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Visuo-spatial construction trajectories in Fragile X Syndrome (FXS) and Autism Spectrum Disorders (ASD)

Ballantyne, Carrie; Nunez, Maria; Manoussaki, Kallia

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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 – 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.

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, Burris & Rivera, 2014; Ballantyne & Núñez, 2016).

Whilst there are reported relative strengths in visuo-spatial *perceptual* tasks in FXS (Ballantyne & Núñez, 2016; Cornish, Munir & Cross, 1999; Hadapp, Leckman, Dykens, Sparrow, Zelinsky & Ort, 1992; Maes, Fryns, Van Wallegham & Van de Bergne, 1994), deficits have been shown in visuo-spatial *construction* tasks (Pegoraro, Steiner, Celeri, Banzato & Dalgalarondo, 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, Dawson, Soulières, Hubert & Burack, 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, Hermelin & Heavey, 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, Steiner, Celeri, Banzato & Dalgarrondo, 2014; Crowe & Hay, 1990; Cornish, Munir & Cross, 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 the 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; Bernatdino, Mouga, Almeida, van Asselen, Oliveria & Castelo- Branco, 2012; Plaisted, et al., 1999; Farran, Jarrold & Gathercole, 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, Annaz, Ansari, Scerif, Jarrold & Karmiloff-Smith, 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 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, Doherty-Sneddon, Hancock & Riby, 2014). School records indicated that the children did not have any other diagnoses. Based on previous findings on their performance on visuo-spatial tasks and more specifically visuo-spatial construction tasks (e.g. Muth, Hönekopp, and Falter, 2014), it was expected that, the HFA group would perform in line with their chronological age matched typical peers. On the other hand, it was expected that the LFA group would show a preference in drawing local stimuli as opposed to global as found in previous visuo-spatial hierarchical studies (e.g. Ballantyne & Núñez, 2016). In regards to the FXS groups, it was predicted that performance would be more in line with their mental age (MA) as previous studies have found that task performance normalises when MA rather than chronological age (CA) is used as a predictor (Ballantyne & Núñez, 2016; Cornish, Cole, Longhi, Karmiloff-Smith & Scerif 2012; 2013; Scerif, Longhi, Cole, Karmiloff-Smith & Cornish, 2012).

As these two types of tasks may tap on slightly different motor skills, the method and results of the trajectories on each measure will be presented separately.

2. Drawing Task

2.1 Method

2.1.1 Participants

Participants were 20 boys with ASD (with mild-moderate ASD symptoms as measured by the Childhood Autism Rating Scale –CARS : HFA, mean age = 13 years 9 months; age range 9 years 3 months to 16 years 2 months), 20 boys with ASD (with severe ASD symptoms as measured by the CARS: LFA, mean age = 10 years 1 month; age range 7 years 7 months to 16 years 1 month), 21 boys with FXS (mean age = 13 years; age range 7 years, 7 months to 17 years 2 months), 19 boys with FXS + ASD (AFXS) (mean age 12 years 7 months; age range 6 years 11 months to 17 year 10 months) and 80 boys with typical development (TD; mean age 9 years 5 months age range 3 years 0 months – 16 years 11 months). See Table 1 for group details. The age range of the TD sample permitted comparisons to be made between disorder and TD trajectories on the basis of either chronological age (CA) or mental age (MA) where disorder groups may have lower MAs. Teachers of the TD group completed the Strengths and Difficulties Questionnaire (Goodman, 1997) to ensure that there were no underlying symptoms that had not received a clinical diagnosis. All other participants had been diagnosed by clinicians and satisfied the diagnostic criteria for ASD according to the DSM-V (APA, 2013) as

recorded on their school record of needs. The Childhood ASD Rating Scale was completed by teachers (CARS; Schopler, Reichler, & Rocher Renner, 1988).

No other diagnoses were noted for the clinical groups.

Nonverbal IQ was assessed by the Raven's Progressive Coloured Matrices (Raven, Raven & Court, 1990). This was then compared to a typically developing group of children (TD). For the TD group, CA predicted 80% of the variance in the RPCM therefore showing predictable performance in line with their chronological age.

2.1.2 Stimuli

Participants had to complete four drawings. Navon style letters were given to the children to copy (large S made up of small X's, Figure 1). The stimuli were presented in A5 booklets. The hierarchical items were separated into global items that were made up of 21 local items. These appeared congruent (same local as global items) or incongruent (different items at local level than global level). The letters C and T were used in the first half of the task and the letters S and T were used for the second half. Participants were given different letter tasks for the drawing and magnet tasks and the trial order was counterbalanced across participants.

2.1.3 Procedure

Participants were shown hierarchical figures as described above in an A5 booklet. Each hierarchical figure was presented individually to the participant. The experimenter emphasised that the stimulus was a large letter made up of smaller letters and that they were required to *copy the figure as accurately as*

possible onto a booklet provided. The figure remained in front of the participant throughout. Although the task had no time constraints, participants did not continue with the task for more than 10 minutes before a break was given.

2.1.4 Scoring

The experimenter and an independent rater scored the quality of all drawings. Raters were not given information about what group the participants belonged to. The rating scale was based on that of Dukette and Stiles (2001) and inter-rater reliability was calculated by a Pearson correlation, $r = .92$. The quality of the global and local items was scored separately using a separate but comparable ordinal 6 – point scale, ranging from 0 -5. In any given stimuli type the maximum points a participant could be awarded was 20. Points were given for accuracy, orientation, spacing and number of elements. For the global scale, points were given when the orientation matched that of the figure presented. Spacing accuracy was defined in terms of the presence and uniformity of spacing between the elements that made up the global form. Within the local scale, points were given for the correct number of elements, spacing between elements, accuracy and uniformity regardless of whether they made up the global form.

