Genetic syndromes in the family : child characteristics and parenting stress in Angelman, CHARGE, Cornelia de Lange, Prader-Willi, and Rett syndrome

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6 Maternal parenting stress in families with a child with Angelman syndrome or Prader-Willi syndrome

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ABSTRACT

Parenting stress was investigated in mothers with a child with Angelman syndrome (AS) or Prader-Willi syndrome (PWS), which are genetically related. Mothers of 24 children with AS and 23 children with PWS (2 – 12 years) completed the Nijmegen Parenting Stress Index–Short, Developmental Behaviour Checklist and Vineland Screener 0-12. Parenting stress was high for 58% AS and 26% PWS cases. For both syndromes, no relationship existed with the child's gender, age, and behavioural problems. In PWS there was no effect of level of functioning. Overall, more mothers with child with AS perceived high parenting stress. When children showed low levels of behavioural problems this difference was contained. However, when children exhibited severe behavioural problems, parenting stress was the same for both syndromes. In AS professional family support is essential, since parenting is stressful for many mothers. In PWS, this is especially the case when behavioural problems are present.

INTRODUCTION

The upbringing of a child, besides being a joyful experience, can at certain times also involve parenting related stress (Deater-Deckard, 2004). Parents with a child with intellectual disability exhibit elevated levels of parenting stress, which tends to be chronic (Hassall & Rose, 2005; Hastings & Beck, 2004; Hatton & Emerson, 2003; Head & Abbeduto, 2007; Olsson, 2008). High levels of parenting stress can have severe implications, such as harsh or withdrawn parenting, and distressed parents are less likely to optimise the child's development (Deater-Deckard, 2004). Parenting stress in families with a child with developmental delays is also associated with negative outcomes for the parent, such as depression (Singer, 2006) and poor physical health (Oelofsen & Richardson, 2006). Children with a developmental disability are particularly susceptible to the influence of a less than optimal family environment (Paczkowski & Baker, 2007; Seligman & Darling, 2007, as cited in Head & Abbeduto, 2007). As such, it is essential to provide the most appropriate support possible in families with a child with intellectual disability when parenting stress is high.

Different theoretical models exist to investigate parental perception, including parenting stress, of the child-rearing experience. Common characteristics of such models

are the incorporation of child characteristics, environmental influences and parental cognitive processes (Hassall & Rose, 2005). In this study, we focus on the perception of maternal parenting stress and the relationship with child characteristics in two different genetic syndromes associated with intellectual disability: Angelman syndrome (AS) and Prader-Willi syndrome (PWS). Both syndromes are caused by changes in the genetic information in the same small area of chromosome 15, and may therefore be called related. In AS the defects are of maternal origin, whereas in PWS they are paternal (Glenn, Driscoll, Yang, & Nicholls, 1997), which results in two distinct (behavioural) phenotypes.

In families with a child with intellectual disability, the child factor most strongly related to parenting stress is the presence of behavioural problems as opposed to, for example, level of cognitive functioning (Hassall & Rose, 2005; Hastings & Beck, 2004; Hatton & Emerson, 2003; Olsson, 2008). Hodapp (1999) states that among children with a genetic syndrome, behavioural problems are also the best predictor of parenting stress. However, he also underlines that children with different genetic syndromes, with their distinct physical and behavioural phenotypes, elicit different reactions from their environment (Dykens, Hodapp, & Finucane, 2000; Hodapp, 1999). It therefore seems important to investigate the relationship between parenting stress and child characteristics for different genetic syndromes separately, since relationships with other child characteristics have been found as well. For instance, higher stress levels in parents of children with Cornelia de Lange syndrome or Joubert syndrome were also related to the child's older age and lower levels of (adaptive) functioning (Farmer, Deidrick, Gitten, Fennell, & Maria, 2006; Sarimski, 1997b; Wulffaert, Van Berckelaer-Onnes, Kroonenberg, Scholte, Bhuiyan, & Hennekam, 2009). Furthermore, a comparison of children with Down syndrome, Williams syndrome, and Smith-Magenis syndrome showed that the influence of child characteristics on stress is syndrome-specific, with different relationships with age and behaviour for the three syndromes (Fidler, Hodapp, & Dykens, 2000). Thus, to provide specific and more individualised support to these families, syndrome-specific investigations are needed.

