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Visuo-spatial construction trajectories in Fragile X Syndrome (FXS) and Autism Spectrum Disorders (ASD): Evidence of cognitive heterogeneity within neurodevelopmental conditions

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ABSTRACT

Background/Aims: There have been discrepancies reported in visuo-spatial construction ability in children with Autism Spectrum Disorders (ASD), fragile X Syndrome (FXS) and those with a comorbid diagnosis of FXS and ASD (AFXS). This study aimed to provide a better understanding of the visuo-spatial processing styles in these heterogeneous neurodevelopmental disorders.
Methods and procedure: Navon-type tasks were used to assess visuo-spatial construction ability across 5 groups of children: typically developing, FXS, AFXS, ASD children who scored low–-moderate (HFA) and ASD children that scored severe (LFA) on the Childhood Autism Rating Scale (CARS). Analyses of their developmental trajectories compared the performance of these groups.
Outcomes and results: Each group produced their own distinct trajectory. HFA achieved higher scores from an earlier age than the TD group, while the LFA group's performance was driven by a bias in local processing. The FXS performance was normalised by using mental age as a predictor while neither mental nor chronological age predicted the AFXS group performance.
Conclusions and implications: The study showed unique processing styles. These findings highlight the importance of taking comorbidity and the severity of symptoms within each condition into account in order to understand cognitive abilities and cognitive profiles.

What this paper adds?

This study used the Navon-task paradigm to explore the visuo-spatial construction ability in children with FXS and with ASD, including a novel task that minimised the fine motor demands. The paper presents the results from a large sample in a wide age-range (160 children aged between 3 and 18 years). Participants were grouped not only as a function of their neurodevelopmental condition but also in terms of the severity of their symptoms and diagnostic comorbidity, as follows: 1. TD (typically developing children) 2. HF ASD – Mild to moderate symptoms 3. LF ASD – Severe symptoms 4. FXS and 5. Comorbid FXS + ASD (AFXS). An analysis of the developmental trajectories of these groups performance showed not only how the "atypical" groups differed from the typical trajectory but also revealed both intra and inter- group differences for the neuro-developmental conditions. The two ASD groups showed different cognitive processing styles in the tasks, a finding that has implications for the current theories on the integration of information in ASD. The FXS group performed closer to what was expected by their mental age, whereas neither mental age nor chronological age predicted the performance of the FXS + ASD group. These findings provide information about the rate of development and age of onset of these skills, rather than the mere absence or presence of an ability, which is valuable to better understand the cognitive profiles of these two conditions and its implications for intervention.

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1. Introduction

In the last two decades, developmental research has focused on cognitive phenotypic outcomes of neurodevelopmental disorders (Karmiloff-Smith, 1998, 2007, 2009). Cognitive delays are not consistent across tasks or throughout development. Recently, findings from a number of developmental studies have highlighted the dynamic role of development in shaping disorder- specific profiles from infancy through to adulthood (e.g., Cornish, Scerif & Karmiloff-Smith, 2007; Hall, Burns, Lightbody & Reiss, 2008). In this context, neurodevelopmental disorders with a clear genetic aetiology and recognised phenotype can help to inform other disorders where the genetic pathway is still unknown, by linking heterogeneous behaviours. Fragile X syndrome (FXS) is such a disorder; it is the result of a cytosine-guanine-guanine (CGG) expansion in the 5′ untranslated region of the *fmr-1* gene and the resulting decreased expression of its associated protein FMRP (Koukoui & Chaudhuri, 2007). Approximately 30% of individuals with FXS display autistic- like behaviours that are significant enough for a comorbid diagnosis of autism spectrum disorders (ASD) and as a result, the syndrome represents one of a small number of known single gene causes of ASD (Miller & McIntosh, 1999). Similarities between the two disorders are mainly shown in the behavioural rather than the cognitive domain (McDevitt, Gallagher & Reilly, 2015). One apparent similarity at the cognitive level however, is that both individuals with FXS and ASD show unusual profiles in visuo-spatial processing (Amso & Scerif, 2015; Gallego et al., 2014; Ballantyne & Núñez, 2016).

Whilst there are reported relative strengths in visuo-spatial *perceptual* tasks in FXS (Ballantyne & Núñez, 2016; Cornish, Munir & Cross, 1999; Hodapp et al., 1992; Maes, Fryns, Van Wallegham & Van de Bergne, 1994), deficits have been shown in visuo-spatial *construction* tasks (Pegoraro, Steiner, Celeri, Banzato & Dalgalarrondo, 2014; Cornish et al., 1999). However, the unusual visuo-spatial processing of individuals with ASD has been extensively researched. ASD individuals show an unusual local/global bias in processing visual information, where 'local' suggests a 'detailed piecemeal' analysis of a visual scene and global refers to the overall meaning of a visual scene (Plaisted, Swettenham & Reiss, 1999; Happé, 1999; Happé & Frith, 2006; Mottron et al., 2006).

