

Special Article

Parental Burden, Coping, and Family Functioning in Primary Caregivers of Children With Joubert Syndrome

Jennifer L. Luescher, BS; Duane E. Dede, PhD; Jill C. Gitten, BS; Eileen Fennell, PhD; Bernard L. Maria, MD, MBA

ABSTRACT

Children with Joubert syndrome have physical and intellectual disabilities. The purpose of this study was to assess the impact of Joubert syndrome on parental burden, coping, and family functioning. Forty-nine primary caregivers were surveyed. Forty-three primary caregivers were mothers and six were fathers; their mean age was 34 years. The following measures were used: Beck Depression Inventory, Child Development Inventory, Caregiver Strain Index, Family Assessment Device, and Ways of Coping Checklist-Revised. The data show that caregiver burden is not related to the severity of the child's illness, but that caregivers report significant burden. Higher burden was associated with the use of palliative coping methods, and family functioning was problematic. The results of this study suggest that for parents of children with Joubert syndrome, degree of parental burden depends more on the parents' coping skills and the level of family functioning rather than on the degree of the child's impairment. These findings highlight the importance of assessing caregiver burden, as well as decreased family functioning or coping abilities, since these problems often can be managed with psychologic intervention. (*J Child Neurol* 1999;14:642-648).

Joubert syndrome is a rare neurogenetic disorder first described in 1969 by Joubert et al.¹ Symptoms such as episodic tachypnea followed by apnea, nystagmus, oculomotor apraxia, hypotonia, ataxia, and developmental delays have been described.¹⁻⁵ The hypotonia and ataxia have been attributed to malformation of the cerebellar vermis.¹ The unusual respiratory pattern, abnormal eye movements, and severe mental retardation suggest more widespread central nervous system involvement, including the brain stem and possibly the cerebral hemispheres.¹ Recent studies have shown a variety of posterior fossa malformations, including the presence of the "molar tooth sign."⁶⁻⁹

Mental retardation, developmental delay, including low levels of expressive and receptive language, and motor delays have been reported in association with Joubert syn-

drome.^{7,10} We previously investigated the relationship between brain structure and neurobehavioral development. The data showed that 94% of subjects were severely impaired according to the Child Development Inventory.⁷ The average developmental age was 63% below chronologic age. Thus, children with Joubert syndrome have multiple physical and cognitive deficits that affect learning, communication, and ambulation. However, little is known about levels of distress in caregivers of such children.

The purpose of this study was to assess psychosocial parameters associated with Joubert syndrome. The first goal was to measure the level of caregiver burden experienced by parents of these children and to see if these burden scores correlated with the level of the child's impairment. It was expected that burden scores would correlate with the use of adaptive coping methods and that the families of children with Joubert syndrome would have disrupted family functioning. We hypothesized that burden was positively correlated with illness severity and negatively correlated with the use of adaptive coping methods, and that family dysfunction was significantly greater in families of children with Joubert syndrome than in families in the normative sample of the Family Assessment Device, especially in the areas of communication, problem solving, and roles.

Received March 24, 1999. Accepted for publication April 16, 1999.

From the Department of Clinical and Health Psychology (Ms Luescher, Ms Gitten, Drs Dede and Fennell) and the Department of Pediatrics (Dr Maria), University of Florida, Gainesville, FL.

Address correspondence to Dr Duane Dede, Department of Clinical and Health Psychology, University of Florida College of Health Professions, PO Box 100165, Gainesville, FL 32610-0165. Tel: 352-395-0294; fax: 352-0096; e-mail: ddede@hp.ufl.edu.

METHODOLOGY

Subjects

The participants were 49 primary caregivers of children diagnosed with Joubert syndrome. The sample included 32 boys and 17 girls diagnosed with the syndrome, ranging in age from 9 months to 17 years. The mean age was 5.34 years (standard deviation, 3.99 years). The sample consisted of 91.8% European-American, 4.1% Hispanic-American, 2.0% Asian-American, and 2.0% African-American children. The caregivers were 87.8% mothers and 12.2% fathers of children diagnosed with Joubert syndrome. Primary caregivers ranged in age from 24 to 52 years, with a mean of 34.0 years and a standard deviation of 6.66 years (Table 1). All parents were members of the Joubert Syndrome Foundation, a parent support group.

