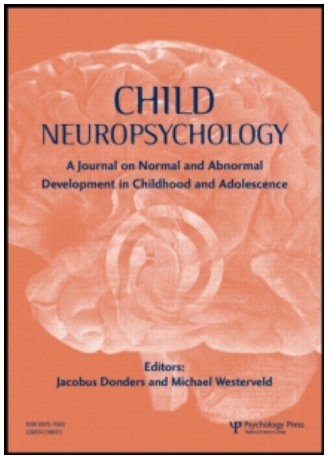

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FACTORS OF BIOLOGICAL RISK AND RESERVE ASSOCIATED WITH EXECUTIVE BEHAVIORS IN CHILDREN AND ADOLESCENTS WITH SPINA BIFIDA MYELOMENINGOCELE

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This study examined differences between healthy children (n=35) and those with spina bifida myelomeningocele (SBM; n=42) on the Behavior Rating Inventory of Executive Function (BRIEF), a measure of executive function behaviors. It also examined whether aspects of biological risk associated with SBM and reserve factors within the family could account for variability in BRIEF scores for children and adolescents with SBM. Patients in the SBM group exhibited more problems than both published norms and a local comparison group of healthy children in metacognition but not behavior regulation. Behavior regulation problems in children with SBM were predicted by parent psychological distress. More shunt-related surgeries and history of seizures predicted poorer metacognitive abilities.

INTRODUCTION

Contemporary theories for understanding the complexities and variability in neurobehavioral outcome emphasize a balance between factors of biological risk and reserve. Satz (1993) introduced the concept of *brain reserve capacity* to explain individual differences in the risk associated with various acquired neurologic insults. Reserve in this sense represents neurophysiological mechanisms inherent in the central nervous system that protect an individual from the disease process, thereby increasing the time before the emergence of clinical symptoms. Building upon Satz's adult-focused model, Dennis (2000) proposed a general structure for predicting neurobehavioral outcomes in children with medical disorders affecting the central nervous system. In her model, a major contributor to neurobehavioral outcome is biological risk, or the cumulative severity of an

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injury or disease process. This may include genetic and environmental effects, as well as any secondary effects of an acquired brain insult and/or treatment of the disorder. In addition, she proposed that the relationship between biological risk and neurobehavioral outcome is moderated by internal and external reserve, defined as factors within the child, family, or social environment that buffer the dysfunction caused by a child's medical condition. Examples of reserve factors include the preinsult status of the child (e.g., gender, IQ), physical and mental health of the child (e.g., difficulties in growth or sleep), school services, rehabilitation resources, and family factors such as parental stress and socioeconomic status (SES). The concepts of risk and reserve may be particularly informative when trying to understand the varied neurobehavioral outcomes of children with complex neurodevelopmental disorders. This paper documents the presence and wide variability of deficits in the executive functioning among children with one such disorder, spina bifida myelomeningocele (SBM), then attempts to predict such deficits with measures of biological risk and reserve.

Spina bifida is a congenital abnormality that appears at birth as an open cyst or mass on the spine. The lesion is caused by a failed closure of one or more vertebrae during the early weeks of gestation. Children with spina bifida myelomeningocele (SBM), the most severe form of spina bifida, often have a multitude of medical problems associated with the disorder, such as urinary, orthopedic, and neurological difficulties. As many as 80–90% of children with SBM also develop hydrocephalus and require a shunt (Reigel & Rotenstein, 1994). Children with SBM are known to have a number of brain abnormalities involving both the gray and white matter (Fletcher, Dennis, & Northup, 2000; Fletcher et al., 2005) including Chiari malformation, elongation of the pons and medulla, agenesis of the corpus callosum, aqueduct abnormalities, and tectal beaking.

As a group, children with spina bifida tend to have IQs in the Low Average to Average ranges, slightly but significantly below those of healthy children (Wills, Holmbeck, Dillon, & McLone, 1990). Despite relatively preserved general intellectual functioning, this population is at risk for impaired cognitive development in such areas as visual-motor integration, pragmatic language, and arithmetic achievement (Barnes et al., 2006; Fletcher et al., 2004; Fletcher et al., 1995; Wills, 1993), as well as elevated rates of internalizing and externalizing behavior problems including increased incidence of attention deficit/hyperactivity disorder (ADHD; Ammerman, et al., 1998; Burmeister et al., 2005; Campbell, Hayden, & Davenport, 1977; Lavigne & Faier-Routman, 1992; Thompson, Kronenberger, Johnson, & Whiting, 1989; Wallander, Varni, Babani, Banis, & Wilcox, 1988). Of note, children with spina bifida are rarely rated as hyperactive, likely as a result of limitations secondary to motor impairments and restricted mobility. In addition, these children appear to exhibit attention deficits that differ from those observed in children with ADHD including deficits on convert and overt attention-orienting tasks that occur independent of motor responses (Burmeister et al., 2005; Dennis, Landry, Barnes, & Fletcher, 2006).

