Ann Ist Super Sanità 2016 | Vol. 52, No. 3: 428-433
DOI: 10.4415/ANN_16_03_15

Parent training education program: a pilot study, involving families of children with Prader-Willi syndrome

Yllka Kodra[1], Loreta A. Kondili[2], Alessia Ferraroni[1], Maria Antonietta Serra[1], Flavia Caretto[3], Maria Antonietta Ricci[4] and Domenica Taruscio[1]

[1] *Centro Nazionale Malattie Rare, Istituto Superiore di Sanità, Rome, Italy*
[2] *Centro per la Ricerca e la Valutazione dei Prodotti Immunologici, Istituto Superiore di Sanità, Rome, Italy*
[4] *Università degli Studi di Roma "Tor Vergata", Rome, Italy*
[5] *Federazione Nazionale Sindrome di Prader-Willi, Rome, Italy*

Abstract

Introduction. Prader-Willi syndrome (PWS) is a rare genetic disorder characterized by severe hypotonia during the neonatal period and the first two years of life, the onset of hyperphagia with a risk of obesity during infancy and adulthood, learning difficulties and behavioral or severe psychiatric problems. This complex disease has severe consequences and difficult management issues also for patients' families. Parents of children with PWS need appropriate psychoeducational intervention in order to better manage their children with PWS. The purpose of this study was the implementation and evaluation of a PWS psychoeducational parent training program.

Methods. The Italian National Center for Rare Diseases implemented a pilot parent training program offered to parents of children with PWS. The intervention's effects was evaluated using questionnaires comprised of 11 items rated on a 7 point Likert scale.

Results. The intervention was offered to 43 parents. The behavior problems management, dietary restrictions, autonomy and relationships were indicated by parents as the priority topics which needed to be addressed. Evaluations, immediately post-intervention and after 6 months, were reported by parents, fulfilling specific questionnaires. 90% of parents involved in the study, appreciated the methodology, 86% felt more informed about PWS, 47-62% felt more capable to better approach behaviour's problems, 20-25% felt better about the child's health situation and future expectations. Feeling more capable to help the child autonomy and relationships were reported in 62% and 63% of parents respectively, which decreased significantly ($p < 0.05$) according to the evaluation 6 months after the intervention. Younger age of parents (< 44 years of age) was significantly correlated with better understanding on how to help the child's autonomy (OR: 0.05; CI: 0.04-0.8) and to better collaborate with the child's teachers (OR: 0.02; CI: 0.001-0.9).

Conclusion. Parent training is a promising intervention for parents of children with behavior's problems. Interventions with a behaviorally oriented program, addressed to parents of PWS affected children, is a useful tool in increasing their ability to manage the problems related to the disease.

Key words
- Prader-Willi syndrome
- rare diseases
- parent training
- parents
- evaluation program

INTRODUCTION

Background

Prader-Willi syndrome (PWS) is a rare multisystem disorder, but it is the most common genetic cause of obesity [1, 2]. This syndrome has specific phenotypes which are characterised by severe neonatal hypotonia; early onset of hyperphagia, development of morbid obesity, short stature, behavioral and severe psychiatric problems and learning disabilities. These patients constitute big medical challenge also for their specific behaviour which is generally characterized by an excessive appetite, temper tantrums, rigidity, oppositional and/or possessive behaviour that bring as consequences several psychiatric problems [3, 4]. In order to meet the medical, developmental, behavioural, and social needs of these patients, a multidisciplinary approach of geneticists, nutritionists, internists, endocrinologists, physical therapists, and psychologists is required [5].

Address for correspondence: Yllka Kodra, Centro Nazionale Malattie Rare, Istituto Superiore di Sanità, Via Giano della Bella 34, 00161 Rome, Italy. E-mail: yllka.kodra@iss.it.