2.2. Results

****Insert Figure 3 here****

2.2.1 Developmental Trajectories

The developmental trajectories are examined in terms of main effects and interactions. A significant main effect refers to a difference between scores at the earliest measurable age (i.e. the age of the youngest participant) and would

refer to a delay at onset. A significant 2 – way interaction of group * accuracy would imply a difference in the rate of development between the TD group and the other four groups. The interaction of hierarchical level, CA or MA, and group was also analysed to investigate whether the groups had a similar developmental relationship in task completion to the TD group – i.e. whether the clinical groups develop in the same way as the TD group. These main effects and interaction were compared to their CA (allowing for experience) or their MA (their developmental level).

2.2.2 Data Analysis

Accuracy data was taken from all participants in the study. Trajectories were analysed using a fully factorial analysis of co-variance (ANCOVA) for the clinical groups, where the within-participants factors hierarchical level (local, global) and chronological age was the co-variant. A direct comparison of each clinical group to the TD group was carried out using an ANCOVA (3x3) with Group (TD group compared to 4 clinical groups) as between- participants factors, within-participant factors of hierarchical level, and age as co-variant. Additionally, some disorder groups were compared to each other to explore detailed similarities and differences. Table 2 displays accuracy scores on each of the drawing tasks.

****Insert Table 2 here****

The HFA group were the highest performing group achieving scores near ceiling across local and global conditions. The LFA group showed a vastly different profile to the HFA group with low scores, high variability, and higher

local than global scores. Although performance for the FXS group was lower, it was consistent across both local and global items. The AFXS group was the lowest performing group overall, with scores close to floor.

2.3.1 TD group

Comparison of the global and local conditions revealed that they both improve significantly with age. There was a difference in local and global from the age onset (main effect: $F(1, 78) = 7.03, p = .01$) and in the rate of increase of accuracy scores with age (interaction of hierarchical level and CA: $F(1, 78) = 7.36, p = .01$). Accuracy performance of the local and global conditions remained reasonably linear until approximately 10:05 years, when the performance of the global condition appeared to increase at a faster rate than the local condition. These are shown in Figure 2a.

2.3.2 HFA group

The accuracy performance of the HFA group improved significantly overall with age (main effect of CA: $F(1, 18) = 5.91, p = .03$) and there were no differences in performance at the global and local levels (main effect of hierarchical level: $F(1, 18) = .15, p = .70$; interaction of hierarchical level and CA: $F(1, 18) = .20, p = .66$).

2.3.3 LFA Group

Analysis of the LFA developmental trajectories of global and local drawings revealed that their performance did not significantly improve with age ($F(1, 18) = .17, p = .69$). There were also no notable differences between performance at

the local and global levels (main effect of hierarchical level: $F(1, 18) = 1.64, p = .22$; interaction of hierarchical level and CA: $F(1, 18) = .45, p = .51$). The LFA group experienced a large amount of variance within their scores. Figure 2c suggests overall higher accuracy scores in the local items, which may be masked by the high variability.

2.3.4 FXS Group

Performance was extremely poor for the FXS group and there was a large amount of variance. As Figure 2d illustrates, the developmental trajectories of global and local accuracy were almost flat, and there was no meaningful improvement with age ($F(1, 19) = .04, p = .85$). There were no significant differences in the performance of local or global accuracy (main effect of hierarchical level: $F(1, 19) = .11, p = .74$; interaction of hierarchical level and CA: $F(1, 19) = .13, p = .72$).

2.3.5 AFXS group

The performance of accuracy scores of local and global drawings did not show any overall improvement with age ($F(1, 17) = 2.44, p = .14$). Performance accuracy also was not significantly modulated by the different hierarchical levels (main effect of hierarchical level: $F(1, 17) = .94, p = .35$), although a trend did appear that indicated that this changed across developmental trajectory, with higher scores emerging in the local condition (interaction of hierarchical level and CA: $F(1, 17) = 4.19, p = .06$).

2.4 Comparisons of groups

Comparisons within and across syndrome groups were also carried out. These included comparisons between HFA and LFA, FXS and AFXS and LFA and AFXS.

2.4.1 HFA and LFA trajectories

The HFA and LFA groups showed a different pattern of accuracy on the tasks, with the HFA group performing more accurately on the global drawing and the LFA group performing more accurately on the local drawings (interaction of hierarchical level x group: $F(1, 38) = 13.34, p = .00$).

2.4.2 FXS and AFXS trajectories

The performances of the two FXS groups' did not show any significant differences either in the developmental relationship between the accuracy of global and local tasks or their score at onset or rate of development. However, the large amount of variance may have obscured any differences in terms of overall accuracy.

2.4.3 AFXS and LFA trajectories

Overall, the comparisons of the AFXS and LFA trajectories revealed the LFA group performed more accurately on global and local tasks (main effect of task: $F(1, 37) = 11.95, p = .00$).

2.5 Changes over MA equivalent

RPCM predicted accuracy scores for only the LFA and FXS group (LFA group: $F(1, 18) = 5.07, p = .04$; FXS: $F(1, 19) = 6.02, p = .02$). Table 3 provides a summary. The HFA group are showing a delay in onset and a delay in the rate of development when using MA as a predictor and the FXS group shows a delay at onset. The only other difference noted is the LFA group is showing a different developmental relationship in task performance. This is reflected by their attention to local items.

****insert Table 3 here****

2.6 Discussion

The hypothesis that the HFA group would show a similar developmental relationship on drawing global and local items to the TD group was upheld and no differences were noted. The other atypical groups all experienced a substantial amount of variance in their scores, which may have masked any trends in performance. This was particularly applicable to the LFA group that appeared performing more accurately in the local drawings than the global, which is similar to young TD children. This performance is consistent with the literature suggesting that visuo-spatial construction is a strength in the ASD profile (Muth et al., 2014; Shah & Frith, 1993; Pring, Hermelin & Heavey, 1995; Mottron, et al., 1999) and the delays shown within the LFA group are perhaps due to the additional overall cognitive delays that they experience. However it is important to note that the LFA and HFA are divided onto groups not based on performance in any cognitive task but rather on their categorisations of severity on the CARS.

