BONE METABOLISM IN CHILDREN WITH AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE (ADPKD)

S. De Rechter[1], J. Bacchetta[2], L. Dubourg[2], P. Cochat[2], M. Van Dyck[1], P. Evenepoel[3], J. De Schepper [4], E. Levtchenko[1], D. Mekahli[1]

[1]Department of Pediatric Nephrology, University Hospital of Leuven, Leuven, Belgium.
[2]Centre de Référence des Maladies Rénales Rares, Hôpital Femme Mère Enfant, Hospices Civils de Lyon, Bron, France.
[3] Department of Internal Medicine, Division of Nephrology, University Hospital of Leuven, Leuven, Belgium.
[4] Department of Pediatric Endocrinology, University Hospital of Brussels, Brussels, Belgium.

Background

Autosomal dominant polycystic kidney disease (ADPKD) is the most common monogenic cause of nephropathy.
It is a systemic disorder associated with extra-renal symptoms such as polycystic liver disease or arterial aneurysms. Clinical data have revealed in a ADPKD adult population that Fibroblast Growth Factor 23 (FGF23) increases while circulating Klotho levels decrease, with a low TmP/GFR even when renal function is normal (Pavik et al, 2012). The mechanisms governing this FGF23 increase are largely unknown. However, it has been demonstrated recently in animal models that the polycystic kidney produces FGF23 but is resistant to its actions (Spichtig et al, 2014).
No data are available in a pediatric population. To fill this gap, we assessed bone metabolism and renal phosphate handling in children with ADPKD in a dual-center study (Leuven – Lyon).

Methods

Children with ADPKD were eligible for inclusion. Blood and urine samples were collected and analysed for parameters of bone and mineral metabolism. Based on normal values according to age, we made percentile (P) charts for phosphatemia and TmP/GFR. We defined hypophosphatemia and low TmP/GFR as values ≤ P5.

Results

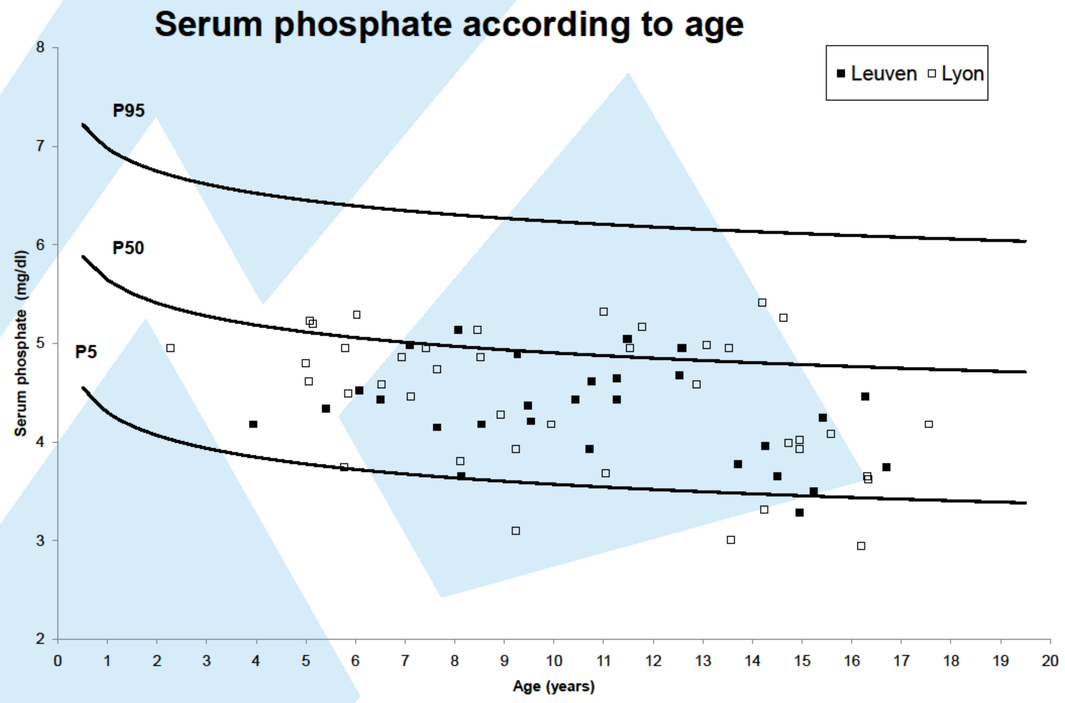
69 patients were included, patient characteristics are shown in table 1. Hypophosphatemia was found in 10% of the children. Low TmP/GFR was observed in 22% of the children. For both, a clear trend towards values within a low-normal range are observed. Parameters of mineral metabolism in the Leuven cohort were as follows: mean serum alkaline phosphatase 341,9 236,7 (U/L); serum PTH 17,1 12,2 (ng/L); serum 25 OH vitamin D 22,4 9,4 (µg/L) and 1,25 (OH)2 vitamin D 74,5 13,9 (ng/L). These values are within normal range except for 3 patients with a 25 OH vitamin D deficiency (<11 µg/L).

