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The impact of Prader–Willi syndrome on the family's quality of life and caregiving, and the unaffected siblings' psychosocial adjustment

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Abstract

Background Prader–Willi syndrome (PWS), a complex multisystem genetic disorder, is characterised by developmental abnormalities leading to somatic and psychological symptoms. Symptoms of PWS include infantile hypotonia and failure-to-thrive, followed by life-long hyperphagia, developmental delays and moderate-to-severe behavioural problems and several physical problems that impact health. This study examined the effects of caring for a child diagnosed with PWS on the mothers and unaffected siblings. We assessed overall family functioning, the mothers' psychological health, the psychosocial and behavioural functioning of siblings, and the quality of life of siblings.

Methods Participants included 12 mothers and 13 siblings of a child with genetically confirmed PWS. Self-report measures administered to the mothers evaluated overall family functioning (PedsQL – Family Impact Module), the mothers' psychological health (Brief Symptom Inventory), and the mothers' perception of the sibling's quality of life (PedsQL – Parent Proxy). Self-report measures administered to the siblings evaluated their perceived quality of life (PedsQL) and symptoms of post-traumatic stress disorder (PTSD) (UCLA PTSD Index).

Results Families/mothers/siblings with children with PWS showed poorer perceived quality of life compared with general inpatient and outpatient samples of children with complex health conditions. Families/mothers/siblings with children with PWS reported difficulties in family functioning, communication problems, and an increased number of conflicts. They appeared to be experiencing significant behavioural distress symptoms, with higher than average levels of depression and feelings of isolation, anger and worry. Ninety-two per cent of the siblings indicated moderate-to-severe symptoms of PTSD. Compared with normative populations, siblings demonstrated poorer quality of life, with mothers perceiving more significant deficits in the sibling.

Conclusion This study reaffirms that PWS affects the entire family system. Mothers and siblings would benefit from psychosocial support due to the multiple challenges of living with and caring for a child/young adult with PWS.

Keywords family impact, parent distress, Prader–Willi syndrome, psychosocial functioning, quality of life, sibling effects

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Note: Due to an oversight, the names of the co-authors were omitted from this article. The correct author list should be M. M. Mazaheri, R. D. Rae-Seebach, H. E. Preston, M. Schmidt, S. Kountz-Edwards, N. Field, S. Cassidy and W. Packman; the article was wrongly attributed to M. M. Mazaheri, R. D. Rae-Seebach, H. E. Preston, M. Schmidt, N. Field, S. Cassidy and W. Packman. This error has been corrected in this version of the article on 22 October 2012 after first publication online on 12 October 2012.

Introduction

Prader–Willi syndrome (PWS) is a multisystem genetic disorder that includes hypotonia causing poor feeding and failure to thrive in infancy followed by developmental delay and intellectual disability, a characteristic behavioural, social and psychiatric pattern, short stature and growth hormone insufficiency, hypogonadism causing small genitalia, cryptorchidism, and incomplete pubertal development, an insatiable appetite leading to obesity if uncontrolled externally, and an increased risk for numerous other medical problems including strabismus, scoliosis, hip dysplasia, seizures, gastrointestinal and skin problems (Prader *et al.* 1956; see Cassidy *et al.* 2011 for review). It is the most common genetic syndrome associated with potentially life-threatening obesity. Estimated to affect 350 000–400 000 individuals worldwide, it has a reported prevalence of 1 in 10 000 to 30 000; it affects both sexes and all races and geographic areas (Whittington *et al.* 2001; Vogels *et al.* 2004). The cause of PWS is complex and related to genomic imprinting; it involves absence of expression of genetic information on the chromosome 15 contributed by the father due to one of three genetic mechanisms (see Cassidy & Driscoll 2009 for review). An individual with PWS is almost always the only affected person in the family.

Consensus clinical diagnostic criteria have been delineated (Holm *et al.* 1993) and diagnostic testing is standard to avoid misdiagnosis (Gunay-Aygun *et al.* 2001). The multiple physical, developmental and behavioural issues of PWS require families to devote considerable time and effort to the care of affected individuals. Management of these manifestations is symptomatic and supportive, and has been discussed in several recent reviews (Eiholzer & Lee 2006; Goldstone *et al.* 2008; Cassidy & Driscoll 2009; Cassidy & McCandless 2010; Cataletto *et al.* 2011; McCandless 2011). The intellectual disability is distinctive, including specific strengths (especially visual-spatial and long-term memory) and weaknesses (most commonly sequential and abstract thinking, short-term memory) that require an individualised approach (Curfs & Fryns 1992; Dykens *et al.* 1992; Roof *et al.* 2000; Whittington *et al.* 2004; Copet *et al.* 2010). However, the most difficult aspects of the disorder for most families include the insatiable appetite and the characteristic behavioural disturbance.