Cornish et al, (1998; 1999) and Pegoraro et al. (2014) found that FXS individuals perform poorest on visuo-spatial construction tasks. The current study found that the performance of the FXS children is not only delayed at onset and in the rate of development but that they also show an atypical pattern of development. However, it was the AFXS group that performed at floor level and it is not clear as to whether the results are a reflection of their construction ability or their fine motor skills.

3. Magnets Task

An issue highlighted in the drawing task was the difficulty experienced in fine motor skill required which meant that a true indication of the groups' visuo-spatial construction ability could be masked. By using magnet cut-outs of the local stimuli, the children were able to construct the hierarchical stimuli, with minimal motor control required. It was expected that the HFA group's performance would be in line with the TD groups and that children in the LFA, FXS and AFXS groups would be significantly less accurate and develop at a slower rate than the TD group in the magnet task.

3.1. Method

Participants (See section 2.1.1)

3.1.1 Stimuli and Procedure

The same A5 booklet (but alternate items) was given to the participants. However, for this study children were given a magnetic white board to place magnet cut-outs of the local stimuli in the correct global configuration. Each hierarchical figure was presented individually to the participant. The

experimenter instructed the participants to place the small items in such a way as to look the same as the picture in the booklet. The children were given all 21 local items to use but were not told that they would have to use all the magnets provided. The figure remained in front of the participant throughout. The task had no time constraints but participants did not continue with the task for more than 10 minutes before a break was given.

3.1.2 Scoring

Scoring was based on that of Dukette and Stiles (2001). However, local and global levels were collapsed as the local items were provided as part of the task, therefore one ordinal mark was given for each trial. Digital photographs were taken of each trial and once again, the experimenter and an independent rater scored each trial. Inter-rater reliability Pearson correlation was $r = .94$. Points were given (an ordinal scale marked from 0-10) for correct orientation of local items and global configuration, accuracy on the number of items used and good approximation of spacing between elements.

****Insert Figure 5 here****

3.1.3. Results

Table 4 displays accuracy scores on each of the magnet tasks. The HFA achieved very high scores. The LFA group scored marginally below that of the TD and HFA groups. The FXS group followed in their performance, however their accuracy scores were half that of the HFA group and the AFXS group performed poorest overall.

****Insert Table 4 here****

3.2.1 TD group

The TD group improved with age ($R^2 = .62$, $F(1, 78) = 129.0$, $p = .00$) although the mean score was below that of the HFA group. The gradient was significantly greater than zero and produced a valid developmental trajectory (gradient 22.58; [21.21, 23.95]).

3.2.2 HFA group

The HFA group showed no improvement with chronological age ($R^2 = .00$, $F(1, 18) = .02$, $p = .89$). However they also achieved the highest scores but as the gradient was not significantly greater than zero, the HFA group did not produce a valid developmental trajectory (gradient $-.01$; [$-.13$, $.11$]).

3.2.3 LFA group

The LFA group produced a developmental trajectory that accounted for quarter of the variance ($R^2 = .25$, $F(1, 18) = 5.98$, $p = .03$) but visual inspection shows that they actually became worse as they got older. This gradient was significantly different from zero (gradient $-.04$; [$-.07$, $-.01$]) and therefore they generated a reliable trajectory but with decreasing performance with CA.

3.2.4 FXS group

The FXS group did not generate a valid trajectory as their scores did not account for a significant amount of the variance ($R^2 = .02$, $F(1, 19) = .42$, $p = .53$). This group had the second lowest rate of performance out of all clinical groups (gradient: $-.03$; [$-.14$, $.07$]).

3.2.5 AFXS group

The AFXS group displayed a similar trajectory to the FXS group as they did not generate a valid trajectory ($R^2 = .10$, $F(1, 17) = 1.79$, $p = .20$). The performance was below that of the FXS group and they produced very little increase in performance rate with age (gradient: .02; [-.01, .04]). The groups' developmental relationships in accordance to CA and MA are shown in Table 5.

****Insert Table 5 here****

3.3 Comparisons of different groups

As the LFA group was the only group to produce a valid trajectory we could not provide a reliable comparison between the clinical groups.

3.4 MA as a predictor

Performance on the RCPM test predicted accuracy level for the FXS group ($F(1, 19) = 4.60$, $p = .05$). This suggests that the FXS group are performing at a typical rate in terms of their visuo-spatial ability. See Table 5.

3.5. Discussion

Overall, the HFA group consistently showed the best performance in terms of accuracy followed by the LFA, FXS and AFXS groups. However the performance of the HFA group was atypical in comparison to the TD group as the HFA group showed no meaningful improvement with age. This is possibly because the scores were close to ceiling from the earliest age of measurement. This in itself is an interesting finding, as the HFA group did not follow a typical developmental trajectory, with a superior performance throughout. Rather than

showing improvement, the LFA group showed a negative trajectory, indicating that performance became worse as they got older. Interestingly, Waterhouse and Fein (1984) also found a decline in post-pubertal cognitive skill development in tasks which included perceptual abilities. The LFA group may be following a similar trend in their results. However, the decline in trajectory was only specific to the magnet task. It is important to note that the categorisation of the HFA and the LFA group was based solely the CARS severity of symptoms. It is therefore possible that the performance of the LFA group was affected by task novelty. Stoet and López (2011) found that children with ASD had more difficulty switching between tasks when the rules are arbitrary in a novel task switching paradigm. Although the design differed greatly from the Stoet and López study, it could be that the relative novelty of the magnet task (they did not fit together in a jigsaw like fashion, but were smaller versions of the global picture participants had to construct) in comparison to the drawing task, was enough to impair performance of the ASD children who had greater severity of symptoms. Further investigations need to be carried out to examine if task novelty affected performance.

A decline in cognitive functioning is also something that is more commonly related with FXS (e.g. de Esch, Zeidler & Willemsen, 2014; Dykens, Hodapp, Ort & Leckman, 1993). The two FXS groups performed at the lowest accuracy rate. The FXS groups appeared to either produce a 'zero' rate of increase with age or show a decline with age. However the AFXS group seemed to show improvement in their scores but were overall, the lowest performing group. In sum, the clinical groups all performed atypically to the TD group on the magnet

task and performance was not normalised when MA was used to in the place of CA to build the developmental trajectories with the exception of the FXS group. This is supported by earlier studies (Ballantyne & Núñez, 2016) and differentiates the FXS group from the AFXS group.

