Cognitive Functions in Congenital Adrenal Hyperplasia

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INTRODUCTION

Early exposure to androgens in congenital adrenal hyperplasia (CAH) is postulated to be linked to changes in cognitive abilities. It has been hypothesized that certain cognitive changes in CAH is a possible

Aim

Assess cognitive functions in children with CAH, and their relation to hydrocortisone (HC) therapy and testosterone level

Subjects and Methods

- **Setting:** Children's Endocrine Clinic , Ain-Shams University Hospital, Cairo, Egypt
- **Study duration:** 1 year (08/2013 – 08/2014)
- **Type of study:** Case control study
- **Subjects:** 30 patients with CAH due to 21 hydroxylase deficiency aged 6 -16 years on glucocorticoid treatment (11 salt wasting, 19 simple virilizing)
- **Exclusion criteria:** co-morbid psychiatric diagnosis
- **Control group:** 20 healthy, age- and sex-matched children
- **Ethical approval:** local ethics committee of Ain-Shams University. All patients and controls or their legal guardians signed an informed consent prior to the study
- **Evaluation**
- Clinical evaluation including auxology
- Calculation of hydrocortisone (HC) daily dose and cumulative dose since starting treatment
- Socioeconomic standard [2]
- Free testosterone level
- Cognitive function assessment was performed using

1. **Wechsler Intelligence Scale – Revised for Children (WISC):** reflects the intellectual performance through verbal, performance, and full scale IQ [3]
2. **Benton Visual Retention Test:** assesses visual perception, memory and visual-constructive abilities [4]
3. **Wisconsin Card Sorting Test (WCST):** a tool for recognizing frontal cortical executive functions (planning – shifting – cognitive flexibility – sustained attention) [5]

RESULTS

- Mean age (SD) of patients was 10.22 (3.17) years
- Eleven males (36.7%), 19 females (63.3%)
- Mean (SD) duration of treatment 114.83 (43.61) months
- Mean (SD) HC dose was 15.78 (4.36) mg/m2/day
- Mean (SD) cumulative HC dose 44,689. 9 (26,892.0) mg
- No significant difference in age, gender, socioeconomic standard, and anthropometric data existed between patients and controls ($p > 0.05$)
- No significant difference in cognitive performance was found when patients were subdivided according to daily HC dose (< 10, 10 – 15, > 15 mg/ m2/day), or according to salt wasting state ($p > 0.05$)

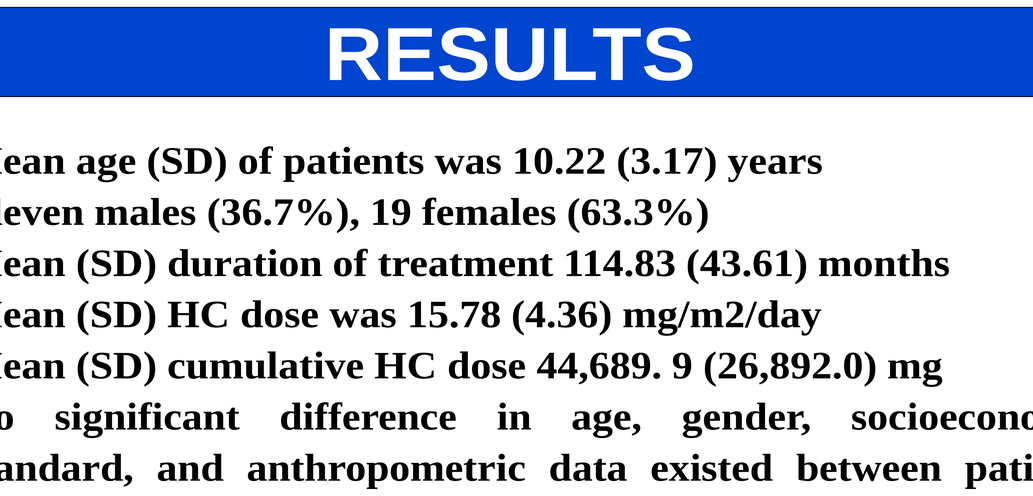

Figure 1: VIQ: verbal intelligence quotient, PIQ: performance intelligence quotient, TIQ: total intelligence quotient
Normal score: > 90, * $P < 0.05$
Patients had significantly lower scores in all domains of WISC test

