ultrasound scan, uroflow, urinalysis and culture, urine Ca/creatinine, and first-morning urine osmolality. Patients <5 years of age, with secondary enuresis, and those who did not show at the follow-up visit were excluded.

Oral desmopressin lyophilisate was recommended to all patients with PMNE and normal bladder capacity. After one month of therapy, initial success was assessed according to ICCS. Correlation coefficients were used to identify variables that were significantly correlated to complete response. ROC analysis was used to determine the urine osmolality cut-off value. Odds ratio and correlation coefficients in favor of complete initial success were analyzed with binary logistic regression.

There were 48 patients with PMNE who received desmopressin and were followed for treatment success. Of tested variables, only lower urine osmolality was found to be significantly in favor of complete response to desmopressin therapy. ROC analysis determined the value of ≤ 814 mOsm/L as a cut-off value for complete success (sensitivity 65% and specificity 75%). The odds ratio for complete success with desmopressin therapy in PMNE patients with first-morning urine osmolality ≤ 814 mOsm/L was 9.086 (95% CI 1.893 – 43.618, $P = 0.006$).

For PMNE patients with high pretreatment morning urine osmolality, an alternative treatment to desmopressin should be considered because of the significantly higher risk of treatment failure.

362 3D PRINTED PEEK EXTRAVASCULAR STENT IN TREATMENT OF NUTCRACKER SYNDROME IN AN ADOLESCENT BOY

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A 14-year-old boy with constant and intense periumbilical pain for the past 24 months caused by nutcracker syndrome (NCS) (visual analog pain scale, 8-9) was treated with 3D printed PEEK extravascular stent. To our best knowledge this is the first European application in a child. 3D model reconstruction showed that the left renal vein (LRV) and the duodenum were contracted at almost the same level via aortomesenteric angle, resulting in LRV compression from the abnormal high-level duodenal compartment. Surgical procedure included a dissection of fibrous bundles which freed the duodenum. LRV was dissected out medially from the renal hilum to the vena cava inferior until it was completely mobilized. A complete resection off the narrow fibrous ring between abdominal aorta and superior mesenteric artery was necessary. 3D printed PEEK extravascular stent via camera port was placed and bound to the narrowed portion of LRV to prevent blood vessel compression. Such stent demonstrates significant advantages over open or other minimally invasive surgery such as endovascular stenting and artificial vessels procedures with expanded polytetrafluoroethylene. It has distinctive personalized design and good rigidity as well as durability which allow vascular growth, preventing stent migration and thrombosis as well. Therefore, it is suitable for both adult and pediatric patients.

363 TWO BOYS WITH C3/DDD GLOMERULONEPHRITIS

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A boy, 16 years of age. During routine analysis hematuria and proteinuria (up to 1.84 g/L) were found along with hypertension (max 160/70 mmHg). Physical examination showed no abnormalities. Total complement activity (classical pathway) of 7 CH50/ml (ref. 48-103 CH50/ml) and total complement activity (alternative pathway) of 0% (ref. 70-105%).

C3 levels were low (median 0.06 g/L) while C4 were within reference ranges.

Anti-C1q IgG autoantibody was 677 U/ml (ref. <52), C3-nephritic factor 11,0% (ref. <10%) and sC5b-9 (terminal complement complex) 1640 ng/mL (ref. 110-252 ng/mL). Complement factor I antigen was decreased 51% (ref. 70-130%) as well as complement factor B antigen of 45% (ref. 70-130%).

These results support the presence of AP dysregulation with overactivation.

Lupus aDNA negative Kidney biopsy revealed C3 glomerulonephritis. Due to nephritic syndrome the boy was treated with pulse methylprednisolone therapy, rituximab and cyclophosphamide. Until present, proteinuria was reduced (up to 0.48 g/L) with microhematuria. Due to a persistently low C3 and elevated sC5b-9, we are now considering Eculisumab as off label treatment option. Genetic analysis ongoing.

Second boy, 5 years of age, was treated for nephritic syndrome in regional hospital. Physical examination showed no abnormalities. As significant proteinuria (3.62g/L) with low C3 was found. A kidney biopsy revealed Dense deposit disease. Total complement activity (classical pathway) 18 CH50/ml (ref. 48-103 CH50/ml) and total complement activity (alternative pathway) 0% (ref. 70-105%). C3 and C4 levels were low (0.1 and 0.06 g/L respectively). C3-nephritic factor was elevated 41,3% (ref. <10%) as well as sC5b-9 (terminal complement complex) of 342 ng/mL (ref. 110-252 ng/mL).

LDH was elevated of 552 U/L. Creatinine level was elevated at its highest of 198 μ mol/L. Activity of the alternative and classical pathways were both deficient. Complement C3 and C4 levels are decreased, while those of factor I and B are in the normal range. The terminal pathway activity marker level is increased. The above results support a complement activation and consumption involving at least the classical pathway. The boy was treated with pulse methylprednisolone therapy, rituximab and cyclophosphamide. After initial therapy elevated serum creatinine levels decreased, as well as proteinuria. C3 stabilized around 0.50, C4 normalizes at 0.40, while proteinuria decreased to 0.9 g/L.

364 URINARY INCONTINENCE

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Functional and morphological anomalies could cause urinary incontinence. The essence of urinary incontinence and