

Fragile X Syndrome – Teaching Communication Skills

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Effective, fluent and enjoyable communication involves a multitude of interrelated, but very different, skills. Unfortunately a number of these vital skills are typically affected in children and adults with Fragile X Syndrome. This can result in communication becoming a difficult and stressful experience for them and for their conversational partners.

However a great deal can be done to remedy the situation. Some difficulties can be anticipated and therefore prepared for. Other difficulties respond to specific treatments or therapies. The situation is also generally improved when there is an understanding of the underlying difficulties and how they impact on the individual's communication skills.

The typical communication profile in Fragile X Syndrome involves a combination of:

1. Delayed early speech and language acquisition
2. Hearing loss, especially during the early years
3. Articulation difficulties
4. Linguistic difficulties
5. Attention and executive function difficulties
6. Hyper arousal
7. Social anxiety

I shall discuss each of these features separately:

1. Delayed early speech and language acquisition

Language is a complex process, requiring a variety of both receptive and expressive skills. When language acquisition is delayed many, if not all, of these skills may be affected. The delay can reflect an individual's overall intellectual development or can be out of kilter with this, with language skills either more or less advanced than non-verbal skills.

Receptive language (or verbal comprehension) involves the recognition of the sound system of a particular language (phonology), the understanding of the meanings of individual words (semantics), the understanding of grammatical structures (syntax), and the ability to process and remember strings of words (auditory memory).

Expressive skills involve the production of the speech sounds of a particular language (phonology and articulation), the use of individual words (vocabulary) and the formulation of grammatical structures at both word and sentence level (syntax). Any, or all, of these skills may be delayed in an individual with Fragile X Syndrome.

2. Hearing loss

Early conductive hearing loss, affecting the transmission of sounds to the auditory nerve, is usually the result of persistent middle ear infections. Even when the child is 'better' the puss that has built up in the middle ear can prevent sound travelling efficiently to the auditory nerve. Not being able to hear properly is likely to affect the individual's ability to pay attention to other people's speech, to understand what they hear, to monitor their own output and to produce clearly articulated speech at an appropriate rate and volume.

3. Articulation difficulties

Articulation difficulties in Fragile X Syndrome that are not directly related to hearing loss may be caused by:

- The typically narrow, highly-arched palate which necessitates a greater effort being made for effective articulation.
- The presence of low oral muscle tone (hypotonia) which limits the strength and range of oral movements.
- The presence of oral dyspraxia which affects the ability to coordinate the complex sequences of movements necessary for the production of words and sentences.

A combination of articulation difficulties, together with a rapid rate of talking and a tendency to 'perseverate' (described later), results in an arrhythmic, somewhat incoherent style of speech termed 'cluttering'.

4. Linguistic difficulties

Linguistic difficulties in Fragile X Syndrome reveal themselves in:

- Problems appreciating underlying or intended meaning. The individual focuses instead only on the literal meaning of phrases. For example, idioms ("Her eyes popped out of her head!"), sarcasm and teasing ("I don't think there'll be space for you!") are taken literally, and underlying suggestions ("What if you...?") or warnings ("*What* did you say?") are misinterpreted.
- Difficulty using language effectively for social purposes, for example for expressing feelings, asking for help, knowing how to address different people or when to approach a particular topic.
- Perseveration, which is the term used to describe the repetition of sounds, words, or phrases, and is different to echolalia where the individual repeats what others have said. The causes of perseveration in Fragile X Syndrome are unclear. Perseveration may be related to problems with word retrieval (with the individual unable to quickly access the word they are searching for), or to problems with attention and impulsivity (with the individual unable to inhibit the repetition), or to problems with planning and organising words and sentences (related to executive function difficulties), or to a combination of these factors. We do know however that perseveration is certainly worse when an individual feels anxious, and anxiety is often a key factor in Fragile X Syndrome.

5. Attention and executive function difficulties

Executive function skills are the skills needed to focus and maintain attention and to organise oneself in terms of thinking through the various stages of a task and then carrying it out. Difficulties in this area are a common feature of Fragile X Syndrome. With regard to communication they affect:

- The understanding of speech, particularly the ability to 'stay with' the conversation.
- The ability to keep to a topic, organise what to say (selecting and presenting relevant information), and maintain the usual style of turn-taking in conversation. Poor executive function skills result in a typically disorganised, 'tangential' style of speech with the speaker frequently digressing from the main topic of conversation, failing to introduce new ideas adequately and presenting peripheral detail before the central facts.
- The ability to think before acting!