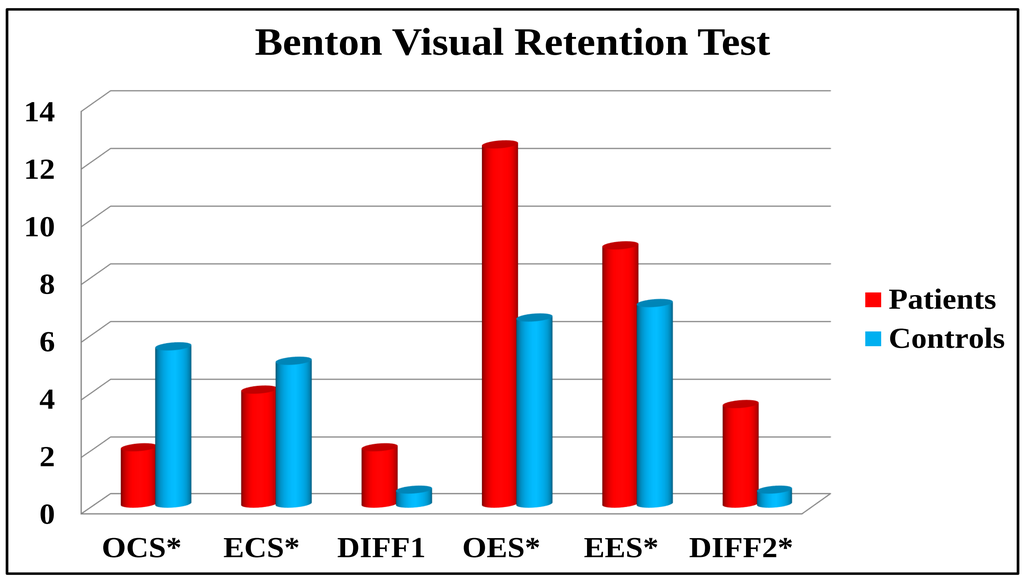

Figure 2: OCS: obtained correct score, ECS: expected correct score, OES: obtained error score, EES: expected error score
Normal score: DIFF < 4, * $P < 0.05$
Patients had significantly higher mean difference between obtained and expected errors
Patients had significantly lower obtained correct scores and higher expected error score

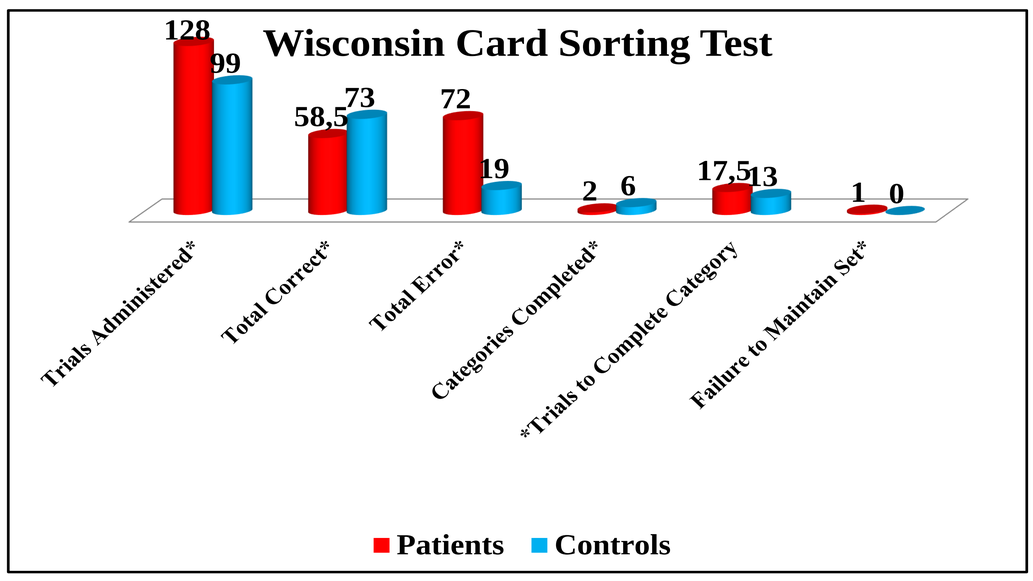

Figure 3: Normal score: categories completed > 6, failure to maintain set: zero
Patients performed significantly worse compared to controls, with less correct trials, more errors, and more failed trials to complete the test categories. * $P < 0.05$

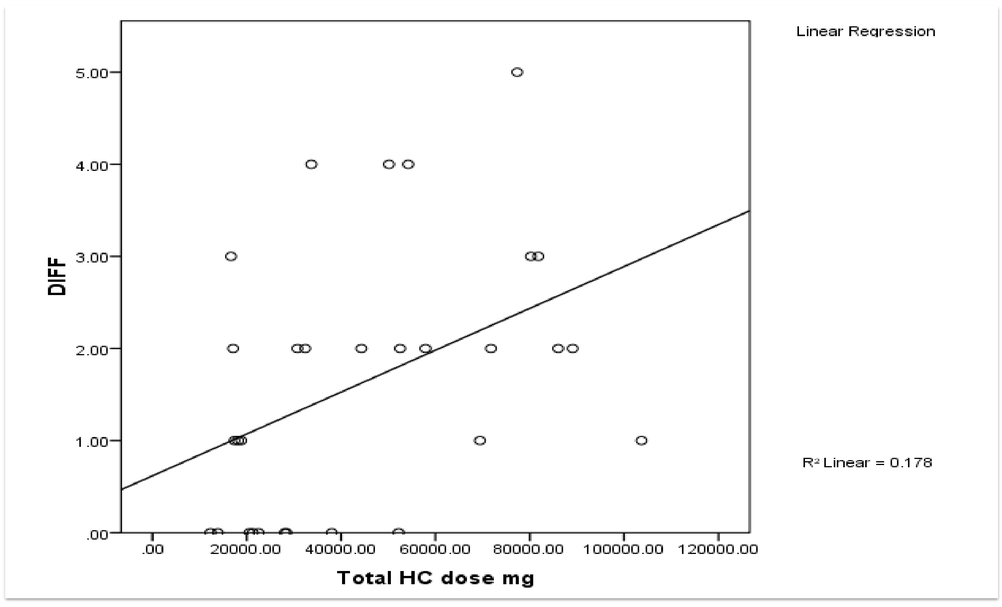

Figure 4: Correlation between Benton Visual Retention Test and Total HC dose ($p < 0.05$)

- **Conclusion:** Cognitive changes affecting intellectual performance, spatial relation, memory for newly learned material, and executive functions exist in patients with CAH. Further studies are warranted for the etiology of such changes
- **Disclosure:** The authors declare no conflict of interests

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