



DEVELOPMENTAL FX
The Developmental &
Fragile X Resource Centre
A NOT FOR PROFIT ORGANIZATION

(This information was compiled by Wendy Bowler of the UK Fragile X Society after a lecture by Tracy Murnan Stackhouse, MA, and Sarah K Scharfenaker, MA, CCC-SLP, Fall 2012, Birmingham, England)

IT TAKES A VILLAGE

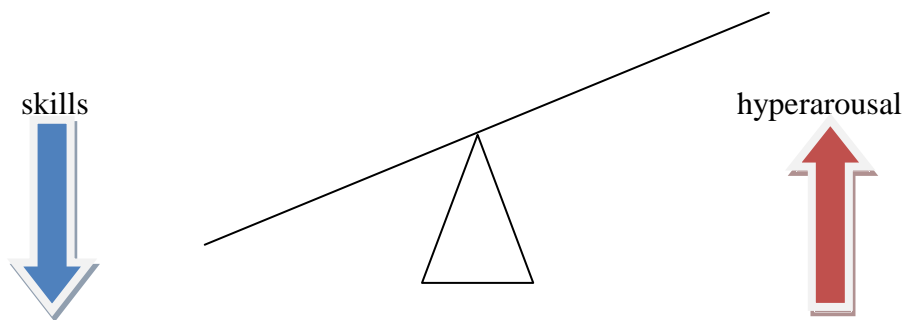
Hyperarousal in Fragile X: What it is and how the unique learning style in fragile X affects it

What is hyperarousal?

Before we think about what hyperarousal is, let's first consider arousal. A certain level of arousal is necessary to perform any task – when you wake up is not the time to be doing high power brain work – your arousal system needs to get going. All individuals will operate best around the middle levels of arousal – alert, but not in a complete state of anxiety. Achieving this middle state of arousal is very difficult for those with fragile X – their base line is much higher to start off with and this is exacerbated by the many events during any one day that will fuel their arousal levels and send them through the roof to the state of hyperarousal.

Fragile X is full of complexities. Children and adults with fragile X have a lot to deal with and often find themselves in a state of hyperarousal or high anxiety. At such times meaningful interaction or engagements is just not possible. They just cannot manage what is going on around them and this can result in complex behaviours or behaviours that “challenge” the rest of us. What we need to do is minimise the stressors that lead to this hyperarousal and thus help individuals with fragile X maximise their potential. The stressors are well known to many families – being overwhelmed and anxious in unfamiliar places, coping with change and transitions etc etc. The behaviours that result from these stressors are not manipulative or volitional, but a direct result of the cognitive and language deficits in those with fragile X.

Hyperarousal is probably the most difficult part of fragile X. The most difficult behaviours result from hyperarousal – behaviours like what appears to be aggression, self harm, high levels of impulsivity. Hyperarousal impacts on everything - as hyperarousal goes up skill levels go down, a bit like a see-saw. Therefore it is something that we need to do something about



On the positive side, it is a field in which the clinical and research world are working well together.

So what does hyperarousal feel like? An intense feeling of anxiety, nervousness. There are physiological symptoms.

1. The body senses something that may be potentially threatening. For someone with fragile X this could be a change or a transition (events that occur all the time in daily life).
2. The brain reacts and there is a stress response – commonly fright, flight, fight.
3. The body prepares – blood flows to the muscles, breathing becomes rapid, heart rate increases, blood pressure goes up, there are changes in metabolism with glucose entering the system.
4. The body realises something is going on which leads to anxious thoughts.
5. Complex behaviours result with learning levels dropping as individuals try to self calm.

People with fragile X are wired for hyperarousal – their arousal levels are naturally higher

- They have a lower threshold for what causes hyperarousal, which means that their system reacts faster.
- When they have got to the top or the spike of those intense feelings of anxiety, it takes them much longer than a typically developing individual to get back to the base line and a feeling of calmness.

These two things together make a lethal combination.

