

SMITH-MAGENIS SYNDROME: MALADAPTIVE BEHAVIORS AND EFFECTS  
ON PARENT STRESS, COPING, AND FAMILY ADJUSTMENT

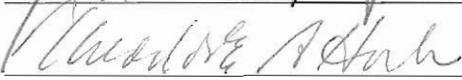
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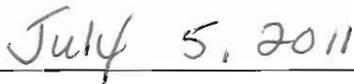


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Smith-Magenis Syndrome: Maladaptive Behaviors and Effects on Parent Stress, Coping,  
and Family Adjustment

A dissertation submitted in partial fulfilment of the requirements for the degree of Doctor  
of Philosophy at George Mason University

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## Table of Contents

	Page
List of Tables.....	iii
Abstract.....	iv
1. Introduction.....	1
2. Methods.....	9
3. Results.....	15
4. Discussion.....	20
5. References.....	27

## List of Tables

Table	Page
1. Table 1. Descriptive Statistics for Age and IQ.....	22
2. Table 2. Instrument Descriptive Statistics.....	23
3. Table 3. Spearman Correlation Coefficients.....	24
4. Table 4. Multiple Hierarchical Regression Summary Results Predicting Parental Stress by Age, IQ and Adaptive/Maladaptive Behavior .....	25
5. Table 5. Regression Summary Results Predicting Family Outcome Measures by Parent Stress.....	26

## Abstract

### SMITH-MAGENIS SYNDROME: MALADAPTIVE BEHAVIORS AND EFFECTS ON PARENT STRESS, COPING, AND FAMILY ADJUSTMENT

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George Mason University, 2011

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Smith Magenis Syndrome (SMS) is a rare genetic syndrome most commonly caused by a microdeletion on chromosome 17 p11.2. It is associated with a pattern of physical, developmental and behavioral characteristics including intellectual disability, sleep disturbance, and a variety of behavior problems. Data were collected on 25 individuals with SMS, 10 males (mean IQ 72; SD 13), and 15 females (mean IQ 68; SD 22.2); ages 1.4-19.4 (mean 6.56; SD 4.71). Parent reports were obtained using the Vineland Adaptive Behavior Scale (VABS; Sparrow, Balla, & Cicchetti, 1984), the Achenbach Child Behavior Checklist (CBCL; Achenbach, 1991), the Family Assessment Device-General Functioning Scale (Epstein, Baldwin, & Bishop, 1983), the Family Crisis Oriented Personal Evaluation Scales (McCubin, Olson, & Larson, 1991), the Parental Stress Index-3rd edition (Abidin, 1995), and the Stress Index for Parents of Adolescents (Abidin, 1995). We found positive linear relationships between maladaptive

behaviors and parent stress, but no association between maladaptive behaviors and overall life stress. We also found that parent stress levels were not associated with measures of family adjustment or specific parent coping strategies.

## Introduction

Smith Magenis Syndrome (SMS) is a rare micro-deletion syndrome of chromosome 17 p11.2 associated with a specific pattern of physical, developmental, and behavioral characteristics (Gropman & Smith, 2010; Smith & Gropman, 2010). New estimates suggest that 1 out of every 15,000 children are born with SMS (Gropman & Smith, 2010; Smith & Gropman, 2010). Originally described as a chromosomal abnormality with facial clefts by Smith and colleagues (Smith, McGavran, Waldstein, & Robinson, 1982), the first article detailing the SMS phenotype was published in 1986 by Smith et al. (1986), which laid the groundwork for the Natural History Study of Smith-Magenis syndrome at the National Institutes of Health. Those affected with SMS have a unique, consistent phenotypic appearance and developmental trajectory.

### *Maladaptive Behaviors*

Maladaptive behaviors are those which impair or compromise normal, healthy, adaptive functioning in everyday situations (Cooper, Heron, & Heward, 2006; Sigafos, Arthur, & O'Reilly, 2003). For individuals with intellectual and/or developmental disabilities (ID/DD), there can be an increased risk of maladaptive behaviors, especially when it is part of the behavioral phenotype of the underlying cause of the ID/DD (Emerson, Moss, & Kiernan, 1999; Rojahn, Schroeder, & Hoch, 2008; Sigafos et al., 2003). The term "behavioral phenotype" was first introduced by Nyhan in 1971 as that

overt behavior which is so indicative of a genetic condition, that its mere presence could demarcate the underlying disorder (Harris 2002; Nyhan, 1972). Further, Nyhan states that these behaviors might be as related to the underlying abnormal neurophysiology as the physiological components of a disorder (such as the dysmorphic faces of Down syndrome, or the limb abnormalities in Cornelia de Lange syndrome), and that "...these stereotypical patterns of unusual behavior could reflect the presence of structural deficits in the central nervous system" (Harris, 2002, pp. 626).

