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Psychosocial problems, coping strategies, and the need for information of parents of children with Prader–Willi syndrome and Angelman syndrome

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Abstract

The aim of the present study was to identify the psychosocial problems of parents of a child with Prader–Willi syndrome or a child with Angelman syndrome. In addition, the strategies these parents apply to cope with these problems as well as their need for information are described. To assess these topics, parents filled in a self-report questionnaire. Both parent groups were found to have a high need for information, high feelings of loss of control, relatively high depressive feelings, particularly in mothers in both syndrome groups. Differences due to the type of syndrome were found on the fear factor. Parents of a child with Angelman syndrome had greater feelings of fear for the negative consequences for themselves, whereas parents of Prader–Willi children were more concerned about the consequences for the child. In general, coping strategies were not found to be different between the parent groups of children who had either type of syndrome.

Keywords: Angelman syndrome; Coping strategies; Informational need; Prader–Willi syndrome; Psychosocial problems

1. Introduction

Parents of a child with a mental handicap are frequently confronted with various kinds of psychosocial and practical problems that sometimes cause high levels of parental distress. These include emotional problems such as fear, depression and worry, reduced satisfaction in their marriage, practical and financial problems, problems related to educational tasks, as well as reduced levels of self-esteem and self-confidence [1–3]. Understanding the specific nature and determinants of these problems as well as the ways parents cope with problems is highly relevant for the development of family interventions and strategies for prevention, education and support. An approach to understanding how parents cope with the education and care of a handicapped child can be based on the transactional theory on coping of

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Lazarus and Folkman [4,5]. According to the stress-coping theory of Lazarus and Folkman [4], the extent to which having a child with a mental handicap is experienced as stressful will depend firstly on the parents' appraisal of the situation of their child and the expected consequences for the family and the family members well-being (primary appraisal). Subsequently, parents will appraise the resources (external and intrapersonal) they have available to cope with the situation, the effectiveness of the different resources and the expected outcomes after implementation of these resources (secondary appraisal). Parents will appraise the efficacy of external resources such as available services, the special education facilities for their child, or opportunities for work. They will also appraise their personal resources and skills in handling the stress for themselves and their family. Based on this appraisal process, parents will apply specific problem-focused and emotion-focused strategies in coping with the situation and with their emotions. According to Lazarus and Folkman [4] coping is seen as a dynamic interaction process between a person and the situation, in which that person constantly tries to keep a balance between his or her own resources and the requirements of the situation.

The appraisal of the situation depends on characteristics of the handicapped child and on characteristics of the parents and family. Characteristics of the child that appear to be clearly related to a higher level of stress in their parents are poor physical health, difficult temperament, low social responsiveness, behavioral problems, limited social skills of the child, and more unusual care taking demands [6,3].

With regard to parental characteristics, research by Schilling and associates [7] indicates that mothers and fathers differ in their perception of their handicapped child's needs and in their own sense of responsibility for addressing those needs. Thus, they may differ in the way they deal with the daily stresses of caring for their child. Other characteristics that have been mentioned as possibly having an influence on experiencing the situation as stressful are age of the parent, social class, income and personality [3]. Also, characteristics of the family that have been mentioned as possibly having an impact on the amount of stress that parents may experience are the life-cycle stage of the family, number of children, age of handicapped child in relation to the others, family integration prior to the birth of the handicapped child, and relations of the handicapped child with the other children [8,3].

The present study focuses on the psychosocial problems of parents with a child with Prader–Willi syndrome or a child with Angelman syndrome, on the strategies these parents apply to cope with these problems, and on their need for information.

Prader–Willi syndrome and Angelman syndrome are rare genetic disorders caused by the loss of function of imprinted genes on the proximal long arm of chromosome 15 (15q11–13) [9]. The estimated prevalence is around 1 in 20,000 for each syndrome. Other studies mention prevalence rates between 1 in 10,000 and 1 in 40,000 live births [10,11]. Both syndromes occur across all ethnic groups.

Prader–Willi syndrome was first described by Prader, Labhart and Willi in 1956 [12]. The syndrome is characterized by severe neonatal hypotonia, feeding difficulties, delayed developmental milestones and a typical physical phenotype including a typical dysmorphic facial appearance with triangular mouth and almond-shaped eyes, short stature, small hands and feet, genital underdevelopment and delayed secondary sexual characteristics [13,14]. There is an increased prevalence of scoliosis and strabismus. In the first months of life severe muscle hypotonia and a failure to thrive are the most prominent features. After a few years the most noticeable characteristic is a developing hyperphagia leading to life-threatening obesity if not controlled. Many of the manifestations are related to functional hypothalamic deficiency [15].