Think what hyperarousal is like for you – when you have to go somewhere new, do something new, work on something you find really difficult. Imagine feeling like that all the time – people with fragile X are very brave!! The difficult behaviours in fragile X that result from hyperarousal are the individual's attempts to get back to organisation, to get away from feeling overwhelmed. To understand and work to the unique learning styles of individuals with fragile X will help to minimise their hyperarousal levels and maximise their ability to learn.

Learning Styles of Those with Fragile X

When working with children and adults with fragile X, it is important that we adapt to their learning styles. They are simultaneous learners i.e. they see things as a whole and not in parts. They are visual learners and good at incidental learning. They like to have a clear understanding of the beginning and end of a task. They have a strong memory for routines, are good at imitating verbal and social skills and have a developed sense of humour. We know a lot about the learning styles of those with fragile X and we need to use this knowledge to maximise learning and minimise hyperarousal

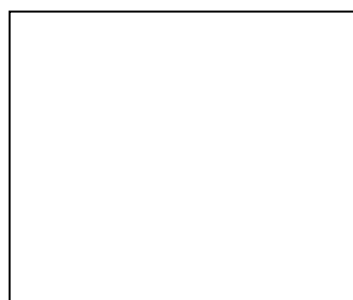
So what works well?

- Showing them the finished product and then giving them all the materials to make the product e.g.

Make the card



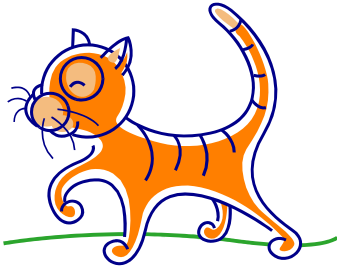
Use



MERRY CHRISTMAS



- The whole word approach is better for teaching children to read e.g.



cat

rather than the phonic approach of c - a - t.

- Visual schedules that let a person know what is expected of them or what is going to happen

 <p>trains</p>	 <p>painting</p>	 <p>cars</p>	 <p>puzzle</p>
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 <p>Bus to town</p>	 <p>Shopping</p>	 <p>Coffee & cake</p>	 <p>Bus home</p>
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- Children will often copy what other children in the class are doing. Imitation skills can be used to teach lots of things.
- They like a well defined beginning and end to activities.
- They do better when their areas of interest are used e.g. counting trains, cars.

What can the abilities of more able individuals with fragile X tell us about the whole spectrum?

The abilities of higher functioning girls (and boys) with fragile X gives us an insight into the strengths and weaknesses of those with fragile X, whatever their degree of learning disability. What are the nuances and characteristics we can make use of?

- Language comprehension can be a relative strength.
- They learn visual systems in the classroom – where things are kept etc. Visual sameness will enhance learning.
- In girls particularly the shyness is very striking.
- The Maths deficits apparent in many girls can also be found in boys.
- Difficulties with spelling and conversation can be linked to problems holding onto all the information and bringing it out in the right order

Tracy and Mouse gave the example of how they had worked with a young girl, who had selective mutism, only speaking to her family in the home. She did have visual strengths and a good memory and Tracy and Mouse were using this strength to get her to communicate using cards with social bids on e.g. “What did you do last night?” To help the young girl feel more confident and not the centre of attention, Tracy and Mouse adopted a technique, which shifted the focus of attention to them. They started chatting to each other and painting their nails at the same time. Gradually the young girl felt able to join the conversation.

Autism and Fragile X

Fragile X does have overlaps with autism. Many interventions/strategies documented in autism literature are helpful when working with individuals with fragile X, but they may need changing a bit.

To qualify for a diagnosis of autism, an individual must show

1. A qualitative impairment in communication
2. A qualitative impairment in making social relationships
3. Restricted interests/repetitive behaviours.

Lots of individuals with fragile X will meet criteria 1 and 3, but not 2. Individuals with fragile X can have a dual diagnosis of autism (about 20-30% of children with fragile X have a dual diagnosis of autism and about 4-6% of children with autism have fragile X). The key difference between the two disorders is that people with fragile X have a social drive; they like to engage with people, though they may avoid people who are too “full on.” They may well also very deliberately avoid eye contact, possibly turning right away. This is not a sign of social indifference and is best managed if the social partner averts their own gaze when approached by an individual with fragile X.

