

Cognitive-behavioral therapy for hyperphagia in an adult with Prader-Willi Syndrome: pitfalls and challenges

Terapia Cognitivo-Conductual para hiperfagia en un adulto con Síndrome de Prader-Willi: obstáculos y retos

Angélica Carrillo-Nieto

National Autonomous University of Mexico (UNAM),
Mexico

Edgar Landa-Ramírez

Emergency Psychology Program/Emergency Department,
Hospital General Dr. Manuel Gea González, Mexico City,
Mexico

Aída Monserrat Reséndiz-Barragán

Department of Obesity and Bariatric Surgery,
General Hospital Dr. Manuel Gea González, Mexico City,
Mexico

Hyperphagia is a typical characteristic in Prader-Willi Syndrome and a clinical problem when not managed appropriately. There is scarce evidence of its treatment. The actual literature targets interventions in well-developed countries, thus leaving a gap for clinical professionals to treat PWS in developing countries where the cultural and socioeconomic conditions are extremely different. The aim of the present study was to assess the effect of cognitive behavioral therapy for hyperphagia within an interdisciplinary treatment conformed by behavioral, pharmacological, dietary and exercise interventions in a 31-year-old Mexican male with Prader-Willi Syndrome and morbid obesity in a developing country context. The main results were the weight of the patient that remained relatively constant, there was a downward trend relevant to his glucose levels, a reduction of his arterial pressure levels, and behavioral changes in regard to exercise that did not maintain overtime. There are considerable limitations inherent to developing country contexts that affect the validity of treatments applied in these settings.

Keywords: Prader-Willi Syndrome; Patient care team; Cognitive Therapy; Social conditions; Developing countries.

La hiperfagia, característica típica del Síndrome de Prader-Willi, es un problema clínico cuando no se maneja apropiadamente. La evidencia sobre su tratamiento es escasa y se focaliza en intervenciones en países desarrollados, dejando un vacío de literatura para los clínicos que tratan el SPW en países en vías de desarrollo con condiciones culturales y socioeconómicas extremadamente diferentes. El objetivo de este estudio fue medir el efecto de la terapia cognitivo conductual para hiperfagia dentro de un tratamiento interdisciplinario conformado por intervenciones conductual, farmacológica, nutricional, y ejercicio en un hombre Mexicano de 31 años con Síndrome de Prader-Willi y obesidad mórbida en un país en desarrollo. Los principales resultados fueron: el peso del paciente se mantuvo relativamente constante, su nivel de glucosa tendió a disminuir, su presión arterial se redujo, y los cambios conductuales relacionados al ejercicio no se mantuvieron. Hay limitaciones considerables, inherentes a los países en desarrollo, que afectan la validez de los tratamientos aplicados en estos contextos.

Palabras clave: Síndrome de Prader-Willi; Atención interdisciplinaria; Terapia Cognitivo Conductual; Condiciones sociales; Países en desarrollo.

INTRODUCTION

Prader-Willi Syndrome (PWS), a genetic neurodevelopmental disorder, affects one in every 29,000 births disregarding race or sex. PWS includes low metabolic rate, hypogonadism, intellectual disability, short-term memory deficits, short stature, small hands and feet, daytime hypersomnolence, and hypotonia during infancy. Behavioral characteristics include tantrums, rigidity, perseveration, argumentativeness, mood lability, skin picking, obsessive-compulsive features, and an increased risk of psychotic symptoms (Radstaake et al., 2011).

Hyperphagia is typical in PWS and closely associated with life-threatening obesity (Martínez, Haqq & Wismer, 2015). Literature about hyperphagia management, mainly obtained in well-developed countries (Heymisfield et al., 2014; Martínez et al., 2015), does not point to straight conclusions, but it recommends interdisciplinary approaches that normally include behavioral modification and cognitive behavioral therapy (CBT). This study examined the effect of CBT for hyperphagia in PWS within an interdisciplinary approach in a non-structured setting in a developing country. The secondary objective was to describe the challenges and pitfalls that were faced regarding this intervention.

METHOD

Case introduction

Charly (pseudonym), a 31-year-old Mexican male, was formally diagnosed with PWS when he was 22 years old by his treating physician, plus hypertension, psychomotor retardation, metabolic syndrome, hypogonadism, morbid obesity, and cryptorchidism. Charly and his mother referred themselves for treatment of hyperphagia, which was associated with his weight 97 kilograms and morbid obesity (Body Mass Index; 52=Extreme Obesity).