Prader–Willi syndrome is said to have a characteristic psychological and behavioural phenotype. Some of the symptoms of Prader–Willi such as insatiable appetite, outbursts of rage, stubbornness, and difficulty in dealing effectively with changes in their daily routines become increasingly apparent and severe with age [13,16,17]. Skin picking and abnormal sleep patterns have been reported in most of the affected individuals as well as difficulties in social relationships with peers and reduced physical activity.

Angelman syndrome is, like the Prader–Willi syndrome, a complex multiple-anomaly syndrome,

which is clinically distinct from Prader–Willi syndrome. Angelman syndrome, which was first described by Angelman in 1965 [18], is characterized by severe or profound learning difficulties and lack of speech, delayed motor development, inappropriate bouts of laughter, an ataxic gait, and jerky voluntary movements. Furthermore, there is the typical facial appearance with a long face and prominent jaw, a wide mouth with widely spaced teeth, thin upper lip, an underdeveloped mid-face and deep-set eyes, and a flat occiput. Among the frequent clinical characteristics are microcephaly, epileptic seizures and an abnormal characteristic EEG pattern with large-amplitude slow-spike waves [19–21].

Because of the typical psychological and behavioral characteristics of a child with Prader–Willi syndrome or a child with Angelman syndrome it is expected that these characteristics will have a specific impact on the parents' interpretation of their situation, resulting in a particular pattern of psychosocial problems and in particular strategies for coping with these problems. In addition to an analysis of the psychosocial problems of parents, this study also focuses on analyzing the parents' need for information. Information available to parents or information received from professionals or from other sources may highly influence the extent to which parents perceive their situation as stressful. Use of information also plays an important role in certain coping strategies, and providing information may enhance the use of specific coping strategies preferred by parents in coping with their problems.

2. Methods

2.1. Participants and procedure

To assess the impact of having a child with Prader–Willi syndrome or a child with Angelman syndrome on the psychological functioning of parents, a self-report questionnaire was sent to all families who were members of the Dutch Prader–Willi/Angelman Parent Association and who either had a child with Prader–Willi syndrome (46 families) or a child with Angelman syndrome (31 families) between 0 and 12 years of age. Families living abroad were excluded from the study. One family was excluded later because their child did not appear to have the Prader–Willi syndrome. In two-parent families both the father and the mother were asked to complete a questionnaire independently of each other. If a questionnaire was not returned after 2 weeks, a reminder was sent to those families.

Questionnaires were returned by 34 *families with a child with Prader–Willi syndrome* (76%). From 28 of these families, both the father and the mother responded. From three two-parent families only the mother responded while three questionnaires were returned by single mothers. In all, 62 questionnaires were returned by parents with a child with Prader–Willi syndrome. Twenty-eight of these participants (45%) were fathers and 34 (55%) were mothers. Forty-three parents (70%) were 40 years of age or younger. From the 34 families 15 had a boy with the Prader–Willi syndrome and 19 families had a girl. The average age of the children was 7.3 years. Most of the children (29) were living with their parents and only five were living in an institution or in sheltered homes.

From the 31 *families with a child with Angelman syndrome*, 22 families (68%) responded. From 15 of these families, both the father and the mother returned the questionnaire while from five two-parent families only the mother responded. Two questionnaires were returned by single mothers. In all, 37 questionnaires were returned by parents with a child with Angelman syndrome. Fifteen of these participants (41%) were fathers and 22 (60%) were mothers. Thirty-one parents (84%) were 40 years of age or younger. From the 22 families nine had a boy and 12 had a girl with Angelman syndrome, while gender was not mentioned by one parent. The average age of the child was 7.2 years. Most of the children (15) were living with their family, four lived in an institution, and three children combined living at home with institutional living or care.

2.2. Measurements

In this cross-sectional study, questionnaire data were collected on psychosocial problems of parents and on strategies parents use in coping with these psychosocial problems. Also, data were collected on general information parents received about the syndrome of their child and about their evaluation of

this information. In addition, questions were asked about the diagnosis of the syndrome, medical supervision of their child, contacts with other parents, need for contact with other parents of a child with the same syndrome and evaluation of these contacts and, finally, sociodemographic information about the parents and their family.