Theory of Mind is also very different across the two disorders – those with fragile X do have an understanding of the feelings and emotions of others. They are also good mimics. Children with fragile X who do not mimic are more likely to have a dual diagnosis of autism and those who do copy aren't.

Anxiety and Fragile X

Anxiety is a key feature of fragile X. Bailey et al 2008 study showed that 70% of males and 50% of females had been clinically treated for anxiety.

The key features of anxiety from a fragile X point of view include

- Social avoidance
- Gaze aversion
- Frequent questions
- Self harming
- Stereo-type behaviours

Some individuals may have a dual diagnosis of anxiety issues alongside fragile X and some may need specific treatments to manage this. In males it can lead to specific phobias like only being in one room in the house, only eating certain foods. In females also there can be high rates of anxiety problems.

Sensory Defensiveness and Fragile X

Many individuals with fragile X have difficulties managing the wealth of information coming to them via their senses. Again their reaction is related to the biology of the sensory system – something out of their control. It is difficult for us to imagine what it feels like to have these difficulties, but Tracy and Mouse organised a really good activity, which I shall try to explain. We were organised in groups of 3. One person was the child, another the person issuing instructions/asking questions and the third the environment. The instruction giver had to keep shouting instructions and asking questions of the child while the person who was the environment had to keep getting close to them, touching, waving in their face etc. At the end the people representing the child had to share their feelings. To a man they found the experience intensely difficult. I was the child in our group and after about half a minute I could not even remember what I had eaten for dinner the previous night.

In the next two sessions Tracy and Mouse talked about ways of working with children to help minimise hyperarousal levels. We will cover these in the next two newsletters.

Calming, Coping and Comforting. Self Regulation and Sensory Based Strategies to Manage Hyperarousal

Thinking back to the article in the last newsletter based on Mouse and Tracy's talk at our conference in September, managing hyperarousal in individuals with fragile X is key to helping them to learn and function. This is because hyperarousal is a disorganised state, leading to diminished language, social and motor skills and sometimes challenging behaviours. It is therefore important to think about what pro-active strategies will minimise hyperarousal and thus help an individual to manage known difficult situations.

How can hyperarousal be managed?

The goal of any intervention is to create a calm, alert, adaptive state. Hyperarousal can be managed in two main ways – via medication and via pro-active strategies in day to day life. A medical practitioner will obviously need to be involved with the prescription of medication, but this article will be looking more at the daily living strategies. These are strategies that help the person manage their responses better so things don't go over the top and get out of hand. They fall into 3 main categories. This article will look at sensory based strategies and next month will cover Tracey and Mouse's last session on routine and language based strategies.

1. Sensory based strategies
2. Routine based strategies
3. Language based strategies

How do we know that an individual is becoming hyperaroused?

General signs include an initial reddening of the ears and face, increased perseveration of language, gaze avoidance and finally gradually moving to shut down.

Stress signs can be subtle, not necessarily linked to the ABC model. The ABC model involves looking at behaviours in terms of what happened before the behaviour (A – antecedent), the behaviour itself (B – behaviour) and what happened after the behaviour (C – consequence). In those with fragile X, what happened before the behaviour is not necessarily the trigger. It is often the case that there are lots of little triggers, each one pushing the anxiety or arousal levels up a bit until the person can bear it no longer. This is a bit like drips of water falling into the cup until it overflows.

Key points to managing hyperarousal in an individual with fragile X

It is helpful to stop whatever the activity is as soon as you see signs of hyperarousal. The next stage is to help the person to recover and the final stage can be to move back to the activity. But how do we do that?

1. Stop what you are doing or what the activity is. Think about yourself and how you are coming across. Don't be too direct or impose yourself verbally or physically. Step back.
2. Take a deep breath yourself. Remember individuals with fragile X are great mimics.
3. Exaggerate your affect. Use positive language and speak slowly and calmly, even lowering your own voice. Try to have an agreed calm down routine – remind the person what this is and give them time to respond. Do not over stimulate the person. If it helps, say nothing at all.

