

Facilitating the Inclusion of Children with Fragile X Syndrome

There is increasing evidence that there are distinctive cognitive and behavioural profiles in several syndromes with associated intellectual disability. Research suggests that there is not, as of yet, a good understanding among teachers of aspects of the learning and behavioural profile associated with children with fragile X syndrome. As a result, there is a poor awareness of strategies and interventions that can maximise the inclusion and participation of these children in mainstream and special education settings. Aspects of the cognitive and behavioural profile associated with fragile X syndrome that teachers and professionals need to be aware of are discussed and outlined.

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INTRODUCTION

There is increasing research evidence which suggests that there are distinctive cognitive-behavioural profiles in several syndromes with associated intellectual disability (Dykens, Hodapp and Finucane, 2000). There is evidence that educators have a poor knowledge of syndrome specific profiles (York, Von Fraunhofer, Turk and Sedgwick, 1999) and as a result may propose educational interventions that are unlikely to best address children's special educational needs. Specific knowledge of these syndrome profiles is critical for professionals on two levels: (a) to promote early diagnosis and appropriate management and interventions for relatively common syndromes and (b) to ensure that research advances are translated into meaningful practice (Lee, Blasey, Dyer-Friedman, Glaser, Reiss and Eliez, 2005). Fragile X syndrome is one such condition where there is an increasing awareness that knowledge of the cognitive and behavioural profile associated with the condition, can help the affected children in reaching their potential in educational settings. Fragile X syndrome is the most common identified inherited form of intellectual disability (Dykens et al., 2000). The prevalence of fragile X syndrome has been estimated to be approximately 1 in 4000 for males (Turner, Webb, Wake and Robinson, 1996), and the prevalence for females affected has been presumed to be approximately half the male prevalence (Sherman, 2002). While almost all affected males will have intellectual disability, only 50-70% of females will have intellectual disability and the remainder may have more specific learning difficulties (Riddle, Cheema, Sobesky, Gardner, Taylor, Pennington and Hagerman, 1998).

PHYSICAL CHARACTERISTICS AND ASSOCIATED MEDICAL CONDITIONS

Though by no means present in all those with fragile X syndrome, there are some distinctive physical features which may be present, especially in postpubertal males.

These physical features include large and prominent ears, a long narrow face, velvet-like skin, hyperextensible finger joints, double-jointed thumbs, hypotonia (abnormal muscle slackness), flat feet and a single palmar crease (Hagerman, 2002). The physical features of the syndrome, often so noticeable in males, may be present in females but usually to a lesser degree. Recurrent otitis media (middle ear infection) is a frequent difficulty for children with fragile X in early childhood, and strabismus (a condition in which there is abnormal deviation of one eye in relation to the other), can be seen in approximately one third of children with fragile X syndrome (Hagerman, 2004). Approximately 20% of children with fragile X will also have epilepsy (Musumeci, Hagerman, Ferri, Bosco, Dalla Bernardina, Tassinari, DeSarro and Elia, 1999).

BEHAVIOURAL PROFILE

Individuals with fragile X syndrome seem to be extra sensitive to sensory stimuli such that they are easily hyperaroused (overexcited) in situations with excess auditory, visual, or tactile stimuli in crowded and/or noisy environments (Hagerman, 2002). This hyperarousal or over-stimulation in social situations is increasingly being recognised as a key feature of fragile X and may be related to the display of challenging and atypical behaviours. Most children with fragile X will engage in hand flapping at some stage in their lives and it may be a reaction to the intensity of social interaction (Cohen, 1995). The most frequent form of self-injury in fragile X syndrome is hand-biting (on back of hand and fingers) and it is most likely to occur following the presentation of a difficult task or a change in routine. Shyness or social anxiety is also a notable feature of children with fragile X (Sobesky, 1996) and is prevalent in both males and females with the syndrome. Tactile defensiveness or extra sensitivity to tactile stimuli is also common in individuals with fragile X syndrome (Hagerman, 2002).