4. General Discussion

This paper investigated the developmental trajectories of two neurodevelopmental conditions in their visuo-spatial construction abilities in order to look for both cross-and-within syndrome differences that can be informative at the cognitive level in terms of the heterogeneity of symptoms that they present. The study compared 5 developmental trajectories where 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. Each group produced a distinct developmental trajectory.

As predicted, in terms of the construction tasks as a whole, the HFA group showed greater accuracy than any other group including the TD group but their performance differed between construction tasks as they displayed a normal rate of development in the drawing tasks but a delayed rate in the magnet task. This finding suggests that the two construction tasks may be employing different cognitive skills. Results of the overall accuracy on the construction tasks complimented those by Bernardino et al., (2012), Charman and Baron – Cohen (1993) and Eames and Cox (1994) who found little difference in drawings by individuals with ASD and typically developing individuals and they concluded that non-savant individuals with ASD conceptually analyse items in much the same way as those without ASD. Nonetheless, by using the trajectory

approach, the current study has shown that even if the performance of individuals with HFA is unimpaired in the construction tasks, it does not follow a typical developmental pathway across the two tasks. This questions the assumptions of similarity of the conceptual approach to visuo-spatial construction by the two groups. In the magnet task, the HFA group showed an initial advantage with respect to the TD group but no improvement in later childhood. Even although the reasons of this developmental difference are unclear, the assumption of similarity in cognitive processing needs to be questioned until further research is carried out.

The LFA group also showed differences between the performance on the drawing and magnet tasks. On the drawing tasks they showed the same developmental relationship and improved at a similar rate as the HFA group. Interestingly however, the LFA group were more accurate in the local rather than the global condition; which is in line with the findings by Ballantyne and Núñez (2016) with the perceptual Navon-tasks. More importantly, this finding indicates that the local advantage often reported as a characteristic of ASD processing style may depend on the severity of the symptoms.

These findings also pose a challenge for the current theories of integration of information in autism. On the one hand, the fact that the HFA group appeared to be performing equally as well on the global and local items can be explained by arguing for separate local and global processing operation systems (so that high local accuracy and global accuracy are not mutually exclusive) as the Enhance Perceptual Functioning (EPF) theory does. On the other, the LFA

group's performance appeared to fit better with the theory of Weak Central Coherence (WCC), which stipulates that localised processing results from a failure to attend to the global level. In line with the HFA results here, Chamberlain, McManus, Riley, Rankin and Brunswick (2013) demonstrated that the superior performance on drawing tasks by ASD individuals is due to Enhanced Perceptual Functioning rather than Weak Central Coherence. However their study, however, did not take severity of ASD symptoms into account, when severity is considered, a similar pattern of results across the two subgroups appears (see Ballantyne & Núñez, 2016). This evidence underscores the need to "look at the whole beyond its parts" in the theories about integration of information in ASD (see López, 2008).

The performance of the FXS groups was consistently below that of the ASD groups across construction tasks. This is consistent with Cornish et al., (1998; 1999) and Pegoraro et al. (2014) who found that visuo-spatial construction showed the poorest performance. The effect of the comorbidity in the AFXS group meant that performance was often close to floor level. It is interesting to note from the performances of the two construction studies that removing the element of drawing normalised the performance of the FXS group in terms of their MA. This is in line with more recent research such as by Scerif, et al. (2012), who found that development showed a plateau when CA was used as a predictor, but showed a meaningful improvement in line with TD matches when MA was used as a predictor. Therefore deficits in construction tasks could be due to motor ability rather than a planning impairment. This provides further

evidence that the two construction tasks may be placing different cognitive demands on the children.

A surprising feature of the AFXS performance (although not statistically measured) was that the children completed the tasks in a very different manner from the other groups. In the construction tasks, children appeared to demonstrate 'closing-in' (a tendency to close in on a model while copying it, see Figure 5). Saglino, D'Olimpio, Conson, Cuppuccio, Grossi and Trojano (2013) observed 'closing in' in typical healthy adults when the task complexity became too great. This is often a behaviour shown in very young children and adults with dementia. It could be that the task demands were too great for the AFXS group, even when it was made simpler by using magnets. This behaviour was demonstrated irrespective of age and is something that requires further investigation.

The current study has provided evidence towards a different processing style in hierarchical visuo-spatial construction tasks depending on the severity of symptoms in ASD and its comorbidity with a FXS diagnosis, as assessed by the CARS. The CARS however is not a diagnostic tool in itself and is teachers rather than clinicians who score the questionnaire. It would be worthwhile following the study up with subgroups divided on the basis of a full scale IQ measure and ADOS scores (Autism diagnostic observation schedule, Rutter, DiLavore, Risi, Gotham & Bishop, 2012). Another factor that should be taken into consideration is that although the use of cross – sectional developmental trajectories do provide an indication of the role of development in visuo-spatial

construction tasks, they cannot replace longitudinal trajectories. However, the use of the trajectory approach has shown that a seemingly poor performance does not indicate an absence of the skill but rather that they might be delayed at onset or develop at a delayed rate.

10.1 Conclusions

In conclusion, this study has provided evidence pointing towards a different processing style in visuo-spatial construction tasks depending on the severity of ASD symptoms on the CARS. It showed important inter - and intra- group differences, which are not evident when testing only ASD or FXS populations in general. It also illustrated the importance of taking note of the subgroups of these populations, especially when there is such a large amount of variability between individuals. The pattern of results of each sub group is distinct, with different strategies for task completion. This study along with others (e.g. Ballantyne & Núñez, 2016) provide evidence towards an intra and inter group approach when considering developmental disorders.

