Nature of the Working Memory Deficit in Fragile-X Syndrome

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Working memory performance in a group of young Fragile X males with FMR-1 full mutation was compared to a learning disabled comparison group comprising Down’s syndrome males and two control groups of mainstream schoolchildren. Performance was assessed on a battery of tasks tapping the three components of working memory—phonological loop, visual–spatial sketch pad, and the central executive. The results indicated that the Fragile X group displayed a general impairment on working memory tasks that cannot be attributed to a single working memory component per se. Instead, the results suggest that Fragile X males have a working memory deficit that may be attributed to how much attentional resource a specific task requires and their overall available executive capacity, irrespective of the working memory subsystem.

Key Words: working memory; fragile-X syndrome; phonological memory; visuo-spatial memory; executive skills.

INTRODUCTION

Working memory deficits in which individuals are unable to hold information in mind (as well as others) have received considerable attention in recent years, in part because of an accelerated growth in “behavioral phenotype” research which has attempted to explain brain–behavior correlates in neurodevelopmental disorders, such as autism (e.g., Bennetto, Pennington, & Rogers, 1996; Hughes, Russell, & Robbins, 1994; Ozonoff, Pennington, & Rogers, 1991), and in part because of the rapid advancing development in

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functional magnetic resonance imaging which allows examination of brain activation in human subjects during performance on working memory tasks (e.g., Cohen, Perlstein, et al., 1997; Courtney, Petit, et al., 1998; Ferreira, Verin, et al., 1998; Smith, Jonides, et al., 1996). The term working memory refers to a brain system that provides temporary storage and retrieval of the information necessary for successful completion of higher cognitive functions, such as language and problem solving. The most current, complete specification of working memory is the model proposed by Baddeley and Hitch (1974), revised by Baddeley in 1986, in which working memory is divided into a three-unit system: the central executive, which is assumed to be an attentional controlling system of limited capacity and is important for skills such as problem solving and planning, and its two slave systems: the phonological loop, which stores and rehearses verbal information and whose capacity is limited by the speed at which the articulation can be performed, and the visuospatial sketch pad, which processes and manipulates visual and spatial information. Several cognitive tasks are reported to tax these different systems. For example, tasks that require immediate verbal recall primarily involve the function of the phonological loop (Daneman & Carpenter, 1980; Turner & Engle, 1989), while tasks that require planning (Shallice, 1982, 1988; Shallice & Burgess, 1991) or organization of sequential material (Morris & Jones, 1990) involve central executive resources.

The extent to which poor performance on measures of cognitive function in children with neurodevelopmental disorders reflects a specific impairment in working memory deficit has recently been addressed in two studies of autistic individuals (Bennetto et al., 1996; Russell, Jarrold, & Henry, 1996). The findings suggested that children with autism are significantly more impaired on working memory tests than normal developing children matched on mental age, but not compared to other children with a similar level of learning disability. Russell et al. (1996) conclude that children with autism and with moderate learning disability may have a less efficient central executive capacity than those of normally developing children because of significant neurological impairments.

The Fragile-X syndrome (Hagerman & Cronister, 1996) has been the subject of considerable interest in recent years, not least because of early reports of its possible association with autism (Brown et al., 1982, 1986; Blomquist et al., 1985; Gillberg, Wahlstrom, & Hagberg, 1985; Wahlstrom et al., 1986; Reiss & Freund, 1990; Turk, 1992). Fragile-X syndrome (FXS) is an X-linked genetic disorder affecting approximately 1 in 3–4000 males (Jacobs et al., 1993; Turner et al., 1996) and represents the most common inherited identifiable cause of learning disability. In common with other genetically disordered groups that present with mental handicap (e.g., Down’s syndrome), cognitive deficits are broad, although weaker nonverbal skills than verbal skills have been suggested by a number of studies (Crowe & Hay, 1990; Hodapp, Leckman, et al., 1992; Freud & Reiss, 1991). Recent findings
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