Behavioral phenotypes are stereotypic patterns of behavior that can be reliably associated as a distinguishing characteristic between groups, such as those with a particular biological condition or disorder (Flint & Yule, 1994; Harris, 1998; 1987). Furthermore, these behaviors are composed of a consistent pattern of motor, cognitive, linguistic, and social abnormalities, all of which are components of the diagnostic criteria. Some of these phenotypic behaviors may be so characteristic (such as the hand-to-mouth stereotypies of Rett syndrome) that they help define a specific disorder before the underlying genetic etiology is known, or may help define an acquired disorder, such as fetal alcohol syndrome (Flint & Yule, 1994; Harris, 2002).

It is important to note, however, that although some behaviors are very characteristic in nature, they may vary within the disorder from individual to individual, based on the environmental cues that evoke or elicit the behavior for that individual, and may also vary with regard to the topography (the form or shape of the behavior itself, i.e. self-injurious behavior in one person may be head-banging, whereas in another it is hand-biting), and magnitude (the intensity or severity of the behavior, such as if the head-

banging is done full force against a floor, or lightly against a cushioned surface), and temporal components (including aspects of frequency and duration) (Cooper et al., 2006). Phenotypic behaviors in a subset of conditions (such as autism spectrum disorder) are usually of a stereotypic nature, characterized by frequency, repetition, inappropriateness, and invariance (Moss, Oliver, Arron, Burbidge, & Berg, 2009), and may include minor to severe self-injurious behaviors, particularly in conditions that have inherent cognitive impairment (Allen, 2010; Byrne & Hennessy, 2009; Rojahn et al., 2008). These self-injurious behaviors may be particularly difficult for families, as they are difficult to stop, uncomfortable to watch, and in some instances, may result in well-intentioned, but erroneous reports of child-abuse.

#### *Maladaptive Behavior in SMS*

The neurobehavioral phenotype in SMS is distinct and complex, and is characterized by high frequency of: outbursts/tantrums, attention-seeking, impulsivity, aggression, hyperactivity, distractibility, toileting difficulties, stereotypies (repetitive or self-stimulatory behaviors), sleep disturbance and self-injurious behaviors (Dykens & Smith, 1998; Finucane & Haas-Givler, 2009; Gropman & Smith, 2010). Many children with SMS tend to exhibit a variety of behavior problems. Using the Child Behavior Checklist (CBCL), Dykens and Smith (1998) reported significantly higher levels of maladaptive behavior in children with SMS (89%), as compared to children with Prader Willi syndrome (71%), and a group of children with etiologically mixed intellectual disability (28%). Specifically, these researchers reported that individuals with SMS had difficulty regulating basic bodily functions (sleeping, modulating activity and affect,

eating and toileting), and tended to engage in self-injurious behaviors and stereotypic behavior (repetitive or self-stimulatory behaviors).

### *Parent Stress and Coping*

Jones and Passey (2005) studied parental stress in 48 British families who had a child with developmental disability and maladaptive behavior. Parents who had an internal locus of control did not report feeling that their lives were controlled by the child's disability, and instead utilized family integration, co-operation, and an optimistic attitude (cognitive reframing) as their primary coping strategies (Jones & Passey, 2005), whereas those with an external locus of control did not report using any one particular coping mechanism.

Several studies on families having children with autism spectrum disorder have found that more parents reported feeling stress and had an increased need for support and a lack of an ability to cope their child than families with typically developing children. Allen (2004) studied 18 married couples who were parents to a child diagnosed with autism. Using the Parent Stress Index (PSI) and Family Crisis Oriented Personal Evaluation Scales (F-COPES), Allen (2004) found that these couples reported an increased amount of stress (92<sup>nd</sup> percentile on the PSI) and the need for coping strategies (70<sup>th</sup> percentile) associated with caring for a child with increased behavioral problems and developmental deficiencies. According to this study, passive appraisal (F-COPES) was the most frequently, and spiritual support the least used, strategy for coping. However, unpublished data collected from a small sample of 41 SMS families (Morse,

Bernert & Smith, 2006), demonstrated a trend for parents increasing the use of spiritual support seeking as the child reaches adolescence and adulthood.