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Figure 1: Congruent and incongruent stimuli for drawing and magnet task

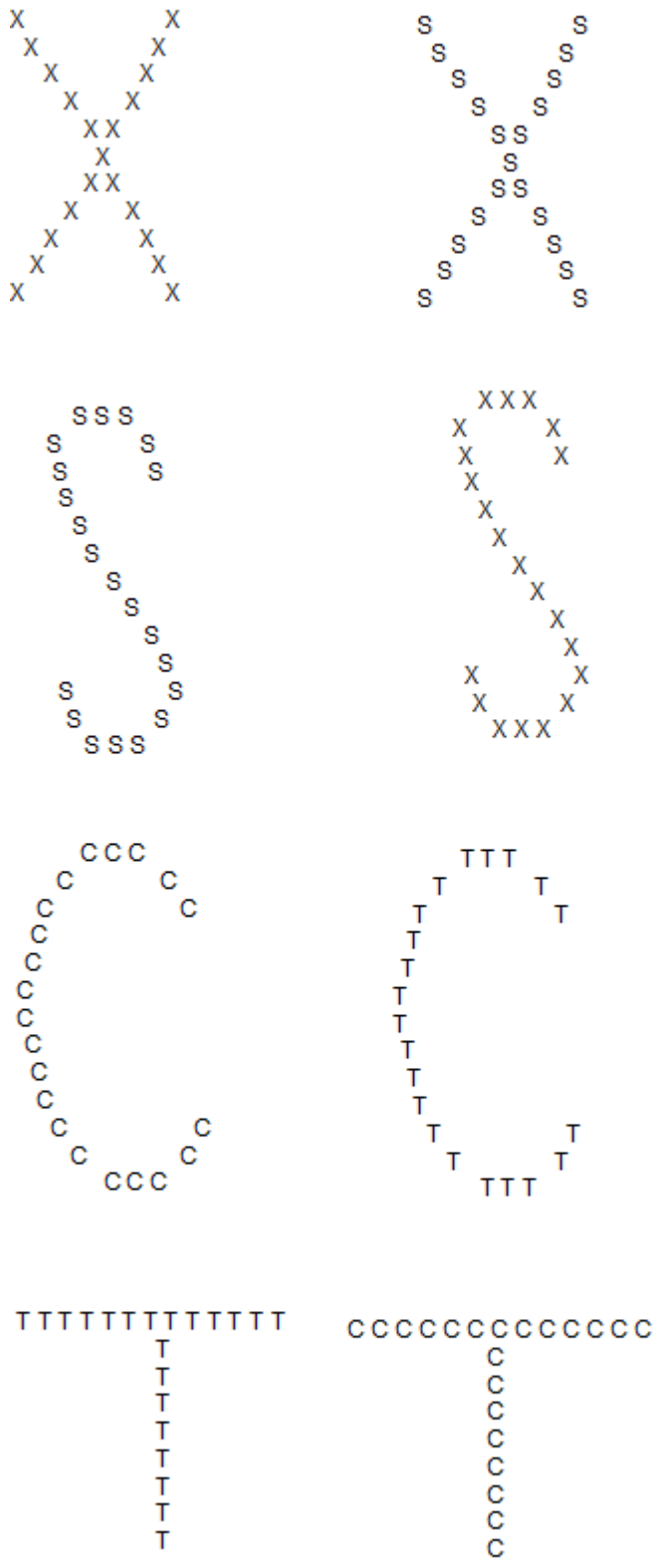


Figure Caption Sheet

Figure 2 a: TD group developmental trajectories of drawing accuracy

Figure 2 b: HFA group developmental trajectories of drawing accuracy

Figure 2 c: LFA group developmental trajectories of drawing accuracy