During the weekends, Charly lives in a non-spacious dwelling with his mother, stepfather, sister, and nephew. He works as a domestic employee for a high-income family. He has a low socioeconomic status and lacks an adequate support net, which has affected his therapeutic adherence to several medical follow-ups. His mother reported that she has always experienced difficulties in controlling Charly's behaviors and hyperphagia and in promoting healthy habits such as adherence to dietary recommendations, physical exercise, and medication taking.

Patient assessment

Sociodemographic data were collected through instruments designed for this purpose. Weekly semi-structured interviews were conducted in a consulting room with the patient, his mother, and sister to determine the history of the hyperphagia and the effects of previous interventions. Diagnoses and poor adherence were confirmed in medical reports. At the beginning of the treatment, the Hyperphagia Questionnaire (HQ) was applied. This is a 13-items hyperphagia-related problems questionnaire that assesses three factors: Hyperphagic Behavior (Cronbach's $\alpha=0.76$); Hyperphagic Drive (Cronbach's $\alpha=0.80$) and Hyperphagic Severity (Cronbach's $\alpha=0.60$; Dykens, Maxwell, Pantino, Kossler & Roof, 2007). Despite the acceptable internal consistency, the HQ requires high commitment from parents or care providers as informants. Unfortunately, we could not get that level of commitment from the patient's mother and sister that is the main reason why assessment with the HQ stopped.

Given these complications, it was decided to evaluate physical variables related to hyperphagia. It was reasoned that if our intervention decreased hyperphagia, it would be reflected in a modification of our patient: weight, glucose level, and blood pressure, as Succurro et al. (2015) described with the metabolic profile of binge eating obese.

Charly had weekly assessments of these variables at his community health center by an external team. These data were collected every session. His weight was measured using a calibrated mechanical physician scale, glucose levels were provided by a simple fingerstick test, and blood pressure, by a sphygmomanometer. Glucose and blood pressure levels were compared with the Mexican Health Department (SSA, 2010, 2017) values that derived from international standards.

The exercise was selected in concordance with other interventions (Messersmith, Slifer, Pulbrook-Vetter & Bellipanni, 2008). A trained-medical team in a controlled setting at an Obesity Clinic at a General Hospital in Mexico City conducted cardiovascular, strength, and elasticity tests.

Informal caregiver assessment

We applied to the mother four self-report instruments adapted and validated for the Mexican population: The Zarit Burden Interview, to evaluate the informal caregiver burden, impact of care, interpersonal relationship, and expectations for self-effectiveness (ZBI; 22 items; $\alpha=.90$; Galindo-Vazquez et al., 2015). The Beck Depression Inventory, to assess the presence and severity of symptoms of depression (BDI; 21 items, $\alpha=.87$; Jurado et al., 1998). The Beck Anxiety Inventory, to assess the presence and severity of symptoms of anxiety (BAI; 21 items, $\alpha=.84$; Robles, Varela, Jurado & Paez, 2001). Finally, the Brief COPE Inventory, to assess her coping strategies (26 items, $\alpha=0.71$; Morán, Landero & González, 2010).

Case conceptualization

Functional Analysis (FA) with stimulus, organism variables, response, and consequences (SORC) model established that the problem behaviors (hyperphagia and skin picking) were favored by biological (satiety dysfunction), cognitive (intrusive thoughts and misinterpretations, and poor executive functioning), environmental (aggressive critics, mockeries, scoldings, punishments, and restrictions) and operant variables. Even though biological and cognitive characteristics of PWS are frequently considered to explain hyperphagia and skin picking (Messersmith et al., 2008; Radstaake et al., 2011), the FA pointed that these behaviors had a self-stimulation function. They also used to follow exposure to stressors (including punishments and scoldings when his mother caught him), which was consistent with the arousal-reduction hypothesis (Radstaake et al., 2011).

About the mother, her coping style was composed of active coping, planning, emotional and instrumental support, religion, and acceptance. She reported no significant levels of anxious, depressive, or burden symptoms. Nonetheless, these punctuations were incongruent with her verbal report, nonverbal language, and lack of implication with treatments.

