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Domain specific attentional impairments in children with chromosome 22q11.2 deletion syndrome

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Abstract

One of the defining cognitive characteristics of the chromosome 22q deletion syndrome (DS22q11.2) is visuospatial processing impairments. The purpose of this study was to investigate and extend the specific attentional profile of children with this disorder using both an object-based attention task and an inhibition of return task. A group of children with the disorder was compared in these tasks with a group of age-matched typically developing children. The children with DS22q11.2 demonstrated impaired spatially based orienting which is consistent with previous findings in this group. Strikingly, the children with DS22q11.2 also demonstrated an improved ability to use object-based cues, relative to the typically developing group. Finally, the children with DS22q11.2 demonstrated an intact inhibition of return system, however, it appears to be delayed developmentally.

Keywords: 22q11.2; Inhibition of return; Spatial attention; Object attention

Chromosome 22q11.2 deletion syndrome (DS22q11.2) results from a 1.5 to 3 Mb microdeletion on the long (q) arm of chromosome 22 (Driscoll, Budarf, & Emanuel, 1992). This disorder encompasses a number of other previously described disorders, such as, DiGeorge syndrome (DiGeorge, 1965), Conotruncal Anomaly Face (Burn et al., 1993) and Velocardiofacial syndrome (Shprintzen et al., 1978), and is one of the most common genetic causes of mental retardation and psychopathology. The currently accepted prevalence rate of DS22q11.2 is at least 1 in 4000 live births (Burn & Goodship, 1996).

The physical manifestations of DS22q11.2 are variable between individuals and include cleft palate, velopharyngeal insufficiency, congenital heart defects, hypocalcemia, and facial dysmorphisms (Emanuel, McDonald-McGinn, Saitta, & Zackai, 2001; Shprintzen, 2005). Recently, a series of studies using magnetic resonance imaging (MRI) have reported widespread brain dysmorphology in children and adults with DS22q11.2. The most consistent finding is an anterior to posterior pattern of greater tissue volume reductions, particularly in white matter (Eliez, Schmitt, White, & Reiss, 2000; Kates et al., 2001; Simon et al., 2005c). In addition to these global changes in brain morphology, a number of specific brain regions appear to be affected including the thalamus (Bish, Nguyen, Ding, Ferrante, & Simon, 2004), the cerebellum (Eliez et al., 2001; Bish et al., 2006), the basal ganglia (Eliez, Barnea-Goraly, Schmitt, Liu, & Reiss, 2002), and the corpus callosum (Shashi et al., 2004; Simon, Bearden, McDonald-McGinn, & Zackai, 2005a).

The neurocognitive profile for individuals with DS22q11.2 is defined generally by an overall delay in cognitive development, including psychomotor and language delays and an IQ in the range of 70–85 (Gerdes et al., 1999; Swillen, Vogels, Devriendt, & Fryns, 2000). Impairments in specific cognitive domains have recently been reported. Bearden et al. (2001) established that one of the

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primary functional impairments in DS22q11.2 is in the domain of visual–spatial processing. The pattern of impairments is likely to include the temporal domain (Debbane, Glaser, Gex-Fabry, & Eliez, 2005), as well as the numerical processing domain (Simon et al., 2005a). Additionally, recent reports have established executive attention and inhibitory control as problematic in this population (Sobin et al., 2004; Bish, Ferrante, McDonald-McGinn, Zackai, & Simon, 2005). While knowledge of these impairments is very important, there is a need to further specify the precise nature of the impairments in this group in order to provide a foundation for cognitive remediation efforts.

