Sustained Attention in Children With Two Etiologies of Early Hydrocephalus

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Several studies have shown that children with spina bifida meningomyelocele (SBM) and hydrocephalus have attention problems on parent ratings and difficulties in stimulus orienting associated with a posterior brain attention system. Less is known about response control and inhibition associated with an anterior brain attention system. Using the Gordon Vigilance Task (Gordon, 1983), we studied error rate, reaction time, and performance over time for sustained attention, a key anterior attention function, in 101 children with SBM, 17 with aqueductal stenosis (AS; another condition involving congenital hydrocephalus), and 40 typically developing controls (NC). In SBM, we investigated the relation between cognitive and parent ratings of inattention and hyperactivity and explored the impact of medical variables. Children with SBM did not differ from AS or NC groups on measures of sustained attention, but they committed more errors and responded more slowly. Approximately one-third of the SBM group had attention symptoms, although parent attention ratings were not associated with task performance. Hydrocephalus does not account for the attention profile of children with SBM, which also reflects the distinctive brain dysmorphologies associated with this condition.

Keywords: spina bifida, aqueductal stenosis, hydrocephalus, attention, continuous performance task

Spina Bifida Meningomyelocele

Spina bifida is one of the most common birth defects in North America, accounting for approximately 19.3 per 100,000 live births in 2004 (Martin et al., 2006). The most common form of spina bifida is a meningomyelocele, identified by an open defect on the spine with protrusion of the spinal cord and meninges at any point along the spinal column (Detrait et al., 2005). SBM is also associated with structural brain abnormalities. The Chiari II malformation of the cerebellum and hindbrain occurs in about 95% of cases (Barkovich, 2005) and produces hydrocephalus requiring treatment with a diversionary shunt. Neuroimaging studies have also identified abnormalities of the midbrain secondary to a small posterior fossa, congenital absence and/or thinning of the corpus callosum, and thinning of the cortical mantle, especially in posterior brain areas (Fletcher et al., 2005). A high proportion of children with SB also have ophthalmic problems that include strabismus (30 to 73%: Biglan, 1990, 1995; Gaston, 1985; Lennertstrand, Gallo, & Samuelsson, 1990; McIlwaine, Musaji, & Buncie, 1993), paralysis of voluntary conjugate vertical eye movements (Gaston, 1985), horizontal or rotatory nystagmus (Gaston, 1985), astigmatism (Gaston; McIlwaine et al., 1993), and optic atrophy (Gaston, 1985).

Despite extensive CNS abnormalities, SBM is rarely associated with mental retardation. Rather, children with SBM have strengths and weaknesses within each of several cognitive and educational domains (see reviews by Dennis, Landry, Barnes, & Fletcher, 2006; Wills, 1993; Yeates, Dennis, & Fletcher, 2008).

Attention in SBM

Attention is an area of particular interest for neuropsychological research on SBM not only because of the pattern of attention strengths and weaknesses, but also because of the unanswered questions. Studies of attention in SBM have assessed either
behavioral (parent rated) or cognitive (neuropsychological test-based) attention, though rarely both. Behavioral and cognitive measures do not always yield parallel results (Gordon, Barkley, & Lovett, 2006), and different components of attention can be dissociated both by rating scales and neuropsychological assessments (Knudsen, 2007). Research based on assessments derived from neuropsychology (Mirsky, Anthony, Duncan, Ahearn, & Kellam, 1991) and experimental cognitive neuroscience (Posner & Peterson, 1990; Knudsen, 2007) have distinguished two overarching attention networks: a "posterior" system concerned with stimulus-driven orienting and focusing, and disengaging from and shifting attention; and an "anterior" system concerned with voluntarily maintaining vigilance and sustaining attention. The posterior system is a largely bottom-up network driven by environmental salience, and is thought to include midbrain structures and posterior parietal areas (Posner & Petersen, 1990; Posner & Raichle, 1994). The anterior system is a top-down regulatory network that involves cognitively driven response control, and is purported to include frontal and parietal regions as well as the reticular nucleus of the brainstem.

These models have been applied to a variety of populations with neurodevelopmental disorders, including SBM. Dennis and colleagues (2005a, 2005b) identified difficulties in stimulus-driven orienting in children with SBM. On a version of the Posner visual orienting and detecting task (Posner, Early, Reiman, Pardo, & Dhawan, 1988), these authors found that relative to controls, children with SBM oriented more slowly to stimuli designed to engage their attention and also were slower in disengaging from these stimuli. In addition, children with SBM and midbrain abnormalities did not modify their response to stimuli to which they had previously oriented. These deficits in focusing, engaging, and inhibition of return are hypothesized to be under the control of attention systems in the midbrain and posterior cortex, and led Dennis et al. (2005b) to conclude that children with SBM had selective impairment of the posterior attention system. In contrast to their poor orienting, children with SBM had relatively intact ability for some response regulatory processes mediated by anterior brain areas.

Sustained attention, defined here as effortfully maintaining an attentional state or vigilance over time, has been investigated in SBM, although the results are inconsistent. Three studies of children with SBM have utilized the Gordon Diagnostic System (GDS; Gordon, 1983), Vigilance task, one type of continuous performance task (CPT) measure of sustained attention. Lollar (1990) found that children with SBM performed below normative levels on the GDS in terms of both "impulsivity and inattention" (p. 39). Loss, Yeates, and Enrile (1998) found that children with SBM made more errors on a composite measure of omissions and commissions, relative to sibling controls; that performance was marginally poorer in children with oculomotor abnormalities; and that GDS performance was a significant predictor of parent-rated behavior problems using the Child Behavioral Checklist (Loss et al., 1998). In contrast, Colvin, Yeates, Enrile, and Coury (2003) found that children with ADHD committed more GDS errors of commission than children with SBM and shunted hydrocephalus, and sibling controls, with no group differences in the number of omissions. Rose and Holmbeck (2007) found that shunted and nonsunted adolescents with spina bifida performed more poorly than controls on cognitive measures of focused attention and planning using the Cognitive Assessment System (Naglieri & Das, 1997). Some of these tasks involve a significant motor component (e.g., underlining, filling in symbols), with other studies of children with SBM finding that requirements for underlining and fine motor speed make these types of measures less than adequate for children with SBM, who typically have significant fine motor difficulties (see Dennis et al., 2006). There are a number of possible reasons for these apparently inconsistent results. These include differences in operational definitions of sustained attention, variations in ADHD status, and SBM group composition.

