

## Original articles

# Parental training for the behavioral management of children with Prader-Willi syndrome: impact on the mental health and parenting practices of the caregiver

*Treino parental para manejo comportamental de crianças com síndrome de Prader-Willi: impacto sobre a saúde mental e práticas educativas do cuidador*

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## ABSTRACT

**Purpose:** we verified the impact of the parent training program for management of children and adolescents with Prader-Willi Syndrome on the mental health caregivers.

**Methods:** the sample was composed by five mothers of children / adolescents with Prader-Willi Syndrome between 6-18 years. The data collection instruments were: a) Questionnaire for verification Mothers' knowledge about the syndrome, b) Inventory Self-Assessment for Adults 18-59 years c) Inventory of Parental Styles / maternal and paternal educational practices; d) Survey of difficulties and concerns about their children. The study was conducted in four phases: pre-intervention, intervention, post-intervention and follow-up.

**Results:** as a result we find that mothers have adopted parental educational practices based on positive monitoring, decreased risk practices.

**Conclusion:** we conclude that there was an improvement in indicators of emotional difficulties of mothers, seized identify and manage factors that harmed the physical health, behavior and school learning of the children and the need to involve other family members in child care.

**Keywords:** Prader-Willi Syndrome; Behavior; Parents

## RESUMO

**Objetivo:** verificamos os indicadores de impacto na saúde mental de cuidadores de um programa de treinamento parental para manejo de crianças e adolescentes com Síndrome de Prader-Willi.

**Métodos:** a amostra foi composta por 5 mães de crianças/adolescentes com Síndrome de Prader-Willi entre 6 a 18 anos. Os instrumentos de coleta de dados foram: a) Questionário para verificação de conhecimentos das mães sobre a síndrome, b) Inventário de Auto-avaliação para Adultos de 18 a 59 anos, c) Inventário de Estilos Parentais/Práticas educativas maternas e paternas; d) Levantamento de dificuldades e preocupações sobre os filhos. O estudo foi realizado em quatro fases: pré-intervenção, intervenção, pós-intervenção e seguimento.

**Resultados:** as mães passaram a adotar práticas parentais educativas baseadas na monitoria positiva, diminuíram as práticas de risco.

**Conclusão:** houve melhora em indicadores de dificuldades emocionais das mães, apreenderam a identificar e manejar fatores que prejudicavam a saúde física, o comportamento e a aprendizagem escolar dos filhos e a necessidade do envolvimento de outros familiares no cuidado do filho.

**Descritores:** Síndrome de Prader-Willi; Comportamento; Pais

## INTRODUCTION

Prader-Willi Syndrome is a genetic disease caused by the lack of expression of genes on a region of the paternal chromosome 15q11-13 involving the genomic imprinting phenomenon. Genes in this region have differential expression according to parental origin, so that the paternal and maternal copy must be present for normal gene expression. Laboratory diagnosis is established in 95% of cases, with a predominance of a paternal deletion subtype in 70% of affected individuals, with uniparental disomy in 25% of cases (both chromosomes 15 are of maternal origin and neither of paternal origin). In other cases occurrences of the imprinting phenomenon or other rearrangements of chromosome 15 are reported. The disease incidence is 1: 10,000 to 30,000 births<sup>1,2</sup>.

The cognitive and behavioral phenotype observed in people with PWS is characterized by frequent episodes of hyperphagia, stealing food, tantrums, lying behavior, mood swings, anxiety, sadness, aggression and auto aggression, stubbornness and repetitive speech, hypersomnia, compulsive behaviors, deficits in cognitive abilities such as attention and executive functions (especially inhibitory control, working memory and self-regulation) and deficits in visuospatial skills<sup>3-7</sup>. Of these described characteristic phenotypes, the problems most difficult to manage are hyperphagia; stubborn behavioral patterns; challenging, aggressive, oppositional and manipulative behaviours; food theft and lying, which become more frequent and severe with increasing age<sup>8-11</sup>.

