

The Fragile X Society

The Educational and Emotional Difficulties Experienced by Girls with Fragile X Syndrome

Presentation by Dr Lesley Powell, Educational Consultant (Children with Learning Disabilities) in Perth, Western Australia to the Fragile X Society National Family Conference October 2005

Today I will be talking about the educational and emotional difficulties experienced by girls with fragile X syndrome and I will follow their story from birth to early adulthood.

I will be focussing on girls with a FULL MUTATION but there have been some reports of PREMUTATION girls suffering from the same sorts of problems usually only associated with a FULL MUTATION. I will be looking closely at this in the future.

If you do have a PREMUTATION daughter who does present with an academic or emotional profile similar to the FULL MUTATION girls then what I am about to talk about may be relevant for your daughter.

Birth to 18

This section mainly addresses the education of girls who carry the FULL MUTATION and have an average or below average IQ, or a mild intellectual disability. These girls are more likely to be educated in a mainstream school setting than their more severely affected sisters. That doesn't mean that we are only talking about mainstream girls. We are talking about girls who will need the necessary skills to live independently. If your daughter can read and hold a proper conversation then she should be able to go to work or college and live an independent life and be happy. That's what most parents want for their children: Independence and happiness.

The difficulties exhibited by the girls in general do not necessarily apply to every girl who carries a FULL MUTATION. They may exhibit all, some or none of the following educational and emotional difficulties. The strategies regarding emotional and social development can also be used with PREMUTATION girls who exhibit social and/or emotional difficulties.

So, what are the problems facing the girls with a full mutation?

Academic Difficulties

- 60% have some degree of intellectual disability
- School-based anxiety
- Mathematics disability
- Poor organisational/planning skills
- Poor comprehension skills relating to "reading between the lines"
- Poor sequencing and planning skills
- Poor co-operative skills when working in a group

Emotional and Behavioural Difficulties

Anxiety

Social Anxiety

Hand Flapping – usually in the early years

Tantrums – linked to Anxiety

Shyness – life long problem

Poor Eye Contact – linked to shyness and anxiety

Low Self-esteem – “I’m stupid, I’ll never be able to it”

Retreating to an interior world - sometimes their life is confined to their bedroom

ADHD - predominantly inattentive rather than hyperactive

Difficulty making friends – often a lack of interest in spending time with peers

Early puberty 8-10 years is not uncommon

Direct and frank speech – honesty is not always the best policy, especially when you hurt someone’s feelings!

Literal reaction to the spoken word – eg, a headmaster told all his students to keep the playground clean and to tell people who drop litter to “pick it up!” Your average girl would not dream of telling others to pick up their rubbish but the little girl with fragile X took him at his word and became the playground litter police!

To give these girls their very best chance in life we need to begin early. Anxiety is the major factor in the difficulties experienced by the girls with a full mutation. This is why we need to start preparing the girls early in life:

Begin at Day One to make your daughter’s world harmonious, safe, understood. If a child understands her world she is less anxious. The world has to make sense to her.

Begin with baby massage. There are hundreds of books on the market. Create a safe home environment with as little stress as possible. An environment where a toddler can explore her world with curiosity rather than fear will reduce anxiety.

Look at the world through her eyes and answer the questions she isn’t asking. So many Mums tell me that their little girl never asked questions. Explain the seasons and how the natural world works. Why it is warm, why it is cold, why the leaves turn red and fall from the trees.

Read to your toddler, play games that will encourage her to take chances and engage with the world rather than viewing it with terror. Games like *Memory snap* and *I spy* help them to develop and extend their concentration skills, take chances and sequence.

Ask your daughter questions similar to the following:

What do the clouds look like? – this develops imagination

What do we need to do today? And, what will we take to nursery? – this develops problem solving, decision making and sequencing skills.

Girls who grow up in a nurturing, loving environment do so much better than girls who grow up in threatening environments. When I visit a family where Mum and Dad are loving and supportive I see girls who are less anxious and on target to reach their full potential. I see many girls, however, a few from families with fragile X and many from other families with a

variety of conditions, who are self-harming because they are living in hostile and threatening environments.

Reducing Social Anxiety In Pre School Girls

When a girl is identified before school age, early intervention strategies can be implemented in a bid to raise a more socially confident child who is able to interact with her peers by the time she begins her formal education.

1. Join a playgroup or other activities for toddlers and observe your child at play.
2. Observe the other children at play and encourage similar experiences for your child.

Remember, very young children don't play together they play along side one another so don't expect co-operative play. Your daughter will get used to being in a group, experiencing social interaction, and listening to, and hopefully participating in, early conversation. Other children will be your daughter's role models.