Many studies treat CPT errors as indicators of response control, with errors of commission representing an index of impulsive responding and errors of omission representing distractibility (Mirskey et al., 1991). Although relevant, measures of omission or commission may arise from several sources (e.g., underarousal, poor stimulus orientation), and do not adequately measure the maintenance of a vigilant state, which is usually understood to involve tracking performance across time blocks or assessing variability over time. Brewer, Fletcher, Hiscok, and Davidson (2001) is the only study in the SBM literature that operationalized sustained attention as performance across time blocks. Using a version of the visual orienting and detection task (VODT; Posner et al., 1988) and a CPT (Halperin et al., 1988), Brewer et al. compared children with ADHD (combined type), children with shunted hydrocephalus (most with SBM), and typically developing controls. Children with shunted hydrocephalus achieved faster reaction time toward the end of the VODT, and performed similarly to controls overall, whereas children with ADHD showed increasing reaction time over time blocks. Children with hydrocephalus, but not children with ADHD, made more errors of omission and commission than controls on the CPT. Unfortunately, Brewer et al. did not address the variable presence of ADHD and subtypes within the group with hydrocephalus. They also combined etiologies of early hydrocephalus, so the results may not be representative of the specific etiology of SBM.

The issue of parent-rated ADHD status is important because not all children with SBM show significant attention problems, and even those rated as inattentive are unlikely to be either hyperactive or impulsive (Vachha & Adams, 2005). For example, Ammerman et al. (1998) found that 33% of children with SBM were rated above clinical cutoff scores for ADHD, especially the inattentive type. Similarly, Burmeister et al. (2005) found that 31% of their sample with SBM showed symptomatology consistent with ADHD, with approximately three-quarters of these of the inattentive type. Colvin et al. (2003) found that children with ADHD had higher rates of inattentive and hyperactive-impulsive symptomatology than children with SBM, who in turn had more inattentive symptoms than siblings. Thus, it is possible that only a subset of children with SBM—those with evidence of behavioral inattention—will show difficulties on a cognitive assessment of attention.

SBM is a common etiology of hydrocephalus, but hydrocephalus also arises from other sources, such as aqueductal stenosis (AS), a rare condition in which the cerebral aqueduct connecting the third and fourth ventricles is congenitally narrowed and produces hydrocephalus, usually at the level of the superior colliculus (Barkovich, 2005). Early hydrocephalus, regardless of etiology, causes ventricular enlargement in a posterior-to-anterior fashion (Van Roost, Solymosi, & Funke, 1995), resulting in decreased posterior brain volumes, with volumes anterior to the genu of the...
corpus callosum comparable to healthy controls (Fletcher et al., 1996; Juranek et al., 2008). However, children with SBM also have abnormalities involving the midbrain, cerebellum, and corpus callosum. Therefore, it is difficult to simply attribute attention problems to the back-to-front thinning associated with hydrocephalus, especially because patterns of cortical thinning are variable within the population (Dennis et al., 1981; Young, Nulsen, Weiss, & Thomas, 1973). Indeed, Dennis et al. (2005a, 2005b) found that children with midbrain anomalies have more significant attention orienting difficulties than children with SBM and no midbrain anomalies. Dennis et al. (2005a) also found small but statistically significant negative correlations of reaction time (RT) to cued targets with right-hemisphere white matter volumes posterior to the end of the corpus callosum, which they interpreted as reflecting vulnerability of the parietal component of the posterior network. Although children with AS typically require shunt diversion of CSF and show structural abnormalities related to hydrocephalus (e.g., corpus callosum hypoplasia), they rarely demonstrate the degree of congenital dysmorphology of the midbrain, corpus callosum, and posterior brain characteristic of children with SBM.

The Present Study

In contrast with previous studies using small samples, no comparison groups, primarily lower spinal lesions, and/or mixed etiologies, the present study assessed both sustained attention and behavioral attentional symptomatology in a large sample of children grouped on the basis of both etiology (SBM, AS, NC) and ADHD subtypes. Comparison of SBM and AS groups clarifies whether hydrocephalus contributes to attention problems in children with SBM. The relation between cognitive (e.g., CPT) and behavioral (e.g., rating scales) has been shown to be weak in other populations (e.g., Gordon et al., 2006), but it has not been explicitly examined in hydrocephalus, where the incidence of ADHD is high but does not occur in every child, and is largely restricted to the inattentive type. Therefore, we examined the relation of attention performance with parent ratings of ADHD symptomatology and specifically addressed whether ADHD status would be related to cognitive attention difficulties. We also evaluated the role of key medical variables (e.g., spinal lesion level, oculomotor, and midbrain abnormalities) on attention to determine whether variability in performance within the SBM group can be attributed to clinical markers. Other studies have evaluated medical variables, though typically in samples of children with only lumbar and sacral spinal lesions. Including children with thoracic lesions, who have more significant brain dysmorphology, provides a more representative sample of children with SBM (Fletcher et al., 2005).

The first hypothesis concerned group differences on the CPT. We hypothesized that children with SBM would have slower and less accurate CPT performance relative to the NC group. Because they have hydrocephalus but not the specific brain abnormalities of SBM, children with AS were expected to perform more like the NC group than the SBM group. Based on the literature, we did not expect group differences in performance over time across blocks of the task, and we did not expect a difference in commission errors, which are usually deemed to reflect impulsive responses.

The second hypothesis evaluated the role of parent-rated attention and neurological/medical complications in the prediction of CPT performance within the SBM group. We predicted that poor CPT performance would be associated with ADHD status and medical complications. We also expected that omission errors on the GDS would be positively correlated with parent ratings of inattention on the Swanson Nolan Achenbach Pelham–IV (SNAP–IV) Parent Rating Scale (Swanson, 1992), and that commission errors would be positively correlated with parent ratings of hyperactivity/impulsivity.

