Nutritional diagnosis improvement in Prader Willi Syndrome without growth hormone

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Introduction

The prevalence of obesity in children with PWS is approximately 40% and in adolescents and adults it reaches 82-98%. Obesity, which can become morbid in many cases, is multifactorial and very difficult to manage. It is very rare to achieve sustainable goals of normal weight or even weight loss. Obesity is often believed to be an inherent feature of PWS, despite established treatments.

The purpose of this work was to describe the feasibility of achieving a better nutritional diagnosis, including normal weight, in individuals diagnosed with PWS, through specific nutritional interventions within the framework of a transdisciplinary treatment and without resorting to pharmacological treatments nor growth hormone.

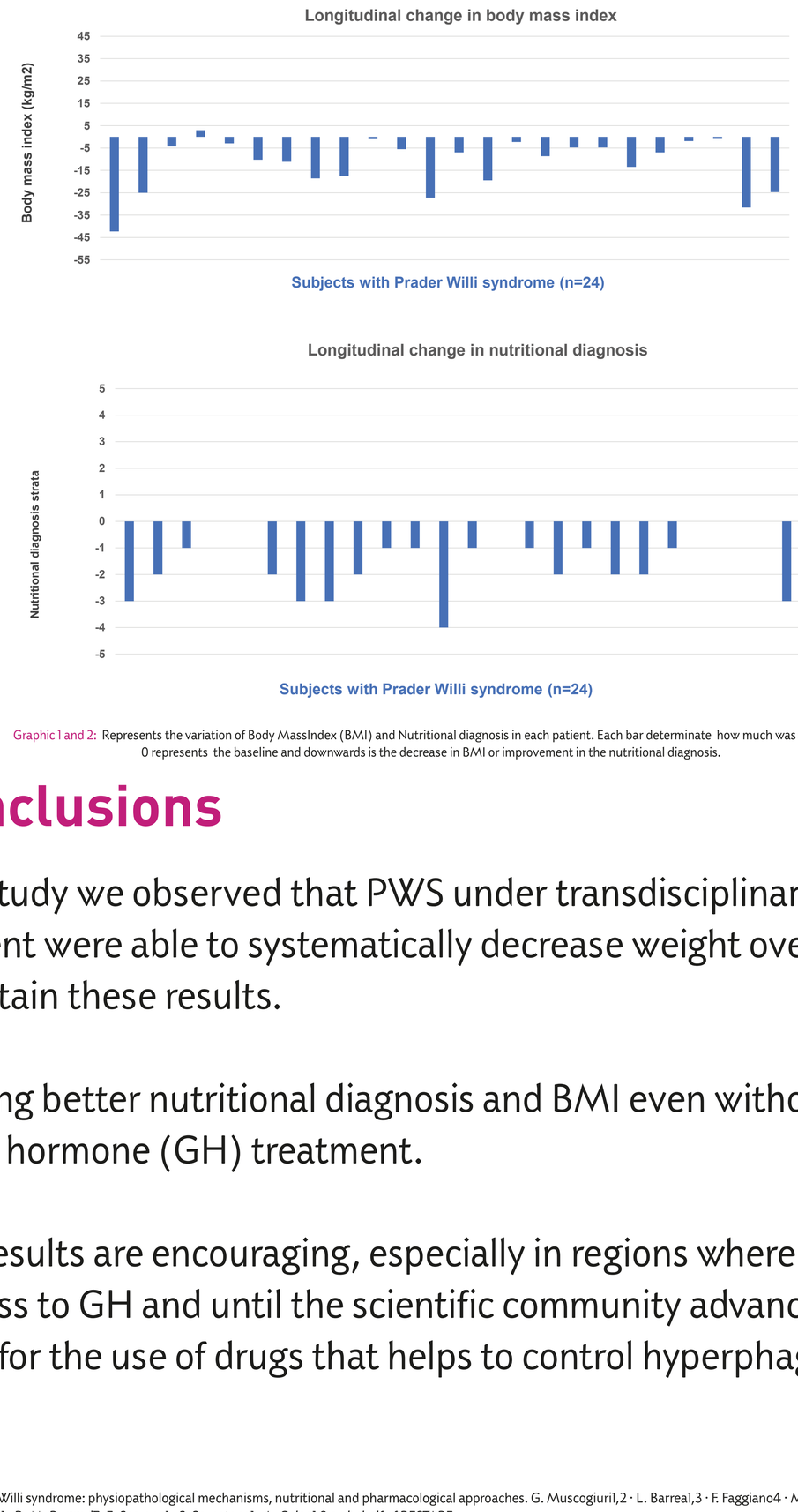
Methods

This retrospective observational study included a sample of PWS individuals of both sexes with confirmed genetic diagnosis, who receive transdisciplinary treatment in a center of rare diseases.

We compared the anthropometric measurements weight, height and body mass index (BMI), at the beginning of treatment, and their last weight measure.
The nutritional intervention includes a hypocaloric eating plan, calculating the energy requirement based on the age, sex and activities of the individual.
It is equally important to implement food safety measures including interventions on food structure, access, and handling. Additionally, raising awareness of eating behaviors of both individuals and their environment.
Nutritional intervention is based on the implementation of a controlled environment through the application of limits on access to food. It also includes setting up or planning a daily structure anticipating changes, and clear and sustainable limits over time.
Moreover, these patients count with scheduled on site treatment, which can be weekly or fortnightly according to their needs.

Results

The sample included 24 patients, 9 women (37.5%) and 15 men (62,5%) , and a mean baseline BMI of 40.2±15.7 kg/m2.
We identified a significant reduction in BMI (baseline 40.2±15.7 kg/m2 vs. follow-up 28.2±6.4 kg/m2, p<0.0001), with no significant differences regarding height (1.44±0.1 mts vs. 1.48±0.1 mts, p=0.09) with a median treatment duration of 52 months (interquartile range 23 – 116 months)
Upon treatment onseat, 20 (83%) patients had a diagnosis of obesity, 9 (38%) of them with grade III obesity; while currently only 6 (25%) patients persist with a diagnosis of obesity, presenting only 2 (8%) grade III obesity, 18 (75%) between normal weight and overweight.
This significant change in BMI and nutritional diagnosis can be seen in the following figures:



Graphic 1 and 2: Represents the variation of Body MassIndex (BMI) and Nutritional diagnosis in each patient. Each bar determinate how much was modified. 0 represents the baseline and downwards is the decrease in BMI or improvement in the nutritional diagnosis.

Conclusions

In this study we observed that PWS under transdisciplinary treatment were able to systematically decrease weight over time and sustain these results.

Achieving better nutritional diagnosis and BMI even without Growth hormone (GH) treatment.

These results are encouraging, especially in regions where there is no access to GH and until the scientific community advances in studies for the use of drugs that helps to control hyperphagia.

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Tratamiento e Investigación de Enfermedades Poco Frecuentes

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