IPN12111-77 PEDİATRİC UROLİTHİASİS AND NEPHROCALCİNOSİS İN TURKEY; AN ANALYSİS OF 1400 CASES

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Objective: We evaluated the demographic features, etiologic risk factors, treatment managements and outcome of children with urolithiasis and nephrocalcinosis.

Methods: A retrospective multicenter study was conducted in eighteen Pediatric Nephrology centers in Turkey. Medical records of 1404 children followed up at least one year with urolithiasis (1299) and nephrocalcinosis (105) were reviewed.

Results: There were 763 boys and 641 girls (1.2/1). The median age at diagnosis was 10 months (range 0.5-216 months), 794 patients (56%) were in first year of life. Consanguinity of patients with urolithiasis and nephrocalcinosis were 26% and 46% respectively. In patients with nephrocalcinosis history of hospitalization in the neonatal period and growth retardation were higher than urolithiasis. Most cases with infantile urolithiasis and nephrocalcinosis were diagnosed incidentally. Microlithiasis (≤3mm) were found in 638 patients (45.4%), 70% of patients with microlithiasis were infants.

Stones were in the pelvis-calyces in 71.4% and lower pole in %45 of patients. The most common stone type was calcium oxalate (67.3%). Hypercalciuria was the most common metabolic risk factor in patients older than 12 months, but in infancy hyperuricosuria was the leading one. Sixty percent of the patients received medical treatment, mostly potassium citrate.

At the end of one year; majority of patients with microlithiasis (92%) showed spontaneous remission. Spontaneous remission was higher in patients without metabolic risk factors. Lower pole stone remission rate was lower than upper pole stone (lower pole 12%, upper pole %82). Resolution was showed in 27% of patients with nephrocalcinosis.

Conclusion: Because of the high rate of spontaneous remission in patients with microlithiasis and stones without metabolic risk factors, these patients should be followed without medical treatment. Patients with lower pole stone should be treated due to low remission rate.

IPN12112-78 A GLOBAL ANTI B-CELL STRATEGY WITH OBINUTUZUMAB AND DARATUNUMAB IN SEVERE PEDIATRIC NEPHROTIC SYNDROME

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Introduction: The efficacy of B-cell depletion and Immunoglobulin adsorption in the treatment of patients with SDNS and SRNS supports the involvement of B cells in the physiopathology of INS. However, rituximab (RTX) targets only CD20 B-cells but not plasma cells. Furthermore, RTX mediated B-cell depletion may paradoxically induce the settlement of autoreactive long-lived plasma cells which may account for some RTX failure. In this pilot study, we investigate in patients with severe SDNS the association of Obinutuzumab (OBZ), a 2nd generation anti CD20 antibody, with Daratumumab (DAR), an anti CD38 antibody with high plasma cell cytotoxicity and an immunomodulatory activity.

Materials and methods: Patients received an infusion of 1000mg/1,73m² of OBZ at D0 and 1000mg/1,73m² of DAR at D15. Oral immunosuppressors were discontinued within two months, and biological monitoring was performed monthly until B cell recovery.

Results: 9 patients with SDNS were included, after resistance to rituximab (n=3) or early relapse after rituximab (n=6). Median ages at INS onset, first RTX and OBZ were 2.9, 7.7 and 10.9 years old, respectively. B cell depletion was achieved in all patients. Median follow-up was 10 months (IQ 8.3-10.3), and all patients remained relapse-free. Six patients had still undetectable peripheral B-cells, B-cells reconstitution occurred at 7.9, 8.1 and 9.3 months in the 3 others. Mild infusion reactions were reported in 2/9 patient during OBZ and 3/9 during DAR infusions. Mild neutropenia (500-1000/mm³) occurred in 2/9 patients. 7/9 patients received IV immunoglobulins because of hypo-IgG. Hypo-IgA was noted in 8 patients and hypo-IgM in all patients. No severe infection was reported.

Conclusion: Global anti-B cell strategy with obinutuzumab and daratumumab induces prolonged peripheral B-cell depletion and INS remission in children with severe SDNS. However, it induces hypogammaglobulinemia and further investigation of the safety and the long-term efficacy of this strategy is needed.

IPN12113-79 BRAIN NETWORK CONNECTIVITY IN CHILDREN AND YOUNG ADULTS WITH SEVERE CHRONIC KIDNEY DISEASE

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Introduction: Children, adolescents and young adults (CAYAs) with severe chronic kidney disease (CKD4+) are at risk of neurocognitive impairments, which may impact psychosocial development, adaptive functioning and