

## Stress and coping in families of children with Smith–Magenis syndrome

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### Abstract

To describe stress and coping in families of children with Smith–Magenis syndrome, the present authors interviewed and received questionnaires from families of 36 children with this disorder. For measures of total stress, and of parent and family problems, the best predictors were the family's number of friends and the child's degree of impairment on the Vineland socialization domain; the single best predictor of parental pessimism was the child's degree of maladaptive behaviour. Although the stress levels of the families of children with Smith–Magenis syndrome are comparable to the levels shown by the families of children with Prader–Willi and 5p- syndromes, these levels are much higher than the stress levels reported by families of children with mixed or non-specific developmental disabilities. Stress levels may be similar across aetiologies involving high levels of maladaptive behaviour, but the correlates of family stress—particularly the moderating role of family friends—seem specific to Smith–Magenis syndrome.

**Keywords** Smith–Magenis syndrome, families, stress, coping, mental retardation

### Introduction

Until about 20 years ago, the families of children with disabilities were thought of as 'problem'

families, with researchers examining divorce in couples, role tensions in siblings, and psychopathology in mothers, fathers and the family system as a whole. In contrast to this earlier focus, more recent research centres on familial stress and coping. Families of children with disabilities are now conceptualized as undergoing increased stresses, but often as coping fairly well (Hodapp 1995). In addition, the family's social supports seem critical, as is the possibility that increased social support from extended family, professionals or friends might alleviate the stresses experienced by these families.

This emphasis on stress and coping leads to a heightened interest in individual differences. If families differ one from another, which aspects of children, parents or families lead certain families to cope well while others have problems? For example, the age of the child may relate to levels of a family's stress. Families may experience more stress when the child is either older (Minnes 1988), or at certain 'critical ages' such as puberty or the entrance into adulthood (Wikler 1986). Conversely, more stress might be experienced by families of children who are more severely delayed (Frey *et al.* 1989) or by families whose children show more behaviour problems (Margalit *et al.* 1987). Other parental and familial variables sometimes associated with lessened stress include increased social support (Beckman 1991), more harmonious marriages, two- versus one-parent families (Beckman 1983), and higher socioeconomic status (Farber 1970). These variables

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probably operate in combination, together predisposing one family to increased stress or another to more successful coping.

Such family issues become more complicated when dealing with different aetiologies. Most existing studies focus on families of children with mixed or non-specific developmental disabilities, yet family stress and coping may change based on the child's specific type of intellectual disability. Indeed, certain aetiologies predispose individuals to specific adaptive and maladaptive behaviours, intellectual or linguistic strengths and weaknesses, and characteristic developmental patterns (Hodapp & Dykens 1994; Dykens 1995), but different aetiologies may also indirectly influence parents, peers and other surrounding individuals (Hodapp 1997, 1999). For example, families of children with Down's syndrome generally show lower levels of family stress than families of children with other disabilities (Seltzer & Ryff 1994; although see also Cahill & Glidden 1996). Conversely, families of children with Prader–Willi syndrome show high levels of stress (Hodapp *et al.* 1997a). In addition, family stress in several genetic disorders seems related to the child's maladaptive behaviours. In both Prader–Willi syndrome and in 5p- (cri du chat) syndrome—two disorders with extremely high levels of child maladaptive behaviour (Dykens & Clarke 1997; Dykens & Kasari 1997)—the child's behaviour problems are the single best predictor of familial stress (Hodapp *et al.* 1997a, b).

Smith–Magenis syndrome is another disorder in which various behavioural characteristics may be associated with increased levels of family stress. First described in 1986, Smith–Magenis syndrome is caused by a deletion on the short arm of chromosome 17 (Smith *et al.* 1986). The disorder affects  $\approx 1$  in 25 000 births, and individuals with Smith–Magenis syndrome often show brachycephaly (i.e. flat head shape) as well as characteristic facial features (e.g. a broad nasal bridge and flat mid-face) (Greenberg *et al.* 1991, 1996). Smith–Magenis syndrome is associated with moderate levels of intellectual disability, hyperactivity, and aggressive outbursts and tantrums, as well as three striking maladaptive behaviours. The first involves a high degree of self-injurious behaviour, including head banging, skin-picking, the pulling off of finger- or toenails, and the insertion of foreign objects into

bodily orifices (Colley *et al.* 1990; Greenberg *et al.* 1991, 1996; Stratton *et al.* 1986). Secondly, individuals with this disorder often show profound sleep disturbances, including reduced REM, difficulties getting to sleep, and extended periods of wakefulness during the night and sleepiness during the day (Dykens *et al.* 1997; Smith *et al.* 1997). Thirdly, when excited many of these individuals engage in an unusual stereotypy, a characteristic 'self-hug' that is performed as often as 100 times per hour (Finucane *et al.* 1994).

