The Fragile X Society

Fragile X Behaviour
– Reducing the Undesirable and Enhancing the Desirable

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Part 1: Clinical aspects of fragile X syndrome

The main issues to be covered in the first part of this article are the psychological aspects of fragile X syndrome. However, physical aspects were the first features to be considered when fragile X syndrome was initially recognised and researched. This is often the case in genetic conditions. But physical features are very variable, they are not unique to fragile X syndrome and are not necessary for a diagnosis of fragile X syndrome. In addition they are present in other genetic conditions and present in many members of the general population. If physical features are present, then they may be important indicators that fragile X syndrome is the cause of the individual's difficulties. They may also have important medical implications. For example, there is a widespread problem with connective tissues in the bodies of people with fragile X syndrome. These, if you like, are the body's "glue" which holds joints together and sticks muscles on to bone. As a consequence many individuals with fragile X syndrome are double-jointed with lax ligaments. But there are positive aspects as well. The skin is said to be very high quality. Soft and velvety are the terms that have been described. This is in contrast to some other genetic conditions, for example Prader-Willi syndrome, where the skin is quite delicate. After puberty in young men with fragile X syndrome the testicles can be quite enlarged. Of more importance, clinically, is the fact that occasionally there may be a heart murmur because of problems with one of the heart's valves being a bit leaky; or there may be a stretching or widening of the main tube carrying blood away from the heart – the aorta. This is one reason why anybody with fragile X syndrome should have a very careful medical examination to exclude these possibilities. As is often the case in genetic conditions, there can be unusual creases, characteristically on the hands. One commonly finds a single crease across the palm. One can also observe them on the feet as well - usually in the form of a mid-line crease going down the sole of the foot from between the first and second toe, a so-called plantar crease. Plantar means the soles of the feet as in plantar warts. Individuals with fragile X syndrome have been said to have largish heads. In reality head size is very

variable. We published a study recently (Turk & Patton, 2000) in which we argued that if you compare people with fragile X syndrome with other individuals who have learning disabilities their heads are not significantly larger. Some other conditions, for example Down's Syndrome, may be associated with having slightly smaller heads. It depends which groups of people you compare. More commonly seen is the largish jaw, which can be quite pronounced. This can have an effect on language difficulties. Protruding ears are also common. The nasal bridge can be quite long and flattened and the palate inside the mouth is often quite high-arched. This is a further reason why people with fragile X syndrome can have expressive language difficulties. But the important thing is that you wouldn't usually know just by looking at somebody that they have fragile X syndrome. The physical features aren't that useful diagnostically. Physical features often become more pronounced in older individuals. There are still many individuals undiagnosed despite this.

It was approximately three decades ago that people began to recognise that physical features are not that useful in diagnosing fragile X syndrome. At that time it was also becoming acceptable to believe that having a particular genetic cause for your learning disability could influence the development of emotional and behavioural problems, as well as intelligence. Since then there has been an enormous mushrooming of research in this field in many conditions, but perhaps in particular fragile X syndrome. It helps to classify information under various headings. Firstly one can consider intellectual functioning followed by speech and language. Attentional deficits are common. When they are combined with marked restlessness, fidgetiness, impulsive behaviour and a ready distractibility they may demonstrate that the individual has attention deficit-hyperactivity disorder (ADHD). Social impairments if combined with marked speech and language problems and obsessional/compulsive tendencies may suggest the presence of an autistic spectrum disorder.

Intellectual functioning

Intellectual functioning can be extremely variable. It is usually in the mild to moderate learning disability range - a rough IQ estimate of about 35 to 70. But just because you have a genetic cause for your difficulties it doesn't mean that you will fall in to the learning disability range. It may be that it's not so much intellectual functioning as other areas of mental functioning that are most problematic. Of more importance than the exact level of intellectual functioning is the "profile" of intellectual strengths and needs. Individuals with fragile X syndrome do not have the same level of skills and abilities across the spectrum of intellectual attributes. Speech and language skills can be relative strengths. However there is a risk that individuals may appear more able than they are. This can produce problems in that an individual is being expected to undertake tasks at school or college, which are way beyond their abilities. It's not surprising that the individual feels very upset about this and perhaps shows distress in the only way possible - behaviourally. They then run the risk of being labelled as having "challenging behaviour" - the focus being on snuffing out the behaviour rather than trying to understand what has been causing it and what message the individual is trying to convey. The main difficulties are usually in the area of what we call performance skills, in particular number work (numeracy), and the area of visuo-spatial abilities. These include, for example, the ability to find the way around places. Intelligence is not a unitary concept. There are many different components to it and any area of intellectual functioning may be delayed to any degree.