Maladaptive child behavior directly and indirectly affects a care-giver's ability to care for a child with developmental disabilities. In a study of 105 preschoolers with developmental disabilities (Plant & Sanders, 2007), mothers reported that care-giving tasks were often disrupted by the child's behaviors. Plant and Sanders (2007) determined key predictors of level of parental stress were tasks disrupted by child behavior and the level of the child's impairment. They found that moderators of care-giving ability and parental stress were mothers' reported level of social support. Finally, they reported that mediators of parent stress and the child's level of impairment were the parent's cognitive appraisal of the responsibilities incurred by caring for a child with developmental disabilities.

In a larger study ( $n = 880$ ) Smith, Oliver, and Innocenti (2001) found similar results, that parents of children with developmental disabilities reported more stress on the PSI, but the researchers also expanded their study to determine what aspects of living with and raising a child with developmental disabilities might be predictive of increased parental stress. Surprisingly, they found that increased parental stress was not as significantly correlated with increased child impairment as it was with factors defined as supportive, such as income, outside social supports, and time spent interacting with the child were better predictors for increased parental stress.

A model to attempt to define the stress unique to parenting a child with developmental disabilities was proposed by Perry (2005). This model classifies the

stressors as either characteristics unique to the child (child characteristics), and stressors not directly attributable to the child (other life stressors), and moderators such as resources (personal (parent's) resources and family system resources), and supports outside of the immediate family (informal social supports, formal supports, and services), and distinguishes between positive and negative parental outcomes (Perry, 2005).

Perry's (2005) model is consistent with previous studies; the more challenging the characteristics of the child, and the fewer or poorer the quality of parental resources, the greater the perceived parental stress, and the more negative the outcome for the parent, and presumably, for the child as well.

#### *SMS and Parent Stress and Coping*

With all of the medical, behavioral, and psychological difficulties inherent in living with SMS, families are often taxed, exhausted, and have limited resources. Raising a child with disabilities increases the demands on the family unit, particularly on the parents (Jones & Passey, 2005; Fiddler, Hoddap, & Dykens, 2000; Frey, Greenberg, & Fewell, 1989). In one of the few studies that examined maladaptive behaviors in children with SMS and parent stress in SMS, Fidler, Hodapp, and Dykens (2000), enrolled 60 children with SMS syndrome, Down syndrome, and Williams syndrome, who were three to ten years of age. They discovered differences between the syndromes on the predictive power of child behavior problems and child age on parental stress. In the Williams syndrome group, both the presence of maladaptive behavior and a young child age predicted family stress; whereas in the Down syndrome group only young child age was predictive of parental stress level. Only in children with Smith-Magenis syndrome was

maladaptive behavior alone predictive for parental level of stress. One explanation for these results might be that behaviors in children with SMS are just as disruptive in older children as they are in younger children, since the maladaptive behaviors and sleep disturbance associated with SMS do not significantly decrease as the child ages.

Therefore, it would appear that family functioning and parent stress would be affected by having to care for a child with disabilities. One specific question is whether maladaptive behavior leads to increased perceived parent stress, and in turn, to decreased family functioning. By learning more about the relationship between the child's maladaptive behaviors and parent stress level, and examining the relationship between types of coping strategies and parent stress, we hope to be able to assist parents to cope with their situational stressors, and to increase the quality of their family life. This study examined the relationship between the severity of child maladaptive behaviors, and parent stress. We anticipated that increased child maladaptive behaviors are associated with increased parents stress, and further, we expected that the more dependent the child is in terms of daily functions and routines, the higher the perceived parental stress. Another question this study sought to address was that of parent coping; more specifically, to see if higher reported parent stress could predict the level of family adjustment, adaptation, and more specifically, the type of coping strategies parents utilized.