Parents and caregivers of people with PWS are routinely faced with two types of problems; the control of access to food to avoid and/or minimize other health problems (such as obesity, scoliosis, and cardiovascular and endocrine illnesses)<sup>12</sup>, and family burden, especially on the primary caregiver, due to the difficulties in the management of behavioral problems that start from early childhood<sup>13,14,15</sup>. Although scientific evidence has shown the neurobiological basis of the behavioral changes and patterns of hyperphagia, interventions which involve giving guidance to parents and caregivers in respect of parenting practices and the management of the children's behavioral problems are considered to be a priority; previous studies report that these interventions can not only improve a range of behavioral indicators in the children but also help support the caregiver in relation to the emotional difficulties they face<sup>16-21</sup>.

Parents and/or caregivers in general are the main agents of change in any type of behavioral management of their children<sup>22</sup>. Parenting practices adapted to the main care demands of a person with PWS may result not only in a reduction in individual behavioral problems related to the syndrome, but also in improving indicators of family relationships and reducing the mental health problems of parents. Studies point to the need to educate and train caregivers in the proper management of overeating and progressive weight gain<sup>23,24</sup>. Evidence shows that providing care is associated with caregiver psychological profiles which are characterized by stress states, anxiety disorders and depression in at least one member of the couple<sup>25</sup>.

In Brazil, the monitoring of people with rare diseases is complex, and although PWS is a syndrome that is associated with moderate to severe levels of intellectual disability and should in fact be treated in Psychosocial Care Centers (CAPS) within the health system, the diagnosis is often delayed, consequently so too are the necessary care actions and psychological interventions<sup>26</sup>. It is also the case that there are many difficulties and inconsistencies in public policies for rare diseases in the country, and only a few regions have interdisciplinary treatment centres for patients and their families, with most of these being concentrated in the south and southeast<sup>27,28</sup>. Thus, access to health services that provide psychological, psychiatric and general medical care are also limited for those affected<sup>29</sup>. In Brazil, studies focused on monitoring people with PWS, and family interventions remain scarce<sup>30,31</sup>. The aim of this study was to assess the impact of parental training for the management of the behaviors of children with PWS on the mental health and parenting practices of the caregiver.

## METHODS

### Sample

This is a cross-sectional study with a non-probabilistic sample of five mothers of children and adolescents aged between 6 and 18 with both a clinical and cytogenetic-molecular diagnosis of Prader-Willi syndrome, confirmed by medical record from the genetics department of the faculty of medicine of the referring institution. The participating mothers were recruited during routine health service appointments for their child. The invitation to take part was made to each mother individually after being given a presentation about the project in a private room. Those who agreed

to participate were then asked to sign an informed consent in accordance with the resolution of the Ethics Committee on Research with Human Beings. The study was approved under the process CEP/UPM No 1195/02/2010 and CAAE No. 0001.0.272.000.10 of the

Universidade Presbiterana Mackenzie. All requirements established by the National Health Council regarding research with human subjects were observed. Tables 1 and 2 describe the characteristics of the sample's participants (caregivers and children with PWS).

**Table 1.** Sociodemographic characteristics of participating caregivers/mothers in relation to age, educational level, marital status, occupation and family income

Mother	Age in years	Level of education	Marital Status	Occupation	Family income in Reais
1	45	High School Completed	Married	Homemaker	1.600,00
2	44	Primary School Incomplete	Married	Cleaner	1.300,00
3	34	High School Completed	Married	Homemaker	2.800,00
4	50	Primary School Incomplete	Married	Homemaker	1.520,00
5	46	High School Incomplete	Married	Homemaker	1.500,00

**Table 2.** Characteristics of children with Prader Willi syndrome in relation to Age, Sex, I.Q, Molecular Diagnosis, Aspects of Assistance and use of Medication

Child	Age in Years	Sex	IQ Estimated by WISC-III and Classification	Molecular Diagnosis	Use of mental health services and medication
1	6	M	51 (Mild intellectual disability)	Deletion 15q11-13	Does not use mental health services. Attendance at genetic service for follow up. Does not use medications.
2	13	M	56 ( Mild intellectual disability)	MUD	Does not use mental health services. Attendance at genetic service for follow up. Uses Phenobarbital and Fluoxetina.
3	14	M	77 (Borderline)	MUD	Does not use mental health services. Attendance at genetic service for follow up. Risperidone and growth hormone.
4	15	M	48 (Moderate Intellectual disability)	Deletion 15q11-13	Does not use mental health services. Attendance at genetic service for follow up. Topiramate, Metformin, Glibenclamide Simvastatin and Enalapril.
5	15	F	59 (Mild intellectual disability)	Deletion 15q11-13	Does not use mental health services. Attendance at genetic service for follow up. Growth hormone.

