The Fragile X Society

Over a five month period in 1996, 50 girls from our members' families took part in Lesley Powell's study investigating the cognitive (learning) functioning profiles of schoolgirls with fragile X. The results of this study were fully reported in Newsletter Issue No 18 Winter 1998 and we are very grateful to Lesley for providing us with this second article which draws on the findings of her study of schoolgirls with fragile X and her earlier study of boys with fragile X.

EDUCATIONAL NEEDS OF A CHILD WITH FRAGILE X SYNDROME by Dr Lesley Powell, Australia

Further to my article in a previous edition where I reported the findings of my research of schoolgirls with Fragile X Syndrome I now take the opportunity to provide some practical recommendations regarding the education of children with Fragile X Syndrome. I have provided cognitive functioning profiles and educational strategies for full mutation boys and girls who have an intellectual disability and full mutation girls who do not have an intellectual disability. I do hope you find them useful.

SECTION 1: FULL MUTATION BOYS AND GHILS WHO HAVE AN INTELLECTUAL DISABILITY COGNITIVE FUNCTIONING PROFILE

These children can usually repeat a series of verbal instructions, and recall a series of items when supplied with a visual cue, as well as their peers of similar intellectual ability. They will have difficulty, however, repeating or carrying out a series of instructions that require motor movements (sequential processing deficit). They will have significantly better visual learning skills, visual awareness, and alertness to the environment (simultaneous processing skills), than their peers who are of a similar age and IQ, but will not perform as well in the academic areas of arithmetic and word recognition.

IMPLICATIONS FOR EDUCATIONAL PRACTICE

Children who prefer to use simultaneous processing methods to solve problems are able to process and integrate parallel pieces of information at the same time by drawing on the knowledge of the environment and relating known facts to concrete objects. Simultaneous processing is particularly important in recognising the shape and appearance of numbers, letters and words, and understanding the overall meaning of a story or situation.

Children who exhibit a sequential processing deficit have difficulty with word attack skills, decoding and phonetics, the rules of grammar, breaking down arithmetic problems into their component parts, following a sequence of steps or rules, and following oral instructions.

These children benefit from the implementation of teaching methods which de-emphasise sequential processing by avoiding tasks which must be completed in a step by step manner, and concentrate on using simultaneous processing problem solving skills for literacy, mathematics, and sequencing.

LITERACY

In the fields of decoding and handwriting, there are several teaching methods which can be used. Decoding is a sequential processing skill which involves breaking a word into separate sounds (phonics) and then sequencing the sounds into a word. All full mutation

fragile X children with intellectual disability need to be taught using a reading method which avoids decoding and concentrates on holistic, visual learning, namely, whole language. Programmes which use a visual-spatial whole word approach to vocabulary building and which concentrate on the shape of the whole word rather than the phonetic structure of the word should be used. Handwriting requires sequential motor and children with fragile X may experience difficulty mastering this skill. Using a computer keyboard requires less motor planning and execution to hit a desired letter than forming a letter with a pen on paper. A computer screen is also related to visual learning and is more suitable for the fragile X child.

MATHEMATICS

Understanding place value, simple addition, and subtraction involves simultaneous processing but the operations of multiplication and division, which rely on verbal rehearsal and memorisation of multiplication tables, focus on sequential verbal strategies. Therefore, beyond simple addition and subtraction, the basic operations call for increasing amounts of sequential processing to carry out procedures in a step by step fashion.

Children with sequencing problems have difficulty perceiving relationships in their mind and understanding the necessary mathematical steps needed to solve a problem. These children rely on real objects in order to learn about the relationships of objects to each other and the manipulation of numbers to explain these relationships.

Programmes should be chosen in which students learn procedures for manipulating real objects to solve real problems with the focus on problem solving rather than on non-contextual memorising. For children who are particularly affected and are unable to cope with conventional mathematics the focus should be on learning maths skills with practical applications: handling money, telling the time and cooking.

SEQUENCING

The following strategies have been suggested by myself and successfully used with boys and girls in Britain and Australia to overcome the difficulties associated with: physically carrying out a series of instructions; reducing anxiety related to sequential planning; and learning a sequenced task.

Carrying out instructions

The most difficult task for all fragile X children appears to be related to physically carrying out a series of instructions relating to motor movements. Once a series of instructions has either been verbally given to the child or modelled for the child they must be reinforced before the series of instructions are performed. For example, if instructions are given to the class in general to begin a task, these could be repeated first by the teacher, then by a fellow student or the fragile X child him/herself. Once the class begins the task, it should be once again reinforced by the teacher who could then check to see that the fragile X child has remembered what she or he must do once their physical performance of the task has begun.

Reducing anxiety related to sequential planning

Much of the anxiety related to the presence of fragile X can be reduced if children are given a simple outline of the day's activities and are reminded to refer to their timetable throughout the day. These children should be encouraged to plan school and family activities in a series of steps listed in the correct order of completion.

Learning a sequenced task

The sequential difficulties exhibited by fragile X children and their relative strength in visual, holistic learning (simultaneous processing), means that they may not respond well to traditional task analysis (or chaining) teaching methods. Task analysis consists of breaking down a whole task into a series of small components which are taught individually and later performed together as a whole task. For children who experience sequencing difficulties and have a holistic approach to learning, performing or learning a task in small unrelated steps may have little meaning. Also, once these small individual steps have been mastered, re-sequencing the task may raise the anxiety levels of these children.

