Differences in visual orienting between persons with Down or fragile X syndrome

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Abstract

The voluntary and reflexive orienting abilities of persons with Down syndrome and fragile X syndrome, at average MA levels of approximately 4 and 7 years, were compared with an RT task. Reflexive orienting abilities appeared to develop in accordance with MA for the participants with Down syndrome but not for those with fragile X syndrome. However, both groups showed delayed voluntary orienting. The group differences in reflexive orienting at the low MA level reinforce the practice of separating etiologies and highlight the contribution of rudimentary attentional processes in the study of individuals with mental retardation.

Keywords: Visual orienting; Attention; Down syndrome; Fragile X syndrome; Mental retardation

1. Introduction

The many differences in the behavioral and neurological profiles of genetically based syndromes associated with mental retardation (Burack, Hodapp, & Zigler, 1988; Dykens, Hodapp, & Finucane, 2000; Hodapp & Burack, 2006) challenge two related myths about attentional deficits among persons with mental retardation. One myth is that attention deficits are defining features, or even a cause, of mental retardation. The second myth is that the same attentional deficits are common across all persons with mental retardation regardless of etiology (for a review, see Burack, Evans, Klaiman, & Iarocci, 2001). However, no evidence of a deficit that is either inherent to mental retardation or common across etiologies is consistently found in studies that meet rigorous methodological and developmental criteria, such as matching groups on developmental level (Iarocci & Burack, 1998). This finding does not imply that attention deficits might not be found within specific etiological groups, but rather that attentional functioning, like all other aspects of cognitive function, needs to be assessed independently for each etiological group (Burack et al., 2001). This issue entails a more fine-tuned, even if more labor-intensive, approach in which the choice of the specific components of attention to be studied is determined by the unique cognitive and behavioral patterns that are characteristic of a specific etiological group.

In one example, the orienting aspect of attention, or the ability to shift attention in relation to cues in the environment, was studied among persons with Down syndrome because of the observation that infants with Down syndrome display difficulties in searching and shifting attention in free play contexts (Krakow & Kopp, 1983; Landry & Chapiske, 1989). These preliminary observations led to the hypothesis that orienting deficits would
be evident later in development among persons with Down syndrome. However, this hypothesis was not supported in three studies in which versions of commonly used experimental tasks of orienting were administered to a group of persons with Down syndrome and a group of typically developing children carefully matched on mental age (Goldman, Flanagan, Shulman, Enns, & Burack, 2005; Randolph & Burack, 2000). The two groups showed similar patterns both in automatically orienting to target stimuli when flashes of light were presented as cues in the locations to the left or right of the center of the screen where the targets were eventually displayed (Randolph & Burack, 2000), and voluntarily utilizing a centrally located arrow (i.e., a symbolic cue) to predict the location of a target to the left or right of the screen (Goldman et al., 2005).

Although discrepant from the initial observations of infants, these findings are convincing because the developmental levels of the participants were between 5 and 6 years, the age range in which orienting abilities approach adult-like levels of efficiency and, therefore, when group differences are most likely to be found (for a review of issues related to the development of participant groups, see Burack, Iarocci, Bowler, & Mottron, 2002; Burack, Iarocci, Flanagan, & Bowler, 2004).

In keeping with the notion that the efficiency of attentional processing may vary across etiological groups, the evidently intact orienting abilities of the persons with Down syndrome may serve as a metric for gauging the efficiency of persons with mental retardation of other etiologies. This comparison strategy serves to diminish the effects of potentially confounding factors such as IQ differences and the disparity between chronological age and mental age that would need to be considered if differences in orienting are found between persons with intellectual disabilities and typically developing persons (Burack et al., 2001; Zigler, 1967, 1969). Thus, the comparison of the orienting abilities of persons with Down syndrome to those with fragile X syndrome may be useful because attentional problems are often cited for the latter syndrome (e.g., Cornish, Sudhalter, & Turk, 2004; Hagerman, 1999; Munir, Cornish, & Wilding, 2000).

Any orienting differences between etiologically homogeneous groupings might be considered within the context of the disparate profiles of cognitive and social skills displayed by the two groups to allow for more fine-tuned mapping of the relation between orienting and other aspects of functioning (see Burack et al., 2002; Cicchetti & Rogge-Hesse, 1982; Hodapp, Burack, & Zigler, 1990).

