



Contents lists available at [ScienceDirect](#)

Infant Behavior and Development



Early information processing among infants with and without spina bifida

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ARTICLE INFO

Article history:

Received 5 August 2008

Received in revised form 5 November 2009

Accepted 19 March 2010

Keywords:

Spina bifida

Infants

Disability

Habituation

Attention orienting

Attention processing

ABSTRACT

This study focuses on the development of early visual information processing among infants with spina bifida (SB) compared to typically developing infants using the habituation–dishabituation paradigm. Analyses were conducted in two stages. First infants were evaluated to determine if 18-month old infants (SB = 47; Control = 40) differed in their ability to shift attention and habituate to two female faces, as well as their responses to composite and novel stimuli. Second, relations between these variables and infant motor and mental functioning were evaluated. The results of the study indicated that difficulties with visual attention skills can be detected as early as 18 months of age among infants with SB. Infants with SB differed significantly from controls on attention getting. Although there were no differences found on habituation and composite tasks, infants with SB differed significantly from controls on their ability to dishabituate. Implications are discussed.

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Spina bifida meningocele (SBM) is a disabling congenital birth defect affecting 0.3–0.5 per 1000 births in the United States (Williams, Rasmussen, Flores, Kirby, & Edmunds, 2005). It is characterized by an open spinal defect that leads to variable difficulties with ambulation and bladder/bowel control depending on the severity and location of the lesion. SBM is accompanied by congenital brain malformations involving posterior cortex, corpus callosum, cerebellum and midbrain (Del Bigio, 1993; Dennis, Landry, Barnes, & Fletcher, 2006; Fletcher et al., 2004). The Chiari type II malformation, present in almost all children with meningocele above the sacral level (Griebel, Oakes, Worley, & Rekate, 1991), is characterized by the brainstem and part of the cerebellum being displaced downward, which can obstruct the flow of cerebral spinal fluid in utero, and cause hydrocephalus requiring shunt diversion after birth (Fletcher et al., 2000).

Children with spina bifida tend to score in the average to low average range on measures of intellectual ability; however, these children have heightened vulnerability for learning disabilities and other cognitive difficulties (Brewer, Fletcher, Hiscock, & Davidson, 2001; Fletcher et al., 1996; Wills, 1993). Dennis et al. (2006) propose a model of neurocognitive functioning for SBM to explain why, as a group, children and adults with SBM have modal core deficits and assets in particular aspects of cognitive functioning. Because SBM involves naturally occurring systematic variability in the genotype, the neural phenotype, as well as the environment of the individual there is considerable variability in motor and cognitive outcomes

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(Batshaw, 2002; Dennis et al., 2006; Lomax-Bream, Barnes, Copeland, Taylor, & Landry, 2007; Taylor, Landry, English, & Barnes, 2010).

One area of particular concern for children with SBM is difficulty in the ability to regulate attention. Research in this area has identified a pattern of visual attention problems with variable strengths and weaknesses related to specific areas of the brain impacted by SBM. For example, school-aged children with spina bifida and hydrocephalus have been found to have greater difficulty with focused attention than sustained attention. Using neurocognitive models of attention, Dennis et al. (2005a) have shown that older children and adolescents with SBM have difficulty with orienting to and disengaging from salient sensory stimuli and these difficulties are related to tectal beaking and volume loss in right posterior regions of the brain. In comparison, they have less difficulty with these attention functions for interesting cognitive stimuli, attention functions subserved by a more anterior neural attention system thought to be under more top-down control. Dennis et al. (2006) proposed that attention orienting is a core deficit in SBM because it arises from a specific set of brain abnormalities (in midbrain and posterior cortex), is demonstrable over a range of cognitive ability levels, is only weakly related to other core deficits in timing and movement, and emerges in infancy before formal education. The study reported here tests the hypothesis that the deficit in attention orienting to and disengagement from perceptual salient stimuli is apparent early in development.

