead to poor processing of sensory/motor information for skilled response – this type of sensory integration problem results in poor coordination, known as motor planning problems or dyspraxia.

- Sensory modulation refers to the manner in which sensory information is responded to and utilized for fundamental brain functions such as arousal, alertness, attention, organization, coping/adaptation and self-regulation.

Individuals with FXS are likely to exhibit both of these types of sensory integration difficulties (Stackhouse, 1998), and these difficulties will impact learning and skill development (including

impacts on domains that are not solely sensory/motor – such as language, attention, and problem solving) – and also impact behavioral adaption.

Sensory Discrimination Difficulties

Sensory discrimination is the ability to distinguish between various types of sensory input, assign meaning to them, and make use of the information for specific skill production. Sensory discrimination problems lead to poor processing of sensory/motor information for skilled response so individuals frequently exhibit poor recognition and interpretation of sensory stimuli. These problems also result in poor awareness of the differences or similarities between sensory stimuli. As a result of sensory discrimination difficulties, individuals with FXS may struggle with refining sensory motor skill development in gross, fine, visual and oral motor skills. Additionally, several decades of research show that sensory discrimination abilities underlie fundamental motor planning circuitry resulting in various forms of dyspraxia, or motor planning deficits (Ayres, 1989 ; Mulligan, 1998). Motor planning, or praxis, is the ability to plan, sequence and execute novel or unfamiliar actions. Dyspraxia refers to a breakdown in this process, which can affect one of all of the areas in motor planning. Individuals with FXS may struggle with motor planning of gross, fine, visual and oral motor skills. Several studies have found motor impairments to be present in individuals with FXS, and motor issues may be related more to cognitive ability than to autism status (Zingerevich C, Greiss-Hess L, Lemons-Chitwood K, Harris SW, Hessl D, Cook K, Hagerman RJ. (2009)). At least 80% of children with FXS have motor functioning that is severely impaired (Friefeld, S. J. and Macgregor, D., 1994).

- Some examples of common sensory discrimination difficulties are:

Difficulties with sensory discrimination within the tactile system may impact Oral-motor skills and as a result, a child with FXS may:

1. Overstuff the mouth; have poor oral control
2. Have trouble brushing teeth or tolerating foreign objects in the mouth
3. Be a picky eater – many child seem to prefer foods with hard textures versus soft or mushy textures

Difficulty with proprioception or vestibular functioning may result in:

1. Trouble climbing stairs or climbing on outdoor toys
2. Trouble learning how to ride a bike
3. Trouble tolerating elevators and escalators
4. Difficulty with balance, reaching out and squat to stand
5. Seeking increased amounts of pressure input, physical contact or physical activity
6. Problem with using too much or too little force

Auditory System

1. Difficulty following directions
2. Difficulty distinguishing between similar sounds
3. Talking too loudly or too softly

Fine Motor

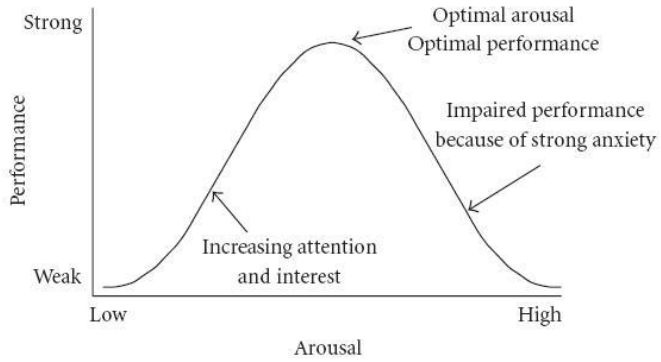
1. Trouble holding a pencil correctly
2. Trouble performing tasks that involve manipulation of small objects
3. Trouble tying shoes or with fasteners and opening things

Sensory Modulation Difficulties

Sensory modulation refers to the manner in which sensory information is responded to and utilized for fundamental brain functions such as arousal, alertness, attention, organization, coping/adaptation and self-regulation. As a result of sensory modulation difficulties, individuals with FXS may struggle with sensory hyper-responding, over activity, poor attention, and poor coping.