**Legend:** M- male; F- female; IQ- Intelligence Quotient; WISC III – Wechsler Intelligence Scale for Children (WECHSLER, 2002)<sup>32</sup>; MUD =Maternal Uniparental Disomy.

## Data collection instruments

**a) Adult Self Report 18 to 59 - ASR/18-59 years:** a self-administered inventory, developed by Achenbach and Rescorla<sup>33</sup>. It aims to check different aspects of adaptive functioning and behavioral and emotional problems of adults aged between 18 and 59. The inventory consists of items that assess skills with issues relating to friends, spouse or partner, family, work and study. The items allow an assessment of the quality of relationships of the adult. They explore the presence of disease, disability or incapacity, concerns or anxieties about family, work, education, and positive aspects or qualities that the person thinks about themselves. The scales of the **ASR / 18-59 years** are distributed as follows:

Profile of adaptive functioning scales: comprised of questions relating to friends, spouse/partner, family, job and study.

Profile of syndrome scales: comprising scales for anxiety/depression, withdrawn, somatic complaints, thought problems, attention problems, aggressive behaviour, rule-breaking behaviors, and intrusive.

Profile of DSM oriented scales: comprising scales for depressive problems; anxiety problems; somatic problems; avoidant personality problems; attention deficit/ hyperactivity problems (inattention and hyperactivity/impulsivity subscales), and antisocial personality problems

Internalized, externalized and total behavioral problems.

Substance Use Scales: Tobacco, Alcohol, Drugs, mean substance use.

**b) Parenting Styles Inventory (PSI):** a self-report inventory that assesses maternal and paternal parenting practices comprising 42 questions related to seven practices<sup>34</sup>. Among these, two are labelled positive practices (positive monitoring and moral behavior) and five negative (inconsistent punishment, neglect, relaxed discipline, negative monitoring and physical abuse). Two forms of the instrument presentation are available a) when parents respond about the parenting practices used with their children; b) when the children respond about the parenting practices used by their parents (paternal parenting practices and maternal parenting practices). The method used in this study was the parents responding about the practices they used with their children.

**c) Questionnaire on the knowledge of PWS:** a questionnaire developed for the study to obtain information on the level of knowledge of the mothers

about the disease of their children. The questions that were asked were: “What do you know about Prader Willi Syndrome?”; “What are the characteristics of the syndrome?”; “How did you get this information?”; “Where do you get guidance?” And “How does your family deal with the syndrome?”

**d) Survey of the main concerns and difficulties faced by the mothers during daily routines with their children:** these data were discussed and handled together with the respective guidelines to mitigate and/or solve *these issues* through the parental training program. To do this, the mothers were given tasks to do at home which was discussed in the subsequent meeting. This homework included tasks such as making a list of the concerns and difficulties they faced during the care of their child. The tasks also covered an exploration of the factors related to the family environment that interfered with the child’s management. For example, support received from family, school aspects, the influences of stress and social and family factors on the behavior of the children and difficulties in the behavioral management of the child, among others. Hereafter follows a description of the training program.

