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ABSTRACTS

Abstracts of the 50th Anniversary ESPN Meeting, Glasgow, September 2017

O-01 ORAL ANTIBIOTIC TREATMENT FOR PYELONEPHRITIS IS SAFE AND EFFECTIVE IF THERE IS A FORMALISED CONTACT TO THE DEPARTMENT OF PAEDIATRICS

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Introduction: We investigated the safety and efficacy of out-patient, oral antibiotic treatment of pyelonephritis in Danish children.
Material and methods: Study population: children >6 months of age with culture positive pyelonephritis admitted to 11 Paediatric Departments, from October 2014 through December 2015. Septic children and those with known urological anomalies were excluded. The patients were treated 10 days with oral antibiotics (amoxicillin-clavulanate or piv-mecillinam). Our protocol included a nurse telephone consultation with the family on day 2 of treatment, and a clinical re-examination of the patient on day 3. The patients all underwent renal ultrasound and in some centers a renal scintigraphy.
Results: 446 children, 388 (87%) females, median age 27.2 months (range 0.6–16.3 years), 361 (81%) of the population had their first episode of pyelonephritis. Urine was obtained by suprapubic puncture in 52 (26% of those <2 years), by clean catch urine in 384 (71% had 2 cultures done) and by clean catheterization in 10 girls. *Escherichia coli* were identified in 410 (92%) patients. The vast majority of patients, 407 (91%) were effectively treated by oral antibiotics, while 39 (9%) were switched to parental antibiotics, causes listed were: lack of clinical improvement (n = 18), vomiting (n = 7), resistance to antibiotics (n = 7) or other problems (n = 7). Renal ultrasound was abnormal in 7% (30/433), and renal scintigraphy was abnormal in 13% (36/283). Data analysis comparing "failure of oral antibiotic treatment" with those who succeeded revealed no significant predictor of treatment failure with oral antibiotics: age (p = 0.826), C-reactive protein (p = 0.064), frequency of girls (p = 0.814), *Escherichia coli* (p = 0.776), first pyelonephritis (p = 0.929), abnormal ultrasound or abnormal scintigraphy (p = 0.599, p = 0.918), respectively.
Conclusions: Treatment of pyelonephritis with oral piv-mecillinam or amoxicillin-clavulanate at home is safe and effective, but formalized contact to the Paediatric Departments is required as 9% of patients need a shift to intravenous antibiotic treatment.

O-02 ROLE OF FETAL MRI IN ANTENATALLY DETECTED RENAL ANOMALIES

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Introduction: Fetal MRI begain to be used as an imaging modality in 1983 and as part of research in Sheffield Teaching Hospitals in 1998. Most papers suggest that fetal MRI is a valuable adjunct or complementatry tool to ultrasound imaging. This is the first known research study from the UK and the only five years duration retrospective review in literature assessing the impact of fetal MRI on antenatally suspected renal anomalies.
Material and methods: This retrospective review was based at the Fetal MRI Department at Royal Hallamshire Hospital, part of the Sheffield Teaching Hospitals NHS Foundation Trust. This study included all referrals for fetal MRI from October 2011 to September 2015. Of the 578 referrals identified, 80 were due to renal concerns. The results of the antenatal USG imaging and fetal MRI were compared with that of the postnatal USG imaging which was considered to be the gold standard in this study.
Results: Out of the eighty referrals, there were four terminations, three stillbirths, nine neonatal deaths and of the rest of the 64 referrals, outcome data was available for 27 pregnancies. Comparable results between antenatal USG imaging and fetal MRI was found in 26% of the pregnancies. Fetal MRI changed the diagnoses in 31.25% and in another 42.5% provided additional information. In 12.5%, the additional information provided could potentially have impacted on the outcome of these pregnancies and of these, 90% of the additional information were about non-renal related systems. The positive predicitive value for fetal MRI was 88.9% as compared to 74% for antenatal USG imaging.
Conclusions: Fetal MRI has therefore made an impact on 44% of the pregnancies that underwent fetal MRI due to renal concerns in the antenatal ultrasound imaging.

O-03 VALUE OF RENAL AND BLADDER ULTRASOUND IN DIAGNOSING VESICOURETRAL REFLUX

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Introduction: Continuous antibiotic prophylaxis reduces the incidence of recurrent pyelonephritis in children with vesicouretral reflux (VUR). However, there is an ongoing debate on which children should be

screened for VUR after a first episode of pyelonephritis. More concrete, the value of renal and bladder ultrasound (RBUS) in diagnosing VUR has been challenged. In this abstract, we present our preliminary findings of a database study in children younger than 4 years of age.

Material and methods: Retrospective analysis of patients younger than 4 years who were followed-up at our department, after a first episode of pyelonephritis, between 2009 and 2015. Patients were included if clinical information concerning index pyelonephritis, and paired RBUS and voiding cystourethrogram (VCUG) were available. Patients were excluded if a VCUG had been performed preceding index pyelonephritis. Descriptive statistics and logistic regression models were used for examining risk factors for VUR, SPSS version 22 was used for performing all analyses.

Results: Data of 399 children were available for analysis. *Escherichia coli* was cultured in 345 cases (86.4%). Hydroureters and duplex ureters were found in 25 renal units (3.1%), any grade of VUR in 179 units (22.4%), dilating VUR in 74 units (9.2%). In our first logistic regression analysis, girls, index infections with non-*E. coli* pathogens, and duplex ureters were risk factors for any grade of VUR. In general, ultrasound parameters were not significantly predictive for VUR. In VUR cases that were missed by RBUS (124), bladder was empty at RBUS in 76 children (73.1%).

Conclusions: Girls, and children suffering pyelonephritis by *non-E.coli* uropathogens are at risk for VUR in children less than 4 years. RBUS is of limited value in diagnosing VUR.

O-04 FIRST-YEAR PROFILE OF BIOMARKERS FOR EARLY DETECTION OF RENAL INJURY IN HEALTHY INFANTS

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Introduction: To analyze first-year profiles of two serum renal biomarkers: creatinine and cystatin C; and five urinary renal biomarkers: Neutrophil gelatinase-associated lipocalin (NGAL), Kidney Injury Molecule-1 (KIM-1), Transforming growth factor beta 1 (TGF-β1), Retinol-binding protein (RBP) and Cystatin C (CyC) in a cohort of healthy infants.

Material and methods: In this longitudinal, prospective and pilot study 24 healthy infants were followed from birth up to one year of age, in a quaternary healthcare institution, by nephrology pediatrics, neonatology and obstetrics team. All of the subjects were free of gestational or delivery complications with normal glomerular and tubular function parameters and normal renal ultrasound findings. The blood samples were collected from the umbilical cord at birth and subsequently, between 3th and 7th day, at 6th and 12th months of age. The urine samples were obtained in the period between 3th- 7th day, at 1st, 2nd, 3rd, 6th, 9th and 12th month of age. Anthropometric parameters and blood pressure were obtained during follow-up. Serum and urine samples were stored at −70 °C, and thereafter analyzed by quantitative enzymatic immunoassay (ELISA).

Results: The cohort is characterized by homogeneous distribution of sex (58.3% boys, 47.1% girls), race and gestational age. 24/45 (53.3%) newborns completed the follow-up, in period from 2011 until 2016. Demographic distributions, main renal function parameters and mean values of renal biomarkers during the first year of life are presented in Table 1.

Table 1: Demographic characteristics, main renal function parameters and Mean values of renal biomarkers during the first year of life in healthy infants.

Cohort Characteristics	**Mean/SD**	
Gestational age (weeks)	38.4 ± 1.4	
Birth weight (grams)	3105 ± 401.5	
Weight at 12 months – boys (grams)	9820 ± 1005.8	
Weight at 12 months – girls (grams)	9433.5 ± 1144.2	
Stature at 12 months – boys (cm)	76 ± 1.8	
Stature at 12 months – girls (cm)	74.9 ± 3.8	
Systolic blood pressure (mmHg)	88.3 ± 1.08	
Diastolic blood pressure (mmHg)	53.4 ± 2.99	
Biomarker	Mean / SD	*p* value
Albuminuria (mg/g urine creatinine)	23.06 ± 3.77	$p = 0.33^{(1)}$
GFR by creatinine (ml/min/1.73m^2)		$p = 0.068^{(2)}$
Neonatal period (3rd to 7th day of life)	69.87 ± 32.33	
6 months	142.21 ± 23.64	
12 months	135.75 ± 35.15	
GFR by cystatin C (ml/min/1.73m^2)		$p = 0.105^{(2)}$
Neonatal period (3rd to 7th day of life)	71.66 ± 24.55	
6 months	97.64 ± 26.49	
12 months	96.7 ± 19.49	
Serum Creatinine (mg/dl)		$p = 0.21^{(2)}$
Umbilical cord	0.62 ± 0.20	
Neonatal period (3rd to 7th day of life)	0.38 ± 0.17	
6 months	0.22 ± 0.04	
12 months	0.27 ± 0.07	
Serum Cystatin C (μg/dl)		$p = 0.47^{(2)}$
Umbilical cord	2.55 ± 0.75	
Neonatal period (3rd to 7th day of life)	1.65 ± 0.55	
6 months	1.20 ± 0.57	
12 months	1.13 ± 0.25	
RBPu (normalized by urine creatinine, in ng/g)		$p = 0.034^{(2)}$
Neonatal period (3rd to 7th day of life)	23.79 ± 67.27	
1st month	1.77 ± 1.66	
2nd month	1.18 ± 1.37	
3rd month	0.74 ± 0.56	
6th month	1.12 ± 1.28	
9th month	2.12 ± 3.11	
12th month	2.48 ± 6.13	
NGALu (normalized by urine creatinine, in ng/g)	1.09 ± 1.51	$p = 0.43^{(2)}$
CyCu (normalized by urine creatinine, in ng/g)		$p = 0.023^{(2)}$
Neonatal period (3rd to 7th day of life)	61.51 ± 58.99	
1st month	10.73 ± 9.34	
2nd month	8.22 ± 5.77	
3rd month	10.87 ± 10.64	
6th month	13.78 ± 14.26	
9th month	22.15 ± 24.86	
12th month	17.74 ± 18.68	
TGFβ1u (normalized by urine creatinine, in pg/g)	1.5 ± 1.5	$p = 0.06^{(2)}$
KIM-1u (normalized by urine creatinine, in ng/g)	2.01 ± 1.8	$p = 0.12^{(2)}$

GFR – Glomerular filtration rate; (1) t-Student Test. (2) Test F (ANOVA) by Tukey's comparisons, statistical significance: $p < 0.05$; RBP – Retinol-Binding Protein; NGALu – Urine Neutrophil Gelatinase-Associated Lipocalin, CyCu – Urine Cystatin C, TGFβ1u – Urine Transforming Growth Factor Beta 1 (TGF-β1); KIM-1u – Urine Kidney Injury Molecule-1;

Conclusions: In healthy infants, the evolution of biomarkers values during the first year of life represents, as expected, renal maturation. KIM-1, TGFβ-1 and NGAL maintained low values throughout the observation period confirming absence of renal impairment. Further studies with more participants are necessary in order to validate these biomarkers for their future use in early renal impairment detection.

O-05 LONG-TERM LIFESTYLE INTERVENTION IMPROVES RENAL FUNCTION IN OBESE CHILDREN AND ADOLESCENTS

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Introduction: Weight loss has positive effects on renal function in adults with obesity. In children, the effect of weight loss on renal function is unknown. The aim of this prospective study was to examine the effect of a lifestyle intervention on renal function in children and adolescents with overweight, obesity and morbid obesity.
Material and methods: 245 children and adolescents (mean age 12.30 (2.61–18.69) years, 40.4% boys, BMI z-score 3.46 ± 0.70) participating in an out-patient lifestyle intervention at the Centre for Overweight Adolescent and Children's Healthcare (COACH) at the Maastricht University Medical Centre were included in the study. Participants with at least 12 months of follow-up (n = 144 (58.8%)) were included in the follow-up study. Mean follow-up period was 15.2 ± 3.9 months. All participants were subjected to detailed history taking, physical examination, anthropometric measurements, blood analysis and blood pressure measurements. Glomerular filtration rate (GFR) was estimated by means of the Schwartz formula.
Results: At baseline, mean estimated glomerular filtration rate (eGFR) was 116.8 (66.1–189.0) ml/min/1,73m^2. 20.8% of the participants demonstrated glomerular hyperfiltration and 2.9% microalbuminuria. After intervention, eGFR decreased significantly from 119.6 to 111.6 ml/min/1.73m^2 in the total group of participants. eGFR normalized in 61% of the participants with glomerular hyperfiltration at baseline. eGFR improvement was positively associated with improvement in diastolic blood pressure, fasting glucose and glycated haemoglobin (HbA1c), and was not associated with changes in BMI z-score. No difference in pre- and post-intervention urine albumin-to-creatinine ratio was found.
Conclusions: After one year of multidisciplinary lifestyle intervention, there is a significant improvement in renal function in children and adolescents with obesity and morbid obesity, which is associated with improvement of cardiovascular risk parameters.

O-06 UTILITY OF PERFORMING VOIDING CYSTOURETHROGRAM (VCUG) WITH ANTIBIOTIC - AN OPEN LABELLED RANDOMIZED CONTROL TRIAL.

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Introduction: Voiding cystourethrogram (VCUG) remains the gold standard for demonstrating the grade of reflux as well as urethral anatomy. Procedure related urinary tract infection (UTI) remains an important concern. There is controversy regarding utility of antibiotic in preventing post VCUG acquired UTI.
Material and methods: The objective of the study was to assess whether antibiotic reduces VCUG acquired UTI. An open labelled randomized controlled trial (RCT) was conducted at a tertiary paediatric nephrology centre. One hundred twenty children (age 2 month to 5 years) undergoing VCUG were randomized into Group A (Antibiotic), or Group B (no antibiotic) in 3:2 ratio. Group A received oral antibiotic (cephalexin if <6 month or cotrimoxazole if >6 month old) a day prior VCUG and continued till a day post VCUG. The primary outcome of interest was incidence of VCUG associated UTI. Urine was checked on day 3 after VCUG and UTI was defined as significant growth of a single organism in a symptomatic child.
Results: Median age of the study population was 8 months (IQR 13 months) with 68% male. Indication for undertaking VCUG was history of UTI (1st UTI in infancy = 43, recurrent UTI = 49) and congenital anomaly of kidney and urinary tract (CAKUT) without any UTI (n = 28). Post VCUG UTI was significantly higher among Group B in comparison to Group A [17% vs 1.4% p = 0.01, OR = 14.2 (95% C.I. = 1.7 to 117)]. Multivariate binary logistic regression analysis found an abnormal pre-VCUG ultrasound scan to be significant independent risk factors for post VCUG UTI (p = 0.02; OR 9.51–95% C.I. 1.43 to 63.4). Number needed to treat with antibiotic to prevent one post VCUG UTI was 6.5, which reduced to 4 if only the group with abnormal pre-VCUG USS were included.
Conclusions: Antibiotic significantly reduces post VCUG acquired UTI especially in those with abnormal ultrasound scans.

O-07 LOWER PREDNISONE DOSING FOR NEPHROTIC SYNDROME RELAPSE: A PROSPECTIVE RANDOMIZED STUDY

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Introduction: Most children with idiopathic steroid sensitive nephrotic syndrome (SSNS) will suffer from more than one relapse after diagnosis. Current steroid regimens on first relapse expose patients to different regimens of daily followed by subsequent every other day steroids, leading to a high cumulative dose. Previous studies have reported on a wide range of prednisone regimens. However there has been no prospective controlled study comparing the efficacy of different prednisone doses to achieve short term remission and its effects on subsequent relapses.
Material and methods: In this single center randomized controlled study, SSNS children were randomized to receive at relapse 3 different prednisone doses: 1-, 1.5- or 2 mg/kg/day. Patients that didn't respond after 14 days were switched to 2 mg/kg. Time to remission (2 consecutive days with urine protein dipstick <1) and rate of treatment failure were recorded. Children were followed for 3 months after this intervention.
Results: We enrolled 30 patients (1 mg/kg/day = 9; 1.5 mg/kg/day = 11; 2 mg/kg/day = 10). Mean age of all cohort was 5.8 ± 3.2 year and disease duration 2 ± 1.5 years, without differences in these 2 parameters between groups. Mean days to response was 8 ± 2.7 (1 mg/kg), 9.5 ± 2.3 (1.5 mg/kg) and 7.1 ± 1.3 (2 mg/kg) without statistical difference between the groups. Two patients, one from 1 mg/kg group and second from 1.5 mg/kg failed to enter remission within the first 14 days and were switched to 2 mg/kg. One responded after 3 days, and the second after 2 weeks. Mean cumulative prednisone doses were 22.6 ± 5.1 mg/kg (1 mg/kg), 34.9 ± 8.1 mg/kg (1.5 mg/kg) and 45.2 ± 3.5 mg/kg (2 mg/kg) (p < 0.05). Time to subsequent relapse was not calculated since 20% of the patients were continue with low prednisone dose or started mycophenolate mofetil.
Conclusions: In conclusion our results support the use of lower prednisone doses in relapses of SSNS. A daily prednisone dose of 1 or 1.5 mg/kg/day is as effective as 2 mg/kg, leading to a lower cumulative dose. The use of lower daily prednisone dose may lower its side effects rate.

O-08 STANDARD VS. EXTENDED COURSE PREDNISOLONE THERAPY FOR THE PRESENTING EPISODE OF STEROID SENSITIVE NEPHROTIC SYNDROME: THE PREDNOS STUDY

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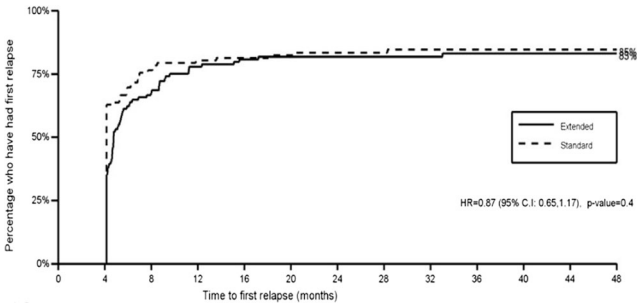
Introduction: The optimal corticosteroid regimen for treating the presenting episode of steroid sensitive nephrotic syndrome (SSNS) remains uncertain. Previous systematic reviews indicating that >3 m of prednisolone reduces risk of relapse and FRNS have not been confirmed by recent large, high quality RCTs.
Material and methods: 237 UK children (146 male) aged 1-15y with newly presenting SSNS were randomised to receive double-blind standard course (SC: 4w 60 mg/m^2 then 4w 40 mg/m^2 AD, total 2240 mg/m^2) or extended course (EC: 4w 60 mg/m^2 then 12w tapering 60 mg/m^2 AD

to 10 mg/m^2 AD, total 3150 mg/m^2) prednisolone and followed for a minimum of 24 m. The study primary end point was time to first relapse. Secondary endpoints included overall relapse rate, use of other immunosuppression, FRNS, AEs, and assessment of quality of life (PedsQL) and behaviour (Achenbach Child Behaviour Checklist (ACBC)). A comprehensive cost-effectiveness analysis (CEA) was performed. Analysis was by intention to treat.