McDonough, Vietze, and Vaughan Jr (1988) suggested that it is important when studying infants with motor difficulties, like SB, who have a varied intellectual range, to use assessments of cognitive functioning that are less dependent on motor functioning. One such assessment for measuring attention is the habituation–dishabituation paradigm. Cohen (1972) identified at least two separate attention processes in visual attention (habituation/dishabituation) tasks, attention getting and attention holding. *Attention getting* is defined as the time it takes the infant to orient to a stimulus, and *attention holding* is defined as the amount of time the infant fixates on the stimulus after turning. He argues that the use of a total fixation time measure confounds these two processes. Therefore, he developed a procedure that subdivides an infant's attention to a visual stimulus into these two separate processes. For the purposes of the current study, attention getting in the habituation/dishabituation paradigm provides an early measure of attention orienting and disengagement.

Habituation/dishabituation paradigms not only measure aspects of attention, but also infant visual information processing more generally. According to Cohen, Friedman, and Sigman (1981), the decrease in looking time during habituation is assumed to result from the infant's growing awareness of, or boredom with, the stimuli. Faster habituation is generally interpreted as more efficient information processing associated with learning about a new stimulus. The dishabituation phase indicates another aspect of information processing, namely discriminative ability (Cohen et al., 1981). Relatively greater amounts of looking to a novel stimulus following a familiar stimulus are generally interpreted as more efficient information processing because infants interpret the stimulus as new compared to the familiar stimuli.

Another component of Cohen's paradigm has been used to evaluate featural versus configural or holistic processing. The procedure typically consists of an infant habituating to two different face stimuli. Then the infant is tested with a composite face, constructed from some of the features of both stimuli combined. The infant is then evaluated to see if he or she treats the composite as a familiar or novel stimulus. Infants with longer fixations to the composite face are thought to be responding to the composite face as a new stimulus, and responding holistically; that is, they are responding to the new relationship among previously seen features rather than responding to the individual features.

The vast majority of infant habituation research has been concerned with the functioning of typically developing infants. However, habituation/dishabituation tasks have also been effective in evaluating various aspects of perception and cognition in preterm infants (Caron, Caron, Glass, Field, & Sostek, 1983; Landry, Leslie, Fletcher, & Francis, 1985), as well as some mixed samples of infants with SBM, Cerebral Palsy (CP), and Downs Syndrome (DS) (McDonough & Cohen, 1982; McDonough et al., 1988).

It is likely that the problems observed with certain forms of visual attention in school-aged children with SBM can be identified in infancy, reflecting the congenital nature of the neural insults with consequences for attention very early in life. The purpose of this study was to investigate early visual attention and information processing skills by comparing infants with SBM and typically developing infants on a visual attention task. Variables of interest include attention getting and attention holding (habituation, dishabituation). Based on the research on visual attention in school-aged children with spina bifida discussed above, we hypothesized that infants with SBM would have more difficulty orienting and disengaging their focus of attention from a salient sensory stimulus (attention getting process) than typically developing children. However, the two groups were not expected to differ on measures of attention holding and rate of habituation, or response to novelty (dishabituation).

1. Methods

1.1. Participants

Children with SBM in the larger study from which this subsample was drawn were referred to the study at birth by treating neurosurgeons and pediatricians in Houston (Memorial Herman Children's Hospital and Texas Children's Hospital), Toronto (Hospital for Sick Children), Hamilton (McMaster Children's Hospital), and London (Thames Valley Children's Center). The sociodemographics of the Texas and Ontario sites were different, with the Houston site including many more children of Hispanic origin, in contrast to the predominantly Caucasian population in the Ontario sample. Children were generally

admitted to the study at 6 months of age. However, a decline in births of children with SBM during the active years of recruitment necessitated that infants with this disorder were permitted to enter the study as late as 18 months of age in a limited number of cases.

Exclusionary criteria included uncontrollable seizure disorders, other known congenital anomalies outside of SBM, and significant sensory impairments (blindness, deafness). In addition, all infants in the study had to have gestational ages from 39 to 41 weeks, birth weight appropriate for gestational age, normal history of pregnancy and birth, Apgar score at 5 min of eight or greater, normal physical examination, and hospital discharge within 5 days of birth. All the typically developing children began the study at 6 months of age. Typically developing children were recruited from well baby clinics, advertisements in newspapers, and local pediatricians. Exclusionary criteria for this group included those listed above, as well a lack of gross sensory or motor abnormalities.