Procedure

Caregivers were recruited for this study either at a national Joubert syndrome conference or by phone from information supplied by a research registry. The Joubert Syndrome Foundation is an international network of more than 125 families of children with Joubert syndrome or other cerebellar abnormalities. The majority of families asked to participate returned assessment packets, either through the mail or at a conference.

Definitions and Measures

Parents completed a battery of questionnaires, including the Beck Depression Inventory, Caregiver Strain Index, Child Development Inventory, Family Assessment Device, Ways of Coping Checklist-Revised, and a demographic questionnaire.

Caregiver Burden

Burden is a multidimensional concept previously operationalized as the negative emotional, cognitive, physical, and financial consequences of providing care for others. Several studies of children with chronic disorders having multidimensional symptoms found that their parents experienced significant levels of distress, as measured by the Parental Stress Index or the Questionnaire on Resources and Stress.^{11,12} In this study, we used the Caregiver Strain Index, a 13-item questionnaire in a yes/no format (mean score, 3.519; SD, 3.497).¹³ The index reflects common stressors that occur during caregiving and taps multiple dimensions in which burden could be experienced (eg, physical health, emotional symptoms, and social activities). The Caregiver Strain Index has high internal

consistency ($\alpha = .86$). Content validity was examined and supported in relation to care recipients' characteristics, caregivers' subjective views of the caregiving situation, and the emotional health of the caregivers.

Research also has shown that elevated levels of caregiver burden increase the tendency for caregivers to become clinically depressed.^{14,15} To assess whether this occurs in Joubert syndrome, the Beck Depression Inventory was used. This inventory is a 21 Likert item self-report measure of depressive symptomatology.¹⁶ Scores of 9 or lower are rated nondepressed, 10 to 15 mildly depressed, 16 to 23 moderately depressed, and scores of 24 or more are considered severely depressed. The Beck Depression Inventory has well-established reliability and validity data.¹⁶

There are conflicting results in the literature on whether illness severity affects caregiver burden and family functioning. When comparing families of children with and without a handicap, the best predictor of parental stress has been found to be the presence of a handicap.^{17,18} Severity of the disorder appears to affect caregiver burden only in conditions that involve cognitive impairments (ie, autism) but not primarily physical disorders (ie, cystic fibrosis).^{14,19} As Joubert syndrome can be fatal and causes both cognitive and physical problems, it is probable that caregiver burden and family functioning would be related to illness severity in families of children with the disorder.

In this study, illness severity was measured with the Child Development Inventory (a 270-item true-or-false parental report measure). This inventory is an updated version of the Minnesota Child Development Inventory, which was originally designed to help identify and describe children with a variety of developmental problems.²⁰ The Child Development Inventory is made up of nine scales: Social, Self-Help, Gross Motor, Fine Motor, Expressive Language, Language Comprehension, Letters, Numbers, and General Development. For each scale, raw scores are converted to developmental age scores; from there, calculations are done to determine how far below chronologic age the child scored. According to the Child Development Inventory manual, the normal range extends to those scoring as much as 20% below chronologic age. Scores 20% to 30% below chronologic age are considered borderline. Scores greater than 30% below chronologic age are considered severely delayed.

Most research supports the conclusion that a child's age does not affect level of parental burden. Although it has been suggested that parental stress is likely to increase over time as children fail to obtain age-related milestones, stress does not appear to correlate directly with age.^{14,18,21,22}

Coping

The literature on coping strategies of caregivers and families dealing with chronic illness has predominately shown that the use of palliative coping methods (eg, wishful thinking, self-blame, and avoidance) is related to poor adjustment, increased depression and anxiety, and poor physical health; such techniques are used most often with stressors perceived to be uncontrollable.²³⁻²⁵ Furthermore, children and adults who use more adaptive, or problem-focused, coping styles (eg, building support systems, information seeking, cognitive restructuring) experience less distress and a lower incidence of depression and anxiety.^{25,26}

Table 1. Demographic Characteristics

Children	
Boys	32
Girls	17
Mean age in years	5.34 (3.99)
Race	
European-American	92%
Hispanic-American	4%
Asian-American	2%
African-American	2%
Primary caregiver	
Mother	87.8%
Father	12.2%
Mean primary caregiver age in years	34 (SD, 6.66)

Parental coping skills were measured using the Ways of Coping Questionnaire-Revised, which is based on the Lazarus transactional model of stress.²⁷ The questionnaire lists a range of both behavioral and cognitive coping strategies, which are rated on a four-point Likert scale ranging from "never used" to "regularly used." Scores from the Ways of Coping Checklist can be divided into five factors: avoidance, blaming self, problem focused, social seeking, and wishful thinking.