The measurement of psychosocial problems in this study was based on a theoretical model of coping with psychosocial problems with life-threatening diseases [22,23]. This model is based on the assumption that uncertainty, negative feelings (fear, depression), loss of control and threat to self-esteem are the four most important psychosocial problems experienced because of a health threat. Parents will try to prevent or to reduce such problems as much as possible. These efforts are called coping strategies.

2.2.1. Uncertainty

Uncertainty was defined as 'the need for information regarding subjects of high relevance to a person'. Uncertainty was measured in two dimensions by scales adapted from Van den Borne and Pruyn [23]. These two dimensions are: (1) 'Need for information about prospects of disease and treatment' and (2) 'Need for information about access to help and about how to solve problems'. Sample items in the first scale included: 'Need for information on the medical condition of my child at this moment' and 'Need for information about the course of the Prader–Willi syndrome (or Angelman syndrome)'. Sample items in the second scale included: 'Need for information on how to talk with others about problems and difficulties related to the handicap of my child' and 'Need for information on how to improve the physical fitness of my child'. The items were scored on a four-point scale, using alternatives: not at all (1); a little bit (2); quite a lot (3); very much (4).

2.2.2. Negative feelings

Fear was measured by a 15-item scale asking questions about the level of concern about problems bothering their child and being concerned for problems bothering (the parents) themselves. Factor analysis revealed two dimensions of fear: (1) 'Fear for negative consequences for the child' and (2) 'Fear for negative consequences for themselves (the parent)'. Sample items in the first dimension included: 'Concerned about worsening of the handicap of my child' and 'Concerned for loneliness of my child in the future'. Sample items for the second dimension included: 'Concerned that other people who are important to me, will let me down because of my child' and 'Concerned for tensions in my family because of worries about my child'. The items were scored on a four-point scale, using alternatives: not concerned at all (1); a little concerned (2); quite concerned (3); very much concerned (4).

2.2.3. Depression

Depression was measured by 10 items from the Zung Depression scale [24].

2.2.4. Loss of control

Loss of control was measured by a scale including 12 statements based on a loss of control scale by Van den Borne and Pruyn [23]. Sample statements included: ''Because of my child, I feel like my hands and feet are tied'' and ''Because of the problems with my child, I have more problems with controlling my emotions''. The statements were scored on a four-point scale, using alternatives: 'does not apply to me at all' (1); 'hardly applies to me' (2); 'applies to me partly' (3); 'applies to me entirely' (4).

2.2.5. Self-esteem

Self-esteem was measured by a Dutch self-rating questionnaire with 12 statements about self-esteem and mood [25]. Sample statements included: 'I am satisfied with myself' and 'I feel that I have little to be proud of'. The statements were scored on a four-point scale, using alternatives: 'completely agree' (1); 'partly agree' (2); 'partly disagree' (3); and 'completely disagree' (4).

2.2.6. Coping strategies

Coping strategies of parents were measured by the short version (26 items) of the Utrecht Coping List (UCL) [26]. Factor analysis on this list revealed three different coping strategies: (1) cognitive and behavioral avoidance; (2) active problem solving; and (3) seeking social support. Sample items from the 'cognitive and behavioural avoidance' scale included: 'If I have problems I tend to give in to

Table 1
Review of scales for psychosocial problems and coping strategies

	Number of items	Cronbach's alfa	Min. score	Max. score	Mean	Standard deviation
Uncertainty about prospects of disease and treatment	11	0.88	11	44	32.685	8.171
Uncertainty about access to help and about how to solve problems	8	0.88	8	32	20.215	6.431
Fear for negative consequences for themselves (the parent)	10	0.86	10	40	15.563	4.960
Fear for negative consequences for the child	4	0.74	4	16	8.243	2.725
Loss of control	12	0.92	12	48	30.275	9.123
Depression	10	0.85	10	40	18.161	5.095
Self-esteem	12	0.84	12	48	38.815	6.374
Coping: active problem solving	6	0.94	6	24	16.192	4.091
Coping: cognitive and behavioural avoidance	11	0.95	11	44	19.020	4.357
Coping: seeking social support	6	0.93	6	24	13.560	4.370

prevent difficult situations' and 'If I expect problems, I tend to withdraw from the situation'. Sample items from the 'active problem solving' strategy included: 'If I have problems, I try to come up with different solutions' and 'In difficult situations I try to intervene immediately'. Sample items from the 'seeking social support' strategy included: 'If I have worries I share these with others' and 'If I have problems, I ask someone for help'. Items were scored on a four-point scale, using alternatives: 'hardly or never' (1); 'sometimes' (2); 'frequently' (3); and 'very frequent or always' (4).