## Method

### *Participants*

Data were collected from 25 parents of individuals with a cytogenetically confirmed deletion diagnosis of SMS [del (17) (p11.2p11.2)] who were participants in the SMS natural history study (protocol 01-HG-0109) at NIH. Parental informed consent was obtained for all participants, and assents for minors were also obtained, when appropriate. Demographic information is presented in Table 1.

### *Assessments and Instruments*

As part of their assessment for the study, parents/guardians completed questionnaires, medical and genetic histories, and were engaged in semi-structured interviews. As part of the enrollment in the Natural History of Smith-Magenis syndrome study protocol participants underwent extensive medical and psychological tests.

#### *Measures of General Intelligence*

Cognitive abilities of adult patients/participants were measured with the *Wechsler Adult Intelligence Scale - Third Edition (WAIS-III)* (Wechsler, 1997). The WAIS-III is one of the most widely used measures of adult intelligence, and may be used to assess cognitive functioning in individuals with cognitive disabilities. The reliability of the WAIS-III has been established with various clinical populations (Wechsler, 1997; Zhu et

al, 2001). Children and adolescents were tested with the *Stanford-Binet Intelligence Scales- 4<sup>th</sup> and 5<sup>th</sup> editions* (Roid, 2003): a test of general intelligence for ages 2.0 through 24 years. It consists of 15 subtests that yield T-scores ( $Mean = 50, SD = 8$ ), and 4 domains (Verbal Reasoning, Abstract/Visual Reasoning, Quantitative Reasoning, and Short-Term Memory) that yield standard scores ( $Mean = 100, SD = 16$ ) as well as one composite standard score. Infants were assessed with the *Bayley Scales of Infant Development (Mental)*-2nd and 3rd editions (Bayley, 2006), which assess cognitive functioning of children from birth to 42 months. It yields an overall standard score (Mental Developmental Index (MDI): ( $Mean = 100, SD = 15$ ), and an overall age equivalent.

#### *Vineland Adaptive Behavior Scale (VABS)*

The VABS (Sparrow, Balla, & Cicchetti, 1984) is a semi-structured interview conducted with the child's parent (usually the mother, or both parents), which is used to assess the child's behavioral functioning in the home environment in the following domains: communication, daily living skills, socialization, and motor skills, and an adaptive behavior composite. It yields standard scores ( $Mean = 100, SD = 15$ ) and age equivalents for each domain, and an adaptive age composite.

#### *Achenbach Child Behavior Checklist (CBCL)*

The CBCL (Achenbach, 1991) is a standardized parent behavior rating scale, which measures the frequency (occurrence) of the behaviors (Not True; Sometimes or Somewhat True; Often True). Using parent report, children and young adults are assessed for behavioral problems and social competencies. It has up to 10 subscales (dependent upon the version for that age group) that yield T-scores ( $Mean = 50, SD = 10$ ). The

subscales consist of the following: social withdrawal; somatic complaints; anxious/depressed; social problems; uncommunicative; hyperactive; delinquent; aggressive; externalizing; and internalizing. Although the CBCL is only for use diagnostically for children ages four to 16 years of age, it was used for all enrolled participants with SMS; this is not considered to be a confound, as relatively few individuals with SMS, and none in this sample, have a mental age equivalent over 16, as measured by the VABS. Only two individuals in this sample had a chronological age over 16 (16.4 and 19.4, respectively), and were examined for effects as outliers.

*Family Assessment Device (FAD)*

The *General Functioning Scale* of the FAD (Epstein, Baldwin, & Bishop, 1983) is a parent report questionnaire of 60 items and consists of six domains of family functioning based on the McMaster Model (Epstein, Baldwin, & Bishop, 1983), and it has demonstrated reliability and validity. The domains include: Problem Solving, Communication, Roles, Affective Responsiveness, Affective Involvement, Behavior Control, and General Functioning. The *General Functioning Scale* consists of the most highly inter-correlated subset of all the items and measures the family members' perceptions of their family and assesses the overall health/pathology of the family. Higher scores (2.00 or above) are considered indicative of problematic family functioning. Various studies have tested for reliability, and found Cronbach alpha levels for the subscales of the FAD between .72 and .90 (Epstein, Bishop, Ryan, Miller, & Keitner, 1993), which is consistent with the original study, which found levels between 0.72 and 0.92 (Epstein, Baldwin, & Bishop, 1983). Test-retest reliability coefficients

range between 0.66 and 0.76, with concurrent validity estimates of the general functioning score exceeding  $r = .50$  (Miller, Epstein, Bishop, & Keitner, 1985).