Family Functioning

When assessing problem areas within family functioning, studies of families with ill children that used the Family Assessment Device appeared more sensitive in detecting dysfunction than global family assessment measures, such as the Family Environment Scale.^{17,28,29} Studies using the Family Assessment Device have shown significant relationships between child disorders and dysfunction in communication, problem solving, and roles within the family. The Family Assessment Device is divided into six subscales: Affective Involvement, Affective Responsiveness, Behavior Control, Communication, Problem Solving, and Roles. There is also a General Functioning scale.³⁰ The Family Assessment Device is a paper and pencil measure consisting of 60 items that can be given to any family member older than age 12 years. The Family Assessment Device has adequate test-retest reliability, moderate correlations with other self-report measures of family functioning, and differentiates families rated by clinicians as healthy from dysfunctional families.³¹

For this study, Family Assessment Device subscale scores were standardized across the entire sample (mean, 0; SD, 1) according to the method described by Yeates et al.³² For both raw and converted scores, higher scores reflect worse family functioning.

All analyses were conducted using SPSS for Windows: Professional Statistics, 7.5. All missing data were replaced with series mean.

RESULTS

Family Characteristics

The mean number of people in each household was 4.15, with a standard deviation of 1.65. Of the parents, 83.7% were married, 10.2% were divorced, 4.1% were separated, and 2.0% were single. Thirteen families had more than one sick child. Data were analyzed from each family only one time. Thirty-nine of 49 primary caregivers (79.6%) said that someone helped them care for the child. Eleven (22.4%) of these families hired help to care for their child. Ten (20.4%) parents received help from someone within the household, while seven (14.3%) caregivers had help from family members outside of the home. Eleven (22.4%) families used combinations of these to care for their child. According to the demographic questionnaire, both primary and secondary caregivers spend the majority of their time caring for and sitting with the patient, as well as working (Table 2).

Caregiver Burden

Mean score on the Caregiver Strain Index was 19.65 (SD, 3.75), indicating significant burden. The caregivers' responses

Table 2. How Parents Spend Time

Activity	Hours Per Week
Patient care	16-21
Occupation	10-15
Sitting with patient	10-15
Housekeeping	9-10
Sitting with other children	6-9
Emotional support	5-6
Errands	5-6

indicated that feeling overwhelmed, physical strain, and family adjustment were the most stressful aspects of caring for a child with Joubert syndrome (Table 3). Mean Beck Depression Inventory score for primary caregivers was 8.19 (in the nondepressed range), with a standard deviation of 6.83. Of primary caregivers, 33 parents scored in the nondepressed range, 9 in the mildly depressed range, 6 in the moderately depressed range, and 1 in the severely depressed range. Pearson correlations conducted on the Caregiver Strain Index and Beck Depression Inventory were significant: $R = .419$; $P = .003$.

These scores support the hypothesis that parents of children with Joubert syndrome have higher levels of burden, but are not depressed. It also supports the results of earlier research, which found that caregivers often might not be clinically depressed, but that a significant correlation appears to exist between caregiver burden and depression. All parents appear to be burdened, but not all parents' burdens manifest themselves affectively.

Illness Severity and Caregiver Burden

Severity of illness was defined as the percentage below chronologic age, as measured by the Child Development Inventory. As a group, the children, whose mean chronologic age was 5.34, scored in the significantly delayed range on all Child Development Inventory subscales. The mean developmental age scores were as follows: Expressive Language, 20 months; Fine Motor, 18 months; General Development, 20 months; Gross Motor, 12 months; Letters, 3 years, 3 months; Language Comprehension, 21 months; Numbers, 2 years, 4 months; Self-Help, 15 months; and Social, 21 months (Table 4). Each score is at least 31% below that expected for age. The Numbers and Letters subscales had

Table 3. Caregiver Strain Index

Strain	Percentage of Primary Caregivers
Felt overwhelmed	69.4
Physical strain	65.3
Family adjustments	63.3
Changes in personal plans	63.3
Work adjustments	63.3
Confining	57.1
Financial strain	51.0
Other family demands	53.1
Patients' behavior upsetting	55.1
Inconvenient	44.9
Sleep disturbed	34.7
Upsetting to find patient changed	10.2