Within the sensory modulation domain, hyperarousal is the most prevalent and troubling problem seen in individuals with FXS (Hagerman, 2002; Reiss, 2008; Stackhouse, 2002). Individuals with FXS tend to become hyperaroused by normal or excessive sensory information. This leads to a tendency to avoid stimulation and to become upset or anxious. Due to associated difficulties with self-regulation this leads to poor adaptability and coping. Many of the challenging behaviors seen in individuals with FXS, including selective mutism, fears and anxiety, withdrawal, perseveration and aggression can be related to hyperarousal (Belser & Sudhalter, 1995).

It is important for professionals and parents working with or raising a child with FXS to understand some of the mechanisms involved in hyperarousal in order to understand how to manage it. Arousal in the brain is well studied and a model for arousal is based on the Yerkes-Dodson law which has established an empirical relationship between arousal state and performance (Yerkes & Dodson, 1908). The relationship demonstrates that in low or high states of arousal, performance is poor with optimal functioning happening in the middle arousal range.



(From Hebb, 1959 as referenced in Diamond, et al, 2007)

This inverted U model establishes one of the pressing issues for individuals with FXS, that when in a hyperaroused state, performance is minimized.

In a state of hyperarousal, the brain is impacted to adjust to the state of (dis)stress that it is in. Often this is associated with an increase in autonomic nervous system activity and in FXS, typically heightened sympathetic arousal (or fear, fight, flight responding). Additionally, when hyperarousal occurs, the brain becomes more narrow in what it processes and tends to shift toward more protective ways of operating.

Hyperarousal produces a lower threshold for responding to sensory and emotional stimulation. With a lower threshold, the system is vulnerable to negative bias in sensory and emotion processing. In individuals, this results in more sensory hyper-sensitivity, which is called sensory defensiveness. It can also produce more negative emotionality. The arousal system also influences attention systems. As the person becomes more hyperaroused, they become more vigilant, distractible and hyperactive. Therefore, inattention and anxiety result.

There is great variation in the ability to learn and utilize skills in individuals with FXS. This variation is in part accounted for by frequent shifts into a hyperaroused state. Importantly, if a person is in a very heightened state of arousal, the brain has a protective mechanism that shifts the person into a more calm state. This can make the individual appear to be in a low state of arousal although they are recovering from hyperarousal and not really relaxed. For the treating occupational therapist, it is important to observe heart rate, respiration and autonomic stress response cues to help determine the state the person is in and to provide the appropriate treatment plan.

- Some examples of common <u>sensory modulation difficulties</u>

  Visual System
  1. Difficulty tolerating bright lights
  2. Difficulty making good eye contact when looking at people
  3. Difficulty looking at written materials for long periods of time

  Auditory System
  1. Difficulty tolerating loud noises such as the vacuum cleaner
  2. Difficulty tolerating noisy places such as the mall, a restaurant or a movie theater

  Touch
  1. Trouble tolerating certain clothes
  2. Trouble going barefoot
  3. Difficulty tolerating light touch
  4. Difficulty working with writing utensils or art materials such as finger paint
  5. Not seeming to notice wet or soiled diapers (hyporesponsiveness)
  6. May be a picky eater, with aversion to temperatures and texture changes
  7. Difficulty tolerate touch or feeling of grooming activities

The sensory integration and sensory processing issues in those with FXS typically are a significant contributor to delayed skill acquisition and to challenging behavior. The intervention

plan will often be broad, including OT utilizing a sensory integrative approach along with general OT clinical practice, as well as coordination of care with medical and educational providers, for a comprehensive approach.

Consideration of Clinical Reasoning to Guide Sensory Integrative Practice

As can be seen in the symptom lists, sometimes there is overlap between sensory discrimination and sensory modulation issues (such as with picky eating). While the pathways are dual and separate to and from the brain, their functions are interrelated. Why is this important to point out? For a general level of understanding, sensory based concerns are often categorized into one large construct called "sensory integration" or "sensory processing". However, for a therapist utilizing this approach, a higher level of neurologically based clinical reasoning skill is required to understand, guide and provide appropriate treatment. The OT must be able to sort out the "what" and "why" of the underlying processing in order to provide comprehensive, efficacious intervention. So, for physicians, parents and collaborating professionals, it is important to find an OT who has the ability to make a discerning and clear assessment of the sensory based issues with specificity of treatment approach reflecting careful clinical reasoning.