1.1. Visual orienting and the experimental task

Visual orienting entails shifting attention from one stimulus to another based on information in the environment (Posner, 1980). Overt attentional shifts are accomplished by eye movements that are directed toward a particular location in space, whereas covert shifts occur independent of eye fixation (Posner, 1980; Turatto et al., 2000). These events are typically measured with tasks that involve visual cues that are processed either deliberately or automatically (Logan & Compton, 1998). A central cue indicates where attention should be focused and serves to initiate a voluntary attention shift to a specific location (Parasuraman & Greenwood, 1998). In contrast, a peripheral cue, in the form of, for example, an abrupt flash of light, initiates an automatic shift of attention to the location and does not require higher-order intentional processes for interpretation (Klein, 1993).

Visual orienting is often studied with reaction time (RT) tasks similar to a task developed by Posner (1980) in which a cue is presented just prior to the appearance of a target stimulus. In this methodology, the validity of the cue can be manipulated such that the location of the target is correctly cued on most trials (valid trials) and incorrectly cued on other trials (invalid trials). This procedure typically results in enhanced performance for conditions with the valid cue (relative to baseline), but impaired performance for conditions with invalid cues (relative to baseline) because attention must be redirected from the incorrectly cued location to the target location in the latter condition. The primary index of attention in this task is the “orienting effect,” which is derived by subtracting the RT on valid trials from the RT on invalid trials (e.g., Akhtar & Enns, 1989; Randolph & Burack, 2000). Shorter RTs on valid trials than on invalid trials are evidence that processing was influenced by the location of the cue in relation to the target.

In order to examine reflexive and voluntary visual orienting and the relations between the two, among children and adolescents with Down syndrome than to MA-matched children and adolescents with fragile X syndrome, we developed a simple detection task based on the location of the target stimulus with two cue types (peripheral or central) that were valid or invalid. The cue validity was based on the direction of an arrow or the location of a flash cue that either correctly or incorrectly predicted target location. The relations between the two forms of orienting were measured during trials on which both cue types were presented. Based on evidence of intact orienting abilities among persons with Down syndrome, we expected the participants with fragile X syndrome to show a less effective pattern of orienting in relation to those with Down syndrome. Further, in order to examine the developmental changes in orienting within groups, we assessed each component in high and low mental age (MA) groups and expected more effective performance among the high MA group.

2. Method

2.1. Participants

The participants with fragile X syndrome (N = 20) were recruited across the United States, although the largest number resided in Wisconsin and surrounding states. The
participants with Down syndrome \((N = 25)\) were recruited from Wisconsin and surrounding Midwestern states.

The parents/guardians of all participants with Down syndrome reported the etiology as trisomy 21. Medical records confirming the karyotype were available for all but four of the participants with Down syndrome. Reports of DNA confirmation of the fragile X full mutation were available for all but two participants, who had cytogenetic confirmation. For those participants with DNA results, two are mosaic (full + premutation) and the rest are considered full mutation. For the fragile X sample, three families have more than one child in the sample.

The performance of the persons with Down syndrome and fragile X was matched on nonverbal scale IQ, as measured by subtests (described below) from the Stanford–Binet, 4th edition (Thorndike, Hagen, & Sattler, 1986). The participants were then divided into high \((M = 7\) years, 5 months; \(SD = 1.87\) years) and low mental age \((M = 4\) years, 3 months; \(SD = .64\) years) groups. The performance of the high functioning persons on the Stanford–Binet was significantly higher than the performance of the low functioning group \((F = 53.27, p < .0001)\), but no differences were noted within groups or across etiologies. The high and low MA groups did not differ with regard to CA \((M = 17.88\) years, \(SD = 3.28)\). Across the two etiological groups, most participants in the high functioning groups were female \((N = 16)\), whereas most participants in the low functioning groups were male \((N = 16)\).

2.2. Standardized cognitive measures

2.2.1. Stanford–Binet, 4th edition

Nonverbal IQ and MA were determined by administering three subtests from the Stanford–Binet, 4th edition (Thorndike et al., 1986): Bead Memory, Pattern Analysis, and Copying. These subtests require few verbal instructions to administer, and the participant responds nonverbally. The Stanford–Binet is a standardized measure of intelligence that is used to test cognitive abilities of children from the age of 2 years through adulthood.

2.2.2. Stimuli and apparatus

The presentation of stimuli, feedback, and data collection were controlled by a Macintosh Color Classic Computer, and the space bar on the keyboard was used to collect responses. The visual stimuli were geometrical shapes. The target location markers were four outlined square boxes (1.2° per side, the lines were black and 2 pixels in width, on a white screen) positioned at the corners of an imaginary square at 2.0° from the center of the screen.