Angelman syndrome is a rare genetic syndrome; birth prevalence is estimated at 1:40,000, but population prevalence rates as high as 1:10,000 have also been reported (Petersen, Brøndum-Nielsen, Hansen, & Wulff, 1995; Thomson, Glasson, & Bittles, 2006). A diagnosis of AS can be based on clinical criteria (Williams et al., 2006) but in the majority of cases can be confirmed by genetic testing (Clayton-Smith & Laan, 2003). The

following features are present in 100% of cases: developmental delay, a movement or balance disorder, severe speech impairment (none or only a few words), and behavioural uniqueness including frequent smiling/laughter, happy demeanour, easily excitable personality often with hand-flapping, and hypermotoric behaviour. In 80% of cases epilepsy is found, as well as an abnormal EEG and delayed head growth. According to the diagnostic criteria, a functionally severe developmental delay will be present (Williams et al., 2006). However, somewhat better cognitive and adaptive abilities have been found, although the majority seem to function on the severe delayed level (Peters et al., 2004; Thomson et al., 2006). A behavioural phenotype is just emerging for AS (Horsler & Oliver, 2006). Frequently mentioned, besides the aforementioned behaviours, are eating problems (e.g. eating inedible things), hyperactivity and attention problems, mouthing objects, and sleep disturbances. Persons with AS have an intense fascination for water and other reflective surfaces. It is still unclear whether there is an increased prevalence of autism spectrum disorders (Clayton-Smith & Laan, 2003; Didden, Korzilius, Sturmey, Lancioni, & Curfs, 2008; Dykens et al., 2000; Horsler & Oliver, 2006; Pelc, Cheron, & Dan, 2008). The clinical picture is most distinct in children between 2- to 16-years-old (Buntinx et al., 1995).

Prader-Willi syndrome has been studied much more extensively, especially concerning behavioural aspects. Its population prevalence is estimated to be between 1:8,000 and 1:52,000 (Åkefeldt, Gillberg, & Larsson, 1991; Whittington et al., 2001). A PWS diagnosis can be based on clinical criteria (Holm et al., 1993), but is preferably confirmed by genetic testing. The development of individuals with PWS takes place in two stages. The first phase of life is characterised by hypotonia, with poor sucking and failure to thrive; motor milestones are achieved later in life. The second phase starts at the age of one to six years; problems with gaining weight turn into life-long problems with overeating. This hyperphagia is due to insufficient functioning of the hypothalamus and can lead to life-threatening obesity; nowadays, most children are placed on a strict diet (Dykens et al., 2000; Goldstone, Holland, Hauffa, Hokken-Koelega, & Tauber, 2008). Intelligence quotients (IQ) for most persons with PWS are in the borderline, mild, or moderate range; a near normal distribution of IQ with a downward shift of 40 points is found (Curfs, 1992, as cited in Dykens et al., 2000; Whittington et al., 2004). The level of adaptive functioning is very often lower than what would be expected according to the IQ due to behavioural problems (Dykens et al., 2000). Apart from food-related problems,

such as hoarding food, other specific behavioural and psychiatric problems can be present. Often mentioned are aggression, oppositional and argumentative behaviours, self-injurious behaviour (skin-picking), stubbornness, and temper tantrums. Obsessive-compulsive symptoms and disorder are highly prevalent in PWS. Furthermore, symptoms of psychoses and affective disorders are frequently described with full-blown co-morbid disorders as well. Results of studies of a heightened risk for autism spectrum disorders and attention-deficit/hyperactivity disorder are contradictory (Cassidy & Driscoll, 2009; Dykens et al., 2000; Dykens & Shah, 2003; Goldstone et al., 2008; Hiraiwa, Maegaki, Oka, & Ohno, 2007; Holm et al., 1993; Walz & Benson, 2002).