The present study examines how various child and family characteristics relate to stresses and supports in families of children with Smith–Magenis syndrome. The present authors' goal was two-fold. Firstly, they related characteristics of the children, and their parents and families to various domains of family stress, and also compared the predictors of family stress in Smith–Magenis syndrome to predictors found in earlier studies of Prader–Willi and 5p- (cri du chat) syndromes. Secondly, they described basic characteristics of stress and support in these families. In these analyses, the present authors also compared stress and support characteristics to families of children with other genetic disorders, and to families of children with mixed aetiologies.

## Method

### Subjects

The subjects were 36 children (16 males and 20 females) diagnosed with Smith–Magenis syndrome. The mean  $\pm$  SD age of the subjects was  $8.35 \pm 4.16$  years and all subjects lived in their family home. The average age at Smith–Magenis diagnosis was 2.75 years.

The children in the present sample predominantly showed moderate levels of intellectual disability. On the Vineland Adaptive Behaviour scales, the overall age-equivalent scores averaged 3.90 years (SD = 1.71) and the average standard score composite was 47.79 (SD = 16.11). These children also showed high levels of maladaptive behaviour: on the Child Behaviour Checklist (CBCL; Achenbach 1991), the subjects averaged a total score of 70 and 28 out of 36 subjects (78%) were above CBCL 'clinical cut-off' scores (i.e. ninety-eighth percentile

score) for children their age. Over half (53%) of the children attended special classes or schools, 25% were mainstreamed full- or part-time, and 22% were in early intervention or other settings.

The families of these children were predominantly middle class. The mean educational levels of mothers and fathers were 14.37 years (range = 12–19 years) and 14.66 years (range = 10–20 years), respectively. Forty-six per cent of families had a combined family income of \$50 000 or more. All but four of the 36 families were intact, two-parent families. The respondents included 33 mothers and three fathers.

### Procedures

The parents of the children with Smith–Magenis syndrome were all members of the Parents and Researchers Interested in Smith–Magenis Syndrome (PRISMS), the disorder's main US parent-professional group. Calls, mailings and solicitations were made to the families of individuals with this disorder. From these solicitations, 41 families responded; these families constituted  $\approx$  66% of the membership of PRISMS at the time of the present study. Out of these 41 families, 36 were raising children aged 18 years or younger. In addition to the mailed questionnaires (described below), parents were called by telephone. The parents were administered the Vineland Adaptive Behavior Scales in 30–45-min telephone interviews. Besides measuring the children's adaptive functioning, the telephone interviews also helped to fill in any missing or unclear responses from the mailed questionnaires.

### Measures

#### *Demographic instrument*

Parents were asked the age, sex and first name of their child, and where and when the child was diagnosed with Smith–Magenis syndrome. They were also asked to list everyone in their immediate family, each person's age and role within the family (e.g. mother and son), and whether that person lived at home. Parents were also asked about their occupations, level of education completed, family income and marital status.

#### *Questionnaire on Resources and Stress—Friedrich edition*

The Questionnaire on Resources and Stress—Friedrich edition (QRS-F; Friedrich *et al.* 1983) consists of 52 Yes/No items which converge on four factors. Factor I, consisting of 20 items, asks about *Parent and Family Problems*. Items include 'Other members of the family have to do without things because of [the child with retardation]' and 'I have given up things I really wanted to do because of [the child with retardation]'. Factor II, consisting of 11 items, asks about parental *Pessimism*. Items include 'I worry about what will happen to [child with retardation]' and 'I am disappointed that [child with retardation] does not live a normal life'. The remaining two factors, *Child Characteristics* and *Physical Incapacitation*, relate more to the child with disabilities; these domains were not used in the present study. This shortened form of the original Holroyd (1974) scales has been widely used in family research (Scott *et al.* 1989).

#### *Family Support Questionnaire*

Adapted from Kazak (1987), the Family Support Questionnaire asks respondents to list 'people who are important to you at this time in your life'. These individuals, who the respondent has called, visited or done things with over the past 4–6 weeks, can be members of one's family, friends or professionals. Room for up to 14 names is provided.