Results: There was no difference in time to first relapse between SC and EC regimens (Figure 1: HR 0.87 (0.65–1.17), $p = 0.4$).



No differences were observed in the proportion experiencing relapse over 24 m (SC 81% vs. EC 80%; $p = 0.9$), developing FRNS (50% vs. 52%; $p = 0.8$) and requiring other immunosuppression (56% vs. 54%; $p = 0.4$). Total prednisolone dose received during the trial was 5446 mg vs. 6645 mg; $p = 0.06$. SAE rates were comparable (25% vs. 17%; $p = 0.1$) and analysis at 16w, 12 m and 24 m showed no significant differences in the incidence of cushingoid features, striae, hypertrichosis, cataract, acne, increased appetite, glycosuria or abdominal pain. There were no differences in PedsQL or ACBC scores. EC therapy was associated with a mean increase in generic health benefit (0.0182 additional QALYs) and cost savings (£3194 vs. £4915).

Conclusions: PREDNOS has not shown any clinical benefit associated with EC prednisolone therapy in UK children, however has shown this to be cheaper and more effective in QALY terms.

O-09 MODIFICATION OF YOUNG ADULT OUTCOMES ON HAEMODIALYSIS- CHANGING "OUR" PRACTICE

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Introduction: Young adults (YAs) with end stage renal disease (ESRD) face unique complex physical, psychological and social challenges making them particularly vulnerable. Those with ESRD on haemodialysis (HD) have an over tenfold increase burden of cardiovascular comorbidity and mortality when compared to contemporaries, significantly reducing their life expectancy. This contrasts with a fourfold increase when compared to older patients and their contemporaries.

All YAs on our St George's renal database were surveyed (Have your say!) in 2014, to ascertain changes they would like to see in the renal unit to improve their patient experience. Peer support opportunities featured in over 70% of responses. Additionally, an adherence assessment of our YAs showed 100% of the YAs on HD had adherence issues. We therefore elected to assess whether providing peer support opportunities could improve haemodialysis outcomes for this patient group.

Material and methods: We targeted our YAs (18–25 years) on haemodialysis population and offered them the opportunity of forming a cohort on the Monday, Wednesday & Friday twilight shift 5.30(6.00) pm to 9.30(10.00) pm, as a preparatory survey identified later dialysis away from the weekend to be the preferable option for the majority of the YAs. Six of our young adults demonstrating low adherence were cohorted and followed up in our Young Adult Clinic.

Results: Attendance and completion of dialysis sessions significantly improved in this cohort. There has also been a reduction in hospital admissions and length of stay among our young adults as well as reduced intra-dialysis weight gain. The Young Adults at St George's hospital are consistently achieving Urea Reduction Ratios (URR) above the Renal Association (RA) Guidelines 65%. URR scores have increased steadily across the young adult cohort population between 2015 and 2017 similarly to Haemoglobin levels. Bone health however has remained suboptimal over this period.

Parameter	2015	2016	2017
Missed Dialysis Sessions	20.8%	4.8%	3.5%
URR	69.9%	77.3%	83.65%
Haemoglobin (mean)	90 g/dL	103 g/dL	107.8 g/dL
PTH (mean)	76.7 pmol/L	51.1 pmol/L	72.2 pmol/L

Table: Demonstrating change in some of the parameters assessed including dialysis attendance, dialysis adequacy, haemoglobin and bone health. Additionally, reported qualitative data shows wellbeing improved as young adults gained awareness of other young adults on dialysis, formed friendships and utilised the opportunity for peer support.

Conclusions: The cohorting of YAs together, on to the monday, wednesday, friday haemodialysis twilight shift, has significantly improved the attendance and completion of dialysis sessions in this patient group. In addition patients have reported improved wellbeing. Bone health however has remained suboptimal and should remain a focus for future improvment. The care of our YAs on haemodialysis was highlighted as excellent practice on the recent National Care Quality Commission (CQC) and peer review visits in 2016.

O-10 EARLY CHANGES IN PD MEMBRANE MORPHOLOGY WITH LOW GDP FLUIDS

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Introduction: The impact of peritoneal dialysis (PD) on membrane integrity and function is incompletely understood, peritoneal changes associated with low GDP fluid usage are largely unknown. Experimental studies indicate a pivotal role of the microvasculature for transport, respective human data is scant.

Material and methods: We obtained 212 standardized peritoneal and 153 omental specimens from 111 children at time of catheter insertion and from 99 children on PD, 84 treated with low GDP fluids for automated quantitative morphometric and molecular tissue analyses.

Results: The mesothelial cell layer vanished with time on PD. Median capillary density increased by 54%, endothelial surface area by 37% within one year of low GDP PD, and remained high thereafter. The submesothelial three-layer vessel configuration dissipated, lymphatic vessel density remained low. The submesothelial zone thickness increased by 45% within

the first year, and slowly thereafter until ≥4 years of PD, when a 3-fold increase in submesothelial fibrosis was observed. Intraindividual comparisons in 24 children reconfirmed these findings. ASMA+, activated fibroblasts and CD45/CD68+ macrophages, VEGF and TGF-ß induced pSMAD abundance, profibrotic miR21 and endothelial mesenchymal transition increased irrespective of PD fluid GDP content. Findings were comparable in 34 patients with history of peritonitis longer than 4 weeks ago. Vasculopathy was more pronounced in children on high GDP fluids. Peritoneal microvessel density correlated with 2 h D/P creatinine and D/D0 glucose at baseline (rho = 0.74/0.78) and while on PD (−0.51/−0.61). By multivariate analyses vessel density predicts PD membrane function.

Conclusions: Peritoneal membrane transport function primarily depends on microvessel density. PD fluids with low GDP content induce early inflammation, EMT, angiogenesis and submesothelial fibrosis, whereas lymphatic vessel density remains low. Minor changes develop subsequently until 4 and more years of PD. These alterations are in line with inferior transport function with low GDP fluids observed in clinical trials within the first months of PD but not thereafter.

O-11 EFFECTS OF HAEMODIAFILTRATION (HDF) VS CONVENTIONAL HAEMODIALYSIS (HD) ON GROWTH AND CARDIOVASCULAR MARKERS IN CHILDREN – DATA FROM THE HDF VS HD (3H) STUDY

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Introduction: Randomised trials in adults suggest that HDF with high convective volumes is associated with reduced cardiovascular mortality. Also, a single centre study has shown improved growth in children on HDF.

Material and methods: We performed a prospective longitudinal study in children on HDF vs conventional HD to determine annualised change in cardiovascular end-points and growth and to determine factors related to dialysis therapy that may improve outcomes.

Results: 179 children (106 on HD and 73 on HDF) were recruited from 28 centres in 10 European countries. There was no difference in age, underlying diagnosis, previous dialysis therapy, dialysis vintage, residual renal function, type of vascular access (AVF in 34 vs 29% on HD and HDF) or blood flow between HD and HDF groups. High flux dialysers and machines with HDF capability were used even in a significant proportion of HD patients (38% and 76% respectively), but ultra-pure water was available only in 57% of HD patients ($p < 0.0001$). HDF patients achieved a median convective volume of 13.3 L/m2; this was significantly influenced by blood flow only, and independent of vascular access type. At baseline, HDF patients were taller (height SDS −1.39 vs −2.21; $p = 0.0005$), and had a higher Kt/V and urea reduction rate. On further assessment of the prevalent dialysis patients (35 on HD and 34 on HDF), HDF patients had a higher haemoglobin and albumin and lower serum phosphate compared to HD patients ($p = 0.04$, $p = 0.02$ and $p = 0.01$ respectively). Serum bicarbonate <18mMol/L was more common in HD than HDF patients ($p = 0.03$). Differences in serum phosphate and bicarbonate persisted after adjusting for centre bias.

Conclusions: This is the largest prospective cohort study on dialysis outcomes in children. At baseline analysis prevalent patients on HDF had significantly lower serum phosphate levels and were less likely to have metabolic acidosis than those on HD.

O-12 HEMODIAFILTRATION IS ASSOCIATED WITH REDUCED INFLAMMATION AND OXIDATIVE STRESS AND IMPROVED ENDOTHELIAL RISK PROFILE COMPARED TO HIGH-FLUX HEMODIALYSIS IN CHILDREN

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Introduction: Randomised trials in adults have shown reduced all-cause and cardiovascular mortality on hemodiafiltration (HDF) compared to conventional hemodialysis (HD), but the mechanisms for improved outcome are not clear and pediatric data is scarce.

Material and methods: We studied non-traditional cardiovascular risk factors of inflammatory (IL-6, hsCRP, pentraxin-3, LP-PLA2), anti-inflammatory (IL-10), oxidative stress (nitrotyrosine), anti-oxidant capacity [Total antioxidant capacity (TAC)] and endothelial (ADMA, SDMA, oxidized LDL) markers in 22 children (13 female, aged between 8.9–15) on HD and HDF in two tertiary dialysis units in London and Istanbul. All children received HD for at least 3 months, and then switched to HDF.

Results: None of the measures, except IL-10 levels, correlated with time on dialysis, suggesting that even a short dialysis vintage of 3 months on HD increases inflammatory and endothelial markers. After 3-months of switching to HDF there was a significant improvement in Beta2 microglobulin (B2m), IL-10, hsCRP, ADMA, SDMA, AGE, ox-LDL and TAC (Table-1). HDF was associated with a significant reduction in ADMA, SDMA, hs-CRP and AGE even in children with a urine output >200 ml compared to those with <200 ml urine per day. The clearance of these markers was not associated with the type of vascular access, but children with a lower blood flow rate had higher inflammatory status (higher IL-6/IL-10 ratio; $p = 0.045$, $r = -0.431$). Children with a higher convective volume (higher than median 12.8 L/m2) had lower Ox-LDL ($p = 0.024$), compared to those who achieved a lower convective volume.

The TAC in HDF was comparable to levels in a cohort of CKD2 patients ($p = 0.08$) but other endothelial markers (ADMA, SDMA, ox-LDL) were significantly higher on HDF than CKD2 ($p < 0.01$ for all).

Table 1: Comparison of the inflammatory and oxidative stress marker between HD and HDF in patients.

n = 22	HD median (IQR)	HDF median (IQR)	p value*
LP-PLA2, ng/mL	333 (294–412)	372 (278–424)	0.465
Nitrotyrosine, nM/ml	28.5 (23.7–56)	32 (24.4–43.7)	0.370
Total antioxidant capacity, mmol/L	0.43 (0.40–0.72)	1.68 (0.42–2.39)	**<0.001**
Pentraxim-3, ng/mL	1.29 (0.49–1.91)	1.09 (0.63–1.66)	0.641
IL-6, pg/mL	3.72 (2.34–8.36)	3.76 (2.37–8.86)	0.499
IL-10, pg/mL	8.93 (3.81–146)	5.73 (4.39–10.4)	**0.030**
Oxidized LDL,ng/mL	278 (203–384)	172 (114–211)	**0.001**
ADMA, μmol**/L**	1.03 (0.92–1.21)	0.85 (0.75–1.02)	**0.001**
SDMA, μmol**/L**	3.54 (2.46–3.54)	2.58 (2.12–3.12)	**0.003**
hsCRP, mg/L	2.80 (1.95–3.16)	1.92 (0.70–2.43)	**0.002**
AGEs, ng/mL	1338 (1221–1490)	982 (1029–1221)	**0.001**
B2 microglobulin, mg/L	38.5 (33–43)	22.5 (16–26.2)	**<0.001**

*Wilcoxon signed ranks test.

HD: hemodialysis, HDF: hemodiafiltration, LP-PLA2: lipoprotein phospholipase A2, ADMA: Asymmetric dimethylarginine, SDMA: symmetric dimethylarginine, AGEs: Advanced glycation end products.

Conclusions: A significant improvement in inflammation, antioxidant capacity and endothelial risk profile is seen even within a short time (3 months) of HDF compared to HD treatment.

O-13 BLADDER FUNCTION AT 10 YEARS OF AGE IN BOYS WITH POSTERIOR URETHRAL VALVES (PUV)

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Introduction: Boys affected by PUV may develop bladder dysfunction. We aimed to evaluate bladder status at 10 years of age.
Material and methods: We retrospectively reviewed non-invasive urodynamics performed at ten years in PUV patients. Data recorded: Bladder capacity (BC) and expected BC (EBC), post- void residual (PVR), uroflow pattern, lower urinary tract symptoms (LUTS) and current bladder. Patients divided in group 1 - large BC (>150%EBC), group 2 - normal BC and group 3 - small BC (<65% EBC) as per ICCS criteria. Incomplete bladder emptying was considered when >10% of BC.
Results: We identified 84 boys born 1997–2006; we excluded those who had bladder augmentation (n = 7), renal transplant (n = 12) or had unavailable information (n = 11). Fifty-four patients were analysed (outcomes shown in table). Three patients practised CIC (two in group 1, one in group 3). Two had recurrent UTIs (one in group 1 with abnormal PVR; one in group 3, normal PVR). Seven patients (13%) demonstrated normal uroflow assessment. See Table. 1.
Conclusions: At 10 years, the majority of boys with PUV (87%) have abnormal bladder function with 54% having incomplete bladder emptying.

Table 1

	Total n = 54	Group 1 Large BC n = 13 (25%)	Group 2 NormalBC n = 29 (54%)	Group 3 Small BC n = 12 (21%)	p
Incomplete emptying	29 (54%)	10 (77%)	15 (52%)	4 (33%)	0.4302
LUTS	18 (33%)	4 (31%)	10 (34%)	4 (33%)	0.769
Bladder medication +/−CIC	12 (22%)	3 (23%)	4 (14%)	5 (42%)	0.088
Bell-shape Uroflow	27 (50%)	9 (69%)	14 (48%)	4 (33%)	0.74

O-14 CLINICAL CHARACTERISTICS OF PEDIATRIC URINARY TRACT INFECTION WITH NEGATIVE URINE LEUKOCYTE ESTERASE TEST

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Introduction: Urine dipstick test for leukocyte esterase is known to be negative in some children with culture-positive UTI. In the present study, we retrospectively reviewed the clinical characteristics of culture-positive UTI with negative or positive urine leukocyte esterase test, which would help us to minimize the risk of missing UTI detection based on the negative result of the dipstick test.
Material and methods: The patients who were diagnosed as UTI based on positive urine culture from April, 2006 through March, 2014 were reviewed. Clinical characteristics including age, gender, laboratory data, clinical course prior to diagnosis and pathogens, were compared between patients with negative and positive urine leukocyte esterase test.
Results: Eighty-five patients (20 females and 65 males) with 86 episodes of UTI were enrolled. Leukocyte esterase tests were negative in 7 UTI episodes, all of which were male cases. No significant differences were recognized in the median age (0.37 and 0.38 years, p = 0.583), peripheral WBC count (18,600 and 15,100/μl, p = 0.275) and CRP (5.19 and 5.70 mg/dl, p = 0.890) between leukocyte esterase negative and positive episodes of UTI. Mean duration between the onset of fever and diagnosis was significantly longer in leukocyte esterase negative episodes than positive episodes (3.43 and 1.37 days, p = 0.015). *E. coli* was the pathogenic bacteria in 87.3% episodes of leukocyte esterase positive UTIs, while Enterococcus faecalis was not detected in those UTIs. Of note, 5 out of 7 cases (71.4%) of leukocyte esterase negative UTIs were caused by Enterococcus faecalis.
Conclusions: Our study suggests that Enterococcus faecalis significantly contributes to leukocyte esterase negative UTIs, and associated with the risk of delayed diagnosis of UTI in children.

O-15 ADOLESCENTS WITH POSTERIOR URETHRAL VALVES: KIDNEY AND BLADDER FUNCTION AT TRANSITION TO ADULT CARE

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Introduction: The short-term prognosis for boys with posterior urethral valves (PUV) has improved in recent decades, but the long-term outcome for kidney and bladder function are not well defined. Boys with PUV are followed from birth till 16–18 years of age by pediatric nephrologists and urologists. The kidney and bladder function of this cohort has not been clearly defined at this sensitive time of transition to adult care.
Material and methods: The data of 41 boys with PUV were analyzed at time of transition to adult care. The average time of observation following valve ablation was 16 years (15-18 years). Outcome measures were defined as kidney and urinary tract damage or loss of function. Kidney damage was assessed by ultrasound (lack of corticomedullary differentiation and hydronephrosis) and kidney function by eGFR (MDRD calculation) with KDOQI categorization of Chronic Kidney Disease (CKD). Bladder function was assessed by bladder diary and uroflow

measurements (presence of increased bladder capacity and significant post void residual (PVR)).
Results: 83% (34/ 41) of subjects developed CKD. The majority (49%) had CKD1 with normal eGFR, the remaining 34% had decreased eGFR (17%-CKD2, 7%-CKD3, 2%-CKD4, 5%-CKD5). Twenty-nine percent (12/41) received antihypertensive treatment. Kidney ultrasound showed loss of corticomedullary differentiation in 60% (25/41) and persistent hydronephrosis in 51% (21/41) of the cohort. Incontinence was reported by 24% (10/41). Uroflow measurements revealed a bladder capacity above 150% of EBC (expected bladder capacity for age) in 19 (46%) and PVR greater than 10% of bladder capacity in 13 (31%).
Conclusions: 83% boys with PUV have CKD and 46% have bladder dysfunction at time of transition to adult care. Adolescent boys with PUV require close surveillance and active treatment when transferred to adult nephrology and urology care due to the high risk of developing both ESRD and bladder insufficiency.

O-16 GDNF CONTROLS URETER LENGTH BY REGULATING COLLECTING DUCT PROGENITOR EXPANSION

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Introduction: Mechanisms controlling ureter length and kidney position in the abdominal cavity are poorly understood. Signaling through RET tyrosine kinase receptor is important for urogenital system (kidneys, associated urine excretion organs and reproductive organs) development, but which of its four ligands is involved in specific processes remains unclear.
Material and methods: We have studied how glial cell-line derived neurotrophic factor (GDNF) impacts the patterning of the whole urogenital system by utilizing a new mouse model endogenously expressing excess GDNF due to disruption of normal 3'untranslated region function. These hypermorphic GDNF mice develop hypodysplastic kidneys and survive approximately two weeks after birth.
Results: We show that GDNF regulates collecting duct progenitors, a newly identified cell population residing in the ureteric bud tips. High GDNF levels expand progenitors at the expense of ureteric trunk elongation due to changes in cell cycle length and progenitor motility. Consequently kidneys fail to ascend to their normal position resembling human pelvic kidneys. Interestingly, postnatal nephrogenesis continues longer in GDNF hypermorphic kidneys than in control mice, suggesting that GDNF could act as a cessation signal for renal differentiation. Furthermore, normal GDNF level is also important for distal ureter remodeling as excess GDNF leads to infertility in both genders due to an imperforate hymen in females and due to an ectopic connection of vas deferens to seminal vesicles in males.
Conclusions: Our findings show that GDNF plays several functions beyond ureteric bud induction and suggest that abnormal GDNF levels may be causative for a portion of CAKUT, pelvic kidney and infertility cases in humans.