This is the first study, as far as we know, to investigate the perception of parenting stress in AS. In PWS two studies on parenting stress have been carried out, in which high stress levels were found (Hodapp, Dykens, & Masino, 1997; Sarimski, 1997a). Furthermore, in PWS, parenting stress appeared to be related to behavioural problems but not to gender, age, IQ, or degree of obesity of the child (Hodapp et al., 1997). For this study we have chosen to report on a relatively homogeneous group: all children are 2- to 12-years-old and are living at home. It is still unclear whether mothers and fathers of children with intellectual disability perceive similar parenting stress levels, since the results are mixed (Hassall & Rose, 2005; Hastings & Beck, 2004; Olsson, 2008). To rule out the unknown effect of gender, only the results for maternal parenting stress are included. Following these choices, the *first aim* of this study was to test the hypothesis that mothers with a child with AS or PWS perceive high levels of parenting stress. The *second aim* was to test the hypothesis that certain child characteristics are related to maternal parenting stress (within-syndrome). The *third aim* was to compare the level of maternal parenting stress between the two syndromes. The investigated child characteristics are: gender, age, behavioural problems, and level of intellectual disability. To our knowledge this is the first study to explore which characteristics of children with AS are related to maternal parenting stress. In PWS, it is expected that there will be no relationship with the child's gender or age, but that there will be a positive relationship with behavioural problems, as described by Hodapp et al. (1997). It appeared that IQ is not related to parenting stress (Hodapp et al., 1997), but the level of adaptive functioning might be a better indicator of the actual functioning of children with PWS. Therefore, adaptive functioning is used to classify the level of intellectual disability and the relationship of this characteristic with maternal parenting stress is explored. With this project we aim to

expand our knowledge about those child characteristics that are of specific relevance to the maternal perception of the child-rearing experience in these two syndromes and also add knowledge about the differences in maternal parenting stress between the syndromes. The ultimate goal is to contribute to better and more specific support for these families.

METHOD

Procedure

With permission of the board of the Dutch PWS/AS Parent Support Group, all its members were invited by means of a letter to participate in the current study. Ethical guidelines of the Royal Netherlands Academy of Art and Sciences (KNAW) were followed to recruit the participants, and written informed consent was obtained from the participants. Of the AS group, 75 parents (53%) joined the project, and 67 PWS parents (30%) reacted positively to the request. In the current study, data were used for children aged 2- to 12-years-old who were living at home, had a definite diagnosis of either AS or PWS, and whose mothers filled out the questionnaires. The percentage and number of participants fitting the criteria were comparable: 24 children with AS (32%) and 23 children with PWS (34%). Parents received the questionnaires by post and were asked to return them in the pre-paid envelope. Parents were requested to identify their child's gene mutation type. If a parent was uncertain about this, written permission was obtained to request this information from the child's medical specialist.

Participants

Twenty-four children with AS (11 boys, 13 girls) and 23 children with PWS (10 boys, 13 girls) and their mothers participated. The distribution of gender did not differ between the syndromes (χ^2 (1) = .03, p = 1.00). The age range was 2 to 12 years (AS M = 8.6, SD = 3.10; PWS M = 7.3, SD = 3.16), and the children of both syndromes did not differ in their age (t (45) = -1.38, p = .18). The following gene mutations were found for the children with AS: in 67% a deletion on the maternal chromosome 15, in 17% a paternal uniparental disomy, in 4% an imprinting defect, in 4% an *UBE3A* gene mutation, and in 8% no gene mutation was found but the AS diagnosis was given by a medical specialist. All children with PWS had gene mutations: in 57% a maternal uniparental disomy, in 35% a deletion on the paternal chromosome 15, in 4% an imprinting defect,

and in 4% a gene mutation was found but further specification of mutation type was absent.

Research instruments

All questionnaires used in this study conform to the official manuals. The following instruments were used.