Two types of fixation cues were presented at the center of the four target location markers. In the peripheral cueing condition, the fixation was a small square (black, 0.03°) that served as a neutral warning cue for the commencement of the trial. In the central and the central combined with peripheral cueing conditions, the fixation was an arrow (black, 0.06° in length) that pointed to one of the four boxes as a predictive orienting cue. One of these fixation cues, and the target location markers, were always present on screen to ensure that the participant attended to the target throughout the trial.

Two types of location cues were presented either alone or together, depending on the condition. In the peripheral cueing condition, the cue was a 30 ms blackening (an additional 2 pixels of black line) of one of the four target location markers. The blackening gave the appearance of a 30 ms abrupt onset flash. In the central cueing condition, the cue was the same as the fixation (i.e., a black arrow that was 0.06° in length). In the conditions with the central arrow cue only and with both the central arrow and peripheral cues, the arrow was followed by the 30 ms blackening of one of the target location markers.

The target was a round disk (black, 0.5° in diameter) that appeared in the center of one of the four target location markers. Following the participant’s response, one of three possible feedback stimuli appeared. If the participant responded correctly, a plus sign (black, 0.1°) appeared. If the participant responded incorrectly (i.e., when no target was present), a minus sign (black, 0.1°) appeared. If the participant failed to respond when a target was present, an empty circle (outline black, 0.5° in diameter) appeared.

2.2.3. Task

One hundred and ninety-two experimental trials, divided into two blocks of 96 trials, were administered to each participant. Trials on which either no cue or no target appeared occurred 5% of the time. Performance on the trials with no cues served as a baseline measure. The trials with no target were used to ensure that participants pressed the spacebar only when the target was presented, thereby indicating that they understood the task and were following instructions. The experimental session lasted 15 min with a break between the two trial blocks.

2.2.3.1. Peripheral cueing condition. These trials were designed to measure reflexive orienting. Each trial began with the four target location markers and the square fixation cue at the center of the screen. The peripheral cue was presented following a 40 ms period during which the screen was blank. The target stimulus was presented randomly in one of four possible target locations. The target remained on the screen until the participant responded by pressing on the spacebar, or until 3000 ms had elapsed. Feedback appeared for 600 ms following the participant’s response. Forty-six of the 192 trials measured reflexive orienting.

2.2.3.2. Central cueing condition. These trials were designed to measure voluntary orienting. Each trial began with the four target location markers and the arrow fixation that appeared in the center of the screen and pointed toward one of the possible target locations. The arrow fixation was the same as the arrow cue and remained on the screen.
until the participant responded. A fixed 850 ms interval followed the feedback phase prior to the onset of the next trial.

2.2.3.3. Central combined with peripheral cueing condition. These trials were designed to measure reflexive versus voluntary orienting. Trials that included the presentation of both central and peripheral cues began with the four target location markers and the arrow fixation that appeared at the center of the screen and pointed toward one of the possible target locations. The arrow fixation was the same as the arrow cue and remained on the screen until the participant responded. The arrow cue was presented following a 40 ms period during which the screen was blank. This procedure was then followed by the presentation of the peripheral cue. The target was presented with 77% probability in the location indicated by the arrow cue. Ninety-six of the 192 trials measured reflexive versus voluntary orienting.

2.2.3.4. Cue validity. Peripheral and central cues provided either correct or incorrect information regarding the location of the target. Among the arrow-cue trials, the arrow correctly indicated the location of the target 77% of the time. Peripheral cues were valid only 25% of the time, thereby making them non-predictive. All possible relations between reflexive and voluntary orienting were measured. The trials examining the relations between the two included valid central cues followed by invalid peripheral cues, invalid central cues followed by valid peripheral cues, and invalid central cues followed by invalid peripheral cues.

2.3. Procedure

Each participant was tested in a quiet room at a university research center. Each participant completed a larger protocol that included several standardized and experimental tests of various aspects of language, cognition, and social cognition, including those described in this article (see Abbeduto & Murphy, 2004, for a description of the remainder of the protocol). The tests were administered to each participant in two sessions. Each session lasted between 1 and 2 h. Depending on participant availability, the two sessions were conducted on different days or on the same day with an hour or more of a break between them. The subtests of the Stanford–Binet, 4th edition were administered at the beginning of the first session and the visual orienting task was presented last in the second session. During the orienting tasks, the participants were seated so that their eyes were approximately 100 cm from the screen. They were instructed to keep one hand on the spacebar.