Importance of Diagnostically Specific Intervention

The FXCRC members have long been proponents of ensuring diagnostically specific intervention approaches for individuals with FXS. As the phenotype of FXS is well specified, it suggests that utilizing the core features of FXS to modify, augment, or enhance interventions available for therapy, special education, behavior intervention, etc., will likely make said interventions more efficacious for those with FXS. As such, in 2006, Stackhouse and Scharfenaker outlined, for case study purposes, how to create diagnostically specific adjustments. This included: knowledge of the phenotype of FXS should guide treatment; a developmental presentation of FXS is emerging in literature, assisting in making intervention FXS specific developmental appropriateness; from phenotypic and developmental information, anticipate core issues and address each from best practice within each field; individually assess and determine goals for each child through play based assessment (as suggested by Scharfenaker, Riley, Stackhouse, Coleman, and Engleman, 2004). Consequently, for individuals with FXS, diagnostically specific modifications are suggested for most interventions, including OT/SI.

Who Do I Go to for Help?

A team approach to the varied issues related to SI and treatment for individuals with FXS is always recommended. Team members typically include, but are not limited to, physicians, psychologists, occupational therapists, speech therapists, physical therapists, and educators. Parents are pivotal members of the team and contribute to the assessment and ongoing treatment of sensory related issues (as well as all other developmental concerns). The intervention, school and medical teams should work collaboratively with the parents to ensure communication, carry over and that necessary support is provided.

Physicians may provide medications that can be helpful for related problems such as ADHD and anxiety. They often refer individuals for necessary services, including occupational, speech, and physical therapy.

Assessment

Traditionally, occupational therapist(s) (OT) do the most work in this field and can perform assessments, create treatment plans and provide intervention. Occupational therapy is a profession that helps people gain, develop, and build skills that are essential for independent functioning, health, and well-being. The primary role of the occupational therapist in pediatrics is to help children play, grow, and develop many of the skills that will enable them to enjoy a satisfying adult life (Case-Smith, Allen, and Pratt, 2001). The OT should assess both sensory discrimination and sensory modulation functions and outcomes. The Sensory Profile (Dunn, 1999) and the Sensory Processing Measure (Parham & Ecker; Kuhanek, Henry and Glennon, 2007) are questionnaire tools available to assess individuals for this condition. Additionally, Tartaglia and Stackhouse have recently found that the most valid and reliable neuromotor assessments for use with individuals with FXS include the Movement Assessment Battery for Children, the Quick Neurological Screening Test and the Berg Balance Scale (Tartaglia, Stackhouse, Cordeiro and McGrew, 2011).

Physical therapists may assess problems that affect gross motor development and muscle tone concerns, such as difficulty with balance, climbing or riding a bicycle. They may also provide orthotics for pronated feet.

Speech therapists may assess oral-motor issues. They will be important collaborators in looking at how language is impacted by basic sensory and motor processes. Co-treatments or other collaborative intervention models are often the optimal treatment for individuals with FXS.

Educators may assess problems with handwriting or other fine motor skills that are academic in nature, such as cutting and pasting or use of a ruler. Additionally, educators will have keen insights into the level of restlessness, anxiety, fidgeting, impulsivity, and distractibility in the child. Additionally, how the individual copes with and adapts to the demands of people and the environment is often best seen in a school setting.

Psychologists may assess cognitive and emotional skills as well as coping and self-regulation. Coordinated treatment planning is often required due to the overlap between domains of functioning and intervention.

Physicians are instrumental in assessing the medical aspects presented by the individual with FXS. Often, there may be medications available to address some of the issues, such as anxiety or attention based difficulties. While there is not currently a medication that will remediate the sensory integrative difficulties, other symptoms can be supported with medication management. Sleep and digestive issues may co-occur within SI related difficulties in FXS and

management requires a team approach, starting with the physician. A team approach to the varied issues related to sensory integration is always recommended.