These recruitment efforts resulted in 165 children, including 91 with SBM and 74 neurologically normal, typically developing children. For the purposes of this study, children's performance at 18 months of age was evaluated. This time point was selected due to the fact that recruitment efforts were complete at 18 months and the habituation/dishabituation task was also given at that time allowing for a larger sample of children. A total number of 154 infants were evaluated at 18 months. However, unexpected technical difficulties with the habituation/dishabituation task apparatus made the task unusable for a period of time and 58 infants were unable to be tested. In addition, some infants were lost due to being fussy or crying during the task making their data unusable ($n=9$). This resulted in a subsample of 87 infants (47 SBM/40C) who completed the habituation/dishabituation task, in addition to the measures completed by the full sample.

In terms of medical variables commonly used to characterize SBM, 42 infants were shunted for hydrocephalus (14 with no shunt revisions, 16 requiring one shunt revision, and 12 requiring 2 or more shunt revisions). There were 7 infants with spinal lesions at or above T12 and 40 with spinal lesions at or below L1, the differentiation at the thoracic level based on genetic and neuropsychological research (Fletcher et al., 2005).

Table 1 provides descriptive data on participant sex, ethnicity, and socioeconomic (SES) status, assessed with the Hollingshead (Hollingshead, 1975) 4-factor scale. The demographic profile of this sample consistent with the epidemiology of SBM, including a greater representation of children who were Hispanic in the SBM group (Northrup & Volick, 2000). There was no significant group differences for sex, $\chi^2(1, N=87)=1.256, p=.262$. However, there were site differences by ethnicity, $\chi^2(6, N=87)=23.22, p<.05$, and SES, $F(1, 87)=10.18, p<.05$, reflecting the separate matching of the SBM and comparison groups in the two sites. The Hispanic subgroup was lower in SES and therefore groups differed on this dimension, $t(75)=3.81, p<.01$. Due to the fact that the Houston site had all the infants of Hispanic origin who were also lower in SES, site was included in the analyses as a covariate.

1.2. Procedures and measures

The 18-month assessment was one of five laboratory visits over the first 3 years of life (6, 12, 18, 26, and 36 months). The habituation/dishabituation task was the first of several measures obtained at each visit, including the Bayley Scales of Infant Development—II (BSID-II). The Bayley results have been previously reported for the larger sample (Lomax-Bream et al., 2007). Table 1 presents the 18-month Psychomotor Developmental Index (PDI) and the Mental Developmental Index (MDI) for the sample who received the habituation task. The BSID-II scores were used as a correlate of the outcome measures to determine the relation of motor and mental functioning to the outcomes.

Table 1
Means and standard deviations by group.

Variable	Control	Spina Bifida
<i>N</i>	40	47
Ethnicity, <i>n</i> (%)		
Caucasian	21 (52)	19 (40)
Hispanic	12 (30)	20 (43)
Other	7 (18)	8 (17)
Gender		
Female, <i>n</i> (%)	45 (52%)	42 (48%)
Socioeconomic status	42.3(12.9)	30.1(14.7)
Bayley scores		
PDI	99.5(10.27)	62.9(16.51)
Motor age equiv.	17.9(2.18)	11.2(4.95)
MDI	101(11.82)	79.8(18.58)
Mental age equiv.	18.3(2.23)	14.5(4.88)

Note: SES reports mean (SD) based on Hollingshead Scale (1975). PDI, Psychomotor Development Index. MDI, Mental Development Index.