Much research has shown that young children with fragile X develop intellectually at the same rate as other children - so that whilst they are behind most children of the same age

intellectually they keep pace with them, develop at the same rate and stay the same degree behind. However as they approach adolescence and puberty the discrepancy between intellectual functioning in people with fragile X syndrome and others widens. This is because of particular problems with the ability to process sequences of information. There are two ways that humans deal with incoming information. There is the early way that children usually use which is called "simultaneous information processing". You look at a whole scene and you say 'that's a farmyard'. Or you see a square with cars and a road, for example, and you say 'it's a town'. As you grow up you increasingly have to use "sequential information processing". This is processing information in sequences, for example the teacher who says to the class "right, class, first put your paints away, then wash your hands, then help me lay the table and sit down for tea". A better teaching approach might be to break down the sequence of commands into a series of specific instructions. Another much researched area relates to "executive functions". These are to do with the ability to execute psychological and intellectual functions, to plan ahead, to deal with sequences of information, to organise information, to think about various different possibilities and solutions of problems, and indeed to be able to switch fluently from one strategy or tactic to another. These are not skills that relate specifically to low intellectual functioning. They are a set of mental abilities that allow one to approach problems fluently. They can be faulty in individuals with any level of intellectual ability.

Speech & language

Speech and language, what we say, how we say it and indeed the degree of gesture and body language we use are largely culturally determined. They are dependent on our upbringing, our background, where we live and whom we live with. Yet there are compelling reports of particular language styles in people with fragile X syndrome. Words like "jocular" have been used to describe the humorous quality of language. "Litanic-like" refers to the up and down swings of pitch. Language can be very repetitive. Some aspects are reminiscent of language heard in people with autism. This was one reason why people initially and erroneously thought there was a specific and direct link between fragile X syndrome and autism. "Cluttering" describes a combination of rapid speech with a lot of rhythm problems ("dysrhythmia"). Individuals know what to say, understand the nature of social communication and want to relate and talk. It's as if the brain's processing of information is working too fast for the articulatory apparatus so the words fall over themselves, resulting in mumbled and mixed up speech. This emphasises the need for careful speech and language input. As part of our London study of boys with fragile X syndrome, we found that language as described above was very common in fragile X syndrome, occurring in about 50% of individuals. However there were only about 10% of people with learning disability generally of the same age that showed this trait. No people with Down's Syndrome demonstrated it.

Social functioning & autistic features

The area that has attracted most research has been the postulated link between fragile X syndrome and autism. There are important qualitative differences between behaviour shown by most people with fragile X and behaviour shown by most people with more typical autism (Turk & Graham 1997). For example, autism is usually associated with social indifference – lack of interest in other people. Fragile X syndrome individuals are usually socially aware, socially sensitive and "tuned in". Social anxiety is more common in people with fragile X syndrome. Eye contact difficulties in autism are usually in the form of indifference to direct eye contact. In fragile X syndrome there is usually "gaze

aversion". Individuals often have to avert their eyes and even head and body on greeting another person. There is often a general sensory sensitivity in fragile X syndrome. Whether it's what you see, hear, taste, feel or smell the sensation can be overwhelming and difficult to deal with, particularly with multiple incoming stimuli. Why the most common manifestation should be gaze aversion remains unclear. Practically, it is important not to insist on a person with fragile X syndrome maintaining eye contact with you. Useful classroom strategies include the use of booths, so pupils don't have inadvertent eye contact with others, and tutoring from the side or behind. The individual can then tolerate the proximity while understanding the nature of social interaction, but doesn't have to deal with aversive eye contact.