In addition to a measure of those individuals constituting the parent's support network, questions are asked about each supporter. Parents are asked about the relationship of this person (e.g. friend, family member or professional), and that person's sex, age and residence (e.g. home, neighborhood, city or other). Parents also describe the kinds of support provided by this person: emotional, informational, tangible or service. This person's degree of helpfulness is rated on a five-point scale and parents also note the types of reciprocal support which they return to the supporter (e.g. emotional, informational, tangible or service).

#### *Child Behavior Checklist*

The Child Behavior Checklist (CBCL; Achenbach 1991) is a parent-report measure asking about 112 common problems of childhood. Parents note

whether each behaviour is 'Not True' (0 points), 'Somewhat or Sometimes True' (1 point) or 'Very True or Often True' (2 points) of their child. The behaviours relate to two 'broad-band' factors, Internalizing and Externalizing behaviours, and nine 'narrow-band' factors. The nine narrow-band factors are Withdrawn, Somatic Complaints, Anxious/Depressed, Social Problems, Thought Problems, Attention Problems, Delinquent Behaviour, Aggressive Behaviour and Other. The CBCL is among the most widely used measures of children's maladaptive behaviour, and has been used to assess non-retarded children (Achenbach 1991), children with mixed aetiologies (Dykens & Cohen 1996), and children with aetiologies such as Smith–Magenis syndrome (Dykens *et al.* 1997), Down's syndrome (Dykens & Kasari 1997) and Prader–Willi syndrome (Dykens & Kasari 1997; Dykens *et al.* 1992).

#### *Sleep Questionnaire*

Specifically developed for Smith–Magenis syndrome, the Sleep Questionnaire asks parents about various aspects of their child's sleep patterns (Smith *et al.* 1997). In addition to describing the time of the child's bedtime and waking, and the number and length of naps during the day, parents also answer 'yes' or 'no' to 32 questions. These questions are grouped into domains of problems: Getting to Sleep (nine items), During Sleep (14 items), Morning Behaviour (six items) and Daytime Behaviour (three items). Total scores for all sleep problems are computed, as are domain scores.

#### *Vineland Adaptive Behavior Scales—Screener Edition*

The Vineland Screener is a shortened form of the Vineland Adaptive Behavior Scales (Sparrow *et al.* 1983) designed specifically for research purposes. It has acceptable reliability, validity and factor structures (Sparrow *et al.* 1995). Interviewers directly ask parents about the child's functioning in four domains: Communication, Daily Living Skills, Socialization and Motor Skills (up to 6 years of age). Like the original Vineland Adaptive Behavior Scales, the Vineland Screener provides age-equivalent and standard scores in each of the four domains.

## Results

### Characteristics of family stress and support in Smith–Magenis syndrome

The families of children with Smith–Magenis syndrome showed high levels of both family stress and family support. Parent and Family Problem scores averaged 9.08 (out of a possible 20), Pessimism 7.33 (out of 10) and total QRS-F scores 25.14 (out of 52). At the same time, however, these families also enjoyed high levels of social support. The average family listed 9.06 supporters, and while family members made up exactly half of these supporters (4.53), family friends (2.62 or 28%) and professionals (1.91 or 22%) were also mentioned. Over three-quarters of families (76.5%) listed one or more professionals in their support system. These professionals mainly included special education teachers (30.5%), physicians (23.7%) and psychotherapists (20.3%), although child care workers/baby-sitters (8.4%), social/caseworkers (5.1%) and clergy (3.4%) were also mentioned. The most common types of support were emotional (85% of all supporters), informational (54%) and service (38%), and families seemed satisfied with all types of support (rating almost 4 on a five-point scale).

### Relationship of child, parent, family and support characteristics to family stress

To identify correlates of family stress in Smith–Magenis syndrome, correlations were first performed between child, parent and family characteristics, and the three domains of family stress. Child characteristics included the child's age, sex, age-equivalent and standard scores on the Vineland Adaptive Behavior Scales, and narrow-band and wide-band scores on the CBCL. Parent and family characteristics included parental educational levels and marital status, the number in family, and family income.

Several child characteristics were related to the three family stress domains. Specifically, the Sleep Questionnaire Morning Behaviours category, involving items such as waking up looking tired, reporting a bad dream during night and awakening in a different room, was related to Parent–Family Problems ( $r = 0.52$ ;  $P < 0.001$ ), Pessimism

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( $r = 0.42$ ;  $P < 0.05$ ) and Total QRS-F score ( $r = 0.47$ ;  $P < 0.01$ ). Adaptive and maladaptive scores were also related to levels of family stress. As shown in Table 1, Vineland Adaptive Behavior Scales Socialization standard scores correlated with all three domains of family stress, and overall maladaptive behaviour (i.e. CBCL total score) correlated with Parent–Family Problems ( $r = 0.47$ ;  $P < 0.01$ ), Pessimism ( $r = 0.57$ ;  $P < 0.001$ ) and QRS-F total score ( $r = 0.49$ ;  $P < 0.01$ ). Various CBCL wide- and narrow-band factors also related to the three domains of family stress.