Table 4. Child Development Inventory

Subscale	Mean (SD)	Developmental Age, months
Expressive language	12.07 (14.87)	20
Fine motor skills	11.07 (8.77)	18
General development	19.55 (17.98)	20
Gross motor skills	6.48 (5.32)	12
Language comprehension	16.74 (15.52)	21
Letters	2.76 (4.17)	39
Numbers	3.83 (4.37)	28
Self-help	12.29 (9.86)	15
Social	16.50 (10.95)	21

the highest scores, but it should be noted that scores of 0 on these scales convert, respectively, to age 18 months and 2 years, 1 month. The area that shows the most impairment is the Gross Motor subscale, which would be expected in children with a cerebellar disorder such as Joubert syndrome. Using the converted developmental ages of the individual children of the General Development subscale of the Child Development Inventory, 93% of the children were severely impaired.

Pearson correlations conducted on the Caregiver Strain Index and each of the scales of the Child Development Inventory were not significant. Pearson correlations for the Beck Depression Inventory and subscales of the Child Development Inventory also were not significant. These results do not support our second hypothesis. The amount of parental distress is not related to illness severity. There was not a significant difference in scores on either the Beck Depression Inventory or Caregiver Strain Index for families with one ill child compared to families with more than one ill child.

In relating the child's age to the level of burden as measured by the Caregiver Strain Index, there was no relationship between age and Caregiver Strain Index score. The mean score for parents of children in all age groups was in a range from 19 to 21, indicating significant burden. In looking at age and the Beck Depression Inventory, the trend appears to show that depression scores increase for parents of children up to age 4. The highest level of depression is among parents of children ages 4 to 6 years. Finally, there is a decrease in the level of depression among parents of older children, with the lowest depression scores in the oldest age group, 12 years and above (Figure 1).

Caregiver Burden and Coping

The mean scores (and standard deviations) for the Ways of Coping Checklist subscales were as follows: Problem Focused (0.42); Social Seeking (0.70); Avoidance, .96 (0.47); Wishful Thinking, 1.43 (0.68); Blame Self, .92 (0.68) (Table 5).

Pearson correlations conducted on the Caregiver Strain Index and the Wishful Thinking subscale of the Ways of Coping Checklist for the primary caregivers were significant: $R = .27$; $P = .05$. The Pearson correlations for the Caregiver Strain Index and all the other subscales were not significant.

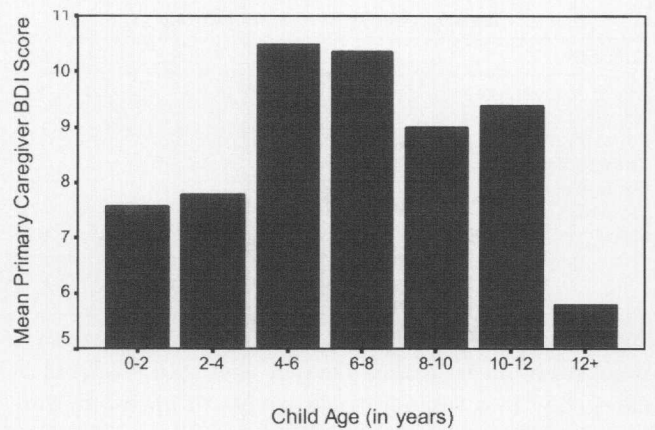


Figure 1. Depression levels in caregivers according to child age.

Pearson correlations conducted on the Beck Depression Inventory and the Avoidance, Blame Self, and Wishful Thinking subscales of the Ways of Coping Checklist for the primary caregivers were significant: $R = .50$, $P = .0002$, $R = .42$, $P = .003$, and $R = .58$, $P = .00001$. The Pearson correlations for the Beck Depression Inventory and other subscales of the Ways of Coping Checklist were not significant. These results support the hypothesis that level of parental burden is significantly related to coping skills. The level of burden is also significantly correlated to the parents' use of palliative coping skills, especially wishful thinking.

Family Functioning

The mean transformed score on the General Functioning Scale of the Family Assessment Device for primary caregivers was 2.17 (SD, 1.0), suggesting significant family dysfunction. The mean for the Roles subscale was 4.08 (SD, .9). The Communication subscale mean was 2.84 (SD, 1.0). The mean for the Problem Solving subscale was 2.54 (SD, 1.0). The mean score on the Affective Involvement scale was 2.06 (SD, 1.0). The mean score for the Behavior Control subscale was 2.01 (SD, 1.0). The mean score for the Affective Responsiveness scale was 1.88 (SD, 1.0) (Table 6).

