

THE FRAGILE X SOCIETY



**FRAGILE X SYNDROME:
EDUCATION AND SEVERE LEARNING DIFFICULTIES**

INTRODUCTION

The effects of Fragile X range from mild to severe learning difficulties, or SLD. This booklet focuses on these severe effects. For a more generalised introduction to Fragile X, readers are advised to see the Fragile X Society's booklet, Fragile X Syndrome: An Introduction to Educational Needs.

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WHAT IS FRAGILE X SYNDROME?

Fragile X Syndrome is the commonest form of inherited mental handicap or learning difficulties. The name comes from the unusual appearance of the X chromosome in those who have the condition. This appearance is due to an abnormally long repeating chemical sequence in the gene which causes Fragile X. If the sequence is long enough, the gene is switched off, and it is this switching off that results in the unique characteristics of Fragile X Syndrome.

Heritability and Diagnosis

Geneticists give a diagnosis of Fragile X if there are more than 200 repeats of the chemical sequence. Fragile X is always inherited but the consequences are highly variable. Since Fragile X invariably affects intellectual ability to some extent, diagnosis is important for the purposes of effective management and intervention.

Identification of affected individuals and carriers has changed dramatically in recent years with the development of reliable genetic testing. Testing is straightforward and relatively quick, and is recommended where there is any doubt at all. Diagnosis brings with it the need to involve and inform extended relatives, however distant.



THE EFFECTS OF FRAGILE X

These range from subtle learning problems with an average IQ, to severe learning difficulties with near total adult dependency. As yet, no link has been established between the size of the change in the gene and the severity of disability. Two individuals, each with the same number of repeats of the chemical sequence can show markedly different educational needs. Assessment of any individual's characteristics and needs can be done only through observation in context.

Children with Fragile X are not themselves physically fragile. Usually they are as robust as other children.

Although relatively few children with Fragile X show severe effects, it has been estimated that as many as one in ten school children who have severe learning difficulties do in fact have Fragile X Syndrome. Although recent research suggests that these estimates need further study, there is little doubt that Fragile X is the predominant single cause of inherited learning difficulties.



SEVERE LEARNING DIFFICULTIES

Children with Fragile X and a severe learning difficulty (SLD) show many of the characteristics of much younger children. This is particularly evident in terms of, for example, language, self help, social interaction and understanding. Most developing children learn these and other skills incidentally. That is, the vast bulk of their progress is not formally, or even informally, taught. It is picked up gradually as they go along, undergoing continual refinement as they learn more and more about the world around them.

With SLD resulting from Fragile X Syndrome, the capacity for this kind of incidental learning is impaired. This is not to say that it is absent altogether, only that the children concerned profit from it to a significantly lesser extent, and at a significantly slower rate.

As noted earlier, the possible disabilities resulting from Fragile X cover a wide range from mild to moderate to severe, and all shades in between so as to form a continuum of educational need. The categories on that continuum are notional in so far as they have no objective definition. Although some children show a fairly uniform set of characteristics, others may show a scatter of widely differing abilities. Even within the category of Severe Learning Difficulties, the spectrum of abilities, needs and strengths includes children and young people who are able to travel and shop without supervision, and others who need to be accompanied and supported at all times. Both situations may exist within one child at different times, and depending on circumstances.

A frequent feature of children severely affected by Fragile X is an incapacity to see the most relevant features of a given situation or task. They are inclined to pick up peripheral aspects almost at random, and then to pursue them exclusively. This is one of the reasons why some children perform badly on tests which are structured to cope with normally developing children.

TYPES OF ASSESSMENT

Effective educational assessment of SLD and Fragile X has five fundamental criteria:

- The aim is to inform the teaching and learning process in relation to the individual child to which it is applied.
- The assessment is related to the context in which the child lives and learns.
- Assessment is an integral part of the teaching and learning cycle.
- Assessment of needs is carried out by the people who have responsibility for meeting them.
- Parents and teachers work together.

Psychometric Tests

These include IQ tests. They present a child with tasks thought to indicate intelligence and then rate them against the scores of non-disabled children. These tests were once used to categorise children for the purposes of type-of-school placement. Psychometric tests do not meet any of the criteria listed above. Use of such tests is not recommended in assessing or intervening in the educational needs of children with Fragile X.

