

THE FRAGILE X SOCIETY



**FRAGILE X SYNDROME:
AN INTRODUCTION TO EDUCATIONAL NEEDS**

INTRODUCTION

The aim in producing this booklet is to provide an introductory guide for teachers interested in Fragile X Syndrome.

The information it contains comes from a variety of published works and experiences and though the booklet refers to children throughout, much of its content is applicable to adolescents and adults in educational and training settings.

The Fragile X Society gratefully acknowledges the help and advice it received from its specialist advisors: Charles Gibb who compiled the booklet and Dr Jeremy Turk for his support and comments.

Photography by kind permission of Piperway School, Leicestershire, Kingswode Hoe School, Essex, Nursery Learning Ltd, Essex, Yeovil College, Somerset and Anastasia, Connor, Joshua, Rebecca, Abhimanyu and Andrew whose photographs appear.

Third Edition reprinted 2003 by
The Fragile X Society
Rood End House
6 Stortford Road
Great Dunmow
Essex
CM6 1DA
Tel: 01371 875100
Family Helpline: 01424 813147
Fax: 01371 859915
Email: info@fragilex.org.uk
Website: www.fragilex.org.uk
Registered Charity No. 1003981

WHAT IS FRAGILE X SYNDROME

Fragile X Syndrome is the most common identifiable cause of inherited learning difficulties (mental handicap). It shows itself in a wide range of difficulties with learning and in developmental delay.

Until recently Fragile X was relatively unknown. In the last few years medical understanding of the condition has developed significantly. Educational knowledge has also developed but large gaps remain. Again until recently, accurate diagnosis was difficult and many individuals went unrecognised. It is possible that without knowing it, many experienced teachers will have come across at least one child with Fragile X.

The genetic origin of Fragile X was not discovered until 1969 when it was found that individuals showing particular mental and physical characteristics had a fragile area on their X chromosome. It is now known that problems with the associated gene are the basis of the syndrome. It should be stressed that children with Fragile X are not physically fragile. They have generally good health and are as robust as other children.



THE EFFECTS OF FRAGILE X ON LEARNING

Fragile X is wide-ranging in its effects, from specific learning difficulties and subtle learning problems with an average IQ, to severe intellectual disability and developmental delay. Although this booklet describes many features of Fragile X which have educational implications for the children and their teachers, it is important to note that any one child may present only a few of the problems described, or may present a whole range of features but in a relatively mild form.

Speech and Language

Speech and language are almost always affected. Delayed development in these areas is often one of the earliest presenting features.

Repetition of words and phrases with up and down swings of pitch is common, as are repetitions of the last word or phrase spoken to them (echolalia), repetitions of words and phrases they have themselves spoken (palilalia), and a tendency to skip from topic to topic while in conversation. Speech can be rapid with poor control of rhythm (dysrhythmia) and words seem to fall over each other with pauses often in the wrong places making the speech difficult to follow.

While many children with Fragile X have difficulties in speaking and being understood, their understanding of speech may be significantly higher than one would expect. Teachers often comment on the surprising abilities of children with Fragile X to take in what is said in the classroom. Characteristically, receptive language skills are generally better than expressive.

The advice and guidance of speech and language therapy services is essential here. An intervention programme should be individualised to suit each child, their interests and current attainments. The programme should also be integrated between school and home. Co-operation between therapists, teachers and families is the approach most likely to meet the child's needs. New skills and concepts can be introduced in 1:1 sessions. These should be regularly practised and reinforced in natural social contexts.

Also important is access to normal language experience. Makaton

sign language, the TEACCH visual structure and PECS (Picture Exchange Communication System) have in a number of cases proved a valuable aid to communication and overall language development.

[Information Overload](#)



Children with Fragile X often find it difficult to make sense of and react appropriately to the welter of information coming to them via their senses. In busy environments they can be overwhelmed by the excessive stimulation, sometimes responding with tantrums, withdrawal, hyperactivity, repetitions and other confused behaviours. Adults need to be aware of situations that may provoke such reactions. When they do occur, a calm and comforting response should be aimed for.

Curriculum Strengths and Weaknesses

It is almost certain that in all cases a degree of curriculum differentiation will be necessary. In a significant number of cases, additional classroom support will be required.

The need for suitably differentiated teaching may be particularly evident in arithmetic and mathematics. Children with Fragile X can find this work exceptionally difficult, particularly where conceptual learning is involved. Handwriting, too, can present significant problems.

Reading and spelling can be relative strengths, with some children



achieving average range performance at least in terms of accuracy. Again though, conceptual learning such as that involving complex understandings and grammatical nuances can prove problematic.

BEHAVIOURAL ASPECTS

Children with Fragile X often show characteristic patterns of behaviour which tend to moderate as the child grows older.

Overactivity/Hyperactivity

Always rushing about, seldom sitting still.

Impulsivity

Not inclined to wait for anything, wanting things straight away, doing first and thinking later.

Inattention

Not sticking to one activity or task for any length of time.

Distractibility

Inability to maintain concentration.