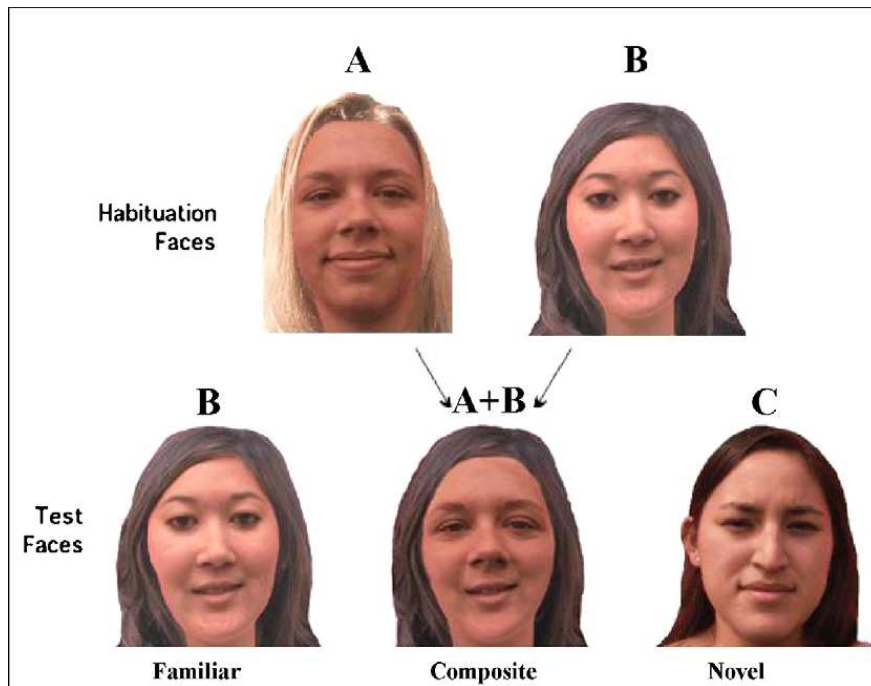


Fig. 1. Types of faces presented during habituation/dishabituation task.

1.3. Habituation/dishabituation paradigm

The habituation/dishabituation paradigm involves presenting the child with a series of photographed, human female faces on a television screen in a room that is otherwise dark (see Fig. 1). Each infant was seated on the mother’s lap facing a panel covering an observation window. All infants were seated on their mother’s lap, leaning back against mother facing forward, with mother offering trunk support to their infant. In this way, infants’ trunk support and positioning was the same for all infants. A 25 cm × 25 cm television monitor placed in a large black wooden box was positioned at a 20° visual angle to the left of the infant’s position (66 cm from the infant’s eyes to center of screen). The stimuli were displayed on the TV screen. At a 20° visual angle to the right of the infant, a 28-W blinking light was used to attract the infant’s attention at the beginning of each trial (along with a beep).

1.4. Stimuli

The rationale for the stimuli used in this study was taken from Cohen and Cashon’s previous work (Cashon & Cohen, 2004; Cohen & Cashon, 2001). Fixation time was recorded by a computer from the time the infant first looked at the stimulus until the infant looked away. An initial practice trial was administered to orient the child. Then a maximum of 20 habituation trials were given to the infant. On each habituation trial one of two female faces were presented. Once a child habituated to the two female faces the habituation trials ended. The computer calculated habituation to have occurred if the child’s fixation time over four consecutive trials decreased to ≤50% of the original fixation time over the first four trials the stimuli were presented. Next, the test trials were presented in the following sequence: (1) a familiar face (one of the two shown during habituation), (2) a composite face (a mixture of the two familiar faces constructed from the internal features of one face and the external features of the other face), and (3) a novel face (a female face not presented earlier). Table 2 presents the trial order and Fig. 1 provides examples of the types of habituation and test faces shown to the infants.

Table 2
 Visual attention procedure trial order.

Trial #	Name	Stimulus
1	Practice	Woman’s face (A)
2–21	Habituation trials	2 women’s faces (A and B) appear until child habituates to both or maximum trial was reached
22	Test trial 1	Familiar face (A)
23	Test trial 2	Composite face (A and B combined)
24	Test trial 3	Novel face (C)

1.5. Procedure

The mothers were asked to sit in front of the television screen with their infant on their lap facing the screen. The nature of the task was briefly explained and each mother was asked to be very still and to refrain from talking or moving her infant during the task. The examiner was then positioned on the other side of an observation window where the computer controls were located. A video camera recorded the child during the task.

Each trial began with the light blinking on and off. Once the infant fixated on the light, the examiner pressed a key on the computer to display the face stimulus and simultaneously turn off the blinking light. When the infant looked at the stimulus, the examiner pressed a key to record this on the computer. Each stimulus remained on until the infant looked away for 1 s. At that point the examiner pressed a key to start the light blinking. When the child looked at the light, the examiner pressed the key on the computer to display a face stimulus again and simultaneously turn off the blinking light, and this cycle continued until the child habituated to the stimulus or 20 trials were reached. On occasion an infant would get distracted. In this case, if a distracted infant did not fixate on a presented stimulus within 20 s, the trial was considered *failed* and the computer repeated the same stimulus trial again. In contrast, the maximum looking time recorded by the computer was 40 s. If a child continued looking at the same stimulus for more than 40 s, the trial was ended and the cycle continued. After the habituation trials were complete, the three test trials (familiar, composite, novel) were presented.