Several studies have utilized traditional, clinic-based tests to document problems with executive functioning in this population, including deficits in higher order planning, mental flexibility, focused attention, ability to shift attention, working memory, and problem-solving skills (Brewer, Fletcher, Hiscock, & Davidson, 2001; Fletcher et al., 1993; Holmbeck et al., 2003; Landry, Robinson, Copeland, & Garner, 1993; Loss, Yeates, & Enrile, 1998; Snow, 1999; Yeates, Enrile, Loss, Blumenstein, & Delis, 1995). However, factors associated with standardized testing in a traditional lab setting may restrict the ability to adequately measure executive functioning, prompting the development of a purportedly more ecologically valid measure (Gioia, Isquith, Guy, & Kenworthy, 2000). One

such measure, the Behavior Rating Inventory of Executive Function (BRIEF; Gioia et al., 2000) is a questionnaire for parents and teachers used to assess the behavioral manifestations of executive functioning in home and school settings of children. Using the BRIEF, Mahone, Zabel, Levey, Verda, & Kinsman (2002) found that parents consistently rated their adolescents with SBM as having worse metacognition (e.g., planning, working memory, organization) than behavioral regulation (e.g., impulse control, emotion regulation), extending prior clinic-based findings.

Biological Risk Factors in SBM

Physical symptoms and brain dysmorphologies associated with SBM have been related to outcome in various neurocognitive domains, consistent with the concept of biological risk in Dennis's (2000) heuristic. Several investigators have documented greater overall cognitive impairment in those patients with higher lesion levels (i.e., lesions affecting the spine more rostrally) than in those with lower lesions (e.g., Bier, Moralse, Liebling, Geddes, & Kim, 1997; Donders, Rourke, & Canady, 1991; Fletcher et al., 2005). The presence of seizures (see Wills, 1993) and central nervous system infection (Donders, Canady, & Rourke, 1990) has also been linked to poorer intellectual outcome in children with SBM. Other factors such as the presence of a shunt (Tew & Laurence, 1975; Yeates et al., 1995) have been examined as sources of variability in neurocognitive outcome. In younger children with shunts, there is limited evidence that the number of shunt revisions affects cognitive or behavioral functioning; however, there is some evidence to suggest that the number of shunt revisions is related to specific cognitive functions such as numeracy in young adults with spina bifida and hydrocephalus (Dennis & Barnes, 2002).

Some findings from previous research on biological risk factors as they relate to neurocognitive outcome have had specific relevance to the domains of attention and executive functioning.Loss et al. (1998) reported that other medical factors such as high lesion level, frequent shunt revisions, a history of central nervous system infection, oculomotor abnormalities, and agenesis of the corpus callosum significantly predicted differences in performance on clinic-based measures of attention and executive function measures after controlling for shunt status. More recently, Fletcher et al. (2005) conducted a qualitative and quantitative volumetric MRI study examining relationships between spinal lesion level, variations in development various brain structures, and neurobehavioral outcomes in children with SBM. They found that higher lesion levels were associated with decreased cerebral and cerebellar volume and specific abnormalities in several posterior brain regions (including the midbrain, tectum, pons, and splenium). Further, they reported that spinal lesion was predictive of attention disorders in their sample of non-Hispanic children. Specifically, children with upper-level lesions were less often rated as hyperactive or impulsive. The authors appropriately concluded that these findings were likely due to the decreased ambulation of the children with upper-level spinal lesions; however, other researchers have also found impairments in basic attentional processes unrelated to motor functioning (e.g., covert orienting, Dennis et al., 2005a; and inhibition of return, Dennis et al., 2005b) thought to be subserved by more posterior brain regions, as those seen in children with SBM. Taken together, prior research suggests that numerous biologic factors may indeed be associated with cognitive outcome in children with SBM and, more specifically, compromised attention and executive functioning.

Reserve Factors in SBM

Certain commonly used measures of reserve in the adult research literature, such as premorbid IQ and other "pre-insult" factors, are not appropriate or accessible in children with congenital central nervous system disorders such as SBM (Dennis, 2000). Instead, more indirect measures of reserve must be considered, such as family functioning, parental characteristics (e.g., parent mental health), and SES. Prior research suggests that low SES has significant predictive utility when examining behavioral dysfunction and verbal cognitive abilities in children with SBM (Bier et al., 1997; Fletcher et al., 2005; Fletcher et al.; 2004; Holmbeck et al., 2003). Children with spina bifida from low SES backgrounds, especially those who are shunted, also demonstrate lower levels of scholastic competence, obtain lower grades in school and score lower than their typically developing peers on teacher-reported measures of academic functioning (King, Shultz, Steel, Gilpin, & Cathers, 1993; Holmbeck & Faier-Routman, 1995). Further, SES has been observed to significantly account for individual performance differences on several clinic-based tasks of attention/executive functioning such as encoding, focusing, and sustaining attention in children with SBM (Loss et al., 1998).