3. Results

3.1. Error data

Two types of error data were analyzed: (a) false alarms (i.e., spacebar presses when the target did not appear) and (b) misses (i.e., failures to press the spacebar when the target was present). False alarms were noted in 20% of target absent trials for both groups. No group differences were noted in participant’s responses to false alarms within the high and low functioning groups or across etiologies; however, the performance of low functioning participants with fragile X was slightly more variable \( (M = .700, SD = .365) \) than that of all other groups \( (M \text{ of the other three groups} = .840, SD = .179) \). Misses were noted for approximately 1% of trials in both MA groups, and there were no differences in the number of such errors across either etiological or MA groups.

3.2. Baseline RT

The baseline RT’s of all participants were assessed by examining participants’ performance on trials in which targets were presented without any cues. The RT’s of both high and low MA groups was similar for such trials, although the performance of lower MA participants tended to be somewhat slower than the RT of the higher functioning groups. Within each MA group, the performance of participants with fragile X and those with Down syndrome did not differ from one another (see Fig. 1).

3.3. RT to peripheral cues

In order to compare the reflexive orienting abilities of high and low functioning participants with fragile X and those with Down syndrome, a repeated-measures ANOVA was conducted for trials with RT to the flash cue as the dependent variable and Group (DS vs. fragile X), MA level (high vs. low), and type of flash (valid or invalid) as the independent variables. These trials included only those in which the flash cue was present or absent and did not include trials combining central and peripheral cues. Significant differences were noted for Flash type \( (F(1,36) = 14.06, p < .006) \), with RT to valid flashes \( (M = 639.425 \text{ (237.02) ms}) \) being faster than RT’s to invalid flashes \( (M = 848.75 \text{ (286.4) ms}) \). A significant three-way (Group × MA level × Flash Type) interaction also emerged \( (F(1,36) = 6.145, p = .018) \), reflecting the RT differences between valid
and invalid trials for all but the low MA participants with fragile X. These findings are indicative of similar reflexive orienting across both MA and etiological groups, except for the low MA participants with fragile X, who, as a group, do not appear to utilize the reflexive cues to orient their attention (see Fig. 2). Simple main effects of group and MA did not attain significance.

3.3.1. RT to central cues

In order to compare the voluntary orienting abilities of high and low MA participants with fragile X and those with Down syndrome, a repeated-measures ANOVA was conducted with RT to the arrow cue as the dependent variable and Group (DS vs. fragile X), MA level (high vs. low), and type of arrow (valid or invalid) as the independent variables. These trials included only those in which the central cue was present or absent and did not include trials combining central and peripheral cues. No significant effects were noted; however, the performance of participants in the high MA groups suggests that they voluntarily oriented their attention (RT to valid arrows was faster than to invalid arrows), whereas the participants in the low functioning group either did not show this effect or showed the reverse effect (RT to invalid arrows faster than to valid arrows) (see Fig. 3).

3.4. Central vs. peripheral cues

Measuring the effect of both cues together provides an index of which type of cue was more salient for detecting a target among these groups of participants. In order to compare the salience of cue type (arrow vs. flash), a repeated-measures ANOVA was conducted with RT to target detection as the dependent variable, and Group (fragile X vs. Down syndrome), MA level (high or low), flash type (valid vs. invalid), and arrow type (valid vs. invalid) as the independent variables. A significant main effect was noted for flash type ($F(1,36) = 9.03, p = .0048$), with RTs to valid flashes being faster than RTs to invalid flashes. A significant arrow by flash type interaction ($F(1,36) = 4.24, p = .0468$) and a significant group by flash type by arrow type interaction ($F(1,36) = 4.165, p = .0487$) also emerged. Analysis of these interactions indicated that for the participants with Down syndrome, RTs were fastest in conditions where both the arrow and flash cues were valid, whereas no differences in RTs were noted in this group when both cues were invalid or when one was valid (either flash or arrow). For the participants with fragile X, RTs were fastest for trials in which the flash cue was valid, regardless of the type of arrow cue (see Fig. 4).

3.5. Exploratory analyses

A 3 way (Group × Flash Type × MA Split) repeated-measures ANOVA was performed to assess the effect of each cue separately, comparing trials in which no cue was presented to trials in which either valid or invalid peripheral flashes were presented. A significant main effect was noted for flash type ($F(2,72) = 9.478, p < .0002$), which indicated
that participants were faster for trials in which a valid flash cue was presented than for trials in which cues were either not present or invalid. A significant flash type by Group by MA level interaction ($F(2, 72) = 5.084, p = .0086$) was also noted. This pattern was related to an increased RT for the low MA participants with Down syndrome on trials on which flashes were invalid.