People with PWS have dysregulation of their appetite-satiety patterns leading to continuous interest in eating (hyperphagia). The cause of the hyperphagia is as yet poorly understood, but is believed to be hypothalamic (see McAllister *et al.* 2011 for review). The individual's interest in food varies radically with age, from lack of interest and sucking abnormalities in early infancy to excessive eating beginning at several years of age (Miller *et al.* 2011). For most of the lifetime there is typically excessive food seeking, eating of unappetising food, hording of food, and stealing food or money to buy food. Thus, initially parents need to be very concerned with maintaining adequate nutrition and growth, and later they are required to assure a very low calorie diet, ample exercise and very limited access to food with constant supervision. If hyperphagia is not externally controlled, morbid obesity with its attendant complications including cardio-pulmonary problems, obstructive sleep apnoea, diabetes mellitus, and chronic oedema will typically result. The early feeding problems and later hyperphagia are potential causes of stress for families (Hodapp *et al.* 1997a; Whittington & Holland 2010).

A distinctive maladaptive behaviour disorder is also characteristic of PWS. Starting shortly after the onset of hyperphagia, affected children are at risk for emotional lability leading to temper outbursts, stubbornness, compulsive and ritualitbursts occur most often when expstic behaviour. Temper ouectations about food are not met or when routines are changed (Whittington *et al.* 2004).

Treatment and family adjustment

Managing symptoms of PWS often requires modifications in the entire family's daily routine, often including round-the-clock supervision, locking of food cabinets and refrigerators, monitoring and regulating food intake, accommodating cognitive delays and behavioural problems, and appropriately responding to, managing and coping with emotional outbursts.

Beyond environmental and behavioural treatments, selective serotonin reuptake inhibitors and atypical antipsychotics have been used with children with PWS (Soni *et al.* 2007). While known to be

effective in affective and/or psychotic disorders, research into their effectiveness in PWS is limited (Whittington & Holland 2010). No specific medication has been found to be uniquely helpful to all individuals with PWS (Butler *et al.* 2006), and taking psychoactive medications at such a young age comes with many risks.

Growth hormone therapy has become standard of care in most developed countries as it has been shown to improve height, body composition, energy levels and physical activity, and thereby normalise appearance and help prevent morbid obesity (McCandless 2011 for review). This requires daily injections and frequent endocrine evaluations. The multiple aspects of medical management for other manifestations of PWS have recently been summarised (Cassidy & Driscoll 2009; McCandless 2011).

Influences on mothers

Multiple studies have indicated that parents/caregivers are at an increased risk for experiencing emotional distress, depression and anxiety when a child/young adult has health concerns (Curfs & Fryns 1992; Sarimski 1997; van Lieshout *et al.* 1998). Hodapp *et al.* (1997a) examined stress-support in 42 families of children with PWS and found that the level of parental stress and pessimism associated with PWS was higher than that in families of children with intellectual disabilities of mixed aetiologies. They noted that parents of children with PWS exhibited higher levels of 'Parent and Family Problems' as demonstrated by high scores on the Freidrich-Stress Questionnaire.

Sarimski (1997) sampled parents of children diagnosed with three genetic syndromes: PWS ($n = 35$), Fragile-X ($n = 30$) and Williams ($n = 35$) with children between the ages of 1 and 12. Using the Society for the Study of Phenotypes Postal Questionnaire, the Parenting Stress Index and the Family Functioning Style Scale, Sarimiski found a high degree of parental stress in the PWS group. van Lieshout *et al.* (1998) also found that parents of children with PWS ($n = 39$) reported more anger, marital conflict, and an overall increase in family stress, than parents of children with Fragile-X syndrome ($n = 32$) and Williams syndrome ($n = 28$).

In a related vein, when using the Rutter Malaise inventory, an instrument that looks at psychological distress or depression, Whittington *et al.* (2004) found that 35% of professional caregivers and 26.6% of parents caring for children with PWS reported 10 or more symptoms. These percentages are considerably high when compared with caregivers of children with other disorders, as research suggests that caregiver depression for individuals with learning disabilities is roughly 3.7% (Einfeld & Tonge 1996).

