‘Over time it just becomes easier…’: parents of people with Angelman syndrome and Prader-Willi syndrome speak about their carer role

Abstract

Purpose: This study investigated two of the stresses experienced by parents caring for offspring with Angelman syndrome (AS) and Prader-Willi syndrome (PWS) in Western Australia, and identified their coping strategies.

Method: Parents of 19 offspring with AS and PWS participated in the Family Stress and Coping Interview which provides a stress level score, and discussion of stressors and coping methods associated with 24 life situations, two of which are reported. All text was examined using directed content analysis.

Results: Family carers (14/19) reported high stress associated with *the initial diagnosis* of AS or PWS in their offspring; and *finding time for themselves*. Stressors identified included lack of quality information about the disorder, time constraints, and physical and emotional tiredness. Parents adopted a variety of coping strategies, including learning about the disorder, accepting the situation, seeking instrumental and social supports, and dealing with problems.

Conclusions: No specific coping strategy was associated with reduced stress. However, parents felt that accurate and timely information during the diagnostic period helped. Parents used family and community support although there were difficulties accessing respite care. Government agencies, service providers, family members, and peer support associations should provide practical and emotional support to assist parents of offspring with AS and PWS, and indeed any form of intellectual disability, across the lifespan.

Implications for Rehabilitation

- Long-term caring for offspring with AS or PWS can involve considerable stress for parents.
- Stress has been associated with poorer health outcomes for parental carers.
- Parents need a variety of practical and emotional supports to cope with stress, including timely access to information.

Introduction

Background

Over the course of the last thirty years significant research has been conducted into the long-term impact of caring for an individual with intellectual and developmental disability (IDD) [e.g., 1,2-7]. The family unit is recognised as a vital component in the life of individuals with IDD, offering support and constancy to people who may require assistance in many or even all aspects of their daily life. However, raising a child and later caring for an adult with IDD can impose major limitations on the lifestyle of primary carers and other family members, and often these limitations are not only physically and mentally demanding, but also life-long [8]. Elevated stress levels, low self-esteem, and social isolation have all been shown to be significant consequences of the carer role [9,10]. Long-term carers are at higher risk of adverse health consequences as their own comorbidities, and those which arise in their offspring with advancing age, pose limitations on their ability to continually provide the necessary level of care and assistance.

Stress and coping in the carer role

Lazarus and Folkman [11] describe stress as experienced when the perceived burden of an event exceeds the resources available to ensure successful management of that event. Within this cognitive-behavioural model, coping is described as the processes applied in attempting to deal with a stressful situation [12]. Given the diversity of coping responses to different stressors, the range of possible outcomes is also highly variable [13]. Research involving the parents of typically developing children and of children with disabilities indicates that coping strategies may moderate stress [3,14], and that coping itself may lead to a more positive appraisal of the event and its consequences [15].

Coping commonly involves either behavioural/physical actions, or cognitive/mental reactions [16] within two main types of coping strategy. The first, problem-focused coping, is aimed at

changing situations arising from the problems or behaviour of the individual and may include concepts corresponding to actions, e.g., 'Restraint', or to thoughts, e.g., 'Planning' (table 1). The alternative strategy, emotion-focused coping, concentrates on the reduction or management of the emotional effects of stress through subjective assessment [2,17,18]. Emotion-focused concepts tend to involve mental reactions, however both 'Seeking social support' and 'Behavioural disengagement' imply some degree of physical/behavioural action (table 1).

Insert table 1 about here

The adoption of problem-focused strategies is reported to have a positive impact on maternal well-being [19], and many of the predominant coping strategies belong to this category [3,20]. Emotion-focused coping is reportedly both rarer and less effective in reducing stress among parent carers [19], although the fathers of children with IDD in an Irish study gained greater benefit from such strategies and used them more often than the mothers in the same study [21].