Figure 2 d: FXS group developmental trajectories of drawing accuracy

Figure 2 e: AFXS group developmental trajectories of drawing accuracy

Figure 2 a

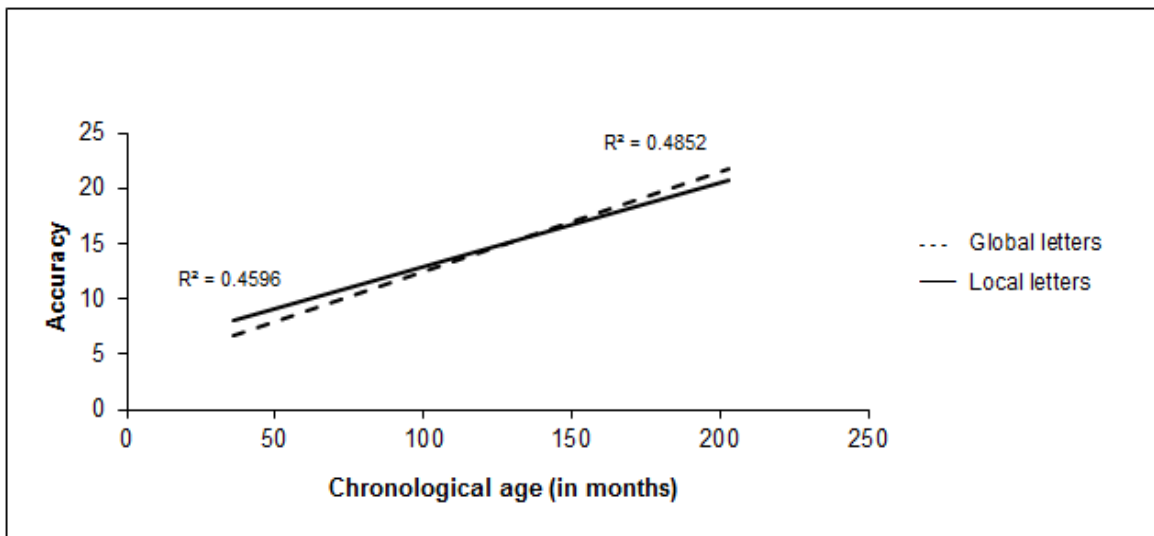


Figure 2 b

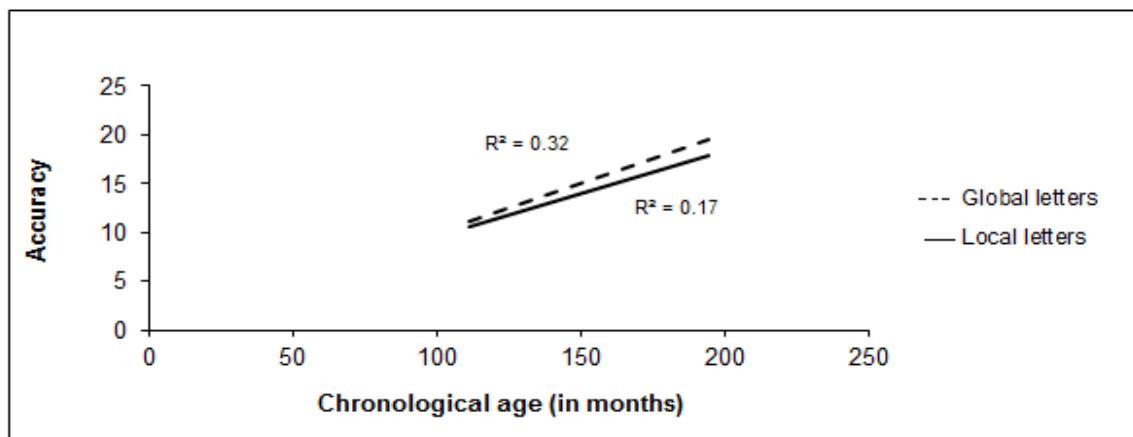


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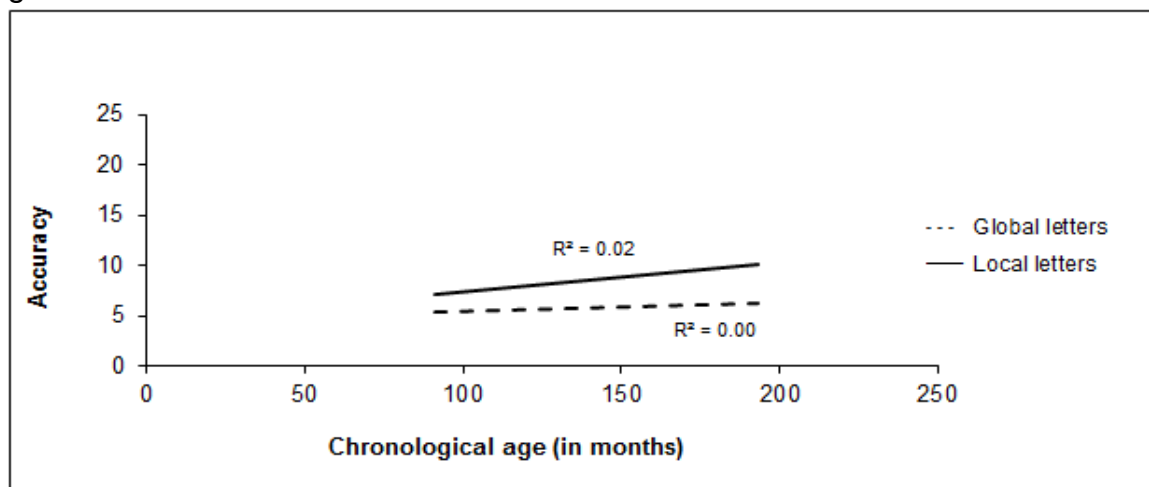


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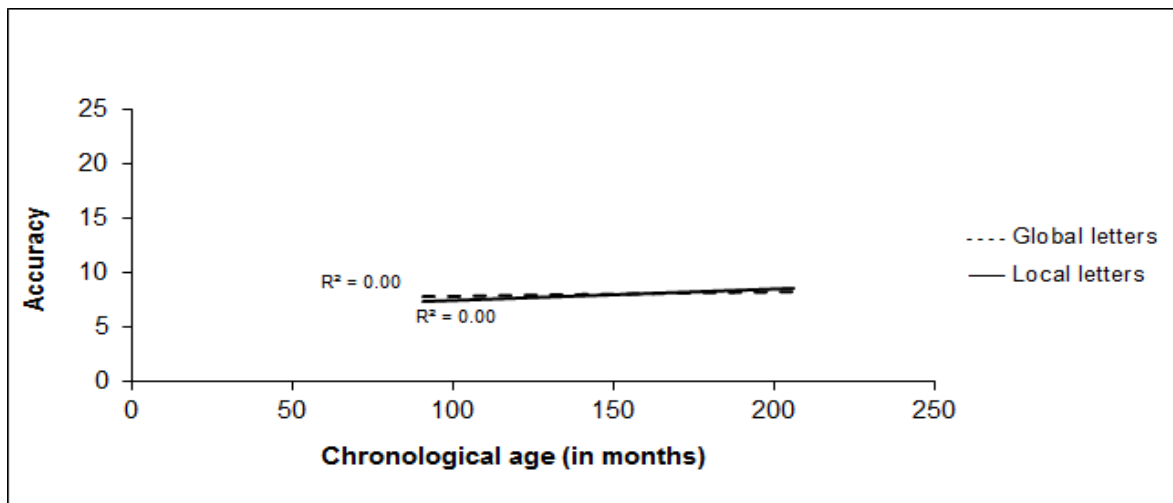