Aspects of the parents themselves and of the family's social support systems also related to family stress. Specifically, mothers who were more educated showed lower levels of Parent–Family Problems ( $r = 0.55$ ;  $P < 0.001$ ), Pessimism ( $r = 0.41$ ;  $P < 0.05$ ) and QRS-F total scores ( $r = 0.48$ ;  $P < 0.01$ ). Several aspects of family support also alleviated family stress. The most important variable here was the number of friends listed in the support system. The number of friends was negatively correlated with Parent–Family Problems ( $r = -0.60$ ;  $P < 0.0001$ ), Pessimism ( $r = -0.40$ ;  $P < 0.05$ ) and QRS-F total ( $r = -0.55$ ;  $P < 0.001$ ).

Using stepwise multiple regression techniques to identify the independent contribution of child, family and support variables, different variables appear important to different aspects of family stress. As Table 2 shows, the number of friends seems the most important variable affecting Parent–Family Problems and QRS-F total scores; in both cases, the number of friends accounts for over 30% of the variance. The socialization standard score also adds another 13–15% of the variance for Parent–Family Problems and QRS-F total. Conversely, the only significant predictor of parental Pessimism was the child's total CBCL score, which accounted for over 30% of the variance (see Table 2).

These correlates of family stress are both the same and different compared to the present authors' earlier studies of families of children with Prader–Willi syndrome (Hodapp *et al.* 1997a) and with 5p-syndrome (Hodapp *et al.* 1997b). Across all three studies, the demographic characteristics of the samples were virtually identical. The mothers of children in the Smith–Magenis, Prader–Willi and 5p-groups all averaged  $\approx 2$  years of college or university education (14.37, 15.0 and 14.34 years of education, respectively); the fathers were also almost identical in education across the three samples

**Table 1** Correlations of stress with adaptive and maladaptive behaviours

	Parent–family problems	Pessimism	Total QRS-F
<i>Vineland standard scores</i>			
Communication	–0.27	–0.43*	–0.29
Daily Living Skills	–0.21	–0.34*	–0.30
Socialization	–0.47**	–0.42*	–0.45**
<i>CBCL wide-band factors</i>			
Internalizing	0.35*	0.49**	0.38*
Externalizing	0.51*	0.54***	0.48**
<i>CBCL narrow-band factors</i>			
Withdrawal	0.40*	0.36*	0.41*
Somatic Complaints	0.20	0.28	0.23
Anxious/Depressed	0.25	0.47**	0.27
Social Problems	0.30	0.48**	0.34*
Thought Problems	0.34*	0.27	0.29
Attentional Problems	0.37*	0.52**	0.39*
Delinquent Behaviour	0.42**	0.47**	0.46**
Aggressive Behaviour	0.48**	0.51***	0.45**
Other	0.41*	0.45**	0.46**

\* $P < 0.05$ ; \*\* $P < 0.01$ ; \*\*\* $P < 0.001$ .

**Table 2** Predictors of three domains of familial stress\*

	Multiple R	R <sup>2</sup>	Change in R <sup>2</sup>	F-value	P-value
<i>Parent and family problems</i>					
(1) Friends	0.617	0.381	–	16.62	0.0004
(2) Socialization standard score	0.728	0.530	0.149	14.67	0.0008
<i>Pessimism</i>					
(1) Total CBCL	0.558	0.312	–	12.24	0.002
<i>Total QRS-F score</i>					
(1) Friends	0.559	0.313	–	12.27	0.002
(2) Socialization standard score	0.667	0.445	0.132	10.41	0.0005

\*For all analyses, the independent variable are: child's age; total CBCL score; Vineland socialization standard score; morning behaviours; and family's number of friends.

(14.66, 14.80 and 14.39, respectively). Across all three groups, from 40% to 50% of families earned over \$50 000 in total family income and the percentages of single-parent families were low (from 6% to 14%).

When examining correlates of family stress, however, friends served to alleviate family stress only in the Smith–Magenis sample. Indeed, although the family demographics and research methods were identical in studies of the three groups, only the Smith–Magenis analyses revealed friends as an important moderator of family stress. In both other groups, maladaptive behaviour was the best—and sometimes sole—predictor of familial stress (see Table 3).