4. Try to have scheduled recovery breaks during an activity e.g.

	Make bed
	Dust bedroom
	Listen to CD
	Hoover bedroom

5. Start and stop each activity in a regulated state

The final aim will be to move to the next level with the person being able to regulate themselves. But how? It is important to always be consistent, to make sure everyone involved is working in the same way and using the same strategies. The interventions need to be linked to the biology of fragile X – they need to manage what is driving the hyperarousal and any subsequent behaviours.

In this section Tracey and Mouse were very much looking at providing individuals with fragile X with a sensory diet that could lead to sensory regulation.

Sensory Diet

A sensory diet is aimed at children and adults who have sensory defensiveness. Tracey started working with sensory diets with children with fragile X in 1989. The aim is basically to treat

hyperarousal by improving sensory defensiveness. It is something that is important in both children and adults.

- A sensory diet is based on making sure that the key trickier events of the day have routinely scheduled into them activities to help the person with fragile X manage them better by calming down the nervous system. We all know that individuals with fragile X like routines. We also all know that one of the most common difficulties for individual with fragile X is managing transitions and so this is a key time to build in calming activities.
- It is important that the interaction between the individual with fragile X and the other person (possibly a parent or a key worker) is modified for the best effect
 - It is best to avoid strong perfumes
 - Think about the voice quality – calm, slow speech.
 - Modify environments as much as possible – think about the decor in an area that is going to be used specifically for calming activities
- How often sensory diet style activities will need to be schedule into the day varies from person to person. Once a good sensory diet is established, individuals can often go longer between “treatment” times. However, Tracey and Mouse did feel that no one with fragile X would be able to go for longer than 3 hours without having something put in place to settle them. They stressed that this was very important in our culture where medication is not so readily used. A sensory diet is therefore the only biological intervention.

Possible Elements of a Sensory Diet

Access to an Occupational Therapist will be useful here to give specific input on what is calming for the particular individual concerned, but possible elements of a sensory diet are

- Deep pressure like a massage.
- Something to give proprioceptive input and activate the system to calm down – lifting, pulling, pushing. Sometimes yoga style activities can be helpful here e.g pushing a ball. Guidance is needed to ensure the right amount of pressure for the right age



- Deep Pressure Proprioceptive technique (DPPT) is sometimes used. It is important to know how to do this technique and you need to be taught by an Occupational Therapist (OT).
- Oral pressure Technique is also helpful, but again you need to be taught how to do this by a trained therapist.

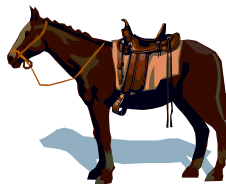
- Simple strategies can also help e.g. for the child to be a monitor and carry a bag of PE equipment to the hall, the box of musical instruments to the music room.
- Fidget toys can help to keep an individual in just the right state



- Oral based exercises like sucking, blowing, chewing, licking, blowing through a straw can be helpful. A biting options box containing things that gives resistance to the mouth when chewed e.g. a chew tube



- Relaxation responses like deep breathing
- Movement based activities e.g. swinging, rocking, dancing, horse riding



Tracey and Mouse gave the example of a family who had used dancing to the Riverdance CD as a grounding activity before dinner.

- Many individuals with fragile X have particular sensory defensiveness around noise and so noise concealing headphones can be used. Individual with fragile X are often socially motivated to take them off when they feel more comfortable



- A sensory visual choice board or key chain with pictures of the options can be used to give an individual a choice of calming activities e.g.