The *Nijmegen Parenting Stress Index-Short* (NPSI-S; De Brock, Vermulst, Gerris, & Abidin, 1992) is an officially translated and adapted version of the Parenting Stress Index by Abidin (1983, as cited in De Brock et al., 1992). It measures parenting stress in families with children from approximately 2 to 13 years. Parents (in this case mothers) rate 25 items on a 6-point scale. All scores on the 25 items are summed to make up the total score. The total score is classified into seven norm categories defining parenting stress level. Dutch non-clinical and clinical norms are available with separate norms for mothers and fathers. The non-clinical norm group is made up of families from the normal population; the clinical norm group exists of parents with a child who is admitted to mental health services. The non-clinical norms for mothers were used in this study. Cronbach's alpha for internal consistency is .95. The NPSI-S shows good criterion validity with accurate prediction of membership of the clinical and non-clinical population. Construct validity is only investigated for the extended version of the instrument: concurrent validity ranges from satisfactory to good, and discriminant validity is considered reasonable (De Brock et al., 1992).

The Dutch version (Koot & Dekker, 2001) of the *Developmental Behaviour Checklist-Primary Carer* (DBC-P; Einfeld & Tonge, 2002) assesses emotional and behavioural problems exhibited over the past six months by children with intellectual disability. Parents rate 95 items on a 3-point scale. A total behaviour problem score and five subscale scores can be computed. A clinical cut-off point is only available for the total behaviour problem score; it has good sensitivity and specificity to distinguish clinical cases (Einfeld & Tonge, 2002). The intra-class correlation for inter-rater reliability is .55 for the total problem behaviour score. Internal consistency (Cronbach's alpha .95) and test-retest reliability (intra-class correlation .86) are high. Construct and criterion validity are satisfactory (Koot & Dekker, 2001).

The *Vineland Screener 0-12 years* (VS 0-12; Van Duijn, Dijkxhoorn, Noens, Scholte, & Van Berckelaer-Onnes, 2009) is a Dutch screening instrument adapted from

the Vineland Screener by Sparrow, Carter, and Cicchetti (1993). The VS 0-12 measures the level of adaptive functioning of children up to age 12 or older persons with comparable levels of functioning. An adaptive behaviour composite score is based on four domains (Communication, Daily Living Skills, Socialisation, Motor Skills). Unlike the Vineland Screener by Sparrow et al. (1993), the Dutch VS 0-12 does not include an optional section on maladaptive behaviour. Parents indicate on a three-point scale for 90 items whether the child exhibits that particular behaviour in everyday life. Good reliability and validity have been established for a normal population. Inter-rater reliability for the adaptive behaviour composite has an intra-class correlation of .98 and test-retest reliability of .95. Cronbach's alpha is .99 (Van Duijn, Dijkxhoorn, Noens, et al., 2009). The VS 0-12 years is an expansion of the VS 0-6 years which has adequate content, construct, and criterion validity (Scholte, Van Duijn, Dijkxhoorn, Noens, & Van Berckelaer-Onnes, 2008). A regression formula was developed based upon normal population data to estimate the adaptive level of functioning (Van Duijn, Dijkxhoorn, Van Berckelaer-Onnes, Scholte, & Noens, 2010). In the first data wave parents did not fill out the VS 0-12 but were interviewed with the Vineland Adaptive Behavior Scales (Sparrow, Balla, & Cicchetti, 1984). However, the interview appeared to be so time-consuming for the parents that it was replaced by the VS 0-12 questionnaire for the other participants. The relevant items from the interview were used to complete the VS 0-12 for the first 13 mothers with a child with PWS.

Data analysis

The data were analysed with SPSS 16.0, and an alpha of .05 was chosen for all analyses. Univariate outliers were given the next highest score plus or minus one, depending on whether the outlier was at the higher or lower end. The Shapiro-Wilks test was used to check whether the data deviated from a normal distribution and parametric tests could be used. The effect sizes for *t*-tests were given by *r* whereby .10 is viewed as a small effect, a .30 medium effect, and .50 as a large effect. For comparison of categorical data Pearson chi-square tests for association were used. If the expected count in one or more cells was less than 5, Fisher's exact tests were used. Phi was used as effect size for categorical data and the same rule of thumb for the size of the effects was applied (Field, 2009).