Attention deficits, restlessness, hyperactivity and fidgeting have been described as some of the most striking and pervasive of the challenging behaviours associated with fragile X syndrome (Bregman, Leckman and Ort, 1988) and may result in some of the children, especially males, receiving a diagnosis of Attention Deficit/Hyperactivity Disorder (ADHD). Girls with fragile X and ADHD usually have less hyperactivity, compared to boys with fragile X, but impulsivity and short attention span can also be a significant problem for girls (Hagerman, 2002). Research has shown that autistic-like features, such as hand flapping, perseveration in speech, shyness, and poor eye contact are seen in the majority of individuals with fragile X syndrome (Turk and Graham, 1997). However, the majority of children with fragile X syndrome do not demonstrate the core social deficits typical of autism (Hagerman, 2002). The percentage of children with fragile X syndrome who also fulfil criteria for autism has ranged from 15% to 28% (Turk and Graham, 1997). The presence of autism with fragile X syndrome is associated with severe language and social deficits, in addition to lower IQ, compared with that of children with fragile X syndrome without autism (Lewis, Abbeduto, Murphy, Richmond, Giles, Bruno and Schroeder, 2006). The typical communication profile of those who have fragile X syndrome may involve a combination of: delayed early speech and language, attention difficulties, social anxiety and atypical language (Taylor, 2004). Atypical language includes tangential

language (off-topic questions and responses or comments), perseverative language (the reintroduction of favourite topics over and over), and repetitive speech (repetition of sounds, words or phrases).

EDUCATIONAL PROVISION AND SUPPORTS

Many children with fragile X syndrome demonstrate strong verbal and behavioural imitation skills (Spiridiglozzi, Lachiewicz, MacMurdo, Vizoso, O'Donnell, McConkie-Rosell and Burgess, 1994) and, because of this, placement in a mainstream setting may be desirable. However, it may be that while they can participate in some of the activities in the mainstream school, they will begin to experience difficulties as the curriculum begins to focus on more abstract topics in the middle and senior primary years (Dew-Hughes, 2004b). It has been suggested that students with fragile X syndrome may benefit from attending schools/centres where ABA approaches are practiced, but Stackhouse and Scharfenaker (2006) argue that a strict ABA approach will need modifications for children with fragile X.

In terms of how personnel who work with children affected by fragile X can best support the student's inclusion and participation, Braden (2002) suggests that during times of high stress a special needs assistant (SNA) can be available to encourage calming activities and direct sensory integration support. Saunders (1999) advocates that the adult working with the child with fragile X syndrome should be as quiet, calm and unobtrusive as possible, avoiding touch or eye contact in order that they do not become a distraction themselves and to reduce the chances of the child becoming too dependent on them. Given the genetic implications of the condition, the inclusion of goals for the family in the child's Individual Education Plan (IEP) may be particularly relevant for children with fragile X. In this regard, Braden (2004b) recommends that an IEP for a child with fragile X should contain both long-term goals and short-term objectives for the family and include the family's resources, priorities, and challenges, including the need for additional money, community resources and emotional support. In a study of educational provision for children with fragile X in Ireland, it was found that although most children with fragile X had an IEP developed for them, only two-thirds of parents reported that they were involved in the IEP development process (Reilly, 2006). In terms of the areas included in the IEP, the areas most frequently included were mathematics, literacy, behaviour and social skills (Reilly, 2006).