O-17 MUTATIONS IN THE LEUKEMIA INHIBITORY FACTOR RECEPTOR (LIFR) GENE AND LIFR DEFICIENCY CAUSE URINARY TRACT MALFORMATIONS

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Introduction: Congenital anomalies of the kidneys and urinary tract (CAKUT) are the most common cause of chronic kidney disease in children. As CAKUT is a genetically heterogeneous disorder and most cases are genetically unexplained, we aimed to identify new CAKUT causing genes.
Material and methods: Whole-exome sequencing (WES) and a trio-based *de novo* strategy were performed in a sporadic CAKUT patient and his healthy parents identifying the leukemia inhibitory factor receptor (*LIFR*) as a novel CAKUT candidate gene. Additionally, *LIFR* mutational screening of a CAKUT patient cohort as well as *in vitro* and *in vivo* characterization of this gene or identified variants thereof were performed in cellular and mouse models.
Results: WES identified a novel heterozygous *de novo* frameshift variant in the *LIFR* gene causing instability of the mRNA in a patient presenting with bilateral CAKUT and requiring kidney transplantation at one year of age. *LIFR* encodes a transmembrane receptor utilized by IL-6 family cytokines, mainly by the leukemia inhibitory factor (LIF). Mutational analysis of 121 further patients with severe CAKUT yielded two rare heterozygous *LIFR* missense variants predicted to be pathogenic in three unrelated patients. LIFR mutants showed decreased half-life and cell membrane localization resulting in reduced LIF-stimulated STAT3 phosphorylation. *LIFR* showed high expression in human fetal kidney and the human ureter, and was also expressed in the developing murine urogenital system. *Lifr* knockout mice displayed urinary tract malformations including hydronephrosis, hydroureter, ureter ectopia, and, consistently, reduced ureteral lumen and muscular hypertrophy, similar to the phenotypes observed in patients carrying *LIFR* variants. Additionally, a form of cryptorchidism was detected in all $Lifr^{-/-}$ mice and the patient carrying the *LIFR* frameshift mutation.
Conclusions: We demonstrate heterozygous novel or rare *LIFR* mutations in 3.3% of CAKUT patients, and provide evidence that *Lifr* deficiency and deactivating *LIFR* mutations cause highly similar anomalies of the urogenital tract in mice and humans.

O-18 AUSTRALIAN RENAL GENE PANELS: RESULTS FROM A NATIONAL DIAGNOSTIC SERVICE

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Introduction: The reducing cost of massively parallel sequencing has enabled rapid incorporation into clinical practice. Judicious use will significantly aid in primary diagnostic investigations.
Material and methods: Seventeen Renal Gene Panels have been developed at the department of Molecular Genetics at Children's Hospital at Westmead by a multidisciplinary group of nephrologists, clinical and molecular geneticists according to reported association with renal phenotypes. Referrals were accepted from Aug 2014. Sequencing was performed using an Illumina TruSight One panel capture on an Illumina HiSeq platform with analysis using NextGene bioinformatics pipeline. Genes of interest were backfilled as required. Pathogenic variants were confirmed with Sanger sequencing.
Results: Forty-three percent of patients had an identifiable disease-causing variant. Where clinical phenotype is clearly defined, the diagnostic rate was high in both adult and paediatric cohorts (Alport's 80% and

88% respectively; Tubular 75% both). However, where a condition displays heterogeneous genetic aetiology, detection rate was generally higher in the paediatric compared to the adult cohort (Complement mediated disease 64% vs. 17% and Ciliopathies 47% vs. 29%). Identification in the nephrotic cohort was even (33% vs. 32%) which reflects a differing threshold for referral by adult and paediatric clinicians.
Conclusions: The use of genetic analysis is now available to Australasian clinicians and renal patients. However, the rate of detection differs according to age at presentation and renal phenotype. Using this information, clinicians can better counsel at the time of consenting patients for diagnostic genetic testing and provide higher fidelity phenotypic information to genetic diagnostic laboratories.

O-19 EXOME SEQUENCING FOR THE IDENTIFICATION OF CANDIDATE GENETIC VARIANTS IN RENAL HYPODYSPLASIA PEDIATRIC PATIENTS

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Introduction: Non-syndromic renal hypodysplasia (RHD: OMIM #610805) is a severe congenital kidney anomaly which is a defect in the number and/or normal differentiation of nephronic units leading to kidney function impairment. RHD accounts for 20–25% of the end stage renal failures causes in children. Although 20 renal developmental genes were associated to RHD, the majority of patients remains without a genetic diagnosis. This study aims to identify new disease-causing mutations in RHD patients using Whole Exome Sequencing (WES).
Material and methods: WES (80X average coverage) was performed with Ion Proton System in the DNA of 20 children with sporadic non-syndromic bilateral RHD, with or without associated upper urinary tract malformations. In 12/20 cases, it was possible to sequence also healthy relatives as control. WES data analysis was done using "QueryOr" variant prioritization platform (http://queryor.cribi.unipd.it/).
Results: Our data analysis highlighted candidate variants in 60% of patients. Four were new mutations, confirmed by Sanger sequencing, in RHD genes (SIX2, SALL1, SLIT2, ROBO2). The other variants were identified both in genes known to be associated to different renal disorder (eg. NPHS2), and as well in genes that are not associated to renal malformations. In 8 patients none candidate variations were up to now identified.
Conclusions: The data obtained underline a larger allelic and locus heterogeneity involved in the RHD determination. These findings suggest that a RHD diagnostic panel must comprised all syndromic and non–syndromic CAKUT genes but also renal cystic diseases genes.

O-20 SOLUBLE UROKINASE PLASMINOGEN ACTIVATOR RECEPTOR (SUPAR) SERUM LEVELS PREDICT PROGRESSION OF KIDNEY DISEASE IN CHILDREN

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Introduction: Renal disease progression rate in CKD children is highly variable. Even within individual age and disease groups, progression rate varies widely, defining a need for informative prognostic biomarkers predicting disease progression and the need for early intervention in an individual patient. Recently, serum soluble urokinase plasminogen activator receptor (suPAR) has been shown to be a strong predictor of incident CKD stage 3 in adults. Here we aimed to determine whether elevated suPAR levels are associated with renal disease progression in children with CKD.
Material and methods: Post-hoc analysis of two prospectively followed pediatric CKD cohorts (ESCAPE trial and 4C Study) including 898 children (mean age 11.9 ± 3.5 years) with serum suPAR level measured at enrollment and longitudinal eGFR measured prospectively. Renal diagnoses included CAKUT (70%), tubulointerstitial nephropathies (10.2%), glomerulopathies (7.7%), post-ischemic (4.7%), and other CKD (6.5%). Mean eGFR was 34 ± 16 ml/min/1.73 m2, median follow-up 3.1 (0 to 7.9) years. The primary renal endpoint was a composite of 50% eGFR loss, eGFR < 10 ml/min/1.73 m2 or start of renal replacement therapy.
Results: 5-year endpoint-free renal survival was 64.5% (95%CI 57.4–71.7%) in children with suPAR in the lowest quartile as compared to 35.9% (95%CI 28.7–43.0%) in those with levels in the highest quartile ($P < 0.0001$). In multivariable analysis, the risk of attaining the endpoint was higher in children with glomerulopathies and increased with age, blood pressure, proteinuria and lower eGFR at baseline. In patients with baseline eGFR > 40 ml/min/1.73 m2, higher log-transformed suPAR levels were independently associated with a higher risk of CKD progression (HR 5.12, 95% CI 1.56–16.7, $P = 0.007$).
Conclusions: Elevated suPAR levels are independently associated with disease progression in children with mild to moderate CKD. Further studies are warranted to determine whether suPAR measurements in pediatric CKD patients will facilitate the identification of children at need for early nephroprotective interventions.

O-21 COLLAGEN (COL4) MUTATIONS IN STEROID-RESISTANT NEPHROTIC SYNDROME (SRNS) IN CHILDREN

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Introduction: SRNS is a genetically heterogeneous glomerulopathy with a high rate progression to CKD. Mutations in *COL4A3–5* genes associated with Alport syndrome (AS) or thin basement membrane nephropathy (TBMN) have been identified in patients with SRNS recently. The aim of the study was to investigate the frequency of *COL4A3–5* mutations in children with SRNS by targeted next generating sequencing (NGS).
Material and methods: 26 children (9 M/17F) with non-familial SRNS were recruited. Renal biopsy revealed FSGS in 22 (84.6%), MCD in 2 (7.7%) and IgAN in 2 (7.7%) patients. A targeted NGS covering 68 genes implicated in SRNS including *COL4A3–5* was applied. All identified by NGS pathogenic variants were confirmed by direct Sanger sequencing.

Results: Mutations in *COL4A3–5* genes were identified in 4 out of 26 (15.4%) children with SRNS (Table 1). Three novel sequence variants in *COL4A3* and *COL4A5* genes were detected with consideration as likely pathogenic by *in silico* studies and AS autosomal dominant and X-linked was diagnosed. Their phenotypes included haematuria in all SRNS children (familial haematuria in 3 out of 4 patients) without hearing loss or ocular disorders. Kidney biopsy shown FSGS (n = 3) and IgAN (n = 1). Electron microscopy (EM) revealed histopathological features of AS, including thickening and lamellation of glomerular basement membranes, and diffuse effacement of podocytes foot processes. Immunosuppressive treatment was not effective in 3 SRNS children with *COL4A3* and *COL4A5* mutations and was stopped. TBMN was diagnosed in one girl with SRNS and IgAN based on identification a previously described pathogenic mutation in *COL4A4* gene. At the last follow up 3 SRNS pts. with AS due to *COL4A3* and *COL4A5* mutations were on ACE inhibitors and had isolated non-nephrotic proteinuria with normal kidney function.

Patients / gender	Age at onset SRNS (years)	Haematuria (familial)	Histological type of SRNS (AS features by EM)	Gene	DNA (protein) ID (dbSNP) or novel (*)	Phenotype
1. 1. F	12.5	+ (+)	FSGS (+)	*COL4A3*	c.4743T>G p.(Phe1581Leu) heterozygous*	AS autosomal dominant
1. 2. F	4.5	+ (+)	IgAN (NA)	*COL4A4*	c.4760C>G p.(Pro1587Arg) heterozygous (rs190148408)	TBMN
1. 3. M	5.0	+ (+)	FSGS (+)	*COL4A5*	c.936+2T>A homozygous*	AS X-linked
1. 4. F	7.5	+ (NA)	FSGS (+)	*COL4A5*	c.2595delA p.(Pro865fs) heterozygous*	AS X-linked

Conclusions: We conclude that *COL4A3-5* genes mutations were the most prevalent mutations identified in children with SRNS by NGS. Genetic analysis in SRNS children important not only for diagnostics, but also for therapeutic decisions, counseling and prognosis.

O-22 TARGETING THE TYPE IV COLLAGEN MISFOLDING IN HEREDITARY NEPHRITIS ALPORT SYNDROME

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Introduction: Alport syndrome (AS) is a hereditary kidney disease that is caused by a mutation in Type IV collagen (COL4A) α3, α4 or α5 gene. Genetic re-expression of α3 gene rescued the phenotype of α3 knockout AS mouse model. Furthermore, the expression level of α345 proteins correlated with the severity of nephritis in clinical study. These results indicated that targeting the causal misfolded protein is a promising therapeutic strategy for AS. However, progress in this research area is hindered by the insufficient knowledge about the regulation of COL4A3/4/5 proteins and the absence of cellular model and tools for AS.
Material and methods: Split nanoLuc-fusion α3/ α5 and α4 were transfected into 293 T cells, and luminescence was assessed in the cell lysate for intracellular trimer and in culture media for secreted trimer. The trimerization of wild type (WT) and some mutant α5 chains were also analyzed in the same way.
Results: Luciferase-based assay showed that WT α345 trimer but not monomer and dimer is detected with high sensitivity. Domain deletion mutant α5 (delCOL, delNC1) had significantly reduced trimerization, and some α5 mutants (*G227S, G325R, G624D, G869R, G1030S, G1107R, G1244D, C1567R, R1569Q, L1649R, R1683Q)* had decreased trimerization compared with WT α5. Moreover, our preliminary data suggested that some chemical chaperones could rescue trimer formation of mutant α5.
Conclusions: The α345 trimer assay system that we developed is applicable for HTS, and could be a powerful tool to elucidate the regulation of causal protein in AS. Although further detailed studies are necessary, the results using chemical compounds suggested that it is possible to correct the trimerization of mutant α5, and targeting α345 in AS becomes more realistic.

O-23 CLINICAL GENETIC TESTING USING NEXT GENERATION SEQUENCING IN STEROID-RESISTANT NEPHROTIC SYNDROME AND ALPORT SYNDROME

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Introduction: Steroid-resistant nephrotic syndrome (SRNS) and Alport syndrome (AS) are rare diseases in children but may lead to end-stage renal failure requiring transplantation. This study reports on the findings and utility of the first clinically-approved gene panel test for SRNS/AS using next generation sequencing (NGS).
Material and methods: A customised gene panel test was developed using NGS of 37 genes associated with SRNS and AS. Copy number variation (CNV) analysis to look for exon deletion and duplications was performed. Clinical data were collected from genetic referral forms and a bespoke proforma.
Results: A total of 240 patients (137 male) with disease onset ≤18 years were referred for diagnostic genetic testing from 12 countries. The presentation was nephrotic syndrome (NS) in 220 and haematuria/AS in 20 patients. The genetic diagnostic rate (GDR) for SRNS patients was 21.1% with pathogenic variants in 11 different genes, most commonly *NPHS1, NPHS2* and *WT1* (Table 1). In two patients, CNV analysis revealed heterozygous novel deletions: exon 23–29 deletion in *NPHS1* and exon 2 deletion in *NPHS2* respectively. In the haematuria/AS group, causative variants were found in *COL4A3* and *COL4A5* representing a GDR of 40%. Importantly, *COL4* likely-pathogenic variants were also found in patients unsuspected for AS. Clinicians reported that genetic testing results assisted decisions about immunosuppression, biopsy and whether to

use a close relative as a renal donor, and informed about risk of recurrence after transplant in SRNS patients.

Table 1: Genes with likely-pathogenic variants in patients ≤ 18 years with SRNS or haematuria / AS identified by NGS gene panel testing

Gene with likely-pathogenic variant	Steroid-resistant nephrotic syndrome (number of patients) ($n = 209$)*	Haematuria / Alport syndrome (number of patients) ($n = 20$)
NPHS1	12	0
WT1	9	0
NPHS2	7	0
LMX1B	4	0
LAMB2	3	0
MYH9	2	0
PLCE1	2	0
ACTN4	1	0
SMARCAL1	1	0
COL4A3	0	3
COL4A4	1	0
COL4A5	2	5
Total	**44**	**8**

*None of the 11 patients with steroid-sensitive nephrotic syndrome had likely-pathogenic variants and so are not shown in this table.

Conclusions: This study demonstrates that gene panel testing in routine clinical practice provides a genetic diagnosis in a significant number of patients presenting with SRNS or suspected AS. The use of CNV analysis identified large deletions in two patients supporting the use of routine copy number analysis. Use of clinical genetic testing after diagnosis of SRNS has the potential to stratify patients and assist decision-making regarding management.

O-24 STAT3 INHIBITION ATTENUATES THE PROGRESSIVE PHENOTYPES OF ALPORT SYNDROME MOUSE MODEL

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Introduction: Alport syndrome (AS) is a hereditary kidney disease caused by mutation of type IV collagen, which eventually leads to end stage renal disease (ESRD). It is important to understand the molecular mechanisms of progressive kidney failure, especially of early glomerular abnormality. In the present study, we sought to determine the critical regulator and the early cell-signaling pathway in AS progression. We took advantage of previous microarray-based studies and identified STAT3 signaling pathway as an activated pathway in AS mice. Here, we aimed to investigate the role of STAT3 in the progression of AS.
Material and methods: Phosphorylated STAT3 expression was assessed by immunoblotting analysis of AS model mice (Col4a5 G5X mutant). To determine the effect of blocking STAT3 signaling, we treated AS mice with STAT3 inhibitor stattic (10 mg/kg *i.p.*, 3 times/week, 10 weeks, $n = 10$). We assessed the renal function (proteinuria, BUN, serum creatinine) and analyzed the glomerular injury score and fibrosis by histological staining. We further analyzed the gene expression of nephritis-associated molecules.
Results: Phosphorylated STAT3 was up-regulated in AS glomeruli and kidney. Treatment with stattic ameliorated the progressive renal dysfunction such as increased levels of proteinuria, BUN and serum creatinine. Histological analysis revealed that stattic treatment ameliorates the glomerular injury, renal fibrosis and inflammatory cell infiltration. Moreover, stattic also significantly suppressed the gene expression levels of renal injury markers (*Lcn2, Kim-1*), pro-inflammatory cytokines (*Il-6, KC*), pro-fibrotic genes (*Tgf-β*, *Col1a1*, *α-Sma*, *Mmp9*).
Conclusions: STAT3 inhibition significantly ameliorated the renal dysfunction in AS mice. Our finding identifies STAT3 as an important regulator in AS progression, and provides a promising therapeutic target for AS.

O-25 DOUBLE FILTRATION PLASMAPHERESIS – 10-YEAR EXPERIENCE AS AN ALTERNATIVE TO PLASMA EXCHANGE

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Introduction: Plasma exchange is a non-selective treatment for a number of conditions. It removes large-molecular weight molecules from the blood and is associated with risks of bleeding, infection and reaction to blood-products. Double filtration plasmapheresis (DFPP) provides a more specific antibody removal treatment, reducing the need for replacement albumin and other blood products. We have been using this therapy for 10-years and submit our experience.
Material and methods: We retrospectively analysed the medical records of all patients in whom DFPP had been performed using the Infomed HF400.
Results: In total, 30 patients were treated with DFPP. Eleven of these following transplantation; of those 9 were for antibody-mediated rejection and 2 for recurrence of focal segmental glomerulosclerosis. Nineteen patients were treated for other conditions including; anti-GBM disease (5), MPGN disease, Wegener's granulomatosis (3), Myasthenia Gravis, Familial Hypercholesterolaemia (2), Guillain-Barré disease, autoimmune limbic encephalitis, pre-ABO incompatible transplantation and Henoch-Schönlein purpura (1). In total, adequate data allowed analysis of 155 sessions of DFPP in 21 patients. Two patients with familial hypercholesterolaemia had >50 sessions, on different secondary filters and weren't included. At the time of treatment median age was 13.5 (2.2–19.2) and median weight 43.3 kg (13.4–82.8). Patients had a median of 5 sessions (1–21). Of the 6 transplant patients analysed, 1 recovered, 1 recovered with CKD stage 4 and the other 4 required dialysis within 3 months. We used previously published European data on efficacy end-points in non-transplanted patients and in our 15 patients; 2 showed "absent" response, 4 "partial" and 9 (60%) "complete" efficacy in their treatments.
One patient had a unit of cryoprecipitate, another had 1 blood transfusion; no other blood products were required. In total 29% of sessions required a fluid bolus.
Conclusions: This study demonstrates that DFPP is a safe and effective method of antibody removal, without the risks of blood product replacement.