6. Hyper arousal

Hyper arousal is the term used to describe increased sensitivity to various sensory stimuli. For individuals with Fragile X Syndrome touch, sound and busy environments can produce an uncomfortably high level of stimulation. The original feeling of excitement can quickly become an unpleasant feeling of panic that is difficult to reduce. Not surprisingly, certain types of social situations can easily become associated with feelings of anxiety.

7. Social anxiety

Social anxiety is a particular feature of Fragile X Syndrome. Typically, individuals are friendly, socially interested and enjoy companionship, yet they are shy with unfamiliar people, anxious in social gatherings, and often unwilling to make direct eye contact.

Anxiety affects concentration and this in turn affects learning. In addition, communication difficulties already mentioned (such as perseveration and a disorganised style of speech) increase. Behaviours associated with anxiety (such as hand-biting, hand-flapping and other repetitive actions) become evident.

In addition to the difficulties described above 15-25% of individuals with Fragile X Syndrome also have autism.

These people find it much harder to make sense of social information in all forms, including gesture, facial expression and tone of voice. They develop more pronounced ritualistic or obsessional behaviours. They are less motivated to communicate and have greater difficulty learning a communication system.

Effective Therapy

A variety of different approaches will be useful in helping an individual with Fragile X Syndrome combat such a wide variety of difficulties and develop their range of communication skills. However there are some important underlying principles that apply to all effective therapies.

Interventions should be:

- **Focussed**

The question that needs to be continually asked is, 'What skill am I trying to teach?' It may, for example, be an appropriate means of attracting attention, the initiating of a request, a slower rate of speech, an aspect of vocabulary, or an ability to listen to and process a certain number of words in a sentence.

- **Collaborative**

There needs to be agreement between home, school and respite care about which particular skill needs to be developed when, and on how and where it will be taught.

- **Developmental**

It is important to teach particular skills in accordance with the normal sequence of their development. For example, with regard to speech, perception of sounds develops before production, and "m" develops before "s". Speech and language therapists are of course very familiar with the normal development of speech and language skills.

- **Skill-based**

It is extremely difficult to teach someone a completely new skill. Instead, start from something the individual already does, an existing skill, (such as grabbing for an object) and guide and develop this into a more appropriate communication (such as pointing to an object).

- **Structured**

Again, beginning with an existing skill or behaviour, move through a series of small, graded steps towards your eventual goal. These steps will need to be well-planned and each consolidated before moving on to the next.

- **Rewarding**

True motivation arises when the learning and the teaching process is rewarding for all concerned. The learner needs to find the actual therapy enjoyable, or at least to gain an immediate reward for their effort, and the teacher needs to see some progress.

- **Functional**

Skills taught need to be useful for, and easily generalised to, real-life situations. For example, situations may need to be engineered so that phrases learnt earlier to describe picture cards can be used in a meaningful context.

- **Flexible**

Teaching often needs to be adapted to suit changing needs and circumstances. For example, a child who has successfully been taught to ask their teacher to go to the toilet at primary school may well need at secondary school to be taught to go to the toilet when the buzzer sounds between lessons and to practice explaining his lateness for his next lesson.

The ideal communication programme

The ideal communication programme begins with a thorough speech and language assessment. The speech and language therapist may use formal and/or informal assessment

methods. They should also take the lead in collating information from as many sources as possible about the individual's social use of language.

Following this process there needs to be agreement on the short-term aims of therapy (i.e. the skills one hopes to achieve during the first few months), on the teaching strategies to be used (for example, physically prompting particular responses or rewarding particular actions), and on who will implement the programme.

As the individual will undoubtedly benefit most from a programme that is implemented throughout his/her day it is only practical for teachers, parents and carers to become the 'therapists'. The speech and language therapist is ideally placed to teach these 'therapists'. This will involve detailed advice and instructions on teaching strategies, regular contact with the student and with those carrying out the communication programme, regular reviewing of the programme and its modification and development as necessary. In this way the individual with Fragile X Syndrome receives far more care and attention than they would using the 'traditional' model of weekly speech and language therapy.

