Pediatr Nephrol (1998) 12: 83–85

Pediatric Nephrology

Letters to the editors

Body growth in primary de Toni-Debré-Fanconi syndrome

Key words: Growth – de Toni-Debré-Fanconi syndrome – Supportive therapy

Sirs,

We read with interest the paper by Haffner et al. [1] on growth in primary de Toni-Debré-Fanconi syndrome. Our experience, although based on a much shorter follow-up, is very different. Four children (2 males, 2 females) with a primary de Toni-Debré-Fanconi syndrome have been followed in our department since 1993. Defined inborn errors of metabolism were excluded. Median age at diagnosis and start of treatment was 2 years (range 1.5–3.1 years) and longitudinal growth was examined for all children over a median period of 2.7 years (range 2–3.5 years). All patients were treated from the beginning with ibuprofen (20–30 mg/kg per day), vitamin D_3, oral phosphate (P), calcium (Ca), and bicarbonate. After the first administration, no further potassium or magnesium supplement was necessary. After institution of therapy, Ca and P were only occasionally low. Plasma bicarbonate, despite supplementation, was low in 15%–36% of the laboratory tests. No child received recombinant growth hormone. Creatinine clearance at last observation was normal in all cases (mean 105 ml/min per 1.73 m^2, range 87–122). Children were seen in our clinic approximately every month for the 1st year and then every 2–3 months.

Growth in our patients always improved after the first observation; height standard deviation score (HtSDS) was, on average, –2.4 (range –3.4 to –1.9) at diagnosis and –1.3 (range –1.7 to –0.7). at the end of follow-up. Comparing HtSDS curves in our patients (Fig. 1) with those published by Haffner et al. [1], HtSDS at diagnosis was <-2 DS in both groups but statural growth in our patients improved progressively.

In our experience, children with idiopathic primary de Toni-Debré-Fanconi syndrome improve their initial height and reach normal percentiles. Therefore, we cannot agree with the sentence "supportive therapy is frequently unable to prevent further loss of relative height." Non-steroidal anti-inflammatory drugs and the subsequent persistent normalization of plasma electrolytes could be the main explanation for the better growth observed in our children, because only one of the patients reported by Haffner et al. [1] took indomethacin regularly.

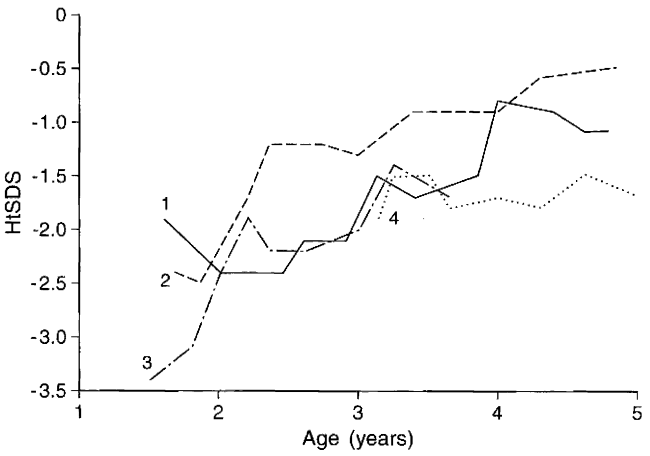

Fig. 1. Changes in height standard deviation score (*HtSDS*) of four children with de Toni-Debré-Fanconi syndrome

M. Greco, A. Sesto, and G. Rizzoni
Division of Nephrology and Dialysis
Bambino Gesù Children's Research Hospital
Rome, Italy

Reference

1. Haffner D, Weinfurth A, et al (1997) Body growth in primary de Toni-Debré-Fanconi syndrome. Pediatr Nephrol 11: 40–45