P641

Influence of salt supplementation on drug therapy in children with congenital adrenal hyperplasia (CAH) due to 21 hydroxylase deficiency aged 0-3 years: Update on a retrospective multicentre analysis using the International-CAH (I-CAH) registry

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Introduction

Early treatment of classic congenital adrenal hyperplasia (21-hydroxylase deficiency) with glucocorticoids and mineralocorticoids will prevent life-threatening crisis. In some centres, additional salt is prescribed in the first year. However, until now the use of salt is controversial and the benefit has not been proven in studies.

Objectives

To evaluate the effect of additional salt supplementation on fludrocortisone (FC) dosage, hydrocortisone (HC) dosage, height and blood pressure in young CAH-children (0 – 3 yrs).

Methods

- Retrospective analysis in July 2018
- 23 centres,
- 13 countries, worldwide

- **355 patients**
- born after 2000
- 21-hydroxylase deficiency
- **2672 visits** at 9 time points (birth, 3, 6, 9, 12, 18, 24, 30, 36 months of age)

154 children without salt treatment (NST) between 46 and 365 days of life (assigned female n=83, assigned male n=71)

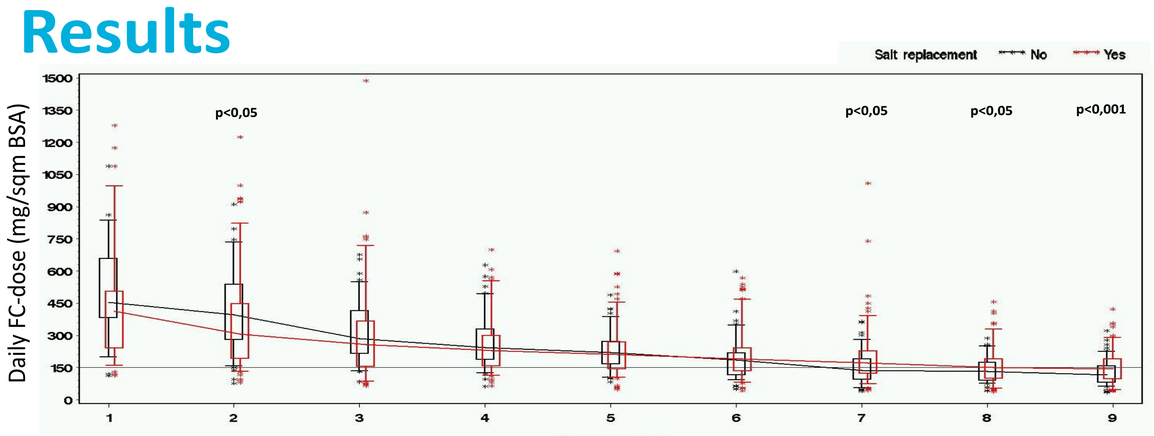
195 children treated with salt (ST) at any time between 46 and 365 days of life (assigned female n=115, assigned male n=80)

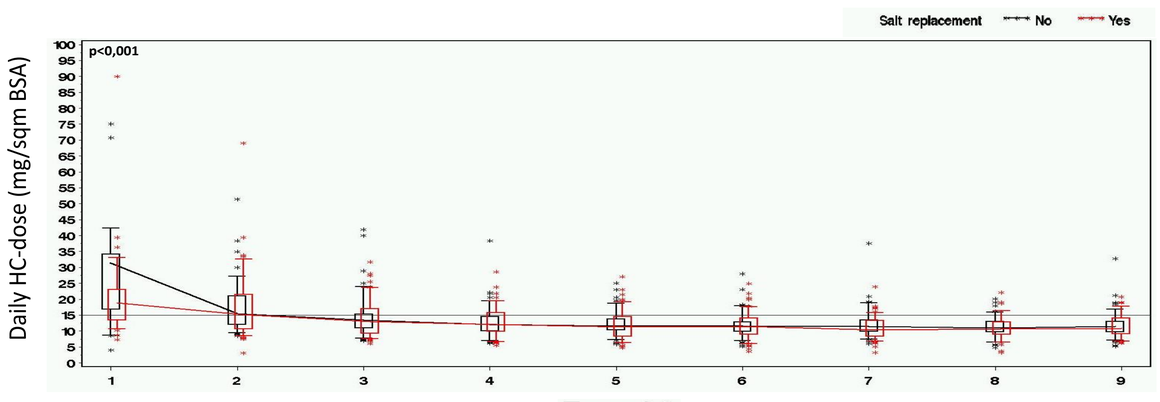
6 children excluded (missing information on salt therapy)