O-26 IL-10 POLYMORPHISMS IN CHILDREN WITH TUBULOINTERSTITAL NEPHRITIS AND UVEITIS

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Introduction: Tubulointerstitial nephritis (TIN) and uveitis syndrome (TINU) is a rare condition with various etiology. Both TIN and TINU

syndrome have been shown to be enriched in patients with certain HLA type potentially reflecting underlying autoimmune mechanisms. It has also been previously shown that Interleukin 10 (IL-10) and tumor necrosis factor α (TNFα) polymorphisms are associated with idiopathic and non-infectious uveitis. IL10 is an important immunoregulatory cytokine inhibiting T cells, monocytes, and macrophages. TNFα is it's counter regulator inducing inflammation. In this study we investigated IL10 and TNFα polymorphisms in TIN/TINU patients.
Material and methods: Four IL-10 and three TNFα single nucleotide polymorphisms (SNP) (rs1800629, rs361525, rs1800896, rs1799724, rs2222202, rs3024490 and rs6703630) were genotyped in 30 pediatric patients with TIN/TINU syndrome. Control group frequencies for these SNPs were obtained from both Illumina Immunochip analysis of 587 Finnish siblings and from 1000 Genomes project Finnish population subset (n = 99). Fisher's exact test was performed for significance of association for SNP frequencies between patients and controls. Finally, all raw p-values were adjusted for multiple testing by the Benjamini-Hochberg method.
Results: A significant increase in the frequency of *IL-10 + 434T* (rs2222202) and IL-10+*504G* (rs3024490) alleles was found in all patients and in TIN/TINU subgroups separately when compared to the Finnish reference population (100% vs. 40% and 76% respectively, p = <0.05). There were no statistical differences in any of the studied TNFα genotypes between TIN/TINU patients and control population.
Conclusions: A significant difference in the frequency of *IL-10 + 434 T* and *+504G* alleles was found in our patient cohort. This genetic variation in the inflammatory mediators may predispose to inflammatory diseases.

O-27 IL-10 AS A POTENTIAL MEDIATOR IN PARENCHYMAL DAMAGE IN CHILDREN WITH CONGENITAL ANOMALIES OF THE KIDNEY AND URINARY TRACT

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Introduction: IL-10 has essential role in the regulation of the tissue healing/regeneration, fibrosis and regulation of immune response. Since, the parenchymal damage is frequently observed among patients with congenital anomalies of the kidney and urinary tract (CAKUT) we aimed to evaluate the relationship between IL-10 serum levels and parenchymal damages in children with CAKUT. Therefore, we also aimed to clarify whether the IL10 rs1800896 polymorphism has implication in the CAKUT pathogenesis.
Material and methods: The quantitative determination of IL-10 in sera was performed by ELISA test in 79 CAKUT patients and 18 age-sex-matched unaffected children from Bulgarian population. Genotyping of IL10 rs1800896 was performed by allele specific-PCR reaction.
Results: Serum IL-10 was elevated in total group of CAKUT cases compared to controls with marginal significance (28.6 ± 36 pg/ml vs. 12.4 ± 8.5; p = 0.066). Patients with renal hypo/dysplasia showed higher IL-10 levels compared to patients with renal agenesis and controls (37.3 ± 49.5 pg/ml; 17.1 ± 9.5 pg/ml; 12.4 ± 8.5 pg/ml, respectively). Cases with high-grade hydronephrosis elevated IL-10 than controls (34.4 ± 32.2 pg/ml; p = 0.01), in contrast to cases with low-grade hydronephrosis. Also, we observed that almost all extremes values of serum IL-10 belongs to CAKUT patients with −1082*GG-genotype of IL10 rs1800896. Among CAKUT patients with parenchymal damage, GG-genotype was associated with higher serum IL-10 (48.7 ± 63.5 pg/ml) compared to AA (26.03 ± 27 pg/ml; p = 0.05) and AG (22.7 ± 27 pg/ml; p = 0.045) genotypes as well as compared to controls with the same genotype (14.1 ± 9 pg/ml, p = 0.03).
Conclusions: IL-10 might be involved into pathogenesis of parenchymal damage in CAKUT. Higher IL-10 level, at least partially determined by −1082*GG genotype may contribute to IL-10 mediated renal damage in renal hypo/dysplasia, high-grade hydronephrosis and reflux nephropathy.

O-28 TOCILIZUMAB (ANTI-IL6 RECEPTOR) AS A POTENTIAL TREATMENT FOR IDIOPATHIC NEPHROTIC SYNDROME

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Introduction: B cell depletion is a recognized treatment of steroid dependant(SDNS) and steroid resistant nephrotic syndrome(SRNS). Interleukine-6(IL-6) is an attractive target as it promotes B-cell differenciation into antibody-forming plasma cells. We report a pilot series of SRNS and SDNS patients treated with tocilizumab(TCZ), a monoclonal anti IL6-receptor antibody, with Intraveinous Immunoglobulins(IVIg).
Material and methods: 17 patients received an initial treatment combining IVIg(2 g/kg) at day 0 and 30 and TCZ(8 mg/kg, or 12 mg/kg if weight < 35 kg) at day 15 and 45 followed by TCZ once a month in patients in complete remission(A) or twice a month if proteinuria persisted(B). Anticalcineurin inhibitors were discontinued 3 months after remission. Treatment objective was a sustained remission after anticalcineurin inhibitors withdrawal and/or after end of the protocol.
Results: Five SDNS and 12 SRNS were included. Among SRNS, five had tacrolimus-dependant remission and 7 multidrug resistance. All patients but the two youngest had previously received rituximab. Eight patients received schedule (A): tacrolimus was successfully discontinued in 4/6 patients initially under tacrolimus. Sustained remission was observed in three patients, with a follow up after M6 TCZ of 15 months, 2 and 1 months. Nine patients received schedule (B): remission of proteinuria was obtained in 4/9 patients in association with tacrolimus. Tacrolimus was successfully discontinued in one patient. Three patients are still under TCZ and tacrolimus for longer than 6 months. Immunophenotyping showed no reduction in memory B cell subsets while under TCZ. Reversible leuconeutropenia occurred in 2/17 patients.
Conclusions: Targeting IL-6 has been modestly efficient in this small pilot series. Effect of IL-6 inhibition on B cells subsets is inconclusive but most patients had received rituximab in the previous months. Longer follow-up is required to assess if it is a valuable treatment option to be discussed in severe NS or as an alternative to B cell depletion.

O-29 SUCCESSFULLY IMPLEMENTING TRANSITION BY ADDRESSING COMMON MISCONCEPTIONS AND USE OF THE 'READY STEADY GO' TRANSITION PROGRAMME

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Introduction: Aim: To evaluate the impact of an educational session on transition and the 'Ready Steady Go' (RSG) transition programme in overcoming common misconceptions about delivering effective transition.
Background: Transition is defined as the purposeful, planned movement of adolescents from child-centred to adult-orientated healthcare systems. Despite the importance of good transition being recognised, delivery is often fragmented, with many young people (YP) feeling unprepared. This failure has a major impact on long-term outcomes. Interpretation of the definition of transition by many Healthcare Professionals (HCPs) and misconceptions about implementation have delayed the provision of effective transition. An educational package expanding on the definition of transition to include 'empowering the YP by equipping them with the skills and knowledge necessary to manage their own healthcare in paediatric and adult services' and provision of the RSG programme helps enable delivery of transition.

Material and methods: Multi-professional 40 min RSG transition educational sessions were delivered over 2 years in 21 centres. Sessions expanded upon the definition of transition, addressed common misconceptions (need for an adult physician, need for specialist clinics, age YP starts transition) and introduced RSG to demonstrate the incorporation of transition into routine clinical practice. Audience response tools polled HCPs perception and understanding of transition at the start and end of these sessions. RSG uptake was subsequently recorded.
Results: Results from 642 responders during 21 sessions demonstrated a difference pre and post session: Need for an adult physician identified before starting transition 71% versus 11%; Need for specialist clinic 35% versus 14%; Start of transition: 34% start at 14 years+, (23% at 16 years+) versus, 95% starting at 11–12 years. Post-education RSG has been widely adopted across sub-specialities, including paediatric nephrology, throughout the UK.
Conclusions: Expanding the definition of transition and use of the RSG programme overcomes common misconceptions and enables the successful implementation of transition.

O-30 ACUTE PEDIATRIC NEPHROLOGY SIMULATION

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Introduction: Simulation-based education is considered one of the best training strategies to enhance the performance of healthcare professionals. Simulation is used intensively now for the training purpose and for the assessment of residents performance. Interprofessional education is the strategy used to boost the collaborative practice among multidisciplinary team for the best quality of care. Using different types of simulation-based education in pediatric nephrology field is limited in the research body. We conducted multiple simulation sessions to improve the training in the acute pediatric nephrology among the residents.The individual and inter-professional performances of the residents in all the simulation scenarios were objectively assessed.
Material and methods: Three acute pediatric nephrology simulation scenarios were conducted over different days to evaluate the residents' performance. The participants included residents, faculties in pediatric nephrology, faculties in different specialties, nurse and some allied health professionals. All the participants were working together in each scenario as a multidisciplinary team while the target population was the residents of different levels. The scenarios were designed by an expert in pediatric nephrology and simulation as well. The learning outcome is measured by two different tools. The first was a post sessions survey (self-assessment). The second tool was direct observation by high quality performance check list (Expert assessment). The simulation station was 15 min followed by 30 min debriefing. The stations were; acute kidney injury secondary to renal hypoperfusion, renal failure with critical hyperkalemia and neonatal hyperammonemia required CRRT. Student t-test was used to evaluate the quantitative portion of the data expressed by the Likert scale of the post simulation surveys. Manual coding by three independent researchers was done to evaluate the text portion of the questionnaire.
Results: The result showed a significant learning outcome from the simulation sessions. The residents were quite interested in the simulation learning compared to the traditional lectures in the same topics. The hands-on practice and rapid effective actions were observed in all of the simulation scenarios. Most of the residents worked collaboratively and effectively within the multidisciplinary team.
Conclusions: This inter-professional simulation demonstrated an effective learning outcome among the residents on the short term. The long term retention and the performance in real life events need to be evaluated in another study to evaluate the simulation effect on the real residents practice and in hence the patients' outcome. It is recommended to implement the inter-professional simulation-based acute and non-acute training to the residents in the pediatric nephrology curriculum and to disseminate this learning strategy to the other pediatric sub-specialties.

O-31 ALL-CAUSE MORTALITY AND CARDIOVASCULAR DISEASE INCIDENCE IN PATIENTS WITH CHILDHOOD-ONSET END-STAGE RENAL DISEASE IN SCOTLAND

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Introduction: End-stage renal disease (ESRD) in children is a rare but serious health problem, which occurs in about 5 to 10 children per million each year, globally. Scottish Renal Registry (SRR) reports that the incidence of new renal replacement therapy (RRT) patients <20 years of age in Scotland in 2015 is 1.2 per 100.000 population. The major cause of mortality in children with ESRD is cardiovascular disease (CVD). Data on long-term survival and CVD incidence among children with ESRD are sparse. Available studies are short-term, are based on single centre experience and include only selected RRT population (either on dialysis, or after transplantation, or patients in specific age groups). Therefore, we aimed to describe the long-term survival and CVD incidence in patients initiating RRT in childhood in Scotland.
Material and methods: We included all patients who started RRT at <18 years of age between 1961 and 2013 registered in the SRR to describe all-cause mortality (mortality cohort). We identified incident CVD through linkage to causes of death and hospital admission records. CVD incidence was defined as the first CV event, either CV death or CV hospital admission, whichever came first. Since causes of death and hospital admission data is available in Scotland from 1981 we included patients who started RRT between 1981 and 2013 to describe CVD incidence (CVD incidence cohort). We used Cox regression analysis to describe associations between primary renal disease (PRD), type of RRT, age at start of RRT and sex and all-cause mortality or CVD incidence. PRD were divided into three categories: congenital anomalies of kidney and urinary tract (CAKUT), glomerulonephritis and "other" (cystic kidney disease, hereditary nephropathy, ischemic renal failure, vasculitis and metabolic disorders). In the all-cause mortality analyses we included patients from the start of RRT until date of death or 31st December 2015, whichever came first. In the CVD incidence analysis we included patients from start of RRT until CVD event or 31st December 2015, whichever came first.
Results: Characteristics of the mortality cohort (N = 479) and the CVD incidence cohort (N = 381) were similar. There were more males (57.2%) than females, the largest group of PRD was CAKUT (48.6%) and the majority of patients initiated their treatment with dialysis (87.9%). In the mortality cohort 126 patients died during a median follow-up of 18.3 years (interquartile range (IQR) 8.7–27.0). The long-term survival was 86% (95% CI 82.9–89.1) at 10 years and 76% (95% CI 72.2–79.8) at 20 years. In the CVD cohort 134 patients developed CVD incidence during a median follow-up of 12.9 years (IQR 5.6–21.5). The overall crude mortality and CVD incidence rates were 1.5 and 2.6 per 100 person-years, respectively. Patients with an 'other' category of PRD had a higher risk of all-cause mortality compared to patients with CAKUT. Receiving dialysis rather than a kidney transplantation during follow-up was associated with a higher risk of both all-cause mortality and CVD incidence. Younger age at initiation of RRT was associated with a higher risk of all-cause mortality, while the reverse was found with respect to CVD incidence.
Conclusions: Type of RRT and age at start of RRT were significant determinants for both all-cause mortality and CVD incidence, while PRD was significantly associated only with all-cause mortality.

O-32 CAUSES OF NON-ELECTIVE HOSPITALIZATIONS IN CHILDREN WITH CHRONIC KIDNEY DISEASE (CKD): FINDINGS FROM THE 4C STUDY

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Introduction: CKD and renal replacement therapy (RRT) are associated with an increased risk for non-elective hospitalizations. The causes for hospitalizations in the pediatric CKD population have not been defined to date. Here, we aimed to analyze the leading reasons for non-elective hospitalizations in the 4C study, a large prospective European cohort of children with CKD.

Material and methods: Patients aged 6–17 years with CKD stage 3–5 or renal replacement therapy (RRT) followed at 55 centers in 14 countries with at least two study visits were included in the analysis. Data about CKD stage, renal replacement therapy (RRT) and hospitalizations were collected 6-monthly.

Results: A total of 662 patients were followed for a total of 2718 patient-years (PY). A total of 138, 80 and 167 non-elective hospitalizations were documented for patients on conservative therapy (CKD; n = 662), dialysis (n = 170) and renal transplant (RTx) recipients (n = 205), respectively.

The leading causes for non-elective hospitalizations in all groups were infections (44.2%, 44.5% and 48.5% in patients on CKD, dialysis and RTx, respectively). These were followed by non-infectious gastrointestinal conditions (17.4%) in patients on CKD, cardiovascular complications (17.5%; 64.3% hypertension) in patients on dialysis and RTx rejection-related hospitalizations (28.7%) in RTx recipients. Urinary tract infections (UTI) accounted for most of infection-related hospitalization in children on CKD, HD and RTx (62.3%, 35.7% and 44.4%) and peritonitis was the leading cause in patients on PD (62.5%). EBV or CMV infections comprised 13.6% of infection related hospitalizations in RTx.

Conclusions: Infectious diseases represent the leading cause for non-elective hospitalizations in children with CKD and on any RRT. UTIs are the major cause for hospitalizations in most children with CKD. Among patients on RRT, children on PD are at highest risk for hospitalizations due to treatment modality-related infections.

O-33 PRE-EMPTIVE KIDNEY TRANSPLANTATION IS ASSOCIATED WITH IMPROVED GRAFT SURVIVAL IN CHILDREN: DATA FROM THE FRENCH RENAL REGISTRY

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Introduction: Kidney transplantation (KT) is the treatment of choice for end-stage renal disease. Preemptive KT is considered to be the optimal treatment of ESRD particularly in children but reports on the results of pediatric preemptive KT are scarce. The objective of this study was to evaluate the impact of preemptive KT on the risk of graft failure in children with ESRD.

Material and methods: We analyzed all first kidney transplants performed in children <19 years in France between 1995 and 2013. A Cox multivariable model with competing risk analysis was used to study the impact of preemptive KT on the hazard of graft failure defined as return to dialysis, retransplant, or death, whichever occurred first.

Results: A total of 1920 pediatric patients were included, of whom 387 (20.2%) received a preemptive KT. Median time of follow-up was 7 years. At 10 years post transplant, graft survival was 85.2% in preemptive KT and 67.1% in non preemptive KT ($p < 0.001$). After adjustment for recipient age and sex, primary kidney disease, donor type (living or deceased donor), donor age, HLA mismatches, and cold ischemia time, preemptive KT was associated with a 45% reduction in the hazard of graft failure when compared with dialysis prior to KT (HR 0.55; 95%CI 0.41–0.73; $p < 0.001$). Patient survival was not significantly influenced by preemptive KT. The impact of preemptive KT on graft failure risk was greater among deceased donor transplant recipients (HR 0.52; 95%CI 0.37–0.72) than among living donor kidney recipients (HR 0.67; 95% 0.31–1.25). Pretransplant dialysis was associated with an increased hazard of graft failure, whatever the duration of dialysis.

Conclusions: Preemptive KT in children is associated with a lower risk of graft failure than KT performed after the initiation of dialysis, and should be promoted when feasible.

O-34 CORRELATION OF ACOUSTIC RADIATION FORCE IMPULSE (ARFI) ELASTOGRAPHY WITH HISTOLOGICAL FINDINGS IN PEDIATRIC RENAL TRANSPLANT PATIENTS

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Introduction: Acoustic Radiation Force Impulse' elastography (ARFI) is a new methodology for determining tissue and organ stiffness by

ultrasound measurement of shear wave velocity (SWV). Data on ARFI in renal transplantation are limited and there are no data on ARFI in children having renal biopsies for suspected transplant rejection. We aimed to correlate ARFI measurements with histological findings in renal transplant biopsies.

Material and methods: SWV was measured using a 9 MHz linear probe (9 L4) and the Siemens Acuson S3000 Helx ultrasound device equipped with Virtual TouchTM Quantification (VTQ, Siemens, Erlangen, Germany) focusing the region of interest on the central parenchyma/columns of Bertin. Data from 38 pediatric renal transplant patients with suspected rejection, having renal biopsies within one week of ARFI measurement, were analyzed (71% boys; mean age 13.3 ± 6.3 years, time since transplantation 3.6 ± 3.8 years, mean donor age 31.4 ± 13.0 years, mean eGFR 43.3 ± 21.4 mL/min/1.73 m2). Histological lesions were graded according to the BANFF 2015 classification.

Results: 13/38 patients had histological signs of chronic-active rejection. In these patients, SWV was significantly increased compared to patients without rejection (3.31 ± 0.79 m/s vs. 2.68 ± 0.49 m/s, $p = 0.005$). Regarding staging of interstitial fibrosis and tubular atrophy (IFTA, ci and ct grading 0 to 3), SWV increased with increasing scores. The difference in SWV was significant for patients with IFTA3 and ct3 compared to those with lower grading ($p < 0.05$). SWV was correlated with the percentage of sclerotic glomeruli ($r = 0.44$, $p = 0.006$). No correlations were found between conventional resistance index measurements and histological findings.

Conclusions: Shear ware velocity, a measure of tissue stiffness, correlates with the histological findings of fibrosis in pediatric renal allografts. Further studies are warranted to determine whether ARFI is of additional use in the assessment of renal graft outcome.

