

The Fragile X Society

Behaviour challenges in children and adults with fragile X syndrome: What help is available?

Presentation by Dr Angela Hassiotis, Senior Lecturer and Honorary Consultant Psychiatrist, Royal Free & University College Medical School, Camden Learning Disabilities Service to the Fragile X Society's National Family Conference, June 2006

Email: a.hassiotis@ucl.ac.uk

Fragile X is the commonest inherited cause of learning disabilities and affects 1 in 3600 males and 1 in 4000-6000 females. This syndrome is associated with behavioural challenges and other mental disorders that can precipitate and perpetuate those behavioural problems. In this article, a version of which I presented at the Fragile X Society annual conference in June 2005, I will talk about the management of those disorders and describe the process by which parents can access help for their child. I will also refer to recent Government policy, which aims to improve services for people with learning disabilities including those with fragile X.

Behaviour challenges

The behavioural challenges that can present in both children and adults with fragile X are due to a host of underlying causes such as the developmental stage the person with fragile X is at, his or her level of learning disability, the presence of the syndrome itself as well as additional disorders and the capacity to learn from and respond appropriately to experiences.

Fragile X can be present alongside other mental disorders such as

- Autism
- Hyperactivity (attention deficit hyperactivity disorder (ADHD))
- Anxiety

The syndrome is also associated with recognised deficits in

- Social interaction
- Speech production and use
- Cognitive skills
- Sensory motor skills.

Autism and fragile X syndrome

2-6% of all children diagnosed with autism, have a fragile X gene mutation. Approximately one-third of all children diagnosed with fragile X also have autism. Lastly, fragile X is the commonest known cause of autism.

The typical features of fragile X i.e. hand biting, hand flapping, poor eye contact, shyness, and social anxiety are probably related to sensory hyperarousal and are referred to as autistic-like features because they can be seen in individuals who have autism but not

fragile X. Most children with fragile X, however, are interested in social interactions and do not meet the diagnostic criteria for autism.

Attentional problems and fragile X syndrome

Usually, children and adults with the syndrome are inattentive, impulsive and distractible. However, hyperkinetic disorder is no more common than in other children without fragile X.

Cognitive patterns for boys and girls with fragile X

There are distinct cognitive patterns and deficits in male and female children and adults with fragile X.

They appear to have good memory for situations, favourite TV shows, videos and songs and may memorize tone as well as words. They score well on adaptive living items, which indicates good ability to learn self-care and household management skills but score poorly on socialization and communication items as well as on complex problem solving, cause and effect questions, and other abstract tasks. Other areas of deficit are visual-motor and construction tasks such as bead sequencing and block designs.

They show strengths in vocabulary and comprehension. They have good short term memory for visual, meaningful material. These strengths can be the basis for achieving reading, spelling, and writing skills. Weaknesses are seen in "executive functioning", that is complex abilities including planning, attending, sustaining effort, generating problem solving strategies, using feedback, self-monitoring, and shifting responses.

Speech patterns

Most children with fragile X learn to speak (some may need augmentative communication devices, e.g. pictures, sign language, computerized boards, etc.). Girls, who are mildly affected by fragile X have good verbal skills, with no particular disorders in articulation, vocabulary (semantics), or grammar (syntax). Language disorders that may affect these females with fragile X are usually in the area of conversational skills (pragmatics).

Ear infections, sensory motor problems with oral motor weakness, lowered cognitive abilities, and attention and behaviour difficulties in social interactions may combine to affect speech and language.

5. Medical problems

Other medical conditions that can be present in children or adults with fragile X are:

- Gastrointestinal difficulties (reflux)
- Epilepsy (in about 10-30%)
- Joint hypermobility (clumsiness, scoliosis)
- Cardiovascular problems (infections)
- Hearing and vision impairment

Interventions

Services should strive to provide a comprehensive, integrated and multidisciplinary treatment programme based on the bio-psycho-social assessment and management model taking into account not only the individual but also his or her social and support network.

The management should be practical, problem-focused, tailored to the individual and for very young children home-based (i.e. Portage system). Below I will give examples of different interventions and professions that may contribute in implementing them.