The level of intellectual disability was estimated on the basis of the level of adaptive functioning as measured with the VS 0-12. For children up to nine years of age, a developmental quotient (DQ) [VS 0-12 developmental age / chronological age x* 100] was computed and the level of intellectual disability was subsequently classified based upon Došen (2005) (see Table 6.1). Children aged 10 to 12 years can no longer obtain a DQ of 100 with the current VS 0-12 regression formula. For the children the classification was based upon the adaptive developmental age (see Table 6.1). It was decided to dichotomise variables, except age, because of the small number of participants. For the NPSI-S the two highest norm categories, high and very high stress, were coded as high maternal parenting stress. Scores for the other norm categories were coded as the low maternal parenting stress group. For the DBC-P, clinical caseness of behavioural problems was used to define groups with high versus low levels of behavioural problems. The level of functioning was dichotomised into profound/severe/moderate intellectual disability and mild/no intellectual disability.

Table 6.1 *Classification of intellectual disability based on Došen (2005)*

Level of intellectual disability	**Developmental quotient**	**Developmental age**
Severe/profound	0 - 35	0.0 - 4.9 years
Moderate	36 - 50	5.0 - 7.9 years
Mild	51 - 70	8.0 - 12.9 years
None	> 70	> 12.9 years

RESULTS

Maternal parenting stress in AS and PWS

Mothers with a child with AS perceived high levels of parenting stress (see Table 6.2). None of them scored in the norm categories very low to below the mean (norm group 35%). The scores of 29% of mothers fell in the category high parenting stress and another 29% in the category very high parenting stress. In PWS, only 9% of the mothers reported stress levels below the mean. The percentage of mothers who scored in the highest two categories (17% and 9%) was somewhat higher than in the norm group. After

dichotomisation maternal parenting stress was coded as high in 58% of mothers with a child with AS and 26% of mothers with PWS.

Table 6.2 *Parenting stress of mothers with a child with Angelman syndrome (n = 24) or Prader-Willi syndrome (n = 23*

Parenting stress NPSI-S category non-clinical norm group		**Angelman syndrome % (*n*)**	**Prader-Willi syndrome % (*n*)**
Category	Percentiles in norm population		
Very low	0% - ≤ 5% (5%)[a]	-	-
Low	5% - ≤ 15% (10%)	-	-
Below the	15% - ≤ 35% (20%)	-	9 % (2)
Mean	35% - ≤ 65% (30%)	29% (7)	22% (5)
Above the	65% - ≤ 85% (20%)	13% (3)	43% (10)
High	85% - ≤ 95% (10%)	29% (7)	17% (4)
Very high	95% - ≤ 100% (5%)	29% (7)	9% (2)

Note. NPSI-S = Nijmegen Parenting Stress Index-Short.
[a] Percentage of total norm population in parentheses

Child characteristics in AS and PWS

The DBC-P provides insight into which behavioural problems were the most prevalent among the children. In AS (*n* = 24) the following 15 items received a score of 1 or 2 in more than 70% of cases: becomes over-excited; chews or mouths objects, or body parts; easily distracted from task; eats non-food items; impatient; likes to hold or play with an unusual object; makes non-speech noises; overactive; poor attention span; poor sense of danger; repeated movements of hands, body, head, or face; sleeps too little, disrupted sleep; stubborn, disobedient or unco-operative; unrealistically happy or elated; unusual body movements, posture, or way of walking. In PWS (*n* = 23), there was more variation in behavioural problems; only six items were scored in more than 70% of cases: arranges objects or routine in a strict order; easily distracted from task; easily led by others; impatient; poor sense of danger; scratches or picks at skin; stubborn, disobedient or unco-operative; upset over small changes in routine or environment.

Substantial behavioural problems (clinical range) were found for approximately half of the children with AS (13, 54%) and a third of the children with PWS (8, 35%).

There was no significant association between type of genetic syndrome and number of behavioural problems (χ^2 (1) = 1.79, p = .24).