#### Family stress in Smith–Magenis syndrome versus other aetiological groups

Figure 1 compares levels of Parent and Family Problems, Pessimism and Total QRS-F scores across various groups examined both by the present and other authors. In addition to comparisons with their studies of families of children with Prader–Willi and 5p- syndromes, the present authors also compared stress levels in Smith–Magenis families to published studies of young children with mixed aetiologies of intellectual disability.

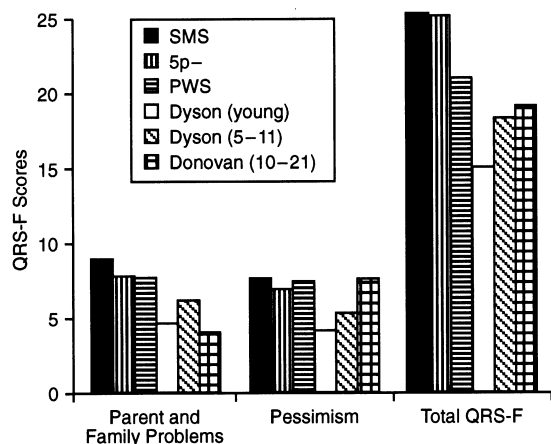
Compared to two studies of young children with mixed aetiologies of disabilities (Dyson 1991, 1993), the Smith–Magenis group showed markedly higher

**Table 3** Predictors of Questionnaire on Resources and Stress—Friedrich edition (QRS-F) total stress in Smith–Magenis, Prader–Willi and 5p- (*cri du chat*) syndromes

Predictor of QRS-F total stress	Percentage of variance accounted for
<i>Smith–Magenis syndrome</i> (n = 36; age = 8.33 years)	
(1) Friends	31.3
(2) Socialization standard score	13.2
<i>Prader–Willi syndrome</i> (n = 42; age = 10.3 years)*	
(1) Maladaptive Behaviour (CBCL)	25.0
<i>5p- (<i>cri du chat</i>) syndrome</i> (n = 99; age = 8.08 years)†	
(1) Maladaptive Behaviour (ABC)	38.2
(2) Communication Standard Score	4.4
(3) Chronological Age	3.3
(4) Number of Supporters	3.0

\*Hodapp *et al.* (1997b).

†Hodapp *et al.* (1997b).



**Figure 1** The Questionnaire on Resources and Stress—Friedrich (QSR-F) edition domain and total scores for families of children with Smith–Magenis syndrome (SMS), Prader–Willi syndrome (PWS), 5p- syndrome and children of mixed aetiologies: young, Dyson (1991); 5–11 years, Dyson (1993); and 10–21 years, Donovan (1988).

levels across all three domains of family stress, even when children with Smith–Magenis syndrome were equated in age to children in the Dyson studies. Examining only the families of children with Smith–Magenis syndrome aged below 7 years, the Parent–Family Problems score of families of children with Smith–Magenis syndrome (8.27) was above the score (4.60) for the families of Dyson’s (1991) children with mixed aetiologies younger than 7 years [ $t(68) = 2.98; P < 0.01$ ]; higher scores in the Smith–Magenis syndrome families also occurred on Pessimism [ $t(68) = 3.68; P < 0.001$ ] and on QRS-F total scores [ $t(68) = 3.74; P < 0.001$ ]. Examining only 5–11-year-olds, the families of children with Smith–Magenis syndrome showed more family stress than families of children with mixed aetiologies of the same-age (Dyson 1993) for Parent–Family Problems [ $t(57) = 2.71; P < 0.01$ ], Pessimism [ $t(57) = 2.50; P < 0.05$ ] and QRS-F total score [ $t(57) = 2.21; P < 0.05$ ]. No differences occurred when comparing families of adolescents with Smith–Magenis syndrome to families of older (mixed aetiology) children (from Donovan 1988).

## Discussion

As the first examination of families of children with Smith–Magenis syndrome, the present study found

that there are high levels of family stress and support, specific correlates of family stress, and levels of stress in Smith–Magenis families which sometimes differ from the levels found in other disabled groups. As such, these findings further the understanding of the ‘indirect effects’ of different genetic disorders and should lead to more finely targeted intervention techniques.

Confronted with children who show a variety of difficult behaviours and sleep problems, families of children with Smith–Magenis syndrome show high levels of family stress. Parent and family problems, pessimism and overall stress levels are all fairly high in these families, but so too are the number of family supporters. Such support comes from extended families, friends and even professionals. For example, consider the finding that a full 76% of families of children with Smith–Magenis syndrome listed one or more professionals in their list of supporters; in Prader–Willi syndrome, only 33% of families listed any professionals within their support system (Hodapp *et al.* 1997a).

