

## Fragile X

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Fragile X is a genetic disorder which can cause a range of intellectual, behavioural, physical and emotional difficulties, however the severity can vary greatly between people with the condition. Fragile X is the single most common inherited cause of intellectual impairment. It is also the most common single gene cause of autism.

Fragile X is an inherited disorder caused by a full mutation in the gene FMR1 which makes protein. This gene was discovered by scientists in 1991 and a DNA based test to diagnose fragile X was developed in 1992. This test is quite accurate, and can detect both carriers and fully-affected individuals.

The symptoms of fragile X can be quite subtle in young children but may include:

- intellectual impairment, ranging from learning disabilities to profound
- attention deficit and hyperactivity
- anxiety and unstable mood
- autistic-like behaviours
- long face, large ears, flat feet and
- hyper-extensible joints, especially fingers.

Both emotional and behavioural problems may also be evident in people who have fragile X.

### **Effects on Developmental Areas**

#### **Social and Emotional Development**

- May experience delays in learning skills of sharing, turn-taking and playing by the rules
- May exhibit frustrations through aggressive behaviours
- May become overly affectionate
- Child may develop self help skills more slowly and need reminders for toileting
- May exhibit immature eating habits

### Physical development

- May be hesitant to explore due to lack of confidence in their own skills
- May experience some sensory defensive behaviours
- May have delayed gross motor and fine motor co-ordination
- May have elongated face, long and prominent ears, high palate, flat feet, hyper-extensible finger joints and soft velvet like skin
- May have seizures
- May have a hypersensitivity to sensory stimuli

### Language and Communication Development

- May have difficulty in communicating
- May have language delay
- May not readily *understand* or *use* appropriate forms of communication including verbal language, body language, facial expression, tone of voice and gestures
- May have difficulty in understanding complex or abstract concepts
- May be unable to communicate wants, express concerns or fears, or be able to answer questions reliably

### Cognitive

- May have learning difficulties
- May not stay long at activities due to low concentration span
- May require instructions, directions etc. to be repeated 2 or 3 times and requires some time to process before responding or acting
- May have delays in skills of concentration, memory and ability to generalise
- May have difficulty understanding concepts of turn taking, sharing, how to enter into play situations

### Fragile X Inclusion Strategies

Each child diagnosed with **Fragile X** will be different and individual. It is important to gain information from the parents as to what characteristics of **Fragile X** their child displays. It is important to work closely with the parents as well as any additional support specialists e.g. therapists who may be involved with the child. It is also important to gain an understanding from the parent as to what is the most important aspect of their child attending your service. What is it that parents hope to gain from using your service? The following inclusion strategies are just some examples which may be applied to support the inclusion process. This list is only the start and it is dependant on a variety of factors such as environment, length of time child is in care, child's interest, likes, dislikes and skills already achieved. The strategies are divided into developmental areas however some strategies overlap and assist in a variety of developmental areas.

#### **Social development**

##### **Social and Emotional Development**

- Assign children set jobs or tasks to take responsibility for such as watering plants, setting up cups/bowls for morning tea etc. Keep the jobs consistent and routine to enable children to plan for these in their day.
- Encourage participation rather than competition during activities.
- Dramatise familiar stories with children using props including costumes, felt board stories etc.
- Consistency is important. If the child finds that every time they are given that direction, the same response is expected, or that every time they react in that way, the same consequence follows, they will learn the appropriate behaviour far more quickly.
- Encourage the child to take some responsibility for their actions.
- Give the child clear information about what is going to be happening, and what will be expected of the child in a new situation giving the child the chance to prepare themselves for the event and to work out how to behave.
- Forewarn the child when an activity has to be packed up or it is someone else's turn. This will allow the child to prepare for and deal with finishing more appropriately.
- Watch for triggers that may increase inappropriate behaviours.

### Physical Development

- Keep things in the same place to assist child to be able to move from one place to another. If you change the environment walk and talk this through with the child.
- Provide finger plays to encourage the use of both hands in a controlled manner as well as developing fine motor skills.
- Provide lock boxes and musical boards to promote finger and wrist movement and rotation.
- When setting up collage table provide clear defined areas for different materials in boxes.
- Provide gross motor activities that enable success for the child e.g. adapting the obstacle course that can accommodate the child's abilities but at the same time provide some challenge.

### Language and Communication Development

- Encourage children to talk about what they are doing by asking open ended questions several times throughout the day. Also, ask children to tell you what you or others are doing.
- Provide positive reinforcement during play times and encourage dramatic play.
- Provide clear directions and break into manageable steps in sequence.
- Use picture or object system for organising the daily events and encourage children to sequence - establish a comfortable routine.
- Provide advance warning if routine is going to change.
- Maintain a calm approach to ensure situations are more likely to be successfully resolved. Being calm when dealing with a child when they are feeling anxious, upset or frightened is more likely to reassure the child, and settle them down more quickly.
- Limit interactive language to the fewest number of information-carrying words at a time. If the instruction contains a number of steps, give one step at a time, wait for the child to comply, and then give the next part.
- Use positive statements by telling the child what you do want them to do as the child may genuinely not know what else to do and the negative statement is of no help to them.
- Allow one person only to give verbal information/directions at a time. If the person requires help to make the child understand or comply, the other adults can help by possibly physically patterning or prompting the child e.g. modelling the desired behaviour in an obvious way.
- Carry out demanding activities at the time the child functions best e.g. first thing in the morning or after lunch.
- Organise the physical setting to aid the child as much as possible.

### Cognitive

- Avoid generalisation.
- Use concrete representation to enhance concept development.
- Focus and reinforce relevant information, aspects, attributes and characteristics.
- Allow the child time to complete tasks and practice skills at own pace.
- Acknowledge level of achievement e.g. “You have placed that piece in the puzzle, well done” rather than just “Good job”.

### Reference

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Umansky, W. and Hooper, S. (1998) **Young Children with Special Needs** Third Edition New Jersey, USA:Prentice-Hall

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