



Further delineation of the executive deficit in males with fragile-X syndrome

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Received 12 January 2001; accepted 16 October 2001

Abstract

This paper presents a detailed analysis of one aspect of performance by young males with fragile-X syndrome (FMR-1 full mutation) who were assessed on a computerised visual search task as part of a larger study examining aspects of attention [Neuropsychologia 38 (2000) 1261]. They were matched on chronological and mental age to 25 boys with Down's syndrome (trisomy 21) and on mental age to 50 mainstream school boys (controls). The controls were further divided into those matched on "poor attention" to the fragile-X boys and a "good" attention group, as rated by the comprehensive teacher rating scale (ACTeRS) questionnaire. Both fragile-X and Down's syndrome boys made significantly more repeated responses on targets (but a lower proportion of errors based on confusion of shape) than the two control groups and these differences were stronger in the fragile-X group. In the single target condition, search was for a single type of target throughout. In the dual target condition, participants were required to alternate between two different targets. Fragile-X boys showed significantly greater inability than Down's syndrome and normal boys to switch attention between targets and both learning-disabled groups were inferior to the control groups. Thus, both learning-disabled groups displayed a weakness in inhibiting repetition and in switching attention from one type of target to another and the impairments were more acute in fragile-X boys. The results provide further support for an attention deficit in this population at higher levels of attention control/executive functioning that involve switching visual attention and inhibiting repetitious behaviour. © 2002 Elsevier Science Ltd. All rights reserved.

Keywords: False alarms; Inhibition; Response switching; Response repetition; Down's syndrome; Fragile-X syndrome

1. Introduction

Fragile-X syndrome is a well-recognised cause of developmental delay in males and to a lesser extent in females. The causative mutation is an expansion of the CGG repeat at the beginning of the FMR-1 gene [9,32]. The clinical manifestations of the disorder have been recognised since 1943 and the world literature is now extensive [12]. Much recent research has been focused on defining the pattern of neuropsychological deficits that associate with fragile-X syndrome, in essence to link genotype to phenotype [1,4,5,15,19,28,29]. At a behavioural level attention problems and hyperactivity represent a prominent behaviour problem in fragile-X, especially in males [2,3,11,13,14,30]. At a cognitive level, the attention deficit has been explored in a series of detailed studies. Munir et al. [23] and Cornish et al. [6] compared the performance of fragile-X males (FX) with that of Down's syndrome (DS) males and normal males, matched for mental age, on a wide range of tasks testing var-

ious aspects of attention. They concluded that the FX group showed specific deficits in "selective attention, divided attention and inhibition, with a particular impairment in the ability to plan and organise search, shift attention from one concept to another, delay responding and inhibit task-irrelevant responses". This pattern of deficit broadly suggests an impairment of executive function. Interestingly, this type of deficit has also been reported as a prominent characteristic in females with FX FMR-1 full mutation. Mazzocco et al. [16–18] found that FX females had pronounced deficits on tasks typically referred to as tapping "frontal lobe" processing compared to controls matched on age and IQ. These deficits appeared to be relatively independent of IQ, in that intellectual level did not significantly affect performance. There is a growing consensus, however that the term "executive function" will need to be defined in more detail if we are to fully understand the causes of behavioural malfunctioning in a variety of clinical syndromes. This term has been used to encompass a number of postulated processes ranging from general notions, such as planning, to more specific processes, such as switching, updating and inhibiting [21]. Nigg [24] has distinguished

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eight different types of inhibition. It is as yet unclear whether features of the FX syndrome (and other syndromes which have been ascribed to executive impairment) can be attributed to a deficit in one or more of the above processes or in some other component of behaviour.