I want	



bubbles



koosh ball



music



swing

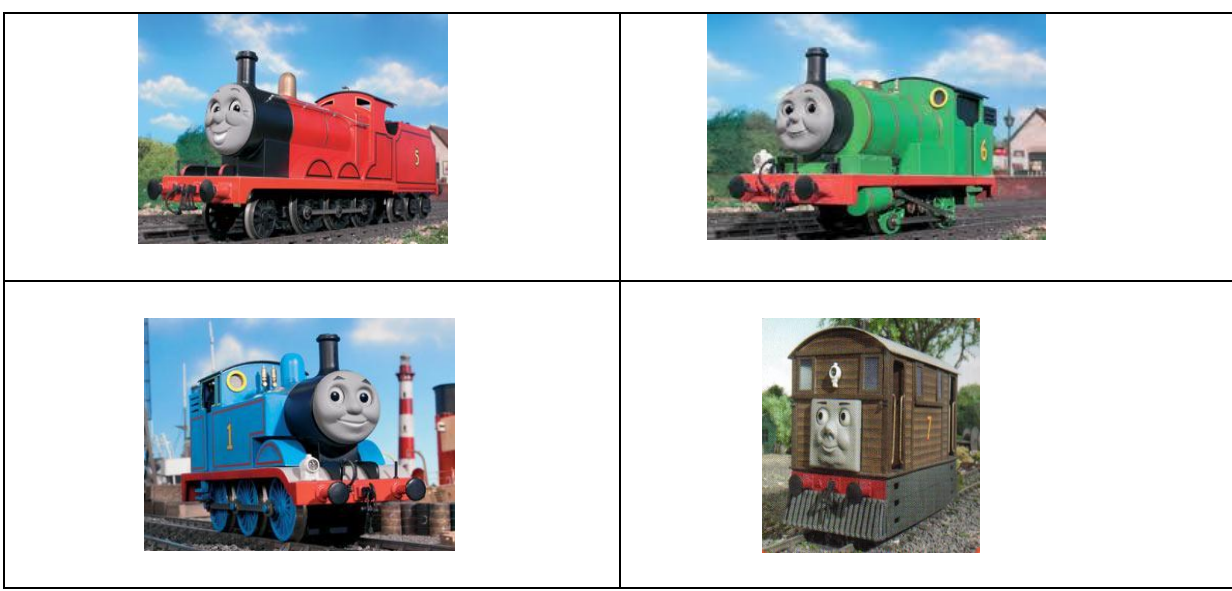
- A cosy corner where an individual can retreat to to calm down and regroup until they are ready to go back to the activity. The types of items to make up a cosy corner include a nice comfy place to sit, calming activities e.g. calming music.



For an adult this may mean time in their own room.

- Fragile X emergency kits can also be useful They will contain things very specific to an individual – things they like doing and are able to do successfully e.g. something to suck, fidget toys as mentioned above, matching games, simple puzzles

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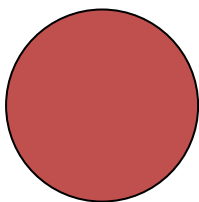


Self Regulation

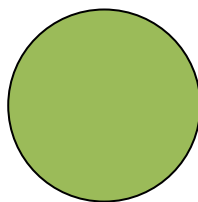
Again self regulation is important for children and adults with fragile X. It is about helping them to get their mind and body ready to do something. Generally speaking this is not something that is taught – we just expect people with fragile X to obey the rules just like everyone else without giving any consideration to thinking about how they are going to cope with the stress.

What we do know is that individuals with fragile X are good mimics. They will watch how we cope with stress – they often need to co-regulate and can follow the lead of another person to self regulate.

Colour cards can be used to help an individual with fragile X signal whether they are “ready” or “not ready” to engage in an activity.



Not Ready



Ready

Five Finger Approach

The idea is then is to help them move from the red to the green. Activities will need to be faded in to facilitate this. Tracey and Mouse have come up with an approach to help with this calming procedure, called the “Five finger Approach”. This is best done with a person modelling each step to the individual with fragile X



1. At the point of meltdown. STOP. Grab the thumb. You can say “Stop” or “I need to settle”



2. Grab the forefinger. Take a deep breath. You can say “Deep breath” or “Blow Hard”



3. Grab the middle finger. What do I need to do? This the time that they identify that they are out of sorts and need to settle down.