PWS has severe consequences and difficult management issues also for patients' families. Parents of children with PWS need appropriate information; first, in how to manage and to support their children and second, in how to adopt an intermediary role with doctors who take care of their children. Based on this requirement, the late 1960s saw a shift in addressing children's problematic behaviours, which focused exclusively on changing the child's undesirable behaviours, to interventions attempted changing parents' behaviour and their empowerment [6]. In particular, in PWS the parent training aims to motivate changes in parents' attitude and subsequently behavior, through the increase of parent's knowledge about the disease and the improvement of parents' communication. The effect of a parent training program is to proximally modify the parent's outcome with the final goal to distally modify the outcome of the affected child. This effect is either related to the disease's features, or to the parents sociodemografic and cultural backgrounds. A pilot parent training psychoeducation program was implemented by the Italian National Center for Rare Diseases in a group of Italian parents with PWS affected children. The main aim of the present study was to describe and evaluate the results of this training program.

METHODS

Sample

Families of children with PWS were selected by a simple random sampling, extracted from a list of 200 members of Italian National Federation of Prader-Willi syndrome, including couples and/or single parent, accordingly with the selection procedure. Acceptance rate was 93% with no differences in the sociodemografic characteristics (age, gender, level of instruction and occupational status) among those who accepted and did not accepted to participate in this training programme. The informed consent to participate in the study was obtained from participants. The work has been carried out in accordance with the Code of Ethics of the Declaration of Helsinki.

Study design and description of the parent training program

The Italian National Center for Rare Diseases implemented a parent training psychoeducation program in which the training methodology was the "active learning" through an interactive discussion integrated with the traditional lecture. A specialised expert psychologist, with good capacities in mediating dynamic and communicative approaches and a parent with previous experiences on parent training method, were also involved. Both professional facilitators participated actively in meetings with parents, in order to guarantee a good collaboration among the participants. In particular, the information, given by the psychologist, was integrated with an active parental involvement in which parents actively demonstrated their specific parenting skill (with active group discussions during each session). The aim of this approach, was to translate the knowledges, explained during the lecture, into a appropriate parenting skill.

In order to tailor the program's content to the parents' individual requirements, the parents involved were asked to fulfill a specific preliminary questionnaire in order to define the information helpful for them. On the basis of parents' needs the training program content was elaborated. It consisted in twelve sessions of a duration of 2.30 hours each. The main objective of each session is reported in *Table 1*.

Program evaluation

In order to evaluate the effects of the program in better PWS management, the parents' feelings were evaluated. The effect of this program in the improvement of the parent's knowledges and attitudes, in managing the child with PWS, was evaluated through specific questionnaires which were fulfilled by the parents in two periods of time:

- post-intervention, directly following the training program and;
- 6 months after the end of the training program in order to evaluate if the effects obtained by the lectures were maintained on time.

The questionnaire comprised 11 items rated on a 7 points of Likert scale as follows: 1 extremly negative; 2 very negative; 3 somewhat negative; 4 neither positive or negative; 5 somewhat positive; 6 very positive; 7 extremely positive.

Moreover, participants were also asked to report, as a free text (as qualitative measure) the most positive aspects of the program and the issues which needed to be improved.

Statistical analysis

For each of the item included in the evaluation questionnaire, the frequency of positive opinions (points 6 and 7), post-intervention and 6 months after, were calculated.

Comparison of the points distribution, used for each item by each parent, post-intervention and 6 months after, were analyzed using the nonparametric Wilcoxon-test. $P < 0.05$ was considered as significant.

The role of sociodemografic factors (age, gender, level of instruction and occupational status) on the parents' evaluation, reported as low (scores = < 5) or high (scores > 5) for each item, was determined by the univariate analysis. A sociodemografic factor that was associated with the better outcome in the univariate analysis was adjusted for the possible effect of the others by the logistic regression analysis. The 17.1 Version of SPSS software was used.