Psychosocial adjustment of siblings

Siblings of individuals with chronic conditions are considered 'a population at risk,' yet there is a lack of consensus about their psychological adjustment (McKeever 1983, p. 210). Healthy children with a chronically ill sibling are at an increased risk of developing emotional and behavioural problems (Beiser *et al.* 2010). In addition, research has demonstrated that psychological functioning, peer activities and cognitive development scores were lower for siblings of children with a chronic illness compared with controls (Sharpe & Rossiter 2002). A review of studies on sibling adjustment to childhood cancer found siblings to have higher levels of anxiety and post-traumatic stress, more behavioural adjustment problems and problems in social competence (Houtzager *et al.* 1999; Sharpe & Rossiter 2002; Cox *et al.* 2003). Additionally, parents reported more school-related problems, with increased absenteeism, and lack of self-confidence than the child reports. In a related vein, an investigation by Hodapp *et al.* (1997b) examined family stress and sibling reactions in families of children with Cri du Chat syndrome. In one part of the study, sibling concerns were examined in 44 unaffected siblings. The major finding was that parents and siblings disagreed on the extent of the siblings' interpersonal concerns. Parents reported that the siblings felt ignored and misunderstood, whereas the siblings themselves rated these concerns at a much lower level.

In a study by Williams *et al.* (2010), secondary data analysis was performed using qualitative data gathered during the baseline of a randomised controlled clinical trial of an intervention for siblings/families of children with long-term conditions, including developmental disabilities. Content analysis identified themes from responses of 151 parents

to an open-ended question on their perceptions of the effects on well siblings of living with a brother/sister with developmental disabilities. Of 363 themes identified, 61.1% reflected negative manifestations of increased risk in well siblings; 1.7% indicated no risk; and 37.2% reflected positive outcomes, suggesting the continued need for potential interventions.

On the other hand, in a survey of 24 unaffected siblings of children with PWS, Waters (1996) reported that siblings did not solely experience negative symptoms. Siblings were willing to help others, developed a greater sense of humour, and thought that having someone in the family with PWS brought the family closer together. Similarly, in a meta-analysis, siblings of children with a chronic condition displayed better psychological functioning, cognitive development, and peer interaction, regardless of the severity of their siblings' illness (Sharpe & Rossiter 2002).

In the current study, we compared families/mothers/siblings with children with PWS to children with chronic conditions. It is uncommon to compare those with a disorder marked by intellectual disability and chronic physical problems to patients with chronic conditions generally. This important and innovative aspect of PWS has not been emphasised before. Intellectual cognitive disability is just one aspect of this complex and chronic condition and it may be difficulty for some families.

Siblings' post-traumatic stress reaction

Living with a chronically ill child can be stressful and distressing. Several studies have reported post-traumatic stress reactions in siblings of children with cancer (Fine 2004; Packman *et al.* 2004) and siblings of chronically ill children (Wasik 2002). Packman *et al.* 1997 reported moderate-to-severe symptoms of post-traumatic stress in siblings of paediatric bone marrow transplant patients. Individuals with PWS are constantly seeking food, so that the home environment must control food access (such as locking of refrigerators and kitchen cabinets), and those with PWS often have severe temper tantrums, are at risk of running away, or have severe compulsive symptoms. Given the broad literature on post-traumatic stress disorder (PTSD) and the many stressors of living with a sibling with these problems, disturbances and life events that can lead to PTSD, it is reasonable and innovative to ask whether PTSD is present in the siblings of individuals with PWS.

Present study

The goal of this study was to better understand the functioning of families/mothers/siblings with children with PWS through determining the level of distress experienced by the parents, the quality of life of the parents, siblings and family unit as a whole, and the siblings' level of distress. In order to achieve this goal, the following research questions were addressed:

1 Do families that have a child/young adult with PWS show significant differences in their overall family functioning as compared with other diagnostic groups or families with children without chronic conditions?

2 Does raising a child/young adult diagnosed with PWS affect the parent's psychological health, as evidenced by their self-reported level of distress?

3 How is the psychosocial-emotional and behavioural functioning of the sibling impacted by living with a child/young adult diagnosed with PWS?

4 What is the parent's perception of the psychosocial adjustment of the sibling, and how does the parent's perception relate to the sibling's report?

Methods

Participants

Participants included 12 families (12 mothers and 13 siblings) of a child/young adult diagnosed with PWS. All of the affected individuals had a genetically confirmed diagnosis of PWS. The majority (11/13) were diagnosed with PWS in the first two years of life, and the entire group ranged from 1–27 years of age ($\mu = 12.1$). All of the reporting parents were mothers, and 8 of 13 siblings were sisters. Approximately 72.7% of mothers reported being homemakers, with 54.5% leaving prior occupations to care for the child diagnosed with PWS. All families were intact, two-parent households, and the participants were geographically diverse. They were recruited through a variety of resources including

advertisements in PWS association state chapter newsletters, list-serves, conferences, and the Genetic Medicine Central California PWS clinic in Fresno, California directed by one of us (SBC) (see Table 1).