Examining the use of coping strategies will allow insight into which strategies are used under certain conditions, how they relate to the type and severity of disability present in people with IDD, and how effective the adopted coping mechanisms are in modifying carer stress and health [22,23]. Information from an analysis of these relationships may assist in reducing carer stress and illness, increasing the length of time that a carer can continue in that role, and promote resilience and mental wellness.

Angelman syndrome (AS) or Prader-Willi syndrome (PWS) each occur at an approximate frequency of one in 10,000 to 40,000 live births [24-27]. Within the spectrum of IDD, these two disorders have been under-researched in terms of carer coping and wellbeing [28-31]. Although AS and PWS have genetic similarities, they are distinct in their physical presentations and behavioural profiles. AS is characterised by seizures, severe intellectual

disability, absent speech, jerky ataxic movements and a generally happy sociable disposition [32,33], whereas PWS is characterised by specific behaviour problems, hyperphagia and obesity, and delayed sexual development [34-36].

Research has shown that family carers generally experience greater levels of stress if their offspring have more severe intellectual impairments, psychiatric comorbidity, or behavioural problems [4,37]. People with either AS or PWS experience significant age-related disability and require ongoing care throughout their lifespan. It is therefore expected that their carers will be subject to high levels of stress [26-28,38].

Aims

The aim of the study was to describe the stressors acting on parents caring for offspring with either AS or PWS, and the coping strategies used by family carers to manage the stress. Effective stress management strategies can support family carers to continue in that role.

Materials and Methods

Study design

This study was designed using mixed methods: quantitative survey data relating to the individual with AS or PWS and carer characteristics; and qualitative interview data relating to stress and coping associated with 24 life situations, two of which are described in this paper. Due to the small number of participants most of the quantitative data were omitted from the analysis.

Recruitment

The sampling frame for the study was all known individuals identified with AS and PWS living in Western Australia (WA) in 2008. The expected number was between 80-100 individuals, based on previous data [26,27]. Staff from the Disability Services Commission of WA and the Genetic Services of WA sent invitation letters on behalf of the study to all individuals identified from their databases with AS and PWS. The Disability Services Commission is the primary support and service organisation for people with IDD in WA, and

Genetic Services WA is the main organisation responsible for the diagnosis, counselling, and treatment of people with inherited conditions and their families. Study information leaflets were also mailed to the convenors of the WA branches of two family support groups, the Angelman Syndrome Association and the Prader-Willi Association, for distribution to their membership and the recruitment of any unidentified cases. Recruitment was on an opt-in basis, with all potential participants required to contact the investigator directly if they were interested in participation. Consent forms and survey questionnaires were posted to participants for completion prior to a face-to-face interview.

Participants

Twenty-one families, all of whom were caring for an individual with AS (n=13) o**r** PWS (n=8), volunteered to participate in the interview but it was not possible to schedule interviews with two of these families within the available time-frame. Some descriptive data are missing due to guardianship issues that prevented five families sharing details of the life of their adult offspring with AS (n=3) or PWS (n=2). Denominators for each item therefore vary with the numbers of respondents for each section of the study protocol.

Most of the families interviewed (13/19) included a member with AS (table 2). Ten of the 19 mothers interviewed were over 50 years of age and the interviews included two mother/father dyads. As few fathers were directly involved, their FSCI scores were disregarded in the analysis.

Insert table 2 about here

Measures

The survey questionnaire contained sections on demographic data, carer information, such as the health of the carer and the amount of care provided, carer satisfaction, a clinical profile of the individual with AS or PWS, and a Food-Related Problems Questionnaire [39].

Participants also completed the Family Stress and Coping Interview (FSCI) [40], a semi-structured interview comprising a 5-point Likert scale assessment of stress level, and a

directed discussion about the role of caring for an individual with IDD under 24 different situations. The open-ended nature of the discussion enables researchers to more effectively identify the coping mechanism/s used by individuals to deal with stress, and to compare the effectiveness of different coping styles [40,41]. Stress ratings are given on a scale of 0 (not stressful) to 4 (extremely stressful), thus giving a possible total range of 0-96.