It seems, therefore, that a deeper investigation into the spatiotemporal domain is likely to generate important data elucidating the nature and extent of processing impairments, as well as possible compensatory strengths in children with DS22q11.2. These could then be used as a basis for intervention and remediation. In this paper we extend our previous analyses into two closely related spatiotemporal aspects of visual attention, that we have previously claimed to be a key foundational cognitive competence for children with DS22q11.2 (Simon et al., 2005a). Specifically we demonstrated that children with DS22q11.2 are impaired in the ability to effectively disengage attention from an invalidly cued location and re-engage attentional processing in a new target location (Simon et al., 2005b). This impairment in visuospatial processing seems to extend into the ability to move between and enumerate greater than four objects (Simon et al., 2005b). Similar in nature to space-based attention, object-based attention involves the ability to use cues within an object to enhance the processing of target information elsewhere within the confines of the same object. While similar in function to space-based attention, object-based attention is thought to be an independent process (Farah, Wallace, & Vecera, 1993; Duncan, 1984), and is likely to depend on a different neurological mechanism (Egly, Driver, & Rafal, 1994a). Since previous investigations of DS22q11.2 have focused primarily on spatial processing, what remains unclear is whether or not the impairments in space-based attention extend into the domain of object-based attention. This is because spatial processing tasks involve target objects, some characteristics of which may influence performance. Without tasks specifically to disambiguate the effects of each, the ability to process different types of information carried by target stimuli cannot be assessed.

Despite the known temporal judgment difficulties recently reported by Debbane et al. (2005), very little is known about the nature of temporal processing in children with DS22q11.2. Inhibition of return (IOR) is a phenomenon that depends on the temporal dynamics of visuospatial attentional processing. Therefore, it is likely to be revealing when used as a vehicle to explore the temporal dynamics of visuospatial attention in DS22q11.2. IOR is a counterintuitive pattern of performance in which attention to recently processed items is inhibited in order to give preference to novel ones. It has been demonstrated that when human observers are provided with a visual cue to the location of a subsequently appearing target, processing of that target is typically enhanced or facilitated (Posner, Rafal, Choate, & Vaughan, 1985). However, several studies have found that facilitation of target processing occurs in typical adults provided that the length of time between the cue and target is short (100–350 ms). When the length of time between the cue and target increases (>500 ms) processing of the subsequent target location becomes inhibited (Posner et al., 1985; Maylor, 1985; Klein, 2000). In other words, the processing of the target location is now delayed rather than speeded up simply as a result of lengthening the SOA. The adaptive function of IOR has been assumed to be the facilitation of efficient visual search in a crowded environment. For example, when visually scanning a crowded scene, inhibiting processing in previously attended locations, if the target does not appear quickly, becomes advantageous (Klein & MacInnes, 1999). Thus, IOR can be seen as optimizing limited visuospatial attentional resources. Thus, changes in the temporal dynamics of IOR in children with DS22q11.2, may be related to impairments shown in other visuospatial tasks.

The purpose of the experiments reported in this paper was to examine whether children with DS22q11.2 have impairments in object-based attention to the same extent as the impairments shown in space-based attention. Additionally, we were interested in exploring whether temporal judgements shown to be impaired in this group (Debbane et al., 2005) extend to impairments of the temporal dynamics of visuospatial attention, as in IOR.

1. Method

1.1. Participants

A total of 30 children aged 7–14 participated in the study, which consisted of the two experiments described below. Of these 30, 15 were children with DS22q11.2, with diagnosis of the disorder confirmed by molecular Fluorescence In Situ Hybridization (FISH) between the ages of 6 and 14. The mean age of the DS22q11.2 was 9 years, 1 month ($SD = 2.37$) and the group consisted of seven females and eight males. Eleven of these children were recruited through their participation in the Velocardiofacial Syndrome Education Foundation Annual Conference (Atlanta, 2004). The other four children with DS22q11.2 were recruited through their ongoing involvement in the "22q and You" clinic at the Children's Hospital of Philadelphia. The remaining 15 children were typically developing children that were age matched to the DS22q11.2 group between the ages of 7 and 14. The control group had a mean age of 9 years, 7 months ($SD = 2.03$) and consisted of eight females and seven males. Control children were recruited from the general population. A t-test to compare the group ages revealed a non-significant result, $t(28) = -.671$, $p = .508$. Both parental consent and child assent was collected prior to all experimentation. All participants were treated in accordance to the ethical