Self-injury can occur in fragile X syndrome, usually biting the base of the thumb in response to anxiety or excitement. Delays in imitative or symbolic play are common. These relate to problems with understanding the "make-believe" components of play. Such skills usually develop over time, unlike autism where there is often a lack of development of these abilities. Hand flapping is the other extremely common behaviour seen in people with fragile X syndrome. In the London study 50% of boys with learning disabilities flapped their hands. In contrast 90% of boys with fragile X syndrome did so. Again it seems to be a response to anxiety and excitement - a way of physically showing your internal feelings. Much confusion persists as to the overlap between fragile X syndrome and autism. There are certain social and language tendencies, which characterise people with fragile X syndrome. Echolalia (repeating what somebody else has just said) and repetitive speech are both commonly heard. Yet there is good understanding of facial expression. We have shown that people with fragile X syndrome are generally good at understanding facial expressions and are good at understanding how people are feeling from the look on their face – skills that are rare in people with autism (Turk & Cornish 1998). People with fragile X syndrome are usually friendly and sociable, even though they may be shy or socially anxious.

If you ask what proportion of people with autism have fragile X syndrome, the answer is probably not many, no more than 2-3%, but a lot more than most known causes of autism. Anybody with an autistic spectrum disorder should be tested for fragile X syndrome. A substantial minority of people with fragile X syndrome have autism. Autism does occur in fragile X syndrome, certainly more than in the general population. A much higher proportion of people with fragile X syndrome don't have autism but show an array of communicatory and repetitive tendencies which may be described as autistic-like. However these individuals are friendly and sociable.

Attentional deficits & overactivity

There have been descriptions going back to the mid 1970's of people with fragile X syndrome having poor concentration spans and being restless, fidgety, impulsive, easily distracted, and overactive. Initially these tendencies were attributed solely to the learning disability and quality of upbringing. However, there is no doubt that people with fragile X syndrome do have more than their fair share of difficulties with inattentiveness and overactivity. In the London study (Turk 1998) there was no difference found between boys with fragile X syndrome and boys with learning disabilities generally in terms of rates of strictly defined hyperkinetic disorder. However boys with fragile X syndrome were found to have poorer attention spans and worse restlessness and fidgetiness than boys with learning disability generally. Thus boys with fragile X syndrome can be inattentive, restless and fidgety without being more overactive than other boys with learning disability

are. The North American term for this is Attention Deficit-Hyperactivity Disorder, predominantly inattentive type. It is treatable medically, psychologically, educationally and socially.

Females with fragile X

Females with fragile X syndrome have been less intensively studied. This is partly because intellectual function is generally higher than in boys and men. Also, they look very normal. However research has shown that females with fragile X syndrome can show physical features similar to those seen in males. These physical features become more pronounced with increasing degrees of intellectual impairment. Approximately a third of females who carry a full mutation have learning disabilities. Females with just a premutation often report more subtle versions of the intellectual, emotional and behavioural problems known to be associated with fragile X syndrome. Such features include shyness, social anxiety, concentration difficulties, numeracy problems and the above-mentioned executive function impairments. Anecdotally there are two aspects that really impress me about girls and women who have fragile X syndrome. The first is their enormous staying power – they will keep on going until they succeed. Perseverance is indeed a virtue. The second is just how resilient they are to the emotional knocks that they inevitably get through their lives. Sadly school is often still an unnecessarily negative experience. There is still frighteningly little knowledge about fragile X syndrome amongst educationalists (York et al., 1999).

Premutation carriers

We are becoming aware that individuals with just premutations can frequently show traits characteristic of fragile X syndrome. We have undertaken detailed evaluations psychiatrically, psychologically and speech and language-wise on boys and young men with premutations (Aziz et al., 1998). Many of them have low levels of intellectual functioning, language difficulties, autistic spectrum disorders and attentional deficits. Most strikingly almost all of them have difficulties with the social use of language; the ability to use language meaningfully in a social context - to get on with and relate to other people, the so-called semantic and pragmatic aspects. Many of the individuals we studied have impaired social use of language in the form of difficulties with their speech being intelligible. There is also often a discrepancy between their ability to express themselves and their ability to understand what's being said to them - receptive aspects. This can lead to the person sounding articulate while not being able to process and work through what has been said to them. Most strikingly some physical features are present in particular enlarged testicular volume. Further research is needed but it seems clear that many individuals with even small premutations experience difficulties similar to those of individuals with full mutations.