Comorbidity and Confounding Issues

Sensory processing/integration issues can potentially restrict attentional, social and cognitive development and lead to behavior problems such as anxiety and outbursts. Much like executive functioning difficulties, sensory integration problems tend to occur in many conditions and the influence on other capacities is evident, even when an additional formal diagnosis is not made. Children with sensory processing problems may resist participating in normal activities that are necessary for development. There is some evidence that this problem becomes more evident with age (Baranek , et al, 2008) and is present in both males and females with FXS.

Some of these problems may be difficult to distinguish from other common problems in FXS such as hyperactivity and anxiety. Additionally, difficulties with motor planning and imitation are associated with the presentation of autism within FXS. As many of the SI based difficulties may contribute to common behavior or associated diagnoses seen in those with FXS, it is important for an OT to be involved in clinical diagnostic and treatment plan decisions for individuals with FXS.

Recommendations for treatment

1. Occupational therapy is often one of the main interventions for children with FXS. These therapists should have some training in the field of sensory integration with knowledge of the brain processing involved in these disorders.
2. Children with FXS should receive routine assessments from occupational therapists.
3. When possible, children with FXS should receive OT one to two times per week during early development, as early intervention is indicated. As children grow into adults, OT can be helpful at times of difficulty, to shore up skills and provides necessary supports and accommodations. Also, when an individual is experiencing a burst in skill acquisition, this can be bolstered by additional therapy. During more “status quo” periods a treatment hiatus may be indicated. The OT, family and physician should work together to determine if the child can benefit from therapy.
4. School occupational therapists may provide beneficial treatment for children but may not be mandated to address all areas of concern since all of these areas are not considered to be educationally related. Parents should consider providing their children with private therapy when the school services are limited.

Traditional Therapies

Occupational therapists may utilize clinic based sensory integration treatment with the adjunct of specific sensory-based strategies to support the child’s functioning. However, it is vital that these strategies are embedded in a multi-faceted occupational therapy treatment plan and not

offered in isolation. For example, a weighted vest or noise reduction head phones may be suggested, but are only tools and not the full treatment. Sensory integration treatment has recently made important advances with the establishment of a fidelity to treatment model that should foster more research into its efficacy (Parham, Roley , et al, 2011). Regardless, Sensory integration treatment is offered in the context of a broader occupational therapy treatment plan. As Case-Smith and Schaff (2012) point out, sensory-based interventions are: (a) based on a thorough assessment; (b) individualized in accord with the child's sensory modulation and sensory discrimination problems; (c) monitored closely to gauge the child's fluctuating responsivity then adapted or adjusted accordingly; and (d) periodically evaluated for their effects on the child's participation in play, school, and home environments. Finally, a key aim is to enable the family and the child to recognize when the strategy is or is not needed in everyday life.

Sensory Integration therapy is offered within the context of occupational therapy. It does not "sit alone". The OT should provide outreach to home and school so that the sensory based strategies learned in the clinic are consistently implemented across the child's day. They should work with other providers to ensure consistency and coordination of care. Additionally, OT's working with children with FXS should include routines and language based supports in addition to the sensory based approaches that match the child's needs. Stackhouse and Wilbarger (1998) have formulated a clinical reasoning model, the S.T.E.P.S.I. approach, which is an acronym for the components of a well formed treatment plan – this acronym stands for Sensation, Task, Environment, Predictability, Self-Regulation and Interaction. These components should be included in clinic based treatment as well as in outreach programming. Some examples include the use of visual schedules and work processes as Task, Environment and Predictability supports. Working on co-regulation (interaction) and self-regulation are pivotal skills for individuals with FXS and should be an active, ongoing part of the treatment.

If a child has difficulty with sensory modulation, an OTR can help create an intervention plan which may include a "sensory diet" – a structured sensory motor intervention plan based on individual needs - to guide him or her through activities that cultivate the ability for successful, organized responses to sensory input through the daily routine. A sensory diet is a component of an overall proactive intervention plan that might include other supports such as increasing organizational supports and routines, visual supports, and modifying interactions and the environment. Sensory diets often include the Wilbarger Protocol (often erroneously referred to as "brushing") and auditory training programs such as Therapeutic Listening.