Posthoc Analyses

We conducted a posthoc regression to explain why, although the parents showed increased levels of distress on the Caregiver Strain Index and did not show increased depression on the Beck Depression Inventory, their Caregiver Strain Index and Beck Depression Inventory scores were correlated. Schumacher and colleagues also found a

Table 5. Ways of Coping Checklist

Subscale	Mean	SD
Social seeking	2.26	0.70
Problem focused	2.04	0.42
Avoidance	0.96	0.47
Wishful thinking	1.43	0.68
Blame self	0.92	0.68

Table 6. Family Assessment Device

Subscale	Mean	SD
Affective involvement	2.06	1.0
Affective responsiveness	1.88	1.0
Behavior control	2.01	1.0
Communication	2.84	1.0
General scale	2.17	1.0
Problem solving	2.54	1.0
Roles	4.08	0.9

significant relationship between burden and depression in their study investigating dynamic and static aspects of caregiving in a sample of caregivers of chemotherapy patients.³³ We chose the following representative scales from our measures to enter into the regression equation to predict Caregiver Strain Index scores: Gross Motor and Social subscales of the Child Development Inventory, the Wishful Thinking subscale of the Ways of Coping Checklist, and the Global Functioning subscale of the Family Assessment Device. These factors explained 15% of the variance of the Caregiver Strain Index ($R^2 = .153$). For the Beck Depression Inventory regression, the Fine Motor and Social subscales of the Child Development Inventory, the Wishful Thinking subscale of the Ways of Coping Checklist, and the Global Functioning and Roles subscales of the Family Assessment Device were selected. These factors explained 43% of the Beck Depression Inventory scores ($R^2 = .434$). The variance of the Beck Depression Inventory scores explained by dynamic factors significantly exceeded the variance of the Caregiver Strain Index scores explained by these factors. Our results seem to agree with those of Schumacher et al that it is the more dynamic aspects of the caregiving situation, coping, and the family situation that are most important in predicting depression.

DISCUSSION

This study describes the largest sample of families (49 families) with children diagnosed with Joubert syndrome thus far in the literature. The data reveal that the parents are receiving help in caring for their children from a variety of sources, including family inside and outside of the home. The majority of families do not have more than one ill child. The results of the demographic questionnaire provide important information about how the parents incorporate child care, occupation, and household tasks into their daily lives. Both primary and secondary caregivers spend most of their time in caring for and sitting with the patient.

Results from the Caregiver Strain Index supported the hypothesis that caregivers of children with Joubert syndrome would have significantly elevated levels of burden. With a population mean of 3.519 (SD, 3.497), scores of 14 or more are considered a high burden on this scale; therefore, the mean score of 19.65 for primary caregivers shows that they are significantly burdened. The results measuring burden with the Beck Depression Inventory were not consistent with the findings of the Caregiver Strain Index. The

majority of primary caregivers scored in the nondepressed range of the Beck Depression Inventory. Although the majority of caregivers in this sample do not report symptoms of depression, 33% do report mild to severe depression. The point prevalence of dysthymia or depression in the general population ranges from 2% to 9%.³⁴ Therefore, our results indicate that a higher percentage of caregivers of children with Joubert syndrome report elevated depression levels compared to the general population. Although the results of the Caregiver Strain Index suggest that parents of children with Joubert syndrome are significantly distressed, the results of the Beck Depression Inventory do not support this finding. The parents do appear to be burdened; however, not all parents manifest their burden affectively. These contradictory results suggest that the Beck Depression Inventory is probably not an accurate measure of overall caregiver burden because depression is only one aspect of burden.

Although the primary caregivers did not have significantly elevated Beck Depression Inventory scores, the Caregiver Strain Index scores were significantly elevated. The Beck Depression Inventory and Caregiver Strain Index were significantly correlated, yet only the Beck Depression Inventory was significantly correlated with the coping measure. Schumacher et al found similar results in a study of family caregivers of persons receiving chemotherapy for cancer.³³ This study used the Caregiver Strain Index and used the depression subscale of the Profile of Mood States as its depression measure. They measured coping skills with a one-item overall assessment of the efficacy of the repertoire of coping strategies. The majority of caregivers were women and had a mean age of 43.8 years (SD, 14.7), similar to our caregivers; however, the patients did not match our patient sample because the majority of their patients were women and had a mean age of 47.4 years (SD, 14.5). The authors found their sample to have significant levels of burden and low levels of depression, similar to our sample. They also found a significant correlation between burden and depression. In conducting regression analyses, the authors found that the majority of burden, as measured by the Caregiver Strain Index, was accounted for by social background and context and disease-related stressors. They found that coping and social support explained more of the variance in depression. The authors concluded that strain was most related to fixed characteristics of the caregiving situation, while the dynamic factors of coping and social skills were more related to depression. The authors further stated that while burden might be an innate characteristic of caregiving, depression could be moderated more easily with successful coping skills and social supports. This suggests that while burden itself can be difficult to treat, depression in primary caregivers of chronically ill patients can be ameliorated by increasing adaptive coping skills and social skills.