The demands associated with a chronic illness such as SBM have been found to increase levels of parent and family stress in comparison to parents of typically developing children (Carr, 1991; Holmbeck, 1997; Wallander et al., 1989). Overall, both mothers and fathers of children with SBM appear to be more vulnerable to maladjustment and stress when compared to parents of children with various other medical conditions (Barakat & Linney, 1995; Wallander et al., 1989;) or typically developing children (Holmbeck, 1997). When considered in combination with research that documents poor behavioral and cognitive outcome for children of parents with identified psychopathology (e.g., Hay, McStephen, & Levy, 2001), these findings suggest a need to examine relationships between parent distress and mental health on cognitive and behavioral outcome in children with SBM.

In summary, prior research has examined various aspects of executive dysfunction in children with SBM, including whether various aspects of risk and reserve may account for the substantial variability in outcome that has been observed within this population. However, nearly all research on executive dysfunction in this population has relied upon clinic-based tests, despite limitations in how well these findings can be generalized to children's "real world" functioning. Further, the one prior study to have used a measure designed to maximize real-world relevance (Mahone, Zabel, Levey, Verda, & Kinsman, 2002) did not examine the factors that might influence outcome in executive functioning. Finally, few authors have used a theory-guided approach, such as that provided by Dennis (2002), to guide their research into factors influencing the neurobehavioral outcome of children with SBM. The current project was designed to investigate the impact of various aspects of biological risk and reserve on parent-completed BRIEF profiles in an attempt to analyze sources of potential variability in executive functioning in children and adolescents with SBM. The *first hypothesis* was that children with SBM would display greater metacognitive than behavioral regulation deficits compared to published norms and a healthy comparison group. The *second hypothesis* was that biological and medical factors associated with SBM would contribute significantly to a statistical model of executive behavior outcome, and reserve variables would further contribute to the prediction of BRIEF scores after accounting for biological risk. Finally, consistent with Dennis's (2000) proposed algorithm, the *third hypothesis* was that measures representing reserve

(e.g., SES and parent distress) would significantly moderate the relationship between biological risk and neurobehavioral outcome.

METHOD

Participants

Participants in the study were 35 children and adolescents with SBM and 42 healthy controls originally recruited for a longitudinal study examining the psychosocial functioning of children and adolescents with SBM (see also Ris et al., 2007). Inclusion criteria for both groups were 1) age between 10 and 17 at the time of the initial evaluation and 2) Verbal or Performance IQ ≥ 70 from the Wechsler Intelligence Scale for Children, Third Edition (WISC-III; Wechsler, 1991) or Wechsler Adult Intelligence Scale, Third Edition (WAIS-III; Wechsler, 1997). Children with lipomeningocele or sacral meningocele were excluded.

Children with SBM were recruited through the Spina Bifida Clinic at Cincinnati Children's Hospital Medical Center, Cincinnati, Ohio and The Children's Hospital, Columbus, Ohio. Participants in the nonclinical comparison group were recruited through area pediatric practices and general advertisements and matched to the patients on SES. BRIEF profiles were examined for validity evidenced by an Inconsistency score ≥ 7 or a Negativity score ≥ 5. Six participants with SBM and one participant from the comparison group were omitted from final analyses because they had invalid BRIEF profiles secondary to either an elevated Inconsistency score ($n=4$) or Negativity score ($n=3$).[1] In addition, one child originally recruited into the comparison group was excluded from analyses for extreme values on the BRIEF; scores for the Global Composite Index and Behavioral Regulation Index were greater than the 99th percentile, and the Metacognition Index scores was greater than the 88th percentile when compared to published norms.

Demographic and IQ information for the SBM and comparison groups are provided in Table 1.

Procedure

In the context of a larger neuropsychological evaluation (not detailed in the current report), parents completed questionnaire measures regarding general demographic and medical background, parental psychopathology/distress, and child executive functioning. Analyses were based upon the report of primary caregivers (mothers in all but two cases, both in the SBM group, in which the father was identified as primary caregiver). To supplement this parent-report information, the medical records of each child in the SBM group were carefully reviewed to assist in coding measures of biological risk.

[1]Multivariate analysis of variance was conducted to examine the potential effect of excluding these cases from analyses. Participants were divided into two groups representing 1) valid profiles ($n=76$) and 2) invalid profiles ($n=7$). Results indicated that parents who completed invalid profiles reported significantly more severe symptoms in their children ($F=4.37$, df=3, $p < .05$). Follow-up comparisons indicated that the groups differed significantly on the Behavioral Regulation Index ($F=9.80$, df=1, $p < .01$), the Metacognition Index ($F=11.16$, df=1, $p < .01$), and Global Executive Composite ($F=12.70$, df=1, $p < .01$) from the BRIEF.

Table 1 Characteristics of the Clinical Sample and Comparison Group.