RESULTS

Parents enrolled in the study

The intervention was offered to 43 parents with children affected by PWS (mean age 44 years: range: 26-64), coming from different Italian regions. The parents' characteristics are shown in *Table* 2. The number of mothers and fathers that followed the course was similar and the overall instruction level was medium to high (5-18 years of school). The mean age of PWS affected children was 13 years (range: 1 months-37 years).

Table 1
Description of the parent training program content

Content	Description
1. Session **Providing knowledge about development of a PWS affected individual: from child to adult age**	Describe clinical features, according to genetic subtypes of PWS, in various life stage. Specifically, focus on feed difficulties, hyperphagia in chilhood; excessive weight gain, obesity and short stature in adolescence; hypogonadism and learning and or attention difficulties in adulthood.
2. Session **Behaviour management of hyperfagia, obesity and its complications**	Address appropriate issues related to the importance of normo-hypocaloric diet and child involvement in physical activities in PWS in order to avoid or decrease obesity and possible respiratory, musculo-schelectrics or other complications.
3. Session **Promote children's cognitive or academic skill, enhancing their school readiness**	Insights on the importance of a specific education programme for children with PWS. Considering its broad clinical variability, specific indications on the collaboration among parents and teachers, in order to elaborate *ad hoc* educational programmes for each child affected by PWS.
4. Session **Promote children's social skill or prosocial behaviour**	Give indications in how parents should teach the affected child to share and cooperate, to use good manners and to get along with peers, siblings and adults. Explain the importance that these different social relationships may have on the behaviour and social development of the PWS affected child.
5. Session **Promote the autonomy and social partecipation of the affected individual to the everyday life activities**	Focus on how the parents could support their child and how the educator could be supported by the parents in the management of the PWS affected child. Indicate the importance of the collaboration with a psicologist educator in order to implement specific interventions to manage autonomies' problems of individuals with PWS.
6. Session **Indicate effective strategies to improve the communication with the PWS affected children**	Address issues related to the limited cognitive ability, in particular to the vocabulary level, capacity of logical argumentation, capacity of understanding and elaboration of different informations. Indicate positive interactions with a PWS affected child *e.g.*, how to use skills that promote parent-child interactions and provide positive attention etc.
7. Session **Behaviour problems in children with PWS**	Provide knowledges about PWS child's behaviour problems and identification of appropiate strategies for their prevention or management. Emphasize the role of psychologists and other professionalsin supporting parents to identify environmental factors that are associated with an increased risk of occurrence of temper outbursts, repetitive and ritualistic behaviours.
8. Session **Rehabilitation**	Address the role of an appropriate rehabilitation program, guided by an accurate functional ability evaluation of the PWS affected child. Give knowledges on how such strategies can be implemented by family members and homes individuals with experience in the PWS management.
9. Session **Psychiatric problems**	Provide knowledges about psychiatrics problems in order to help parents to distinguish the behaviour challenges from psychiatric illness. Focus on the management of affective disorders with particular interest in affective psychosis. Address the possibility of deterioration of an existing behaviour pattern or the onset of new behaviours challengesthat should be considered for psychiatric evaluation.
10. Session **Provide scientific updated information about PWS**	Discuss scientific research in the field of PWS, specifically management of behaviour and affective disorders, possible changes in the environment factors related to the syndrome, therapies known to be effective in the treatment of mood disorders and or psychotic illness. Address new therapeutic research and possibilities on the treatment of patients with PWS.
11. Session **Comunication of diagnosis to sibilings**	Focus on how to use appropiate manners to communicate the PWS diagnosis of the affected child to slibings and give indications on when and how to explain to siblings the particular attention and care for the affected child. Address how this approach should consider the character and the age of the affected child and of the respective siblings.
12. Session **Sexual development of an individual affected by PWS**	Address the importance of the sexual counselling since the childhood, providing specific indications on how to start sexual education in the early stages of the life. Focus on the particularities of sexual development in order to support, in a individualised manner, the variability of this handy-cap in a single person that need, either affective, or sexual relationship.

