

FINAL HEIGHT OF A GROUP OF PATIENTS WITH CONGENITAL ADRENAL HYPERPLASIA

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Introduction Congenital adrenal hyperplasia (CAH) is a group of autosomal recessive disorders characterized by impaired cortisol synthesis with an incidence of 1:15 000 births. The most common form is attributable to mutations in CYP21A2, the gene encoding the adrenal steroid 21-hydroxylase enzyme (P450c21). Because of that, cortisol synthesis is impaired which leads to a corticotropin stimulation of the adrenal cortex, with accumulation of cortisol precursors that are derived to sex hormone biosynthesis. Aldosterone production may also be decreased (salt wasting forms). The final height of children with these disorders seems to be compromised. That can happen because of the disease itself or because an excessive dose of corticoids used during treatment. This work evaluates the final stature SDS of a group of patients with CAH and correlates it with 17-hydroxyprogesterone levels (17OHP) and corticoid dose at 10 years of age and at puberty onset.

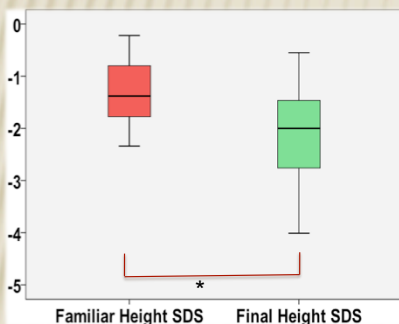
Methods Selection of children with CAH diagnosed from 1983/08/01 to 1995/08/01, followed in an outpatient clinic of paediatric endocrinology, with all necessary data in their clinical files: height, weight, 17OHP and corticoid dose at 10 years of age, puberty onset and at the last medical evaluation, parents height. Data was analysed in SPSS 20.0, with descriptive statistics, Wilcoxon test, Spearman's correlation coefficient and Mann-Witney U test (significance level: 0.05).

Results

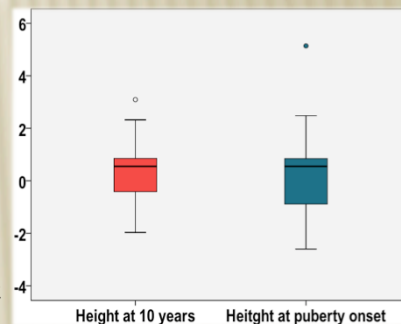
n=19 children

♂ = 8
♀ = 11

13 children were diagnosed in the first year of life
13 children had salt wasting forms of CAH



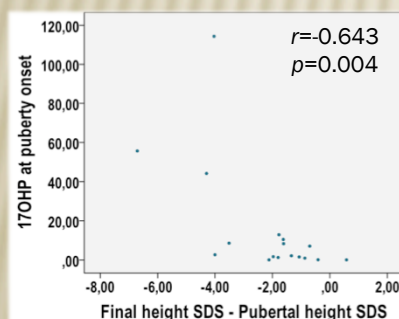
Graph 1: The mean final height SDS of these children was -2.07 (from -4.01 to -0.55; standard deviation, SD=0.98). Their mean familiar height SDS was -1.33 (from -2.34 to -0.22; SD=0.64). This difference was statistically significant ($p=0.003$). In centimeters, their mean height was 155 (from 141 to 171 cm; SD=7.44)



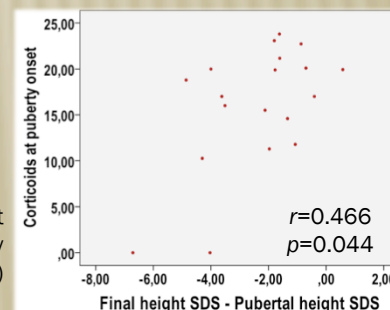
Graph 2: At puberty onset, these children's mean height SDS was 0.33 (from -2.6 to 5.14; SD=1.76). When they were 10 years old, their mean height SDS was 0.29 (from -1.69 to 3.09; SD=1.26).

Final height (SDS)	Moment	17 OHP (ng/mL)	r (Spearman's correlation coefficient)	p	Hydrocortisone dose (mg/m ²)	r (Spearman's correlation coefficient)	p
-2.0 SDS (-4.01–0.55)	10 years of age	4.5 (0.47-7.34)	-0.77	0.75	16 (9.38-28.70)	-0.08	0.75
	Puberty onset (Median=9.5; min-max=7-13.4)	4.9 (0.06-7.34)	-2.26	0.29	17 (0.00-23.80)	-0.003	0.99
	Last evaluation	4.9 (7.27-27.39)	-0.30	0.21	18.18 (0.10-153)	-0.70	0.77

Table 1: The final height SDS had no statistically significant correlation with 17OHP and corticoid dose used at puberty onset, at 10 years of age or at the last clinical evaluation.



Graph 3: The difference between the final height SDS and the pubertal height SDS was negatively correlated with 17OHP (ng/mL) at puberty onset.



Graph 4: The difference between the final height SDS and the pubertal height SDS was positively correlated with corticoid (hydrocortisone – mg/m²) dose used at puberty onset.

Conclusion

In this group of children the final height SDS was inferior to their target familiar stature, because of the poor growth after puberty onset: their height SDS when they were 10 years old and on their puberty onset were better than the final one. There wasn't any correlation with 17OHP and corticoid dose used during treatment. When the growth after puberty onset was analysed, it was positively correlated with corticoid dose and negatively correlated with 17OHP levels on puberty onset. There are many studies reporting that high doses of corticoids used in these children may impair their growth. But, in this study, to achieve better levels of 17OHP, higher doses of corticoids were used. So, maybe that could have helped to achieve a better growth before puberty and could have prejudiced the growth after puberty onset.

References

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