Speech and Language Therapy

Speech therapists are integral members of the Paediatric, Child and Adolescent Mental Health or Learning Disability service multidisciplinary teams. Their role is to assess and treat speech deficits and devise communication strategies. They may design goals that are carried out in the classroom, or in the day care, and work alongside a variety of other professionals. Preferably they administer tests or work with the person in a calm, quiet room, and if the child is too young or anxious should be accompanied by a familiar person. Calming strategies may need to precede therapy. These may include exercises, such as wall push-ups or deep pressure massage.

An audiological examination should precede a speech and language assessment to rule out hearing loss. Other assessments include a parent or formal carer interview, an oral-motor examination, testing of both receptive (listening) and expressive (speaking) vocabulary and syntax, and assessment of speech (sound production), voice, and fluency. It is also important to assess how the child or adult interacts with peers or family members.

Helpful communication strategies include use of visual resources across the age-span. For example, pictures of the day's activities in sequence, calendars, pictures of classmates and their weekly jobs, and pictures of vocabulary being taught in the classroom/college. For older children, visual cues can include signs or signals for proper behaviour (e.g., hand over mouth for "quiet mouth"). Pictures and logos provide visual cues for linking language to reading. Dramatization can also help with the comprehension of story sequences and plots.

Sensory Integration and Occupational therapy

Occupational therapists are also part of the multidisciplinary team. They can work with sensory integration disorders, arousal levels, tactile defensiveness, fine motor weaknesses, and oral-motor disorders. The occupational therapist tries to determine what settings are optimal for best performance by the child or adult with fragile X and what alterations can be made in the environment to help. The occupational therapist may observe the individual during transitions from one activity to another, when unexpected events take place and assess posture seating and movement.

Lighting adjustments, noise reduction, or private, calming spaces may prove helpful to a person overwhelmed by too much sensory stimulation. Other distracting techniques include provision of breaks in the day, time in a quiet space, playing a computer game or listening to music or a story on headphones.

Psychological interventions or when problems arise

Another profession that is important in the management of behavioural challenges in children and young persons with fragile X is psychology. Psychologists provide assessments of cognitive ability as well as assessment and management of behavioural and mental disorders. Therefore, they can provide behavioural plans to help children with fragile X to cope with everyday demands of home, school, and community. Psychologists

devise behavioural techniques to deal with specific problems such as poor eye contact, hand flapping, and lack of awareness of social cues, which may cause difficulties in peer interactions, making inclusive educational placements more of a challenge.

Psychologists may work with speech therapists and/or occupational therapists to help implement a specific behavioural programme. For example, in a classroom,

- The child sits near the teacher and away from distractions, short tasks with the opportunity to move around often, visual cues for sequences of events, and interactive lessons, that do not always involve sitting and listening.
- Some unusual circumstances (such as a loud assembly) may call for time in a quiet room, with calming items such as a beanbag chair and a videotape, rather than the scheduled event.

Other psychological interventions may target difficulties within the family system (family therapy) or the individual (counselling, cognitive-behavioural treatment) in order to address emotional disorders or skills training.

Behavioural Treatment of Aggression

Significant aggression such as impulsive hitting occurs in approximately 20-30 % of children and adults with fragile X. It is always important to try and understand the function the behaviour plays in the sequence of an episode. For instance, aggressive behaviour may provide an escape from something that is frightening or threatening. The message is that if one wants to escape a situation and lacks the ability to communicate that need, it works to become aggressive.

A functional behaviour assessment requires observing the behaviour and then collecting data related to the behaviour across different settings.

Proactive behavioural strategies

Those include reduction in the level of environmental noise/sound, provision of nonverbal cues and feedback. It may be necessary to remove the person from stressful events, allow the person to sit in the back section of a room, allow additional processing time, provide calming activities and encourage breaks and "down time".

Pharmacological treatment for aggression

It is paramount to consider the reason for the aggressive behaviour before instigation of pharmacological treatment. The choice of medication depends on the underlying cause. Particular drugs can help certain conditions. For example:

- methylphenidate (Ritalin) is recommended for children (current advice is up to the age of 18 years) who suffer with attention deficits.
- Clonidine could be helpful to hyperarousal to stimuli.