Throughout the interviews with the mothers and fathers of people with AS and PWS one thing was clear: regardless of any reported stress, parents were eager to share stories of their life and that of their offspring. For many families it was the first opportunity they had been given to express their feelings of disappointment, frustration, anger, love and even joy to someone who was not family, a peer family carer, or a medical or support staff member. The idea that their thoughts could be made available to a wide audience seemed to open the hearts and voices of this remarkable group of people. It is clear that the family carers of people with IDD deserve to have their opinions heard as they work towards ordinary life goals, and for their lives to be as happy, healthy and long as possible.

Data analysis

The survey data were analysed using Microsoft Excel. The interviews were transcribed verbatim and the texts managed in NVivo v8.0 (QSR International) which supports mixed methods research and content analysis. A directed content analysis (CA) was conducted on the interview transcripts utilising a deductive approach [42]. The aim of directed CA is to expand or refine extant theory [43], and in the present study it was used to gain an understanding of the stress factors acting on the family carers of the people with PWS and AS, and of the coping methods they used to combat these stressors.

The literature on stress and coping was examined for common themes and concepts that could be applied as the initial coding categories within either the problem-focused or emotion-focused top order categories (table 1). Interview text that correlated with a specific concept was assigned to one or more of these sub-categories.

Separate sub-coded items emerged from the interview data that corresponded to specific stressors. These items were helpful in indicating the most common sources of stress in greater detail, regardless of the actual situation or event that was being considered at that point of the interview. The coding of interview transcripts was undertaken by one author (AT) and then independently verified by another author (EG) for consistency and reliability of coding. Any differences identified in the use of the coding model were discussed to enable consensus to be reached. Minor changes were then made to the coding sub-categories and the remaining pre-analysed transcripts were revised to adhere to these coding categories before the remaining transcripts were encoded.

Ethics and consent

Ethics approval was obtained from Edith Cowan University Human Research Ethics Committee (1721), the WA Department of Health (#EC 2007/02) and King Edward Memorial Hospital Human Research Ethics Committee (1409/EW), and from the Disability Services Commission of WA. Separate consent was sought for the survey and the interview components of the study. Family carers of a minor with AS or PWS, or who were the legal guardian of their adult offspring, consented on behalf of their child and for themselves. Any adult with AS or PWS adjudged capable of self-consent signed a consent form and completed the sections of the survey that related to them. Data relating to an adult with AS or PWS who did not have a legal guardian and could not provide their own consent were not eligible for inclusion [44].

Results and discussion

Family Stress and Coping Scale

Total scores on the FSCI scale were highly variable and ranged from a low of 1 to a high of 64, with an overall mean of 38.2 (median = 39.0). Mothers caring for a family member with AS reported lower mean stress levels (mean 32.6: median 33.0: range 1-53) than those caring for a member with PWS (mean 50.3: median 53.0: range 31-64).

Across the entire cohort a mean exceeding two, signifying higher stress levels, was scored for half the FSCI items (figure 1). Parents reported less stress associated with explaining about their offspring's condition, the cause of the condition, dealing with friends and family, dealing with medical professionals, dealing with legal professionals, deciding on the appropriate level of integration for their offspring, considering short-term housing, maintaining personal friendships, their offspring's sexuality, accessing emotional support, help with day-to-day care of their offspring, and time apart from their offspring.

Insert figure 1 about here

Family Stress and Coping Interview

This paper discusses the effect of two situations, the initial diagnosis and finding time for oneself, on the stress of family carers, and the coping strategies used to deal with that stress.

The initial diagnosis of AS or PWS

The initial diagnosis was the single most stressful situation within the interview schedule, although the mothers of people with PWS (mean score 2.5) reported less stress than the mothers of people with AS (mean score 3.2). Regardless of the condition and age of their offspring, most mothers (13/19) found this item considerably or extremely stressful.

