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ASOCIACIÓN MADRILEÑA
PARA EL SÍNDROME DE
PRADER-WILLI



POLYSOMNOGRAPHY IN ADULT INDIVIDUALS WITH PRADER-WILLI SYNDROME TREATED WITH GH

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INTRODUCTION: Patients with Prader-Willi Syndrome (PWS) are at risk of a variety of abnormalities of breathing during sleep, such as obstructive and central sleep apnea (OSAS) and abnormal ventilatory responses to hypoxia and hypercapnia. A respiratory dysfunction sometimes concomitant with the treatment with GH has been reported as a possible cause of sudden unexpected death in PWS young subjects. We studied a group of adult PWS subjects who performed repeated complete sleep studies during GH treatment (IGF-1 basal value mcg/l: 94 ± 13 and 305 ± 39 $p < 0.0001$ and 328 ± 32 $p < 0.0001$ after 6 and 12 months respectively).

METHODS: 13 PWS patients, 11 del15 and 2 UPD, 6 females and 7 males, aged 26.9 ± 1.2 years, treated with hormonal replacement (mean dosage 0.064 ± 0.004 mcg/kg/week) were recruited for the study. All patients underwent an adaptation night and then a full-night polysomnography (PSG). Parameters registered were EEG EOG; ECG; respiratory effort by thoracic and abdominal strain gauges, nasal air-flow, snoring nose, oxyhaemoglobin (SaO₂) using a pulse oximeter with finger probe; Macrostructure of sleep analysis (Sleep Stages) according to Rechtschaffen and Kales' criteria was performed. Respiratory parameters included: apnea/hypopnea index (AHI), basal oxygen saturation (basal SaO₂), lowest oxygen saturation (lowest SaO₂), time % spent at oxygen saturation below 90%, average minimum SaO₂ during desaturations. The sleep studies were performed before hormonal treatment and after 6 months and 1 year of treatment.

RESULTS: 4 subjects (2f/2m) resulted positive for OSAS at the basal polysomnography and the two males were treated with CPAP. BMI was unchanged after 6 months and 1 year (basal: 46.3 and 46.4 and 45.8 after 6 and 12 months respectively). AHI was unchanged in 11/12 subjects after 6 and 12 months (basal 7.03, 6 months 7.06 and 12 months 6.7), increased in one subject after 12 months ($1.9 \rightarrow 17.65$ $p < 0.001$). Average minimum SaO₂ during desaturations remained unchanged. On the contrary, lowest SaO₂ progressively lowered ($76.8\% \rightarrow 68.3\%$, $p < 0.05$) and the time % spent at oxygen saturation below 90% increased after 12 months ($24.7 \rightarrow 35.6$, $p < 0.05$).

CONCLUSIONS: A one year's GH treatment does not seem to be associated with a worsening of AHI in adult PWS subjects. Further observations may be suitable to confirm the registered SaO₂ alterations. Our data suggest the opportunity of periodical saturimetric nocturnal respiratory controls both in PDW OSAS and non OSAS adult subjects.