Method

Participants

Participants in the two clinical groups (SBM and AS) were children and adolescents (ranging in age from 7 to 17 years at the time of evaluation) in a larger study of neurobehavioral function in SBM and related disorders (Fletcher et al., 2004) recruited from clinics and local neurosurgical practices in Houston (44 children with SBM, 12 with AS) and Toronto (57 with SBM, 5 with AS). Participants with SBM were identified at birth, whereas those in the AS group were diagnosed in the first year of life. All participants had been treated for hydrocephalus, usually with a shunt; hydrocephalus in two participants with AS had been treated with third ventricle ventriculostomy. The typically developing group (NC; 15 children from Houston, 25 from Toronto) was composed of age-matched children recruited through local advertisements and posters.

All children were English-speaking and had Verbal Reasoning and/or Abstract/Visual Reasoning subtest scores greater than 70 on the Stanford-Binet Test of Intelligence (4th ed.; Thorndike, Hagen, & Sattler, 1986). Exclusion criteria for all children were neurological disorders unrelated to SBM or AS, severe psychiatric disorder, uncontrolled seizure disorder, uncorrected sensory disorder, or inability to control the upper limbs. Additional exclusion criteria for the NC group were identified neurobehavioral disorders such as learning disabilities or ADHD. The study was approved by the human participants review boards at all institutions, and, prior to participation, participants and their parents gave informed assent and consent, respectively.

Sociodemographic and IQ information on participants (101 children and adolescents with SBM, 17 with AS, and 40 NC) is presented in Table 1. The groups did not differ significantly on gender or age at test (p > .05). When race was analyzed as a comparison of Hispanic and non-Hispanic participants, the groups did not differ significantly, p > .05, but a clear trend for greater representation of Hispanics in the group with SBM is apparent, which is not surprising because SBM occurs most frequently in Hispanics. Because the Hispanic subgroup with SB is generally economically disadvantaged, children with SBM had lower socioeconomic status (SES) than children in the typically developing group, F(2, 154) = 3.20, p = .04. As expected, children with SBM demonstrated a significantly lower IQ composite score than children with AS, who in turn performed significantly below the NC group, F(2, 155) = 40.87, p < .0001.

Information on clinical markers and MRI findings in children with SBM and AS is presented in Tables 2 and 3. Children with SBM and AS did not differ from each other on variables related to hydrocephalus, including number of shunt revisions, history of shunt infection, history of seizures, presence of oculomotor abnormalities, or corpus callosum status (all ps > .05). As expected, children with SBM and AS differed on clinical variables related to
etiology, including ambulatory status, bladder function, and neural abnormalities unrelated to hydrocephalus. Some cell sizes were too small for statistical analysis, so proportions are presented. For ambulatory status, no children with AS were classified as partially ambulatory or unable, whereas 79% of children with SBM fell in these two categories. No children with AS had abnormal bladder function, in contrast to over 95% of children with SBM. A similar pattern was found for presence of a Type I or II Chiari malformation: only one child with AS (7%) had a Chiari II, in contrast to over 95% of children with SBM. As expected, less than 20% of children with AS, but 81% of those with SBM, had the tectal beaking malformation, the predominant midbrain anomaly. Tectal beaking refers to the appearance of the roof of the mesencephalon when it is compressed and assumes a beaked appearance (Barkovich, 2005). It is part of the cascade of anomalies associated with the Chiari II malformation, all of which are a mechanical consequence of brain development in a small posterior fossa (McLone & Knepper, 1989).

Measures
SNAP–IV. This scale consists of 90 items consistent with ADHD and oppositional defiant disorder (ODD) criteria in Diagnostic and Statistical Manual of Mental Disorders (4th ed. [DSM–IV]; American Psychiatric Association, 1994), as well as items from the Conners Index Questionnaire (Conners, 1984), IOWA Conners Questionnaire (Loney & Milich, 1982), and items from DSM–IV disorders that may overlap with criteria for ADHD. Children are rated on a 4-point scale ranging from 0 (not at all), 1 (just a little), 2 (quite a bit), to 3 (very much) on each behavioral item. We focused only on the 18-item DSM–IV based inattention and hyperactivity/impulsivity scales to identify children with

Table 1
Sociodemographic and Handedness Information by Etiology Group

<table>
<thead>
<tr>
<th>Group</th>
<th>Control</th>
<th>SBM</th>
<th>AS</th>
</tr>
</thead>
<tbody>
<tr>
<td>N</td>
<td>40</td>
<td>101</td>
<td>17</td>
</tr>
<tr>
<td>Age (years: M ± SD)</td>
<td>12.41 ± 2.61</td>
<td>12.40 ± 2.67</td>
<td>12.69 ± 2.57</td>
</tr>
<tr>
<td>Gender—n (%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>20 (50%)</td>
<td>53 (52%)</td>
<td>8 (47%)</td>
</tr>
<tr>
<td>Female</td>
<td>20 (50%)</td>
<td>48 (48%)</td>
<td>9 (53%)</td>
</tr>
<tr>
<td>Handedness—n (%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Right</td>
<td>36 (97%)</td>
<td>72 (78%)</td>
<td>11 (73%)</td>
</tr>
<tr>
<td>Left</td>
<td>1 (3%)</td>
<td>20 (22%)</td>
<td>4 (27%)</td>
</tr>
<tr>
<td>Ethnicity—n (%)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Caucasian</td>
<td>30 (75%)</td>
<td>74 (73%)</td>
<td>15 (88%)</td>
</tr>
<tr>
<td>Hispanic</td>
<td>2 (5%)</td>
<td>18 (18%)</td>
<td>2 (12%)</td>
</tr>
<tr>
<td>African American</td>
<td>2 (5%)</td>
<td>4 (4%)</td>
<td>0</td>
</tr>
<tr>
<td>Asian American</td>
<td>5 (13%)</td>
<td>3 (3%)</td>
<td>0</td>
</tr>
<tr>
<td>Other</td>
<td>1 (3%)</td>
<td>2 (2%)</td>
<td>0</td>
</tr>
<tr>
<td>Socioeconomic status (M ± SD)</td>
<td>46.25 ± 11.97</td>
<td>40.46 ± 12.92</td>
<td>44.12 ± 12.42</td>
</tr>
<tr>
<td>Stanford-Binet composite IQ (M ± SD)</td>
<td>108.38 ± 9.69</td>
<td>88.93 ± 12.27</td>
<td>100.94 ± 13.79</td>
</tr>
</tbody>
</table>

Note. SBM = spina bifida meningomyelocele; AS = aqueductal stenosis.