Such support by family, friends and professionals seems particularly helpful to families of children with Smith–Magenis syndrome. Indeed, the main predictor of overall stress and of parent–family problems was the size of the family’s support system. The larger a family’s support system, the lower the levels of family stress. Only on parental pessimism did the family’s number of friends fail to independently account for significant amounts of variance. On both other stress measures, the number of family friends was the strongest single predictor of lower family stress.

In addition to a family’s number of friends, certain aspects of the child with Smith–Magenis syndrome were also associated with family stress. The degree of the child’s impairments in socialization was related to both parent–family problems and overall stress levels. Thus, children who showed greater delays in following rules, making friends and playing by themselves (the Vineland Socialization domain) had families with more parent–family problems and higher overall stress scores. Similarly, pessimism scores were higher in families in which children with Smith–Magenis syndrome showed higher CBCL maladaptive behaviour scores. Therefore, both a family’s social support and the specific

characteristics of the affected child related to family stress.

These correlates of family stress are both similar and different compared to those found in families of children with different aetiologies of intellectual disability. In Prader–Willi and 5p- (cri du chat) syndromes, the child's behaviour problems are the main correlate of familial stress. In Prader–Willi syndrome, temper tantrums, over-eating, skin-picking and other, non-food compulsive-like behaviour are common (Dykens *et al.* 1996; Dykens & Cassidy 1996), whereas children with 5p- syndrome show extremely high rates of hyperactivity (Dykens & Clarke 1997). In both cases, behaviour problems override the effects of the child's age, sex or degree of impairment as predictors of familial stress.

In Smith–Magenis syndrome, however, the picture is not so clear. Like children with Prader–Willi and 5p- syndromes, these children also show extremely high levels of maladaptive behaviour, mainly involving tantrums, sleep disturbance, toileting difficulties, and a combination of self-injurious and stereotypical behaviours (Smith *et al.* 1998a, b), but while maladaptive behaviour was the sole predictor of parental pessimism (accounting for 31% of the variance), maladaptive behaviour did not predict either parent–family problems or overall stress scores. Why the number of friends, in particular, should so strongly relate to two of the three stress measures remains unclear. Although the present authors can only speculate, friends may provide parents of children with Smith–Magenis syndrome a respite from child care duties (which may allow parents to catch up on sleep), or possibly, to confide their problems. Yet why friends are so strongly associated with lessened stress for families of children with Smith–Magenis syndrome, but not for similarly stressed families of children with Prader–Willi or 5p- syndromes, remains a question for future studies.

So too are there both similarities and differences present in the levels of familial stress in families of children with different disabilities. In studies of children with Prader–Willi (Hodapp *et al.* 1997a) and 5p- (cri du chat) syndromes (Hodapp *et al.* 1997a), the present authors found similarly high levels of familial stress to the Smith–Magenis families. At the same time, families of children with

these three disorders experience much greater stress levels than the families of children with mixed or unknown aetiologies (Dyson 1991, 1993). Even though these three aetiological groups each consisted of middle-class, married and well-educated samples, families experienced greater levels of stress in response to children with these three genetic aetiologies. Nevertheless, family stress is not a response to genetic aetiology *per se*: compared to families of children of the same age with other aetiologies, families of children with Down's syndrome generally display lower levels of familial stress (Hodapp 1996).

The present study also has several limitations which should be noted. Firstly, the present sample was recruited through PRISMS, the main US parent–professional group in Smith–Magenis syndrome. Like all parent groups, PRISMS is comprised primarily of middle class, well-educated and intact families. Although the high response rate in the present study argues against a large number of Smith–Magenis cases known to the parent group but not examined in this study, the subject group was relatively advantaged nevertheless. One might argue that stress levels in less-advantaged Smith–Magenis families might be even higher. Secondly, the present authors did not directly examine the families in face-to-face interviews, relying instead on postal questionnaires and telephone interviews. Although using neither method may be ideal, both parent-group families and telephone interviews constitute reasonable compromises when examining relatively rarely occurring syndrome across a large geographical area.

Finally, the practical implications of the findings of the present study should be emphasized. If parents and families of children with Smith–Magenis syndrome (but not of other syndromes) are helped, mostly by friends, these types of supportive relations might be fostered. Parent groups may play some role in this process; indeed, parent groups such as PRISMS might be especially important in newly discovered, less well-known syndromes where few parent-to-parent support networks exist. In terms of this type of family research, the job becomes determining what, exactly, are the operative mechanisms underlying family stress and support in Smith–Magenis and other syndromes. Only then can we implement more specific programmes of



intervention, programmes that are tailored directly to families of children with each specific aetiology.

