INTRODUCTION

Type 1 Narcolepsy (NT1) is a rare paediatric disorder characterized by excessive daytime sleepiness and cataplexy. The cause is a selective loss of hypocretin-secreting neurons, probably with an autoimmune etiology [1]. Hypocretin deficiency could affect the neuroendocrine system, causing alterations in energy balance, eating behaviour, glucose metabolism and modulation of the hypothalamic-pituitary axis. In particular, obesity, GH deficiency and Central Precocious Puberty (CPP) are endocrine complications already reported in literature for these patients [2].

AIM

The aim of this study was the evaluation of endocrine-metabolic and auxological aspects at diagnosis and during follow-up in children affected with NT1. Secondary objective was to evaluate of the effect of Sodium Oxibate (SO) in the trend of the endocrine-metabolic parameters and the identification of possible prognostic factors that could identify patients at risk for endocrine complications.

RESULTS

98 patients fulfilled inclusion criteria and were recruited for the study: 53 boys and 45 girls. Auxological parameters at diagnosis and during follow-up are reported in Table 1. Median follow-up was 36 ±16 months. A diagnosis of CPP was found in 21% (21/98) and 11 subjects started treatment with GnRH analogue according to the guidelines [5].

Obesity was found in 33% (33/98), while 25% (25/98) was found Overweight. A significative increase in baseline insulin (uIU/ml) values was found in obese subjects (15.1 ± 13.4) compared to others (12.7 ± 10.5) (p<0.05).

Others laboratory parameters were found in the normal range. Stature (SDS) at diagnosis and during follow-up is reported also in Table 1. Mean IGF-1 levels was 297.8 ± 133.3 µg/L (SD 0.4 ± 1.0).

Patients treated with SO at 12 months of follow up were 46/98 (46.9%). The BMI trend is reported in Graphic 2. At 36 months of follow up, we find a significative difference in BMI SDS between SO treated vs untreated patients (0.0 ± 1.3 vs 1.3 ± 0.4) (p<0.003). 62 patients reached their final height (mean CA 15.9 ± 2.5) with SDS of 0.5 ± 1.1 in boys and 0.3 ± 1.2 in girls. Target parental height was 0.2 ± 0.8 SDS in boys and 0.3 ± 0.7 SDS in girls.

CONCLUSIONS

The results of our study confirm an increased frequency of CPP and obesity in NT1 patients compared to general population [1]. A predictor sign of CPP seems to be an earlier age at onset of NT1.

According to a previous study [6], we demonstrate that treatment with SO leads to a significative improvement on BMI, which also persist at 36 months of follow up.

To our knowledge, these are the first results regarding the final height in a large series of patients diagnosed with NT1 in pediatric age.

Our results appear in contrast with the hypothesis of a GH deficiency, in fact, we find IGF-1 levels and stature (SD) in the normal range, as the final height where available.

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REFERENCES


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