- Antidepressants can treat excessive anxiety or obsessive-compulsive symptoms, mood stabilizers such as lithium or carbamazepine (also an antiepileptic drug) are useful if there is an underlying mood disorder (i.e. bipolar affective disorder) and antipsychotic medication (i.e. risperidone) is indicated if aggression and hyperactivity is the result of psychosis.

Social interventions

Additional services that are necessary components of the overall management of a child or adult with fragile X are:

- Respite
- Education
- Vocational training
- Life planning

Research

Recent research of service use of a sample of adolescents with any type of learning disabilities showed that a significant minority visit their GP frequently to ask for support regarding the child's behavioural challenges. Over half of all recognised cases (55%) and a third of non cases (38%) show such challenges.

About a third of young persons with a learning disability and mental disorders receive medication. Over two thirds (76%) of those who have a diagnosed mental disorder receive ongoing health and social services input. However, treatment plans are piecemeal and professionals other than psychiatry and nursing are scarce. This snapshot shows that problems may start early, are ongoing and that for a minority of families recognition of the problem is essential in order to receive appropriate help.

Gaps often mentioned in service provision are:

- limited respite options,
- poor community support,
- scarce employment opportunities for the young person,
- lack of flexibility of support
- exclusion from school which is used as a remedy for inappropriate behaviour

Transition

Transition is defined as the time from 14 to 18 years old when social services have a statutory obligation to assess the young person with learning disabilities needs to ensure a planned transfer to the adult learning disabilities services. This is perceived as a complicated process that few parents can master and may create anxiety about the future of the young person particularly where local resources are overstretched or the planning phase has not been done properly. Anecdotal information indicates that there is considerable confusion and lack of clarity about each service funding responsibilities.

Policy context

Two main government policies address the issues of inequality, service gaps and personalised planning. These are "Valuing People" (Department of Health, 2001) and the "National Service Framework for Children, Young People and Maternity Services" (NSF,

Department of Health, 2004). The policies provide guidance about service development and an overhaul of the conceptual framework for service provision to both children and adults with learning disabilities of any aetiology. An aspect of the NSF for children is the development of mental health teams specialising in various childhood problems such as mental health of children with special needs.

The implementation group associated with Valuing People has recognised that the risk to develop mental disorders is greater in those with learning disabilities. In order to promote awareness of emotional problems in young persons themselves and their parents it has produced two leaflets, which describe the symptoms of common disorders such as anxiety and depression. The leaflets called “all about feeling down” and “meeting the emotional needs of young people with learning disabilities”, are directed to the person with learning disabilities and their family or formal carers respectively.

What happens as the child grows up?

Many parents as well as professionals find the maze of service provision particularly difficult to navigate. Service structure and interface can be hard to negotiate and not infrequently parents may be under the impression that they have to look after their child without any external support. Regardless of the reason for the child’s learning disability there are common principles, which underpin the treatment approach to many of the problems that have already been discussed in this article.

As a clinician working with young people around the time of their transition to adult learning disabilities services and their families it is my practice to advise that they should become familiar with their local services; find out what they are and where they are based. Most services have accessible leaflets explaining their roles and they can translate this information to languages other than English.

Services are funded in many different ways and although they appear to offer similar input, access may be through different pathways. Most services have eligibility criteria, which means that they can accept referrals for only one group of individuals but not others. There are several personal stories whereby parents have succeeded in helping their adult son or daughter with a learning disability to live a more independent and fulfilling life within the limitations of his or her disability. It is equally true, though, that there is a “postcode lottery” when it comes to what might be on offer in many parts of the UK. Many services these days try to find new models of day care away from the traditional day centre, although some form of such provision may still be necessary for those with severe learning disability.

The internet, the Royal College of Psychiatrists and syndrome societies (The Fragile X Society) are good sources of information and facts which can then be discussed with the professionals involved in a person’s care plan. Such resources are important in suggesting ways in which to deal with behavioural challenges, e.g. sleeping problems, some maintain a help-line or may have other useful links.

