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Information Sheet 48

Fragile X Syndrome

By Dr Leonie Star

Leonie Star is a freelance writer who lives in Sydney. Her much loved grandson has both fragile X and autism. She has researched and written about fragile X, educational and training needs as related to government programs and links with autism in the journal of The Fragile X Association of Australia. Leonie was President of The Fragile X Association in 2004 and 2005.

Todd is a five year old boy who has a significant mental impairment with speech delay. He dislikes loud noises and very bright lights and has poor fine and gross motor skills. He is also subject to constant ear infections. Emily is an eight year old girl who seems to have normal intelligence except for a difficulty in learning mathematical concepts. However, she is shy and moody and becomes extremely anxious in some situations.

What do these children have in common? They both have fragile X syndrome, which is the most common inherited form of mental impairment worldwide, second only to Down syndrome as a cause of mental retardation. Yet outside those families who are affected by it, surprisingly few people know about it. It affects approximately 1 in 3,800 males and 1 in 4,000 females. A genetic condition, 1 in 809 males and 1 in 129 females are carriers. In an affected family, children can either be born with fragile X, be carriers or be totally free of the syndrome. The likelihood of a carrier giving birth to a child with fragile X is about 50%.

Males are generally more seriously affected than females, mostly suffering mental retardation ranging from mild to severe. They can also exhibit anxiety. Two thirds of affected females show some mental impairment, as well as suffering from serious, often crippling, anxiety. There are some medical problems associated with the syndrome but they generally do not shorten life.

Accurate DNA testing, called 'DNA Studies for fragile X syndrome' is now available and covered by Medicare. Any medical practitioner can refer a patient to a geneticist or developmental paediatrician to order this test.

People who should be tested for fragile X are any male or female with an intellectual disability (borderline to severe), developmental delay or learning disability of unknown cause; individuals with a family history of undiagnosed intellectual disability or fragile X syndrome; any male or female with autism-like characteristics (because of the increasing evidence of a link between fragile X and autism) and any individual with a previous cytogenetic test that was negative or inconclusive (because previous cytogenetic tests for fragile X were not always accurate).

Pre-conceptual testing to determine a woman's carrier status is also recommended if there is a personal or extended family history of any of the above criteria.

Most people affected with fragile X show some signs of being different from their peers. A major problem is self-regulating behaviour and lack of impulse control – attention-deficit disorders and hyperactivity are commonly associated with the syndrome. Emotions can spiral out of control, resulting in tantrums.

However, it should be understood that behaviour problems are often caused by anxiety.

Females with fragile X, whether or not intellectually impaired, can be extremely shy and lack the social skills of their peer group. Some affected people find eye contact extremely difficult, rarely looking you in the eye even if they know you well. This trait can persevere into adulthood.

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We raise funds to help children from birth to 18 years by offering a range of services including the following.

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- Early childhood intervention and support for very young children.
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- An assessment and consultancy service for families who are concerned about their young child's development.
- Specialist early childhood teaching and therapy.

School Age Services for children from Kindergarten to Year 12 who have low support needs.

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- Small group tuition and therapy.
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- Outreach programs.
- The Ronald McDonald Learning Program for seriously ill children and the Reading for Life Program for children falling behind in their reading.

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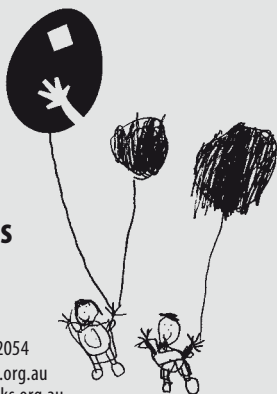
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Learning Links has branches in six Sydney locations at Peakhurst, Penshurst, Fairfield, Miller, Dee Why and Randwick. We also offer some services to children in country NSW, the ACT, Victoria and New Zealand. A complete list of branch locations and contact numbers is on the back cover.

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In young children, toilet training is often delayed and many suffer from disturbed sleep. Children may bite their hands or clothing when anxious. Repetitive behaviour and repetitive and poor speech patterns are often found. One sign of the syndrome is so-called 'flapping' when the person is agitated or excited – arms are raised and the hands flap continuously. Flapping often moderates as a child matures.

People with fragile X usually have a good sense of humour and can crack up at jokes they understand or situations they find comical. They generally have pleasant personalities and can be very social. However, communication may be difficult, which is particularly hard when a child is upset and cannot express the reason. They have excellent memories and because they are such good mimics, learn a lot by imitation. This is why it is essential that they are provided with the right role models wherever possible.

For preschool children, early intervention programs are vital, particularly where there is a late diagnosis. Parents aware of developmental delay are advised to arrange early intervention as soon as possible, whether or not they have received a diagnosis of fragile X. Most children make advances when they receive speech therapy, occupational therapy with sensory integration and behavioural therapy.

Some primary school children are integrated into regular classes at local schools. Alternatively, they may be enrolled in special classes at local schools or in special schools.

Parents may favour the idea of integration into regular classes as children are exposed to good peer group role models. As children with fragile X learn by mimicking those around them, those educated with their peers pick up useful regular behaviour patterns.

When children are integrated into regular classes, some curriculum differentiation may be needed. Many children benefit from the assistance of a teaching aide for at least part of the day.