Measures

Brief Symptom Inventory

The Brief Symptom Inventory (BSI) (Derogatis 1993) was used to assess each mother's psychological distress symptoms using the Positive Symptom Total (PST), the Global Severity Index (GSI) and the Positive Symptom Distress Index (PSDI). The measure asks each participant to rate 53 items on a 5-point Likert scale of distress. The BSI has moderate test-retest reliability, strong convergent validity and strong internal consistency, with alpha coefficients ranging from 0.71 (psychoticism factor) to 0.85 (depression factor) (Derogatis 1993).

The UCLA PTSD Index for DSM-IV, Child Version

The UCLA PTSD Index for DSM IV, Child Version (Pynoos *et al.* 1998) was used to measure symptoms of post-traumatic stress reaction in the siblings. The UCLA index is a 20-item scale designed to screen for symptoms of post-traumatic stress associated with exposure to traumatic experiences. The UCLA index is psychometrically sound (Rodriguez *et al.* 2001a, 2001b) and indices are keyed to DSM-IV criteria (traumatic event, re-experiencing symptoms, avoidance symptoms, increased arousal symptoms). Responses are based on a 5-point Likert scale (0 = None; 1 = little; 2 = some; 3 = much; 4 = Most). Scoring includes an overall score for severity of PTSD symptoms as well as scores related to the DSM-IV indices. The score sheet provides instructions for calculating a total PTSD severity score, and severity scores for each of the DSM-IV B, C and D subcategories. When criterion A is met, children who meet criteria B, C and D (using endorsements of 'much of the time' and 'most of the time' as indicating symptom presence) are scored as having a likely diagnosis of DSM-IV 'full' PTSD. Where criterion A is met, children meeting criteria for only two symptom subcategories are scored as 'partial' PTSD likely. In the current study, an analysis of siblings' endorsement for each

Table 1 Demographic and background information parents and unaffected siblings

	Parents of children with PWS (*n* = 11)
Variable	**Per cent**
Gender of participants	
Female	100.0
Male	0.0
Age of mother (years)	
30–40	27.2
41–50	58.3
51–55	16.6
Age of father (years)	
30–40	18.1
41–50	72.7
51–55	16.6
Ethnicity	
Latino	8.3
Asian	8.3
Caucasian	83.3
Education of mother	
High school diploma or equivalent	18.2
1–3 years college	27.3
4 or more years of college	18.2
2 or more years graduate school	36.4
Education of father	
High school diploma or equivalent	27.2
1–3 years of college	18.2
4 or more years of college	18.2
2 or more years graduate school	36.4
Mother's occupation	
Homemaker	72.7
Work outside home	27.3
Father's occupation	
Work outside home	100.0
Family income	
$26 000–40 999	18.2
$41 000–60 999	27.3
$61 000–80 999	18.2
$101 000–200 000	18.2
$200 000+	18.2
Family structure	
Two-parent home, first marriage	100.0
	Unaffected siblings (*n* = 13)
Variable	**Per cent**
Gender	
Female	61.5
Male	38.5
Race	
Latino	9.1
Asian	9.1
Caucasian	81.8
Age range	
12–19 years	
Age of sibling at diagnosis with PWS (years)	
0–4	16.7
5–10	33.3
11–15	16.7
16–20	16.7
21–25	8.3
26–30	8.3
Variable	**PWS child data (*n* = 12)**
Age of child at diagnosis	
At birth	44.4
1 month to 2 years	44.1
2–5 years	11.1

PWS, Prader–Willi syndrome.

item on the PTSD scale was conducted. When criterion A was met, siblings who met criteria B, C and D (using endorsements of 'much of the time' and 'most of the time' as indicating symptom presence) were scored as having moderate-to-severe symptoms of post-traumatic stress reaction. Where criterion A was met, siblings meeting criteria for only two symptom subcategories were scored as reporting mild symptoms.

Pediatric Quality of Life Inventory – Parent Proxy

The Pediatric Quality of Life Inventory (PedsQL) measures quality of life in children ages 2 to 18 (Varni *et al.* 2001) in the following areas: (i) physical health, (ii) emotional health, (iii) social functioning, and (iv) school functioning. Items are reverse scored and linearly transformed on a scale from 0 to 100 (0 = 100, 1 = 75, 2 = 50, 3 = 25, 4 = 0) so that higher scores indicate better health-related quality of life (HRQoL) (Varni *et al.* 2002). The PedsQL has high internal consistency with alphas for the full 23-item scale approaching 0.90 (Varni *et al.* 2001). Validity has been demonstrated using the known-groups method. The PedsQL distinguished between healthy children and children with cancer as a group (Varni *et al.* 2002). The PedsQL – Parent Proxy was used to evaluate each parent's perception of the HRQoL of their healthy child/young adult.