For each parent total scores were calculated for each of the scales. A review of scales for psychosocial problems and coping strategies is presented in Table 1.

Reliability analysis of the scales for psychosocial problems and coping strategies reveals satisfactory Cronbach's alphas, ranging from 0.74 for 'fear for negative consequences for the child' to 0.95 for coping: 'cognitive and behavioral avoidance'.

3. Results

In Tables 2–8 the scores on the individual items of each scale measuring psychosocial problems of parents of both children with Prader–Willi syndrome and children with Angelman syndrome are presented. In Table 9 psychosocial problems of parents with a child with Prader–Willi syndrome and parents of a child with Angelman syndrome are compared. For each of the syndromes, psychosocial problems of fathers and mothers are compared in Table 10.

3.1. Uncertainty

Items about uncertainty of parents defined as '*the need for information about prospects of disease and treatment*', are presented in Table 2. In general, both parents of Prader–Willi and Angelman children show high needs for information. Most parents have a high need for information about how their child may develop in the future, about the possible course and consequences of the syndrome, about the purpose and the role of education for their child and about prevalences and life expectancies. On many issues more than half of the parents up to nearly all of the parents have these needs for information. Comparing parents with a child with Prader–Willi syndrome to parents with a child with Angelman syndrome reveals a significant difference ($t = 3{,}462$; $P = 0.001$) between both groups, with Angelman parents having higher needs for information than Prader–Willi parents (see also Table 9). Differences were also significant ($t = 3{,}144$; $P = 0.002$) when only parents of families with two parents to take care of the children were selected for analysis.

Table 2
Uncertainty about prospects of disease and treatment

Quite a lot/very much need for information about	Parents (%)	
	Prader–Willi ($N = 61$)	Angelman ($N = 37$)
How my child can develop	79	95
Possible consequences of the syndrome	72	87
The use or purpose of education	71	81
The possible course of the syndrome	66	84
Numbers about the syndrome (prevalence, life expectancy)	65	79
The use or purpose of medication	64	81
The use or purpose of dieting[a]/speech training[a]	54	81
The use or purpose of physical therapy	50	81
The cause of the syndrome	45	67
The medical condition of my child at this moment	41	54
How to get information material or literature about the syndrome	33	58

[a] Syndrome specific topic.

Table 3
Uncertainty about access to help and about how to solve problems

Quite a lot/very much need for information about	Parents (%)	
	Prader–Willi ($N = 61$)	Angelman ($N = 36$)
What is the best way to take care for my child's physical fitness	69	78
What is the best way to deal with my child	58	69
Which caregivers can supply me with information	56	68
In which way other parents deal with their child's disability	43	57
What is the best way to talk to people who are close to me about problems and difficulties concerning my child's disability	41	33
What caregivers can do	38	69
How I can talk to or deal with the doctor	28	35
How to contact other parents of a child with the syndrome	13	39

Table 4
Fear for negative consequences for the child

Quite/very much concerned about	Parents (%)	
	Prader–Willi ($N = 59$)	Angelman ($N = 37$)
Disappointments my child has to cope with in the future	41	19
Worsening of my child's disability	29	19
Loneliness of my child in the future	27	19
Tension in the family because of worries about my child	26	16

Uncertainty was also measured with respect to '*the need for information about access to help and about how to solve problems*' (Table 3). Both groups of parents show fairly high needs for information about how to deal with and to take care of their child, about where to go for information, how other parents

Table 5
Fear for negative consequences for themselves (the parent)

Quite/very much concerned about	Parents (%)	
	Prader–Willi ($N = 59$)	Angelman ($N = 37$)
Coming to the end of my patience in assisting my child	17	16
My child not getting sufficient care in the future	15	33
Not being taken seriously when I talk about problems with my child	15	11
Important people abandoning my child	9	22
I will always depend on other people because of my child's disability	7	22
Asking other people for help	7	5
Visiting other people with my child	5	19
Losing my job or social position because of the time I need for my child	5	3
Other people will not be able to or don't want to help me in assisting my child in the future	3	14
Important people abandoning me	3	14