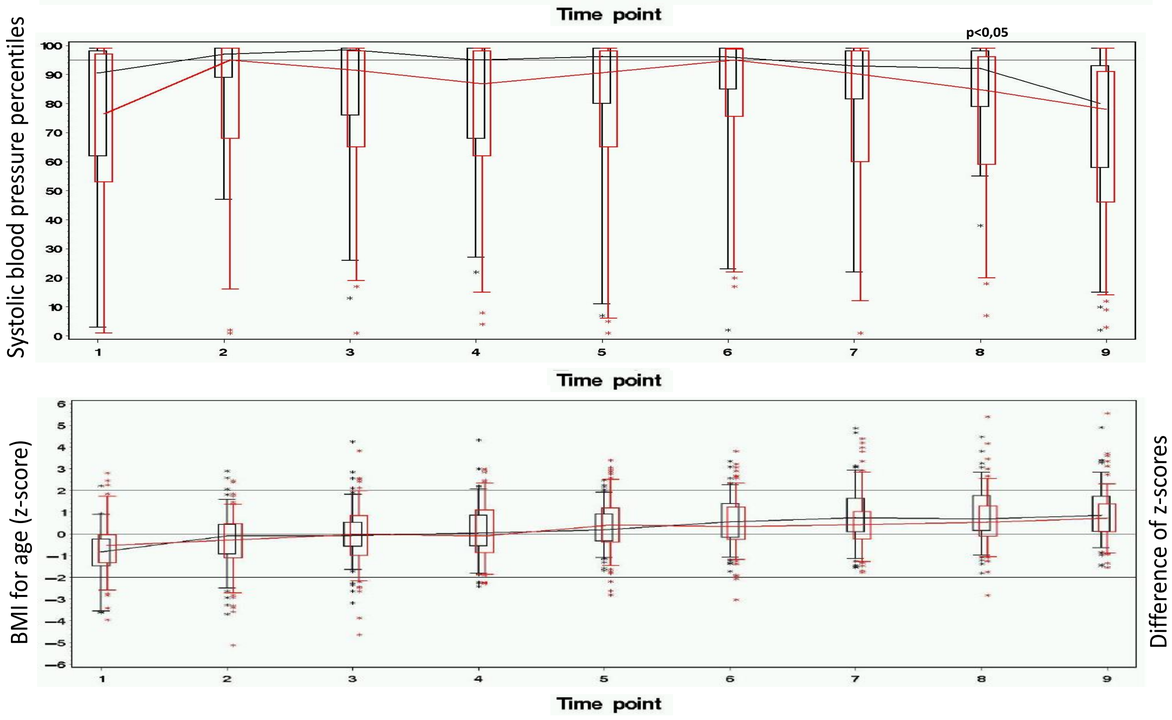
Conclusion

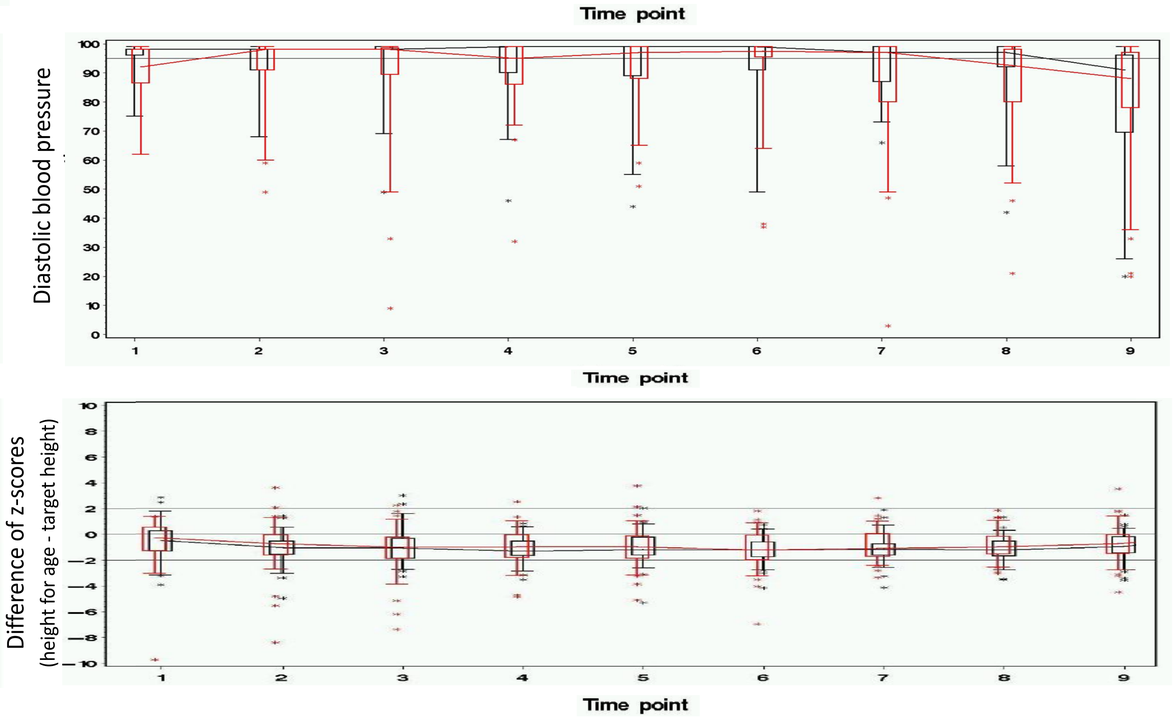
- Additional salt supplementation during the first year of life in CAH patients reduced FC-dose until 136 days and might reduce the risk of systolic hypertension. Although all CAH children showed a high percentage of hypertensive blood pressure readings, possibly due to FC-therapy or measurement method in young children.
- Independent from salt treatment there is a risk of decreased height gain and increasing BMI in all CAH children from 0-3 years of age which should imply further studies on HC-treatment and dosage in young children with CAH.

Results









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Adrenals and HPA Axis

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