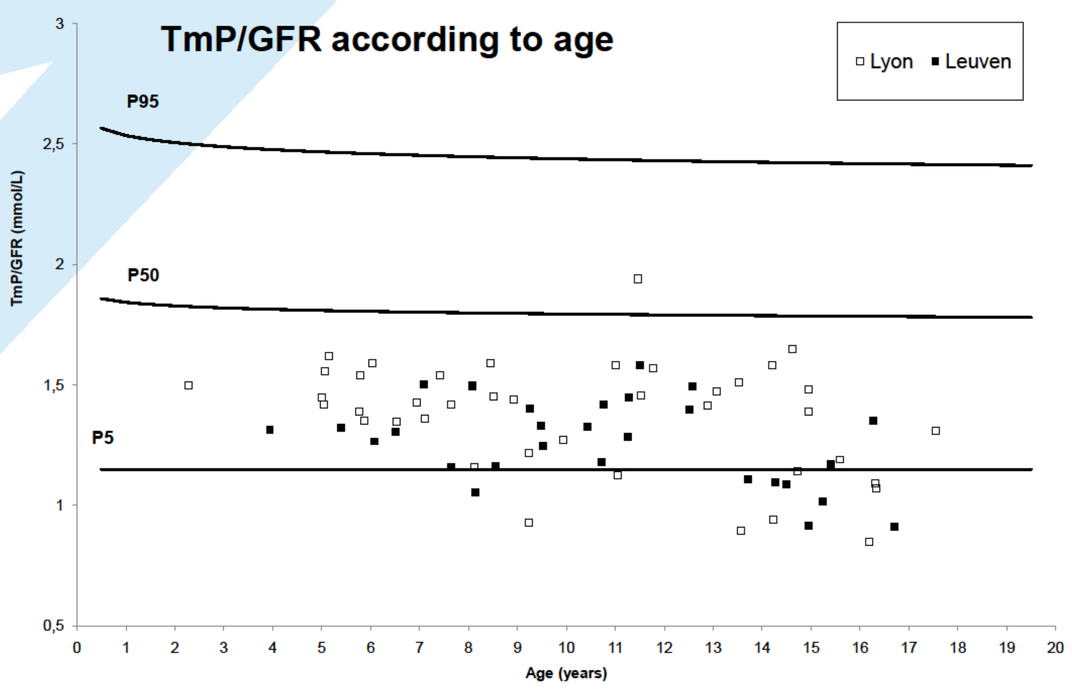
Conclusion

This is the first report highlighting hypophosphatemia in combination with phosphate leak in ADPKD children with normal eGFR. As PTH levels were in the normal range, these data point to an abnormal FGF23 metabolism and as such confirm recent data in adult ADPKD patients. Further studies are required to elucidate the underlying pathophysiology and to investigate potential clinical consequences.

	cohort Leuven	cohort Lyon	Total
Number of patients	28	41	69
Male/Female	20/8	22/19	42/27
Age (y)	11,3 (3,9-16,7)	9,9 (2,3-17,5)	10,7 (2,3-17,5)
Weight (kg)	40,5 ± 16,9	37,9 ± 15,4	39 ± 15,9
Height (cm)	146,4 ± 22	143,8 ± 24,2	144,9 ± 23,2
BMI (kg/m²)	18,1 ± 2,7	17,4 ± 2,5	17,7 ± 2,6
eGFR (ml/min/1,73 m²)	125,1 ± 18,0	119,2 ± 21,0	121,6 ± 19,9
Nephromegaly (unilateral)	9/28	NA	
Nephromegaly (bilateral)	13/28	NA	

Table 1: Patient characteristics (values in mean standard deviation or median (min – max) NA= not available)





stephanie.derechter@uzleuven.be

UZ Leuven | Herestraat 49 B - 3000 Leuven | www.uzleuven.be tel. 0032 (0)16 33 22 11

UNIVERSITY HOSPITALS LEUVEN

51ERA-EDTA 19--SP

C) Renal development and cystic diseases.

Stéphanie De Rechter

DOI: 10.3252/pso.eu.51era.2014

Poster presented at: 51 ERA-EDTA CONGRESS AMSTERDAM THE NETHERLANDS MARCH JUNE 2014

PosterSessionOnline.com