4. Grab the ring finger. What are we going to do about it? What is going to happen to help the person calm down. This needs to be something that really helps that person and you need to be aware what it is so that you can help them to move onto the calming strategy.



5. Execute the plan. Ok now we know what the strategy is, time to do it.

A Relaxed Body

So the final aim is to get the mind and body back to a relaxed state – ready to move onto the next activity or event, but what does a relaxed state look like. Tracey and Mouse came up with this analogy

S soft face, soft shoulders

T take 5 slow breaths

O open up the chest

P posture (calm)

Using Language and Routine Based Strategies for Managing Hyperarousal and Anxiety and Teaching to the Fragile X Learning Style

Tracy and Mouse's last session was about the use of language and routine based strategies and how best to teach and work with individuals with fragile X.

Routine Based Strategies

In this session Tracy and Mouse drew attention to 4 very important areas that match the learning styles of individuals with fragile X

- Schedules
- Visually based systems
- Work systems with a very clear beginning and end
- Supported transitions

We all know that individuals with fragile X are stressed by transitions whilst liking routines. This points to the need for keeping things as predictable as possible with routine based strategies. These include

- Visually structured routines throughout the day – visual schedules.
- Visual strategies for specific skill development

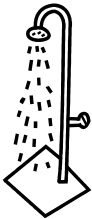



Visual Schedules

To a certain extent we are all dependent on visual structure and schedules – how many of you use a calendar to remember engagements? Visual structures are even more important for children and adults with fragile X – they are individuals who like to know what is going to happen and their visual skills are their strengths. Even when they know the routine, many still do better with a visual schedule there to confirm it. Having a set routine in place and being able to see clearly what that is can significantly reduce hyperarousal and thus free up layers of thinking.


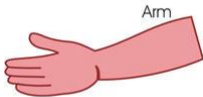




The example above shows the use of pictures to create a schedule, but that is not the only medium that can be used. It is important that whatever is used is something relevant to the skill level of the person – something they can understand. Objects, photos, symbols, pictures and written text all have their place. Sticky back Velcro is a good tool to use to stick the pictures to the schedule. It enables pictures to be removed easily to give closure and show that that particular part of the schedule is finished.

Sometimes it may be appropriate to have a schedule or timetable for the day and within that have sub schedules about particular activities within the day e.g.








			
shower	breakfast	trip to town	dinner

Part of the sub schedule of shower could be

 Head ©kidsfront.com	 Arm ©kidsfront.com	 Chest ©kidsfront.com	 Legs ©kidsfront.com
wash face	wash arms	wash body	wash legs

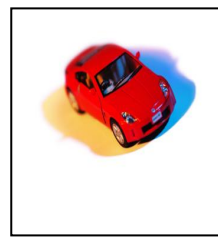
Tracy and Mouse spoke about a family where the adult male children had been able to progress to managing themselves in the shower by having a schedule rather than relying on their father to prompt them about what to do.

Sub schedules can also be used at school e.g.

Main schedule of the day	Sub schedules. Put in 1 at a time			
 Register				
 Literacy	 Circle Time	 Drawing	 Writing	 Circle Time
 Playtime				



Some families have also found it helpful to have a picture of a particular interest on the back of the schedule. This can be introduced as a topic of conversation to distract if anxiety levels start to go up.

Some families have also found it helpful to have pictures or photos to demote what a child can do when they get home to ease the transition from the car into the house. These choice pictures can be put on the back of the seat in front of the child.



Indeed Tracy and Mouse were referring to the importance of schedules for “down” or “relaxation” times as well as for times that are naturally more structured.

Sometimes a firstthen...style schedule can be used to encourage someone to do a particular activity e.g.

First	Then
 <p data-bbox="344 1106 485 1149">Drawing</p>	 <p data-bbox="979 1106 1091 1149">Trains</p>

Key questions to think about when devising a routine based intervention or schedule can include

- What am I supposed to be doing?
- How long will it last?
- How do I know when it's done
- What should I do next?

The schedules above give very clear messages about what is happening now and what is going to happen next. Visual timers can act as prompts to how long an activity is to last.