A degree of ambivalence between being relieved to have a diagnosis and sadness due to the condition was reflected in the present interviews. The concept of grief as a stressor, and feelings of disappointment because of changed expectations were also voiced. These feelings were variously expressed as:

> ***'So the actually 'having a label' was good, um, but the actual label was fairly devastating…' (Child, AS)***
>
> ***'Some people grieve for life. Some people grieve and accept and move on quicker than the other individual….in our case it's a pretty much of an ongoing grief because…you feel labelled every day…' (Child, PWS)***

'…just the realisation that what you perceive as normal is not or no longer will apply, and that your dreams and hopes for your child's future…have all been turned upside down…' (Child, AS)

Considerable dissatisfaction was expressed within the present study regarding the process of disclosure, with a perceived lack of support, and a general lack of knowledge available at the time of diagnosis. Anger and dissatisfaction were clearly articulated as:

'…when we got the diagnosis, the paediatrician was really off-hand and she gave us no back-up support systems to speak to.' (Adult, PWS)

'…the information that was out there was out-dated and any documentation of cases of Prader-Willi were of severe cases and so all of the documentation we read was extremely negative.' (Adult, PWS)

It was apparent that feelings of distress caused by the diagnosis persisted in many carers, often for years. Respondents who spoke of improved perceptions of the diagnosis, although it often did not reduce their stress score, attributed this to a range of coping factors such as their improving knowledge of the condition (Seeking Instrumental Support), their acceptance of the diagnosis (Reframing), and the emotional support they received (Seeking Social Support), especially from family members (table 3).

Insert table 3 about here

Meeting your own personal needs

The mean scores for this item were 3.7 (PWS) and 2.5 (AS). Insufficient time for personal matters was the general stressor most frequently mentioned in regard to this situation. Several mothers also talked about their tiredness or even exhaustion resulting from disrupted and insufficient sleep:

'That's right up there because there is no time. You're just down at the bottom when you can fit yourself in.' (Child, AS)

205 ***'I didn't have time, didn't have the energy. I was so tired. I mean, you know what***
206 ***Angels are like – they don't sleep.' (Adult, AS)***

207 Another dominant theme within this item was the issue of babysitters or respite care (Seeking
208 Instrumental Support, table 4). Most carers experienced considerable difficulty accessing
209 respite care as, and when, needed. However, some of those who obtained respite care still
210 reported stress associated with managing the arrangements:

211 ***'So if you had a special event coming up, to get a sitter for it….And then we had a sitter***
212 ***coming in. That was stressful in itself as well.' (Adult, AS)***
213 ***'…[husband] and I go out together and do separate things, sometimes together, four***
214 ***hours a week….[however] it falls to me to organise it…' (Adult, PWS)***

215 Putting aside one's own needs to concentrate on their offspring's needs (Suppression) was the
216 most commonly reported coping strategy for this item. Making light of or learning from the
217 situation (Positive Appraisal) were also reflected in responses from mothers (table 4).

218 *Insert table 4 about here*

219 **Discussion**

220 Previous studies have identified a range of factors that predict successful coping in families
221 with a child with IDD [12,17,45], including the use of a variety of coping strategies, adequate
222 personal and couple time for parents, supportive friends and families, and feelings of self-
223 efficacy. Similarly, parents of people with IDD in Western Australia reported that stressful
224 situations could result in a new outlook for themselves, and strengthen their social and
225 instrumental support networks [46]. Other studies have, however, found that the use of
226 coping strategies had a minimal effect on carer well-being [4,47].