Table 6
Depression

Often/very often or always the case	Parents (%)	
	Prader–Willi ($N = 57$)	Angelman ($N = 37$)
I have got the feeling things don't come as easy as they used to do	54	43
I view the future with concern	32	27
I am more irritable than before	30	19
I feel hunted, not able to sit still	19	19
I don't enjoy life	18	11
I am not as clear-headed as before	16	8
I am tired for nothing	16	14
I am subject to crying fits	11	8
I feel dejected	9	5
I feel lonely	5	3

deal with the disability of their child, and how to talk to other people about problems and difficulties with their child.

3.2. 'Fear for negative consequences for the child' and 'fear for negative consequences for parents'

On the first dimension of fear four items indicate a fear for negative consequences of the syndrome for the child. A number of parents are very concerned about the possible disappointments with which their child may be confronted in the future: (1) the loneliness of their child; and (2) a worsening of the child's disabilities. A comparison of the two parent groups shows a significant difference in these kinds of concerns ($t = -2{,}430$; $P = 0.02$) (Table 9). The Prader–Willi parent group has a higher concern about possible negative consequences for the child than the Angelman parent group. This was also found in an analysis based on only two-parent families ($t = -2{,}581$; $P = 0.01$).

On the second dimension of fear, '*negative conse-*

Table 7
Loss of control

Applies to me partly/entirely By having a child with the syndrome:	Parents (%)	
	Prader–Willi ($N = 59$)	Angelman ($N = 35$)
I have to spend (much) more time on the education of the child with the syndrome	77	89
My thoughts often wander to concerns about my child	66	81
I feel like my hands and feet are more tied	61	83
I am not able to handle my affairs as well as before	56	89
I have more special financial burdens	50	64
I am not the person I used to be	50	56
I cannot have the same leisure activities as before	44	66
I have less control of my emotions	43	57
I depend more on other people as before	38	64
I pay less attention to my job or social position	33	54
I make appointments with other people less easily	25	47
I have less time to run the household properly	22	46

Table 8
Self-esteem

I agree partly/completely that	Parents (%)	
	Prader–Willi ($N = 57$)	Angelman ($N = 37$)
I don't hate myself	95	95
I don't feel I have little to be proud of	91	100
I would not like to be another person	82	92
I don't incline to think I am useless	81	100
I don't feel sometimes I am good for nothing	79	97
I am satisfied with myself	70	89
I take up a positive attitude towards myself	70	86
I am not a pessimist	68	81
I don't get in the depths easily	67	76
I am not easily discouraged	63	68
I am not a tense person	51	49
I am not a self-doubting person	46	54

quences for the parents themself', both groups have a lower level of fear as compared to 'fear for negative consequences for their child'. Some parents expressed a fear of losing their patience with their child, fear of not being taken seriously when talking to others about their child, fear of needing to depend on other people because of their child's disability, and fear of visiting other people with their child (Table 5).

A significant difference ($t = 1{,}984$; $P = 0.05$) in 'fear for negative consequences for themselves' between both groups was found, with Prader–Willi parents being less concerned than Angelman parents (see also Table 9). This significant difference disappears when only parents of families of two adults taking care for the children are selected for the analysis.

For both parent groups, percentages on the item 'I have got the feeling things don't come as easy as they used to do' are relatively high. Between 20 and

Table 9
Means of psychosocial problems, differences between parents

	Prader–Willi		Angelman	
	Parents	*N*	Parents	*N*
Uncertainty about prospects of disease and treatment	30.72	61	35.93***	37
Uncertainty about access to help and about how to solve problems	19.30	61	21.77	36
Fear for negative consequences for themselves	14.78	59	16.81*	37
Fear for negative consequences for the child	8.76	59	7.41*	37
Loss of control	28.22	59	33.74**	35
Depression	18.32	57	17.92	37
Self-esteem	37.59	57	40.70*	37

** $P \leq 0.01$.
*** $P \leq 0.001$.
* $P \leq 0.05$.

Table 10
Means of psychosocial problems, differences between fathers and mothers within the syndrome