	SBM	Comparison
N	35	42
Males/Females (*n*)	15/20	19/23
Age (years), *M* (*SD*)*	13.00 (2.56)	11.81(1.98)
Handedness		
Right/Left	32/3	34/8
Race		
Caucasian/African American	32/3	41/1
VIQ, *M* (*SD*)**	95.26 (16.21)	110.69 (13.28)
PIQ, *M* (*SD*)**	81.97 (13.41)	106.79 (12.07)
FSIQ, *M* (*SD*)**	87.97 (14.37)	109.60 (12.23)
CPT		
Number of Omissions[b]	9.41 (9.20)	9.62 (11.61)
Number of Commissions[b]	19.41 (8.79)	19.26 (7.41)
Children's Category Test[c]**	42.43 (9.08)	52.90 (9.70)
SES[a], *M* (*SD*)	47.24 (10.60)	50.86 (9.17)

[a]Hollingshead Four Factor Index; [b]Raw scores; [c]*T*-score; PIQ = Performance IQ; VIQ = Verbal IQ.
*$p < .05$, **$p < .001$.

Measures

Biological Risk. Based on a review of the literature, eight biological risk variables were selected for inclusion in the statistical analyses:level of functional spinal lesion (thoracic, lumbar, sacral), presence of seizures (yes/no), history of central nervous system infection (yes/no), number of shunt revisions, age in days when first shunt was placed, number of oculomotor deficits present, presence of Arnold-Chiari II malformation (yes/no), and handedness (left/right). Information regarding the factors related to biological risk for each of the participants with SBM was obtained from background questionnaires and chart review.

Reserve. The construct of reserve was represented in this study by measures of socioeconomic status (SES) and parental mental health/distress. SES was calculated using the Hollingshead Four Factor Index of Socioeconomic Status (Hollingshead, 1975), based on parental education and occupation information obtained from the baseline questionnaire. SES was calculated for each participant's mother and father. The highest score for the family was used for all analyses.

Each participant's primary caregiver completed the Symptom Checklist-90-R (SCL-90-R; Derogatis, 1983), a self-report instrument designed to reflect the psychological symptom patterns of psychiatric and medical patients. Consistent with contemporary research use of the SCL-90-R, the composite Global Severity Index (GSI) assessed overall parental mental health or distress. The GSI has a mean of 50 and standard deviation of 10, and higher scores reflect greater symptom severity. A summary of biological risk and reserve information for the SBM group is provided in Table 2.

Executive Functioning. Executive functioning, as evidenced in daily life, was measured with the parent-report BRIEF (Gioia et al., 2000). The BRIEF is comprised of eight clinical scales (Inhibit, Shift, Emotional Control, Initiate, Working Memory, Plan/Organize, Organization of Materials, Monitor) and two validity scales (Inconsistency and

Table 2 Factors of Biological Risk and Reserve for the SBM Sample (n=35).

Biological Risk parameters	*n*	Mean	*SD*	Range
Lesion Level				
Thoracic	5	–	–	–
Lumbar	23	–	–	–
Sacral	7	–	–	–
Presence of a Seizure Disorder	31	–	–	–
Presence of a Shunt	31	–	–	–
History of Shunt Infection	3	–	–	–
Arnold Chiari II Malformation	18	–	–	–
Number of Shunt Revisions	31[a]	1.31	1.78	0–9
Age at 1st shunt (days)	31[a]	9.37	10.29	0–42
Oculomotor deficits	35[a]	.89	1.05	0–4
Measures of Reserve				
Global Severity Index/SCL-90-R[b]	35	48.49	9.28	30–64
SES[c]	34	47.24	10.60	27–66

[a]Number of subjects with SBM on which data were available for this variable; [b]Symptom Checklist 90-R; [c]Hollingshead Four Factor Index.

Negativity). Three composite indexes are derived from the eight clinical scales: the Global Executive Composite, Behavioral Regulation Index, and Metacognition Index. The BRIEF has gathered solid support for its validity, as evidenced in several studies including a special issue of *Child Neuropsychology* (2002, 4). In addition, however, two clinic-based tests of executive function/attention, Conners's Continuous Performance Test (CPT; Conners, 2000) and the Children's Category Test (Boll, 1993), were included in preliminary analyses of the current study to add further support for construct validity of the BRIEF given that it is a relatively new measure.

Statistical Analyses. Preliminary *t*-tests were conducted to compare demographic and test variables in the SBM and nonclinical comparison groups (see Table 1). Age and Full Scale IQ differed significantly between the groups and were consequently used as covariates in analyses examining differences between the SBM and the comparison group. Significant group differences were also observed for the Children's Category Test (Boll, 1993), with the SBM group performing more poorly than the nonclinical comparison group ($p < .01$). Pearson product-moment correlations, calculated for the entire sample to examine the relationship between BRIEF Index scores and other clinic-based measures of executive functioning/attention, indicated a significant relationship between BRIEF MI and the Children's Category Test total *T*-score ($r = -.23$, $p < .05$). No relationship between BRIEF Indexes and the CPT were noted. In addition, because preliminary analyses suggested that age at first shunt and number of shunt revisions were both markedly skewed, these underwent a square root transformation prior to entry into main analyses.