PedsQL – Family Impact Module. The Family Impact Module (Varni *et al.* 2004) was completed by mothers in order to measure how the child's illness affects the family unit. The 36-item, self-report measure is comprised of 6 scales. Preliminary results have found strong internal consistency scores for the Module Scales (a = 0.82–0.97), Family Functioning Summary (a = 0.90) and the Total Scale (a = 0.97) (Varni *et al.* 2004).

PedsQL – Child/Teen Report. The Child/Teen Report (Varni *et al.* 2001), a 23-item measure, was completed by siblings in order to measure HRQoL. Research has illustrated high internal consistency on the PedsQL, Child/Teen Report, with scores ranging from a = 0.80 to a = 0.88 (physical health a = 0.80, psychosocial health scale a = 0.83, total score a = 0.88).

PWS interviews. The family interview, a 48-item semi-structured interview, was administered to the mother and included: background information, medical and psychosocial information, and sources of social support. Items included questions about the initial reaction to the PWS diagnosis, the stress associated with caring for and raising a child/young adult with PWS, the impact on finances, career decisions, relationship with mother's partner, sibling relationships, and any positive effects associated with having a child/young adult with PWS. The sibling interview, a 17-item semi-structured interview, focused on questions about feelings about the impact of PWS on the sibling as well as family relationships. The interview items were reviewed by a panel of experts consisting of one geneticist, one genetic counsellor, one paediatric psychologist, and two masters level graduate students in clinical psychology.

Quantitative analysis

One-sample *t*-tests were used to compare the mothers' level of distress on the BSI with normative data and to compare scores from the PWS sample on the Family Impact Module of the PedsQL with results in Varni *et al.* (2004). In addition, one sample *t*-tests were used to assess whether the mother's perception of the sibling's psychosocial adjustment was significantly different from PedsQL – Parent Proxy norms; and, to assess whether the sibling's perceived HRQoL, as measured by the PedsQL – Teen Report, was significantly different than norms. A descriptive analysis of the siblings' endorsement for each item on the PTSD scale was conducted.

Qualitative analysis

All responses to the interviews were transcribed. A qualitative case study methodology was used to explore the experiences of families/mothers/siblings with children of PWS. Case studies are useful when one needs to understand a unique situation in depth. A case study examines phenomena in a real-life context when the boundaries between the phenomena and the context are not clearly defined and multiple sources of data are used (Yin 1993). Such an approach allows an investigator to retain the

Table 2 Pediatric Quality of Life Inventory Version 2.0 Family Impact Module

Domain	PWS sample ($n = 13$)	CCH (inpatient) sample ($n = 11$)	REACH (outpatient) sample ($n = 12$)
Physical	48.72 (18.89)	82.99 (17.36)	53.03 (17.26)
Emotional	43.46** (16.51)	78.33 (18.26)	64.48 (26.59)
Social	49.04 (24.45)	85.42 (17.34)	61.93 (25.99)
Cognitive	54.62** (23.05)	88.75 (12.81)	74.09 (18.95)
Communication	37.18* (21.95)	73.61 (24.58)	52.15 (24.67)
Worry	39.62** (19.42)	69.17 (21.09)	56.82 (25.52)
Daily Activities	48.79 (26.47)	85.14 (24.75)	51.89 (31.48)
Family Relationships	56.15** (20.73)	83.75 (23.07)	78.95 (27.62)
Parent HRQoL	48.94** (16.33)	83.75 (15.55)	62.94 (19.83)
Family Functioning	52.64** (17.58)	84.27 (20.47)	68.81 (24.11)
Total Impact Score	47.12** (13.42)	81.0 (17.06)	62.49 (17.26)

Standard deviation presented in parentheses. Higher values equal better health-related quality of life.
**P*-value is significant at <0.05 level; ***P*-value is significant at <0.01 level. Comparison is between PWS sample and REACH sample.
PWS, Prader–Willi syndrome; CCH, long-term care convalescent hospital; REACH, at home with their families; HRQoL, health-related quality of life.

holistic and meaningful characteristics of real-life events. In the current study, the experiences of psychosocial adjustment occurred within the life context of families living with PWS, a chronic health condition. Thus, a multiple case study approach was used to explore differences and commonalities between cases (Yin 2003). This approach facilitated the identification of factors that affect these families and their unique responses. Interviews elicited information regarding sources of stress and support for mothers, and the impact of PWS on family and sibling relationships.

The authors examined and coded responses provided by all of the mothers and siblings. Discrepancies in coding were discussed and reconciled. After analysing all of the interviews, we selected four participants (2 mothers and 2 siblings) to illustrate differences and similarities between cases. Themes were identified and illustrative quotes were selected to explicate our findings.