The local bias shown in individuals with ASD has often been demonstrated through construction tasks, such as the block design task or drawing task (Van Eylen, Boets, Steyaret, Wagemans, Noens, 2015; Shah & Frith, 1993; Pring et al., 1995, Mottron, Belleville, & Menard, 1999). There is however a drawback in using these tasks, due to the added processing demands they place on the individual which are not specifically visuo-spatial. For example, drawing is a complex task which depends on the successful integration of complex functions, such as planning and motor control, and therefore employs several cognitive abilities to ensure task completion (Sommers, 1989). Nevertheless, studies looking at performance on drawing tasks were central to the development of key theories within the ASD literature such as weak central coherence (WCC; Frith, 1989) and enhanced perceptual functioning (EPF; Mottron & Burack, 2001). WCC posits that localised processing results from a failure to attend to the global or meaningful items of a stimulus, whereas EPF argues that global and local processing operate independently of each other.

Impairments among FXS populations have been shown on drawing, pegboard and block design tasks (Pegoraro et al., 2014; Crowe and Hay, 1990; Cornish et al., 1998, 1999). Cornish and colleagues highlighted dissociations between visuo-spatial perception and visuo-spatial construction tasks, finding a deficit among the FXS group in the latter and a relatively strong performance in the former. It is not surprising that these tasks were harder for the FXS individuals, as construction tasks such as drawing require a more detailed analysis of the stimuli than perceptual attention and detection tasks. Additionally, the reproduction of a correct configural formation of the stimuli would necessitate the formation of the correct spatial arrangement of the parts in relation to each other.

An alternative way to assess visuo-spatial hierarchical processing is the well-known Navon task (Navon, 1977). This task uses hierarchical stimuli to assess visual attention. Traditionally the stimuli are made of letters, with a large global letter that can either be congruent or incongruent to the smaller, local letters. Navon (1977) found a global processing bias in typically developed adults, a finding which is inconsistent and seems to depend more on the experimental paradigm (e.g. Wang, Mottron, Peng, Berthiaume & Dawson, 2007; Plaisted et al., 1999). Nonetheless, studies assessing hierarchical visuo-spatial processing in developmental disorders have utilised the Navon task to demonstrate perceptual and construction abilities (Ballantyne and Núñez, 2016; D'Souza, Booth, Connolly, Happé, Karmiloff-Smith, 2015; Bernardino et al., 2012; Plaisted et al., 1999; Farran et al., 2003). These have provided varying results but evidence towards unique processing styles (see e.g., López, 2008).

Using Navon stimuli, Ballantyne and Núñez (2016) found that performance on hierarchical tasks among individuals with FXS and ASD was dependent upon diagnosis and severity of ASD symptoms. However, these were assessed based on their performance in perceptual, rather, than construction tasks. Therefore, it remains unclear whether and how these two developmental disorders differ in their construction abilities. The current study aimed to investigate visuo-spatial construction ability in ASD and FXS in order to examine how the developmental pathways differ. In line with Ballantyne and Núñez (2016) and other cross-syndrome comparison studies (Thomas et al., 2009; Dimitriou, Leonard, Karmiloff-Smith, Johnson & Thomas, 2015), developmental trajectories were built to compare change in performance across age observed in each group.

The current study utilised two different Navon-type tasks as follows. (1) A drawing construction task that is similar to those used in research in other developmental disorders such as Williams syndrome (Farran et al., 2003). (2) A novel magnet construction task with cut-outs as the local items for the purpose of minimising the impact/confound that any fine motor impairment may have on task performance.

The ASD group was split into a high functioning group (HFA) and a low functioning group (LFA) based on the severity of symptoms as measured by the Childhood Autism Rating Scale (CARS). The FXS group was divided was depending on whether FXS was present with or without ASD (as measured by the CARS) and a group of FXS + ASD (AFXS as measured by severity of ASD symptoms on the CARS). This is in line with recent research that showed differences in group performance depending on severity of ASD symptoms using measures such as the CARS (Ballantyne & Núñez, 2016; Riby & Hancock, 2009; Gillespie-Smith et al., 2014). School records indicated that the children did not have any other diagnoses. Based on previous findings on their performance on