## Reply

**Key words:** Primary de Toni-Debré-Fanconi syndrome – Body growth – Acidosis – Potassium

Sirs,

Greco et al. describe impressive catch-up growth in four growth-retarded children with primary de Toni-Debré-Fanconi syndrome (FS) after initiation of supportive treatment; this is in contrast to our results. From the shorter observation time compared with our study, we assume that the Italian patients were studied more recently, benefiting from the more vigorous treatment schedules introduced in the last decade. Therapeutic control was insufficient in many of our patients observed earlier, as demonstrated by decreased blood levels of potassium, phosphate, and bicarbonate in 31%–50% of laboratory tests (Table 3 of our paper). Conversely, the patients of Greco et al. only occasionally had low phosphate and bicarbonate levels, and none required ongoing potassium supplementation. This agrees with the observation that in our 2 patients who reached the tallest height at last observation, phosphate, potassium, and bicarbonate levels were rarely subnormal.

It seems that the primary reason for the marked stunting in our patients compared with the Italian children with FS is the more severe impairment of renal tubular function. This made it difficult to maintain an optimal electrolyte and acid-base balance and would explain the growth retardation more marked at the time of diagnosis in our population [mean height standard deviation score (SDS) –4.0 vs. –2.4]. Our extensive literature search (cited on page 43 of our paper) confirmed that growth retardation is already pronounced in most cases at the time of diagnosis. Figure 1 demonstrates that at the time of reporting and/or at last observation only 3 of 20 boys and girls with FS reported in the literature had a height above the 3rd percentile. The height percentile improved in only 2 of 8 patients followed longitudinally. The slightly younger age of the Italian patients at the start of treatment (<2 years: ¾ vs. ⅜ in our series) might have contributed to the difference in height SDS at the last observation.

Among the many factors contributing to growth failure in FS, extrarenal manifestations of the disease (gastrointestinal, neurological) were detected in 6 of our 9 patients, but were apparently absent in the patients of Greco et al. We cannot comment on the suggestion of the Italian authors that a non-steroidal antiinflammatory medication (ibuprofen) is the main explanation for better growth in their children with FS, because we did not use this drug.

We believe that in any chronic renal disease an adequate account of body growth can only be expected by following the patients from the apparent onset of disease until attainment of adult height, including the pubertal period.

We take this occasion to draw attention to a printing error in Table 1 of our paper: The superscript "a" under "height at last observation" in patient no. 4 should be replaced by "e".

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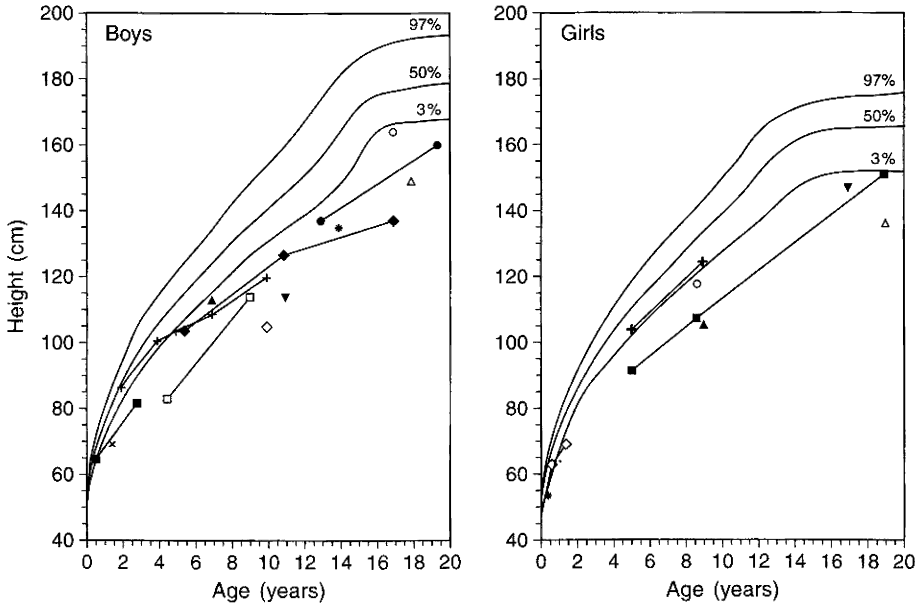


**Fig. 1.** Height data of 12 boys and 8 girls with primary de Toni-Debré-Fanconi syndrome reported in the literature. Each patient is represented by a symbol. Eight patients were followed longitudinally. Normal percentile curves are according to Prader et al. (1998) (Helv Paediatr Acta [Suppl] 52: 1–125)