* Significant difference between groups at p < .05.

Table 2
Clinical Markers by Etiology Group

<table>
<thead>
<tr>
<th>Group</th>
<th>SBM</th>
<th>AS</th>
</tr>
</thead>
<tbody>
<tr>
<td>No. shunt revisions (M, SD)</td>
<td>2.02 (2.49)</td>
<td>2.07 (2.58)</td>
</tr>
<tr>
<td>History of shunt infection</td>
<td>17 (17%)</td>
<td>1 (6%)</td>
</tr>
<tr>
<td>History of seizures</td>
<td>20 (20%)</td>
<td>4 (24%)</td>
</tr>
<tr>
<td>Presence of oculomotor abnormalities</td>
<td>37 (37%)</td>
<td>7 (41%)</td>
</tr>
<tr>
<td>Ambulatory statusa</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Normal</td>
<td>2 (2%)</td>
<td>16 (94%)</td>
</tr>
<tr>
<td>Independent</td>
<td>19 (19%)</td>
<td>1 (8%)</td>
</tr>
<tr>
<td>Partial</td>
<td>39 (39%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Unable</td>
<td>41 (41%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Abnormal bladder functiona</td>
<td>95 (94%)</td>
<td>0 (0%)</td>
</tr>
</tbody>
</table>

Note. SBM = spina bifida meningomyelocele; AS = aqueductal stenosis.

* No statistical analyses were performed due to inadequate cell sizes.

Table 3
MRI Findings by Etiology Group

<table>
<thead>
<tr>
<th>Group</th>
<th>SBM</th>
<th>AS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Corpus callosum status</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Normal</td>
<td>7 (7%)</td>
<td>1 (6%)</td>
</tr>
<tr>
<td>Hypoplastic</td>
<td>44 (44%)</td>
<td>11 (65%)</td>
</tr>
<tr>
<td>Some agenesis</td>
<td>38 (38%)</td>
<td>4 (24%)</td>
</tr>
<tr>
<td>Tectal dysmorphology</td>
<td>72 (81%)</td>
<td>3 (18%)</td>
</tr>
<tr>
<td>Chiari malformationa</td>
<td></td>
<td></td>
</tr>
<tr>
<td>None</td>
<td>2 (2%)</td>
<td>14 (82%)</td>
</tr>
<tr>
<td>Type I</td>
<td>2 (2%)</td>
<td>0 (0%)</td>
</tr>
<tr>
<td>Type II</td>
<td>74 (73%)</td>
<td>1 (6%)</td>
</tr>
</tbody>
</table>

Note. SBM = spina bifida meningomyelocele; AS = Aqueductal stenosis.
attention problems. Although the gold standard for identifying children with ADHD is a structured interview, which also takes into account the other diagnostic criteria (age of onset, behavior displayed in multiple contexts), formally diagnosing ADHD was not a primary objective of the study and a structured interview would have been time consuming and expensive. The SNAP-IV has seen extensive validation as a screener for attention problems, showing sensitivity to math difficulties (Fuchs et al., 2006) and relating well to the results of structured interviews. In the National Institute of Mental Health-sponsored ADHD multimodal clinical trial (MTA Cooperative Group, 1999), the SNAP-IV was the primary outcome measure and there was good concordance of the SNAP-IV parent ratings and the structured interview used to identify children with ADHD. Test–retest reliability is .77 to .80, and the measure is sensitive to ADHD treatment effects (Pelham, Fabiano, & Massetti, 2005).

ADHD status (see Table 4) was defined by a rating of 1.78 or higher on the Inattention scale or 1.44 or higher on the Hyperactivity/Impulsivity scale of the SNAP-IV questionnaire (Swanson, 1992) completed by parents and scored according to Swanson et al. (2002). Children were dichotomously categorized as having, or not having, significant ADHD symptomatology, on the basis of the cutoffs above (hereafter, ADHD status). Although the cell sizes are too small for quantitative analysis, examination of sociodemographic, IQ, MRI, and clinical markers by ADHD status did not reveal trends indicative of differences across subtypes.

GDS. The GDS Vigilance Task is a continuous performance task of sustained attention. Numbers 0 to 9 appear in quasi-random fashion on a console with a central screen at a rate of one per second, and children are instructed to press a button every time a 9 appears immediately following a 1. Variables recorded include number of times the child does not press the button following a 1/9 combination (omission errors), number of incorrect presses (commission errors), and mean reaction time (in milliseconds) for correct responses. Scores are provided for total performance as well as within each of three 3-min blocks (containing 15 targets each). Commission variability is equal to the standard deviation of the three block commission error scores and is a measure of consistency in impulsive responding. The GDS has been shown to discriminate between ADHD and learning disabilities (Aylward, Verhulst, & Bell, 1990). GDS scores accurately classified 90% of children with DSM-based ADHD diagnoses and 52% of children with no ADHD diagnosis (Mayes, Calhoun, & Crowell, 2001). The GDS is moderately correlated with parent and teacher ratings of attention problems, and performance improves with administration of stimulant administration (Nichols & Waschbusch, 2004; O’Laughlin & Murphy, 2000).