The *first hypothesis* was tested with several sets of analyses. First, mean scores for each of the BRIEF Indexes (Behavioral Regulation Index [BRI] and Metacognition Index [MI]) were compared to published normative data by means of single-sample *z*-tests. Second, multivariate analysis of covariance (MANCOVA; adjusted for age and IQ) was conducted to examine differences between the SBM and nonclinical comparison groups on these indexes. In both of these sets of analyses, when significant group differences emerged on a composite index, pairwise comparisons (with a Bonferroni correction for multiple tests) were calculated to

examine differences on the individual subscales in that composite. Next, a series of chi-square analyses compared the rate of children who scored above the 95th percentile (according to published norms) across the SBM and comparison groups. Odds ratios were also reported as measures of the strength of associations between group membership and clinically elevated scores on the BRIEF MI and BRI Indexes. Finally, paired *t*-tests were also used to examine within-group differences for the SBM group on scaled scores for the MI and BRI.

Hypothesis two was tested via a combination of theoretically driven hierarchical and stepwise multiple regression procedures to examine the contributions of risk and reserve variables to executive functioning within the sample of children with SBM. Separate regression equations were developed to predict each BRIEF composite (MI and BRI). Given the large number of risk and resilience variables and relatively small sample size, each regression equation was developed in several steps. In the first step, age was entered to control for developmental differences in the SBM group beyond that accounted for by standardization. Stepwise regression was employed in the second step to statistically determine which factors of biological risk (level of lesion, presence of seizures, presence of central nervous system infection, number of shunt revisions, age of first shunt, presence of oculomotor deficits, presence of Arnold Chiari II, handedness) best improved prediction of BRIEF composite scores beyond age. Parent GSI from the SCL-90-R and SES were entered in a stepwise procedure in the third step to determine if the addition of reserve variables improved prediction beyond that accounted for by age and biological risk.

To test *hypothesis three*—that there would be a moderating effect of reserve variables (e.g., SES and parent psychological distress) on the relationship between biological risk and BRIEF scores—additional multiple regression analyses were performed to examine the unique contribution of interactions calculated among the child's age, biological risk, and each reserve factor to the overall regression model. It is recognized that examining the significance of interactions through multiple regression is problematic given the large number of biological risk factors being evaluated and the relatively small sample size. Therefore, the interaction effect was computed using a composite rating of overall "biological risk" that was based on a system previously developed by Hommeyer, Holmbeck, Wills, and Coers (1999). The biological risk composite calculated for the present study included the following parameters: 1) lesion level (1=sacral, 2=lumbar, 3=thoracic), 2) number of shunt revisions, 3) presence of seizures (0=no, 1=yes), 4) presence of shunt infection (0=no, 1=yes), 5) presence of Arnold-Chiari II malformation (0=no, 1=yes), and 6) number of oculomotor deficits recorded from the child's medical chart. The biological risk severity score ranged from 2 to 15 (M=4.97, SD=2.43) with higher scores indicative of greater risk. A combination of hierarchical and stepwise regression was utilized in which all main effects were entered in the first step (i.e., age, biological risk, and parent GSI). Two-way interactions among the variables were entered on the second step (i.e., age × biological risk, parent GSI × biological risk, age × parent GSI) and subjected to stepwise entry. A total of four hierarchical linear regression analyses were conducted each looking at the moderating influence of either SES or parent psychological distress on the two BRIEF Indexes.

RESULTS

Hypothesis 1: SBM Scores vs. Norms and the Comparison Group

Table 3 summarizes the BRIEF Index and subscale standardized scores for the SBM and comparison groups. None of the mean subscale or index standard scores reached

Table 3 Means and Standard Deviations for all BRIEF Scales and Indexes.

BRIEF parameters	SBM *M* (*SD*)	Comparison *M* (*SD*)
Global Executive Composite	58.46 (8.21)	48.71 (8.94)
Behavioral Regulation	51.77 (9.66)	47.69 (8.07)
Inhibit	48.80 (9.06)	47.45 (7.38)
Shift	54.17 (9.49)	48.40 (9.88)
Emotional Control	53.46 (13.28)	48.74 (7.97)
Metacognition**	61.51 (9.07)	49.50 (9.96)
Initiation**	62.49 (11.07)	49.12 (9.05)
Working Memory**	63.71 (11.29)	48.98 (9.77)
Plan/Organize**	62.11 (11.23)	49.86 (10.03)
Organization of Materials*	55.83 (9.01)	50.00 (10.05)
Monitor	54.11 (7.66)	49.88 (9.58)

**$p < .001$, *$p < .01$; Single Sample Z-test for SBM group.

clinical significance as described in the BRIEF manual ($T > 65$; Gioia et al., 2000). However, the SBM group average on the MI was statistically different from the normative mean of 50, $z = 6.81$, $p < .001$ (2-tailed), though the mean BRI score was not, $z = 1.05$, $p > .10$. Follow-up analyses indicated that the following subscales in the SBM group had mean scores that differed from norms at $p < .01$: Initiation, Working Memory, Plan/Organize, and Organization of Materials.