CLASSROOM ACCOMMODATIONS TO FACILITATE INCLUSION

Appropriate accommodations in the classroom environment are likely to lessen the potential for the emergence of challenging and atypical behaviour and promote a positive learning environment for the child with fragile X. The use of a visual schedule/timetable can reduce anxiety often experienced by children with fragile X at times of change and transition (National Fragile X Foundation, 2004). A visual schedule/timetable was reported to have been used with 69% (n=20) of the children in Reilly's 2006 study. Braden (2004a) and Saunders (1999) suggest the following strategies for classroom organisation and instruction when working with children with fragile X:

- **The room is as free from distractions as possible, both in terms of visual distractions, displays and wall mountings.**
- **Attending behaviours may improve and the need for hyper-vigilance may decrease if a student with fragile X is positioned at the front of the classroom with easy access to a door or exit.**
- **Frequent breaks can help avoid attention difficulties and lack of concentration.**
- **The amount to be copied from printed or written material should be limited and visually presented materials should be simplified to eliminate a cluttered or excessively stimulating format.**
- **It may be useful to provide a quiet, distraction-free safe area that the child can work in and retreat to, if the activities of the room become overwhelming.**
- **There should be sufficient flexibility in the class to allow the child with fragile X to leave their seat and move around the room periodically.**

TEACHING STRATEGIES

Studies in the 1980s identified a particular processing style for males with fragile X that has formed the foundation from which intervention strategies have been developed (Braden, 2002). The studies and subsequent research (e.g. Powell, 2004) suggest that males and most females with fragile X syndrome perform better on tasks that require a simultaneous approach, as opposed to a sequential approach to processing information. The relative weakness in sequential processing is shared by children with Prader-Willi syndrome (Dykens, Hodapp, Walsh and Nash, 1992) but not by children with Down syndrome (Pueschel, Gallagher, Zartler and Pezullo, 1986). This style of acquiring information is counter to most traditional teaching methods, which rely on sequential or step-by-step approaches.

Information presented using a simultaneous approach resembles a “visual whole” or “gestalt”. The use of diagrams, pictures and visual associations can help capitalise on this processing style and facilitate the memorisation of novel tasks. The implications of the weakness in sequential processing mean that children with fragile X are likely to have difficulty with phonics and decoding words, breaking down mathematic or science problems into their constituent parts, interpreting the parts or features of a design or drawing, comprehending the rules of a game, following verbal instructions and recalling specific details and the sequence of a story (Hodapp and Ricci, 2002). As well as the strength in simultaneous processing, the National Fragile X Foundation (2004) suggests that cognitive strengths in males and some females may include:

- **verbal learning and single word vocabulary especially for subjects of interest;**
- **receptive vocabulary;**
- **memory for situations and for favourite TV shows, videos and songs;**
- **mimicry/imitation and incidental learning (e.g. observing modelled behaviours);**
- **adaptive functioning for life skills such as self-care, household management, and cooking;**
- **computer skills**

The National Fragile X Foundation (2004) suggests that cognitive weaknesses among males with fragile X may include:

- **higher level thinking and reasoning skill;**
- **visual-motor tasks such as handwriting;**
- **quantitative skills including arithmetic abilities because of visual-spatial problems, sequencing difficulties and poor concepts of quantity and number;**
- **socialisation and communication, especially in novel settings.**

Children with fragile X are likely to benefit from learning that is linked to, or associated with, a bigger whole. Closely linking new learning to children's interests and previously well-learned information will capitalise on their good long-term memory skills (Kemper, Hagerman and Altshul-Stark, 1988) and their desire for an association with a whole. Social stories (Gray, 1994) may also be useful in helping children with fragile X overcome anxiety about specific events or occasions (Scharfenaker, O'Connor, Stackhouse, Braden and Gray, 2002). Children with fragile X seem to perform better on tasks that are familiar and repetitive (Braden, 2002) and they have good memory for experiences that are visual, contextual and directly experienced.

