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IMPROVED LINEAR GROWTH IN PATIENTS WITH CLASSICAL CONGENITAL ADRENAL HYPERPLASIA

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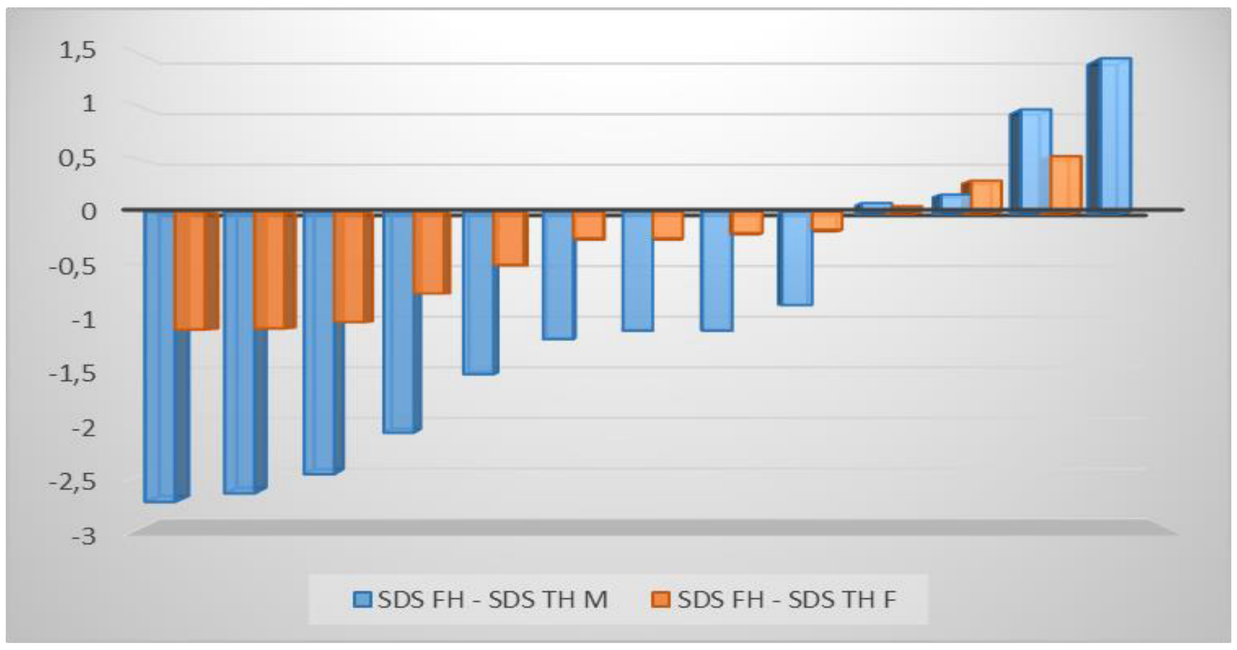
OBJECTIVES

Background: Poor linear growth is still one of the main concerns in children with congenital adrenal hyperplasia (CAH). An impairment of linear growth, adversely affecting final height, has been related both to overtreatment with glucocorticoid replacement therapy and to poor control of adrenal androgen levels.

Objective and hypotheses: to define factors that influence linear growth and final height in patients with classical CAH.

METHODS

The medical records of 25 patients (12 females, 13 males) followed from early infancy until adulthood in our Institution were analyzed and the clinical observations were divided into **4 groups according to age and puberty** (0-2 years, 2 years up to puberty onset, pubertal years, post-pubertal years). Differences in the mean dose of hydrocortisone and average levels of Δ4-androstenedione and 17-hydroxyprogesterone between the 4 groups were evaluated using one-way ANOVA. Multivariate analysis was used to study factors independently affecting final height.



	0-2 years	prepuberty	puberty	postpuberty	p
Hydrocortisone dose mg/m²/day	23.3 ± 7.5	16.5 ± 3.6	18.1 ± 4.4	15.9 ± 6.4	<0.001
Δ4-androstenedione ng/ml	0.74 ± 1.5	0.74 ± 1.4	3.7 ± 3.4	2.9 ± 3.4	<0.001
17OH progesterone ng/ml	24.5 ± 90.1	15.8 ± 58.6	26.8 ±35.9	23.4 ± 44.6	0.098

RESULTS

Mean final height was 167.8±7.1 cm in males (-1.3±1.1 SDS) and 158±6.5 cm in females (-0.8±1 SDS).

Final height corrected for parental height was -1.01±1.3 in males and -0.37±0.5 in females.

Mean total pubertal growth spurt was 23.1±4.6 cm in males and 19.8±6.5 cm in females. Significantly higher **doses of hydrocortisone** were required during the first two years of life and during puberty ($p<0.05$).

Multivariate analysis showed that final height was adversely affected by the average dose of hydrocortisone ($p<0.001$) and by the mean level of Δ4-androstenedione ($p<0.045$) during follow up.

CONCLUSIONS

The main goals of therapy in children with CAH are the control of glucocorticoid deficiency and the suppression of adrenal androgen hyper-secretion. Particularly during puberty, the daily dose of hydrocortisone should be maintained as low as possible to obtain a normal pubertal growth spurt and optimize final height. Final height of CAH patients followed from early infancy, particularly females, seems to be now less impaired than previously reported.

References

Study	FH sds tot	FH M	FH F	FH-TH sds	FH-TH M	FH-TH F	Spurt tot (cm)	Spurt M	Spurt F
OPBG 2014	-1.1±1.1	-1.3±1.1	-0.8±1	-0.71±1.1	-1.01±1.3	-0.37±0.5	21.5±5.7	23.1±4.6	19.8±6.5
Brunelli 2003	-1.55±0.3	-1.5±0.2	-1.6±0.4	-0.95±0.3					
Balsamo 2003	-1.3±0.4	-1.5±0.6	-1.1±0.2	-0.86±0.3	-1.04±0.3	-0.6±0.1	17.15±4.1	19.1±3.5	15.2±4.6
Sarafoglou 2014	-0.8±1.1	-0.9±1.1	-0.7±1.2	-0.55					
Han 2014		-1.11±1.09	-1.05±1.14		-2.13±1.69	-1.03±1.2			
Mothusamy 2010	-1.38			-1.03					
Bonfig 2009	-0.95±0.2	-1.1±0.2	-0.8±0.1	-0.67±0.2	-0.9±0.1	-0.45±0.2	15.2±2.1	16.9±1.1	13.4±0.4
Eugster 2001	-1.37			-1.21					

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