Frax E

Individuals with Frax E, if anything, look even more normal than many young people with Frax A – the technical name for what we usually call fragile X syndrome. Frax E is caused by a similar genetic problem even closer to the tip of the X chromosome. Few people with Frax E have been identified. This is partly because the test is only very occasionally done. It is also because Frax E is almost certainly rarer than Frax A. We don't know quite how rare. It used to be labelled as a mild form of Frax A (Barnicoat et al., 1997). We are currently building a clinical series of people with Frax E. Early indications are that there are a wide range and severity of difficulties (Turk et al., 2000). You can be mildly affected with perhaps low average intellectual functioning, mild speech and language difficulties

and concentration problems. Conversely you can have severe learning difficulties, autism and hyperactivity. Frax E should be suspected in anybody with unexplained intellectual, social, language or obsessional tendencies, particularly with a family history of boys and men being affected more than girls and women, when the test for Frax A has proved negative.

Practical implications

Physical features can be very variable (Turk & Patton 2000). Large head circumference is not useful in making a diagnosis. Vision and hearing problems are frequent. When present they should be diagnosed and treated vigorously. However they are not diagnostic of fragile X syndrome. Psychologically there may be marked inattentiveness and restlessness, even in the absence of overactivity. Shyness and social anxiety are common, as is gaze aversion. But people with fragile X syndrome by and large lack the more profound psychological deficits often seen in autism, namely difficulty in understanding faces and emotions, and difficulty in understanding that people have different perspectives and experiences - so-called theory of mind (Garner et al., 1999).

Educationally, we need to make teachers aware of number difficulty, visio-spatial problems, sequential information processing difficulties, social anxiety as a cause of behavioural disturbance, and gaze aversion. Expressive language may be cluttered and understanding may lag behind. Anxiety, often social, is a potent trigger for challenging behaviour. Often it is communicatory and ritualistic/obsessional aspects rather than social and symbolic problems that characterise people with fragile X syndrome.

Questions

Q. Can you have fragile X and Fragile E or are they two totally separate things?
A. No, you have one or the other. Two different genes cause them. The genes are very close to each other on the X chromosome. They looked very similar on the old chromosome test. New DNA tests can differentiate them. The term fragile X syndrome is usually used to describe Frax A. Frax A is more common than Frax E but they can both cause substantial developmental difficulties.
Q. If you've got a positive test for Fragile A, would they look for Fragile E?
A. No, it's a different test. The test for Frax E is only rarely undertaken. It may be worth doing if a family present with individuals affected in ways which suggest Frax A, yet all the tests come back negative.

Part 2: Enhancing Desirable Behaviour and Reducing Undesirable Behaviour

It is very difficult if not impossible to write a recipe book along the lines of treatment for challenging behaviours. It is important to adopt a particular framework, a structure, an approach to how you start thinking about behavioural difficulties and their possible causes. These are usually multiple rather than single. There are many different possible causes of behavioural disturbance, not just in people with fragile X syndrome but in human beings generally. Firstly there are "constitutional" factors. Is there something like a behavioural phenotype present? Is there fragile X syndrome or indeed one of the increasing number of genetic conditions that we now recognise where there are particular profiles of developmental and behavioural problems and vulnerability. There are also aspects active in all of us, in particular our temperament. Some people are more active and impulsive than

others, or more sociable, more motivated and so on. There are important interactions between these constitutional factors and your experiences. A further important biological cause of behavioural difficulties is prescribed medications. Also, epilepsy can influence behaviour, as can many other illnesses. Much behaviour may be appropriate for the developmental stage that the individual has reached. How you appraise others, situations and your own experiences can be very important as well.

