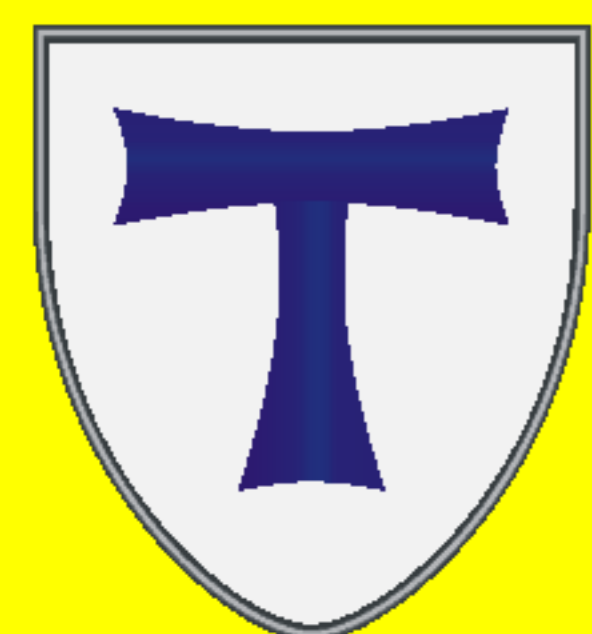


Effectiveness of rhGH treatment in a boy with nephrogenic diabetes insipidus



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Background

The majority of children with primary nephrogenic diabetes insipidus grow below the third centile [1-3].

Objective and hypotheses

Effect of rhGH treatment on growth in a patient with primary nephrogenic diabetes insipidus.

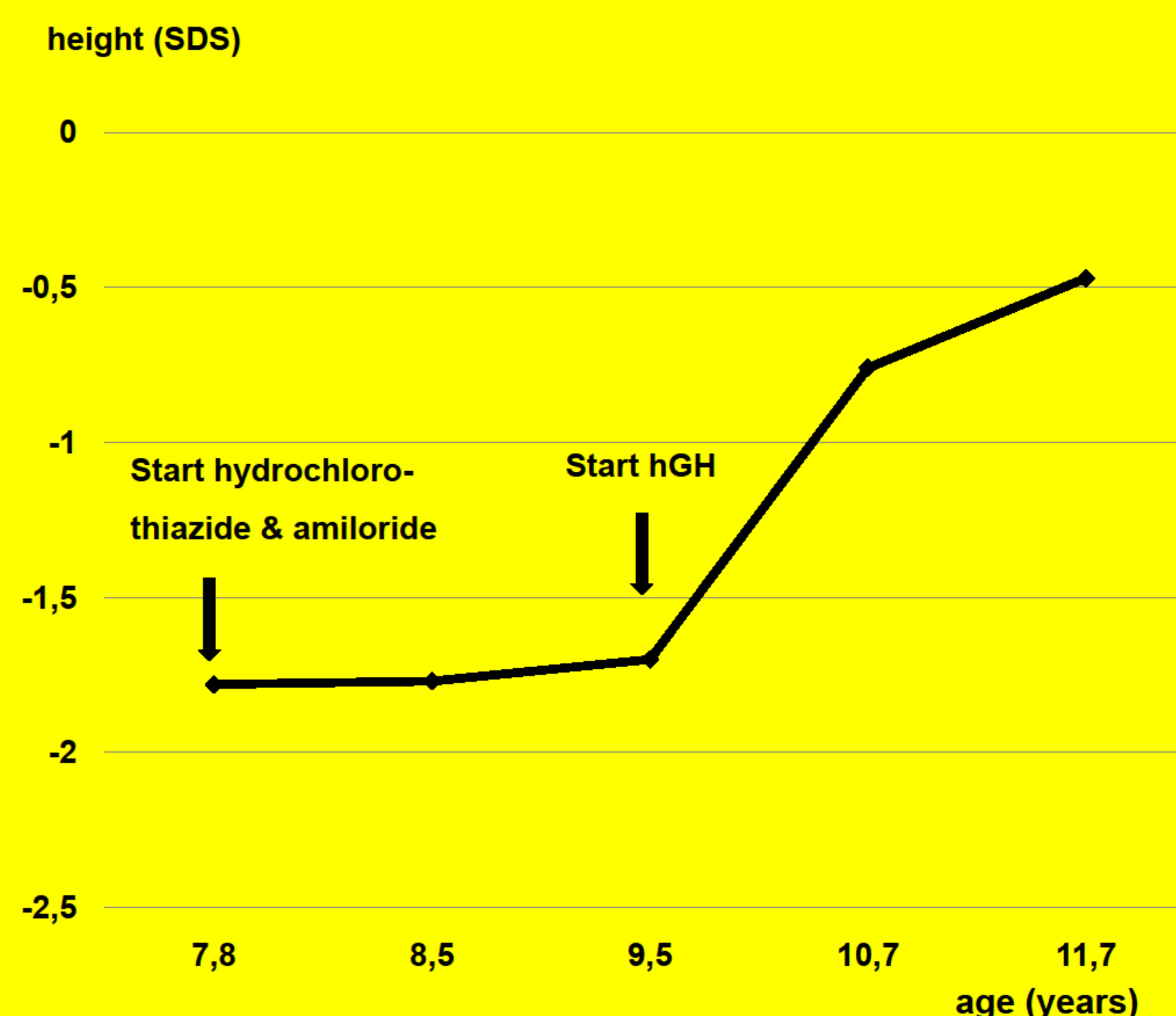
Methods

Case report.

Results

The patient is an 11-yr and two month old Caucasian boy of unrelated healthy parents. At the age of 7 ys and 9 month he was admitted to our hospital for evaluation of polydipsia and polyuria. His body height was 116.0 cm (-1.8 SDS). Urine volume was 4165 mL/ 24h (5.2 L/m²). During a water deprivation test, urine osmolality was below 200 mosm/L while plasma sodium and plasma osmolality increased to 140 mmol/L and 305 msom/L, respectively. Administration of desmopressin revealed no increase in urine osmolality and a mutation in the Aquaporin 2 gene was found during molecular analysis (c.732del C in exon 4 of the AQP2 gene). Treatment with hydrochlorothiazide (2 mg/kg/d) and amiloride (0.2 mg/kg/d) led to a decrease of urine output to 2800 mL/ 24h (3.5 L/m²). At the age of 9 1/2 ys his height was 125.3 cm (-1.7 SDS). Levels of IGF-I and IGFBP3 were 90 µg/L (-2.1 SDS) and 2.0 mg/L (-1.4 SDS), two GH stimulation tests revealed low GH levels below 8 ng/mL and GH treatment was started with 0.8 mg per day s.c. (0.029 mg/kg). He showed a good catch-up growth. At the age of 11 ys and 8 months his height was 144.0 cm (-0.47 SDS). Levels of IGF-I and IGFBP3 were within the normal range (198 µg/L (-0.28 SDS) and 3.29 mg/L (-0.52 SDS), respectively).

Figure 1



Conclusions

Our patient had a GH deficient state and rhGH treatment induced impressive catch-up growth. GH deficiency should be investigated in short children with nephrogenic diabetes insipidus.

Literature

1. Lejarraga H, Caletti MG, Caino S, Jiménez A. Long-term growth of children with nephrogenic diabetes insipidus. *Pediatr Nephrol* 2008;23:2007-12.
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