## Parental Training Program

The meetings throughout the study were conducted in a private room with appropriate physical conditions to meet the needs of the group, with ample space, chairs and sound insulation. The researcher made use of a computer and a digital projector. The training program covered the following themes: educational training on the general characteristics of Prader-Willi syndrome: cognitive phenotype, behavioral and psychiatric disorders associated with PWS; inclusion and school context; the relationship between the disciplinary and parenting practices of parents and the appropriate and inappropriate behaviors of children and adolescents with PWS; guidance on behavioral management strategies and the influence of family stress and social and family factors on the behavior of children. These things were discussed with the mothers in eleven three-hour meetings. Each meeting had tasks that were discussed at the subsequent meeting. The tasks had the following objectives: to answer questions or concerns arising from reading the psychoeducational information guide about the syndrome that was given to the mothers<sup>35</sup>, highlight examples of cognitive abilities preserved in the child, schedule a meeting with the educational team of the school and talk to teachers, inform and guide the staff on eating behaviors, identify and describe familiar

situations that illustrate the use of positive and negative parenting practices, identify at least three appropriate and inappropriate behaviors of the child and use their training to classify them as examples of either internalizing or externalizing behaviors, list and prioritize inappropriate behaviors they wish to reduce or extinguish and appropriate behaviors they wish to maintain or increase, record instances of family stress situations and how they could these situations, identify the family members who exercise the greatest positive or negative influence on the child's behavior and guide family members, and finally raise any doubts or concerns and complete an evaluation of the training. The training had a 100% attendance record with all given tasks being completed satisfactorily.

### Data collection procedures

This study was conducted in four phases with a total duration of 24 months. Phase 1 was the pre-intervention assessment (duration: 1 four-hour meeting) in which the data collection instruments for the children/adolescents and mothers were completed; Phase 2 was the intervention (duration: 11 three-hour meetings with an interval of 20 days between them), with the completion of the parental training program described above; Phase 3 was the post-intervention assessment (duration: 1 three-hour meeting), with the reapplication of the instruments used to collect data on the children/adolescents and mothers; Phase 4 was the follow-up (duration: 1 three-hour meeting) with an interval of three months between them, and reapplication of the same instruments used to collect data on the children/adolescents and mothers.

### Data analysis procedures

The questionnaire data on the mothers' knowledge of the syndrome, and the data on the main concerns and difficulties they faced during their daily routines with their children were analyzed qualitatively. The responses of the mothers to the questionnaire on their knowledge of the syndrome were grouped together for each question and tabulated for better data visualization. For the difficulties and concerns identified by the mothers, we chose to include them in a qualitative way in the data analysis of the standardized instruments. That is, they were analyzed together with the results of the behavioral inventories which evaluated the emotional and behavioral problems of the caregivers, as well as the inventory of parenting styles.

The results from the application of the behavioral assessment tools were analyzed by means of specific standardizations. For the generation of the behavioral profiles from the ASR/18-59, the program *Assessment Data Manager 7.2*<sup>33</sup> was used. The behavioral data from the ASR were analyzed according to the number and classification of problems in the clinical, borderline or normal groups according to the manual of the instrument<sup>33</sup>. The interpretation of the PSI data was conducted according to the manual of the instrument<sup>34</sup>. Descriptive data analyses were conducted comparing the phases of pre-intervention, intervention, post-intervention and follow-up.

## RESULTS AND DISCUSSION

The information on the mothers' knowledge of the general aspects of the syndrome in the pre-intervention phase is shown in Table 3. The results of the interviews with the mothers to check their knowledge of the syndrome show that, in general, the information they have on the disease of their children is limited and mainly based on the key clinical indicators, whether behavioral or clinical, namely, hyperphagia, body weight and the physical characteristics. This lack of knowledge of the mothers is probably associated with how they handle the various behavioral difficulties of their children, underestimating or not recognizing problems as being immanent to the syndrome. In fact, as shown in Table 3, when they were asked about their usual management of the condition, the responses indicated that mothers had adopted parenting strategies assuming that the affected child had the same characteristics as a child with typical development. Previous studies highlight the need for caregivers to understand the development of the phenotype's characteristics in different age groups throughout the condition; changes related to behavior and socialization being some of these. For example, in early childhood acceptable standards of features such as social interaction, tantrums, stubbornness, motor difficulties and ability to make friends are often observed. From adolescence to adulthood, aggressive behavior, lying and psychiatric symptoms in the obsessive-compulsive spectrum are commonly observed and lead to many emotional impacts, both for the child and for the parents<sup>9,36</sup>. Goldstone and colleagues also highlighted the need for the family to know precisely the specific characteristics of the behavioral and psychiatric symptoms which are often associated with the syndrome<sup>37</sup>. Lack of knowledge can undermine the role of the carer, and may exacerbate

many behavioral problems and symptoms associated with PWS, particularly aggression, stubbornness, depression, lying and child and parental stress <sup>38</sup>.