1.6. Attention measures

1.6.1. Attention getting

The attention getting outcome measure was obtained from the video tapes of the task. Individual tapes were time stamped. Researchers then coded the habituation trials on the tapes by noting the time on the tape from when the infant looked at the blinking light to the time on the tape when the infant looked at the face stimulus (reliability coefficient $>.9$). Thus this measure captures time to disengage from a salient sensory stimulus to attend to a cognitively interesting stimulus (face). From this coding, attention getting times were able to be calculated for each trial by taking the time it took the infants to move their head and/or eyes from the light to the face stimulus. Because infants may be able to shift their attention more quickly after experience with the contingency between the blinking light and face appearing, we were also interested in comparing stability in shift times across the task.

1.6.2. Attention holding

Based on Cohen and Cason's previous work (Cohen & Cason, 2001), four attention holding outcome variables were calculated. *Habituation Ability* was the average of the fixations from the last four habituation trials, divided by the average of the fixations on the first four habituation trials. *Habituation Status* (whether or not a child habituated to the stimulus) was indicated by a Habituation Ability score of $<.5$. *Composite* looking time to assess holistic versus featural processing was calculated by taking the fixation time on the composite test face, divided by the composite fixation plus the infant's fixation time to the familiar test face. Finally, *Novel* looking time (dishabituation) was calculated by taking the infant's fixation time on the novel test face, divided by the novel fixation time plus the infant's fixation time to the familiar face during the test trial.

1.7. Data analysis

Data were analyzed in two stages. First, groups were evaluated to determine if they differed on each of the attention outcome variables. Depending on the distribution, generalized estimating equation (GEE) (Liang & Zeger, 1986) or General Linear Modeling (GLM) procedures were used. Separate follow-up analyses were then conducted to evaluate the relation of motor and mental functioning on each of the outcome variables, using infants' BSID-II scores at 18 months of age, with site included as a covariate in these analyses. Shunt status was considered as covariate for the analyses. However, because no significant differences were found on the outcomes related to shunt status (0, 1, 2 or more shunt revisions) it was dropped from further analyses. The group with spinal lesions at T12 and above was too small ($n = 7$) to include this variable in analyses. All analyses were performed with SAS software (SAS, 2006).

Infants varied in the number of trials taken to habituate. Therefore, the first and last three trials were selected for the attention getting analyses. All infants were included regardless of the number of trials taken (range = 6–20 trials). A generalized estimating equation (GEE) (Liang & Zeger, 1986) approach was used to evaluate if group would predict attention getting, and would predict the trial patterns between the first three and last three habituation trials. The distribution was a negative binomial with a log link function due to data being positively skewed. The mean attention getting score of the first three trials and the mean of the last three trials were estimated for each child. This approach allowed an evaluation of possible differences in infant attention getting over the habituation trials while controlling for site differences.

2. Results

2.1. Attention getting

The GEE indicated a significant difference in shift time between the two groups, $\chi^2(1, N = 73) = 5.27, p = .021$. As expected, this was greater among children with SBM (mean 1.275 s) than typically developing children (mean = .687 s). All reported means for GEE analyses are back transformed from the original transformed variable and reported in original units for ease of interpretation. There was no significant difference in early and later shifting between groups, $\chi^2(1, N = 73) = 1.72, p = .189$. This indicates that although it took longer for children with SBM to disengage from the light and orient to the face than typically developing children, neither group's attention getting changed over time (SBM = 1.266 early to 1.284 later; control = .534 early to .883 later). Follow-up analyses revealed a significant relation of motor performance on attention getting, $\chi^2(1, N = 73) = 5.86, p = .015$, with higher motor scores being associated with shorter shift times. Moreover, differences by group and by site were not significant in the model after controlling for motor performance. The relation of mental functioning to attention getting was not significant, $\chi^2(1, N = 73) = 2.88, p = .089$.