	Prader–Willi				Angelman			
	Fathers	*N*	Mothers	*N*	Fathers	*N*	Mothers	*N*
Uncertainty about prospects of disease and treatment	29.67	28	31.61	33	36.13	15	35.79	22
Uncertainty about access to help and about how to solve problems	17.31	28	20.99*	33	21.93	15	21.64	21
Fear for negative consequences for themselves	13.00	27	16.28**	32	15.37	15	17.79	22
Fear for negative consequences for the child	8.04	27	9.38	32	7.35	15	7.45	22
Loss of control	25.08	26	30.69*	33	32.33	15	34.80	20
Depression	16.12	26	20.16**	31	15.73	15	19.41*	22
Self-esteem	39.27	26	36.18	31	42.60	15	39.41	22

* $P \leq 0.05$.
** $P \leq 0.01$.

30% of the parents view the future with concern, are more irritable than before, and have a feeling of being hunted.

In a comparison of both parent groups no significant differences on depressive feelings were found. An analysis of variance with type of syndrome and sex of the child as factors revealed a significant interaction effect ($F = 7{,}373$; $P = 0.008$) on depression. Parents of a boy with Angelman syndrome have the highest score on depression, much more than parents of a girl with Angelman syndrome. Differences on depression between parents of boys and girls with Prader–Willi syndrome were very small.

3.3. Loss of control

Many parents feel a high level of loss of control indicated by feelings like ‘their hands and feet are more tied’, not feeling able to handle their affairs as before, thoughts that wander to concerns about their child, and more financial burdens (Table 7). A significant difference ($t = 3{,}164$; $P = 0.002$) between both parent groups was found, with parents of

children with Angelman syndrome reporting more loss of control (Table 9). The difference remains significant ($t = 3{,}196$; $P = 0.002$) for parents of families consisting of two parents only.

3.4. Self-esteem

As suggested by the findings in Table 8, having a child with one of the syndromes did not seem to have a high negative impact on the parent's self-esteem. On two items all respondents of the Angelman parent group 100% agreed partly or completely with the esteem statements ('I don't feel I have little to be proud of' and 'I don't incline to think I am useless'). Relatively low are responses for both groups on the items about being a tense person and being a self-doubting person. A comparison of both groups of parents indicated that the Angelman parent group scored significantly higher ($t = 2{,}371$; $P = 0.02$) on self-esteem than did the Prader–Willi parent group (Table 9). Differences were even more significant for two-parent family cases only ($t = 2{,}745$; $P = 0.007$).

In Table 10 differences between fathers and mothers are presented for both syndromes. Within the Prader–Willi parent group, mothers scored significantly higher than fathers on uncertainty about access to help and about how to solve problems, on fear for negative consequences for themselves, on loss of control, and on depression. Considering parents of children with Angelman syndrome no significant differences were observed except for mothers who scored significantly higher on depression than fathers.

3.5. Coping strategies of parents

Coping strategies of parents were measured with respect to 'cognitive and behavioral avoidance', 'active problem solving', and 'seeking social support'.

Concerning the parents coping with psychosocial problems, no significant differences in frequency of following different coping strategies between parents of children with Prader–Willi and Angelman syndrome were found. In an analysis of variance the scores of parents on each coping strategy were dichotomized into those scoring low and those scoring high. In this way three dichotomized variables, one for each coping strategy, were created to be used as factors in analyses of variance. One of the analyses revealed a significant interaction of parent gender and active problem solving as factors on self-esteem ($F = 4{,}144$; $P = 0.045$). This interaction effect was significant only for cases of two-parent families. Fathers who had a high score on the coping strategy 'active problem solving' had high feelings of self-esteem. Mothers, both low and high on active problem solving, had equal levels of self-esteem.

A second analysis showed a significant two-way interaction effect of type of syndrome and cognitive and behavioural avoidance as factors on fear for negative consequences for the parent as the dependent variable ($F = 5{,}285$; $P = 0.024$). Parents of children with Prader–Willi syndrome who cope with their problems through cognitive and behavioural avoidance have a high level of fear for negative consequences for themselves in comparison to parents of Prader–Willi children who do not avoid their problems. For parents with a child with Angelman syndrome, who on average have a relatively high fear of consequences for themselves, no differences between those scoring high and low on avoidance were found.

4. Discussion

The results indicate a great need for information for parents with respect to many different aspects that may impact on the development and future prospects of their child.