Particular strategies

Effective communication programmes need to be tailor made for the individual. It is extremely important that they are founded on a thorough speech and language assessment and are implemented under the guidance of a speech and language therapist. However, there are some useful general pointers and strategies. These can be summarised under the headings of developing verbal comprehension, clear speech, non-verbal communication and social interaction.

Developing Verbal Comprehension

- If you have any worries about an individual's response or lack of response to sound, ask for a hearing assessment.
- Before speaking make sure you have the individual's full attention.
- Rather than slowing down your rate of speech, use short sentences with pauses between them.
- Comment on the immediate focus of attention or the activity in hand, rather than on people not actually present or past or future events.
- Tailor activities to suit the individual's attention span.
- Develop attention through general imitation and turn-taking games, as well as through specific listening activities.
- Reinforce key words with gestures and/or pictures.
- Don't assume an individual understands what has been said just because they respond appropriately. Take account of situational cues that they may be using to help them understand what is going on or is about to happen, for example the keys you pick up and

jangle before you open the door. These situational cues can either be used to advantage or consciously 'masked' to help with assessment.

- Gradually extend the amount of verbal information (the key words) that needs to be understood in order to carry out a request, for example:
0 key words (if the individual is used to tidying things away) - 'Throw that (the used tissue you gesture towards) in the bin.'
1 key word - 'Throw the tissue (as opposed to banana skin) in the bin.'
2 key words - 'Put the apples (as opposed to the oranges) on a plate (as opposed to the table).'
3 key words - 'Get me the box (as opposed to bag) that's in the cupboard (as opposed to the drawer).'
- Start teaching the more concrete verbal concepts and gradually move towards the more abstract, for example from 'cow' or 'cushion', to 'big' or 'soft', and then to 'last' or 'different'.
- In addition to direct telling or showing, encourage the individual to work things out actively by the process of deduction, by giving them a choice of objects or pictures, for example:
'Which one is the penguin?' (squirrel/penguin/horse)
'Which one is for wearing?' (ice cream/chips/scarf)
'She's painted a picture.' (girl painting a picture/ girl showing finished picture)
'The man's been bitten by the dog.' (man bitten by dog/ dog bitten by man)
- Clarify ambiguous language and intentions by explaining what you mean when, for example, you say, 'Keep an eye on the baby', 'Hold your tongue!' 'You better lie low for a while.'
- Use humour wherever possible (jokes, puns, 'silly' drawings) but explain what you mean.

Developing Clear speech

- Increase the strength and control of oral muscles through chewing, sucking, blowing and oral games. Try blowing ping-pong balls into goals across the table.
- Under the explicit guidance of a speech and language therapist you can reduce hypersensitivity with oral massage, using gradual desensitisation and deep pressure. A word of caution - this involves using your fingers to massage areas inside the child's mouth and if done incorrectly is likely to cause more problems!
- Teach sound discrimination through picture pairs of rhyming words, for example, "ball/tall", "fall/wall", "doll/dot", "dog/dodge".
- The individual's phonological (speech sound) system can be shaped and developed. However this requires a speech and language therapist's specialist knowledge of normal phonological development as well as of the particular phonological processes being used by the individual.

- Slow down the rate of speech through the use of rhythm and rhyme. For example using action and nursery rhymes with younger children, and repeating favourite, useful or amusing phrases in a rhythmical manner with older individuals.
- Teach syllable awareness, for example “hos-pi-tal”.
- Teach awareness and use of emphasis, for example the difference between ‘I don’t want to go *out* with you’ (I want to *stay in* and play Monopoly), and ‘I don’t want to go out with *you*’ (I want to go out with *Tamsin*).
- Encourage self monitoring skills by focusing on and reinforcing *the better attempts*.
- Play word finding games such as making lists to fit a particular category (vehicles/sounds/feelings, etc), or completing sentences (spiders crawl – birds.....)
- Practice selecting and organising information in terms of key points and peripheral details. This can be done by playing guessing games such as “What am I?” (where one person describes characteristics of an object and the other tries to guess what the object is), and also by using either pictures or written words (if the individual has some reading skills) to illustrate key concepts and so ‘guide’ a narrative.