Your daughter may gravitate towards the boys. This is because little boys are less complicated than little girls. Boys' conversation is literal and they tend to "do" rather than "say". That's fine, but try and steer her towards a mix of boys and girls.

Getting Ready For School – Literacy

The following strategies will prepare the girls for formal reading lessons. Use them to teach your daughter to think about the text as they listen. Ask very young children to:

1. Look at the cover and title of a book and ask your child to predict what will happen in a story (make a silly prediction yourself!)
2. Give them an opportunity to reassess their prediction as you read
3. Ask the child to retell the story (sequencing)
4. Discuss why the characters did certain things (comprehension)
5. Think of a different ending (make it lots of fun)

The use of context clues can be taught from a very early age by encouraging very young children to look for clues in the illustrations in children's books. You can help promote the use of context long before a child attends school simply by allowing them to finish a sentence, phrase, or nursery rhyme for you; or by making amusing errors in sentences that they can correct. For example, the pig said Moo! In other words, getting your children ready for school and literacy through play.

Getting Ready For School - Maths

Fragile X girls usually have great difficulty mastering mathematics. The following are pre-maths skills that must be mastered before formal lessons begin.

1. Counting
2. Number Recognition
3. One to One Correspondence
4. Mathematical Language, for example over, under, through, more than, less than, equal

- to, left, right, up and down, before and after.
5. Sand and Water Play: weights, measures and estimation skills.
 6. Sorting and Grouping: sorting objects into groups by colour, shape, animals, transport etc.
 7. Classification: sorting groups into more specific classifications. For example, sorting a group of transportation toys into groups of buses, planes, cars, trucks and trains.
 8. Sharing equally: sweets, cake, blocks etc.

Formal Education

My research found that girls aged 9 and under were able to perform simple sums involving addition, subtraction, and multiplication but many of the older girls, when presented with quite simple calculations, had commented that they used to be able to do those types of sums but had forgotten the procedure. Thus, the difficulties with mathematics associated with the full mutation are sometimes not evident until the girls are aged nine years and over.

Many of the girls under nine years were also confident about their mathematical ability whereas the older girls were not. Anxiety and poor self-esteem with regard to mathematics were constantly noted despite my reassurances.

After the age of nine, children are taught problem solving where they must apply their basic mathematical skills to abstract situations. They must tackle complex fractions, decimals and geometry. The research showed that many of the older girls had been so confused by complex mathematical concepts that they no longer retained their basic, rote maths skills. They could no longer tackle division and multiplication. Anxiety now prevented them from attacking their mathematical problems with confidence. The logical solution to this is to limit their mathematical learning to what is relevant to their lives. This does not mean that very able students learn only basic mathematics. It means that girls who obviously experience mathematical difficulties should not be required to struggle with concepts that will have no significance in their adult lives.

It is also known that children with fragile X work better with 'concrete materials' or a visual representation but this is just not possible in a real life setting. Concrete materials can be used to teach new concepts and skills but at some stage the girls must reach the level where they can perform addition, subtraction, multiplication and division without reliance on concrete materials or visual cues for assistance. Reliance on concrete materials often indicates that the child does not understand the underlying concept of the mathematical task at hand.

The teaching focus at the primary level, therefore, must concentrate on learning the basic mathematical concepts related to functional mathematics followed by consolidation of those skills at the secondary school level. Girls who have no mathematical difficulties should of course continue with their secondary mathematics studies, but there is no advantage in insisting that girls who are bewildered by higher order mathematical concepts continue, especially if this results in a situation where they may lose the ability to perform simple tasks that will be needed over the course of their adult lives. Also, to continue with concepts that are too difficult guarantees failure, social anxiety and low self-esteem.

The sensible approach is to revise basic mathematics skills and concentrate on academic areas of interest and ability that will promote high self-esteem and allow the student to either excel or at least be on a par with their peers.

Mathematics: Strategies For Parents

1. Make sure that your daughter knows the basics and continually revise them so that she does not forget them.
2. Inform teachers of your daughter's difficulties and make it clear that you are not concerned if she does not understand abstract mathematical concepts, for example, algebra or how to multiply fractions. Insist that teachers concentrate on, and continually reinforce, the basic mathematics skills listed below.
3. Involve your daughter in simple mathematical tasks at home. For example, setting the table and shopping are functional mathematical tasks.
4. Impress on your daughter that the difficulties that she has with mathematics are not her fault and you only expect her to work to the best of her ability and to achieve her highest level of expertise in the subject. No girl can do more than fulfil her potential. The following is a list of mathematical skills your daughter needs to learn and retain. If your child can go further and still retain the basic knowledge then of course allow her to do so. (I have never met a girl with a full mutation who loves maths!)

