

Therapeutic education contributes to minimize excess weight in Prader-Willi syndrome

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INTRODUCTION:

Prader-Willi syndrome (PWS) is a complex genetic disorder characterized by hyperphagia with progressive obesity, dysmorphic features, hypotonia, mental retardation, behavioural abnormalities and endocrine dysfunctions as hypogonadism and growth hormone (GH) deficiency. PWS is the most commonly identified genetic cause of obesity.

METHODS:

We reviewed 5 cases of confirmed PWS (3 female and 2 male patients, aged between 8-32 years old) evaluated at the Endocrinology Department of Iași between January 2008 – July 2013. Clinical and hormonal data were documented.

RESULTS:

All patients had specific clinical features of PWS and genetic confirmation. They were all born in non-sanguine couples, four of them with low birth weight. After the first year of life they presented hyperphagia with rapid and important weight gain, except for one case where hyperphagia began in adolescence (at 13 years of age). The major weight gain was until late adolescence (> +6 SD). Nutritional counselling, with detailed information about diet, exercise, and subsequent adverse consequences of obesity, was offered to patients and family in order to ameliorate their eating behaviour. After that, the 3 patients that reached adulthood achieved a mean weight at +1.5 SD (under rigorous alimentation). Paradoxically, all patients had a higher height than expected (mean +1.5 SD) in childhood and adolescence, even though three of them had low IGF1. Two patients had their first endocrinologic examination in adulthood and presented a satisfactory final height (>155cm)

CONCLUSIONS: Therapeutic education of the family, performed as early as possible, is an important determinant of the disease evolution and diminishes the potential weight gain. Although most patients with PWS have morbid obesity it seems that caregivers involvement in weight management may lead to favorable results.