




## Theme: Pediatric Nephrology

 **Kidney disease progression in autosomal recessive polycystic kidney disease** (*J Pediatr.* 2016 Jan 28. pii: S0022-3476(15)01669-8. doi:10.1016/j.jpeds.2015.12.079.)

This study aimed to define glomerular filtration rate (GFR) decline, hypertension, and proteinuria in a well-defined autosomal recessive polycystic kidney disease (ARPKD) cohort and compared with two congenital kidney disease control groups (hypoplastic/dysplastic disorders and obstructive uropathies). The overall rates of GFR decline did not differ significantly in subjects with ARPKD vs controls. There were no significant differences in rates of hypertension or left ventricular hypertrophy, but subjects with ARPKD had a greater percent on  $\geq 3$  blood pressure medications, more angiotensin-converting enzyme inhibitor use, and less proteinuria. The authors conclude that the relatively slow rate of GFR decline in subjects with ARPKD and absence of significant proteinuria suggest that these standard clinical measures may have limited utility in assessing therapeutic interventions, and highlight the need for other ARPKD kidney disease progression biomarkers.


 **Metabolic risk factors in children with asymptomatic hematuria** (*Pediatr Nephrol.* 2016 Feb 25. [Epub ahead of print])

The aim of this study was to identify possible urinary risk factors for hematuria in children. The authors retrospectively evaluated clinical onset, family history, and metabolic risk factors of 60 children with idiopathic hematuria but without renal stones or other pathologic conditions that could explain the hematuria. A family history of stone disease was found in 63% of the children. At least one urinary metabolic abnormality was present in 49 patients, while 11 patients had no metabolic abnormality. The most common urinary risk factor was idiopathic hypercalciuria (single or associated) 43.5% of patients, followed by hypocitraturia (single or associated) in 31.7%. The authors also found hyperoxaluria and, less frequently, hypomagnesuria and hyperuricosuria. The authors concluded that asymptomatic idiopathic hematuria in pediatric patients may often be linked to different urinary biochemical abnormalities, similar to what is observed in pediatric kidney stone-formers.


 **Association of hypercalciuria and hyperuricosuria with vesicoureteral reflux in children.** (*Clin Exp Nephrol.* 2016 Jan 28. [Epub ahead of print])

This study was conducted to determine the association between hypercalciuria and hyperuricosuria with vesicoureteral reflux (VUR) in children. One-hundred children with VUR were compared to 100 healthy children in terms of hypercalciuria and hyperuricosuria. Hypercalciuria and hyperuricosuria frequencies, and also urine calcium/creatinine and urine uric

acid/creatinine ratios, were significantly higher in the case group compared to the control group. A positive correlation was observed between hypercalciuria and hyperuricosuria and severity of VUR. The present study showed that there is association between hypercalciuria, hyperuricosuria and VUR in children. The authors recommend adopting measures to prevent the development of urolithiasis in VUR patients.

 **Calculation of prednisolone dose in nephrotic syndrome** (*Pediatr Nephrol.* 2016 Jan 12. [Epub ahead of print])

Body surface area (BSA)-based prednisolone dosing for childhood nephrotic syndrome (NS) leads to higher cumulative prednisolone doses than body weight (BW)-based dosing. Since the clinical effects of this higher dosage have not been evaluated in prospective studies, this parallel-group open-label randomized clinical trial enrolled 100 children with idiopathic NS, to receive BW-based or BSA-based prednisolone dosing by block randomization in a 1:1 ratio. There was no significant difference in the time taken for remission in the BW group versus the BSA group; similar results were observed on subgroup analysis in new-onset and infrequently-relapsing NS. The incidence of hypertension was higher in the BSA group on per-protocol analysis. The relapse rates in the two groups per 6 months on follow-up were comparable. The study concludes that clinical outcomes with BW-based dosing are equivalent to BSA dosing-related outcomes, although cumulative prednisolone doses are lower in the former. The practice of BW-based calculations for prescribing prednisolone in NS seems to be a reasonable approach.

 **Systemic corticosteroids and immune system in nephrotic syndrome** (*Eur J Pediatr.* 2016 Jan 30. [Epub ahead of print])

Nephrotic syndrome is known to be related with complex immune disturbance including T and B cells dysfunctions. Steroids induce neutrophilic leukocytosis concomitant with lymphopenia and eosinopenia, leading to immunosuppression. This study aimed to investigate the effects of corticosteroids on the various compartments of immune system in relation to timing of initiation and persistence of therapy. Pediatric patients with idiopathic nephrotic syndrome treated with 2 mg/kg/day prednisolone and healthy controls were enrolled. The study showed that T cell subsets and proliferation are susceptible to corticosteroids more than B cells; however, the reversibility is faster with dose reduction in corticosteroids. The change of B cells and B cell subtypes (CD27<sup>+</sup> memory) shows prolonged effect of corticosteroids on B cells which may alter antibody production even after three months of cessation of corticosteroids.

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