Treatment

Given the lack of knowledge about the mechanisms underlying hyperphagia and about the efficacy of drugs for controlling this problem in PWS nowadays, there is not a gold standard for treating hyperphagia in this population. Despite the lack of evidence for treating hyperphagia in PWS, there is a consensus for implementing multidisciplinary care (pharmacological treatment, behavioral approach, and nutritional treatment) focused on parental education and weight control. Consequently, behavior management, dietary restriction, and exercises are fundamental in the management of obesity in PWS (Crinò, Fintini, Bocchini, & Grugni, 2018; Ho & Dimitropoulos, 2010).

The intervention pursued as the main objective to reduce hyperphagia. The cognitive-behavioral treatment components involved: control of stimuli, assertivity, differential reinforcement, shaping, token economy, behavioral contract, diaphragmatic breathing, self-instructions, problem-solving, decisional balance, and social skills. See table 1 for the treatment outline. Intervention sessions were held usually once a week when Charly and his mother met directly with the therapist. The intervention was planned to be implemented across all of Charly's environments (i.e., workplace and home) with the collaboration of his mother. The treatment took place throughout 13 sessions, ranging from 90 to 120 minutes ($M = 85.38$, $SD = 11.26$) during five months. All techniques were presented to Charly and his mother, excepting diaphragmatic breathing and self-instructions, which were strategies managed individually with Charly.

Meanwhile, Charly started psychotherapy; he was introduced to an interdisciplinary team to receive integral management in an Obesity Clinic at a General Hospital in Mexico City. The team was composed by an endocrinologist, psychiatrist, exercise physician, psychologist, and nutritionist. His nutrition plan consisted of a diet of 1,500 calories distributed in 27% proteins, 25% lipids, and 48% carbohydrates in 5 meals a day. The weight goal was 42.5 kilograms. The physical activity plan was to walk 10 minutes with a 5-minute warm-up and a 5-minute cool-down five days a week. Each week he had to walk 1 minute more, exercising at 85 percent of his maximum heart rate. Also, a strength routine was indicated two days per week: 18 push-ups, 15 squats, and 15 abdominal crunches increasing two repetitions each week. Six stretching exercises were included.

Design and data analysis

This study follows the recommendation from Virués y Moreno (2008). We used a clinical case report with two phases: phase "A" consisted of the assessment before treatment sessions, 10 interview sessions were needed to complete the FA and case conceptualization gave that Charly and his mother did not respond well to the interview,

Table 1.
Session History

Number	Session topic	Mother components	Patient components
1	Introduction, Information.	Psychoeducation (PWS, CBT), treatment plan.	Psychoeducation (PWS, CBT), treatment plan.
2	Introduction, Information.	Behavioral contract, agreements of stimuli control, and token economy program.	Behavioral contract, token economy program.
3			Behavior records. Positive reinforcement of adaptative changes and little weight reductions (300/200 grams per week).
4	Behavioral modification (mother). Treatments adherence (patient).	Training in positive reinforcement and differential reinforcement techniques.	
5			Problem-solving.
6	Behavioral modification (mother). Diaphragmatic breathing (patient).	Adjustments on the token economy program.	Training in diaphragmatic breathing.
7	Behavioral modification (mother). Social skills and treatment adherence (patient).	Follow-up of the techniques already trained.	Self-instructions. Training in assertive communication.
8	Treatment adherence (mother).	Problem-solving. Psychoeducation of the cognitive model. Cognitive restructuring. Recommendation to start individual psychotherapy.	
9	Social skills.	Training in: active listening, negotiation and appropriate expression of anger and displeasure.	
10	Social skills and treatments adherence.		Negotiation. Decisional balance. How to face critics, say no, and set limits.
11	Risky stimuli avoidance and reduction of the hyperphagic drive.		Control of stimuli plan. Self-instructions. Diaphragmatic breathing. Problem-solving. Decisional balance. Mindfulness.
12	Termination	Review. Therapist reminded suggestion to start individual psychotherapy.	Review. Relapse prevention.

Note: Positive reinforcement of adaptive behaviors and practice of presented techniques were constant during all sessions. The training of abilities included resources like psychoeducation, modeling, role-playing, and shaping.

Table 2.
Physiological measures.