Social Anxiety

Children with Fragile X can be reluctant to give eye contact and will tend to avoid other people's gaze. This can be particularly evident in situations the child perceives as stressful. Such social anxiety is often a trigger for what can be perceived as challenging behaviour in the classroom.

For a child with Fragile X, difficulty in making or receiving eye contact is not a sign of social indifference. They find eye contact uncomfortable in social situations such as meeting new people, or when they feel they are the centre of attention. Consequently, care should be taken not to force eye contact upon them as this will simply exacerbate their anxiety. Teachers may find that once a child feels comfortable in the classroom eye contact is more likely to follow. Clearly, "making eye contact" would not be an appropriate IEP target for such children.

Mimicry

Tendency to copy humorous or slang expressions and bad language.

Memory Retention

Generally short but can be very good over the long term in a topic where the child's interest has been engaged.

Liking for Routines

Can become upset or confused when familiar routines are broken or things around them are changed, such as the furniture or layout of a classroom.

Repetitive Behaviours

Such as hand flapping and hand biting can occur, usually at times of distress, anxiety or excitement.

Children with Fragile X are not 'badly behaved' in the commonly understood meaning of the term. Nevertheless, they can still present teachers with management problems. They may, for example, get up on impulse and start meandering around the classroom, or they may react vigorously and inappropriately to events around them. It can be easy to misinterpret these and other behaviours as wilful and uncooperative.

While we might expect such behaviours from a child with Fragile X that does not mean we have to accept them. Teachers need have no worries about insisting on appropriate behaviour in circumstances where the step is small enough and the child is capable of reaching it. That is part of the learning process. As with other curriculum areas, however, longer term and supported practice will probably be required, and some individuals may require a permanent behaviour management strategy to be in place.



SCHOOL PLACEMENT

Some children with Fragile X, should, with the kind of differentiation or additional support mentioned earlier, manage within the normal run of mainstream. This is likely to be particularly so for younger children. But for others, additional resourcing will be required. In many cases special schooling may be best placed to meet their educational needs.

Given the facility for mimicry that many children with Fragile X show, mainstreaming is, wherever possible, the preferred option since the children tend to learn a great deal from the models around them. This is particularly important in respect of the development of language and the development of appropriate standards of behaviour.

The decisions about which type of school placement is most suitable can only be made on an individual basis where a child's educational needs are matched to the range of needs a particular provision aims to address. There are no absolutes in such decisions. For *any* child any form of schooling will create needs as well as meeting them. The Fragile X Society takes the view that placement decisions should be made on the basis of an informed judgement in respect of more needs being met than created.



CLASSROOM ENVIRONMENT

Classrooms by their very nature are busy, stimulating environments. Excessive stimulation can over-excite a Fragile X child and lead to tantrums, over-activity, withdrawal, repetitive behaviour and further disrupted speech patterns.

An environment that controls for over-stimulation can avoid many of these potential problems. For example:

- Minimising auditory and visual distractions.
- Working from a position behind or alongside the child to reduce the need for eye contact.
- "Packaging" work in at most 15 minute blocks to retain attention.
- Providing a personal workstation.
- Establishing and maintaining links with parents (perhaps a home/school book) to keep informed of developments.
- Teaching methods which involve relatively few distractions.
- "Time-out" to a familiar place or activity to give the child a break when necessary.
- Establishing familiar and consistent classroom or individual routines, keeping the child informed of impending changes.
- Making sure the child knows why and how things are to happen. For example, when a child says, "Can we read a book?", rather than saying "In a minute", it is better to explain: "We'll have to wait until we finish our water experiment so the book doesn't get wet".



NUMBER WORK

The problems here are believed to be with the kind of reasoning required and with the sequential nature of the thinking. However, other Fragile X characteristics - such as difficulty with attention control - do have a significant part to play.

Children with Fragile X are apt to lose sight of what it is they are being asked to learn, or even not see it in the first place. They tend to go down blind alleys, to attend to wrong or irrelevant aspects of the task. This difficulty becomes greater with more complex and involved teaching methods because the opportunities for following the wrong paths are much more numerous. Direct, unequivocal teaching methods are likely to produce better results. For many children with Fragile X aspects of maths may present exceptional difficulties. Teaching should concentrate on practical applications, such as handling money, telling the time and so on.

Direct teaching methods should not be confused with breaking down all learning into small steps as a matter of course. While there will always be value in such approaches for particular aspects of learning, children with Fragile X respond better and engage more if a complete picture of the learning to be undertaken is presented to them. Moreover, as outlined in a later section, visually presented learning can have significant benefits.



READING

It should not be assumed that because of their speech and language difficulties children with Fragile X will be poor readers. In fact, and as noted earlier, reading and its various sub skills can be a relative strength in Fragile X.

However, perhaps related to the speech and language difficulties, the children do learn better with visually based approaches than with auditory based approaches. For example, learning a sight vocabulary is usually significantly easier than learning phonics.

Reading schemes tend to be more effective the more visually based they are. As if to underline this, children with Fragile X often have a remarkable capacity for learning logos and environmental signs ('Bus Stop', 'Post Office'). This strength is often used as a basis for reading activities and learning. Paired reading has inherent features which make it suitable for Fragile X. In particular, it involves teaching alongside the child and so reduces the risk of uncomfortable eye contact. Teaching alongside is a good general approach to working with Fragile X children.