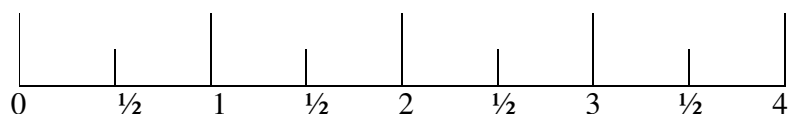
The following lists of basic mathematical skills are guides for teachers and parents.

Hierarchy of Basic Mathematics Skills

1. Addition
2. Subtraction
3. Calendar Skills: days of the week, months of the year, seasons, calculating dates.
4. Counting by twos, fives and tens.
5. Measurement: centimetres, metres, kilometres, perimeter and area.
6. Basic Geometry: shapes, angles, parallel lines.
7. Money
8. Basic Multiplication
9. Use of a calculator to perform complex addition, subtraction, multiplication and division.
10. Basic Fractions: halves, quarters, thirds, eighths.
11. Basic Division: using one digit denominators
12. Simple addition of fractions with the same common denominator or denominators divisible by 2, 4, 3, 5, or 10.
13. Simple Problem Solving - writing algorithms for story problems relating to real life situations. For example: Three people are going to MacDonald's and they want two hamburgers each.
 $3 \times 2 = 6$ (hamburgers).

It is better to teach fractions using the number line method that allows whole numbers and fractions to be seen visually and in context without removing sections. For example, the following figure is a number line representing whole numbers and halves.

Number Line



Literacy

Most girls read well and some read extremely well. If a girl does have reading difficulties the best method of instruction is to teach the four main reading strategies for word recognition, fluency and understanding. These are: sight words, phonics, context and comprehension.

Sight words provide a base for word recognition because they are recognised instantly by students. The more words a child recognises by sight alone the more fluently they read. The Fry Instant word lists comprise three lists of 100 words each.

Sight words

The first 10 words make up approx. 24% of all written material.

The first 100 make up approx. 50% of all written material.

The 300 words make up about 65% of all written material.

These can be learnt with flash cards, playing word bingo, and writing simple sentences using these words. Many of the words can be used to teach word families, for example, light, night, fight, might, fright. Many simple everyday words are irregularly spelt so need to be recognised by sight rather than sounded out. For example, enough and eight.

Phonics

Using phonics (association of specific sounds with specific symbols) rarely works with boys with fragile X because of their totally holistic approach to word recognition. This does not appear to be the case with the girls. Phonics instruction should include: consonant blends, for example, bl, gr, st, str, etc; long and short vowels and the use of mute 'e', for example mat has a short vowel while mate has a long vowel because of the additional e. Double consonants that create a new sound, for example, ch, sh, ph, ck, th and; double vowel combinations, for example, ee, ea, oa, ow and ai.

Context

Context helps the reader in identifying an unknown word and deciding which pronunciation of a word to use in a sentence. For example,

I read yesterday

I will read today

Comprehension

If a child does not understand what she is reading then she is simply decoding or attacking each word in isolation. Make sure she understands what she is reading.

Anxiety and Social Behaviour

It is well documented that many girls with fragile X, with or without learning difficulties, suffer social anxiety resulting in poor social skills including: gaze avoidance; difficulty conversing with, and initiating contact with their peers; organising social outings; isolation (isolating themselves as well as exclusion by peers); understanding the use of specific language in specific social contexts and; blatant honesty - it is not always the best policy to tell the truth!

Parents face a real dilemma. They desperately want their daughters to be socially comfortable and competent, to be relaxed and enjoy social interaction and, to seek and maintain friendships. Conversely, parents often realise that their daughter does not seek social interaction nor does she miss the closeness of friendship. Unfortunately, in reality these girls are expected by society to live actively in the world, gain employment, perhaps marry, raise children and be productive, successful members of society.

Do parents leave them in isolation where they feel comfortable? In many cases, yes, especially when the girl has not been diagnosed until she is struggling to cope with the social demands of adolescence. It is very difficult to teach high level social skills to girls and young women who are already experiencing social anxiety or bewilderment at the behaviour and multi-layered conversation skills of their peers.

As well as dealing with her own difficulties, girls with fragile X often have to make allowances for one or more affected siblings. Research carried out in Australia by Karen Anderson (2001) identified several negative effects on siblings with special needs brothers and sisters including:

1. Feelings of isolation
2. The burden of care taken on by siblings as children and in adult life
3. Pressure to achieve - to be the "superchild"
4. Jealousy relating to the amount of attention the affected sibling receives and the unacceptable behaviour that they exhibit without punishment.
5. Fear of developing or passing on the gene (a very real fear for girls with fragile X).
6. Guilt at not having the disability. During my research, sisters who had not inherited the fragile X gene often expressed these feelings of guilt.