Important preventative activities include early identification and early intervention. These involve early assessment of the individual's profile of strengths and needs, and the early development of a multi-disciplinary intervention support package. Speech and language therapy assessment can be crucial. One of the most elegant interventions is to help an individual to communicate, by whatever means, thereby relieving frustration and diminishing consequent anger and disturbed behaviour. Occupational therapy is also often invaluable in terms of dealing with motor problems as well as overwhelming sensory experiences. The importance of an appropriate school environment should never be underestimated. There should ideally be sufficient suitably trained and experienced staff, as well as a consistent and predictable routine with minimum of changes and distractions.

In terms of helping autistic features, the most important aspect is appropriate educational input. Occasionally a highly specialist school for pupils with autism is needed. Very often having staff with the correct specialist awareness and training will suffice. Sometimes judicious use of medication can complement psychological, educational and social interventions. This is most frequently used for hyperactivity and inattentiveness. There are also useful medications available for obsessive-compulsive problems, depression, severe sleep disturbance and sometimes severe self-injury.

Step by step to a behavioural programme

Define objectively which behaviours are problematic. Something you can't see and witness, although it may be real, is difficult to rate and deal with. If something can be seen or witnessed it's easier. Prioritise the behaviours you wish to tackle, first in terms of need to be addressed, but then in terms of likely response to intervention. Start with something that you are most likely to be successful with. Being successful makes one feel better and more able. If you feel good, you feel more able to tackle more difficult problems. So choose the behaviour that's likely to respond, not necessarily the most problematic.

Undertake a "behavioural analysis". Draw three columns on a sheet of paper. Label the middle column B for "Behaviour". Label the left-hand column A for "antecedents" - the things that have been happening up to and including the onset of the behaviour. Label the right hand column C for "consequences" - what happened as a result of the behaviour. Document repeated events of the behaviour, the antecedents; the who, the what, when, where and the consequences, what happens as a result. Through this process very often the apparently inexplicable takes on meaning as a pattern emerges. For example, an individual always tantruming when a particular staff member is around, or crying whenever it's mealtime.

Develop a plan based on your findings and their likely meaning. Behaviour may be attention seeking. Conversely it may have a solitude seeking function or may serve the purpose of avoiding demands. It is important to work on maximising the time spent in useful activities rather than simply trying to diminish the undesired behaviour. The more time spent on positive activities, the less time there is for negative ones.

Ensure all who come in contact with the individual are aware of the programme and are "signed up" to ensuring it works. Unless people are doing things as similarly as they can, which includes even the wording and the phrasing and the intonation of the voice, whether it's at home or hostel or school or respite care centre or wherever, then it will be confusing for the individual involved. The plan has to be crystal clear to all concerned. It has to be practical. It has to be manageable. It is important to rate what's happening. Respond to what's happening. If things aren't going well think about why, but give things a certain amount of time to work first.

At its simplest there are just four ways of influencing behaviour for better or worse. You can add or subtract a positive or a negative consequence. This applies to all of us. You can add a positive reinforcement, or "reward" to enhance behaviour. I am positively reinforced for going into work by getting some money at the end of each month. If my employers stopped giving me money I shall in all probability rapidly stop going into work and look elsewhere. You can positively reinforce behaviour in which case it is likely to increase in frequency and intensity. You can add or remove a positive consequence. It may be tangible like money or sweets or may be something less tangible like social attention or a lot of affection. Theoretically you could add a very negative consequence, a punishment, in order to reduce the frequency and intensity of behaviour. Such techniques have ethical as well as scientific disadvantages. Often people will be behaving in a particular way because they find an experience unpleasant. This may be an insistence on their eye contact or being exposed in a classroom to people or activities one can't cope with. Subtracting a negative consequence – negative reinforcement – is the final possibility. An example is a parent with young child in a supermarket at the checkout where there are arrays of sweets. The child starts screaming. The parent tries to hold out and not to give in, knowing what's wanted. After a long time, probably longer than the time before, the parent finally gives in and gives the child Smarties. The child stops screaming. By having waited increasing durations of time to give the sweets, the parent is teaching the child that if you don't get what you want first and you persevere, then you'll get what you want. So if you're going to give in, give in immediately. Don't wait. The second important component of the above anecdote is that mother has positively reinforced the child's screaming behaviour by giving a nice consequence in response to the screaming. Thirdly mother's sweet-giving behaviour is being negatively reinforced. By giving sweets the aversive consequence of the child screaming is removed.

