

Nutritional requirements in Prader Willi Syndrome patients under two years of age treated with growth hormone

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BACKGROUND

Prader Willi Syndrome (PWS)

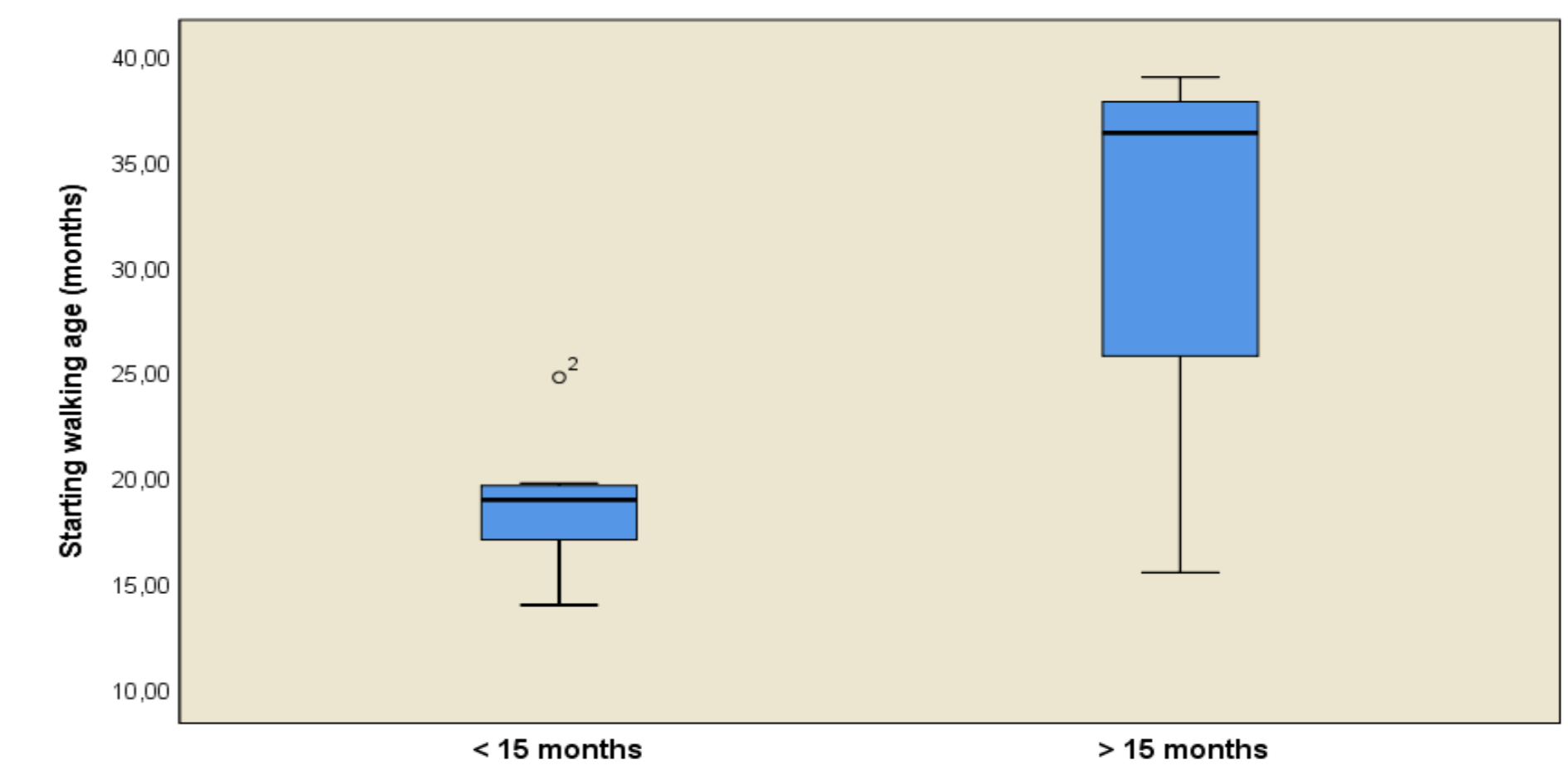
- ❖ **Genetic disorder:** lack of expression paternal Cr 15, q11.2-q13 genes
- ❖ **Clinical features:** hypotonia, cognitive and behaviour disabilities, altered body composition, hyperphagia, hypogenitalism

Treatment with growth hormone (GH)

- ❖ **Clinical improvement:** body composition, cognitive and motor functions
- ❖ GH accepted by FDA in 2000, EMEA 2001 → PWS guide on 2013 recommends early introduction of GH (4-6 months)
 - **Study Parc Taulí: 2-year longitudinal observational study in PWS treated with GH under 2 years of age**
 - Safe
 - Clinical improvement → Better results <15 months: precocious start of speech and walking (JPED 2019 Corripio et al doi.org/10.1515/jped-2018-0539)

Hyperphagia and obesity

- ❖ **Dietetic recommendations:** 25% decrease in the caloric intake is usually recommended to avoid obesity
- ❖ **Hypothesis:** caloric needs in PWS patients under treatment with GH may be higher the first 2 years of life



AIM To study the variation of **BMI** in patients with PWS < 2 years treated with GH and to determine their **caloric intake**

METHODS

RETROSPECTIVE OBSERVATIONAL STUDY

TREATMENT GROUP (G1):

- ❖ SPW <2 years GH treatment

CONTROL GROUP (G2):

- ❖ Historical cohort SPW <2 years **without** GH treatment

Comparing at 9, 12, 15, 18 and 24 months

Studied variables:

- ❖ Weight
- ❖ Height
- ❖ BMI
- ❖ Caloric intake (G1)

RESULTS



CONCLUSIONS

- ❖ **Growth Hormone treatment affects body mass index** in Prader Willy Syndrome patients under 2 years of age
- ❖ It's important to **adjust the caloric intake** of this patients in order to adequate to their **actual needs**
- ❖ It is required to verify these results