Procedures

Cognitive and behavioral assessments. Children were administered the Stanford-Binet and GDS Vigilance Task as part of a longer assessment battery. Parents completed the SNAP-IV. In two cases, parent SNAP-IV was not returned, but teacher forms were available for these children. Teacher ratings for these two children were well below clinical cut points, so they were included in the “no ADHD” group. Sample sizes vary for each measure due to inability or declining to participate. Each child’s status on medical and clinical marker variables was obtained via parent interview and confirmed through examination of medical records, or through structural MRI scans that were coded by two pediatric radiologists (Fletcher et al., 2005).

MRI scan procedures. The MRI scans used by the radiologists to code clinical markers were obtained on comparable General Electric Signa 1.5 tesla magnets in Houston and Toronto. After an initial sagittal scout (spin-echo T1-weighted localizer, FOV 24 cm, TR 500ms, TE 14ms, 256 × 192 matrix, 3 mm with a 0.3 skip, 2 repetitions), two separate whole brain T1 and T2 coronal acquisitions were obtained to ensure adequate estimation of CSF versus gray and white matter. One series was a 3D spoiled gradient-echo with contiguous 1.7 mm coronal images, FOV 24 cm, TR 18ms, TE 3ms, Flip angle 25 degrees, 124 locations, 256 × 256 matrix, 1 repetition. The second series was 3D fast spin-echo T1-weighted sequence, FOV 24 cm, TR 4000ms, TE 102ms, ETI 16, 256 × 256 matrix, 1 repetition with contiguous 1.7 mm coronal images.

Data Analysis

Variables of interest were explored for violations of statistical assumptions. This exploration excluded five outliers for number of commission errors greater than 50 or commission variability across blocks greater than 10 on the GDS. In addition, square-root transformations of commission errors and commission variability were employed to meet homogeneity of variance assumptions. Because the GDS norms may be outdated and not representative of the current population, raw scores were used in all analyses.

Age at evaluation had a significant positive relationship with task performance and was used as a covariate in most analyses to increase power. Intellectual ability was also significantly correlated with GDS scores (r = .24 to .34, p < .01), but was not included as a covariate. Analysis of covariance (ANCOVA) is inappropriate when IQ differences are an inherent characteristic of the disorder. This problem is compounded when IQ is correlated with the neurocognitive measure. These issues have been discussed extensively in the statistical (Evans & Anastasio, 1968; Lord, 1967, 1969) and neuropsychological (Adams, Brown, & Grant, 1985; Tupper & Rosenblood, 1984) literature. These articles and simulations show the difficulties associated with ANCOVA when used to “equate” nonrandomized groups that are unequal, or to exert statistical control whose objective is to eliminate consideration of the covariate as an explanation of the result. In the case of IQ, doing so artificially raises IQ scores in SBM and artificially lowers IQ scores in controls, essentially creating situations that do not exist in nature. The fact that

Table 4

<table>
<thead>
<tr>
<th>ADHD Classification by Etiology Group</th>
<th>Group</th>
</tr>
</thead>
<tbody>
<tr>
<td>N</td>
<td></td>
</tr>
<tr>
<td>No ADHD—a (%)</td>
<td>36 (90%)</td>
</tr>
<tr>
<td>ADHD—a (%)</td>
<td></td>
</tr>
<tr>
<td>Inattentive type</td>
<td>3 (8%)</td>
</tr>
<tr>
<td>Hyperactive/Combined type</td>
<td>1 (2%)</td>
</tr>
</tbody>
</table>

Note. SBM = spina bifida meningomyelocoele; AS = aqueductal stenosis; ADHD = attention-deficit/hyperactivity disorder.

*No children in this group had clinical identifications of treatment for ADHD.

...
IQ scores are related to group membership diminishes the utility of IQ as a covariate in any case, particularly because IQ cannot “cause” group membership. A similar argument might be made for SES, although here, SES was also not related to the dependent measures, further diminishing its utility. Given these reasons, we did not covary for either IQ or SES.

Variations of ANCOVA were used to test Hypothesis 1 with etiology (SBM, AS, NC) as the between-subjects variable and age at evaluation as the covariate. Although clinical diagnosis of ADHD was an exclusionary criterion for the NC group, four children met criteria for significant ADHD symptomatology on the parent ratings. In addition, five children in the AS group met these criteria. To maximize power, analyses in Hypothesis 1b were initially run with these children included. To address concerns that the inclusion of children with significant symptomatology in the control groups might hinder interpretation, analyses were also run without these children. The results were similar except where noted.

For Hypothesis 2a, hierarchical regression was used, whereas Hypothesis 2b was tested with correlational methods. Post hoc group comparisons were evaluated via Fisher’s least significant difference (LSD), which controls alpha at .05 while maximizing power in the special case of three-group hypothesis tests (Levin, Serlin, & Seaman, 1994; Maxwell & Delaney, 2004).

Results

Hypothesis 1a: Group Comparisons of Sustained Attention

Repeated measures ANCOVA of number of omissions, number of commissions, and reaction time (covarying age) indicated that there were no significant Interaction × Block interactions and no main effects of block, indicating that children in each group performed similarly over the course of the task (ps > .05). Table 5 shows the omnibus F values for these analyses. Figure 1 graphically displays these results for RT over trial blocks and also shows the group with SBM divided by ADHD status. A simple comparison of these subgroups of SBM was not significant, p > .05. Given these results, we conducted a univariate ANCOVA for each of the dependent variables, collapsing across blocks.

Hypothesis 1b: Omission and Commission Errors

Table 6 shows the scores on the GDS by etiology group. A one-way ANCOVA for total omission errors (covarying age) revealed an overall model effect, F(3, 154) = 15.12, p < .0001. There was a main effect for etiology over and above the contribution of age, F(2, 154) = 9.68, p = .0001. Follow-up comparisons revealed that the AS and NC groups did not differ (p > .05), but both groups outperformed participants with SBM (p < .025). Effect size calculations indicated a medium difference (d = .72) between the NC and SBM groups, and between the AS and SBM groups (d = .60). Thus, children with SBM were more inattentive than either comparison group.