Within occupational therapy, specific focus on skill development is included. This may include fine motor skill development, such as for handwriting or washing dishes. It may also include learning to dress, to use a vacuum or to complete a work process. It could also include learning how to wait, keep your body still, or cope in the face of frustration. Occupational Therapists devise treatment plans for both the underlying components (muscle skill, sensory processing, social or cognitive function) as well as for the occupational outcomes (such as being an independent worker, a friend, a student, or a family member) (AOTA Practice Act, 2002).

Some occupational therapists also use some non- traditional therapies

Protocolized sensory interventions, such as the Wilbarger Protocol; Astronaut Vestibular Activation Training; Auditory therapies that utilize specialized auditory inputs to make a child less sensitive to noise or to enhance auditory processing, Feldenkrais and cranial-sacral therapy are all examples that parents often mention as being accessed for their children with FXS.

Research is still extremely limited in FXS for most interventions in general, and for non-traditional therapies there is only anecdotal information about the potential benefits of these approaches.

Additional Therapies

1. Medications may be used to help with related inattention or hyperactivity, anxiety, autonomic symptoms, and aggression.
2. CBT (Cognitive Behavioral Therapy) – is often suggested for individuals with anxiety disorders. This treatment is efficacious for children and adults with typical cognitive and language skills. Clinicians experienced in working with individuals with cognitive impairments may be able to adapt this intervention for a select group of those affected by fragile X, although the treatment results may be constrained by the cognitive and developmental level of the patient.
3. Behavioral therapy and intervention are not designed to treat sensory integrative and sensory processing problems directly. Sometimes, the behaviors that result as a part of the complexities of fragile X, including the sensory based issues, may require a behavioral approach. The OT and the behavior therapist should collaborate to explore why a child might be having behavior problems and help the parents or school develop strategies to improve the situation. It should be noted that a directed behavioral intervention cannot improve the underlying motor planning issue, as practice and reward alone do not treat this type of sensory integrative problem. Likewise, hyperarousal can often confound directed behavioral intervention.
4. Non-traditional therapies such as cranial-sacral therapy and hippotherapy are becoming more common. Leisure pursuits such as yoga, dance, martial arts, and other sports are often utilized as well. Current evidence is not available for these less common and complimentary interventions. However, anecdotal case reports may suggest an emerging role for these and other treatments when matched to the individual needs assessment.

Home Life

Therapies should be carried over from home to school to therapy and this should be facilitated by all team members. Families often require support to ensure routines are established to best support functioning related to the complexities of raising a child with fragile X, which includes specific supports related to the sensory integration and sensory processing concerns.

Occupational therapists are an excellent resource for assessing, suggesting and assisting to implement home-based programs, accommodations and modifications.

When possible, parents should be proactive in preparing their children for daily challenges. Learning to live within the bounds of sensory integrative and sensory processing issues may mean limiting exposure to too much intensity until a child can manage these situations. A goal of OT intervention will be to move the family from restricted or limited ways of living, to a more fully realized experience. Rather than avoiding input, it is imperative to obtain the right tools and methods to approach and "live life to the fullest". This is the "tag line" for the profession of occupational therapy.

Common Questions and Answers

1. **Do we always recommend occupational therapy?**
   We recommend occupational therapy if we suspect fine motor delays or the different types of sensory integration/processing problems reviewed above. Sometimes we recommend therapy before the school does because we know that these tendencies exist in fragile X and we would like the child to receive intervention as early as possible. We recommend OT across the lifespan, since children and adults may benefit.

2. **If a child is receiving occupational therapy for 30 minutes per week or less through the school or early intervention system, would we try to obtain more therapy for the child from a private OT?**
   Parents may consider whether private OT would benefit their child.

3. **Do schools address sensory processing issues?**
   Some schools might but the focus may be largely on fine motor problems and not necessarily on sensory integration problems.

4. **Will my child grow out of hyperarousal?**
   Hyperarousal, as the most prevalent and defining characteristic in fragile X is a life-long challenge. Learning to proactively manage hyperarousal allows the person to grow into themselves, not out of the problem.

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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)*