Developing Non-verbal communication

Many individuals with Fragile X Syndrome are very slow to acquire speech and a few, particularly those with autism and severe learning disability, never do develop really useful speech. Although speech is undoubtedly what we all aspire to, it is important to remember that during normal development enjoyable, meaningful interaction always precedes the acquisition of speech, with infants and their carers engaging readily in non-verbal ‘dialogue’. The interaction itself is far more important than the actual means of communication. Non-verbal communication takes various forms:

- Conventional non-verbal communication is easily understood and is generally used alongside speech in the form of pointing, frowning, sighing, shrugging, nodding, etc.
- Alternative communication ‘systems’ (such as signing, using picture or symbol boards, or object or picture exchanges) may not necessarily be easily understandable to the uninitiated and need to be specifically taught.
- Finally there is the personal repertoire of actions and behaviours that a non-verbal individual naturally develops and that serves as a guide to their feelings and desires. These actions (for example squealing, bringing objects, biting their clothes) may or may not be intentional, may or may not be appropriate and may or may not be effective in communicating what the individual means. However they can be shaped into communication that is more intentional, more appropriate and more effective. This can be done by:
- *Anticipating and redirecting*. For example, if arrival home is always heralded by screaming and opening kitchen cupboards, a set of pictures depicting what the individual prefers to eat and drink can be prepared in advance and he can be taught to hand one of these to his carer in exchange for what he wants.

- *Treating the unintentional as if it were intentional*, so that the individual comes to realise that a particular action in a particular situation causes something particular to happen. For example, initially inadvertent eye-contact can be shaped into a 'request' for another spoonful, another verse of a song, another push on the hammock, etc
- *Physical prompting*, whereby for example the grab is shaped into a point.
- Enjoyable repetitive games or actions in which a particular format is used to encourage the individual to respond in a particular way in order to keep the activity going. This format (*action -pause just before reaching the climax – individual's response – action continuing*) tends to occur naturally in action rhymes. These typically build towards a particular word and associated action (such as "Ring-a-ring-a-roses...we all *fall down!*", "Humpty-Dumpty...had a great *fall!*", "...*pop* goes the weasel"). A gradually increasing tempo followed by a pause just before the climax is reached can be used to elicit, for example, a particular sign or gesture, eye contact and/or vocalisation. The 'action rhyme' is often even more effective when it is a spontaneous creation arising from a spontaneous interaction, for example when bouncing a child on your knee (e.g. "Higher and higher and higher and *up* you go"), pushing a swing, spinning a swivel chair, or playing chase (e.g. "one, two, three, four, I'm coming to *get you!*").

Developing Social interaction skills

- As we have seen, social anxiety is a salient feature in Fragile X Syndrome. It is therefore particularly important to take notice of behaviours (such as avoiding eye-contact, biting hands or clothes, hand flapping or rocking) that may indicate rising stress levels. By recognising these behaviours, situations that are likely to be difficult can be anticipated and therefore avoided, adapted or prepared for.
- The individual can themselves be taught to recognise and to monitor their own arousal or anxiety levels. A visual reminder or cue, in the form for example of traffic lights, a thermometer or a scale of emotions, can be helpful.
- An effective calming down procedure should be taught that the individual can manage for themselves, such as playing with a particular toy, looking at a particular book, listening to a piece of music, or engaging in some rhythmical routine or strenuous physical activity (such as lifting, carrying or pushing).
- The intensity of a social interaction can be reduced by avoiding face-to-face conversation and teaching, and by focussing on cooperative and group activities rather than on the individual's speech or actions.
- A clear, modelled framework for social interactions should be provided, for example for making requests, sharing an activity, giving an apology, refusing politely, making a suggestion, etc.
- Opportunities to practice these interactions need to be especially engineered and supported.

- Real life situations, such as asking for or giving directions, explaining a problem, offering a point of view, can be rehearsed using prepared scripts and visual prompts.
- Relating to other people is directly influenced by how we feel about ourselves. It is therefore vital to foster the individual's sense of achievement and self-worth. This can be done through constant reminders (visual, verbal and tangible) of their various interests, successes and enjoyable past experiences. Photo albums, collections of memorabilia and wall displays are all useful.

You will see progress

Fragile X Syndrome with its various associated difficulties cannot of course be 'cured'. However with sensitive and knowledgeable management there is an excellent chance that the difficulties will decrease and that skills and enjoyment of life will increase. Once you are able to appreciate more fully the difficulties of someone with Fragile X Syndrome you can begin to understand the world from their perspective. Then, with patience, flexibility and some creative thinking you will, I am sure, see progress.

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