The results of the Child Development Inventory demonstrate that the majority of children with Joubert syndrome are significantly delayed and that the greatest delays are in the areas of gross motor, self-help, and fine motor skills. These results are expected in children with cerebellar mal-

formations. Correlations between the measures of burden and child impairment were not significant, suggesting that child impairment level cannot predict parental burden. This is not what was expected from the results of earlier studies. In previous studies, severity of disorders involving cognitive impairments was correlated with levels of parental burden. The results of this study suggest that the degree of burden in parents of children with Joubert syndrome is more dependent upon the parents' coping skills and the level of family functioning than on the child's degree of impairment. Degree of parental burden also does not appear to be related to whether or not the family has more than one child with Joubert syndrome. The properties of the child's illness itself do not appear to determine the parents' level of burden; rather, parental functioning seems to be determined by the parents' and family's ability to adjust to the presence of chronic illness. This is encouraging in that mental health professionals potentially can improve skills in these areas, while characteristics of the child's illness remain inflexible.

Problem-focused and social-seeking strategies were found to be used significantly more often by caregivers than avoidance, wishful thinking, and self-blaming coping skills. Avoidance and self-blaming coping skills were used rarely by caregivers. These results are positive in that problem solving and social-seeking strategies are adaptive coping strategies that are associated with better adjustment than are palliative coping methods (avoidance, wishful thinking, and self-blame). The results demonstrate that the use of palliative coping methods is positively correlated with caregiver burden. The Caregiver Strain Index score for primary caregivers correlates with scores on the Wishful Thinking subscale of the Ways of Coping Checklist. The Beck Depression Inventory score correlates with the Avoidance, Blame Self, and Wishful Thinking subscales for primary caregivers. These results support previous findings in the literature that palliative coping methods are associated with higher levels of burden; that is, if the parents had better coping skills they would be less burdened and less depressed.

Standardized scores above 2 for the Family Assessment Device suggest family dysfunction. Elevated scores were found for all subscales except Affective Responsiveness. The highest scores were on the Roles, Communication, and Problem Solving subscales, as expected based on previous research. Therefore families of children with Joubert syndrome have increased levels of communication, problem solving, and role difficulties. This implies that families make significant adjustments in their duties and are often overwhelmed by these demands. In previous studies of families with healthy and developmentally delayed children, positive family functioning has been associated with decreased parental stress. Important family characteristics for adapting to crisis, such as caring for an ill child, include cohesion, integration, and adaptability.³⁵⁻³⁷ These characteristics, as well as

family expression of emotions and supportiveness, have been shown to be related to better psychologic adjustment.^{24,25}

In summary, parents of children with Joubert syndrome have increased burden. Furthermore, this burden is associated with the use of palliative coping methods and problems in family functioning. It is not surprising that coping relates to burden. As Thompson and coworkers indicated, uncontrolled stressors, such as illness, are related to palliative coping.²⁵ Coping style and family functioning contribute more to the parents' level of burden than does the level of the child's functioning. These findings will be very helpful in directing counseling for families of children with Joubert syndrome by helping the parents to obtain adaptive coping styles and improve family functioning in the areas of communication, problem solving, and roles. These results are encouraging because, while the child's level of impairment is difficult to change, coping skills can be taught and improved in parents. Also, while improving coping skills will be helpful in dealing with parental burden, improved skills also will reduce the caregivers' susceptibility to depression.