Parents particularly have a great need for information about the possible consequences of the syndrome for their child, about how their child may develop in the future, and how education might contribute to this development. As compared to the parents with Prader–Willi syndrome the need for information of parents with a child with Angelman syndrome in this respect is significantly higher. A possible explanation for this higher need of Angelman parents may lie in the fact that, because of the relatively recent foundation of the Angelman Parent Association, little educational material has been developed. For Prader–Willi syndrome much more and better education material is available.

With regard to the emotional burden, some parents are very much concerned about the negative consequences of the syndrome for their child, with parents of a child with Prader–Willi syndrome being most concerned. Parents particularly worry about how the child will cope with challenges because of the syndrome, about a possible worsening of disabilities and worry about future loneliness of their child. Parents may also be concerned about negative consequences for themselves such as the fear of losing their patience in caring for their child or fear for not being taken seriously when talking to others about the problems they have with their child. These concerns were found to be somewhat higher in parents with a child with Angelman syndrome.

Many parents also feel a high level of loss of control indicated by a feeling like 'their hands and feet are more tied', not feeling able to handle their affairs as before, thoughts that wander to concerns about their child, and more financial burdens. Parents with a child with Angelman syndrome have a significantly higher level of loss of control compared to parents of a child with Prader–Willi syndrome.

The results also indicate a relatively high level of depressive feelings, particularly in the mothers, irrespective of the type of syndrome. Constant feelings that things don't come as easy as they used to, a constant feeling of concern about the future, feeling hunted, and feelings of more irritability predominate.

Having and caring for a child with the Prader–Willi syndrome or Angelman syndrome does not seem to have a great negative impact on feelings of self-esteem, although parents with a child with Prader–Willi syndrome were found to have a significantly lower level of self-esteem.

Although the parents of both a child with Prader–Willi syndrome or with Angelman syndrome experience various psychosocial problems, the type of handicap and related behavioral problems of the child appear to have distinctive psychosocial consequences for the parents. While parents with a child with Angelman syndrome have somewhat higher feelings of fear for the negative consequences for themselves, the parents with a child with Prader–Willi syndrome appear to have a higher fear for negative psychosocial consequences of the syndrome for the child itself. A possible explanation for this could be that children with Prader–Willi syndrome as compared to children with Angelman syndrome, on average, are less mentally handicapped, and consequently have a higher level of awareness of their condition. Because of this higher awareness, parents may anticipate and fear that their child in the future will be confronted with its limitations more often and more intensely.

Fear of negative consequences for the parents themselves is not only related to the type of syndrome, but also to the strategies on how parents cope with problems. In coping with the problems of a child with Prader–Willi syndrome particularly, parents who cope with these problems by trying to avoid them, through cognitive and behavioral strategies, have a high fear of possible consequences for themselves. This might also indicate that some parents of a child with Prader–Willi syndrome do struggle with accepting the problems characteristically related to the Prader–Willi syndrome.

There are some indications that mothers as compared to fathers experience a higher burden (a higher level of depression). In particular, mothers of a child with Prader–Willi syndrome score higher than their partners on fear for negative consequences for their child, a much higher level of depression and more loss of control and more uncertainty about access to help and possibilities for solving problems. It could be that mothers particularly are more frequently confronted with the burden of coping with a high level of (behavioral) problems, such as managing the insatiable appetite and compulsive-obsessive behaviour of their child with Prader–Willi syndrome.

4.1. Practical implications

The results from this study indicate a high need for adequate education and information for parents with a young child with one of these syndromes. In this education, information to parents about (behavioural) consequences of the syndrome, about future prospects related to treatment, and about child education, child development and care, are particularly important. Also, helping parents understand that specific behavioral problems are a direct consequence of the syndrome and teaching parents techniques for coping with these problems may well support the parents' coping strategies. This teaching

of parents should be done before the problems in the child emerge.

As the burden of care for the child appear to have the strongest impact on the mothers, educating the fathers as well as the mothers may contribute to less avoidance of problems by fathers and a better way of coping with problems and support for the child by the family. Also, facilities or organisations that may provide periodical relief from heavy care duties, particularly for mothers, may help to prevent the development of depressive feelings.

Finally, for some parents contacts with other parents with a child with the same syndrome may help them to learn that they are not the only ones with a child with special challenges. In such contacts, as for example organised by the Dutch Prader–Willi and Angelman association, parents may also learn particular strategies that have been found to be effective by others in coping with similar problems.

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