Dear Sir,

The term ‘cone-shaped epiphyses’ (CSE) refers to the secondary centers of ossification in the hands and feet which have the shape of a cone indenting the metaphyses. These CSE can be identified radiographically in the hands and feet and hence they are considered as a subgroup in a large group of peripheral epiphyseal dysostosis. They might be found as a normal variant in healthy children [1], however there are several diseases in which those CSE were described, among them cleidocranial dysostosis, chondroectodermal dysplasia, tricho-rhi-no-phalangeal syndrome, osteopetrosis and others [2]. Some authors revealed the association between CSE and chronic renal disease in children (with or without retinitis pigmentosa) [3]. This association has been termed by Giedion as the ‘cono-renal syndrome’ [4]. In most cases the histology of the kidney biopsy reveals nephronophthisis [4].

Recently a 12-year-old girl, of Dnie origin, was admitted to our hospital due to weight loss and severe anemia. A family history revealed 2 older brothers who died because of ‘renal disease’. One of them died about 5 years previously, being 9 years old, in another hospital after several months of dialysis. No records were available from the other hospital. On admission the girl was found to be severely anemic and severely azotemic. The blood tests gave the following results: Hgb 4.0 g/dl, Hct 14.7%, urea 198 mg%, uric acid 8.8 mg%, creatinine 9 mg%, Na 135 mEq/1, K 4.2 mEq/1, Ca 7.9 mg%, P 6.2 mg%, pH 7.07. Urine specific gravity was 1.006. Her blood pressure on admission was 180/110 mm Hg. On physical examination there were no findings consistent with cerebellar ataxia. Eye fundus examination was normal. A kidney biopsy was not done.

The girl was sent for an X-ray of her hands to look for signs of secondary hyperparathyroidism. The films showed several cone-shaped epiphyses (fig. 1, 2). An X-ray of her feet was performed as well and cone-
Fig. 3. CSE were demonstrated at the base of all phalanges of the toes excluding those of the 1st toe. They are most prominent at the base of the proximal phalanges.

Fig. 2. Close-up of index finger of right hand. Typical cone-shaped epiphysis is noted at the base of middle phalanx. The obliquity of the distal middle phalanx causes ulnar deviation of the inter-phalangeal joint. This finding is considered as a hallmark in peripheral dysostosis [2].

Fig. 4. A longitudinal sonographic image of the right kidney (between arrows). The kidney is relatively small and its cortex is echo-genic. These findings are consistent with chronic renal disease. A cortical cyst (C) is identified in the upper pole.

References

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