*Family Crisis Oriented Personal Evaluation Scales (F-COPES)*

The F-COPES (McCubbin, Olson, & Larson, 1991) was developed to identify effective problem-solving and behavioral strategies used by families in response to problems or difficulties. The 30-item scale assesses the ways in which a family internally and externally handles difficulties. Subscales which measure the type of coping strategy employed by the parents include: Social Support Seeking, Cognitive Reframing, Spiritual Support Seeking, Acquiring and Accepting Help (Mobilizing), and Passive Appraisal. Higher scores are considered indicative of more adaptive family coping. Cronbach alpha levels range from .71 to .86, and test-retest reliability coefficients range from .61 to .95 (McCubbin, Olson, & Larson, 1991).

*Parental Stress Index-3<sup>rd</sup> Edition (PSI)*

The PSI (Abidin, 1995) was used to assess the parental stress of parents with children from one month to 12 years, and was theoretically derived from the concept that parent stress is a direct function of child, parent, and situational variables. The full scale consists of 101 items that fall into six child domains (Distractibility/Hyperactivity, Adaptability, Reinforces Parent, Demanding-Ness, Mood, Acceptability) and seven parent domains (Competence, Isolation, Attachment, Health, Role Restrictions, Depression, and Spouse), and also yields a Parent (Total) Stress Score, and a Life Stress Score; the percentile score is used in lieu of a standard score. Chronbach alpha levels for

the long form range from .70 to over .90, with test-retest reliability coefficients in subsequent studies ranging from .55 to .96 (Abidin, 1995).

#### *Stress Index for Parents of Adolescents (SIPA)*

The SIPA (Sheras, Abidin, & Konold, 1998) scale is the version of the PSI for parents of adolescent's ages 11 – 19 years. It consists of 112 items and the adolescent characteristics consist of the subscales: Moodiness/Emotional Lability Social Isolation/Withdrawal, Delinquency/Antisocial, Failure to Achieve or Persevere; whereas the parent characteristics include: Life Restrictions, Relationship with Spouse/Partner, Social Alienation, and Incompetence/Guilt. The SIPA also yields a Parent (Total) Stress Score, and a Life Stress Score; the percentile score is used in lieu of a standard score. Chronbach alpha levels for the SIPA range from .81 to over .90; test-retest reliability coefficients for all subscales range from .74 to .91 (Sheras, Abidin, & Konold, 1998).

For the analyses, the Parent (Total) Stress and the Life Stress Scores from both the PSI and the SIPA were used, depending on the child's age. Parent Stress for the PSI/SIPA includes the items which comprise *all* the domains/subscales listed above. These Parent Stress items includes questions which relate to the impact having this child has had on the parent, such as decreased time to spend with friends, feeling incapable or guilty, decreased intimacy between spouses, and increased fighting between spouses. Life Stress for the PSI/SIPA includes items separate from the domains/subscales, and these questions relate to specific life events, such as divorce, increased debt, employment problems, and legal problems.

#### *Procedure*

Full scale intelligence quotient scores from the Stanford Binet IV and V, the

WAIS-III and the Bayley were included to determine patient sample parameters. All cognitive batteries were administered by contracted NIH licensed psychologists. Parent rating scales and the VABS were administered to the parents at the time of visit by either a contracted NIH licensed psychologist, or a trained and supervised research assistant. The majority of the informants were the mothers, with input from the fathers when available.

## Results

Data were analyzed using SPSS software. First, we generated descriptive statistics of the dependent and independent variables (see Table 2).

### *Behavior Problems and Parent Stress*

Spearman correlation coefficients for all variables of interest were computed and are reported in Table 3. As age and cognitive level (IQ) were both significantly correlated with the CBCL Externalizing score, and IQ was significantly correlated with the CBCL Total score, we controlled for them in the respective hierarchical linear regression analysis, as outlined below. The results for all regression analyses are presented in Table 4.