Results of preliminary questionnaire

The management of the behaviour problems, weight, dietary restrictions, autonomy and social integration were indicated as the top five priority topics by the involved parents. In 42 (98%) parents, the reasons for the program willing were to receive more information about the syndrome, to have practical advices about the disease's management and to share experiences with other parents and professionals. Twenty five (58%) parents had not participated in the previous vocational training programs.

Post-intervention evaluation

The effect of the program, according to the data reported in the fulfilled questionnaires, immediately post-intervention and 6 months after, are reported in *Table 3*. The Global Opinion about the training program (items 1-3), was generally very good and positive scores were reported. These scores were similar, also in the evaluation performed 6 months after the end of the training program. Specifically, all participants wanted to participate in similar educational training programs in the future; 95% of them would recommended the course to

Table 2
Summary of parents' characteristics (n. = 43)

Category	Number	%	Mean age (SD)
Type of participants			
Couple parents	36	84	43 (9)
Single parent	7	16	45 (13)
Instruction			
>12 years	11	26	47 (10)
8-12 years	22	51	43 (9)
5-8 years	10	23	42 (13)

other parents with the same problems, 90% appreciated the methodology and the approach used and 86% felt more informed about PWS.

Regarding the program's effect in the improvement of the parent's attitudes related to PWS, less positive opinions were reported. Specifically, according to the feeling of how to cope with the PWS behaviour's management (*Table 3* items 5-9), only part of parents (47%-62%) felt more capable to help their child accordingly. Only 40 (25%) of parents felt better regarding their child's situation (*Table 3* item 10) and only 41 (20%) felt to have more expectation for the future of the PWS affected child (*Table 3* item 11).

At the evaluation, obtained 6 months after the intervention, significant lower points were used in feeling more capable to help the child to be more autonomous and in feeling more capable to collaborate with the school (*Table 3* items 7 and 9, respectively).

Younger age parents (< 44 years) felt significantly more able to help the child to be more autonomous (Adjusted OR: 0.05: 0.04-0.8) and to collaborate with the child's teachers (Adjusted OR: 0.02: 0.001-0.9) compared to older age parents. Gender, education levels and occupational status did not show any effect on the overall evaluations (data not shown).

The open questions confirmed that the most positive aspects of the program, were the delivery methods, the acquisition of new concepts, information about the syndrome and the possibility to share experiences with other parents with similar problems. Introduction of more practical advice and skill acquisition in order to better face problems related to the syndrome were reported as topics that needed to be improved in the future educational programs.

CONCLUSIONS

Parents of children with rare diseases experienced stress, especially related to lack of competence, social isolation and emotional demands [7]. As a consequences, the parents' empowerment process is a necessity for families of patients with rare diseases, because they are chronic and require a complex management and a multidisciplinary approach [8]. It has been reported that sharing information with parents about their children's cognitive and emotional development could improve their understanding about the factors that influence the child behaviour, could increase their sensitivity about

Table 3
Evaluation of immediately and six months post-intervention effects of the training course by the parents' point of view

	Post-intervention positive opinions N. (%)	Post-intervention evaluation mean score (SD)*	Six months after evaluation mean score (SD)	P
Global opinion about the programme				
Item 1 agree with the methodology	42 (90.5)	6.2 (0.6)	6.1 (0.7)	0.28
Item 2 desire to participate in the similar courses in the future	42 (100)	6.8 (0.4)	6.5 (0.6)	0.09
Item 3 recommend the course to other parents	42 (95)	6.6 (1.3)	6.6 (0.6)	0.86
Parenting knowledge				
Item 4 feeling more informed about PWS	42 (86)	6.4 (0.7)	6.1 (0.7)	0.12
Parenting attitude regarding the PWS affected child				
Item 5 feeling more capable to manage the child problematic behaviours	40 (57.9)	5.5 (1.0)	5.2 (1.1)	0.09
Item 6 feeling more capable to prevent the child problematic behaviours	42 (47.3)	5.4 (0.9)	4.2 (0.6)	0.13
Item 7 feeling more capable to help the child to be more autonomous	42 (61.9)	5.5 (0.8)	4.6 (1.5)	0.02
Item 8 feeling more capable to help the child to improve the relationships	42 (61.9)	5.4 (0.9)	5.4 (1.0)	0.74
Item 9 feeling more capable to collaborate with the child's teachers	37 (62.5)	5.7 (0.8)	3.7 (3.4)	0.02
Item 10 accept betterthe child's situation	11 (25.6)	5.0 (1.0)	4.6 (1.5)	0.79
Item 11 having more expectation for the future of the child	9 (20.9)	4.7 (1.3)	4.6 (1.2)	0.80