Quantitative results

Family functioning

As shown in Table 2, the means and standard deviations of the mothers on the PedsQL – Family Impact Module are shown, and compared with results found in a comparable study utilising the same measure (Varni *et al.* 2004). The families in the Varni *et al.* (2004) measure were parents of children with complex chronic health conditions, such as cerebral palsy or birth defects, who either resided in a long-term care convalescent hospital (CCH) or at home with their families (REACH). Varni *et al.* (2004) found that individuals in the home (REACH) had lower quality of life than those residing in the hospital (CCH). In the current study, it was found that the PWS families showed poorer perceived quality of life than the REACH sample in Emotional and Cognitive Functioning, Worry and Communication subscales. Furthermore, the PWS sample had significantly poorer HRQoL than the REACH sample in the overall domains of Parent HRQoL, Family Functioning, and Total Impact Score.

Parent's psychological health

On the BSI, the mothers showed elevated means, signifying increased levels of distress symptoms, as compared with normative data (Derogatis 1993). Significant differences were found in the Obsessive-Compulsive, Depression, and Hostility subscales, and the Positive Symptom Total.

Psychosocial adjustment of the unaffected siblings

When examining differences between the sibling self-report PTSD total score means to means from a school based sample of 695 children 12–18 years (Pat-Horenczyk *et al.* 2007), and a sample of 52 non-bereaved siblings of children with cancer (Fine 2004), a significant difference was found ($t_{10} = 2.20$, $P = 0.05$ and $t_{10} = 4.07$, $P = 0.01$ respectively). The first *t*-test approached the 0.05 level. Notably, 92% ($n = 12$) of siblings in the current study endorsed moderate-to-severe symptoms of post-traumatic stress reaction, while one reported mild symptoms.

Quality of life

When compared with normative populations, mothers from the PWS sample had a significantly different perception of the siblings' psychosocial adjustment and HRQoL compared with parents of other healthy children ($P < 0.01$) on all scales of the PedsQL – Parent Proxy. The mothers from the PWS sample reported similar scores on the PedsQL – Parent Proxy to the parents of children who have been diagnosed with cancer. The only significant difference was for Psychosocial Health (PWS Sample: mean = 60.38, Oncology Sample: mean =70.31, $P \leq 0.05$).

Siblings' responses on the PedsQL – Teen Report revealed that their perceived quality of life in the School Functioning domain was significantly lower than healthy controls ($P < 0.05$), and the PWS siblings' scores in the Psychosocial Health domain approached significance ($t_{12} = -2.038$, $P = 0.064$). When comparing the siblings' self-reported quality of life to the parents' report, results demonstrated that the PWS parents perceived the siblings' HRQoL to be poorer than the siblings believed. Significant differences between mothers and siblings were found in School Functioning ($P \leq 0.05$), Psychosocial Health ($P \leq 0.01$) and Total Score ($P \leq 0.01$).

Qualitative examples

The mother's psychological health and family functioning were described in interviews which demonstrated the diversity of distress experienced by these mothers.

Mrs X demonstrates a mother who is having difficulty coping, with higher reported distress symptoms on the BSI and poorer social support. When asked about her own psychological health and what she does to take care of herself, she replied:

> I feel stressed out. Raising a child with PWS has been a lot of work. I often feel depressed, and cry a lot. Having a child with PWS definitely makes it harder to meet family obligations [because] I am always stressed out about keeping [PWS child] away from food. I don't have a social life because no one understands [PWS] and can't relate to the stress involved. I can't go to a party, or out in public. Sometimes it feels like life is all work and no fun. I try to stay involved in work. I also take antidepressants, and go to therapy for one hour a week.

Mrs Y demonstrates a mother who does not seem as overwhelmed by the difficulties of caring for a child with PWS and has a stronger support system than Mrs X, though she is still experiencing distress. Mrs Y reports about stressors she experiences as a parent of a child with PWS:

> I definitely feel more stress, more anxiety, more worry, and more concern about the future. It's almost like constantly being on guard. It's hard to relax because you don't know what's coming next. House cleaning, getting repairs done, things like that are difficult. I try to make sure that we are still meeting the obligations of our other kids. It has affected the time we spend on their needs . . .
>
> There are definitely negatives—[no] personal time, how much time I can spend with my other kids. It has affected our whole lives. However, there are quite a few positives. [PWS child] is a great kid. We've met a lot of people who are great, wonderful people. It has made me not take things for granted, and focus on things that matter.

Family contexts, severity of the PWS manifestations, as well as intra- and inter-personal variables may vary between siblings in the sample. Also, siblings may vary in their feelings about living with a sibling diagnosed with PWS. When Mrs X was asked her views regarding the relationship between her two daughters, she stated:

> My daughter is jealous of the PWS child because she is beautiful. I think sometimes she feels embarrassed by the PWS child. She has told me in the past that she doesn't want to take care of the PWS child when she is older.