According to the VS 0-12 the adaptive level of functioning ranged in AS (n = 23; for one person, data were missing) from 0 to 2.76 years and in PWS (n = 23) from 0.28 to 8.40 years. On the basis of these data, the level of intellectual disability was estimated. All children with AS were categorised as having a severe/profound intellectual disability. In PWS, 15 children (65%) were categorised as having mild or no intellectual disability, and 8 children (35%) were categorised as having moderate/severe/profound intellectual disability.

Maternal parenting stress within and between AS and PWS

In AS there was no significant association between high or low levels of maternal parenting stress and the child's gender (Fisher's exact p = 1.00). There was no difference in age of the child between mothers with high versus low levels of stress (t (22) = .65, p = .52). No association was found between the level of maternal parenting stress and a high versus low amount of behavioural problems (Fisher's exact p = .70). Since all children with AS had a severe/profound intellectual disability, no association with level of maternal parenting stress could be investigated.

In PWS there was no significant association either between maternal parenting stress and the gender of the child (Fisher's exact p = .18). There was no difference in the child's age between the mothers with high versus low levels of stress (t (21) = -1.56, p = .13). No significant association was found between maternal parenting stress and behavioural problems (Fisher's exact p = .13). Level of maternal parenting stress was compared for children functioning on a moderate/severe/profound level versus children functioning on a mild/no intellectual disability level. There was no significant association (Fisher's exact p = .62).

A comparison was also made between the two syndromes with regard to the level of maternal parenting stress. As shown in Table 6.2, 58% or 14 AS mothers reported high levels of stress, while in PWS the comparable figure was 26% or 6 mothers. These figures suggest that mothers of a child with AS more often perceive high stress than mothers of a child with PWS. Statistical testing confirmed this hypothesis, (χ^2 (1) = 5.00, p = .03). With Φ = -.33, this was a medium effect.

A further analysis revealed that the behavioural problems of the children played a mediating role in the maternal perception of stress in both syndromes, as is shown in Tables 6.3 and 6.4.

If the children had no behavioural problems (see Table 6.3), a comparable picture emerged for the total group. Compared to mothers with a child with PWS, significantly more mothers with a child with AS reported high levels of parenting stress (Fisher's exact p = .01). In the subgroup of children without behavioural problems, this effect can be described as large with Φ = .52.

Table 6.3 *Distribution of maternal parenting stress for children with AS (n = 11) and PWS (n = 15) without behavioural problems*

		Maternal parenting stress		
		Low	**High**	**Total**
AS	*N*	4	7	11
	% within syndrome	36%	64%	100%
	% within maternal parenting stress	23%	78%	42%
PWS	*N*	13	2	15
	% within syndrome	87%	13%	100%
	% Within maternal parenting stress	77%	22%	58%
Total	*N*	17	9	26
	% within syndrome	65%	35%	100%
	% within maternal parenting stress	100%	100%	100%

Note. AS = Angelman syndrome; PWS = Prader-Willi syndrome.

However, when the children had behavioural problems (see Table 6.4), there was no association between the perceived levels of maternal parenting stress and the two syndromes (Fisher's exact p = 1.00), implying that the levels of stress perceived by the mothers are equal for AS and PWS when coping with a behaviourally difficult child is involved.

Table 6.4 *Distribution of maternal parenting stress for children with AS (n = 13) and PWS (n = 8) with behavioural problems at a clinical level*

		Maternal parenting stress		
		Low	**High**	**Total**
AS	*N*	6	7	13
	% within syndrome	46%	54%	100%
	% within maternal parenting stress	60%	64%	62%
PWS	*N*	4	4	8
	% within syndrome	50%	50%	100%
	% Within maternal parenting stress	40%	36%	38%
Total	*N*	10	11	21
	% within syndrome	48%	52%	100%
	% within maternal parenting stress	100%	100%	100%

Note. AS = Angelman syndrome; PWS = Prader-Willi syndrome.