O-35 ASSOCIATION OF ANGIOTENSIN II TYPE 1 RECEPTOR ANTIBODIES WITH GRAFT HISTOLOGY, FUNCTION AND SURVIVAL IN PAEDIATRIC RENAL TRANSPLANT RECIPIENTS

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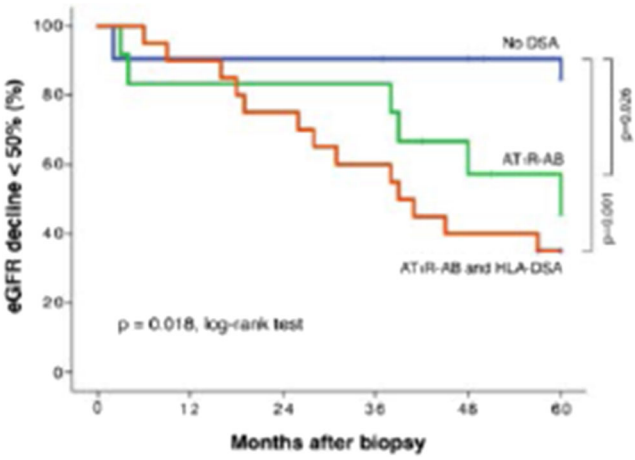
Introduction: The predominant role of antibody-mediated rejections (ABMR) presupposing premature graft dysfunction is comprehensively described. The main causes are donor-specific HLA-antibodies (HLA-DSA). However, not all cases could be attributed to HLA-DSA. Therefore, we found it of interest to investigate the association of non-HLA angiotensin II type 1 receptor antibodies (AT_1R-AB) with graft histology, function and survival.

Material and methods: 62 patients with a low immunological risk profile and graft biopsies for clinical indication more than one year post-transplant were included. Serum samples at the time of indication biopsy were analysed for the presence of HLA-DSA and AT_1R-AB. HLA-DSA with an MFI value >500 were classified as positive (LABScreen Luminex Kits (One Lambda)). The cut-off value for AT_1R-AB was defined as 10 U/mL (ELISA, CellTrend GmbH).

Results: 32 of 62 patients (52%) were positive for AT_1R-AB at the time of indication biopsy. After stratification according to different histologic entities, the ABMR positive group (including biopsies with suspected ABMR, BANFF'15) revealed significantly higher AT_1R-AB levels compared to the group with T-cell mediated rejection or the group without rejection (median 12 vs. 9 or 8 U/mL; both comparisons $p < 0.05$). Based on ROC-curve analyses, the optimal cut-off to differentiate between ABMR and non-ABMR biopsy results was 9.5 U/mL (ROC-AUC 0.74). AT_1R-AB positivity (>9.5 U/mL) was associated with a significantly decreased graft survival 5 years post-biopsy ($p = 0.025$). Furthermore, AT_1R-AB positive patients more frequently reached the end-point 'graft function deterioration', defined as an eGFR-decline of more than 50% compared to baseline values before indication biopsy ($p = 0.004$). Sub-classifying the antibody positive group by HLA-DSA and AT_1R-AB status revealed no significant difference between the only AT_1R-AB positive and the AT_1R-AB/HLA-DSA double positive groups regarding graft function or graft survival.

Conclusions: AT_1R-AB positivity at the time of indication biopsy is associated with a significantly increased risk of graft function deterioration.



O-36 IN VITRO TACROLIMUS EXPOSURE IN HUMAN PROXIMAL TUBULE CELLS RESULTS IN DIFFERENTIALLY INCREASED CTGF EXPRESSION IN RELATION TO THE PHARMACOGENETIC BACKGROUND FOR CYP3A5 AND ABCB1

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Introduction: The interplay between CYP3A enzymes and the P-gp pump (*ABCB1*) is important for drug metabolism and toxicity. Genetic variation in *CYP3A5* and *ABCB1* are important in tacrolimus (Tac) nephrotoxicity. We show for the first time, the implications of Tac exposure in a novel human PTC model incorporating the genetic variation in *CYP3A5* and *ABCB1* on the function of these genes and the production of pro-fibrotic cytokines.

Material and methods: Eight clones of conditionally immortalized PTC (ciPTC) with different combinations of *CYP3A5 (rs776746)* and *ABCB1(rs1045642)* genotypes were incubated with vehicle, 50 ng/mL and 300 ng/mL Tac for 24 and 72 h. qRT-PCR and western blot were performed for *CYP3A5*, *ABCB1* and *CTGF* expression. Functional *CYP3A5* expression was assessed by differential midazolam (MDZ) hydroxylation using LC-MS and P-gp activity by calcein efflux.

Results: -Baseline mRNA, protein and functional expression of *CYP3A5* was higher in ciPTC with the **1* versus **3/*3* allele. Increasing Tac concentration resulted in decreasing 1'OH MDZ hydroxylation ($p = 0.01$). **-**Baseline *mRNA* and calcein-AM efflux was higher in *ABCB1 3435 TT* vs. *CC/CTs* (mean ΔCt *ABCB1:* 2.25 vs. 2.97 and Δfluorescence: 23.4% vs. 45.3%, respectively; $p = 0.001$). A progressive decrease in calcein efflux (50 ng/mL: 79.1%; 300 ng/mL: 68.8%; $p < 0.001$) was observed for both variants. Prolonged incubation resulted in decreased mRNA and protein expression ($p = 0.01$) in *CC/CTs* only. Calcein-AM efflux remained higher in TTs (29.90% vs. 41.26%; $p = 0.016$). **-**Both relative *CTGF* mRNA and protein expression increased with Tac concentration (50 ng/mL: 0.73; 300 ng/mL 0.79; $p = 0.02$). Prolonged exposure nor *CYP3A5* genotype had a significant effect on mean relative *CTGF* expression. However, subgroup analysis demonstrated lower *CTGF* expression after 72 h in **1* carriers vs. non-carriers (0.58 vs. 0.81; $p = 0.03$). *CC/CTs* demonstrated a significant decrease after 72 h, while *CTGF* expression in the *TTs* was significantly higher than *CC/CTs*.
Conclusions: -Tac exposure in human PTC has no direct effect on the regulation of gene expression but results in decreasing functions of CYP3A5 and P-gp. -Tac exposure in human PTC results in a concentration-dependent increase in *CTGF* expression (pro-fibrotic cytokine). -Prolonged Tac exposure results in increased *CTGF* expression in PTC with lower *CYP3A5* functional expression and higher P-gp function which might correspond with an increased risk for renal fibrosis in patients.

O-37 EVIDENCE FOR "SHRUNKEN PORE SYNDROME" IN CHILDREN

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Introduction: Cystatin C (CysC), a LMW protein marker for kidney function, has been linked to mortality independently of kidney function in adults. Elevated levels of CysC compared to serum creatinine (Crea) may reflect diminished filtration of CysC due to a smaller size of glomerular pores - the "Shrunken Pore Syndrome" (SPS). In adult populations, around 8% fulfill the criteria of SPS. This study aims to compare the elimination characteristics of the LMW protein GFR markers CysC, beta-trace protein (BTP) and beta-2 microglobulin (B2M) with creatinine in children using a gold standard GFR measurement.
Material and methods: Retrospective study of 459 inulin clearance (Cin) studies with simultaneous determination of Crea, CysC, BTP and B2M. Establishing eGFR equations in a subset of 153 of these studies. Analysis of the remaining 306 studies for the presence of SPS defined as eGFRLMW-P/eGFRCrea <0.6. Comparison of Cin, eGFRCrea and the three eGFRLMW-P in relation to the eGFRCysC/eGFRCrea ratio.
Results: Cin, eGFRcreat and the three eGFRLMW-P gave comparable results as long as eGFRCysC/eGFRcreat was above 0.6. Beyond this point, eGFR decreased for CysC, BTP and B2M, while eGFRCrea increased. Of note, Cin did not change indicating that filtration of all three LMW markers was diminished compared to inulin while eGFRcreat overestimated GFR. Based on eGFRBTP SPS was observed in 5.6%, on eGFRCysC in 5.2% and on eGFRB2M in 4.2% of the patients.
Conclusions: The proportion of children meeting the criteria of SPS is comparable to adults. In SPS patients, filtration of the three LMW-protein markers is impaired compared to inulin suggesting abnormal filtration rather than an extra-renal mechanism. At the same time, eGFRcreat is increased, most likely due to diminished creat synthesis. Taken together, these findings support a link between SPS and muscle wasting caused by retention of pro-inflammatory cytokines of similar size.

O-38 ELEVATED MYOCARDIAL WALL STRESS PREDICTS LEFT VENTRICULAR REMODELLING IN CHILDREN WITH CHRONIC KIDNEY DISEASE

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Introduction: In children with chronic kidney disease (CKD) myocardial wall stress (MWS) is elevated independent of raised blood pressure and before development of left ventricular hypertrophy. We hypothesised that elevated MWS may predict development of ventricular remodelling over time in children with CKD.
Material and methods: In seventy-nine children (10.7 ± 3.0 years) including 56 children with CKD, transthoracic echocardiography and carotid tonometry were performed at baseline and after an average of 24.4 ± 11.0 months follow-up. Endocardial and epicardial volumes were obtained from Tomtec wall tracking analysis. Left ventricular mass (LVM) was calculated from M-mode. Central aortic pressure during systole was used to estimate LV pressure and was calibrated by mean and diastolic blood pressure (BP). Myocardial wall stress was calculated from LV volume and pressure measurements.
Results: There was a significant increase in height (0.11 ± 0.10 m, $p < 0.001$), weight (9.9 ± 9.5 kg, $p < 0.001$), systolic (8 ± 11 mmHg, $p < 0.001$) and diastolic BP (10 ± 11 mmHg, $p < 0.001$). eGFR reduced significantly (−15.7 ± 1.8 ml/min per1.73cm^2, $p < 0.001$) in CKD group. Among all subjects, change in LVM/EDV was associated with baseline MWS ($\beta = 0.263$, $p = 0.003$) after adjustment for age, gender and blood pressures. LVM/EDV changed by −0.185 ± 0.041, −0.033 ± 0.038 and 0.014 ± 0.043 units respectively across tertiles of baseline mean MWS.
Conclusions: Elevated MWS is an independent predictor of concentric left ventricular remodeling in children with CKD.

O-39 THE FGF23 AND KLOTHO AXIS IN PEDIATRIC CHRONIC KIDNEY DISEASE - A PROSPECTIVE COHORT STUDY

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Introduction: Chronic Kidney Disease-Mineral Bone Disorder (CKD-MBD) is common in pediatric kidney disease patients and a risk factor for future cardiovascular disease (CVD). Fibroblast growth factor-23 (FGF23) and Klotho are novel key players in CKD-MBD, and have been suggested to be involved in the development of CVD.
Material and methods: We prospectively analyzed 74 pediatric patients, 31 with CKD and 43 transplanted (CKD-T) patients annually for 3 years. We assessed longitudinal patterns and predictors of FGF23 and Klotho and associations to cardiac remodeling and function examined by echocardiographic pulse wave Doppler (PWD) and color-coded tissue Doppler imaging (cc-TDI).
Results: The prevalence of high FGF23 levels (≥95th percentile) was 60% in CKD and 42% in CKD-T patients, despite a low prevalence of hyperphosphatemia and normal Klotho levels. Low GFR at baseline was a predictor for high mean log FGF23 during the follow-up in CKD and CKD-T patients ($p < 0.001$). A high log FGF23 z-score longitudinally was borderline significantly associated with elevated left ventricular mass index (LVMI) in CKD patients ($p = 0.06$). In addition, high log FGF23 ($p = 0.008$) and low log Klotho ($p = 0.02$) over time were associated with a worse left ventricular diastolic function (cc-TDI e′/a′) in CKD-T patients.

Conclusions: In pediatric CKD and CKD-T patients, FGF23 increase and Klotho decrease with progressing renal failure, despite well controlled phosphate levels. Following adjustments, both high FGF23 and low Klotho were strongly associated with a worse left ventricular diastolic function longitudinally. The potential role of FGF23 and Klotho in cardiac morbidity in pediatric CKD requires further investigation.

O-40 ANTHROPOMETRY AND CLINICAL OUTCOMES IN PAEDIATRIC RRT - RESULTS FROM THE ESPN/ERA-EDTA REGISTRY

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Introduction: We aimed to study associations of height and body mass index (BMI) and access to transplantation and risk of death during the entire course of paediatric RRT.
Material and methods: We included all children <20 years starting RRT in 31 European countries between 1995 and 2014 for whom anthropometric data were reported to the ESPN/ERA-EDTA Registry.
We defined short and tall stature as a height SDS < −1.88 and >1.88. BMI was expressed according to height-age.
Associations with outcomes were assessed using Cox regression models with time-dependent covariates and adjusted for country, age, sex, primary renal disease, and treatment modality, where relevant.
Results: After a median follow-up of 4.9 years [IQR: 2.3–7.9], 279 patients out of 6255 patients had died. Causes of death were known for 69% of patients, and infection- or cardiac related deaths were most common (both 24%). Compared to children with normal stature, all-cause mortality risk was higher among short children (aHR: 2.05, 95% CI: 1.58–2.67), but not among tall ones. Short patients particularly showed a higher risk of infection-related death (aHR: 3.78, 95% CI: 2.10–6.80). Compared to normal weight subjects, underweight patients showed a higher all-cause mortality risk (aHR: 1.94, 95% CI: 1.35–2.78) and a lower access to transplantation (aHR: 0.80, 95% CI: 0.69–0.92). We also found a trend towards lower transplantation rates for obese patients. (aHR: 0.86, 95% CI: 0.74–1.01). Furthermore, obese patients were more likely to die from cardiac causes (aHR: 3.09, 95% CI: 1.35–7.08) compared to normal weight patients.
Conclusions: We found a higher mortality risk among paediatric RRT patients with short stature and those who were underweight. Furthermore, extremes in BMI were associated with a lower access to transplantation. Our results highlight the need for careful nutritional management and timely intervention in these patients.

O-41 UBIQUITIN CARBOXYL-TERMINAL HYDROLASE L1 IS A PODOCYTE TARGET OF IGG ANTIBODIES IN IDIOPATHIC NEPHROTIC SYNDROME

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Introduction: The efficiency of B cell-depleting treatments highlights the involvement of B cells in INS. This study searched for identifying antibodies (Abs) directed against podocytes in patients with INS.
Material and methods: The study included 42 patients sampled at various stages of INS, and 38 controls. Fractions of plasma obtained by size exclusion chromatography were tested on cultured podocyte adhesion; 2/ specificities of IgG Abs contained in the plasma fraction of interest were studied through immunoprecipitation of a podocyte lysate then identification of cognate antigens by liquid chromatography-mass spectrometry.
Results: Cultured podocyte detachment was observed with one specific plasma fraction in 16/34 INS relapsing patients, 1/11 INS patients in remission and 0/25 controls. IgG were isolated from this specific plasma fraction in 3 INS relapsing patients (all detaching cultured podocytes), the 3 same INS patients in remission (all not detaching cultured podocytes) and in 3 controls, then used to immunoprecipitate a podocyte lysate. Comparative proteomic analysis allowed selecting 5 proteins according to statistical and biological criteria. Specific Abs were tested and only anti-Ubiquitin Carboxyl-Terminal Hydrolase L1 (UCHL1) IgG led to podocyte detachment. Pre-incubation of either anti-UCHL1 IgG Abs or plasma fractions with recombinant UCHL1 prevented podocyte detachment. Plasma levels of anti-UCHL1 IgG Abs were increased in 18/42 relapsing INS patients over the highest level of 38 controls (median = 0.20 AU/μg of total IgG;IQ0.15–0.29;range0.07–0.85). For those 18 patients, the level of anti-UCHL1 IgG Abs in 43 samples available at various stage of the disease was confirmed to be significantly higher in relapse (n = 23;median = 1.22 AU/μg of total IgG;IQ0.92–1.90) compared to remission (n = 20;median = 0.51;IQ0.33–0.77;$p < 0.001$). In those INS patients, proteinuria correlated with anti-UCHL1 IgG Ab level ($n = 43$,$r = 0.57$,$p < 0.001$).
Conclusions: 1/ UCHL1 is involved in the adhesion of cultured podocytes 2/ UCHL1 is a target of circulating IgG Abs in a subset of relapsing INS patients.

O-42 A SINGLE LOW DOSE SCHEDULE OF RITUXIMAB IS NON-INFERIOR TO HIGH DOSE AND MULTIPLE DOSE SCHEDULES IN THE TREATMENT OF STEROID SENSITIVE FREQUENTLY RELAPSING NEPHROTIC SYNDROME

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Introduction: Rituximab is an emerging and effective treatment for children with steroid dependent or frequently relapsing nephrotic syndrome. The optimum dosing schedule for Rituximab has not been established. We hypothesised that a single low dose of 375 mg/m^2 would be non-inferior to higher or multiple doses in reducing the frequency of disease relapse and time to B-cell reconstitution.
Material and methods: This was a multicentre, retrospective, observational cohort study of children with a diagnosis of steroid sensitive frequently relapsing nephrotic syndrome. Data were extracted from clinical records on the dates of diagnosis, treatment and relapses; the use of concomitant immunosuppression; and lymphocyte subset profiling pre-and post-rituximab administration. The primary outcome was an absence of clinically confirmed relapse 12 months following Rituximab administration. Secondary outcomes were median time to relapse, probability of being relapse free at 6 and 24 months, time to reconstitution of CD19$^+$ B cells and the introduction of additional or ongoing immunosuppression.
Results: 60 patients received 143 courses of Rituximab. Patients in group 1 received a higher total dose of 1.5 g/m^2, group 2 received an intermediate dose between 750 mg/m^2 – 1 g/m^2 and group 3 (103 courses) received our current low dose regimen of a single dose of 375 mg/m^2. There was no difference in event-free survival at 6, 12, or 24 months between groups. Of those who relapsed, the median time to relapse was

317 days in group one and 299 days in group three. The median time to reconstitution of B-cells was not significantly different between groups at 175, 226 and 196 days for groups 1, 2 and 3 respectively.
Conclusions: We conclude that usage of a single low dosage regimen of Rituximab in the management of frequently relapsing nephrotic syndrome does not affect the time to B-cell reconstitution or the probability of relapse at 6 and 12 months in our cohort of patients.

O-43 IS RAAS BLOCKADE EFFECTIVE IN NPHS2 GLOMERULOPATHY?