When Mrs Y was asked about her perceptions of her unaffected child's relationship with her child with PWS, she reports:

> He helps by making sure that the PWS child sticks to his diet, being responsible for himself, cleaning up and doing his chores. He feels bad that the PWS child has to deal with PWS.

In addition, siblings may react to stress associated with the impact of PWS in different ways. While some siblings are able to negotiate stressful situations associated with the impact of PWS with resilience, others may not be as successful.

Sibling A, a 15-year-old female, is an example of a sibling who is having difficulty negotiating the stress of living with an individual with PWS:

> Overall, my outlook on life is quite a bit worse compared with other people I know. I definitely don't see myself as an optimist . . .. I feel like I have more serious things to think about and I worry a lot more about what can or could happen to people. I used to take medication for anxiety. I try to keep my feelings to myself. Everyone is busy with the child with PWS. They care but have more pressing issues. I'm worried about upsetting my parents and a lot of people can't relate to my experience.

On the other hand, Sibling B, a 16-year-old female, is an example of an adolescent who does not seem as overwhelmed by the difficulties of living in a family with a sister with PWS:

> The hardest thing about living in a family with a sister who has PWS is that I can't let my guard down. I never know when she'll have a fit or blow up. If there were no caregivers or if my sister with PWS wasn't gone for most weekends, it would be a lot harder. At least one night we get to eat as much as we want or have sleep-overs. It used to make me mad but now I just try to prepare myself for the inevitable blow-up. I don't usually talk about how I feel and sometimes try to forget the whole thing. When I do talk, I talk to my mom or my counsellor because I want advice on how to handle my sister with PWS' bossiness or meanness.

Discussion

The information gained through this study suggests that PWS affects the entire family system. Our results demonstrate that these mothers' quality of life is adversely affected by the multiple demands inherent in caring for a child/young adult with PWS. When compared with parents of individuals diagnosed with diverse chronic conditions, the mothers of children/young adults with PWS report significant difficulties across several domains on the Family Impact Module. Specifically, mothers reported higher levels of emotional stress, poorer quality of life, difficulties with family communication, more worry, and increased family conflicts than parents of children/young adults with other chronic medical conditions.

The present study is consistent with the few published studies assessing the psychological impact of caring for a child/young adult with PWS. In fact, other researchers have found adverse functioning in parents, citing higher levels of stress and marital discord (Hodapp *et al.* 1997a). As speculated by James & Brown (1993):

> PWS families live with inordinate stress, and the quality of family life is impacted by the stress associated with the constancy of care and supervision demands. (p. 253)

Results also demonstrated that mothers in this study had elevated means on the BSI compared to normative data (Derogatis 1993). Significant differences were found in the Depression, Hostility, and Obsessive-Compulsive subscales, and the Positive Symptom total, which suggests that these mothers are significantly more distressed, i.e. show more behavioural symptoms on the BSI when compared with non-clinical samples. Specifically, mothers reported trouble with cognitive functioning, anhedonia, lack of motivation and anger. Furthermore, mothers in this sample reported significantly higher number of overall distress symptoms than would be expected in the general population, as exhibited by their elevated Positive Symptom Total.

In terms of sibling psychosocial functioning, it is notable that 92% of siblings reported moderate-to-severe symptoms of post-traumatic stress reactions. Our results indicate that siblings of children/young adults with PWS experienced more post-traumatic stress symptoms than normative data (healthy siblings), or siblings of children/young adults with cancer. The siblings in the present study reported increased arousal, avoidance, hypervigilance, feelings of anger, sadness when reminded of their sibling's illness, startle responses, sleep problems, and pessimism about the future. These findings suggest that the experience of living with a brother or sister diagnosed with PWS is stressful to siblings in ways typically associated with traumatic events.

In fact, siblings and parents of a child/young adult with PWS experience a wide variety of potentially traumatic events related to the impact of a chronic condition (Kazak 2006; Kazak *et al.* 2006). This can include the need for ongoing therapy, doctor's appointments, daily injections if on growth hormone or if they have insulin-requiring diabetes, medical emergencies, hospitalisation, the need to assure constant supervision to avoid access to food, and behavioural difficulties (i.e. temper tantrums and food seeking). In addition, obesity has been the leading cause of mortality in individuals with PWS. Just as a diagnosis of cancer or another serious medical condition can represent a life threat (a core concept of traumatic stress), the normal activity of eating can pose a life-threatening risk for individuals with PWS. Lack of a cure and increased impairments that frequently occur during the lifespan of individuals with PWS could evoke helplessness and fear in siblings. Other potentially traumatic events siblings may experience include the death of other children/young adults with PWS known to the siblings, medical emergencies and hospitalisation of the individual with PWS, and concerns about having to care for their sibling after their parents are unable to do so.