VISUAL APPROACHES TO LEARNING

For children with Fragile X we recommend that visual approaches are incorporated into teaching methods wherever possible and that visual aspects of activities are exploited.

One of the most commonly used examples is a picture guide to the day's activities which enables the child to see the plan for the day and feel more comfortable in knowing what is in store. Each activity is ticked off or otherwise closed when it is done. In this way, the child also knows where he or she is in the day. This can be a stark example of the difference between auditory and visual presentation for the child with Fragile X: simply telling the child what the day holds will do little more than confuse the child and provoke anxiety. A similar effect is evident in skills learning; the child will have a better chance of success where individual skills are divided into stages and presented visually.



COMPUTERS

Computers are effective learning tools to use with children with Fragile X. They allow for endless repetition of tasks, they are consistent and they have large memories. They give immediate responses, and, especially important for Fragile X, they are not personally threatening.

WORK HABITS

Given their distractibility, children with Fragile X make better progress if what they are to do in order to succeed is both clear to them and obviously attainable within a reasonable time span. There needs to be an immediate end in sight, a point where success can be identified. In thinking about this, it should be remembered that children with Fragile X like to succeed just as much as other children. The problem for them is that it can be that much harder to achieve and that they need more teacher planning and support to get it. Praise is especially important to children with Fragile X, and they respond extraordinarily positively to evidence of their own success. Their problems and sensitivities can be exacerbated if they end up doing little more than practising errors.

Work packages should be designed to achieve teaching goals in a direct manner. For example, if the goal of a particular lesson is neat handwriting, it would be counterproductive to insist that, additionally, all the words are spelled correctly. Spelling, or any other requirement is best dealt with as a separate goal. They can come together when the skills are established.

Teachers often express concern that the children are not doing as much as they should or that a child has not made as much progress as his or her classmates. This is not something to worry about if the teacher is confident the child's attention has been usefully engaged at points during the day. Like all children, those with Fragile X need opportunities to explore for themselves the things they like, in their own way and at their own pace.

GIRLS AND BOYS

Girls are usually more mildly affected than boys. Nevertheless, they can share all the characteristics noted earlier. In addition they are more likely to show particular social, emotional and communication difficulties related to,

- Organising their thoughts, planning ahead and shifting from one topic to another.
- Extreme shyness and anxiety in social situations, especially unfamiliar ones.
- Oversensitivity to perceived rejection or criticism.
- Difficulty picking up social signals - verbal or otherwise - when trying to join in, making it difficult for them to make friends despite an eagerness to do so.
- Difficulty seeing the consequences of their own social behaviours.

In many respects girls present extra special needs within Fragile X. The main issue for teachers is that awareness of these sensitivities needs to be taken into account in curriculum and other work.



THE WIDER WORLD

The longer term aim of education is to establish for children a place in society where they can be valued and feel themselves to be worthwhile. It is important not to lose this perspective as we design curricula and develop priority goals for the learning for individual children.

If it is true that no child ever had learning difficulties except in the presence of a curriculum, then it follows that the nature of adult planning and priorities are a vital consideration in teaching children with Fragile X.





FURTHER INFORMATION ABOUT FRAGILE X

If you would like more information please contact the Fragile X Society (full details given on the back cover of this booklet).

The Fragile X Society has a number of publications describing specific aspects of Fragile X including:

- Educational approaches
- Speech and language
- Behaviour
- Adulthood
- Genetics of Fragile X

The Society's Booklets:

- Fragile X : An Introduction
- Fragile X : An Introduction to Educational Needs
- Fragile X : Education and Severe Learning Difficulties

are available FREE to Fragile X families, carers and individual professional enquirers.

The Society's Video

- "Fragile X Syndrome"
awarded the British Medical Association's
Bronze Award for Educational Merit
- explains the features and behaviours of Fragile X
- gives examples of intervention and teaching strategies
- explains the genetics and inheritance patterns of Fragile X
- discusses the benefits and implications of diagnosis

This 20 minute documentary video costs £5 (inc. p&p) from the Society.

THE FRAGILE X SOCIETY

was formed in May 1990 and our aims are to:

- provide support and information to Fragile X families
- raise awareness of Fragile X
- encourage research into all aspects of Fragile X.

HOW DO WE ACHIEVE THESE AIMS?

through our

- National Support and Information Service
- National Helplines and Family "Link" members
- Newsletters, publications and video
- Conferences, talks and presentations
- Media campaigning and distribution of literature
- Arranging for our family members to take part in research

MEMBERSHIP OF THE SOCIETY

is FREE to Fragile X families and carers and we welcome as associate members those with a professional interest in Fragile X.

The Fragile X Society

Rood End House

6 Stortford Road

Great Dunmow

Essex

CM6 1DA

Tel: 01371 875100

Family Helpline: 01424 813147

Fax: 01371 859915

Email: info@fragilex.org.uk

Website: www.fragilex.org.uk

Registered Charity No. 1003981