Standardised Tests

These include tests of general development. They aim to measure a child's performance on a set of tasks indicative of average development, and then place that child in terms of a developmental stage. For example, one sixteen year old child with Fragile X who scores the same as an average three-year-old, would be said to be functioning at a three-year-old level. Although these tests contain more relevant tasks than IQ tests, and although they are routinely used outside education, their relevance to the educational process is limited in terms of criteria for effective assessment.

Criterion Referenced Assessment

In this, a teaching and learning programme is established for a child, for example – fastening a zip unaided. Assessment of progress may then be gauged by a separate task related to the main aim which for the example given could be – zipping a jacket on a tabletop. This principle can be applied to a whole range of appropriate learning. The problem it embodies is that by, in effect, determining progress in one area by assessing something different, it removes assessment from the teaching and learning cycle.

Curriculum Referenced Assessment

This functions in the same way as Criterion Referenced Assessment except that it combines assessment with teaching and learning. If, for example, an aim of a particular intervention has been to increase a child's appropriate use of a particular word, then the teacher and parent judge progress in the normal course of events. The key is that the intervention and assessment take place in the same context. This is the approach recommended for children with Fragile X and Severe Learning Difficulties.



A DEVELOPMENTAL CURRICULUM

Many of the educational needs of children with Fragile X revolve around three main headings:

- **Developmental Progress** This is what, in normal circumstances, occurs mainly automatically and naturally through contact with the world around. By and large, developing children learn, for example, to talk, walk, understand, dress, eat and so on with relatively little in the way of deliberate instruction. For non-disabled children, such progress is largely incidentally acquired. For children with Fragile X and SLD, this capacity to interpret and to learn will be significantly impaired.
- **Life Skills** These are artificial in the sense that they are to do with the structures of modern life. Learning to use a toaster or a computer keyboard is not something that would develop mainly naturally. With non-disabled children, although a greater degree of adult input is required, they are still able to speed the process of learning by drawing on their developmental strengths. In Fragile X that input needs to be greater still, and to be planned, systematic and protracted.
- **Social and Behavioural Development** Young children think the world revolves around them. They make demands and make their wants known without much concern for the expectations of, or constraints upon others. As children get older, they learn that in order to get what they want, they have to conform to adult expectations and a set of social rules. If children's demands are met without their learning to conform, they will become increasingly unwelcome in any social group, and less efficient at getting what they want. It is a mistake to regard a sixteen-year old with Fragile X and SLD simply as an older version of a young child who has not yet learned how to behave towards others in order to have their needs met. It is also wrong to think the sixteen-year old incapable of learning different behaviour. They do learn social behaviour, but more slowly and in a different way.

A **Developmental Curriculum** is a device intended to teach deliberately the things which most non-disabled children learn incidentally. The only way to find out if any particular child with Fragile X and SLD will learn a particular skill or behaviour is to try to

teach it to him or her.

If, however, they do not learn it, that doesn't mean they are incapable. It might have been inappropriately taught or assessed, or the building blocks necessary for learning were absent. Some skills are sequential and should be learned in the right order. Many children will need to practise skills, or reinforce them with different materials, environments and companions. They may need to rehearse social skills and language for meeting people or shopping, before tackling this in public. The transferability of skills should also be emphasised; fetching one's coat or tidying up applies equally at home, school or visiting friends.



SOME KEY COMPONENTS OF A DEVELOPMENTAL CURRICULUM FOR FRAGILE X AND SLD

Early Development

- Attention Control
- Motor Imitation
- Sound Imitation

Self Help

- Drinking And Eating
- Personal Hygiene
- Dressing
- Leisure Activities
- Shopping
- Making Food

Life Skills

- Personal Safety
- Social and Personal Relationships
- Using Domestic Electrical Equipment
- Transport - road sense, maps, buses, trains
- Telephone, public and private, and emergency services
- Warning and other notices
- Newspapers
- Holidays

These are only a sample of the many skills which any individual child may need. They do not represent a universal curriculum which should be taught to all children with Fragile X. Rather they are a set of core building blocks which may appear in a child's learning programme, depending on that child's needs.

Movement Co-ordination

- Seeing
- Fine Movement
- Big Movements

Communication and Language

- Non-Verbal Communication
- Auditory discrimination
- Understanding Speech
- Using Speech

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.

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