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Introduction: Antiproteinuric therapy with renin-angiotensin-aldosterone system (RAAS) antagonists is an accepted pharmacological approach in hereditary nephropathies. Partial responsiveness to calcineurin inhibition (CNI) has anecdotally been reported. Here we explored changes in albuminemia during RAAS blockade with and without intensified immunosuppression (IIS) in a cohort of children with *NPHS2* glomerulopathy.
Material and methods: We performed a longitudinal PodoNet registry analysis of 40 children with steroid resistant nephrotic syndrome (SRNS) caused by *NPHS2* mutation and documented periods of RAAS antagonist therapy with and without co-treatment with intensified immunosuppression (IIS; 79% CNI) administered before the genetic diagnosis was made. Serum albumin and estimated (e)GFR were assessed before IIS and/or RAAS blockade, during combined RAAS/IIS treatment and during RAAS blockade after IIS discontinuation.
Results: RAAS blockade was started shortly after first disease manifestation (median 2.4 (IQR 0.9–6) months). In 16 patients RAAS antagonists were initiated simultaneously with IIS, whereas in 24 patients RAAS antagonists were added after an average IIS treatment duration of 2.8 (1.2–5.3) months. Combined IIS/RAAS blockade did not change serum albumin significantly (mean 19.7 ± 6.2 - > 20.2 ± 5.6 g/l; $p = 0.19$). Within 12 months after IIS discontinuation but continued RAAS blockade serum albumin increased slightly to a mean of 21.6 ± 6.2 g/l ($p = 0.02$). eGFR decreased by 7 ml/min*1.73m^2 during this period. During subsequent follow-up under continued RAAS blockade (total duration 4.0 (IQR 2.8–5.6) years) serum albumin increased further to a mean of 22.5 ± 5.8 g/l ($p = 0.06$), paralleled by a median eGFR loss by 18 (6–36) ml/min per year. The changes in serum albumin and eGFR were weakly correlated ($r = -0.14$).
Conclusions: Neither IIS nor RAAS blockade nor the two interventions combined appear effective in increasing serum albumin to a relevant degree in patients with *NPHS2* glomerulopathy. Rather, serum albumin gradually increases over time as eGFR deteriorates.

O-44 RISK FACTORS FOR CARDIOVASCULAR COMORBIDITIES IN CHILDREN WITH STEROID-SENSITIVE AND STEROID-RESISTANT NEPHROTIC SYNDROME

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Introduction: The aim of this study is to evaluate cardiovascular (CV) comorbidities and endothelial dysfunction markers and to define risk factors in children with idiopathic nephrotic syndrome (INS) and eGFR >60 ml/min/1.73m^2.
Material and methods: Forty-nine children (19 girls, 30 boys; 5.04 ± 3.68 years) were included in this study. Patients were categorized as steroid sensitive (SSNS; 63%) and steroid resistant (SRNS; 36%) according to definition of KDIGO. Aortic pulse wave velocity (PWV), carotid intima media thickness (cIMT), left ventricular mass (LVM) and ambulatory blood pressure measurements (ABPM) were done; and standard deviation scores (SDS) according to age and height were correlated with anthropometric, cumulative drug dosage, clinical and laboratory parameters including urine protein measures, lipid profile, von-Willebrand factor and sUPAR levels.
Results: Only 13 and 14% had high PWV-SDS and MAP-SDS, respectively; but 73.9% of patients had high cIMT-SDS, 80% had left ventricular hypertrophy (LVH). There were no significant differences between SRNS and SSNS patients for in CVC parameters. BMI-z score was significantly correlated with PWV-SDS (height), mean 24-h arterial pressure (MAP)-SDS (height and age) and LVMI. Corticosteroid exposure for the last 12 months was significantly correlated with cIMT-SDS (age; r: 0.39, p 0.007) and LVMI (r: 0.33, p 0.036). Current cyclosporine dose was significantly correlated PWV-SDS (age), MAP-SDS (age). cIMT-SDS was also significantly correlated with mean platelet volume (r: −0.425, p 0.003). Among patients with LVH 86.2% and 86.7% had normal MAP-SDS and PWV-SDS, respectively. Multivariate analysis revealed high dose corticosteroid exposure for the last 12 months (>35 mg/kg) as the independent risk factor for LVH (RR: 11.02; 95% CI: 1.02–119.49; $p < 0.05$). Serum sUPAR level was correlated MAP-SDS (height) (r: 0.298; p 0.056). vWF level was higher in patients who had more than one CV-abnormality (84.6 ± 39.4% vs.59.7 ± 24.0% p 0.08).
Conclusions: In this cohort, SRNS and SSNS groups were comparable for CV comorbidities. There were associations between CV abnormality and exposure to corticosteroids and cyclosporine for the last 12 months rather than cumulative drug doses and 24-h mean arterial pressure.

O-45 ECULIZUMAB DOSING REGIMEN IN ATYPICAL HUS: POSSIBILITIES FOR INDIVIDUALIZED TREATMENT

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Introduction: Recent studies indicate that eculizumab is often given in excess to aHUS patients. Individualization of treatment is thus highly requested, however, data on pharmacokinetics and pharmacodynamics of eculizumab remain limited.
Material and methods: Serum eculizumab and complement activity (CH50) were measured by in-house ELISA-based methods. In total, 209 samples were taken from 11 patients before the eculizumab infusion in the induction (weekly), maintenance (2-weekly) and tapering (every 3, 4 and 5 weeks) phases of therapy. Statistical analysis was performed using linear mixed models.
Results: The trough eculizumab levels increased with each additional dose during the induction phase (depending on body weight). During maintenance, high eculizumab concentrations of up to 772 μg/mL were observed. The levels decreased with each following dose during tapering (3- and 4-week intervals), however three patients maintained target eculizumab levels over long time periods (30–48 weeks). At intervals of 6–8 weeks target eculizumab levels were no longer attained. Serum samples with eculizumab concentrations ≥50 μg/mL showed full complement blockade.
Conclusions: Our data provide essential insight for optimization of eculizumab dosing schemes and lessening of therapy burden for the patients and cost of the treatment.

O-46 IMPROVED OUTCOMES FOR PAEDIATRIC PATIENTS WITH ATYPICAL HAEMOLYTIC URAEMIC SYNDROME (AHUS) RECEIVING LONG-TERM ECULIZUMAB TREATMENT DURING ON-TREATMENT PERIODS COMPARED WITH OFF-TREATMENT PERIODS

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Introduction: To assess thrombotic microangiopathy (TMA) risk and long-term outcomes in paediatric aHUS patients on eculizumab treatment compared with off-treatment periods.
Material and methods: This is a long-term, observational, follow-up study of paediatric patients with aHUS treated with eculizumab in previous clinical studies (NCT01522170; March 2016 data-cut). TMA rate, change in renal function and targeted serious adverse events (TSAE: serious infections, meningococcal infection, sepsis, leukopenia, infusion reactions, renal or hepatic impairment and malignancy) were assessed.
Results: Thirty-nine patients with a median age of 8.0 years (range 0.0–17.0) at first eculizumab infusion were enrolled in the study. No on-treatment data was collected from four patients who stopped treatment prior to this study. In the primary studies, median time between aHUS diagnosis and first eculizumab dose was 3.1 (range 0.0–191.4) months. Median follow-up duration in this study was 40.6 (range 3.1–84.7) months.
Seventeen (44%) patients had at least one off-treatment period, of whom nine (53%) restarted treatment. During eculizumab on-treatment periods, the protocol-defined TMA manifestation rate was 8.6/100 patient-years vs 29.3/100 patient-years for off-treatment periods (Table). In a post-hoc analysis, when TMA manifestation excluded patients who only had a change in one laboratory criterion, the rates were 2.3 and 22.8 per 100 patient years during on- and off-treatment periods, respectively. When on-treatment, two patients (1.6/100 patient years) developed meningococcal infections vs none off-treatment. Two deaths occurred, one on-treatment (possibly related to treatment) and one off-treatment. No other TSAEs were identified.
Conclusions: This is the largest study of paediatric patients with aHUS treated with eculizumab to date. Eculizumab was generally well-tolerated in paediatric patients throughout the study. Patients in the on-treatment periods had a reduced rate of TMA manifestations compared with off-treatment periods.

O-47 IMPROVED OUTCOMES WITH PROMPT MANAGEMENT OF ANTI-FACTOR H (FH) ASSOCIATED ATYPICAL HEMOLYTIC UREMIC SYNDROME (AHUS) IN CHILDREN: TRENDS FROM NATIONWIDE DATABASE

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Introduction: Patients with anti-FH antibodies, comprising one-half of aHUS in children in India, are usually managed with plasma exchanges (PEX) and immunosuppressive agents. We report the characteristics of patients diagnosed in last 5-years to those before 2012 to assess the impact of early diagnosis and protocolized therapy.
Material and methods: Of 684 patients in the nationwide database, 366 (53.5%) had anti-FH associated aHUS. In addition to intensive PEX over 4–6 weeks, induction therapy comprised of prednisolone and IV cyclophosphamide/ rituximab over 4–5 months, followed by maintenance with MMF/azathioprine for 2-years. Clinical features and outcome were compared in patients presenting during 2005–12 (n = 117) and 2012–17 (n = 249). Adverse outcome was eGFR <30 mL/min/1.73 m^2 or death.
Results: Anti-FH disease was diagnosed four-times more often in last 5-years. Patients with anti-FH HUS were older, had severe illness and better outcomes than those without antibodies. Median decline of antibodies was 74, 88 & 84% after 3, 5 & 7 PEX, respectively (P = 0.08); titers were similar in those receiving IV cyclophosphamide or rituximab (generalized estimating equation, GEE; P = 0.6). Combined PEX & induction immunosuppression was associated with improved long-term outcomes (HR 2.7; P = 0.001); maintenance therapy reduced risk of relapses (HR = 2.7; P = 0.008). Patients presenting in last 5-years showed less oliguria, seizures and hypertension, indicating benefits of early diagnosis and therapy (Table). Prompt PEX and immunosuppression enabled faster hematological & renal recovery. Adverse outcome at 3-months was decreased, with better 4-years renal survival. Patients with relapse (n = 27) had higher antibodies in remission than those who did not (n = 91) during 5-years follow up (GEE, P = 0.015); titer >1300 AU/ml at 6-months predicted relapses.

Table | Clinical features and outcomes of anti-FH associated aHUS

	2005–12 N = 117	2012–17 N = 249	P
Age	8 (6–10)	7.5 (6–10)	0.9
Prodrome	69 (59%)	159 (64%)	<0.0001
Oligoanuria, days	10 (5–15)	5 (2–10)	<0.0001
Extrarenal symptoms	55 (47%)	139 (56%)	0.12
Stage 2 hypertension	82 (70%)	118 (47%)	<0.0001
C3, mg/dl	65 (48–85)	70 (54–87)	0.16
Anti-FH, AU/ml	2170 (950–7250)	3400 (1180–11,70-6)	0.03
Days to PEX	19 (8–33)	12 (6–23)	0.026
Days to immunosuppression	34 (20–56)	22 (12–33)	0.001
Days to hematological remission	38 (23–55)	25 (17–36)	<0.0001
Duration of dialysis, days	31 (10–51)	11 (4–29)	<0.0001
Duration of PEX, days	36 (18–59)	28 (14–42)	0.006
CKD stage 2–3 @3-months	17 (16.5%)	24 (17.6%)	0.81
Adverse outcome @3-months	33 (32%)	24 (17.6%)	0.009
Adverse outcome @4-years	46 (39%)	65 (26%)	0.3

Median (IQR) or N (%)

Conclusions: Findings from this large nationwide database suggest that prompt recognition and specific therapy for anti-FH associated HUS, with PEX and immunosuppression, is associated with satisfactory outcomes without necessitating the use of complement inhibitors.

O-48 GENOMIC REARRANGEMENT EVENTS WITHIN COMPLEMENT FACTOR H RESULTING IN ATYPICAL HAEMOLYTIC URAEMIC SYNDROME

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Introduction: Atypical Haemolytic Uraemic syndrome (aHUS) is the triad of microangiopathic haemolytic anaemia thrombocytopenia and acute kidney injury. aHUS most commonly occurs due to inherited mutations in the alternative complement cascade, the most frequent mutations are in complement factor H (*CFH*). Mutations in *CFH*, resulting in aHUS, predominantly reside in the C-terminus, resulting in impaired surface binding. *CFH,* along with five highly homologous *CFH* related genes (*CFHR1–5*), is located within the Regulators of Complement Activation cluster. Due to the high degree of sequence homology, this region is prone to genomic rearrangement resulting in hybrid genes (*CFH::CFHR1* and *CFHR1::CFH*). CFH::CFHR1 hybrids result in the exchange of the C-terminus of CFH with the C-terminus of CFHR1 (impairing surface binding), whilst CFHR1::CFH hybrids lead to replacement of the C-terminus of CFHR1 with the C-terminus of CFH (impairing complement regulation).
Material and methods: All patients referred to the UK's National aHUS centre with a diagnosis of aHUS ($n = 984$) underwent Sanger sequencing of CFH to identify point mutations, and multi-ligation probe amplification (MLPA) to identify genomic rearrangement. Genomic rearrangement events generating CFH::CFHR1 hybrid were identified due to loss of CFH Ex.22/23 and gain of the corresponding region of CFHR1, Ex.5/6. CFHR1::CFH hybrids were identified due to loss of CFHR1 Ex.5/6 and gain of CFH Ex.22/23.
Results: Sanger sequencing identified 86 patients with pathogenic variants in *CFH*. Analysis of the MLPA data revealed an additional 40 patients with either *CFH::CFHR1* or *CFHR1::CFH* hybrids.
Conclusions: Genomic rearrangement of CFH is a common cause of aHUS. These events may not be identified by standard Sanger sequencing as the regions used to sequence are frequently involved in the rearrangement, resulting in alelle dropout. MLPA analysis can be used to identify genomic rearrangements, through addition or loss of CFH Ex.22/23 and CFHR1 Ex.5/6 sequence. This data highlights the need to perform MLPA analysis in patients with aHUS.

O-49 TARGETED EXOME SEQUENCING OF A COHORT OF 204 PATIENTS IDENTIFIES PBX1 AS A NOVEL GENE INVOLVED IN MONOGENIC CAKUT

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Introduction: CAKUT (Congenital Anomalies of the Kidney and Urinary Tract) are major causes of chronic kidney disease in children. They are phenotypically and genetically heterogeneous diseases. Monogenic causes of CAKUT in humans, as well as in mouse, have been identified, with more than 50 genes reported as mutated, mostly in syndromic forms. Most of the mutations are heterozygous, with autosomal dominant inheritance and variable expressivity. The most frequently mutated genes are *HNF1B, PAX2, EYA1* and *SIX1,* all encoding transcription factors. Many of the other genes are mutated in only few patients and their implication is sometimes elusive.
Material and methods: We developed a targeted exome sequencing strategy (« cakutome ») focusing on 330 genes, either known to be involved in CAKUT or being candidates (genes whose knock-out in mouse lead to CAKUT, genes involved in cellular processes/signaling pathways relevant for kidney development), in a cohort of 204 unrelated CAKUT cases, 45% of which were severe fetal cases.
Results: This approach allowed us to identify heterozygous loss-of-function mutations/deletions in *PBX1* (Pre-B-Cell Leukemia Transcription Factor 1), a gene reported to play a crucial role in kidney development in the mouse, in 5 cases with syndromic (4 patients) or isolated (1 fetus) renal hypodysplasia. We showed that all the mutations (including a nonsense, a frameshift and a splice mutation and 2 large deletions encompassing *PBX1* and additional genes) occurred *de novo*. *PBX1* is thus a novel gene involved in monogenic CAKUT in humans.
We also identified pathogenic mutations and copy number variations in known CAKUT genes: heterozygous mutations/deletions in *HNF1B* (9 cases), *PAX2* (9 cases), *EYA1* (5 cases), *GATA3* (3 cases), *ANOS1* (2 cases) and *CHD7* (1 cases), and biallelic mutations in *KIF14* (2 fetuses with renal hypodysplasia and microcephaly), thus providing a genetic diagnosis in 15% of the cohort. The rate of deletion (removing one or several exons) was quite high (47%).
Our results also led us to call into question the role of some variations in *SOX17* and *DSTYK* recently reported as pathogenic in CAKUT.
Conclusions: This targeted exome sequencing strategy thus proved to be efficient and cost-effectiv, and allowed the identification of *PBX1* as a novel CAKUT gene.

O-50 GENOMIC IMBALANCES IN CHILDREN WITH CHRONIC KIDNEY DISEASE

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Introduction: Chromosomal microarrays are routinely utilized for genetic testing of developmental delay/intellectual disability, autism spectrum disorders or multiple congenital anomalies. Here, we studied the prevalence of DNA copy number variations (CNVs) in a large cohort of European and Turkish children presenting with chronic kidney disease.
Material and methods: 986 consecutive patients enrolled in the ESCAPE and 4C studies were genotyped using 2.4 million SNP Illumina microarray. Data was interpreted in accordance to ACMG Practice Guidelines using the Nexus Copy Number software. Filtering criteria included the size of imbalance, overlap with known benign CNVs, gene content, and verification against Decipher and ISCA databases.
Results: 890 (90.3%) samples were eventually eligible to detailed genotype-phenotype correlations, of these 74 (8.3%) were classified as having a definite pathogenic genomic aberration and another 11 as having a likely pathogenic CNV. In addition, 34 patients were found to have a heterozygous variant in a known AR gene associated with a hereditary kidney disorder. Definite diagnoses were made in 4.7% of individuals with CAKUT, 2.8% of patients with a glomerulopathy and 27.9% of those with a tubulointerstitial disorder.
The most frequent imbalances were chromosome 2q13 homozygous deletions of the *NPHP1* region (n = 37), 1q21.1 deletions including the *CHD1L* gene (n = 7), rearrangements at 22q11.2 (n = 7) and 17q12 deletions encompassing the *HNF1B locus* (n = 6). Small, including single gene, rearrangements were reported for 56 genes including *CTNS, PKD1, TSC2, ROBO2, NOTCH2* and *TFAP2A loci*.
Conclusions: Clinical diagnosis was revised in 10 CAKUT cases (HNF1ß nephropathy (n = 5); nephronophthisis (n = 5)) and in one case of clinically suspected Bardet-Biedl syndrome (nephronophthisis). The detection of a genomic imbalance allowed for reverse phenotyping in most cases, resulting in clinical interventions such as evaluation for extra-renal involvement and implementation of multidisciplinary care.
Genomic imbalances account for a significant portion of children with CKD and their diagnosis has important implications for genetic counseling and clinical management.

O-51 CLINICAL MANIFESTATIONS OF SYSTEMIC OXALOSIS IN PRIMARY HYPEROXALURIA TYPE 1: WHEN DO WHICH CLINICAL MANIFESTATIONS OCCUR

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Introduction: Description of the manifestations of systemic oxalosis in a large European cohort of patients with Primary Hyperoxaluria type 1 (PH1) and analysis of eGFR, plasma oxalate and other potential clinical thresholds for the occurrence of systemic oxalosis.
Material and methods: Review of all PH1 patients registered in the OxalEurope database, which now includes more than 900 PH patients. We have performed a sub analysis of patients in whom data of sufficient detail was obtained. A more comprehensive dataset will be available in the near future.
Results: Of the 132 included patients, 51 (38.6%) were found to have at least one manifestation of systemic oxalosis. Bone disorders represented the most frequent manifestation (18.9% (25/132) at diagnosis and cumulatively 30.3% (40/132) at follow up), followed by cardiac (3.8%, 15.2%), cutaneous- and vascular (3.8%, 15.4%), ophthalmologic (7.6%, 12.9%), neurological (4.5%, 8.3%) amongst other manifestations. We found 31 different combinations of symptoms. The majority of manifestations (94%, 48/51) were found in patients with an eGFR <15 ml/min/1.73 m2. PH patients were not routinely screened; e.g. 26.5% had not undergone any ophthalmologic evaluation. We report the first patient with manifestations of oxalosis, an eGFR above 50 ml/min/1.73 m2 and plasma oxalate level below 30 μmol/l.
Conclusions: This study highlights the heterogeneity of systemic oxalosis. The high number of reported systemic manifestations of oxalosis might be an underestimate due to the large number of non-systematically screened asymptomatic patients. Evidence of systemic oxalosis in a patient with moderate CKD warrants attention. Our results challenge the current assumption that systemic deposition of oxalate starts when the plasma oxalate level is >30 μmol/l and the eGFR <40 ml/min/1.73 m2. Therefore, we stress the value of annual screening for systemic oxalosis in PH1 patients with CKD2+.