MANCOVA revealed a significant group main effect on the MI, $F(1, 73) = 10.15$, $p < .01$ ($\eta_p^2 = .12$), with the SBM sample scoring worse on average than the comparison group. No significant group differences were observed for BRI, $F(1, 73) = 2.05$, *ns*. Follow-up analyses within the MI domain indicated that the SBM group fared worse than the comparison group on Initiate, $F(1, 73) = 11.97$, $p < .01$ ($\eta_p^2 = .14$), Working Memory, $F(1, 73) = 11.84$, $p < .01$ ($\eta_p^2 = .14$), Plan/Organize, $F(1, 73) = 7.38$, $p < .01$ ($\eta_p^2 = .09$), and Organization of Materials, $F(1, 73) = 5.45$, $p < .01$ ($\eta_p^2 = .07$). Group difference on the Monitor scale was not significant, $F(1, 73) = 1.72$, *ns*. Similar findings emerged in exploratory analyses when only Full Scale IQ (FSIQ) (not age) was entered as a covariate, though a small group main effect was observed on the BRI when only age (not FSIQ) was entered as a covariate, $F(1, 74) = 5.64$, $p < .05$.[2]

Results of chi-square tests indicated that the rate of abnormal scores (i.e., scores above the 95th percentile according to published norms) was greater in the SBM group than the comparison group on the MI, chi-square = 5.53, $p < .05$ (odds ratio 5.93, 95% C.I. 1.17–30.08), but not on the BRI, chi-square = 0.57, $p > .10$ (odds ratio 2.49, 95% C.I. 0.22–28.62). Follow-up analyses further indicated that individuals in the SBM group more often had abnormal scores on the following subscales, $p < .01$: Initiate, Working Memory, and Plan/Organize.

Finally, within the SBM group, scores for MI were significantly higher than for BRI, $t(34) = -5.42$, $p < .01$. In contrast, the MI and BRI scales were quite comparable within the comparison group. In summary, findings supported hypothesis one.

[2]Two-way interactions were calculated in order to test for group differences in the effects of age and IQ on BRIEF Index scores. No significant interactions were found for BRIEF BRI (group × age: $F = 2.57$, $p = .11$; group × FSIQ: $F = 0.94$, $p = .34$) or BRIEF MI (group × age: $F = 2.30$, $p = .13$; group × FSIQ: $F = 1.08$, $p = .30$).

Hypothesis 2: Evaluation of Biological Risk and Reserve

The simple correlations among all the variables used in the regression analyses are summarized in Table 4. A summary of the hierarchical linear regression analyses for the prediction of BRIEF Indexes including standardized regression coefficients (β), amount of variance accounted for by each step in the analyses (R^2), adjusted R^2, and changes observed in R^2 and F is displayed in Table 5. For the equation predicting BRI, the overall R was significantly different from zero, $F(2, 32) = 8.25$, $p = .001$, with 34% (30% adjusted) of the variability in BRI scores predicted by child age and parent GSI. Beyond age, only GSI added significantly to the prediction of BRI scores, R^2 Change = .22, $p < .01$. None of the biological risk measures nor SES made significant contributions to the model beyond

Table 4 Simple Correlations Among Variables of Biological Risk, Reserve, and BRIEF Clinical Indexes for SBM Group.

	Metacognition[a]	Behavioral Regulation[a]
Age	–.30	–.35*
FSIQ	–.13	.02
Lesion Level	.09[c]	.31[c]
History of Seizures	.38[c,]*	.04[c]
History of Shunt Infection	.28[c]	.19[c]
Age at 1st Shunt[b]	–.04	–.08
Number of Shunt Revisions[b]	.26	.01
Oculomotor Deficits	–.24	–.17
Arnold Chiari II	.02[c]	–.01[c]
Dominant Hand	.12[c]	–.02[c]
Parent GSI	.19	.54**
SES	–.05	–.00

*$p < .05$; **$p < .01$.

[a]Standard Scores ($M = 50$); [b]Square root transformation; [c]Spearman's Rho calculated and reported; GSI = Global Severity Index, SCL-90-R; SES = Hollingshead Four-Factor Index.

Table 5 Hierarchical and Stepwise Regression of Biological Risk and Reserve Variables on BRIEF Indexes: Summary of Steps and Unique Variance.

Step	Variables Entered	β	R^2	R^2 Change	F Change (df1, df2)	F
Predicting the Behavior Regulation Index						
I	Age	–.35*	.122	.122	4.58* (1, 33)	
(II)	(None Significant)	(no biological risk variables contributed to the equation beyond age, so no predictors were entered in Step II)				
III	GSI	.48**	.340	.218	10.59** (1, 32)	8.25**
Predicting the Metacognition Index						
I	Age	–.30	.089	.089	3.22 (1, 33)	
II	Shunt Revisions	.36*	.210	.121	4.90* (1, 32)	4.25*
	Presence of Seizures	.34*	.323	.113	5.17* (1, 31)	4.93**
III	None significant	(no reserve variables contributed to the equation beyond age, so no predictors were entered in Step III)				

*$p < .05$, **$p < .01$.