In special classes in regular schools and in special schools, classes are smaller and most teachers will have been trained in Special Education, although perhaps not specifically how to teach those with fragile X. For teachers who find a child with fragile X in their classroom, information is easily available to assist them (see notes at the end of this article). Most techniques can be incorporated into the ordinary teaching day with little difficulty.

Because the child either cannot or will not communicate about the day's events, open and constant communication between parents and teachers is essential. In regular classes in local schools, a successful way of ensuring both parents and teachers know what is happening is to institute a 'Communications Book'. Both parents and teachers write short notes about the day's happenings in the book, which is sent home and then comes back the next day. In this way adequate communication is maintained and incipient problems can be addressed.

Making sure that the child with fragile X understands what is going on is vital.

Because some children are either non-verbal or are not able to communicate freely, it is sometimes assumed that their understanding is extremely limited. This is not always the case. The child should be given every opportunity to understand why and how things are to happen. For example, when a child says, 'Can I do some painting', rather than saying 'In a minute', it is better to explain that 'We'll have to wait until everyone has finished reading so the books won't get paint on them'.

Children with fragile X need highly structured routines and must be carefully prepared for any changes. Many children find it useful to carry small cards with them charting the program for the day – these can either be read by the student, if able, before school so he/she goes off prepared, or read to him/her.

One of the constant questions asked by children with fragile X who are verbal is 'And then?' They need to know exactly what is going to happen and what is going to follow, so that the answer 'I don't know, we'll wing it' is likely to cause not only confusion but actual distress. Once the known program is followed, parents may find that the child can remember what to take to school on what day – which day is 'library day' and which day is 'sports day'.

If the change in routine involves a new situation, or if the child seems unable to grasp repetitive routines, 'social stories' often help. A social story consists of a series of simple line drawings in sequence which break down the activity into its component parts. Social stories are used to describe expected behaviour; often behaviour improves as anxiety of the unknown is removed.

For instance, if a social story is designed to assist the child to be quiet in the library, the words accompanying the frames could be 'first we line up with Mrs Taylor, then we walk quietly to the library, then we give back last week's book to Mrs Green, then we pick a new book from the shelves, then we sit down without talking and read it, then we have it stamped by Mrs Green, then we quietly take it back to the classroom.' Although constructing this story might initially seem like a lot of effort, with practice social stories can be composed very quickly. The story should be read through with the child before the event, not during it.

The best way to educate a child with fragile X is to use the gestalt or 'whole picture' approach.

The child needs to see the activity as a whole. If making biscuits, assemble all the ingredients, show the baking tin, discuss how the oven will be used and display a completed biscuit. Do all this before beginning to cook and anxiety about what is to come next will not interfere with the task. It is most important for a child with fragile X to see the end result of any activity before it is begun.

People with fragile X are visual learners. Reading is best taught by the 'whole word' method rather than by phonics. Good memory skills will help the child build up a reasonable vocabulary.

Wherever possible, verbal learning should always be supplemented with visual, by way of pictures or signs.

Teaching should be done in blocks of no more than 15 minutes, less for a very young child. When teaching children with fragile X in a one-to-one situation, teachers should not face them but preferably stand behind them, so reducing the strain of eye contact.

Nearly all people with fragile X suffer from sensory overload – information impinging on them through their senses is felt far more acutely than in regular people. Placing a child with fragile X into a regular classroom means that he or she will be exposed to a busy, stimulating and even noisy environment. The more that auditory and visual stimulation can be reduced or eliminated, the better the child will respond.

Providing 'time out' will give the child a necessary break from the generally stimulating atmosphere – this can be in a special corner of the classroom, perhaps behind a screen, where the child can be alone for a few minutes, or can take the form of a task to be performed outside the classroom, such as taking a note to the office.

The institutional choices for secondary education are the same as for primary education. Many special courses are available in secondary schools and modified certificates can be attained. Some people with fragile X proceed to tertiary education, usually at the TAFE level. The government provides training courses to suit individual needs, with those in the hospitality area being particularly popular. People with fragile X generally make good workers, are reliable in the workplace and easy to get on with.

Sources for further information

Fragile X Association of Australia

10 Geddes Street
Balgowlah Heights NSW 2093
Telephone Support: 0409 987 012
Office: 02 9949 8456 (Thursday/Friday)
Email: fragilexassociation@optusnet.com.au
www.fragilex.org.au

The recently published booklet *Explaining Fragile X Syndrome* is available in multiple copies at no cost from The Fragile X Association of Australia.

Websites

The Fragile X Association of Australia's website includes lists of books helpful to teachers. The booklet *Explaining Fragile X Syndrome* is available on the site electronically. Some of the information in this article comes from this website.

See also the sites of The Fragile X Alliance, run from Victoria, www.fragilex.com.au; the National Fragile X Foundation (US), www.fragilex.org; and The Fragile X Society (UK), www.fragilex.org.uk.

Clinics

Both the Fragile X Association of Australia and the Fragile X Alliance run clinics for children and adults. For more information see their websites.



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Early Childhood Services
– all enquiries to Head Office

School Age Services
– contact your local branch

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– contact your local branch

All other enquiries
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