Similarly, it is important to check the website of the Department of Health for new legislation and guidance on policies. For example, there are new bills for mental capacity

(i.e. ability to consent to treatment) or the Mental Health Act. Outcomes of social inspection reports on many residential providers are now available online.

Parents should be aware of their entitlements, e.g. direct payments, respite facilities, other supports that can ensure that caring after a loved one does not become a burden. Research indicates that parents of children with long term problems, for instance tourette syndrome, showed significantly increased caregiver burden (Cooper et al, 2003).

The community learning disabilities services (CLDS)

Most areas across the UK have such teams (CLDS). They are multiprofessional although the “postcode lottery” means that some are better staffed than others. Counselling is one of the interventions not readily available in many teams. They are part of Primary Care Trusts, Mental Health Trusts or stand alone learning disability trusts (very few of those still in operation UK wide).

CLDS receive referrals from many sources (GPs, paediatricians, other psychiatrists) including self referrals.

CLDS assess and treat adults aged 16 years and over with a learning disability in the community. Professionals work with individuals at various settings, e.g. home, day centre, school/college depending on what the issues are. The professionals within the CLDS can help not only the individual but also their families and formal carers. CLDS deal with many problems such as behavioural challenges and mental disorders, communication and speech problems, assessment for diagnostic purposes, motor disorders, problems in social skills and psychological difficulties. The service provides staff training and support on many topics such as treatment of epilepsy, administration of medication, how to deal with sexuality, coping strategies for people with autism.

In some areas CLDS may also employ community support workers whose role is to support the individual in activities of daily living and increase participation in the community and recreational opportunities. Health professionals and care managers (used to be called social workers) are more often than not based in the same locality.

I would like to conclude the article with the words of Peter Szatmari, a Canadian Child Psychiatrist and expert in autism and developmental disabilities. The extract is from his book “a mind apart”.

“...There are many interventions that have proven to be helpful, and many of those involve parents working with professionals to facilitate social and communicative development. Above all, acceptance of the diagnosis will call forth from parents a career as forceful advocates for their child, for they will have to advocate with service providers, with teachers and with members of the community for more services, for better understanding and for greater inclusion in the community”.

References and useful websites

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Szatmari P (2004). A mind apart: understanding children with autism and asperger syndrom. The Guilford Press, NY, pp183

Further reading available from The Fragile X Society

Treatments and Services for Individuals with fragile X and their families by Dr Jeremy Turk, 1997

Practical Management of Psychiatric, Psychological and Behavioural Problems in young people with fragile X syndrome by Dr Jeremy Turk, 1994

Modifying Behaviour – Reducing the Undesirable & Enhancing the Desirable by Dr Jeremy Turk and D Smith, 2000

Using Medication for Problem Behaviours in Individuals with Fragile X Syndrome by Dr Jeremy Turk, 1999

Problems of Adulthood in Fragile X Syndrome – a Mental Health Perspective by Dr Andrew Levitas, 2002

Diagnosis and Pharmacological Treatment of Psychiatric Disorders in Adolescents and Adults with fragile X syndrome by Dr Andrew Levitas, 1994

Fragile X Syndrome – Teaching Communication Skills by Catherine Taylor, 2004

Helping your child to make sense of their world by Claire Wolstencroft (workshop) 2004

Living with Fragile X – Sensory Integration Therapy (conference) 1999 & 2000

After School – What Next? (conference) 1996

Transition from School to Local FE College (conference) 2005

Further Education in mainstream and specialist colleges (workshop) 2004

Adult Living (conference) 2000 & 2001

Adult Life – the experience of daily life of nine gentlemen, 2004

Girls & Young Women with fragile X (workshop) 2004

Educating Children with Fragile X Syndrome edited by Dr Denise Dew Hughes,
RoutledgeFalmer, London

The Fragile X Society

Rood End House

6 Stortford Road

Great Dunmow

Essex

CM6 1DA

Tel: 01371 875100

Email: info@fragilex.org.uk

Website: www.fragilex.org.uk

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