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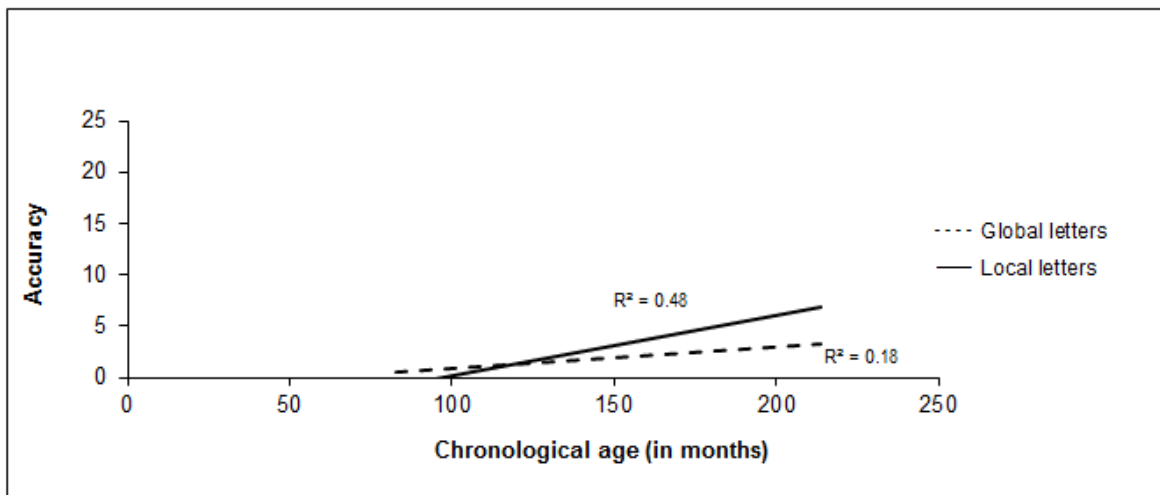
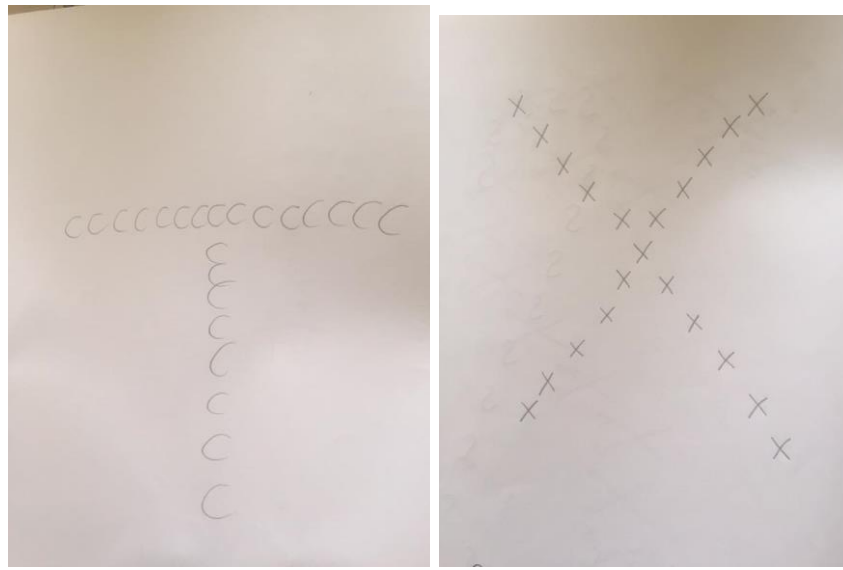


Fig. 3. a: TDGlobal T Local C drawing, b: HFA Global S Local X drawing, c: LFAGlobal C Local T drawing, d: FXSGlobal X Local X drawing, e: AFXS Global T



Local T drawing.

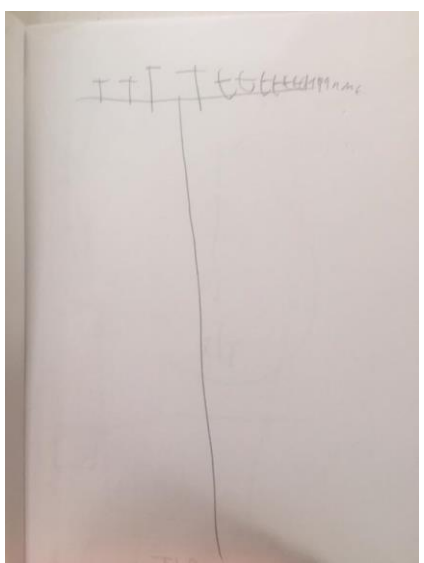
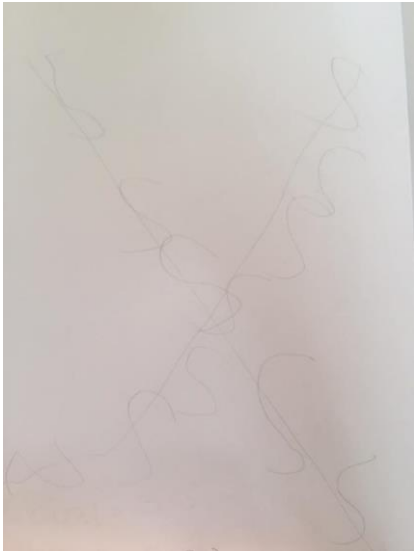
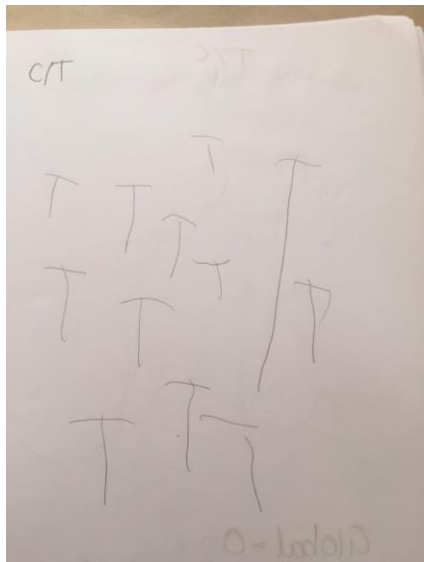


Figure Caption Sheet

Figure 4: Accuracy scores of all groups on the Navon magnet task.