DISCUSSION

To our knowledge, this is the first study to investigate the perceived parenting stress of mothers with a child with AS and to compare the stress between mothers with a child with AS and those with PWS. In line with the *first* hypothesis, the child-rearing experience is related to high levels of maternal parenting stress. Specifically, many more mothers with children with AS reported high stress levels as measured by the NPSI-S (58%) compared to the normal population (15%). In PWS, parenting stress was high for 26% of mothers.

The *second* aim was to investigate the relationship between maternal parenting stress and child characteristics. For AS, gender, age, and behavioural problems were assessed. No relationship was found between maternal parenting stress and these child characteristics. The lack of variation in level of intellectual disability prevented a comparison for that characteristic. The most prominent pattern in families with a child with intellectual disability, and in most genetic syndromes, is higher parenting stress when more behavioural problems are present (Hassall & Rose, 2005; Hastings & Beck, 2004; Hatton & Emerson, 2003; Hodapp, 1999; Olsson, 2008). This was, however, not applicable to AS; mothers with a child with a low amount of behavioural problems

reported the same amount of parenting stress as mothers whose child displayed a clinical amount of behavioural problems. Thus, other child characteristics might be related to parenting stress in this syndrome. It could be that difficulties with communication, both to make things clear to the child and to interpret the child's intentions, is a stress inducing and prominent characteristic. Also, the low level of functioning of the child in general could make the upbringing more stressful. To investigate this hypothesis, a control group with children with the same level of functioning and without speech is needed.

In PWS, maternal parenting stress was not related to the child's gender or age. This is in line with earlier research on PWS (Hodapp et al., 1997). The level of intellectual disability, based on adaptive functioning, was not related to maternal parenting stress in PWS. This result strengthens and extends our knowledge based on Hodapp et al., who found no relationship between parenting stress and IQ. Also, there was no relationship with behavioural problems, and this is at odds with what others have found (Hodapp et al.). There are several possible explanations for this difference. We used an instrument specifically developed for children with intellectual disability. As a proportion of the participants functioned in the borderline range to normal functioning, it might be that some characteristic behavioural problems were not measured by this questionnaire. However, the DBC-P appeared more relevant for the participants with intellectual disability. Another explanation for the difference in results between our study and Hodapp et al. could be the age composition of the two samples. In the current study families with *children* participated, whereas Hodapp et al. included adolescents as well. Steinhausen, Eiholzer, Hauffa, and Malin (2004) found DBC-P behavioural problems in PWS to be more prevalent in the age group 13-29 years compared to the age groups 2-7 and 7-13 years. Thus, more prominent behavioural problems in adolescents could give rise to the different results for the relationship between parenting stress and behavioural problems. However, further studies with different age cohorts are needed to confirm this hypothesis.

The *third* aim was to compare stress levels between mothers with a child with AS and those with PWS. Overall, more mothers with a child with AS reported high stress levels due to the child-rearing experience. However, the presence of a clinical behaviour problems was a mediating factor for maternal parenting stress in the two syndromes. Among children with low levels of behavioural problems, mothers with a child with AS perceived more stress. When the child had a clinical amount of behavioural problems, there was no difference in parenting stress between mothers with a child with AS and one

with PWS. Thus, it can be said that mothers with a child with AS have overall high stress levels, whereas mothers with a child with PWS experience this only when their child has significant behavioural problems. This result could be added to the knowledge that parents with a child with AS have higher levels of loss of control compared to parents with a child with PWS (Van den Borne et al., 1999). Although the syndromes are genetically related, they differ in many respects, such as the level of functioning and behaviour. AS in general seems to be stress-inducing, whereas in PWS more specific behavioural problems relate to stress. We hypothese that some of the most prominent characteristics of AS, severe/profound intellectual disability and absence of speech, might explain why raising a child with this syndrome is a heavy burden, independent of the presence or absence of behavioural problems.