2.2. Attention holding

2.2.1. Habituation

Group differences in habituation to a familiar stimulus at 18 months of age based on the Habituation Status variable (SBM = 50%, control = 67%) failed to reach statistical significance, $\chi^2(1, N = 87) = 2.89, p = .089$. In addition, the GLM analysis revealed that groups did not differ on their overall ability to habituate based on the Habituation Ability variable, $\chi^2(1, N = 87) = 1.51, p = .219$. These findings indicate that infants with SBM in this study performed as well as their typically developing peers in habituation, both in the overall ability to habituate to two face stimuli and to reach Habituation Status. Follow-up analyses were conducted to evaluate the relation of infants' 18-month motor and mental performance on the Habituation Ability variable. The models revealed no significant relation for either mental, $\chi^2(1, N = 86) = 2.14, p = .143$, or motor functioning, $\chi^2(1, N = 86) = .62, p = .431$, on Habituation Ability after controlling for etiology and site differences.

2.2.2. Composite

GLM analyses identified no significant group difference in response to the composite test stimulus, $F(1, 86) = .52; p = .474$. Furthermore, no significant relation was found for mental, $F(3, 85) = .72; p = .542$, or motor scores, $F(3, 85) = 1.27; p = .291$, and looking time to the composite face when etiology and site were included in the models.

To further understand the composite finding as it relates to featural versus holistic processing, a 2 (habituated versus did not habituate) \times 2 (SBM versus control) \times 2 (familiar test versus composite test) ANOVA was conducted. This analysis revealed only a main effect of fixation time between familiar and composite test trials ($\eta^2 = .049; F(1, 83) = 4.68; p = .033$). The combined sample had an average looking time of 3.55 s on the familiar test stimulus versus 4.66 s on the composite, suggesting that both groups perceived the composite face holistically rather than as a set of features to which they had been previously habituated.

2.2.3. Novel/dishabituation

GLM analyses identified a significant group difference for fixation on the *novel* test stimulus (dishabituation), $\eta^2 = .5; F(1, 86) = 4.32; p = .041$, with typically developing children having longer fixation rates (LSMEAN = .67) than those with SBM (LSMEAN = .57). To further evaluate this finding, separate GLM analyses were conducted to determine the relation of mental and motor functioning on novel fixation, controlling for site differences. The model including mental performance in relation to novel fixation time was not significant, $F(1, 85) = 1.99; p = .122$. However, the model for motor performance showed a significant total effect, $\chi^2 = .11; F(1, 85) = 2.94; p = .038$. Type III Sums of Squares showed that motor performance was significantly related to novel fixation time above and beyond etiology and site ($\eta^2 = .05; F(1, 85) = 4.22; p = .043$). This indicates that, despite the fact that typically developing children had higher motor scores overall, children with higher Bayley motor scores demonstrated longer novel fixation rates in both groups.

3. Discussion

The purpose of this study was to investigate early visual attention and information processing skills using Cohen's habituation/dishabituation paradigm. The results of the study indicated that difficulties with visual attention skills can be reliably detected in SBM at 18 months of age. In regard to *attention getting*, infants with SBM took significantly longer than typically developing infants to shift or disengage their attention from a salient sensory stimulus, a blinking light, to the face stimulus. This finding is consistent with difficulties identified in school-aged children with SBM, who have difficulty orienting to and disengaging from salient sensory stimuli (Dennis et al., 2005a). Together, our study and these school-age studies suggest fundamental problems with visual attention to salient environmental events, or difficulties at the level of automatic stimulus-driven orienting and these deficits are discernable very early in development.

Given that the children were 18 months of age, with trunk support provided by parent, it is unlikely that the longer disengagement times were related to fundamental motor difficulties at the level of head turning. However, difficulties in

disengagement/attention getting were found to be related to motor, but not mental scores on the BSID-II. Attention orienting and disengagement in older children with SBM is related to midbrain anomalies and posterior brain volume loss (Dennis et al., 2005a, 2006). The posterior parietal lobe is also implicated in the coordination of visual–motor functions (Andersen & Buneo, 2002) which are assessed by many of the tasks on the BSID-II motor scale. Attention getting and motor functioning may be related in this study through a common neural substrate. A test of this hypothesis awaits further study.