Figure 4

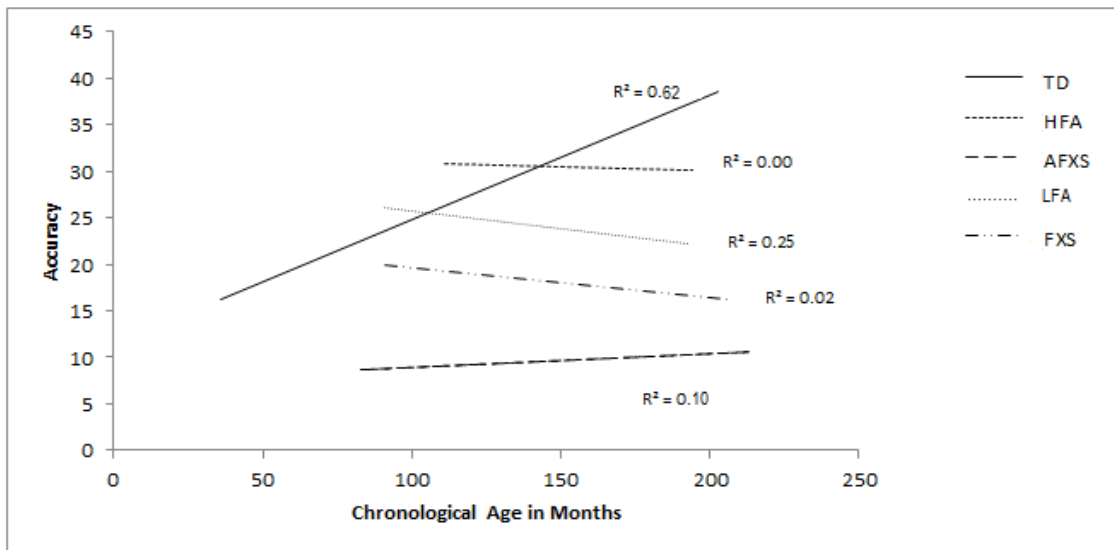


Fig. 5. a: HFA Global S local S magnet task, b: LFA Global S Local S magnet task, c: FXS Global X Local S magnet task, d: AFXS Global T Local C magnet task.

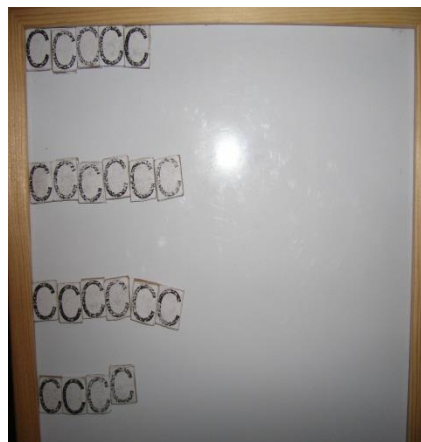
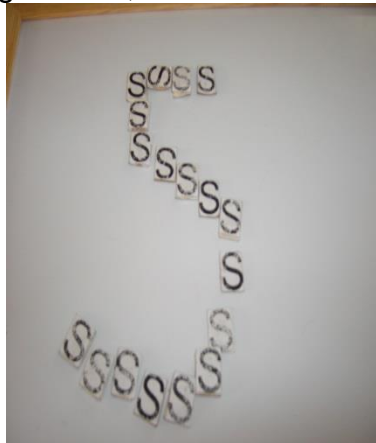


Table 1: Participant details for all groups

Group	N	Mean/SD (yrs & months)	Age Range	CARS score ranges	Mean RCPM test age (yrs & months)	Mean BPVS standard score (months)
TD	80	9:5 ± 4:0	3:0-16:11	NA	10:1± 4:5	121 ±49
AFXS	19	12:7 ± 2:9	6:11-17:10	30-35	7:11± 0.7	56 ±23
FXS	21	13:0 ± 2:8	7:7-17:2	18-27	7:11±.05	72 ±24
HFA	20	13:9 ± 1:5	9:3-16:2	30-35	9:9± .09	94±31
LFA	20	10:1 ± 2:4	7:7-16:1	38-50	7:11± 0.3	46 ±12

RCPM: Raven's Progressive Coloured Matrices

BPVS: British Picture Vocabulary Scale

Table 2: Summary of accuracy and standard deviation on drawing task

Group	Navon Accuracy			
	Global	SD	Local	SD
TD	14.10	6.26	14.26	5.36
HFA	16.15	3.39	15.0	4.12
LFA	5.6	6.31	8.1	6.03
FXS	8.10	6.38	8.1	5.27
AFXS	2.11	4.03	3.42	4.23

Table 3: Developmental relationships between CA and RCPM Navon Drawing Task across all 4 clinical groups

	Main effect of group	2-way*	3-way*
CA			
FXS	$p = .14$	$p = .02$	$p = .22$
AFXS	$p = .00$	$p = .25$	$p = .00$
HFA	$p = .28$	$p = .80$	$p = .01$
LFA	$p = .00$	$p = .26$	$p = .11$
RCPM			
FXS	$p = .00$	$p = .00$	$p = .24$
AFXS	$p = .18$	$p = .23$	$p = .12$
HFA	$p = .00$	$p = .53$	$p = .62$
LFA	$p = .41$	$p = .17$	$p = .02$

A significant 2 way interaction indicates that the clinical group was developing at a delayed rate

A significant 3 way interaction indicates that the clinical group were performing the task in a different developmental relationship to the TD group.

Table 4: Summary of accuracy and standard deviation (SD) on magnet task

Group	Mean	SD
TD	27.19	8.04
HFA	30.35	4.52
LFA	24.70	2.20
FXS	17.76	7.28
AFXS	9.74	1.66

Table 5: Developmental relationships between CA and RCPM test age on Navon Magnet Task across all 4 clinical groups

	Main effect of group	2-way*
CA		
FXS	$p = 0.16$	$p = 0.00$
AFXS	$p = 0.00$	$p = 0.00$
HFA	$p = 0.22$	$p = 0.02$
LFA	$p = 0.18$	$p = 0.00$
RCPM		
FXS	$p = 0.06$	$p = 0.2$
AFXS	$p = 0.00$	$p = 0.02$
HFA	$p = 0.26$	$p = 0.16$
LFA	$p = 0.31$	$P = 0.29$

A significant 2 way interaction indicates that the clinical group was developing at a delayed rate