*SD = Standard Deviation. PWS: Prader-Willi syndrome.

the children's needs, and could help them in developing realistic expectations about the child behaviour [9].

The needs of parents, related to the management of a diversely able child, have been previously classified in the need for information, partnership and normality. Different parent training programs have been implemented and a number of meta-analysis that test the efficacy of parent training programs have been published [10-13].

In this study we evaluated the results of a pilot training program, conducted by the Italian National Center for Rare Diseases, for parents of PWS affected children. This educational training program was not empirically defined; the parents' needs were firstly addressed in order to subsequently organise a tailored content of program. A bio-psycho-social model was used in order to evaluate parents' needs and their empowerment in this complex disorder. This parent training program was an intervention that combined passive teaching with an active skill acquisition and active parent's involvement. As it was observed, by the parents program evaluation, the main effect of the course, was the increase of the knowledge about PWS, which was maintained at least 6 months after the intervention. The mean of the scale points for each item, related to the "Global opinion about the program", and "Parenting knowledge", at the end of the course and 6 months after, was very high. Of the parents involved in this program, 40% stated that wanted honest information about the overall potentially problematic features related to PWS. This was consistent with the finding that parents of children with chronic diseases often prefer to know all possible negative aspects related to the disease, in order to be better prepared for the future [14].

The effect of the training course in changing the parents' behaviours, which means changes in feelings about how to manage the affected child in various life aspects, was less compared to the parenting knowledge. This effect was further reduced in the following 6 months after the end of the course for the parent's feeling on the child autonomy and on the school relationships. This indicates that changes in the knowledge may not ensure changes in the attitudes and in the behaviour. This lower effect could be partly related to the complexity of the PWS in itself, which consequence is the difficulty of the parent to promptly modify the attitude related to the affected child. On the other side, the program content, regarding the parent attitudes, could have been sufficiently explanatory, but less effective because of the lack of the practice component with the affected child during the course. In fact, parents stated that the program's content should have had more practise components. We are aware of this limit which was due to the wide range of ages (from 1 months to 37 years) of the children affected by the PWS, whom parents were involved in this parent training program.

Regarding to the effect of socio-demographic factors in the parents' evaluation, the younger age was positively related to the feeling of being more able to help the child in being more autonomous and in improving the school's relationships. Although the need to know everything about their child's condition, could have a negative impact on the younger parent, because of the insufficient experience in the disease management and the fear of what this syndrome could cause, they preferred to have both positive and negative information, in order to be better prepared for the future.

This study is considered a first attempt in a family centred intervention aimed to empower the parents of the affected children with the complex disorder of the PWS. The results are promising, however, to better evaluate the effect of such programs in parents' skills and behaviours a "no treatment" control group is necessary.

In conclusion: parent training is a promising intervention for parents of children with behavior problems. The implementation of behaviourally oriented program with practices, based on real-life situations, is a need and a useful tool, required by parents in order to better manage their children affected by PWS.

