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Evaluation of medical treatment in the first 2 years of life with a new Dutch national longitudinal registry for children with congenital adrenal hyperplasia (CAH)

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Introduction

Recently, a national database (NEORAH) has been developed to register yearly data from all children detected with CAH in the neonatal screening program from 2002 onwards. So far longitudinal data of 105 children have been registered (roughly 65% of Dutch CAH patients) to evaluate medical treatment and long term effects in CAH. A national CAH working group developed guidelines for diagnostics and follow up in CAH. Aim of our current study is to evaluate the dosage of hydrocortisone, fludrocortisone and salt supplementation in CAH children in the first 2 years of life.

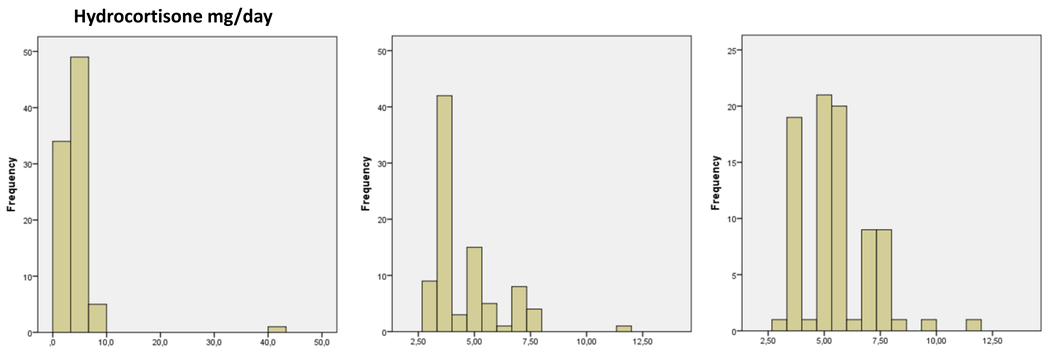
Methods

This is a descriptive study in 105 CAH children. The hydrocortisone, fludrocortisone and salt supplementation was evaluated at the age of 6 months (T 0,5), 12 months (T1) and 24 months (T2).The treatment approaches of different centers were compared. Length and weight gain in the first 2 years are evalueated.

Results

T 0,5: (n=95): median HC dosage (n=89) 4 mg/day; interquartile range (IQR) 2 mg/day. FC (n=82) median 93,75 mcg/day ; IQR 37,5 mcg/day and NaCl (n=46) median 500 mg; IQR 400 mg/day.
T 1: (n=90) median HC dosage (n=88) 4 mg/day; IQR 1 mg/day. FC (n=84) median 62,5 mcg/day; IQR 30,3 mcg/day and NaCl (n=25) median 375 mg/day; IQR 400 mg/day.
T2: (n=88) median HC dosage (n=84) 5,4 mg/day; IQR 2,14 mg/day. FC (n=78) median 62,5 mcg/day; IQR 37,5 mcg/day. No salt medication was used after the first year of life.
No significant differences between centers are observed at all T's.
Mixed models were used for evaluation of length and weight gain. This showed a decline in length of 0,19 SD/year and a decline in weight of 0.34 SD/year.

A



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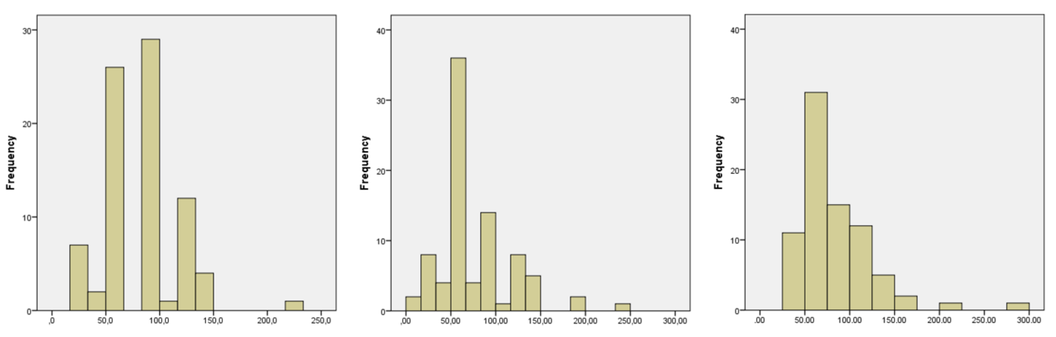

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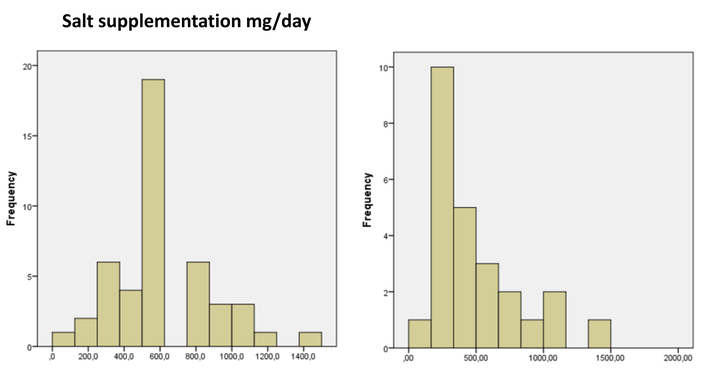

A: hydrocortisone dosage at T0,5; T1 and T2.
B: fludrocortisone dosage at T0,5; T1 and T2.
C: sodium supplementation at T0,5 and T1
On x axis: daily dosage. On y axis: number of patients (frequency)

Conclusion

Our preliminary analysis showed a stable slightly supraphysiological HC dosage over the first 2 years of life. FC shows a more widespread range in dosage suggesting a more individualized treatment approach without a clear difference between national centres. In the first 2 years the patients show a worse length and weight gain than expected. This effect seems greater with a higher dosage of HC. Further studies will focus on the relation between different genotypes, medication dosage and biometrical data.
Our longitudinal database enables us to establish a more standardized care and long term treatment evaluation.

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