References

1. Joubert M, Eisenring JJ, Robb JP, Andermann F: Familial agenesis of the cerebellar vermis: A syndrome of episodic hyperpnea, abnormal eye movements, ataxia, and retardation. *Neurology* 1969;19:813-825.
2. Bolthausen E, Isler W: Joubert syndrome: Episodic hyperpnea, abnormal eye movements, retardation and ataxia, associated with dysplasia of the cerebellar vermis. *Neuropediatrics* 1977;8:57-66.
3. Kendall B, Kingsley D, Lambert SR, et al: Joubert syndrome: A clinico-radiological study. *Neuroradiology* 1990;31:502-506.
4. Holroyd S, Reiss AL, Bryan RN: Autistic features in Joubert syndrome: A genetic disorder with agenesis of the cerebellar vermis. *Biol Psychiatry* 1991;29:287-294.
5. Lewis SM, Roberts EA, Marcon MA, et al: Joubert syndrome with congenital hepatic fibrosis: An entity in the spectrum of oculo-encephalo-hepato-renal disorders. *Am J Med Genet* 1994;52:419-426.
6. Maria BL, Hoang KB, Tusa RJ, et al: "Joubert syndrome" revisited: Key ocular motor signs with magnetic resonance imaging correlation. *J Child Neurol* 1997;12:423-430.
7. Gitten J, Dede D, Fennell E, et al: Neurobehavioral development in Joubert syndrome. *J Child Neurol* 1998;13:391-397.
8. Maria BL, Rosainz LC, Nufeld JA, et al: "Molar tooth" sign in Joubert syndrome, abstract. *Ann Neurol* 1998;44:576.
9. Maria BL, Quisling RG, Rosainz LC, et al: "Molar Tooth" sign in Joubert syndrome: Clinical, radiologic, and pathologic significance. *J Child Neurol* 1999;14:368-376.
10. Steinlin M, Schmid M, Landau K, Boltshausen E: Follow-up in children with Joubert syndrome. *Neuropediatrics* 1997;28:204-211.
11. Fuller GB, Rankin RE: Differences in levels of parental stress among mothers of learning disabled, emotionally impaired, and regular school children. *Percept Mot Skills* 1994;78:583-592.
12. Hodapp RM, Dykens EM, Masino LL: Families of children with Prader-Willi syndrome: Stress-support and relations to child characteristics. *J Autism Dev Disord* 1997;27:11-24.
13. Robinson BC: Validation of a caregiver strain index. *J Gerontol* 1983;38:344-348.
14. Walker LS, Van Slyke DA, Newbrough JR: Family resources and stress: A comparison of families of children with cystic fibrosis,

- diabetes, and mental retardation. *J Pediatr Psychol* 1992;17:327-343.
15. Carpiniello B, Piras A, Pariante CM, et al: Psychiatric morbidity and family burden among parents of disabled children. *Psychiatr Serv* 1995;46:940-942.
 16. Beck AT, Ward CH, Mendelson M, et al: An inventory for measuring depression. *Arch Gen Psychiatry* 1961;4:561-570.
 17. Dyson LL: Families of young children with handicaps: Parental stress and family functioning. *Am J Ment Retard* 1991;95:623-629.
 18. Dyson LL: Response to the presence of a child with disabilities: Parental stress and family functioning over time. *Am J Ment Retard* 1993;98:207-218.
 19. Bouma R, Schweitzer R: The impact of chronic childhood illness on family stress: A comparison between autism and cystic fibrosis. *J Clin Psychol* 1990;46:722-730.
 20. Ireton HB: *Child Development Inventory Manual*. Minneapolis, Behavior Science Systems, 1992.
 21. Flynt SW, Wood TA, Scott RL: Social support of mothers of children with mental retardation. *Ment Retard* 1992;30:233-236.
 22. Beckman PJ: Comparison of mothers' and fathers' perceptions of the effect of young children with and without disabilities. *Am J Ment Retard* 1991;95:585-595.
 23. Frey KS, Greenberg MT, Fewell RR: Stress and coping among parents of handicapped children: A multidimensional approach. *Am J Ment Retard* 1989;94:240-249.
 24. Sloper P, Knussen C, Turner S, Cunningham C: Factors related to stress and satisfaction with life in families of children with Down's syndrome. *J Child Psychol Psychiatry* 1991;32:655-676.
 25. Thompson RJ Jr, Gil KM, Gustafson KE, et al: Stability and change in psychological adjustments of mothers of children and adolescents with cystic fibrosis and sickle cell disease. *J Pediatr Psychol* 1994;19:171-188.
 26. Hatton DL, Canam C, Thorne S, Hughes AM: Parents' perceptions of caring for an infant or toddler with diabetes. *J Adv Nurs* 1995;22:569-577.
 27. Vitaliano PP, Russo J, Carr JE, et al: The Ways of Coping Checklist: Revision and psychometric properties. *Multivariate Behav Res* 1985;20:3-26.
 28. Hamlett KW, Pellegrini DS, Katz KS: Childhood chronic illness as a family stressor. *J Pediatr Psychol* 1992;17:33-47.
 29. Oates RK, O'Toole BI, Lynch DL, et al: Stability and change in outcomes for sexually abused children. *J Am Acad Child Adolesc Psychiatry* 1994;33:945-953.
 30. Epstein NB, Baldwin LM, Bishop DS: The McMaster Family Assessment Device. *J Marital Fam Ther* 1983;9:171-180.
 31. Miller IW, Bishop DS, Epstein NB, et al: The McMaster Family Assessment Device: Reliability and validity. *J Marital Fam Ther* 1984;11:345-356.
 32. Yeates KO, Taylor HG, Drotar D, et al: Preinjury family environment as a determinant of recovery from traumatic brain injuries in school-age children. *J Int Neuropsychol Soc* 1997;3:617-630.
 33. Schumacher KL, Dodd MJ, Paul SM: The stress process in family caregivers of persons receiving chemotherapy. *Res Nurs Health* 1993;16:395-404.
 34. American Psychiatric Association: *Diagnostic and Statistical Manual of Mental Disorders*, 4th ed. Washington, DC, American Psychiatric Association, 1994.
 35. Olson DH, Sprenkle DH, Russel CS: Circumplex model of marital and family system: I. Cohesion and adaptability dimensions, family types, and clinical applications. *Fam Process* 1979;18:3-28.
 36. McCubbin HI, Joy CB, Cauble AE, et al: Family stress and coping: A decade in review. *J Marital Fam Ther* 1980;42:855-871.
 37. Nihira K, Meyers CE, Mink IT: Home environment, family adjustment, and the development of mentally retarded children. *Appl Res Ment Retard* 1980;1:5-24.