Author' contributions

YK, DT formulate the research question, participate in the study design, carried out data analysis and drafted the initial manuscript. FC supervised and coordinate the data collection process. AF, MAS and MAR participate in the process of data collection. LAK participate in data analysis and data interpretation. All authors critically reviewed the manuscript and approved the final manuscript submitted.

Acknowledgements

The authors would like to thank the Italian National Federation of Prader-Willi syndrome and parents of children affected by Prader-Willi syndrome for their participation. We are also grateful for the help of the professionals: Graziano Grugni, Laura Bosio, Deborah Ollero, Alessandra Picco, Irune Achutegui, Teresa Grimaldi Capitello, Letizia Ragusa, Pia Massaglia, Antonino Crinò, Teresa De Toni, Carla Fermariello.

Conflict of interest statement

No conflict of interest.

Received on 22 January 2016.
Accepted on 9 May 2016.

REFERENCES

1. Cassidy SB, Schwartz S, Miller JL, Driscoll DJ. Prader-Willi syndrome. *Genet Med* 2012;14(1):10-26.
2. Bridges N. What is the value of growth hormone therapy in Prader-Willi syndrome? *Arch Dis Child* 2014;99(2):166-70.
3. Holm VA, Cassidy SB, Butler MG, Hanchett JM, Greenswag LR, Whitman BY, Greenberg F. Prader-Willi syndrome: consensus diagnostic criteria. *Pediatrics* 1993;(2):398-402.
4. Chen C, Visootsak J, Dills S, Graham JM Jr. Prader-Willi syndrome: an update and review for the primary pediatrician. *Clin Pediatr (Phila)* 2007;46(7):580-91.

5. Goldstone AP, Holland AJ, Hauffa BP, Hokken-Koelega AC, Tauber M and on behalf of speakers contributors at the Second Expert Meeting of the Comprehensive Care of Patients with PWS. Recommendations for the diagnosis and management of Prader-Willi syndrome. *J Clin Endocrinol Metab* 2008;93(11):4183-97.
6. Bandura A. *Principles of behavior modification*. New York: Holt, Rinehart & Winston; 1969.
7. Dellve L, Samuelsson L, Tallborn A, Fasth A, Hallberg LR. Stress and well-being among parents of children with rare diseases: a prospective intervention study. *J Adv Nurs* 2006;53 (4):392-402.
8. Aymé S, Kole A, Groft S. Empowerment of patients: lessons from the rare diseases community. *Lancet* 2008;14(371):2048-51.
9. Cedar Bruce, Levant RF, Ronald F. A meta-analysis of the effects of parent effectiveness training. *Am J Fam Ther* 1990;18(4):373-84.
10. Kaminski JW, Valle LA, Filene JH, Boyle CL. A meta-analytic review of components associated with parent training program effectiveness. *J Abnorm Child Psychol* 2008;36(4):567-89.
11. Dretzke J, Frew E, Davenport C, Barlow J, Stewart-Brown S, Sandercock J, Bayliss S, Raftery J, Hyde C, Taylor R. The effectiveness and cost-effectiveness of parent training/educationprograms for the treatment of conduct disorder, including oppositional defiant disorder, in children. *Health Technol Assess* 2005;9(50):iii, ix-x, 1-233.
12. Van den Hoofdakker BJ, van der Veen-Mulders L, Sytema S, Emmelkamp PM, Minderaa RB, Nauta MH. Effectiveness of behavioral parent training for children with ADHD in routine clinical practice: a randomized controlled study. *J Am Acad Child Adolesc Psychiatry* 2007;46(10):1263-71.
13. Lundahl B, Risser HJ, Lovejoy MC. A meta-analysis of parent training: moderators and follow-up effects. *Clin Psychol Rev* 2006;26(1):86-104.
14. Barlow JH, Cullen LA, Rowe IF. Comparison of knowledge and psychological well-being between patients with a short disease duration (< or = 1 year) and patients with more established rheumatoid arthritis (> or = 10 years duration). *Patient Educ Couns* 1999;38(3):195-203.