Notably, in the second meeting of the training program it was found that the mothers focused much of their principal concerns on a possible cure for hyperphagia, which shows a lack of knowledge about the

disease and its evolution and, as shown in previous studies, a lack of knowledge on issues related to the management of behavioral problems, including eating at night, skin picking and psychiatric problems such as obsessive-compulsive symptoms and hallucinations, which make this behavioral management even more difficult <sup>38</sup>

**Table 3.** Mothers' knowledge of general aspects of the syndrome at the pre-intervention phase.

Mother	Knowledge about Syndrome	Cause of Syndrome	Characteristics of Syndrome	Sources of information about the Syndrome	Sources of Information for Management	Usual Management of Child
Mother 1	Children are fat	Chromosome problem	Obesity, delay in school, small feet and hands.	Through a medical geneticist	None	Treat the same as a normal child
Mother 2	Weight increase disease	Does not know	Fatness	TV	None	Treat the same as a normal person
Mother 3	Genetic illness which causes obesity and behavioral problems	Failure of chromosome 15 in the formation of the fetus	Obesity, behavioral disorders, compulsive eating	When the child underwent surgery	Child's Psychologist	Mother knows she needs to control food.
Mother 4	Children are fat	Children are fat	Children are fat	Through a medical geneticist	Hospital Clinic	Treat the same as a normal person
Mother 5	Disease of obesity	Chromosome error	Small hands and feet reduced height.	A medical geneticist	USP Hospital Clinic	Treat naturally

During the training program, the mothers reported their major concerns about their child to be the following: irreversibility of obesity; possibilities for independence and autonomy in adulthood; difficulties in controlling satiety and eating behavior; possible cures for the disease, especially of hyperphagia; concerns related to the production of medicines to control hyperphagia and comorbidities associated with the syndrome. During the sessions, the mothers highlighted the difficulties they faced in their daily routines, and felt that taking part in the program had helped them to ease some of their difficulties, for example: a) the daily routine before meals; insecurity, distress, sadness, anger, worry and anxiety about their child's lack of satiety; b) daily routine after meals; distress, worry and fear, especially in relation to the food intake of their child and obesity; c) concerns about satiation of their children, and the sense of calm they felt to see their child satisfied, even temporarily. Mothers also reported feelings of sadness and frequent crying; c) family difficulties; overload due to care of the affected

child, feeling isolated in respect of coping with the child's illness, marital problems, the child's exposure to excessive amounts of food from other family members and relatives. Families with members with PWS should establish appropriate strategies to control eating behavior stimuli, as well as a family strategy that promotes the mental health of all family members by trying to avoid disagreements among its members, and supports the appropriate management of the person affected by the syndrome <sup>10</sup>.

The main concern of the mothers was in relation to a possible cure for hyperphagia in an attempt to mitigate the main symptom of the disease. However, there were also other concerns that focussed on the problems that are typical of the syndrome phenotype and that weigh significantly on the caregiver, such as eating during the evening, skin picking and obsessive-compulsive spectrum behavior problems<sup>1, 39</sup>.

Table 4 shows the distribution of the types of parenting practices according to the caregivers.

**Table 4.** Distribution of mothers according to the use of parenting practices in the pre- and post-parental training phases.

Classification of Parenting Styles	Classification in Pre-intervention stage				Classification in Post-intervention stage			
	Excellent	Regular above average	Regular below average	Risk	Excellent	Regular Above Average	Regular below average	Risk
Positive Monitoring	1	0	3	1	3	2	0	0
Moral Behavior	0	2	2	1	1	3	1	0
Inconsistent Punishment	0	1	0	4	0	3	2	0
Negligence	1	0	3	1	2	1	2	0
Relaxed Discipline	0	0	3	2	2	2	0	1
Negative Monitoring	0	2	1	2	1	2	2	0
Physical Abuse	1	0	2	2	3	0	1	1
PSI Total	0	1	2	2	3	1	1	0