The findings suggest that professional support for families with a child with AS is needed, because stress levels are high in a large proportion of mothers, which can have a negative influence on parenting behaviour (Deater-Deckard, 2004). In PWS, the need for support is more prominent when the child exhibits substantial behavioural problems. In that case, parents should get additional support to manage the behavioural problems, which may result in reduced parenting stress (Hastings & Beck, 2004). It seems important to provide parents with information on parenting stress as related to their child's syndrome. Parents with a child with AS or PWS have a substantial need for information on other child-related issues (Van den Borne et al., 1999). Information on parenting stress in young families might give a realistic description of family life and consequently might better prepare them for future challenges. Wigren and Hansen (2003) reported that parents with a child with PWS mainly wanted general information and support as opposed to family-directed support. It is important that future studies measure parenting stress *and* the desire for support of parents simultaneously. If both components are studied concurrently, professional care and parental satisfaction with this care might be improved. Furthermore, for families with a child with one of the two syndromes, professional support should be a continuous process, since the perception of stress is not related to the child's age. Professional aid is presumably also needed during adolescent years, since Hodapp et al. (1997) found no relationship between parenting stress and the child's age for children with PWS from 3 to 18 years.

There are some limitations of the current project. First, we used only a limited set of child characteristics to relate to maternal parenting stress, while important

environmental and parental characteristics also influence the stress process; for example the socio-economic status of the family and parental cognitions (Hassall & Rose, 2005; Perry, 2004). In addition, measuring positive outcomes among parents is also crucial because it has been shown that there is a large variation in parental experiences with raising a child with intellectual disability. Many parents adapt well to the highly specific demands of parenting a child with disability and, for instance, experience personal growth (Hassall & Rose, 2005, Hatton & Emerson, 2003; Head & Abbeduto, 2007; Olsson, 2008). When more of the relevant child, parental, and environmental characteristics are included in an analysis, a more coherent description of these families will be obtained. Second, causality could not be established because of the cross-sectional nature of the study. For persons with intellectual disability in general the results are mixed whether the child's behaviour problems cause parenting stress or whether there is a bi-directional effect (Hassall & Rose, 2005; Hastings & Beck, 2004; Olsson, 2008). To investigate the causality of relationships, a longitudinal study is needed (Hatton & Emerson, 2003). This is an important aim since it can refine the design of family support. Third, the information with regard to the child's behavioural problems and parenting stress was provided by the same type of informant; that is the mother. This may have influenced the results. Further studies are needed with additional informants like fathers and/or teachers to assess independently of the mother the child's behavioural problems and to relate these findings to the behavioural problems the mothers report and the stress they perceive. Fourth, like other studies of rare genetic syndromes, the small number of participants results in a lack of statistical power. According to Cohen (1992) with an alpha of .05, preferred power of .8 and 26 participants, large effect sizes are needed to obtain statistically significant outcomes with chi-square tests (1 *df*). Results should thus be interpreted with caution. Finally, participants were gathered by the Dutch PWS/AS Parent Support Group. Parents who belong to such support groups are very often highly motivated and from middle to high socio-economic background (Dykens, 1999), and thus may not be representative of all Dutch families with a child with AS or PWS. In addition, only a proportion of all members of the support group agreed to participate. Families in this self-selected sample may have additional specific characteristics which unfortunately remain unknown. However, concerning the children's behaviour we assume to have had a representative sample of children with AS and PWS. The behavioural problems most frequently encountered in this study showed roughly the same pattern as in other studies of the AS

and PWS behavioural phenotypes. It is, however, remarkable that the item on overeating was not scored for more than 70% of the children with PWS, which is contrary to expectations. Possibly parents are so used to this behaviour, as it is a core symptom of the syndrome, that they do not report it any more. In sum, although the behaviour of the children seems representative, caution is needed concerning generalisation of the results as these may be biased by the selection procedure.

In conclusion, this study contributes to our knowledge about the maternal perception of raising a child with AS or PWS. In clinical practice these results can guide the intervention process and ultimately optimise the development of children with these syndromes and the families they grow up in. We should aim to capture the interplay of a lot of different factors to better approach the situation in real life. We agree with Olsson (2008) that it is most important to focus on the processes that lead to different outcomes in families and to include negative and positive outcomes at the same time. Why do some families adapt well to their specific situations? Unraveling these complex processes can provide important clues for clinical practice.