The present paper presents a detailed analysis of one aspect of performance in the study described by Munir et al. [23] in an attempt to elucidate further the impairment presented by the FX group in that study. The task required a computer display to be searched for specified target shapes, and the mouse button to be clicked after positioning the mouse pointer over the targets. Both FX and DS participants produced a very large number of false alarms in this task (i.e. responses to non-target items). However, such responses can arise for a number of reasons, such as poor discrimination of targets from non-targets, poor ability to locate the computer mouse on the target at response, impulsive responding, repetitive responding, etc. Further investigation may help to clarify the nature of the FX (and DS) impairment. Did these participants, e.g. make more false alarms of the same kind as the control groups, performing like controls, but less efficiently, or did their false alarms arise from some impairment specific to their condition and manifest different characteristics from those demonstrated by the control groups?

Closer analysis of the nature of the errors and of differences between groups would help to pinpoint the precise nature of the inferior performance in the learning-disabled groups and the underlying neurological deficits. In particular, it might be possible to be more precise about a vaguely defined deficit in executive function. For example, a specific problem in shape discrimination would implicate a very different underlying problem from a difficulty in switching targets when alternation between different types of target is required. Current research is attempting to clarify the differences between different clinical groups which have been loosely ascribed to general deficits in executive function [25,26], and the present analysis attempts to contribute to these ends in the case of FX syndrome.

2. Method and procedure

2.1. Participants

These have already been described in detail elsewhere [23] and will only be outlined briefly here. The study involved four groups of participants: (1) 25 boys with FX syndrome (age range 8.06–15.09 years; mean age 10.88 years); (2) 25 boys with Down's syndrome (age range 7.04–15.09 years; mean age 11.17 years). However, for present purposes, data were only available from 12 participants in this group, due to loss of some data in the interval between the original and the present analyses; the mean MA and performance on the task were, however, very similar in this subgroup to those for the larger group; (3) 25 control boys matched on *mental* age, and therefore, biologically younger

than the FX group (age range 5.02–10.09; mean age 7.58 years). These boys were individually matched to members of the fragile-X group on age and degree of hyperactivity and poor attention abilities as rated by their teachers on the comprehensive teacher rating scale (ACTeRS) [31], and therefore, were referred to as the "poor attention" control group; and (4) 25 control boys also matched on *mental* age, and therefore, biologically younger than the FX group (age range 5.02–10.09 years; mean age 7.97 years), but rated by teachers as demonstrating average to good attention ability and low hyperactivity; they were referred to as the "good attention" control group. The control groups were drawn from five different schools, two secondary and three primary.

All groups were matched on verbal mental age (VMA) using the British Picture Vocabulary Scale (BPVS) short form [8]. Teachers of the FX syndrome, Down's syndrome and control boys completed a shortened form of the ADD-H comprehensive teacher rating scale (ACTeRS) [31]. Of necessity, these ratings were provided by a number of different teachers, who had direct experience of each child.

2.2. Materials

The visual search task has been described in detail elsewhere [23]. In brief, a display was presented consisting of a river, trees and a variety of different coloured "holes" on a green background. The single target was either a vertical black ellipse (target 1) or a horizontal pinkish-brown ellipse (target 2). There were 25 target shapes randomly positioned around the display among 100 shapes in all.

The instructions were: "you need to find the king monster, by first finding all the hidden monsters, who are hiding behind the black eggs standing upright/the pink eggs lying on their sides". In all cases, the 20th target located was always the king, but the program ended if the mouse was clicked 50 times without locating 20 targets. When a target was clicked on, a square red monster face appeared and remained visible throughout the remainder of the run. When the 20th target was located, a larger version of the face, with a crown, appeared. Two runs were completed after demonstrations and practice, the first with target 1 and the second with target 2. In the dual target task, targets were both vertical black ellipses and horizontal pinkish-brown ellipses. There were 15 examples of each of these. The child was required to click alternately on the black targets and pinkish-brown targets.

The measures taken were hits, false alarms, time per hit in seconds and mean distance travelled between hits. Results on these measures have been reported previously. For the present analysis, the errors (false alarms) on the single target visual search were subdivided into the following mutually exclusive categories: repeated responses on targets (other than returns to targets following responses to other shapes), returns to a previously detected target, responses to targets of the same shape, but a different colour from the target, responses to targets of a different shape, but the same colour, responses to targets differing in both shape and

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