#### *CBCL Total Behaviors*

The first hierarchical regression analysis was conducted using the CBCL Total score, child age, and IQ as the respective independent variables, and PSI/SIPA Total Parent Stress as the dependent variable. Age and IQ were entered in Block 1, and the CBCL Total score in Block 2. Maladaptive behaviors (CBCL Total Score) did significantly predict perceived parent stress levels over and above age and IQ: CBCL Total  $\beta = .76$ ,  $t(20) = 2.69$ ,  $p = .015$ , and explained a significant proportion of variance in parent stress levels ( $R^2 = .24$ ,  $F(1,19) = 7.23$ ,  $p < .015$ ).

#### *CBCL Externalizing Behaviors*

The second hierarchical regression analysis was conducted using the CBCL Externalizing behaviors score, child age, and IQ as the respective independent variables, and PSI/SIPA Total Parent Stress as the dependent variable. Age and IQ were entered in Block 1, and the CBCL Externalizing score in Block 2. Externalizing behaviors did significantly predict perceived parent stress levels over and above age and IQ: CBCL Externalizing  $\beta = .55$ ,  $t(20) = 2.31$ ,  $p < .05$ , and explained a significant proportion of variance in parent stress levels ( $R^2 = .19$ ,  $F(1,19) = 5.33$ ,  $p < .05$ ).

#### *CBCL Internalizing Behaviors*

Simple regression was conducted to determine if CBCL Internalizing behaviors were predictive of Total Parent Stress. CBCL Internalizing behaviors were predictive of parent total stress  $\beta = .43$ ,  $t(21) = 2.23$ ,  $p < .05$ , and explained a significant proportion of variance in parent stress levels ( $R^2 = .19$ ,  $F(1, 22) = 4.99$ ,  $p < .05$ ).

#### *Vineland Adaptive Behavior Composite*

The third hierarchical regression analysis was conducted using the Vineland Adaptive Behavior Composite (VABS ABC) score, child age, and IQ as the respective independent variables, and PSI/SIPA Total Parent Stress as the dependent variable. Age and IQ were entered in Block 1, and the VABS ABC score in Block 2. The child's adaptive functioning level was not predictive of total parent stress  $\beta = .31$ ,  $t(20) = .81$ ,  $p > .05$ , and did not explain a significant proportion of variance in parent stress levels ( $R^2 = .03$ ,  $F(1, 19) = .66$ ,  $p > .05$ ).

#### *Maladaptive Behaviors and Life Stress*

##### *CBCL Total Behaviors*

The fourth hierarchical regression analysis was conducted using the CBCL Total score, child age, and IQ as the respective independent variables, and PSI/SIPA Life Stress score as the dependent variable. Age and IQ were entered in Block 1, and the CBCL Total score in Block 2. Maladaptive behaviors (CBCL Total) were not predictive of overall life stress  $\beta = .46$ ,  $t(20) = 1.40$ ,  $p > .05$ , and did not explain a significant proportion of variance in parent stress levels ( $R^2 = .09$ ,  $F(1, 19) = 1.96$ ,  $p > .05$ ).

#### *CBCL Externalizing Behaviors*

The fifth hierarchical regression analysis was conducted using the CBCL Externalizing behaviors score, child age, and IQ as the respective independent variables, and PSI/SIPA Life Stress score as the dependent variable. Age and IQ were entered in Block 1, and the CBCL Externalizing score in Block 2. Externalizing behaviors were not predictive of overall life stress  $\beta = .11$ ,  $t(20) = .39$ ,  $p > .05$ , and did not explain a significant proportion of variance in life stress levels ( $R^2 = .01$ ,  $F(1, 19) = .15$ ,  $p > .05$ ).

#### *CBCL Internalizing Behaviors*

Simple regression was conducted to determine if CBCL Internalizing behaviors were predictive of Life Stress. Internalizing behaviors were not predictive of life stress  $\beta = .32$ ,  $t(21) = 1.61$ ,  $p > .05$ , and did not explain a significant proportion of variance in life stress levels ( $R^2 = .11$ ,  $F(1, 22) = 2.58$ ,  $p > .05$ ).