O-52 FAMILIAL MEDITERRANEAN FEVER IN PATIENTS DIAGNOSED WITH FABRY DISEASE PREVALENCE

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Introduction: Familial Mediterranean Fever (FMF) is the most common hereditary periodic fever syndrome, affecting the populations surrounding Mediterranean region. Mainly it is mostly seen in Turks, Armenians, Arabs and Sephardic Jews. It is an autoinflammatory disease, which is inherited OR with recurrent abdominal pain, peritonitis, which can be watched with arthritis and skin lesions, lasting in 6–72 h, characterized by amyloidosis in time. Disease responsible gene locus is located on chromosome 16 and is called the short arm MEFV. Colchicine is the standard treatment to prevent attacks and amyloidosis, which is a serious complication of the disease.Fabry Disease (FD) has X linked transition and is a lysosomal storage disease characterized by progressive accumulation of glycosphingolipids in various tissues and organs, caused by the mutation in the GLA gene and alpha-galactosidase A enzyme deficiency. The enzyme activity should be checked when the disease is suspected clinically.Ten years of time between diagnosis and the onset of symptoms in these patients is due to the presence of organ damage and systemic symptoms which can be confused with systemic diseases. Therefore if there is an ethnicity with in FD patients which have these symptoms, mistakenly placed diagnosis of FMF and the FD does not mind too often. Because it can be prevented, it is important to initiate early treatment. This study was aimed to create awareness for the FD.
Material and methods: One hundred patients which are diagnosed FMF according to Tell-Hashomer criterias and have MEFV gene analysis at Celal Bayar University Hospital Pediatric Nephrology Department are included in this study.
Gal A activity analysis Measurement of enzyme activity in dried blood spot samples was performed via DBS method using filter paper containing DBS as a source of DNA. Blood samples were collected before dialysis in hemodialysis patients and at any time in peritoneal dialysis patients. Four drops of the blood were transferred to a filter paper, allowed to dry at room temperature, and kept at 2–4 °C until analysis. The enzyme activities were calculated in μmol/L/h or pmol/spot*20 h. Patients with values <1.2 μmol/l/h or <200 pmol/spot*20 h were considered to have low α-Gal A activity.
GLA mutation on genetic analysis In patients with low α-Gal A activity, screening of the GLA mutation was performed using DBS cards based on Sanger sequence analysis. This assay uses PCR amplification followed

by Sanger DNA sequencing to detect mutations in the GLA gene that cause Fabry disease. The gene encoding GLA is found on Xq22, and spans 13 kb of genomic DNA (7 exons, cDNA of 1290 bases). The GLA gene encodes a 429 amino acid protein, of which the first 31 residues form a lysosomal signal peptide. Classical phenotypes are typically caused by misssense, nonsense, severe splicing mutations and large gene defects. Variant phenotypes are typically caused by splicing defects that express residual enzyme activity. The coding sequences and flanking intronic sequences (minimum of 20 base pairs) of exons 1–7 of the GLA gene are amplified from purified genomic DNA and sequenced in the forward and reverse directions. Sequencing of a single exon is available for targeted mutation analysis. Patient sequences are compared to the reference DNA sequence (GenBank Accession: X14448, NM_000169.1).

Results: Thirty-five percent of the patients in this study were male and 65% female. Male / female ratio was 1 / 1.8 was found. The mean age of onset of symptoms is 5.89 ± 3.35. FMF diagnosed family member was found in %59 of patients. The most common symptoms of patients with initial symptoms include fever (89%) and abdominal pain (94%). Then follows arthralgia (51%), arthritis (20%), chest pain (30%), erysipelas (9%). When the M694 V is the most common allele frequency rate of 40.5% and respectively others are E148Q 11%, V726A 7%, M680 5.5%, R202Q 5%, R761H 4.5% found. The most common genotypes are respectively M694 V homozygotes, E148Q heterozygotes and M694 V heterozygotes. The main symptoms that are common with other gastrointestinal symptoms of Fabry patients which are encountered during the course of the disease are nausea %28, diarrhea %13 and constipation %15. Incidence of neurological symptoms are headache 32%, akroparesthesis 23% and tinnitus 16%. Alpha-galactosidase A enzyme activity in 2 male patients from the study group were significantly lower but in the GLA gene analysis it wasn't found a mutation that belong to FD.

Conclusions: Sometime at first in FD patients which have FMF like symptoms are unnecessaryly treated with colchicine medication. Therefore patients which are diagnosed as FMF and do not well respond to colchicine treatment should be carefully re-examined clinically. Because FD can be prevented, it is important to initiate early treatment.

O-53 FEASIBILITY AND TOLERABILITY OF SUSTAINED LOW EFFICIENCY DIALYSIS (SLED) IN CRITICALLY SICK PEDIATRIC PATIENTS: A MULTICENTRIC RETROSPECTIVE STUDY

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Introduction: Sustained low-efficiency dialysis (SLED) has emerged as an alternative to CRRT in the management of hemodynamically unstable adult patients with AKI. This was a retrospective record review from three major centres in the country from Jan 2010 to June 2016. This is the largest ever data on Pediatric SLED published till date. The objective of the study was to document the SLED practices in these centres, and to look at the feasibility and tolerability of SLED in critically sick pediatric patients.

Material and methods: All pediatric patients undergoing SLED in the collaborating centres were included in the study and the basic demographic data, prescription parameters and outcomes were recorded.

Results: From January 2010 to June 2016, a total of 68 children received 211 sessions of SLED at three major centres in the country. PRISM score at admission in patients was 13.33 ± 9.15. Fifty seven patients were ventilated (84%). Most of the patients had one more organ system involved in addition to renal (n = 64; 94%). Heparin free sessions were done in 153 sessions (72%). There was no statistical change in mean blood pressure in all patients before or after sessions of SLED. Though there was no statistical difference in oxygenation index of patients pre and post SLED, there was an improvement in serum bicarbonate. Premature terminations had to be done in 27 sessions (13% of all sessions), out of which 7 sessions had to be terminated due to circuit clotting (3.3%). Intradialytic Hypotension or need for more ionotrope escalation was seen 31 sessions out of which terminations were done in 20 sessions. Twenty nine patients died due to sepsis and MODS.(Table 1).

Table 1 - Patient characteristics by survival status

Parameters	Value (68 patients; 211 sessions)
Premature session termination	27 (13%)
Sessions with temperature alteration	7 (3.3%)
Pre SLED mean BP (mm of Hg)	82.31 ± 13.76
Post SLED mean BP (mm of Hg)	80.81 ± 14.54
Pre SLED mean number of inotropes	1.19 ± 1.25
Post SLED mean number of inotropes	1.16 ± 1.25
Pre SLED bicarbonate (mg/dl)	17.85 ± 4.64*
Post SLED bicarbonate (mg/dl)	21.19 ± 3.78*
Post SLED hypophosphatemia (sessions;%)	6 (3%)
Post SLED hypokalemia (sessions;%)	59 (28%)
Pre SLED oxygenation index [n = 61 patients]	14.56 ± 5.91
Post SLED oxygenation index [n = 61 patients]	11.75 ± 6.45

*$p < 0.05$.

Conclusions: SLED appears to be a feasible and also tolerable method of providing renal replacement in critically ill pediatric patients.

O-54 RUNNING A SUCCESSFUL PAEDIATRIC HOME HAEMODIALYSIS SERVICE: OVERCOMING TREATMENT RISKS AT HOME

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Introduction: Home haemodialysis (HHD) has clear advantages, including reduced cardiovascular morbidity, liberalisation of fluid and dietary restrictions and improved quality of life compared to conventional in-centre dialysis. However, haemodialysis is not a risk free treatment and managing these risks at home along with addressing carer burden are essential for a successful HHD service. We describe the measures instituted in 2 paediatric centres to address the potential risks of HHD treatment.

Material and methods: A review of treatment complications as well as measures implemented to minimise these risks at both centres was undertaken.

Results: To date, a total number of 34 patients have participated in the HHD service at both centres. Treatment complications have been minimal with 2 cases of line sepsis in the past 3 years; 2 cases of suspected air

embolism, 2 thrombophlebitis episodes and no line dislodgements or other fistula related complications or deaths. Three patients have returned to in-centre haemodialysis. Fourteen dialysis machines have required replacement, due to equipment failure, in the past 3 years. A robust HHD plan with parameters to seek medical help and emergency procedures along with basic life support training is given in every case. Due to proximity of local hospitals to patient homes, each patient has community children's nursing support, a named consultant with an open access policy and provision of emergency protocols at a local hospital. Ambulance services are now routinely notified with a medialert alarm system being incorporated in some homes.
Conclusions: Whilst HHD has clear benefits for patients and families, treatment risks are real and a robust training programme with supportive and emergency measures must be in place to ensure patient safety and minimise carer burden. Risks of treatment at home can thus be outweighed allowing a successful and sustainable paediatric HHD service.

O-55 A RANDOMIZED, DOUBLE-BLIND, PLACEBO-CONTROLLED STUDY TO ASSESS THE EFFICACY AND SAFETY OF CINACALCET IN PEDIATRIC SUBJECTS WITH CHRONIC KIDNEY DISEASE AND SECONDARY HYPERPARATHYROIDISM RECEIVING DIALYSIS

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Introduction: This study evaluated the efficacy of cinacalcet for reducing plasma intact parathyroid hormone(iPTH), the impact on albumin-corrected Ca(cCa) and phosphorus(P); safety and tolerability of cinacalcet.
Material and methods: Eligible subjects (6to < 18 years) on chronic dialysis ≥ 2 months, with PTH > 300 pg/mL, Ca ≥ 8.8 mg/dL and $P \geq 4.0$ mg/dL (6to < 12 years) or ≥3.5 mg/dL (12to < 18 years) were randomized 1:1 to cinacalcet or placebo stratified by age group (<12and ≥ 12 years). Cinacalcet was started at ≤0.20 mg/kg/day and titrated (max 4.2 mg/kg/day) once every 4 weeks over 24 weeks followed by a 6-week efficacy assessment phase (EAP). Plasma iPTH, serum cCa, and P were collected bi-weekly during titration and EAP.
Results: 43/100 planned subjects were enrolled and received ≥1 dose of investigational product (22 cinacalcet; 21 placebo). The study was terminated early after consultation with regulatory authorities following a fatality in a subject with severe hypocalcemia receiving cinacalcet. The cause of death was determined to be multifactorial, but a causal role for hypocalcemia could not be excluded. Baseline demographics and disease characteristics were balanced. 54.5%(12/22) cinacalcet and 19.0%(4/21) placebo subjects achieved ≥30% reduction from baseline in mean PTH during the EAP (study primary endpoint), with a difference of 35.5%(95%CI: 8.76%, 62.24%), based on last observation carried forward imputation (CMH test $p = 0.017$). Numerically reduced cCa and similar P were observed in subjects who received cinacalcet. Adverse events (AE), including hypocalcemia, were reported in similar proportions in the cinacalcet and placebo arms during the double-blind phase.
Conclusions: The observed treatment effect of cinacalcet in reducing plasma iPTH was clinically meaningful and statistically significant despite early termination of the study. The observed AEs were generally consistent with the known safety profile for cinacalcet.

O-56 HEMODIALYSIS IN SMALL CHILDREN - DATA FROM THE INTERNATIONAL PEDIATRIC HEMODIALYSIS NETWORK

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Introduction: Hemodialysis in infants and toddlers is considered a reserve technique and has not been evaluated in detail.
Material and methods: We compared treatment characteristics and outcomes of hemodialyzed children younger than 3 years with older children and adolescents dialyzed via CVL and prospectively followed by IPHN.
Results: Among 395 patients, 46 (12%) started chronic HD before their 3rd birthday (median 1.5, range 0.1–2.9 yrs). During 576 months a total of 87 CVL were placed in these infants, thereof 86%into the internal jugular vein. Compared to children aged 3 years and older, younger patients had longer weekly dialysis duration (12.1 ± 3.8 vs 11.4 ± 2.8; $p = 0.02$) and more frequent sessions (3.8 ± 3.5 vs 3.0 ± 2.9/week; $p < 0.001$). Neither blood flow (157 ± 62 vs 157 ± 49 ml/min/m^2), Kt/V (1.79 ± 0.82 vs.1.65 ± 0.59 per session), nor the prevalence of untoward HD effects differed from older patients (intradialytic hypotension 34 vs 27%, vomiting 12 vs 8%, seizures 1 vs 3% of sessions).
The interdialytic weight gain was lower (2.8 ± 2.3% vs 3.7 ± 3.5%; $p < 0.001$) but systolic blood pressure higher in the infant group (2.48 ± 1.6 vs 0.79 ± 2.54 SDS, $p < 0.001$), at a similar prevalence of anuria and no difference in LV mass index SDS. Hyperphosphatemia and hyperparathyroidism were less common in the infant group ($p < 0.0001$). There were no differences in hemoglobin, transferrin saturation, malnutrition prevalence or serum albumin. EPO requirements were higher in younger children (415 vs 256 IU/kg/wk., $p < 0.0001$). Hospitalization days (3 vs 15 per 100 days), catheter dysfunction and the rates of infection were significantly higher in the infants than in older children(2.7 vs 0.9 and 1.8 vs. 0.7 per 1000 catheter days; $p < 0.001$).The mortality rate did not differ between the groups.
Conclusions: HD is a safe and effective technology for infants, but associated with higher morbidity and access related complications.

O-57 EUROPEAN SURVEY ON DIAGNOSIS AND MANAGEMENT OF CRESCENTIC GLOMERULONEPHRITIS IN CHILDREN

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Introduction: Crescentic glomerulonephritis can be caused by different pathogenic mechanisms with severe glomerular damage. Different

approaches can impact the success of the management of crescentic glomerulonephritis. Our objective is to obtain data on the etiology, diagnosis and management of crescentic glomerulonephritis across Europe.
Material and methods: A web-based questionnaire was prepared by ESPN Working Group for Immune Mediated Renal Disorders and questionnaire results were evaluated.
Results: One hundred active members of ESPN from 19 countries participated in the study. Ninety-one percent of them are working in a university hospital and they have required facilities for the diagnosis of crescentic glomerulonephritis. Crescentic glomerulonephritis etiology is related with immune-complex mediated in 89.61% of the patients, pauci-immune mediated in 83.12 of them, anti-GBM nephritis in 58.44% of them and 20.78% of all patients are related to other etiologies. Ninety-seven percent of them takes advantage of pathological findings for the diagnosis and they prefer pathological findings to clinical and other laboratory findings. Glucocorticoids (100%) and cyclophosphamide (75.31%) are the first choice of treatment for induction therapy. Plasma exchange (66.67%), mycophenolate mofetil (29.63%), rituximab (27.16%) and azathioprine (1.23%) are other treatment modalities. Mycophenolate mofetil is mostly used medication (85.19%) for maintenance therapy followed by glucocorticoids (83.95%), azatioprine (41.98%), rituximab (22.22%).
Conclusions: Although approaches for diagnosis and management of crescentic glomerulonephritis are similar across Europe, a consensus will increase the success of the management of the crescentic glomerulonephritis.

O-58 URINARY NEUTROPHIL GELATINASE-ASSOCIATED LIPOCALIN (NGAL) AND SERUM CYSTATIN C MEASUREMENTS FOR EARLY DIAGNOSIS OF ACUTE KIDNEY INJURY IN CHILDREN

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Introduction: Acute kidney injury (AKI) is common in critically ill children with significant mortality and morbidity. Serum creatinine is insensitive and late biomarker compared to newly proposed AKI biomarkers.
Material and methods: Prospective study in pediatric intensive care unit (PICU) over three months to compare between serum cystatin-C(Cys-C) and urinary neutrophil gelatinase-associated lipocalin (uNGAL) as AKI biomarkers at multiple time points with pRIFLE classification in diagnosing AKI.
Results: Forty children were recruited. Twenty two of them developed AKI according to pRIFLE criteria. There was difference between AKI and non-AKI in age; mean ± SD 38.1 ± 39.5 vs55.5 ± 58.0 (p value 0.29). Post cardiac surgery renal insult was the main cause of AKI (27.3%). There was 2-fold increased risk of incident AKI in those patients with high baseline uNGAL at PICU admission and almost 4-fold increased risk in patients with high baseline cystatin-C at PICU admission. uNGAL levels were highly predictive of AKI during the follow-up period, Area Under the Curve(AUC 0.76, 95% CI: 0.61–0.92) The cutoff point with the highest correctly classified proportion was 223 ng/ml (≥ 12 centiles) which correctly predict 80.0% patients with AKI, with a corresponding sensitivity of 72.7% and a specificity of 89.9%. AUC for serum cystatin-C was 0.86 (95% CI: 0.75–0.97), and the highest correctly classified proportion was 1009 ugL (≥ 13 centiles); 75% of patients with AKI, with a corresponding sensitivity of 63.6% and a specificity of 88.9%.
Conclusions: uNGAL and serum cystatin-C predicts AKI early in critically ill children.

O-59 ECULIZUMAB TREATMENT ON RENAL FUNCTION IN PAEDIATRIC AHUS PATIENTS WITH KIDNEY TRANSPLANT: DATA FROM THE AHUS GLOBAL REGISTRY

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Introduction: To analyse the need for dialysis in paediatric patients with atypical haemolytic uraemic syndrome (aHUS) and kidney transplant (KTx) who received eculizumab either pre- or post-transplantation.
Material and methods: Data from the Global aHUS Registry (NCT01522183) on eculizumab-treated paediatric patients (<18 years at most recent KTx) with at least 1 KTx and ≥1 year of observation post-KTx were analysed for the incidence of dialysis events post-transplant. Patients were grouped by timing of eculizumab treatment: ongoing treatment (≥1 dose) up to time of KTx (pre-KTx; n = 18) vs treatment initiation post-KTx (n = 19).
Results: As of August 2016, 1286 patients with aHUS were enrolled in the registry; data from 37 paediatric KTx patients are reported here. Patients had a median age at diagnosis of 5.2 years, 78% were male, 30% had a family history of aHUS and 16% had ≥2 KTx. Demographics and clinical characteristics were similar between treatment groups (Table). aHUS diagnosis was made prior to KTx in 31/37 patients and after KTx in six patients (all in the eculizumab treatment post-KTx group). One of 18 patients treated with eculizumab pre-KTx required short-term (lasting <3 months) dialysis (incidence rate: 1.3/100 patient-years). Six of 19 patients receiving eculizumab post-KTx required 16 periods of dialysis following KTx (incidence rate: 4.4/100 patient-years). Two of these six patients received dialysis for more than 3 months.
Conclusions: We provide the first real-world data on the association of the timing of eculizumab treatment on the need for post-KTx dialysis in paediatric patients with aHUS. This descriptive, retrospective analysis suggests that eculizumab pre-KTx may decrease the incidence of dialysis post-KTx when compared to eculizumab post-KTx in paediatric patients with aHUS. Further studies are required to confirm whether initiating eculizumab prior to transplant reduces dialysis events in children with aHUS following KTx.
Acknowledgments: We wish to thank the patients and registry investigators.