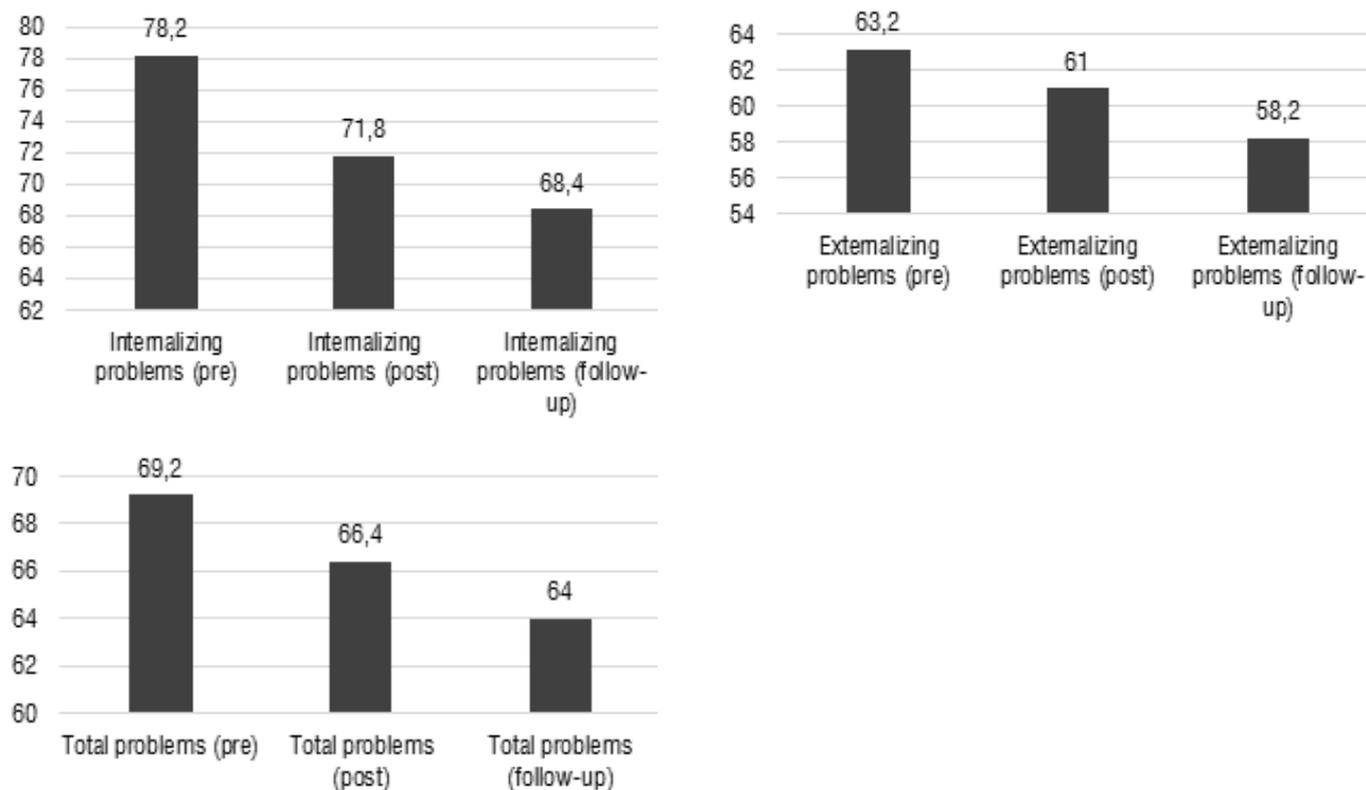
**Legenda:** PSI: Parenting Styles Inventory