There are three phases to most behavioural programmes. The first can be entitled 'the honeymoon effect' whereby the new novel approach hasn't been done before. Everybody is excited including the client, and things go well. There then follows the "extinction burst". Things have become boring. The client may resent the programme. However, stopping the programme at this stage conveys a message that if the programme is disliked then worsening behaviour can lead to its termination. If you are going to commence a programme you need to carry on for a reasonable time. Having weathered the storm of the extinction burst the negative behaviour should start to diminish - slowly.

When you have established a consistent and successful reinforcement programme you then need to start weaning the individual off the need for regular and predictable reinforcers. The first step is to shift from regular reinforcement to intermittent reinforcement. Give the reward only every second time the positive behaviour occurs, then every third time and so

on. The next stage is to reward *on average* every fifth (or however many) times the desired behaviour occurs, with no guarantee as to when rewards will happen.

Questions

Q. When you talk about a child being 10 but maybe having the age of a 5 year-old, do you then approach that child as if it is a 5 year-old or do you try and get them to the stage of a 10 year-old? So do you have a child who you treat as a 5 year old but you try and give them special skills like going on a bus on their own or do you say "you shouldn't really be 5, you should be trying to be 10"?
A. A bit of both, I guess, is the answer. You need to start at a level that makes sense to the individual, which they can understand and appreciate. You also need to give the individual your respect, consistent with their chronological age. You need to be encouraging them to develop skills expected of someone of that age. It is demeaning and inappropriate for a 30 year-old to be carrying around a big, soft teddy bear the whole time. If individuals like that comfort they can be weaned on to smaller and smaller teddy bears, and finally have one of those ones on a little key ring in their pocket.

Q. My question is about self injury. My son is 9 and he seems to take some satisfaction hurting himself. He tends to scratch his legs and arms until they bleed and then whilst they are healing he can make them bleed again easily. He tends to do it at night when he's in bed and when he's calm, not when he's angry. It's a calming thing. But his legs and his arms are just scarred now. I try not to react and say anything, but I don't know why he does it and how to stop it.
A. It sounds like with appropriate support you need to go through the sort of framework I have discussed. From your brief account it seems to me that you've tried obvious things based on the idea that your son is attention seeking. It doesn't seem that he is. Maybe there are self-stimulatory aspects. One thing that strikes me is just how tolerant of pain many people with fragile X are. Maybe their need for very intense sensation might be part of a sensory integration problem. Self-injury may be the means of getting a necessary degree of feeling through whatever sensual modality it is. It sounds as well that there is a comforting aspect to the self-injury. Part of this is biochemical; injury leads to the release of chemicals similar to morphine which counteract pain and keep one calm. It may also be that your son is self-reinforcing his behaviour. One can try response prevention-type approaches, for example wearing gloves to counteract the stimulating sensation of nails on skin. Maybe tucking him in to bed very tightly might produce sufficient pressure to satisfy his need for high levels of incoming sensations. Rather than thinking 'what's the treatment for scratching himself in bed' it may be better to ask 'what is the function of this behaviour'? It sounds like you have made a start along that path, but there is quite a way to go in really understanding why he's doing that. Maybe with professional help you need to try various theories.

Q. When fragile X children get older, teenagers, will they become teenagers like Harry Enfield's Kevin type thing?
A. Adolescents, whether they have fragile X syndrome and learning disabilities or not, should be striving for autonomy and independence, testing out the boundaries and being frustrated by how dependent they still are on their parents. This is even more of a challenge for somebody with disabilities. Whatever developmental level individuals have, they will pass through an adolescent phase. Often it's very difficult, if not impossible, to tease out which behavioural aspects are attributable to learning disability, which to fragile X syndrome and which to being an adolescent.

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