A one-way ANCOVA for total number of commission errors (square-root transformed; covarying age) revealed an overall model effect, F(3, 154) = 17.48, p < .0001. There was a main effect for etiology over and above the contribution of age, F(2, 154) = 3.86, p < .03. Follow-up comparisons revealed that AS and NC groups did not differ from one another (p > .05), but the NC group outperformed participants with SBM (p < .01). Effect size was larger between the NC and SBM groups (d = .41) and between the NC and AS groups (d = .39) than between AS and SBM (d = .02). Thus, both groups with hydrocephalus committed more commission errors than the typically developing controls, which was not expected.

A one-way ANCOVA for overall mean RT (covarying age) revealed an overall model effect, F(3, 154) = 14.65, p < .0001. The main effect for etiology was not significant over and above the contribution of age, p > .05. However, when children in the AS and NC groups with significant ADHD symptomatology were excluded, analysis revealed that the NC-noADHD group outperformed participants with SBM, p < .05, suggesting that the initial lack of a group difference was associated with slow reaction times in NC children with clinically significant parent ratings. With all children in the analysis, there was a small effect size between children with SBM and those with AS (d = .29). With NC and AS children with ADHD symptomatology excluded, there were small-to-medium effect size differences between the NC-noADHD (M = 43.86, SD = 7.51) and SBM groups (M = 46.96, SD = 7.97; d = .37) and between AS-noADHD (M = 42.75, SD = 8.32) and SBM (d = .48).

The one-way ANCOVA for commissions variability (square-root transformed; covarying age) revealed an overall model effect, F(3, 154) = 8.41, p < .0001. The main effect for etiology was not significant over and above the contribution of age, p > .05. However, Cohen’s d indicated a small difference (d = .27) between the groups with NC and SBM; the effect size between AS and SBM was also small (d = .16). When children in the AS and NC groups with significant ADHD symptomatology were removed, analysis revealed that the NC-noADHD group (raw M = 0.73, SD = 0.74) outperformed participants with AS-noADHD (raw M = 1.50, SD = 1.68), p < .02, due to higher mean variability in children with AS and low ADHD symptomatology. In both models, the SBM group was not significantly different from either the NC or AS groups.

Hypothesis 2a: Impact of ADHD Status and Medical Variables on SBM Performance

We examined the impact of ADHD status and medical variables (lesion level, tectal beaking, and oculomotor abnormalities) on the summary GDS variables within the SBM group with a series of hierarchical models, built in three steps. First, the age covariate
was entered. In the second step, ADHD status was entered, and finally, the medical variables were entered as a block in the third step. Only 84 children with SBM were included in these analyses due to missing data on medical variables. For each of the dependent variables, the beta-weights and $R^2$-change for each step are shown in Table 7.

For total omission errors, age was significant, $F(1, 82) = 11.87$, $p < .001$, with a corresponding $R^2$ of .13. ADHD status was not significant when added at step two, $F(1, 81) = 3.32$, $p = .07$. At the third step, the increase in predictive power was not significant, $R^2$ change .02, $F(3, 78) = .68$, $p = .57$. In the final model with all predictors, the only unique predictor was age, $p = .0002$, such that older children made fewer omission errors.

For total commission errors, age was significant, $F(1, 82) = 22.47$, $p < .0001$, with a corresponding $R^2$ of .22. ADHD status was not significant when added at step two, $F(1, 81) = 3.12$, $p = .08$. At the third step (medical variables), the increase in predictive power was significant, $R^2$ change .08, $F(3, 78) = 3.10$, $p < .04$. In the final model with all predictors, the only unique predictor was age and tectal beaking, $p < .01$, such that number of errors decreased with age, and children with tectal beaking committed significantly fewer commission errors than those with a normal tectum. The direction of this difference was unexpected; however, follow-up examination of the data revealed that the distributions of dependent variables as well as medical variables within SBM may be driving this difference. In particular, there were five participants who did not meet criteria as outliers with a very high number of errors (range = 18 to 28) and the proportion of children with SBM but no tectal beaking is small (18%); both of these factors may have contributed to the finding. Two of the children with a high number of errors were in the group with no tectal beaking, while three were in the much larger group with tectal beaking. Without these five cases, we found that the difference between beaking and no beaking was no longer significant. In addition, given that children with SBM and no tectal beaking were significantly older than those with it, and the negative relation of age and commission errors, covarying for age in this model may have artificially inflated error scores in children with no beaking. When the model was run without age as a covariate, tectal status was no longer a significant predictor of performance, even where the five cases were retained.

For overall mean RT, age was significant, $F(1, 82) = 16.85$, $p < .0001$, with a corresponding $R^2$ of .17. ADHD status was not significant when added at step two, $F(1, 81) = 1.97$, $p = .16$. At the third step (medical variables), the increase in predictive power was not significant, $R^2$ change .05, $F(3, 78) = 1.88$, $p = .14$. In the

Table 6
Mean Gordon Task Scores by Etiology Group (Hypothesis 1b)

<table>
<thead>
<tr>
<th>Gordon variable</th>
<th>Control M (SD)</th>
<th>SBM M (SD)</th>
<th>AS M (SD)</th>
<th>SBM-Control</th>
<th>SBM-AS</th>
<th>AS-Control</th>
</tr>
</thead>
<tbody>
<tr>
<td>Omission errors$^{ab}$</td>
<td>2.68 (2.67)</td>
<td>6.21 (5.70)</td>
<td>3.24 (3.63)</td>
<td>.72</td>
<td>.60</td>
<td>.11</td>
</tr>
<tr>
<td>Commission errors$^{ac}$</td>
<td>2.58 (3.02)</td>
<td>4.81 (5.77)</td>
<td>4.71 (7.45)</td>
<td>.41</td>
<td>.02</td>
<td>.39</td>
</tr>
<tr>
<td>Mean RT$^a$</td>
<td>45.15 (8.24)</td>
<td>46.96 (7.97)</td>
<td>44.65 (8.57)</td>
<td>.22</td>
<td>.29</td>
<td>-.06</td>
</tr>
<tr>
<td>Commission variability$^{cd}$</td>
<td>0.73 (0.71)</td>
<td>1.02 (1.09)</td>
<td>1.19 (1.52)</td>
<td>.27</td>
<td>-.16</td>
<td>.43</td>
</tr>
</tbody>
</table>

Note. Cohen’s $d$ effect sizes computed using raw scores and pooled standard deviations. Scale for RT is given in deciseconds. SBM = spina bifida meningomyelocele; AS = aqueductal stenosis; RT = reaction time.