The participants in Karen Anderson's study were siblings who had no disabilities. The problems are much worse for girls with a full mutation who have problems of their own to deal with. To lessen the effect of coping with severely affected siblings:

Ease The Burden

1. If at all possible, place your daughters in a different school from that of your severely affected sons and/or daughters.
2. Do not expect them to care for their affected siblings.
3. Discuss the issue of guilt with all family members.
4. Do not expect more from girls academically than they are capable of achieving.
5. Encourage them to participate in activities away from the home environment.

Social Skills Training - The School Years

This section will cover: Making Friends, Shyness and Conversation, Working in a group, Making Decisions, and Sequencing and Planning.

Making Friends – Strategies for Parents

1. Encourage your daughter to choose friends with similar interests or a similar personality. A shy child may not seem to be the best role model but the most popular or cleverest child in the class or group may overwhelm your daughter.
2. Once your daughter reaches school age you may have to take the initiative and invite girls over to play who are similar to your child - quiet, shy and gentle. Later, you can encourage your daughter to invite children to play.

Making Friends - Strategies for Teachers

1. Encourage the girl in your class with fragile X to participate in class activities and contribute to discussion.
2. Gently guide her towards friendships with other girls in the class, especially quiet, compassionate girls.
3. Recruit a girl from a higher grade with similar interests to become her mentor. In Australia this has resulted in firm friendships.
4. Watch for signs of creativity or interest in art, music, dance or drama and encourage areas of talent. Many girls excel in these areas where they can “be someone else”.
5. Encourage eye contact in non-threatening situations but do not insist on eye contact when a child is anxious or upset.

Shyness and Conversation - Strategies for Parents

1. Talk to your daughter about eye contact and shyness. Persevere gently and point out that there is no need to be shy with people who have proved to be trustworthy.
2. Encourage her to talk to you about her feelings.
3. Role-play social situations over and over again and then act them out in public. This should include ordering in a restaurant, eating in public, asking for directions, buying a ticket at the cinema, shopping, using public transport, and what to do if she is lost or anxious.
4. Role-play conversation for different social situations.

If your daughter remains shy and anxious and if the anxiety increases with age then medication may be required.

Working in a Group

Group work at school is excellent for girls with fragile X. Students are usually set special tasks or have individual roles within the group and this enables girls to work on their own area but still work co-operatively within a group. Teachers need to be conscious of group dynamics and place the child in a group of children who will interact with her and act as positive role models without overwhelming her with their actions or personalities.

Making Decisions

Girls with fragile X find it very difficult to make decisions especially if they are in a public place and anxious about the social setting. For simple decisions such as deciding what to order in a restaurant, your daughter can carry a special coin to flip if she really cannot decide between two choices. Decision making, however, is often more complicated than deciding what to order.

Always discuss a variety of options with your daughter and give her an alternative plan of action. If as a parent you say that you will pick her up at a particular time make sure she knows what to do if you don't arrive on time. If possible, provide her with a mobile phone which provides security for children and young adults who find themselves in situations where things have not turned out the way they had planned or expected and they cannot decide what to do or are anxious about decision making.

Sequencing and Planning

1. Provide a written list, or sequence of pictures, of tasks to be completed before school, getting ready for bed, or how to tidy her room etc.
2. Play a nightly game of remembering and sequencing the day's events.
3. Ask the teacher to quietly explain the day's activities each morning to reduce her level of anxiety. With boys and severely affected girls, parents and teachers use a variety of strategies, including photographs and pictures to provide a visual plan of the day's activities to avoid anxiety. These girls also need the security of knowing what the day's activities are and what they will be doing next to avoid or reduce anxiety.
4. When you are planning a family activity, for example a picnic, ask your daughter to help you plan the picnic and sequence as many different activities as possible.
5. Model thinking aloud. This is a form of Metacognition, or, thinking about what you are thinking! Teach your daughter to talk to herself and ask herself specific questions such as: What equipment do I need to do this activity? How will I go about it? What will I do first, second and third?

Further Reading:

Other papers by Dr Lesley Powell available from the Fragile X Society:

Girls with Fragile X : the Big Picture
Conference, December 2001

Cognitive Functioning Profiles of Schoolgirls with Fragile X Syndrome
Newsletter No 18 Winter 1998

Educational Needs of a Child with Fragile X Syndrome
Newsletter No 20 Winter 1999

Chapters by Dr Lesley Powell in 'Educating Children with Fragile X Syndrome' edited by Denise Dew Hughes, published by Routledge Falmer:
Educating Girls with fragile X syndrome
Emotional and Social Difficulties of women with fragile X syndrome

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