#### *Vineland Adaptive Behavior Composite*

The sixth hierarchical regression analysis was conducted using the Vineland Adaptive Behavior Composite (VABS ABC) score, child age, and IQ as the respective independent variables, and PSI/SIPA Life Stress as the dependent variable. Age and IQ

were entered in Block 1, and the VABS ABC score in Block 2. The child's adaptive functioning level was not predictive of life stress  $\beta = -.64$ ,  $t(20) = -1.68$ ,  $p > .05$ , and did not explain a significant proportion of variance in life stress levels ( $R^2 = .13$ ,  $F(1, 19) = 2.81$ ,  $p > .05$ ).

### *Family Stress and Coping*

To determine if total perceived parent stress predicted overall family functioning, regression analyses were conducted, using the Parent Total Stress Percentile scores from the PSI/SIPA as the predictor variable, and the FAD Global Functioning score as the outcome variable. To determine if total perceived parent stress predicted the need for, and reliance on, specific coping strategies, regression analyses were conducted, using the Parent Total Stress Percentile scores as the predictor variable, and the F-Copes Total score, and respective subscales as the outcome variables. Parent stress levels were not predictive for either family adjustment (FAD GFS), nor was it predictive for total parent coping (F-COPES Total score), or coping style (F-COPES subscales). The results for all regression analyses are presented in Table 5.

## Discussion

In summary, this study showed that there was a positive linear relationship between child maladaptive behaviors and parent stress; however, there was no such association between maladaptive behaviors and life stress. It is possible, however, that this differential result was merely due to the small sample size. In fact, correlation coefficients in Table 3 suggest that internalizing maladaptive behavior contributes to both parent stress and to life stress. Interestingly, as strongly as externalizing behavior contributed to parent total stress, it was far from being correlated with parent life stress.

Overall, these findings are consistent with anecdotal reports by parents of how disruptive a child's maladaptive behavior can be to the family. Furthermore, the finding that externalizing behavior problems were positively correlated with age is consistent with previous findings of developmental asynchrony by Finucane, Dirrigl, and Simon (2001).

It is important to mention that chronological age and IQ were highly negatively correlated ( $r = -.93$ ). Parents report an increasing gap in abilities between the SMS child and typically developing children; however, subjective impressions do not constitute empirical evidence. A developmental regression is not typically seen in SMS, yet our results clearly evidence a significant trend for decreased IQ in this population. As these individuals do continue to gain skills, just at a slower rate than their typically-developing

counterparts, it may be more informative to use mental-age equivalents rather than IQ when describing cognitive abilities in SMS.

Surprisingly, we found that parent stress levels were not associated with measures of family adjustment, or with specific parent coping strategies. This lack of significant association in the latter two relationships may be due to the small sample size, and the restricted range of our measures.

Relative weaknesses of this study include the small sample size, the lack of a control group with age/sex/cognitive function- matched subjects, and an inherent self-selection bias (only subjects who sought medical attention and had parents who consented to participate in a research study), and limited availability of complete and time-matched data sets for all instruments.

Future longitudinal studies are warranted to examine developmental changes among SMS across these behavioral outcomes, accounting for parental coping styles, and overall parent stress, and adjusting for both age-related changes and cognitive function.

Table 1. Descriptive Statistics for Age and IQ

Gender	<i>N</i>	Age			IQ		
		Mean	<i>SD</i>	Range	Mean	<i>SD</i>	Range
Males	10	5.22	1.78	n.a.	72.1	13.32	55-90
Females	15	7.46	5.81	n.a.	68.43	22.19	39-104
Total	25	6.56	4.71	1.4 – 19.4	69.96	18.74	39-104

Table 2. Instrument Descriptive Statistics

Instrument	<i>N</i>	Mean	<i>SD</i>	Range
CBCL Total T-score	24	64.71	9.82	47-83
CBCL Externalizing	24	59.46	12.24	37-85
CBCL Internalizing	24	57.25	9.33	40-78
VABS Adaptive Behavior	24	58.33	11.74	30-77
Parent Total Stress Percentile	25	72.64	24.09	25-99
Parent Life Stress Percentile	25	48.16	31.37	0-98
FAD Global Functioning Score	23	3.30	0.44	2.25-4.00
F-Copes Total Score	24	103.46	9.43	87-128
F-Copes Acquiring Social Support	24	28.08	5.64	19-39
F-Copes Cognitive Reframing	24	31.00	3.64	21-38
F-Copes Spiritual Support Seeking	24	13.00	3.59	6-18
F-Copes Mobilizing Support	24	14.58	2.39	9-19
F-Copes Passive Appraisal	24	16.79	2.11	11-20