Habituation did not differentiate infants with SBM from typical developing infants, consistent with our hypothesis. No differences were identified in the proportion of infants that actually habituated to the familiar face stimuli per group. There were also no differences in infants' ability to habituate. In addition, there was no separate relation of motor and mental functioning to infants' ability to habituate. These findings suggest that once the attention of infants with SBM was engaged to the face they were as skilled as typical developing infants in sustaining their attention to this cognitively interesting stimulus. This is consistent with recent studies showing intact sustained attention in older children and adolescents with SBM (Swartwout et al., 2008), and with studies of learning in individuals with SBM which show intact implicit learning in tasks involving both the eyes and upper limbs (Colvin, Yeates, Enrile, & Coury, 2003; Edelstein et al., 2004; Salman et al., 2005).

We also did not find group differences on infants' fixation time to a composite face stimulus. To better understand this finding, we evaluated the difference between the composite face stimulus and the familiar face stimulus. Infants in both groups looked longer at the composite face stimulus. This suggests that all infants, regardless of group, responded holistically to the stimulus and were able to integrate the composite face features together as a whole. These findings suggest that infants with SBM process faces similarly to typical developing infants. Although they have deficits in some aspects of visual perception, face perception appears to be intact (Dennis, Fletcher, Rogers, Hetherington, & Francis, 2002) as it is in some other neurodevelopmental disorders that impair visual perception (e.g. William Syndrome) (Jernigan, Wang, & Bellugi, 1995).

Infants' ability to dishabituate was significantly different between the groups. Typically developing children looked significantly longer at the novel face stimulus than children with SBM, suggesting that they were better able to process the novel face as something new. Surprisingly, motor functioning was significantly related to dishabituation, so that infants with poorer motor functioning in both groups demonstrated less ability to recognize a stimulus as novel (dishabituate). These findings suggest that motor functioning may play a role in cognitive processes of dishabituation but not habituation. This is consistent with the emphasis other researchers have placed on the influence of early motor development on some perceptual cognitive skills (Bushnell & Boudreau, 1993; Gibson & Collins, 1982; Thelen & Smith, 1995).

The differences between infants with SBM and typical developing infants in dishabituation, but not habituation, suggest that it may be fruitful in other studies to investigate the type of learning and memory processes involved in habituation versus dishabituation and their relation to latent assets and deficit in learning and memory of older children and adults with SBM (Dennis et al., 2007).

4. Conclusions

The current study leads to two important conclusions. The first is that attention difficulties observed among children with SBM by school age can be documented and monitored early in life. There are important parallels between the infant habituation paradigms and covert attention paradigms used with older children that indicate continuity in development, especially for attention processes under stimulus control (Colombo, 2001). The findings are consistent with the proposal put forward by Dennis et al. (2006), that deficits in attention orienting and disengagement are core deficits associated with SBM; this study demonstrates their presence in infancy before formal education and across a range of cognitive ability levels associated with the sample in this study.

It is important to highlight that children with SBM in this study demonstrated relatively intact ability for some aspects of attention, including the ability to sustain attention to cognitively interesting stimuli (habituation). This is consistent with similar research finding that children with SBM had intact ability to sustain attention overtime compared to typically developing peers (Swartwout et al., 2008). Thus, the ability to sustain attention thought to be subserved by a more anterior attention system appears to be relatively intact in infants and older individuals with SBM.

This study had limitations, including the inability to obtain the habituation measures on the entire cohort. In addition, this study was limited by the inability to capture infant's orienting to an interesting stimulus (face) separate from disengagement from a sensory stimulus (blinking light). We were also unable to relate brain measures (e.g. volumetric measures from Magnetic Resonance Imaging) to the cognitive measures of attention, given the young age of the children. Nonetheless, this study makes an important contribution to our developing understanding of the early difficulties that can be identified among infants with SBM, as well as further delineating a unique pattern of strengths and weaknesses related to the neurobiology of the disorder.

The next step is to follow this cohort into school-age and directly assess performance on attention tasks derived from neurocognitive models of attention (Dennis et al., 2005a, 2005b) in relation to earlier performance on the habituation paradigm used in this study. Insight into the children with SBM's difficulties in attention and their relation to other cognitive processes and academic outcomes may be important for informing both early and later intervention programs and cognitive and academic support.

Acknowledgments

This research was supported by Grant PO1HD35946 funded by the National Institute for Child Health and Development and the National Institute of Neurological Disease and Stroke. We extend appreciation to Patricia Rosenlund from Easter Seals in McAllen, TX, for continued support and assistance throughout this project. Most important, we have considerable gratitude for the time and effort of the families who volunteered to help us with this study.

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