In order to assess the effect of each cue separately, a 3-way (Group × Arrow Type × MA Split) ANOVA was performed comparing no cue trials to valid and invalid arrow trials. No differences were noted in RT, which suggests that the arrow was not used as a cue by either group at either MA level. For both the high functioning groups, however, there was a non-significant trend for the RT’s to be faster for the valid arrow than for the invalid or no arrow conditions.

4. Discussion

The voluntary and reflexive orienting abilities of persons with Down syndrome and fragile X at two MA levels were assessed within an orienting paradigm. Reflexive orienting was elicited through the presentation of an abrupt onset peripheral flash cues, whereas voluntary orienting was measured by shifts in attention initiated in response to predictive central arrow cues. Relative uses of reflexive and voluntary shifts of attention were gauged with a task condition in which both arrow and flash cues were presented in a single trial. Group differences, both with regard to diagnosis and MA were noted in the participants’ use of the reflexive and voluntary cues, as well as in how these cues interacted when they were in competition with one another.

Reflexive shifting was indexed by a faster RT to detecting targets cued by valid than invalid flashes. Among typically developing children, this process is well-established by the age of four to five years, (Akhtar & Enns, 1989; Lane & Pearson, 1983; Pearson & Lane, 1990), although improvements in reflexive shifting of attention are noted throughout middle childhood (Akhtar & Enns, 1989; Enns & Brodeur, 1989; Pearson & Lane, 1990). The average MA of the participants in the current study was greater than four years, and thus, all participants should have shown evidence of reflexive orienting. The high MA groups of participants reflexively oriented their attention in response to peripheral cues alone, displaying faster RTs when the flash location coincided with the target location (valid flash) than when the flash location did not coincide with target location (invalid flash). Among the lower MA group, the participants with Down syndrome demonstrated reflexive orienting to targets. The low MA participants with fragile X showed no evidence of reflexive orienting as there were no differences between RTs to valid and invalid flashes. The lack of difference in RT to valid and invalid flashes for the low MA participants with fragile X suggests either that their attention was not drawn to the flash or that the flash was not being used to anticipate the location of the target.

The finding of intact reflexive orienting in the low MA participants with Down syndrome, but not in those with fragile X syndrome, suggests that there is an exceptional delay in the development of reflexive orienting in the latter group. No reflexive orienting in low MA participants with fragile X syndrome, coupled with the presence of reflexive orienting in higher functioning participants with fragile X syndrome, indicates that this delay appears to be a transient developmental lag in reflexive orienting in this group. In contrast, participants with Down syndrome were on par with their developmental level in their ability to reflexively orient their attention.

Both high and low MA participants with Down syndrome and fragile X showed significantly faster cue detection to valid flashes than to invalid flashes, but this effect held only for the involuntary cue. Neither the high nor the low MA participants used the voluntary cue to orient their attention, and the lower functioning group demonstrated cue facilitation for invalid targets. Among typically developing children, improvements are noted in the development of voluntary attention after the age of 6 years (Brodeur & Enns, 1997). It is thus probable that the lack of use of voluntary attentional cues in the low functioning groups is related to their developmental level; however, the lack of a voluntary orienting effect in the higher functioning group of participants is indicative of an exceptional delay for both participants with Down syndrome and those with fragile X syndrome.

In the conditions in which voluntary and involuntary cues were presented simultaneously, group differences were noted in the way in which the cues interacted, suggesting that cue preferences differ across these groups. When both voluntary and reflexive cues were presented simultaneously, facilitation effects were noted only for the participants with Down syndrome. They had faster RT’s for trials in which both the voluntary and reflexive cues validly identified the target. In contrast, the participants with fragile X showed no RT differences in their detection of the target regardless of the combination of cues. Thus, youth with fragile X syndrome seem to ignore all of the cues, and just wait for the onset of the target.

In summary, the reflexive orienting abilities of persons with Down syndrome appear to develop in accordance with mental age functioning, whereas the development of reflexive orienting among persons with fragile X syndrome appears delayed in relation to developmental level. In contrast, both groups were delayed in their voluntary orienting abilities, showing no evidence of orienting their attention in response to a voluntary cue. Further research should examine whether the finding of no voluntary orienting is present throughout the lifespan of these two groups, or whether these abilities are delayed but eventually do develop.

Delays in the voluntary control of attention will affect many aspects of social interaction and learning. Findings of differences in the developmental trajectories of the development of reflexive and voluntary orienting across etiological groups whose phenotypes are both associated with
mental retardation, supports the notion of syndrome-specific profiles of attentional functioning. The findings of the current study highlight the importance of separating etiologies in the study of individuals with mental retardation and point to the presence of exceptional delays in the development of voluntary orienting among persons with fragile X syndrome.

References