Parenting practices that shaped the PSI scores showed an increase in positive parenting strategies (principally in positive monitoring) and a decrease in negative ones (especially neglect, relaxed discipline and negative monitoring), with the latter changed from a level of risk to a regular level. It should be noted that only mother number 4 continued practices associated with relaxed discipline and physical abuse in the risk classification in the post-intervention phase. Parenting styles unsuitable for management of the children's behavioral problems are generally associated with high levels of stress in the caregivers. Physical punishment is one of the practices that affects the psycho-emotional development of the children and adolescents and, generally, is practiced by parents in an attempt to reduce undesirable behaviors. However, the behavioral problems are not mitigated. The relationship between this type of parenting practice and aggressive and challenging behavior in children with the syndrome has been consistently confirmed in the literature<sup>40</sup>. Caregivers need counseling as much as their children. This can help to reduce levels of parental stress and mitigate the strain on carers. Supervision is required for the majority of the time due to the child's hyperphagia, and the other behavioral problems associated with the syndrome such as stubbornness, defiance, lying, limitations related to intellectual disabilities and aggressive behavior<sup>40</sup>.

Figure 1 shows the weighted scores (T) of the mothers' scales for internalizing, externalizing and total behavioural problems of the ASR/18-59 in the pre, post and follow up phases. A decrease in the number of problems evaluated by the three scales was observed throughout the study.

The data in Figure 1 show evidence of a reduction in emotional and behavioral problems during the different phases of the training program. It is likely that this reduction is associated with the development of a better understanding of the child's illness, and the fact that being part of the group has contributed to indicators of social support. Even though the number of participants in our study is small, the results are in line with the data of Skokauskas et al<sup>40</sup>, who reported that the parents of children with PWS can develop anxiety symptoms but guidance on how to manage their problems can minimize many affective symptoms as the caregiver learns to identify and solve the problems, thereby reducing somatization and emotional difficulties<sup>40</sup>.

The improvement in the indicators evaluated by the PSI (Table 4) shows that there was a recognition by the mothers of the different factors that harmed the physical health and behavioural patterns of their children. Improvements were also found in the emotional aspects of mothers when they properly adopted the guidelines offered in the parental training (including an improvement in nutritional indicators for children, as shown in the nutritional data made by the same group and published by Lima and colleagues<sup>32</sup>.) The results suggest, from the records of mothers, that they began to monitor more systematically other members of the family and those who lived with the child who offered food to them. There was also a recognition by the mothers of inappropriate parenting practices. Wulffaert and colleagues in a study on maternal stress in families with children who have PWS reported that professional support is essential for families because of the high stress levels found in the mothers of those with the syndrome<sup>20</sup>.



**Figure 1.** mothers' weighted scores for (T) internalizing, externalizing and total behavioral problems in the ASR/18-59 inventory at the pre-intervention, post-intervention and follow-up phases.

## CONCLUSION

The findings indicate a recognition by the mothers of several factors that harmed the physical and mental health of both their children and themselves. They started to check and supervise in a more direct and systematic manner relatives who lived with their child, to prevent them offering food to them, and came to recognize more clearly the negative influence of conflict within couples on the externalizing and internalizing behavioural problems of the children affected by the syndrome. Reductions in emotional and behavioral problems, assessed by the ASR/ 18-59, show that the program eased several indicators of these problems in mothers and that there were significant improvements in parenting practices in managing the behavior of the children.

Given the lack of studies published in Brazil on parenting intervention models and PWS, we consider this to be a pioneering study in the country. The program that was used contributed to an increase in knowledge about the syndrome, an improvement in parental management strategies in relation to their children and improvements in the mothers' mental health. The results of this study in relation to mental

health care could be used to replicate the program and provide services to this population through, for example, the health systems' Centers for Psychosocial Care; these are services that typically provide care for people with PWS due to disability intellectual. It emphasizes the importance of multi and interdisciplinary interventions for people with PWS and their families. It is also important to note in regard to intervention that the earlier this is, the greater the benefits, both for the individual with the syndrome and for families and caregivers, will be.

In respect of suggestions for such interventions in Psychosocial Care Centers or other mental health services, some limitations of this study could be targets for future work. A comparison of the impact of behavioral intervention programs and diet between different age groups, and between groups of different genetic subtypes is suggested. It is also recommended that other members of the family should be included in interventions, and not just the primary caregiver. Studies assessing interventions in school settings should also be conducted, as well as interventions with physical trainers and nutritionists, aiming at a more pronounced weight loss.

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