

PWS in Brazil: 6 months follow-up in a referral center

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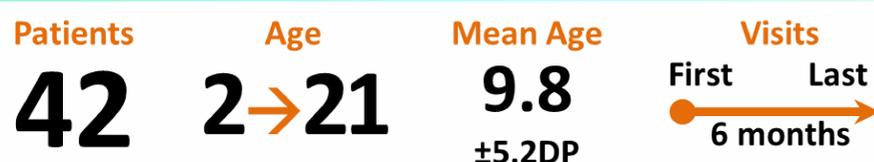
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BACKGROUND

Prader-Willi Syndrome (PWS) patients have been followed in our country in different ways but we do not have a society or a referral center to spread adequate information about the disease.

In January 2015, we started a PWS referral center in Sao Paulo University to promote a better care for patients and families and to support them with a multidisciplinary team.

METHOD



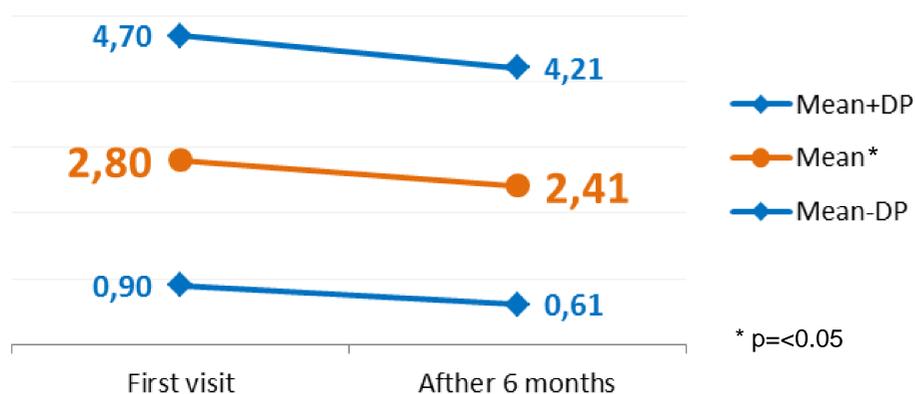
Body mass index-SDS (BMI-SDS) was evaluated at the beginning and after 6 months and these data were compared. The following items were analyzed:

- 1) Use of growth-hormone;
- 2) Metabolic profile: LDL, triglycerides, HbA1c, fasting glucose and insulin levels;
- 3) Polysomnography.

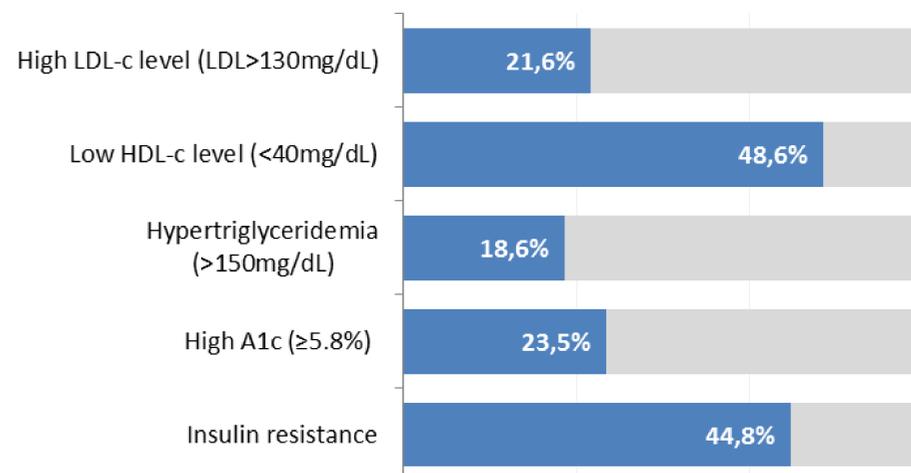
All patients received orientation in diet (900 Calories/day independent of weight), physical activity and behavior. Our team is composed by pediatric endocrinologist, dietician, nurses; neurologist specialized in sleep disorders and otorhinolaryngologist.

RESULTS

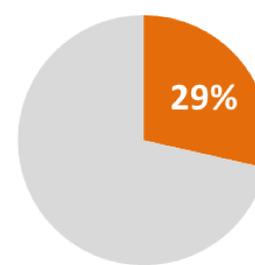
BMI - SDS



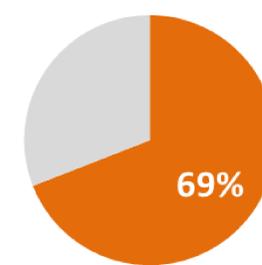
Metabolic Profile



rhGH first visit



rhGH last visit



The reason that thirteen patients were not in use of rhGH is due to polysomnography alterations and patients were waiting for surgery or CPAP.

Polysomnography revealed that 47.8% patients had an apnoea-hypopnoea index (AHI)> 5 events/hour, 20.8% had O₂ saturations under 92% and 56.5% had reduced sleep efficacy.

CONCLUSION

Most of our patients could lose weight with the correct approach in diet, behavior and physical activity. The use of rhGH was increased after the beginning of the clinic and the benefit of this therapy is well known in literature. Alterations in polysomnography were a major problem revealed during follow up and the correct approach of the multidisciplinary team is essential to support this disorder.

