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Follow-up and Prevalence of Precocious Puberty in Children with Classical Congenital Adrenal Hyperplasia diagnosed by Neonatal Screening

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Introduction and objectives

In children with classical congenital adrenal hyperplasia (CCAH) linear growth allows monitoring metabolic control. Precocious puberty has increased in recent years. There are few studies in patients with CCAH diagnosed by neonatal screening about this subject. CCAH screening in our province became obligatory since 2010. Between 2010-2017, 1.260.814 newborns were evaluated . The incidence of CCAH was 1/21.370.

The objective was to analyze linear growth and prevalence of precocious puberty in a group of children with CCAH detected by neonatal screening.

Methods

Thirty-two patients (F:15, M:17) with CCAH diagnosed by extracted 17-ohp levels and molecular analysis were included. Thirty of them presented salt wasting form (SW), the other two were simple virilising (SV) forms.

They were evaluated at start of treatment, at 6 and 12 months of age, and then annually. Twenty one (F:9; M:12) of them started puberty and seven reached final height.

We analized chronological age (CA) at start of treatment (CAST), z-height, z-BMI, hydrocortisone dose (HCd) and bone age (BA) by Greulich and Pyle, up to the age of five years. We calculated ΔBA at start of puberty and one year previous; ΔBA-CA at start of puberty.

Final height was compared with mid parental height (MPH).

Statistical analyisis was conducted using R version 3.5.1. Repeated measures ANOVA and Spearman correlation were performed.

Results

Median CAST was 18 (10;22) days.

Mean height, BMI and BA and median HCd are shown in table 1.

Table 1

	Start	6 month	1st year	2nd year	3th year	4th year	5th year
Mean height SDS	-0.9±1.5	-1.8±1.85	-1.6±1.67	-1.15±1.27	0.71±1.13	0.47±1.23	0.11±1.28
Mean BMI SDS	-1.46±1.21	0.24±1.27	0.67±1.42	0.68±0.91	0.89±0.96	0.97±1.23	1.33±1.40
HCd (mg/m2/day)	36.00 (32.8;43.55)	20.84 (19.12;22.09)	17.94 (16.12;19.52)	16.00 (14.44;19.20)	15.13 (13;19.91)	14.52 (11.83;16.83)	14.00 (10.79;18.09)
Mean BA SDS			0.87±0.33	1.89±0.85	2.59±1.03	3.88±1.33	5.36±2.43

No significant differences were found between variables analyzed by sex. Negative correlation was found between HCd and height (r=-0.27, p<0.0001).

During follow-up, a group of 21 patients started puberty, 6 boys and 4 girls at 11±0.9 and 9.5±0.21years, respectively.

Six boys and five girls (34%) presented precocious puberty at 6.4±1.85 and 6.72±0.35years, respectively.

Media ΔBA at start of puberty and one year previous was 3.05 years in boys and 1.8 years in girls (Figures 1 and 2).

Media ΔBA-CA at start of puberty was 4.4 years in boys and 3.5years in girls.

They are in treatment with GnRH analogue and haven't reached final height yet.

Final height in seven of them (F:2; M:5) was -1,17±0,6 SDS, at -0.75±0.79 SDS below MPH (Figure 3).

Figure 1

BA at diagnosis and one year previous in boys

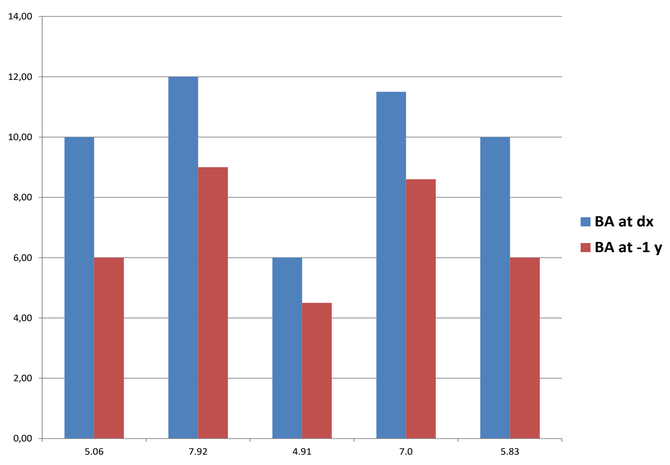


Figure 2

BA at diagnosis and one year previous in girls

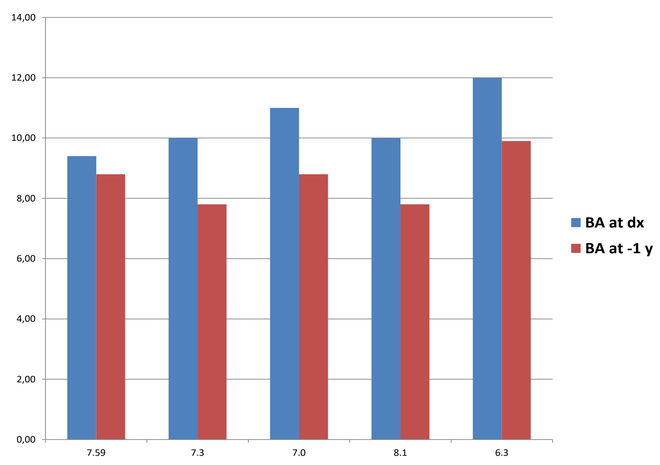
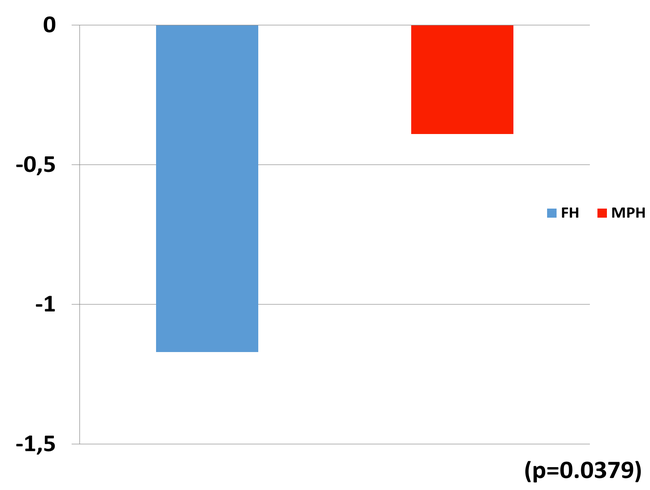


Figure 3

Final height and mid parental height SDS



(p=0.0379)

Conclusions

All patients showed normal BA up to the age of 5 years, but later on, BA progressed rapidly in patients who developed Precocious Puberty.

Final height was normal, but slightly lower than mid parental height in children with normal puberty.

In spite of early diagnosis in this group of patients, precocious puberty was frequent, suggesting that other factors besides compliance are important. Other markers of good metabolic control and treatments are needed to improve outcome.

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