	Pre Intervention			Intervention			Post Intervention	1 month follow-up	3 month follow-up	6 month follow-up														
Weight*	97.8	99.0	98.9	98.8	100	99.7	99.0	99.0	99.0	98.3	96.0	99.3	97.6	97.9	98.3	97.8	97.5	98	98.8	98.8	99.3	99.0	102.3	
BMI**	52	53	53	53	54	53	53	53	53	53	51	53	52	52	53	52	52	53	53	53	53	53	53	55
Glucose Level	191	111	106	101	121	93	112	135	-	-	88	100	105	104	100	112	90	-	99	99	-	98	97	

Note: *=Kilograms. **=Body Mass Index. The missing data because of unavailability of resources of the patient and community health center is marked with a dash.

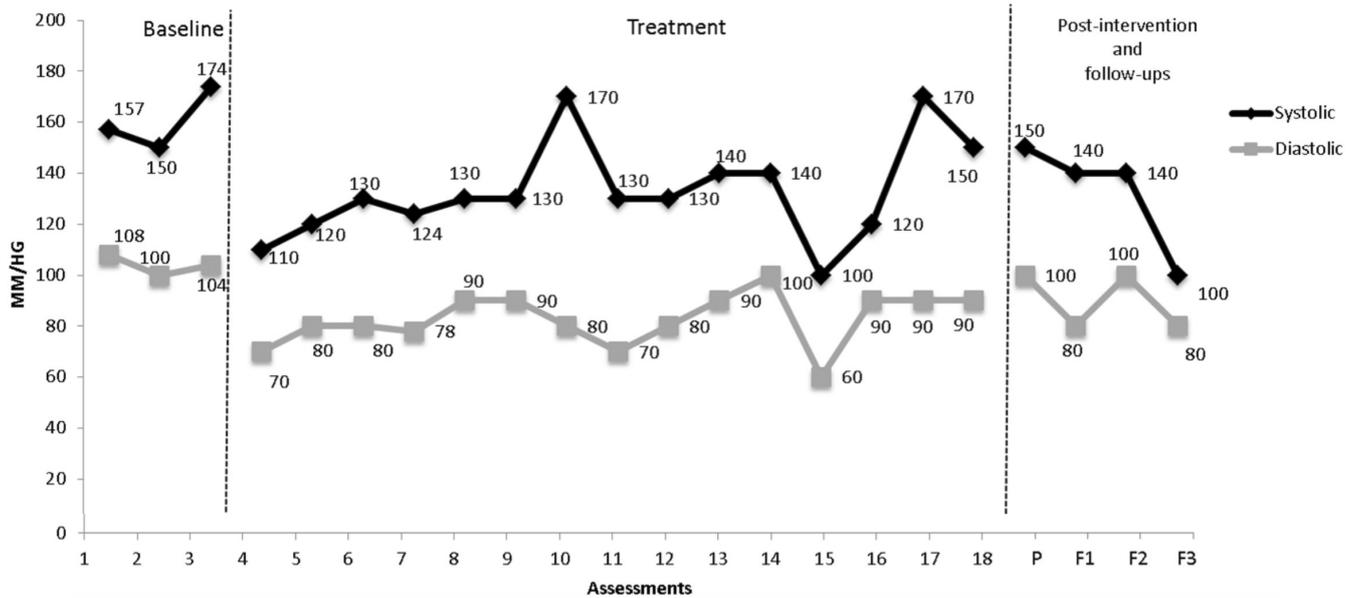


Figure 1. Arterial pressure along with treatment and follow-ups.

they gave tangential and confusing answers, his mother demanded immediate solutions to their actual problems (sometimes unrelated to Charly) and they have considerable difficulties in recalling relevant information in detail. Phase “B” consisted of the intervention described before.

Visual analysis of data between phases: A visual examination was conducted to judge the consistency and reliability of the intervention effects through a graphical analysis of data (Kazdin, 2011). Behavioral increases or decreases during the intervention and follow-ups were assessed and compared with baseline data.

RESULTS

As possible clinic results of the treatment, some changes in hyperphagia behaviors can be enlisted. These changes were identified from observation session through the session and from the verbal reports of Charly and his mother: at 8th session, Charly stopped eating at inadequate moments (e.g., when staying seated to take a rest). At the 11th session, he started to wash his teeth three times a day as a stimulus control strategy to prevent hyperphagia behaviors. At the 12th session, Charly stopped walking in front of food establishments (e.g., groceries, little restaurants) that he frequented.