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### References

- Achenbach T. M. (1991) *Manual for the Child Behavior Checklist—4-18—and 1991 Profiles*. University of Vermont, Burlington, VT.
- Beckman P. J. (1983) Influence of selected child characteristics on stress in families of handicapped infants. *American Journal of Mental Deficiency* **88**, 150–6.
- Beckman P. J. (1991) Comparison of mothers' and fathers' perceptions of the effect of young children with and without disabilities. *American Journal on Mental Retardation* **95**, 585–95.
- Cahill B. M. & Glidden L. M. (1996) Influence of child diagnosis on family and parental functioning: Down syndrome versus other disabilities. *American Journal on Mental Retardation* **101**, 149–60.
- Colley A. F., Leversha M. A., Voullaire L. E. & Rogers J. G. (1990) Five cases demonstrating the distinctive behavioral features of chromosome deletion 17 (p.11.2) (Smith–Magenis syndrome). *Journal of Pediatrics and Child Health* **26**, 17–21.
- Donovan A. (1988) Family stress and ways of coping with adolescents who have handicaps: maternal perceptions. *American Journal on Mental Retardation* **92**, 502–9.
- Dykens E. M. (1995) Measuring behavioral phenotypes: provocations from the 'new genetics.' *American Journal on Mental Retardation* **99**, 522–32.
- Dykens E. M. & Cassidy S. B. (1996) Prader–Willi syndrome: genetic, behavioral, and treatment issues. *Child and Adolescent Psychiatric Clinics of North America* **5**, 193–27.
- Dykens E. M. & Clarke D. J. (1997) Correlates of maladaptive behavior in persons with 5p- (cri du chat) syndrome. *Developmental Medicine and Child Neurology* **39**, 752–6.
- Dykens E. M. & Cohen D. J. (1996) Effects of Special Olympics International on social competence in persons with mental retardation. *Journal of the American Academy of Child and Adolescent Psychiatry* **35**, 223–9.
- Dykens E. M., Finucane B. M. & Gayley C. (1997) Cognitive and behavioral profiles in persons with Smith–Magenis syndrome. *Journal of Autism and Developmental Disorders* **27**, 203–11.
- Dykens E. M., Hodapp R. M., Walsh K. K. & Nash L. (1992) Adaptive and maladaptive behavior in Prader–Willi syndrome. *Journal of the American Academy of Child and Adolescent Psychiatry* **31**, 113–16.
- Dykens E. M. & Kasari C. (1997) Maladaptive behavior in children with Prader–Willi syndrome, Down syndrome, and non-specific mental retardation. *American Journal on Mental Retardation* **102**, 228–37.
- Dykens E. M., Leckman J. F. & Cassidy S. B. (1996) Obsessions and compulsions in Prader–Willi syndrome. *Journal of Child Psychology and Psychiatry* **37**, 995–1002.
- Dyson L. L. (1991) Families of young children with handicaps: parental stress and family functioning. *American Journal on Mental Retardation* **95**, 623–9.
- Dyson L. L. (1993) Response to the presence of a child with disabilities: parental stress and family functioning over time. *American Journal on Mental Retardation* **98**, 20718.
- Farber B. (1970) Notes about sociological knowledge about families with mentally retarded children. In: *Social Work and Mental Retardation* (ed. M. Schrieber), pp. 118–24. John Day, New York, NY.
- Finucane B. M., Konar D., Haas-Givner B., Kurtz M. D. & Scott L. I. (1994) The spasmodic upper-body squeeze: a characteristic behavior in Smith–Magenis syndrome. *Developmental Medicine and Child Neurology* **36**, 78–83.
- Frey K., Greenberg M. & Fewell R. (1989) Stress and coping among parents of handicapped children: a multi-dimensional perspective. *American Journal on Mental Retardation* **94**, 240–9.
- Friedrich W. N., Greenberg M. T. & Crnic K. (1983) A short-form of the Questionnaire on Resources and Stress. *American Journal of Mental Deficiency* **88**, 41–8.
- Greenberg F., Guzzetta V., de Oca-Luna R. M., Magenis R. E., Smith A. M., Richter S. F., Kondo I., Dobyns W. B., Patel P. I. & Lupski J. R. (1991) Molecular analysis of the Smith–Magenis syndrome: a possible contiguous-gene syndrome associated with del (17) (p.11.2). *American Journal of Human Genetics* **49**, 1207–18.
- Greenberg F., Lewis R. A., Potocki L., Glaze D., Parke J., Killian J., Murphy M. A., Williamson D., Brown F., Dutton R., McCluggage C., Friedman E., Sulek M. & Lupski J. (1996) Multi-disciplinary clinical study of Smith–Magenis syndrome (deletion 17p11.2). *American Journal of Medical Genetics* **62**, 247–54.
- Hodapp R. M. (1995) Parenting children with Down syndrome and other types of mental retardation. In: *Handbook of Parenting*, Vol. 1. *How Children Influence Parents* (ed. M. Bornstein), pp. 233–53. Erlbaum, Hillsdale, NJ.