- Individuals can start to become more self reliant by making their own visual schedules – a list of the things they have to remember to do e.g.

Before I go to bed	
1	Switch off TV
2	Switch off lights
3	Lock door

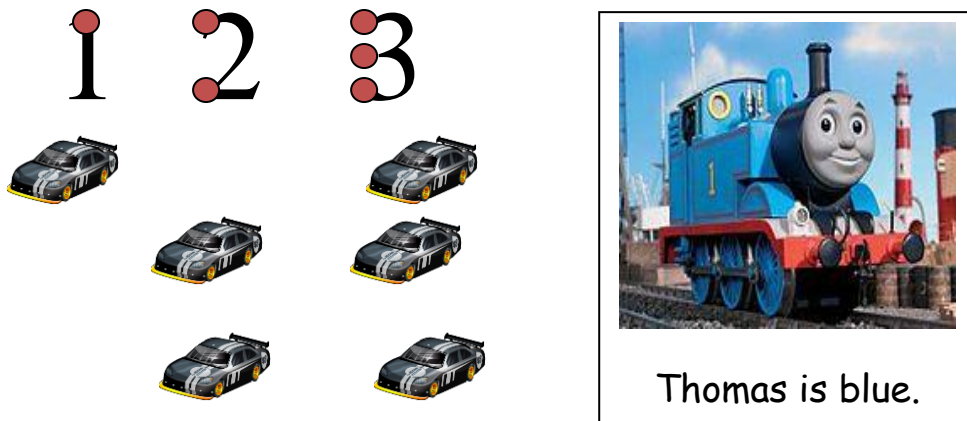
Visual Structures for Learning

When teaching or working with children or adults with fragile X, it is important to keep hyperarousal or anxiety levels down and to go with their learning styles – this is the only way they will learn/manage. We need to keep them in the frame of mind when they will maintain interest on the task rather than withdraw. But how?

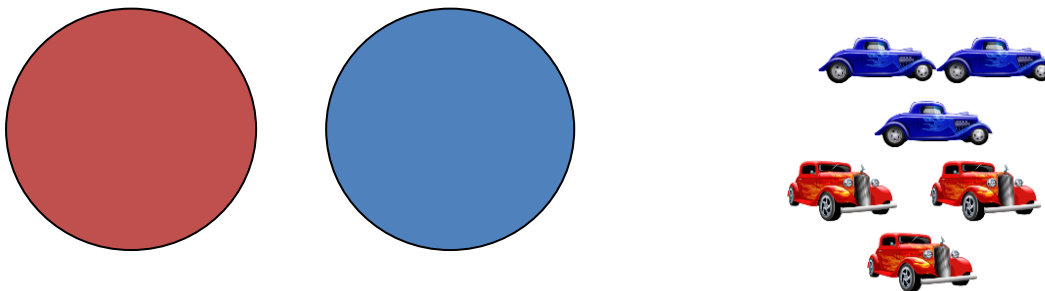
Individuals with fragile X are stronger visual learners and so we need to match strategies to this. Indeed visually based systems are pivotal when new skills are introduced. Having tasks with a clear beginning and end is also useful as individuals with fragile X do like to complete things. Attention is often enhanced by the use of high interest areas.

Strategies include:

- Counting objects of interest or reading books about high interest areas



- Colour matching a fixed set of items.



- Adults may be able to learn how to do a particular job by watching a video that shows the whole task (as mentioned in the first of these articles on Tracy and Mouse's talks people with fragile X do learn by seeing the whole). A schedule of the constituent parts can then be made to remind them how to attack the task on a daily basis.

Individuals with fragile X are also good mimics. Other useful strategies could therefore include

- Video modelling, DVDs and role play.
- Video modelling using someone they know can be an excellent way to model a desired behaviour e.g. to teach use of the five finger approach highlighted in the last article, to teach an individual to greet someone appropriately. Once the latter is learnt it is best to practice with familiar people first before moving the skill out into the community.
- Peer modelling is also a good way to teach desired behaviours.