$^a$ Significant difference between SBM and Control groups. $^b$ Significant difference between SBM and AS groups. $^c$ Means presented are raw scores; square-root transformation was employed in analyses. $^d$ Calculated as the standard deviation of the three block scores of commission errors.
Table 7

Hierarchical Regression Analysis Relating Medical Variables to Gordon Task Performance in Children With SBM (Hypothesis 2a)

<table>
<thead>
<tr>
<th>Step and predictor</th>
<th>Omission errors</th>
<th>Commission errors</th>
<th>Mean RT</th>
<th>Commission variability*</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>β</td>
<td>ΔR²</td>
<td>β</td>
<td>ΔR²</td>
</tr>
<tr>
<td>1. Age</td>
<td>−.36</td>
<td>.13</td>
<td>−.46</td>
<td>.22</td>
</tr>
<tr>
<td>2. ADHD status (yes/no)</td>
<td>.19</td>
<td>.03</td>
<td>.17</td>
<td>.02</td>
</tr>
<tr>
<td>3. Medical variables:</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Lesion level (high/low)</td>
<td>−.05</td>
<td>.01</td>
<td>.03</td>
<td>.06</td>
</tr>
<tr>
<td>Tectal beaking (present/absent)</td>
<td>−.13</td>
<td>−.31*</td>
<td>−.15</td>
<td>−.35*</td>
</tr>
</tbody>
</table>

Note. Scale for RT is given in deciseconds. SBM = spina bifida meningomyelocele; RT = reaction time.

* Calculated as the standard deviation of the three block scores of commission errors.

*p < .05.

The final model with all predictors, the only unique predictors were age and oculomotor abnormalities, ps < .05, such that RT decreased with age, and children who had oculomotor abnormalities responded more slowly than those who did not.

For commission variability, age was significant, F(1, 82) = 11.16, p < .002, with a corresponding R² = .12. ADHD status was not significant when added at step two, F(1, 81) = .77, p = .38. At the third step, the increase in predictive power was significant, R² change = .12, F(3, 78) = 4.12, p < .01. In the final model with all predictors, the only unique predictors were age and tectal beaking, such that variability decreased with age, and children with tectal beaking demonstrated less variability. Again, the direction of this difference was unexpected, but follow-up analyses revealed that when the model was run with extreme cases excluded, tectal status was no longer a significant predictor of performance. Furthermore, as with commission errors, tectal beaking was no longer significant when the model was run without covarying for age.

Hypothesis 2b: Cognitive Performance and Behavioral Ratings

As predicted, in the SBM group, cognitive inattention (total omissions) was significantly correlated with parent ratings of inattention (r = .21, p = .01) but not with ratings of hyperactivity/impulsivity (r = .13, p = .12); conversely, cognitive impulsivity (total commissions) was significantly correlated with parent ratings of hyperactivity/impulsivity (r = .22, p = .006) but not with ratings of inattention (r = .13, p = .09). However, the strength of the significant correlations was weak overall. These findings are concordant with findings in other populations that ratings and test performance are related but do not seem to measure the same constructs.

Discussion

In this study, we investigated some unresolved issues in the attention profile of children with SBM. We studied the relation between sustained attention over time and speed and accuracy of task performance; the relation between performance on neuropsychological tests and parent ratings of attention, the delineation of medical variables within SBM groups, and the contribution of hydrocephalus to the attention profile identified in SBM.

Relative to controls and children in the AS comparison group, children with SBM were not significantly different in their ability to sustain attention over time. They showed consistent performance across the duration of the task, with no significant changes in errors or reaction time between blocks. Although there may be alternative explanations for the finding that performance does not vary significantly over time or across groups, the current results, when taken with previous findings on the VODT (Brewer et al., 2001), suggest that inability to sustain performance over time may be unique to children with primary ADHD. The current study did not employ an ADHD group with no SBM, and the subsamples of children with significant symptomatology were not large enough to replicate the ADHD finding. Nevertheless, the current results do support the hypothesis that, regardless of ADHD status, assessments of the ability to sustain attention over time do not robustly differentiate children with SBM from children with AS or typically developing controls. Although Loss et al. (1998) cited impaired “sustained” attention in children with SBM compared to sibling controls, their operational definition of sustained attention was number of errors, which, as noted earlier, may be less sensitive indices of sustained attention.

In terms of errors, children with SBM committed more omission errors than both groups and more commission errors than children in the NC group. The present findings were consistent with those of Lollar (1990); Loss et al. (1998), and Brewer et al. (2001) in showing more errors of omission and commission on CPTs in mixed etiologies of spina bifida and AS than the respective comparison groups. The significant difference in RT was also found in Brewer et al.; this variable was not included in other studies. However, the present findings differ from those of Colvin et al. (2003), who found no significant differences between children with SBM and sibling controls on errors of omission or commission. This difference may be due to the fact that Colvin et al.’s control group was composed of siblings, or that the SBM sample was less severe (i.e., they did not include any children with lesions above T12). Our hypothesis that commission error rates would be similar across groups was not supported. In fact, both hydrocephalus groups made more commission errors than the typically developing group.
It is unclear whether error rates (as opposed to performance over trial blocks, or sustained attention), are strong assessments of the anterior attention system. However, most studies using the CPT do find differences on these variables in children with SBM, so to the extent that error rates are associated with the anterior attention system, both the anterior and posterior attention systems are implicated across studies of SBM. This finding is interesting in light of data suggesting that children with SBM generally show more errors on a variety of tasks involving problem solving that also require maintenance of attention, such as the Wisconsin Card Sorting Test and the Tower of London. However, Fletcher et al. (1996) found that error rates were higher on initial components of the task, but improved over time, which may reflect the fact that such tasks involve error correction. It is well-established that children with SBM correct and improve performance when feedback is presented about errors (Colvin et al., 2003; Edelstein et al., 2004). Further, these types of tasks are self-paced, unlike the CPT, which requires regular responding over an extended period of time. That children with SBM improve with error correction on self-paced tasks is not consistent with a response control deficit, the key feature of the anterior attention system. Finally, if stimulus orienting is impaired, which is well documented in SBM (Dennis et al., 2005a, 2005b), it is not surprising that error rates are higher on a variety of tasks because many children with SBM will be less activated because of impairments in the posterior brain attention system.