The 22nd Annual Carrell-Krusen Symposium

A Call for Abstracts

Abstract Deadline: Nov. 19, 1999

The 22nd Annual Carrell-Krusen Symposium, to be held Feb. 23-25, 2000, at Texas Scottish Rite Hospital for Children in Dallas, focuses on the treatment of neuromuscular disease and changes in current clinical practice. Guest lecturer will be Arthur K. Asbury, M.D., Van Meter Professor of Neurology Emeritus, Department of Neurology, University of Pennsylvania School of Medicine, Hospital of the University of Pennsylvania.

Abstracts for submission should be prepared on a single sheet of plain white paper. Place the complete title, in upper case, on the first line followed by the name and city location of each author underneath. Limit abstract titles to 65 characters. Skip one line and indent three spaces to begin abstract text. Abstracts must be double-spaced and one paragraph in length, with a maximum of 300 words. At the bottom of the page, give name, academic and position titles, mailing address and phone and fax numbers of the presenting author. Mail original, 10 copies and a computer disk labeled with the software package and file format to: Susan T. Iannaccone, M.D., Department of Neurology, Texas Scottish Rite Hospital for Children, 2222 Welborn Street, Dallas, TX 75219, or call 214/559-7830 for information. Accepted abstracts will be published in the *Journal of Child Neurology* and must not have been presented or published before the meeting.

A cover letter included with the abstract and signed by all authors must contain the following text: "The author(s) has (have) read and agree with the content of this abstract submitted for the 2000 Carrell-Krusen Symposium and warrant(s) the material is (1) original work of the author(s), (2) does not violate my copyright proprietary or personal rights of others, (3) is factually accurate and contains no matter libelous or otherwise unlawful, (4) has not been, nor will be, published or presented elsewhere prior to the 1998 Carrell-Krusen Symposium, and (5) hereby transfers, assigns or otherwise conveys all copyright ownership of this abstract to the *Journal of Child Neurology* and B.C. Decker, Inc. In addition, the author(s) agree(s) to acknowledge all commercial support for options, royalties, consulting fees and honoraria for speaking material support and other financial arrangement(s) with the manufacturer(s) of any commercial product or service relating to the abstract by any author has been described fully in this cover letter."

The University of Texas Southwestern Medical Center at Dallas, the accredited sponsor, is jointly sponsoring this program with Texas Scottish Rite Hospital for Children.

For more information call: 214/559-7830.