The TEACCH structure can be useful as a learning tool. The key here is the movement from left to right and the completion of activities. Various activities are arranged in a pile on the left of the table. Each activity should be in a separate container and it should be very clear from the contents

of the container what the activity is and when it will be finished e.g. a puzzle with the pieces tipped out, a hand writing pattern sheet and a pencil. Shoe boxes can make good containers as can zippy bags. The number of activities that an individual is able to do at any one time can be gradually increased, but it is good to start with an easy activity, gradually build up the difficulty of the tasks and finish with a reward activity. For example if 4 activities were being set

1. Easy task
2. More challenging task
3. More challenging task
4. Reward activity.

The child picks up each container in turn, completes the activity in it and then puts the container on the right to indicate completion. Sometimes there is a finished box on the right hand side to affirm this completion.

Change

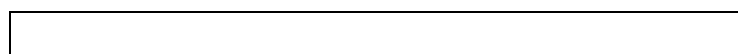
It is well documented that individuals with fragile X find changes to normal routines difficult to manage. It can help to practise small changes on a good day. This can help to prepare individuals for the possibility of change.

Language Based Strategies

- Pictures of events can help to prompt a child to be able to talk about what they have done.
- Looking at books or pictures that contain high interest material can also be used as a calming mechanism or as a conversation starter.
- Home/school books can be very useful in that they can give both teachers and parents/carers an idea of what has happened and thus help them initiate a conversation about it. Remember getting an individual with fragile X to complete a sentence can sometimes be a better than asking a direct question.
- Indeed direct questions can often prompt social anxiety so saying “Wow you look hungry I bet you would like some...” can get a better response than “What do you want to eat?”
- Language priming can be a way of helping a child take part in a lesson e.g. “You really liked this book because..... On a scale of 1-10 I bet you’d give it a.....”
- Side dialogues can be a way of introducing change, possibly with two adults and one taking on the role of the individual with fragile X e.g.
Adult 1 “Time to get finish with the cars and go outside”
Adult 2 “I want to keep playing”
Adult 1 “Let’s think what can we do to finish the game? Let’s drive all the cars into the garage. The first person to drive the car will be...”
Adult 2 “Me I want to drive the first car into the garage”

Side dialogue can be used in families e.g. with a parent talking to a sibling rather than directing the instruction at the child with fragile X.

- A worker/carer can also introduce a change by talking to themselves within the hearing of the person with fragile X e.g. “I know I am supposed to be driving to school, but I have to get some petrol. I know that is different, But I have to get the petrol first. Then I can drive to school.”
- Social stories with pictures have proved useful in many circumstances e.g. to manage a situation e.g.





Going to the cafe



John likes to go to the cafe



Sometimes the cafe is noisy







If the cafe is noisy John can put on his headphones.



Then John can finish his coffee.

- Rhythmically based instructions can also be helpful. Tracy and Mouse used the following example to show how this technique had been used to stop a young girl mouth stuffing. As each instruction was give an icon is moved over the appropriate picture

 <p>Jenny will take a slow bite</p>	 <p>Jenny will take a slow bite</p>	 <p>Jenny will take a slow bite</p>	 <p>Jenny will take a drink</p>
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This can then be moved onto the rhythmical phrases, using the pictures as above if necessary

Take a bitTake a bite.....Take a bite..... Drink

And Finally

Knowing about fragile X is the key to guiding treatments and ways of working with individuals affected by fragile X. The presentation of fragile X remains the same across the whole lifespan – different issues will wax and wane as individuals grow up, but they will always be there.

Everyday use of the strategies covered in this and the previous two articles will help the management of hyperarousal in those with fragile X. We need to anticipate and address the core issues for each individual person to teach them how to relax and be as calm as possible. None of this is a blanket programme – the assessment of individual needs is crucial to determine the strategies/treatments to meet those needs.

Useful Websites given by Tracy and Mouse

www.fragilex.org

www.marciabraden.com

www.developmentalfx.org

www.do2learn.com

www.teacch.com

www.TasksGalore.com

www.watchmelearn.com

www.thetvteacher.com