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- Hodapp R. M. (1996) Down syndrome: developmental, psychiatric, and management issues. *Child and Adolescent Psychiatric Clinics of North America* **5**, 881–94.
- Hodapp R. M. (1997) Direct and indirect behavioral effects of different genetic disorders of mental retardation. *American Journal on Mental Retardation* **102**, 67–79.
- Hodapp R. M. (1999) Indirect effects of genetic mental retardation disorders: theoretical and methodological issues. *International Review of Research in Mental Retardation* **22**, in press.
- Hodapp R. M. & Dykens E. M. (1994) Mental retardation's two cultures of behavioral research. *American Journal on Mental Retardation* **98**, 675–87.
- Hodapp R. M., Dykens E. M. & Masino L. L. (1997a) Families of children with Prader–Willi syndrome: stress-support and relations to child characteristics. *Journal of Autism and Developmental Disorders* **27**, 11–24.
- Hodapp R. M., Wijma C. A. & Masino L. L. (1997b) Families of children with 5p- (cri du chat) syndrome: familial stress and sibling reactions. *Developmental Medicine and Child Neurology* **39**, 757–61.
- Holroyd J. (1974) The Questionnaire on Resources and Stress: an instrument to measure family responses to a handicapped family member. *Journal of Community Psychology* **2**, 92–4.
- Kazak A. (1987) Families with disabled children: stress and social supports in three samples. *Journal of Child Abnormal Psychology* **15**, 137–46.
- Margalit M., Shulman S. & Stuchiner S. (1987) Behavior disorders and mental retardation: the family system perspective. *Research in Developmental Disabilities* **10**, 315–26.
- Minnes P. M. (1988) Family stress associated with a developmentally handicapped child. *International Review of Research in Mental Retardation* **15**, 195–226.
- Scott R. L., Sexton D., Thompson B. & Wood T. A. (1989) Measurement characteristics of a short form of the Questionnaire on Resources and Stress. *American Journal on Mental Retardation* **94**, 331–9.
- Seltzer M. & Ryff C. (1994) Parenting across the lifespan: the normative and nonnormative cases. *Life-Span Development and Behavior* **12**, 1–40.
- Smith A. C. M., Dykens E. M. & Greenberg F. (1998a) Sleep disturbance in Smith–Magenis syndrome (del 17 p.11.2). *American Journal of Medical Genetics* **81**, 186–91.
- Smith A. C. M., Dykens E. M. & Greenberg F. (1998b) Behavioral phenotype of Smith–Magenis syndrome (del 17 p.11.2) *American Journal of Medical Genetics*, **81**, 179–85.
- Smith A. C. M., MaGavran L., Robinson J., Waldstein G., Macfarlane J., Zonona J., Reiss J., Llahr M., Allen L. & Magenis E. (1986) Interstitial deletion of (17) (p.11.2) in nine patients. *American Journal of Medical Genetics* **24**, 421–32.
- Sparrow S. S., Balla D. & Cicchetti D. V. (1983) *The Vineland Scales of Adaptive Behavior—Manual*. American Guidance Service, Circle Pines, MN.
- Sparrow S. S., Carter A. S. & Cicchetti D. V. (1995) Vineland Screener: overview, reliability, validity, administration, and scoring. Unpublished manuscript, Yale University.
- Stratton R. F., Dobyns W. B., Greenberg F., DeSana J. B., Moore C., Fidone G., Runge G. H., Feldman P., Sekhon G. S., Pauli R. M. & Ledbetter D. H. (1986) Interstitial deletion of (17) (p11.2). Report of six additional patients with a new chromosome deletion syndrome. *American Journal of Medical Genetics* **24**, 421–32.
- Wikler L. (1986) Periodic stresses in families of mentally retarded children: an exploratory study. *American Journal of Mental Deficiency* **90**, 703–6.

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