Therefore, reverse task analysis (or reverse chaining) should be used to teach basic skills. With this method the whole task is modelled repeatedly for the child. Once the child is familiar with the task they should be encouraged to carry out the final step of the task so that the task is always being completed by them. Later, the child can complete the final two steps, then the final three steps until he or she can confidently complete the task from beginning to end. If a task has many sequenced steps, beginning with the first step can result in anxiety and frustration for the child with no sense of achievement.

SECTION 2: FULL MUTATION GIRLS WHO DO NOT HAVE AN INTELLECTUAL DISABILITY

COGNITIVE FUNCTIONING PROFILE

These girls often have a significant impairment in their ability to verbally or physically repeat a series of instructions with or without a visual cue. They may also experience difficulties mastering mathematical skills relating to numeration, fractions, geometry and symbols, addition, subtraction, multiplication, division, numerical reasoning, word problems, missing elements and measurement. These girls can also exhibit frontal lobe dysfunction with deficits in attention, planning, abstract thought processes and the ability to retrieve information from an abstract cue, and mental flexibility.

IMPLICATIONS FOR EDUCATIONAL PRACTICE

These girls do not have an intellectual disability and are educated, therefore, in mainstream classrooms. Thus, they may not have access to special education services, except where it is deemed necessary by individual schools who have the resources to offer remedial instruction in relation to mathematics. The deficits exhibited by these girls, however, are more complex than the classification of "learning disability in mathematics" normally suggests. Educational management of their condition, therefore, is multi-dimensional and requires more than the implementation of a remedial mathematics programme. The following four areas may require remediation: sequential processing, mathematics, spatial awareness and frontal lobe function (executive function which is the ability to carry out a plan of action, and to correct and/or change the plan if it is not working).

SEQUENTIAL PROCESSING DEFICITS

The education management of sequential processing deficits and basic mathematics skills discussed in Section One are also relevant for these girls,

If difficulties with reading are observed, a highly structured multi-sensory programme should be used by teachers. The Toe by Toe programme developed in Britain, is of particular value (Cowling & Cowling, 1993). This programme can be ordered by writing to £38 Green Road, Baildon, West Yorkshire BD17 5HL 201274 598807, FAX 01274 582630 website www.toe-by-toe.co.uk

SPATIAL AWARENESS DEFICIT

The following is a list of general strategies that can be used by teachers to avoid spatial confusion and subsequent anxiety in the classroom.

- Limit the amount of work to be copied from written material.
- Make sure that worksheets are well spaced. The work that is normally presented to the class on one sheet can be halved, enlarged and printed on both sides of the page.
- Simplify visually presented work to eliminate clutter.
- Provide visual cues, for example, colour coding to organise written work.
- In mathematics, provide ruled columns which will enable the child to keep separate the units, tens, hundreds and thousands columns in sums.

MATHEMATICS DISABILITY

Girls may require diagnostic assessment of their mathematical performance and remedial instruction in any areas where difficulties are detected. The discussion in Section One relating to mathematics, is also relevant for this group. There are, however, two additional areas of concern that may require attention.

First, recent research suggests that known skills should be constantly revised if these girls are to retain the basic skills needed to perform complex operations in upper primary and secondary schools. Second, the inflexibility of thought and action, attentional problems, and poor sequencing and planning skills exhibited by many girls should be noted by teachers of mathematics, who can then cue the child to stay on task, change direction when necessary, and teach and later reinforce planning and sequencing techniques.

FRONTAL LOBE DYSFUNCTION - CLASSROOM MANAGEMENT

A combination of direct instruction and the use of self-questioning strategies is often the most beneficial remedial approach.

- Encourage participation in activities that promote autonomy and require organisation, planning and judgement (for example, a class presentation).
- Promote self-awareness and goal setting by training the student to estimate the time needed to complete a task.
- Provide a systematic outline for planning in general.
- Promote self-questioning at a specified time during the performance of a task.
- Allow the child to use formal problem-solving outlines.
- Encourage the child to follow a written plan.
- Reduce choices and options to a minimum. Students with executive function problems are overwhelmed by multiple options because they tend to get caught up in irrelevant detail and cannot recognise priorities.
- Provide a well-structured and predictable classroom environment to avoid student anxiety. Teachers should provide students with schedules of daily and weekly activities, including dates when assignments are due and examination dates. Stu-

dents may need to be reminded to check these schedules periodically throughout the day.

- Provide the student with a personal organiser or "memory book". Use can be encouraged through modelling and directed practice with daily entries (e.g. activities, completed tasks, assignments and appointments). Even when long-term memory is good, students may require frequent cueing to carry out activities appropriately.
- Set clear goals and make sure that the student understands those goals.
- Provide clear, concise explanations and illustrations of the subject matter.
- Ask frequent questions to ascertain whether the student understands the requirements of the task.
- Space learning over time. Organise practice of skills according to a written schedule.
- Identify the main idea in new information, draw associations between new information and past learning and draw inferences about the significance of new information.

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