

Executive functioning & social understanding in boys with fragile X syndrome

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Background

Parents of boys with fragile X syndrome have reported problems with attention and hyperactivity and several studies have reported that boys with fragile X syndrome perform poorly on tests of attention. Social difficulties (e.g. difficulties in social interactions with peers and social withdrawal) have also been found in boys with fragile X syndrome. There is some overlap between the problems with social interaction displayed by boys with fragile X syndrome and those observed in children with autism. Many boys with fragile X syndrome display autistic features and some boys with fragile X syndrome also have a diagnosis of autism. Several researchers have investigated how difficulties with 'executive functioning' might underlie the observed social deficits in autism. In the following section, we outline executive function and what is known about executive functioning in boys with fragile X syndrome.

Executive function

Executive function is the *set of abilities* that are involved in planning and guiding behaviour in order to achieve a goal in an efficient way. These include the ability to disengage from the external context; inhibit inappropriate responses; plan and generate actions; monitor your own performance and make use of feedback; and flexibly shift attention. These abilities enable us to perform everyday activities such as getting to work or supermarket shopping. When supermarket shopping, for example, you might plan your route around the store and choose a trolley or a basket according to how many items you plan to purchase. If the goods you intended to buy are unavailable, you can engage in mental flexibility and alter your plans, perhaps substituting items. You may engage in inhibitory control and pick up items you need rather than items you like (if you were unable to do this you might well end up with a trolley full of chocolate). Finally, you are likely to be using working memory, to update which items you have picked up and which items you still need to get.

Executive functions have been studied in boys with fragile X syndrome and individuals with autism. Boys with fragile X syndrome and individuals with autism have difficulties with executive function. Boys with fragile X syndrome have particular difficulties on tests that measure mental flexibility (e.g., the ability to shift to a different thought or action as the situation demands), inhibition (e.g., the ability to inhibit responses to irrelevant stimuli while pursuing a goal) and working memory (e.g., the temporary storage and retrieval of information). The research findings suggest that mental flexibility is influenced by general intellectual ability in males with fragile X syndrome and a similar relationship has been found in children with autism on tests of mental flexibility and tests of planning (e.g., the ability to 'look ahead', construct a plan, monitor, evaluate and revise the plan accordingly). Bennetto & Pennington (2002)

suggest that these difficulties are consistent with some of the behavioural problems that are shown by males with fragile X syndrome. They suggest that difficulties with attention, impulsivity and transitioning between activities may all be related to deficits in executive function.

Social understanding and functioning

A deficit in executive function may also relate to some of the more social impairments that have been observed in fragile X syndrome. Executive function may enable us to step back from our own perceptions of events and enable us to see events from other peoples' perspectives. In order to understand other peoples' perspectives we need to have the ability to inhibit our own knowledge and beliefs. Understanding other peoples' perspectives is important for understanding social situations and successful social interaction. For example, when we decide to share a rude joke with our partner but not with a stranger we have just met, we are probably guided by what we imagine others' expectations and evaluations to be. Without the ability to perspective take, social interaction is likely to become awkward, inappropriate and even challenging to understand.

The Study

This study focused on boys with fragile X syndrome. We were interested in the reasons why boys with fragile X syndrome find social interaction difficult. One explanation might be that they have problems with perspective taking. Other studies have explored this, but so far the findings have been inconclusive (see Garner et al, 1999 or Cornish et al, 2005). One difficulty is that often tests of perspective taking (also known as theory of mind or belief-reasoning) make substantial demands upon working memory and inhibitory control, which boys with fragile X syndrome also have difficulty with. Our study was designed to help us separate out potential difficulties with perspective taking from difficulties with working memory and inhibitory control.

Two groups of boys with fragile X syndrome participated, with families located all across the United Kingdom. Parent ratings were used to identify a group with few autistic features and a group with many autistic features. For comparison purposes, we also recruited a group of boys from a school for children with moderate learning difficulties who did not have fragile X syndrome or autism. Table 1 outlines the main characteristics of our groups.

Table 1. Participant Characteristics.

Group	Number of participants	Age	Verbal mental age	Verbal IQ
Fragile X - few autistic features	15	13 years 8 months	6 years 11 months	53.9
Fragile X - many autistic features	15	12 years 5 months	6 years 8 months	53.4
Intellectual Disability	15	13 years 9 months	6 years 11 months	55.8

Findings

Our findings suggest that children with fragile X syndrome do have a clear impairment on belief-reasoning tasks that is disproportionate to their general level of intellectual

disability. Importantly, this was the case whether or not the children with fragile X syndrome had many autistic features, suggesting that theory of mind difficulties in fragile X syndrome are not merely an artifact of the high co-morbidity of fragile X syndrome and autism. However, boys with fragile X syndrome also perform worse on working memory control trials. This means that although theory of mind difficulties are likely to be an important aspect of the fragile X syndrome clinical profile, it is probable that the deficits in theory of mind understanding stem from general information processing deficits (e.g., working memory).

Clinical Implications

There are some issues identified relating to theory of mind and executive functioning that may be of value to consider in a clinical setting and to explore in future research. There is evidence that theory of mind is impaired in boys with fragile X syndrome and it is likely that this deficit plays a critical role in the social difficulties associated with fragile X syndrome. Interventions aimed at improving theory of mind understanding in young children with fragile X syndrome may therefore help to improve the developmental outcomes of these children.

Clinical work that aims to address the social anxiety observed in adolescents and adults with fragile X syndrome may need to focus upon theory of mind understanding. Moreover, interventions aimed at improving theory of mind may also need to target the difficulties relating to working memory so that the client can benefit from therapy. Hence, interventions aimed at improving executive functioning are particularly important. Within the adult executive function literature, deficits in executive function are associated with a poor response to treatment, including treatment aimed at addressing difficulties other than those relating to executive function (e.g., physical therapy). Impairments in executive function have been shown to affect a client's ability to engage in and benefit from therapy. Strategies such as verbal self-instruction, self-regulation training and metacognitive strategies have led to improved planning, problem solving, goal management and self-regulation abilities in adults, on both specific tasks and in general functioning. In a clinical setting, it is unlikely that boys with fragile X syndrome will be referred specifically for executive functioning deficits. Nonetheless, it may be necessary to target the development of executive functioning skills in boys with fragile X syndrome before the issues for which the referral was made can be addressed.

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