Note: In each prediction equation, age was entered in step 1, the biological risk factors were entered in stepwise fashion in step 2, and the reserve factors were entered in stepwise fashion in step 3.

age, nor were any of these variables associated with BRI in zero-order correlations (Table 4). For the regression predicting MI, the overall R was significantly different from zero, F (3, 31)=4.925, $p < .01$, with 32% (26% adjusted) of the variability in MI scores predicted by age, (square root transformed) number of shunt revisions, and history of seizures. Beyond age, only number of shunt revisions and history of seizures significantly added to the prediction model, R^2 change=.12, .11, respectively, $p < .05$. None of the other risk factors nor either of the reserve factors contributed further to the regression model, nor did they display significant zero-order correlations with MI (Table 4).

In summary, findings were mixed with respect to hypothesis two; although there was evidence that some aspects of medical risk were predictive of metacognitive difficulties and some aspects of reserve were predictive of behavior regulation difficulties, a more complete model including both risk and reserve was not supported in the present data.

Hypothesis 3: Moderating Effects of Reserve Variables

None of the two-way or three-way interactions calculated among child's age, variables of reserve (SES and parent psychological distress), and the biological risk composite significantly added to the variance accounted for in BRIEF MI or BRI beyond that accounted for by the main effects. In the prediction model for MI, the interaction between biological composite and SES approached significance (p=.06). However, all other significance values for two-way interactions ranged from $p > .20$ to $p > .90$.

DISCUSSION

This study was designed to investigate factors of biological risk and reserve on executive functioning in a sample of children and adolescents with spina bifida myelomeningocele. Initial analyses were largely confirmatory and set out to examine the BRIEF profile that emerged for young patients with SBM. Mean scores obtained on each of the BRIEF subscales (e.g., Initiate, Working Memory) did not reach clinical significance for the SBM group as a whole. However, as predicted, the mean scores were significantly worse than published norms and those of a comparison group on subtests measuring several metacognitive skills. Specifically, parents of children with SBM reported concerns about their children's abilities to 1) work independently and initiate tasks, 2) retain and use information in working memory, 3) plan for future tasks or goals, and 4) maintain an organized approach to their activities and materials. Patients with SBM did not differ from the normative or comparison groups in their abilities to self-monitor, inhibit, or regulate their behavior. Indeed, within the SBM group, parent concerns about metacognition were significantly greater than their concerns about behavior regulation. These differences in metacognition between SBM and healthy controls were not attributable to differences between the groups in age or general intellectual level.

These findings are consistent with previous research examining the BRIEF in children and adolescents with SBM (Mahone et al., 2002) as well as other studies in this population examining attention and working memory, constructs often considered to be overlapping with executive function. Specifically, other investigators have found increased incidence of attention deficit/hyperactivity disorder in children with SBM compared to the general population, with a majority of those children presenting with symptoms consistent with primarily inattentive subtype indicating greater concerns with inattention/distractibility, planning, and initiation than behavioral regulation or impulsivity (Ammerman, et al.,

1998; Burmeister et al., 2005). Second, the parent-reported concerns of working memory problems observed in this sample are consistent with other studies that have found working memory deficits on clinic-based measures (e.g., Children's Paced Auditory Serial Addition Task, CHIPASAT) in children with SBM compared to nonclinical comparison groups (Boyer et al., 2006). Further, these aspects of attention/executive function are highly related to deficits in attention orienting, which have been put forward as being among the core cognitive deficits in individuals with SBM (Dennis et al., 2006).

Attention and working memory, as two aspects of the broader construct of executive functioning, have been thoroughly examined in the literature on children with SBM; however, less is known about other aspects of executive functioning such as planning and organization. Specifically, some studies examining the performance of children with SBM on clinic-based measures of executive functioning have concluded that central deficits in processing speed, for example, accounted for group differences in executive functioning (Fletcher et al., 1996). Based on these observations, it has been hypothesized that executive dysfunction in individuals with SBM may be better classified as posterior attention system problems consistent with the nature of congenital brain malformations associated with SBM (Burmeister et al., 2005). The current findings, however, highlight potential difficulties in planning and organization, through assessment with the BRIEF that may not be captured by traditional, cognitive measures of executive functioning and warrant further investigation.

A statistical model for the prediction of executive function outcome set by biological risk factors and moderated by variables of family reserve was not supported by the current analyses; however, specific variables of risk and reserve were each directly associated with aspects of executive functioning measured by the BRIEF. No measured medical or biological risk factor significantly contributed to a prediction model of BRIEF Behavioral Regulation Index. In fact, behavior regulation problems in children and adolescents with SBM were best predicted by parent self-report of general psychological distress and child age, variables of environmental/family reserve. Parents of younger children, and those who endorsed higher levels of distress reported more concerns of behavioral dysregulation and inhibitory control in their children. Interestingly, the age effect was observed even for age-normed BRI standard scores, suggesting developmental variation unique to children and adolescents with SBM that is not observed within the normative population. In other words, although most children show improved behavior regulation with age (e.g., Gioia et al., 2000), children with SBM may show a more dramatic developmental shift in behavior regulation skills. Given the lack of previous evidence to suggest that children with spina bifida exhibit such a differing developmental pattern with regard to behavioral regulation, it is possible that this latter observation is an artifact specific to the sample of children with SBM examined in this study rather than a finding that can be generalized to a broader population of children and adolescents with spina bifida.