227 The diagnosis of a disability in an infant or child affects the emotions and attitudes of family
228 members. The associated stress is initially related to the process of obtaining a specific
229 diagnosis, next to the realisation that the child is unlikely to have a 'normal' life, and finally
230 leads to feelings of guilt and grief [4]. The central theme identified around a diagnosis of

disability by Hallberg *et al*. [48] was "ambivalence between relief and sorrow". Similar feelings of grief and relief were expressed by participants in this study. During the diagnostic period there was considerable use by families of professional and agency assistance, and of family support. Some carers also referred to positive feelings associated with the caring role, and spoke of the personal growth that resulted from learning to cope with adversity; attitudes that have been associated with more effective family function [12]. Emotional support from personal or professional sources should be available for families with a child with IDD. Clear and accurate information regarding their child's diagnosis and prognosis is considered a vital component of the adjustment process for the parents of a child recently diagnosed with IDD [2,49,50]. Information of this nature may be provided by professionals, or by other parents who have experienced similar circumstances, e.g., the members of a relevant support group. Studies within the last decade reported some parents were unhappy with the manner in which the diagnosis and prognosis were delivered [49,51], and by the quality of the information offered about the disorder [2]. Within this study, references to poor attitudes from medical staff and scant information at diagnosis came in equal parts from family carers of all ages, and therefore they were unlikely to reflect changes in clinical knowledge or procedure across specific eras, but suggest a consistent perception of insufficient support for families regardless of time period.

With regard to the initial diagnosis of their offspring's condition, carers referred both to learning about the disorder and of adjusting their expectations for their offspring's future. This may reflect the ambiguity of the situation: the actual condition cannot be changed and therefore attitudinal change is required, but the process of diagnosis can be modified by the use of resources such as information. The receipt of adequate information, whether from professional or personal sources, has previously been identified as crucial to family adaptation to the stress of having a child with IDD [12]. The present study therefore supports

the recommendation that ongoing information services should be provided to families who care for people with AS and PWS.

Most of the mothers who shared their experiences generally found it difficult to find time or energy to look after their own needs. Greater stress associated with personal time, as experienced by mothers with adult offspring, may be a reflection of their realisation that the time for their child's independence has passed by. Regardless of the supports available, including respite care, many carers needed to put considerable effort and planning into arranging time for themselves. In common with reports from Canada and the UK [52,53], most carers in this study experienced considerable difficulty accessing respite care as, and when, needed.

In the interview texts no instances were identified relating to some concepts generally allocated to emotion-focus coping, such as distancing/denial, which have been associated with increased, rather than decreased distress [23,54,55]. Some studies have identified denial as a common response to stress [56], however there has been little consistency in reports of the effect on carers of the strategy [57]. The self-selective recruitment method for the current study may have failed to include individuals who commonly use distancing/denial coping methods. If use of these strategies is indeed conducive to high stress [54], then people who adopt them with greatest frequency are likely to be more overwhelmed by their caring role and therefore may decline to participate in a study such as the present research. Family carers who adapt poorly to their child having a physical or cognitive disability reportedly use fewer different coping strategies than carers who adapt well [12]. The extensive use of different strategies within this study group may be indicative of an extremely poor response rate from people who were not coping well.

The Family Stress and Coping Interview is a relatively new instrument [3,17,40] and has the advantage of supplying both a numeric stress level and a textual description of stress and

coping. Other measures use a variety of coping-related statements with responses regarding the use of these strategies, and/or the efficacy of the strategies [21,58,59]. The results from these instruments can be used effectively for quantitative analysis, including factor analysis, however they lack the richness of the narrative collected by the FSCI.

Conclusions

It is clear from the present study that the family carers of people with PWS and AS in WA experience considerable amounts of stress, over long periods of time. However, there was no evidence among the small sample represented that the use of specific coping strategies either reduced or increased perceived stress scores. Family members spoke of feeling better after gaining knowledge and accepting the situations. A number of steps are recommended to help support the family carers of people with AS, PWS and other IDD in their caring role:

- Clear, accurate and timely information on their offspring's condition, prognosis, and the available support services should be supplied at the time of diagnosis and across the lifespan.
- Services, such as accommodation support and respite care, should be available for people with AS and PWS on an on-going basis.
- Families should be encouraged to seek both practical and emotional support from peer organisations, family, and friends, as well as from formal service providers.
- Carers should be encouraged and supported in taking time to themselves to enable them to continue in their role.

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Declarations of conflict of interest

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