CLASSROOM INSTRUCTIONS

In terms of language, children with fragile X typically have delays in both receptive and expressive language (e.g. Sudhalter and Belser, 2004), so it is important that language should be kept concrete and simple. In order to reduce anxiety, side-on dialogue or teaching may be beneficial for children with fragile X (National Fragile X Foundation, 2004). An understanding of the benefits of side-on teaching or dialogue needs to be appreciated in the classroom context, as insistence on eye contact for children with fragile X may cause undue distress and may lead to behavioural disturbance (Turk and O'Brien, 2002). In only 33% (n=8) of cases, parents interviewed in Reilly's (2006) study reported that side-on teaching was used with their children. Teachers of children with fragile X should avoid direct pressure on the child (time limits, questions in front of others, closed questions, eye contact or insistence on collaboration), as this often causes an adverse reaction and can be counter-productive (Saunders, 1999).

MATHEMATICS

There have been few published studies of the mathematical performance of children with fragile X syndrome, but performance on maths-type tasks on tests of cognitive functioning suggests a particular weakness in this area (Freund and Reiss, 1991). In terms of strategies for teaching maths to children with fragile X, Braden (2004a) suggests that teachers should:

- **use concrete manipulative materials to teach concepts and mathematical operations;**
- **use visual cues such as diagrams, illustrations and visual patterns whenever possible to reinforce mathematical concepts;**

- **allow extra time to reduce possibility of provoking performance anxiety;**
- **use patterning and repetition whenever rote memory tasks are required;**
- **employ the use of a number line for teaching addition and subtraction as students see numbers as "wholes" and part of a larger picture.**

READING

The difficulties in sequential processing that boys, and some girls, with fragile X experience have led some authors (e.g. Braden, 2002) to conclude that learning to read via a phonics based approach will prove very difficult. As a result, interventions to promote reading have emphasised strategies that capitalise on strengths in simultaneous processing. The National Fragile X Foundation (2004) suggests that programmes utilising visual cues to teach letter sounds or words may be useful, or approaches that emphasise sight words may be effective. Braden suggests the following strategies for promoting reading with boys with fragile X:

- **focusing on development of sight words;**
- **visual approach with patterns;**
- **words paired with pictures;**
- **whole word labels on objects;**
- **phrase cards of familiar greetings and sayings.**

The National Fragile X Foundation (2004) suggests that, although students with fragile X may understand what they read, it might be difficult for them to answer direct questions about the content, and in this regard cloze techniques may help increase general comprehension.

THERAPEUTIC INTERVENTIONS

Many children with fragile X have complex needs which will need intervention from professionals working in a number of health and education disciplines. Scharfenaker et al. (2002) point out that because of the multifaceted needs of students with fragile X, an integrated approach to intervention and education that combines the expertise of parents, speech and language therapist, occupational therapist, paediatrician, psychologist, special education teacher, and classroom teacher will be most beneficial. The combining of occupational therapy and speech and language therapy has been identified as a primary recommendation to support students with fragile X (Scharfenaker et al., 2002). Difficulties with sensory integration are common among children with fragile X (Schopmeyer and Lowe, 1992), and amelioration of these difficulties may require the intervention of a professional, such as an occupational therapist or physiotherapist with a good knowledge of sensory integration therapy. Scharfenaker et al. (2002) assert that it is critical for therapists and educators working with individuals with fragile X syndrome to understand that many behavioural problems result from sensory-integrative deficits and are not intentional acts on the part of the child.

SUMMARY

Dew-Hughes (2004a) cautions that the increased "scientific diagnosis of learning difficulties", via medical and genetic methods, has revived the spectre of 'within child deficits' and may lead to a return to the medical model of special educational needs. Hodapp (2004) also notes that rarely are aetiology-related behaviours found in every person with a particular syndrome and in this regard not all children with fragile X syndrome will be shy and anxious, and not all will display hyperactive behaviours. However, a knowledge of interventions that have been shown to work for the majority of children with fragile X is important and should be considered along with other sources of data (e.g. assessment data) when formulating recommendations for educational interventions. In this way a 'label' such as fragile X can become "an enabler" (O'Brien, 2002) and ensure that educational professionals propose and implement interventions that will lead to positive outcomes for the affected children.

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