Similar findings regarding parent distress and executive functioning have been observed in a sample of children with traumatic brain injuries, in which parent ratings on the BRIEF were found to predict parental psychological distress, perceived family burden, and general family functioning (Mangeot, Armstrong, Colvin, Yeates, & Taylor, 2002). The researchers concluded that the behavioral manifestations of deficits in executive functioning in children were burdensome and distressing for parents and families. However, other causal relationships are possible. For example, Taylor et al. (2001) proposed that children and parents affect each other in bidirectional fashion. This leaves open the possibility of multiple points of clinical intervention, with improvements in behavior regulation

having potential effects on parent functioning, and reductions in parental distress having potential effects on child behaviors. Importantly, however, present findings may not be unique to SBM, and indeed behavior regulation problems in this group were not reported to occur significantly more often than is typical in the general population.

Metacognition appears to be a much more salient area of deficit for children with SBM, but little is known about the predictors of metacognitive deficits in this population. In the present study, neither of the two reserve variables (parent psychological distress or SES) was associated with metacognitive skills on the BRIEF. However, two biological risk variables, number of shunt-related surgeries and history of seizures, emerged as significant predictors of poorer metacognitive abilities. Patients who underwent more surgeries to revise their shunts and those with seizures had more parent-reported problems with planning, organization, and problem-solving abilities. Dennis and colleagues (2006) have expounded upon her algorithm and recently proposed a detailed model of neurocognitive function in spina bifida. Specifically, they discuss the moderating role of secondary central nervous system insults in understanding variability in cognitive and behavioral outcome. Increased number of shunt-related surgeries and seizures could be considered examples of such secondary insults; however, there is limited evidence that either variable is related to specific cognitive or behavioral outcomes in children. Of note, several studies have documented negative effects of shunt revision surgeries in young adults in areas such as motor speech (Huber-Okrainec, Dennis, Brettschneider, & Spiegler, 2002), numeracy (Dennis & Barnes, 2002), and independent life skills and quality of life (Hetherington et al., 2006; Hunt, Oakeshott, & Kerry, 1999).

Further research is needed to better understand the degree to which these risk factors reflect primary disease factors (e.g., underlying disease severity) or more readily modified treatment factors (e.g., surgical approach to shunt revision, antiepileptic medications). For example, it is possible that these factors reflect greater neurologic compromise in children with SBM; however, this hypothesis needs much more investigation. In addition, future research should examine these factors as potential moderators as Dennis and colleagues (2006) suggest.

From an immediate and practical standpoint, clinicians who are working with children with SBM should be alert not only to the potential presence of metacognitive difficulties but also to the increased risk for such difficulties in children who have experienced multiple shunt revisions or who have seizure disorders. A number of classroom accommodations and interventions have been described for supporting children with metacognitive problems (e.g., Dawson & Guare, 2004), and the availability of a clear link between a child's neurological history and their neurocognitive status can justify the use of such strategies.

Several limitations to this study warrant mention. First, the same person, the primary caretaker, completed all parent-report measures, which may introduce problems with shared-method variance. As a result, the associations between ratings on the BRIEF and parent psychological distress may have been inflated by reliance on a single informant. That this did not universally occur across all BRIEF indexes argues against a simple method variance effect; nevertheless, future researchers are encouraged to include multiple informants (e.g., teachers) in their data collection. Second, findings based on comparisons between the SBM and the nonclinical comparison groups on the BRIEF were limited by the possibility of sampling biases given the significant differences between the groups on age and FSIQ and should be interpreted in this context. The two groups were originally matched on age; however, when several patients with SBM were excluded due to having

subtypes other than myelomeningocele, Verbal or Performance IQ scores below 70, or having invalid profiles, the final comparison group was one year younger, on average, than the clinical group. The comparison group also exhibited a higher Full Scale IQ that likely reflects the types of families who responded to the request for volunteer participants. Of note, the groups did not differ with respect to SES and, although not an optimal solution, attempts were made to control for effects of group differences in the analyses. Further, the modest sample size limited the power to statistically evaluate the numerous biological risk factors associated with SBM and the complex interaction effects necessary to examine potential moderating effects of reserve variables on the relationship between biological risk and executive functioning. The continued use of multivariate techniques will be essential in order to isolate the effects of the numerous potential biological risk factors, especially when so many of the factors affecting neurocognitive development in children with SBM are interrelated. Such techniques require large samples to be adequately powered, suggesting that future research move beyond single institution to multisite collaborative research protocols, such as those used in the present project or in the collaboration between researchers in Houston and Toronto (e.g., Dennis, Fletcher, Rogers, Hetherington, & Francis, 2002). Methodological limitations notwithstanding, the present study confirmed prior findings of metacognitive difficulties among children with SBM and took an important step towards identifying aspects of biological risk and environmental reserve that serve to buffer or to exacerbate such difficulties.

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