According to Charly’s physiological measures, weight maintained relatively constant. Relevant to glucose, his mean baseline level was 127.2 mg/dl; 104.9 mg/dl, on average along with treatment; and finished at 97 mg/dl. (See Table 2). It is worthy of remark that there was a reduction of his arterial pressure levels, reaching normality at the six-month follow-up. Figure 1 displays the function of arterial pressure across time.

Concerning his exercise plan adherence, there were behavioral changes that did not maintain over time due to various difficulties. Indeed, this lack of increase in physical activity that was observed in Charly is reported in the literature; this point will be taken in the discussion section.

Primary caregiver

No technique was implemented by his mother, even when half of the sessions were dedicated to reviewing her performance during the week, to motivate and mobilize her to solve the difficulties she had and to create several strategies to increase her participation in the treatment. Individual psychotherapy was offered to her, but she rejected because of her lack of interest. The intervention continued with Charly individually since the 10th session.

DISCUSSION

This study was an outpatient intervention targeting hyperphagia in an individual with PWS with an ample set of pitfalls and challenges, such as: lack of scientific reports of PWS treatments in developing countries, the cultural and socioeconomic conditions of Mexico and Charly's family, PWS intellectual and biological characteristics, tangential and confusing speech of Charly and his mother, and low sensibility of the HQ. Even if our intervention was not efficient to reduce weight, it did favor its control, lead to a diminishment in glucose and arterial pressure, and an increase in physical activity.

Similar findings regarding weight and physical activity have been described in a systematic review with obese adults with intellectual disabilities (Spanos, Hankey & Melville, 2016). Concerning his exercise plan adherence, there were behavioral changes that did not maintain over time due to various difficulties. Indeed, this lack of increase in physical activity that was observed in Charly is reported in the literature. Spanos et al. (2016) founded that intellectually disabled adults in a weight loss maintenance intervention, did not show a statistically significant decrease in the percentage of time spent in sedentary behavior, neither a statistically significant effect increase in time spent in light physical activity nor in walking between baseline and 12 months of intervention.

Several pitfalls were faced. It was detected that literature commonly focuses on PWS children or adolescents, leaving adulthood as a forgotten life stage, and it is also produced in developing countries, so the external validity of the founded evidence was critically low. Other pitfall related to the context of our intervention was that in Mexico, there are no centers or clinics oriented to treat PWS and within poverty, lack of public policies, a needy public health system, and absence of trained professionals, it is less possible to find resources to develop multidisciplinary treatments (Crinò et al., 2018; Ho & Dimitropoulos, 2010).

Besides the genetic predispositions and the restrictiveness of living arrangements (Spanos et al., 2016), other obstacles were found such as a social desirability bias in our patient's mother, a lack of a validated coping inventory for Mexican caregivers (a general limitation for Mexican researchers), and the invalidity of the HQ. Based on this, it was important to draw upon physiological measures given that systematic observation was not possible.

Moreover, treatment adherence was affected by very demanding working conditions of our patient, violent behaviors, and wrong diet/exercise advice that he received from his mother, and his lack of social and economic resources. On another side, it is possible that after a 30-year nonstop caring, the physical and emotional health of the mother were severely deteriorated as it frequently occurs with mental disabled patients caregivers' which can explain her scarce collaboration (Ayuurebobi, Korley, Poku & Owusu-Agyei, 2015).

As Spanos et al. (2016) pointed, people with intellectual disabilities are more likely to experience health inequalities consequent to ineffective health systems. Based on this, sociocultural and economic features of patients and their caregivers must be included in the case conceptualization to increase the probabilities of success of clinical treatments (Glasgow, Davidson, Dobkin, Ockene & Spring, 2006).

This study was an exercise to encounter challenges as the invalidity of research already reported, methodological limitations regarding evaluation and the sociocultural characteristics of the family of our patient. The extent to which this intervention will be effective with other patients with similar characteristics is not known. But based on the evidence we have presented, it seems for us that to intervene (even when the environmental factors represent relevant obstacles) is better than nothing. We encourage other colleagues to consider the strategies we use as an alternative to improve the life quality of their patients and to develop future research that could show us other ways to favor the physical and mental health of persons living with PWS.

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