Packman *et al.* (1997) reported similar findings using the UCLA PTSD scale in a sample of 44 siblings of paediatric bone marrow transplant patients. In that study, one-third of donor and non-donor siblings reported moderate-to-severe post-traumatic stress symptoms. Another investigation of healthy siblings of chronically ill children revealed that almost half of the 61 healthy siblings in the sample had elevated levels in one or more of the trauma subscales from the Trauma Symptom Checklist for Children (Wasik 2002).

Further light is shed on sibling psychosocial functioning by examining HRQoL scores. Comparisons between the mothers' Parent Proxy reports of the PedsQL and normative data revealed significant differences across every domain, indicating a lower HRQoL for siblings of those with PWS. Interestingly, the mothers' reports were more similar to the means published for parents of children/young adults with cancer than to means of the normative sample. This latter finding demonstrates the mothers' awareness of the impact that living with a child/young adult diagnosed with PWS can have on the healthy sibling.

Finally, an analysis was conducted to compare the siblings' self-reported HRQoL to their respective mother's report. The results showed that the mothers consistently reported more problems in functioning and health across every scale of the PedsQL than the siblings did. In fact, the mothers' means on all of the subscales were lower than the siblings' means, indicating that the mothers view the siblings' HRQoL as poorer than the siblings themselves did. The mothers and siblings were found to have statistically different perceptions of the siblings' functioning in the Psychosocial Health, Social and School Functioning domains. The Total Score was also significantly different, indicating that mothers and siblings had different beliefs about the sibling's overall HRQoL. The discrepancy between child/young adult and mother reports in our study is consistent with other studies that utilise quality of life measures (Lambert & Boneh 2004). In our study, siblings reported a higher quality of life than mothers in all domains, indicating, perhaps, the differences between siblings and mothers in their understanding of PWS and the sequelae of the condition. In a similar vein, Hodapp *et al.* (1997b) investigated family stress and sibling reactions in families of children with Cri du Chat syndrome and found that parents and siblings disagreed on the extent of siblings' interpersonal concerns with parents rating concerns at a higher level. The discrepancy in the answers given between siblings and parents suggests that 'while inextricably linked, each family member could experience the same condition very differently' (Lambert & Boneh 2004, p. 606).

In reading about the personal experiences of Mrs X and Mrs Y it becomes clear that mothers of children with PWS face a wide array of challenges, with varying levels of perceived social support. The impact that caring for a child/young adult with a chronic condition has on the various relationships within each family are all shown to be affected by these challenges, and can in turn contribute to each family member's perceived quality of life. The qualitative findings from the sibling interviews suggest that siblings are distressed in various ways and also face multiple challenges. While some siblings are able to cope relatively well with stressful situations associated with the impact of PWS, others may be less successful.

Contributions and limitations

The purpose of this study was to determine the quality of life effects and level of distress in families of children/young adults diagnosed with PWS. The study's findings confirm that the multi-dimensional manifestations of PWS do significantly and adversely impact the family, mothers and unaffected siblings. A notable strength of this study was the use of a comprehensive psychosocial battery as well as qualitative data to evaluate the perspectives of mothers and siblings. The use of a measure of post-traumatic stress disorder and comparison with people with other chronic health conditions rather than with other developmental disabilities in families with PWS is, to our knowledge, unique to this study.

A limitation is the small sample size, as is often the case when doing research on a rare condition. Thus, the results should be interpreted with caution. The PTSD findings are based on comparisons between small and large samples and it is possible that the standard deviations may well be different. Another limitation is that a majority of the families attended either national or regional PWS conferences. These families may not represent all families of children with PWS, because the sample represented mothers who were actively seeking knowledge and support from other clinicians and families of children with PWS. Since all reporting parents were mothers, there may be bias when comparing results with a group of parents of both genders (e.g. Gerstein *et al.* 2009). Future research should include younger siblings in order to facilitate the understanding of similarities and differences at different ages and stages of development.

Recommendations for families affected by PWS

Based on two recent studies by Packman *et al.* (2005, 2008) of siblings of children with cancer, research shows that camps [Special and Important Brothers and Sisters (SIBS): Okizu Foundation] have been effective in increasing siblings' emotional well-being. Camps for siblings of children with PWS might be equally beneficial, as research has shown similarities between the impact of PWS and the impact of paediatric cancer on siblings. The goals of SIBS camp are 'to provide siblings with peer support, validate their feelings and bolster their self esteem' (Packman *et al.* 2008). Based on these reported findings, it would be prudent to include opportunities for parents to also attend a similar camp or alternative experience.

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