The data clarify a long-standing question about the relative contributions of hydrocephalus and disorder-specific brain abnormalities in the cognitive performance of children with SBM. Inclusion of the AS group in the present study allowed comparison of children with SBM to those with hydrocephalus but few congenital brain abnormalities. The AS group was indistinguishable from the NC group on variables measuring overall responsivity (omissions, commissions, and reaction time). Compared to children with SBM, children with AS performed significantly better on number of omission errors. For mean RT, although AS–SBM comparisons were not significant, the effect size ($d = .48$ when AS children with significant ADHD symptomatology were excluded) suggests that children with AS did respond faster. For commission errors, performance in the AS and SBM groups was similar. Overall, these results indicate that impaired performance on measures of inattention cannot be attributed solely to hydrocephalus and may reflect the impact of the signature brain dysmorphologies found in children with SBM. In studies of stimulus orienting, these anomalies are clearly related to individual differences within the group with SBM (Dennis et al., 2005a, 2005b).

The second hypothesis was that the presence of medical complications and ADHD symptomatology would predict poorer task performance for children with SBM. In the current sample, the only medical variable that significantly predicted performance was oculomotor abnormalities, which increased RT. Loss et al. (1998) also found that oculomotor abnormalities predicted Vigilance Task and other attention variables. Oculomotor abnormalities also were associated with poorer performance on the posterior attention variables. The sample size was not large enough to tease out effects of etiology that might help evaluate the basis for this association. However, Salmon, Sharpe, Lilikas, Steinbach, and Dennis (2007) found that impaired eye movements were found in children with SBM who had the Chiari II malformation and nystagmus, suggesting that deficits are related to hindbrain malformations rather than the effects of hydrocephalus. Finally, lesion level was not a significant predictor of performance. However, more children with upper than with lower spinal lesion defects have low IQ scores (below 70) and would have been excluded from this sample on the basis of this IQ cut-off (Fletcher et al., 2005).

The presence of significant ADHD symptomatology did not predict task performance, indicating that, at least in children with SBM, variability in cognitive attention task performance is not related to behavioral attention problems. Task performance was only weakly correlated with accompanying parent ratings (omissions with inattention, commissions with hyperactivity/impulsivity). These findings are concordant with findings in other populations that ratings and test performance are related but do not seem to measure the same constructs (Gordon et al., 2006). One limitation of the current study is that the SNAP–IV is a rating scale measure of ADHD symptomatology. It is possible that group composition, and therefore results, would have been different if a standardized clinical interview that applied the full range of ADHD diagnostic criteria had been employed. Given the high percentage of children with SBM who display significant symptomatology, more detailed assessment of clinical attention is warranted.

Children with SBM differed from the typically developing children on IQ and SES. Because these differences are an inherent characteristic of SBM, statistical efforts to control these variables would not permit an evaluation of whether the differences on the CPT are due to inattention or to IQ and SES (Adams et al., 1985; Lord, 1967). The latter is not a likely explanation because SES was not correlated with the Gordon variables. However, it could be argued that the differences in CPT performance were due to the differences in IQ (even if the effect of IQ as a possible covariate had no effect on the CPT differences). A closer examination reveals that IQ is not a likely explanation of the results. First, consider the simple argument that the differences are due to some general intellectual factor that is measured by the IQ test and the CPT. In this scenario, we are saying that children with SBM do not do well on either task because of this general factor. However, consider the causal direction of the argument, which is from IQ to CPT. In fact, just as vascular injury to the left hemisphere causes aphasia and reduces verbal IQ in adults (Hebb, 1949), and few would seriously consider recommending ANCOVA for verbal IQ differences in aphasia, the brain insult associated with SBM reduces IQ and CPT performance. Invoking a general factor based on a cognitive composite like an IQ score to explain a more specific cognitive problem is difficult to justify.

This leads directly to the second scenario, which is how to explain the relation of IQ and CPT in children with SBM. The IQ test measures skills that are not measured by the CPT and represents a broader composite of ability. As such, differences in IQ are likely to be larger than differences in a narrow measure of ability, like attention. If IQ were a general explanation for the attention results, children with SBM (who have a lower than average population IQ) would perform more poorly on all attention measures, and they do not do so. The articulated model of anterior and posterior attention used here includes a number of specific attention measures, but not IQ; therefore, understanding attention is facilitated by a comparison of the attention measures children with
SBM can and cannot perform, not by IQ. For these reasons, we did not attempt to “correct” scores for differences in IQ or SES, and we believe that doing so would have resulted in unrealistic adjusted scores and blurred interpretation of the findings.

In sum, the performance profile of children with SBM on the CPT suggests partial support for the hypothesis of more intact anterior attention systems, especially on CPT variables associated with sustained attention, which is typically moderated by anterior brain structures. However, children with SBM are slower and make more errors. More important, these differences were detected in comparison not only with typically developing children, but also with children with AS, both of whom performed better on most measures. The relative paucity of key attention deficits in children with hydrocephalus but without many of the key brain dysmorphologies of SBM suggests that some of the attention deficits in SBM are related to the brain stigmata of the SBM etiology and are not general effects of hydrocephalus. Our findings are important to the assessment and management of attention skills in children with SBM, because they further delineate a unique pattern of expected strengths and weaknesses. These findings also point to future research directions, as they indicate that various etiologies of hydrocephalus may have quite distinct effects on key neuropsychological functions.

References


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