Table 3. Spearman Correlation Coefficients

	Age	Sex	CBCL Internalizing	CBCL Externalizing	CBCL Total T-score	IQ	Parent Total Stress Percentile	Parent Life Stress Percentile	F-Copes Acquiring Social Support	F-Copes Cognitive Reframing	F-Copes Spiritual Support Seeking	F-Copes Mobilizing Family Support	F-Copes Passive Appraisal	F-Copes Total Score	FAD Global Functioning Score	VABS Adaptive Behavior Composite
Age		.091	.049	.501	.595**	-.927**	.215	.113	-.218	.370	.173	-.069	-.016	.219	.376	-.794**
Sex			.343	.391	.275	-.122	.329	-.091	.104	-.321	-.018	.068	-.161	-.129	.007	.110
CBCL Internalizing				.600**	.606**	-.199	.575**	.425*	.120	.019	.146	-.014	.100	.292	.132	-.037
CBCL Externalizing					.890**	-.569**	.643**	.026	.284	.061	.171	.111	-.135	.333	.304	-.360
CBCL Total T-score						-.777**	.694**	.214	-.047	.097	.145	.017	-.053	.225	.300	-.550**
IQ							-.381	-.210	.309	-.248	-.188	.052	.014	-.110	-.283	.818**
Parent Total Stress Percentile								.326	-.009	.026	-.286	.099	-.353	-.079	.224	-.253
Parent Life Stress Percentile									-.221	.140	-.261	-.162	-.057	-.101	.210	-.199
F-Copes Acquiring Social Support										-.045	.219	.375	-.418*	.469*	-.167	.131
F-Copes Cognitive Reframing											.127	.006	.166	.537**	.177	-.097
F-Copes Spiritual Support Seeking												-.134	.270	.619**	-.265	-.054
F-Copes Mobilizing Family Support													-.115	.328	-.027	.054
F-Copes Passive Appraisal														.264	.062	.077
F-Copes Total Score															-.002	-.068
FAD Global Functioning Score																-.256
VABS Adaptive Behavior Composite																

\*. Correlation is significant at the 0.05 level (2-tailed).

\*\* . Correlation is significant at the 0.01 level (2-tailed).

Table 4. Multiple Hierarchical Regression Summary Results Predicting Parental Stress by Age, IQ and Adaptive/Maladaptive Behavior

Predictor	Parent Total Stress		Life Stress	
	$\Delta R^2$	$\beta$	$R^2$	$\beta$
Step 1	.12			
Age		-.09		-.05
IQ		-.42	.04	-.24
Step 2				
CBCL Total Score	.24**	.76	.09	.50
Step 1	.12			
Age		-.09		-.05
IQ		-.42	.04	-.24
Step 2				
CBCL Externalizing	.19*	.55	.01	.11
Step 1	.13			
Age		-.05		-.24
IQ		-.40	.02	-.27
Step 2				
VABS Adaptive Composite	.03	.31	.13	-.64

Note. \*  $p \leq .05$ , \*\*  $p \leq .01$ , \*\*\*  $p \leq .001$

Table 5. Regression Summary Results Predicting Family Outcome Measures by Parent Stress

Parent and Family Outcome Measures	PSI/SIPA Parent Stress	
	$R^2$	$\beta$
FAD Global Functioning Score	.06	.24
F-Copes Total Score	.10	-.32
F-Copes Social Support	.06	-.24
F-Copes Cognitive Reframing	.00	-.06
F-Copes Spiritual Support	.09	-.29
F-Copes Mobilizing Support	.02	.13
F-Copes Passive Appraisal	.11	-.33

Note. \*  $p \leq .05$ , \*\*  $p \leq .01$ , \*\*\*  $p \leq .001$

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## Curriculum Vitae

Rebecca S. Morse received her Associate of Arts degree in Music Education from Frederick Community College in 1999, her Bachelor of Arts degree in Psychology from Hood College in 2003, and her Master of Arts degree in Thanatology from Hood College in 2005. She was employed as a research assistant/research coordinator at the National Institutes of Health from 2002 to 2010, has been on adjunct faculty at Hood College since 2006, and received her Doctorate in Psychology from George Mason University in 2011.