O-60 CARDIOVASCULAR COMPLICATIONS IN ATYPICAL HAEMOLYTIC URAEMIC SYNDROME IN CHILDREN

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Introduction: We estimated the incidence of cardiovascular complications (CVC) in children with atypical HUS(aHUS) and their outcomes depending on the type of therapy.

Material and methods: One hundred four patients with aHUS were examined at the age of 2.5 months-17 years from 1999to2017. Genetic screening was performed in 17(16.3%) patients, among which in 2 cases CFH mutation was detected, in 1 case - CFI and C3 mutations, in 8 - complement genes polymorphisms. In 8(7.7%) cases anti-CFH antibody HUS was diagnosed. Echulizumab therapy was performed in 65(62.5%) patients.
Results: CVC in the acute period of aHUS were detected in 74(71.2%) patients: arterial hypertension(AH) in 52(70.3%), left ventricular dilation(LVD) in 27(36.5%), left atrial dilatation - 3(4.1%), decreasing of ejection fraction(EF) - 15(20.3%), ischemic manifestations and coronaropathy - in 6(8.1%). In 7(9.5%)cases cardiac glycosides were required. Two patients died in the acute period and seven in the remote one. CVC were associated with severity of acute kidney injury(AKI):creatinine and proteinuria were higher than in the group without CVC(439.9 ± 250.4 vs 307.9 ± 280.2 μmol/l, 6.3 ± 5.2 vs 1.6[0.3,3.3],$p < 0.05$). Eculizumab was assigned to 49(66.2%) patients with CVC, of whom 22 patients(44.9%) in the acute period of the aHUS, 27(55.1%)-in hematologic remission, but persistent organ damage.Among patients with aHUS manifestation before 2012,CVC were detected more often than in the group of children who fell ill after 2012 because of access to eculizumab:85%vs50% ($p < 0.05$). Among 7 patients with aHUS and CVC who did not receive eculizumab, only 1 with AKI was resolved; 2 children died because of the recurrence of aHUS, in 4 - persistent AH remained. In 10 cases, the intensity of LVD decreased, LF was normalized. Chronic CVC were formed in 48(64.9%) patients with aHUS, 12 of them did not receive eculizumab: AH - in 42(87.5%), LVD in 16(33.3%), LV hypertrophy - 5(10.4%), EF < 60%-in4(8.3%), ischemic manifestations - 2(4.2%), ventricular extrasystoles - 2(4.2%), dilatation of the aorta, valve stenosis, compaction of the walls - 7 (14.6%). Three patients died from the CVC: 2 - from acute cardiovascular failure, 1 - rupture of aortic aneurysm.
Conclusions: CVC are frequent extrarenal manifestation of the aHUS. The high frequency of CVC chronization in children with aHUS emphasizes the need for timely targeted therapy. The differentiation between CVC caused by TMA and secondary phenomena due to volume overload and/or hypertension is important, but not always clearly.

O-61 DETECTION OF CONVERTASE-STABILIZING FACTORS IN PATIENTS WITH COMPLEMENT-MEDIATED RENAL DISEASES

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Introduction: The autoantibody C3 nephritic factor (C3NeF) plays a pathogenic role in C3 glomerulopathy (C3G) by stabilizing the key enzyme of complement activation, the C3 convertase. However, reliability of currently used assays to detect C3NeF is limited. Recently, we developed a method to measure convertase stability in whole human serum and we now optimized the method for simple detection of convertase-stabilizing factors such as C3NeF in large patient cohorts.
Material and methods: Convertase stability was measured in a hemolytic assay using the C5-blocking agent eculizumab to separate the alternative pathway (AP) into two steps: formation of C3/C5 convertases by test sera in step 1 (a time-variable step) and formation of lytic membrane attack complexes in a standardized second step for readout. Samples of 15 controls, 33 patients with (suspected) C3G or closely related disorders, and family members with Factor B (FB) mutation (p.Lys323Glu) and atypical hemolytic uremic syndrome (aHUS) were analyzed.
Results: Healthy controls were tested to define the normal convertase activity profile: maximal convertase activity was reached at $t = 10/15$ min and activity returned to background levels from on $t = 30$. When serum or purified Ig fraction containing C3NeF was added to control serum, convertase stability was increased at $t = 30$ min ($P < 0.001$). Thus, detectable convertase activity at $t = 30$ min or later was chosen as a marker for presence of convertase-stabilizing factors such as C3NeF. In our cohort, 17 out of 33 (52%) patients showed increased convertase stability. Interestingly, prolonged convertase activity was also detected in an aHUS family and segregated with the FB mutation in affected and non-affected family members.
Conclusions: We present optimization of a simple, reliable, and cost- and time-effective assay for detecting convertase-stabilizing factors (C3NeF and some mutations) in patients with various complement-mediated renal diseases. This study may give insight in disease pathogenesis and treatment strategies in these patients.

O-62 CLINICAL OUTCOMES IN CHILDREN WITH HENOCH–SCHöNLEIN PURPURA NEPHRITIS WITHOUT CRESCENTS

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Introduction: Henoch-Schonlein purpura is the most common vasculitis in children. Its long-term prognosis depends on renal involvement. The management of Henoch-Schonlein purpura nephritis (HSPN) remains controversial. This study reports the prognosis of children with HSPN presenting with class 2 ISKDC nephritis.
Material and methods: All children with HSPN Class 2 diagnosed between 1995 and 2015 in four pediatric nephrology centers were included and clinical and biological data were collected from the medical files. The primary endpoint was the remission of proteinuria defined as a proteinuria <200 mg/L.
Results: Ninety-two children were included with a median follow-up time of 36 months. Twenty-eight percent had nephrotic syndrome, 31% proteinuria > 3 g/L, 52% proteinuria between 1 and 3 g/L and 18% proteinuria <1 g/L. Forty-seven percent received treatment with oral steroids alone, 37% received methylprednisolone pulses followed by oral steroids, 18% have not been treated with steroids. Eighty-five percent reached remission during follow-up but 12% of them did not maintain complete remission over time so that only 75% remained in complete remission by the end of the follow-up. Univariate analysis found a linear increase of the likelihood of remission with initial proteinuria ($p = 0.009$). This trend was not found in the multivariate analysis after adjusting for treatments as patients with higher proteinuria were most often treated with steroids.
Conclusions: Our study underlines that one fourth of the patients with HSPN class 2 remains proteinuric and thus carry the risk of developing chronic kidney disease on the long term. This finding together with the better outcome of patients who have received steroids is in favor of treating those patients.

O-63 STEROID DEPENDENT AND FREQUENTLY RELAPSING NEPHROTIC SYNDROME IS ASSOCIATED WITH PERSISTENT DISEASE INTO ADULTHOOD

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Introduction: We conducted a retrospective clinical database on children with idiopathic nephrotic syndrome (INS) to evaluate the long-term clinical course and outcome.
Material and methods: Patients with new-onset INS admitted to the paediatric departments in the central and northern region of Denmark between January 1998 and December 2015 were included in the study. Patients were identified by a search on ICD-10 codes in the Danish National Patient Register. All patients (n = 128) identified from the search were asked to provide with written informed consent, and data were obtained from the medical charts. Active disease above 18 years was defined as relapse within 12 months or ongoing immunosuppressive treatment.
Results: The annual incidence of INS in this cohort was 1.9/100,000 children aged 0–14 years. Mean age at debut was 6.9 years (range 1.6–14.5 years), and the male:female ratio was 1.2:1. Mean follow-up was 9 years. A total of 84% (92/110) patients were classified as steroid sensitive (SSNS), of whom 60% (55/92) were either steroid dependent and/or frequently relapsing (SD/FR). Of the 31 patients with SSNS who were above 18 years at last follow-up, 29% were still suffering from active disease. In the group of SD/FR patients, 27% (15/55) had reached the age of 18 years at last date of follow-up, of whom 53% (8/15) still had active disease.
Conclusions: The study presents clinical and long-term outcome data on an unselected Danish population of children from a well-defined geographic area. We found that more than half of patients with SD/FR continue to have active disease into adulthood suggesting that the long-term prognosis of childhood SSNS is much less favourable in this subtype.

O-64 GENETIC AND CLINICAL CHARACTERISTICS OF PATIENTS WITH C3 GLOMERULOPATHY

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Introduction: C3 glomerulopathy (C3G) is the recently defined pathological entity. Similar to atypical haemolytic uremic syndrome, it is associated with uncontrolled alternative complement pathway activation. Its presentation and prognosis remain largely unidentified.
Material and methods: Presentation, progression and outcome of 19 patients (9 females, 10 males) with histopathological diagnosis of C3G were investigated. Variations in the genes encoding complement regulatory proteins (CFH, CFI, C3, MCP, CFHR5, DGKE, thrombomodulin, plasminogen) were also searched.
Results: Mean age of pathological diagnosis was 12.3 ± 3.6 years. At the time of biopsy nephrotic range proteinuria and non-nephrotic range proteinuria was observed in 6 and 3 patients, respectively. Microscopic or macroscopic hematuria was noted in 18 patients (94.7%). At the time of presentation, eight patients had oliguria/anuria. Low C3 level was present in 15 patients (78.9%). C3 nephritic factor was positive in three of four patients. Genetic analysis revealed at least 1 variant that might be associated with the disease in 15 patients (78.9%). Six patients (31.6%) had more than one variant in more than one gene encoding complementary regulatory proteins. Mean duration of follow-up was 1.8 ± 1.9 years and long term follow-up was available for 18 patients. At last visit, five patients were under ACEi and/or ARB without any immunsuppressive treatment and 13 patients were under oral steroid treatment and MMF has been used concomitant with steroids in four of them. Two patients were under cyclophosphamide and steroid treatment and one patient was under steroid and cyclosporine treatment. Eculizumab has been administered to four patients and continued. In two patients, eculizumab was given one and three doses, respectively. As a result of these treatment regimes, mean serum albumin was 3.9 ± 1.0 g/dl and mean GFR was 111 ± 43.8 ml/min/1.73 m2 at last visit. In eculizumab group (n = 6) mean serum albumin and mean GFR was 3.5 ± 1.26 g/dl and 88.4 ± 32.5 ml/min/1.73 m2, respectively. In non-eculizumab group (n = 12) mean serum albumin and mean GFR was 4.1 ± 0.9 g/dl and 123.6 ± 45.3 ml/min/1.73 m2, respectively.
Conclusions: Variants in genes encoding complementary regulatory proteins is common in patients with C3G and eculizumab in selected patients and other treatment regimes seem to be effective. *C3G Study Group also contains authors; Esra Baskın, Oğuz Söylemezoğlu, Mehmet Bülbül, Nur Canpolat, Osman Dönmez, Gürkan Genç, Nilüfer Göknar, Umut Kavakçı, Birsin Özçakar, Alper Soylu.

O-65 EFFICACY AND ACCEPTABILITY OF ADV7103, AN INNOVATIVE PROLONGED-RELEASE ORAL ALKALISING FORMULATION IN DISTAL RENAL TUBULAR ACIDOSIS (DRTA) PATIENTS

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Introduction: A new prolonged-release granule formulation, ADV7103, has been developed in order to obtain an age-adaptable form achieving sustained physiological blood pH (blood bicarbonate levels ≥22 mM) with a 2-daily intake. Improvement of palatability and gastro-intestinal (GI) tolerability were also targeted as they are critical factors affecting compliance. The objective of this study was to evaluate the efficacy and acceptability of ADV7103 with two daily doses (morning and evening) in dRTA patients, in comparison with their current standard of care (SoC), frequently requiring several daily administrations.
Material and methods: Adult and paediatric dRTA patients (n = 37, 30 evaluable for first evaluation criterion) were included in a multicentre (N = 13), open-label, non-inferiority, sequential study. They received their SoC and then ADV7103 at appropriate doses, both during 5-day periods. Bicarbonataemia and kalaemia were measured. Palatability, easiness of administration and swallowing, and GI tolerability were evaluated using 100-mm visual analogue scales or 5-point facial hedonic scales. Descriptive analyses were performed overall and by age-subset for both products.

Results: The efficacy of ADV7103 was shown through its ability to correct metabolic acidosis (Fig. 1). Normal blood bicarbonate levels were attained in most patients with doses of ADV7103 ranging from 0.75 to 8.45 mEq/kg/day. Mean doses of 1.7, 2.3, 3.8 and 6.1 mEq/kg/day ADV7103 were given, respectively, in adults, adolescents, children, and infants. Non-inferiority of ADV7103 vs. SoC or baseline literature data was consistently demonstrated (per protocol, intention-to-treat, and sensitivity analyses), which also allowed demonstrating superiority ($p < 0.0047$). Kalaemia was normal with both treatments. Palatability, easiness of administration and GI tolerability were improved with ADV7103, as compared to the SoC. Easiness of swallowing features were maintained with ADV7103.
Conclusions: These results support ADV7103, a prolonged-release combination of potassium citrate and potassium bicarbonate, for registration as the first alkalising product ever for first-line treatment of dRTA.

O-66 CHRONIC KIDNEY DISEASE AND RENAL TRANSPLANT OUTCOMES IN PATIENTS WITH CYSTINOSIS

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Introduction: North American Pediatric Renal Trials and Collaborative Studies (NAPRTCS) was stablished in 1987 with the goal to study transplant patients (pts). In 1992 the database was expanded to include dialysis pts. and since 1994, data collection began on pts. with CKD. As of 2015, the NAPRTCS has data on over 19,000 children with CKD, on dialysis and with renal transplants. Cystinosis is a rare genetic disease. There are little data on outcomes of CKD, dialysis and kidney transplantation in cystinosis pts.
Material and methods: Retrospective analysis of NAPRTCS database of registered cystinosis pts. with CKD, dialysis and renal transplant. Data on cystinosis pts. are compared to other pts. in NAPRTCS registry who have non-recurrent, ie non-immunologic forms of potential graft loss. Specific aims were to describe the natural history of cystinosis, to investigate potential differences in CKD, dialysis and transplant pts.
Results: CKD data from 117 cystinosis pts. were compared to 2109 pts. with CKD from other causes. There was a statistical significance in growth delay in cystinosis pts. and use of growth hormone (rGH). Cystinosis pts. have significantly lower blood pressure (BP), poorer height Z-score, lower calcium, phosphate and higher alkaline phosphatase levels. Cystinosis pts. progressed to ESRD faster than other pts. with CKD. There were no statistically significant differences in gender, serum creatinine, bicarbonate, PTH level, anemia and erythropoietin use. Dialysis data from 95 cystinosis pts. were compared to 1472 dialysis pts. from other causes. There was statistical significance in BP, height and weight Z-score, growth deficit, the use of rGH, calcium and PTH levels. Dialysis survival was better in cystinosis pts. There is no statistical difference in gender, dialysis modality, anemia, erythropoietin use, albumin, phosphorus level and time to transplant while on dialysis. Data obtained from 231 transplant pts. with cystinosis vs. 2844 other causes, revealed statistical significance in height and weight Z-scores. There was no statistical difference in gender, graft survival, donor source (LRD vs. DD), preemptive transplant, time to first rejection, number of transplants.
Conclusions: CKD and dialysis cystinosis pts. differ from other CKD and dialysis pts. Cystinosis pts. progress to ESRD faster than other pts., growth deficit in cystinosis pts. is significantly worse and rGH is more commonly used. Calcium and PTH levels are lower and alkaline phosphatase is higher in CKD cystinosis pts. Dialysis survival in cystinosis pts. is better. Transplant cystinosis patients have no difference in donor source, preemptive transplant, graft survival and time to first rejection.

O-67 THE ROLE OF MACROPHAGE ACTIVATION MARKERS IN THE THERAPEUTIC MONITORING OF NEPHROPATHIC CYSTINOSIS: A MULTICENTER LONGITUDINAL STUDY

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Introduction: Cystinosis is an autosomal recessive lysosomal storage disorder characterized by cystine accumulation and early renal damage. Strict compliance to the cystine depleting agent cysteamine is necessary for more efficient treatment. Leucocyte cystine is the current therapeutic monitor. Although highly specific, its use is hindered by many technical difficulties and its availability in only few laboratories all over the world. Recent evidence suggests that inflammatory cells play an important role in the pathogenesis of cystinosis and its rapid progression to ESRD. Macrophage activation markers, such as chitotriosidase and several cytokines have been linked to disease severity and response to cysteamine therapy in cross-sectional studies. We aim to assess the longitudinal clinical value of these markers as potential therapeutic monitors in a large cohort of cystinosis patients.
Material and methods: Fifty four patients (19 children and 35 adults) were recruited from the cystinosis clinics in Leuven (Belgium), Nijmegen (Netherlands) and Traunstein (Germany). Patients were followed-up for two years during which, clinical and laboratory data were regularly collected from hospital records. Every three months, plasma samples were obtained to analyze macrophage activation markers including chitotriosidase. These markers were correlated with leucocyte cystine concentration and with other parameters of renal disease such as, serum creatinine and urinary albumin/creatinine ratio.
Results: Cystinosis patients showed large variation in compliance/response to cysteamine therapy. Average leucocyte cystine concentrations during the previous two years ranged from 0.65 to 5.8 nmol ½ cystine/mg protein. During the first year, plasma chitotriosidase activities ranged from 2 to 834 nmol/ml plasma/h in cystinosis patients (reference range < 55 nmol/ml plasma/h). Chitotriosidase activities correlated with individual cystine measurements ($r = 0.432$, $P = 0.002$). More importantly, the correlation was stronger with the average cystine values ($r = 0.582$, $P < 0.001$).
Conclusions: Chitotriosidase activity seems to correlate with long-term cystine accumulation and can be a candidate for the therapeutic monitoring of cysteamine therapy in nephropathic cystinosis.

O-68 FRENCH COHORT OF TRANSIENT ANTENATAL BARTTER SYNDROME WITH MAGED2 MUTATIONS

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Introduction: MAGED2 was recently identified in an X-linked severe and transient form of antenatal Bartter's syndrome associated with polyhydramnios and prematurity as well as in idiopathic polyhydramnios in the male offspring. An inappropriate expression of the sodium-chloride transporters NKCC2 and NCC is disclosed.

Material and methods: MAGED2 was screened by Sanger Sequencing in a series of 42 cases with transient or antenatal Bartter's syndrome and no pathogenic variant in *SLC12A1*, *KCNJ1, CLCNKB* and *BSND* genes.

Results: We found 17 symptomatic cases from 16 families harboring *MAGED2* variants including: three nonsense, three missense, three frameshift, three splice-site, two small in frame deletions and one complete deletion of *MAGED2* gene. Only two variants were previously reported, p.Arg446Cys and p.Ala490_Ala493del.

Severe polyhydramnios occurred in all pregnancies, between 18 to 27 weeks of gestation requiring serial amniocentesis (one to eleven) and indomethacin treatment in 5 cases. One case resulted in medical termination of pregnancy. In four cases, polyhydramnios was present in previous or later pregnancies. All the infants (16) were born preterm with gestational age at delivery between 26 and 33 weeks. All presented a Bartters syndrome with severe polyuria (median diuresis was 15 mL/kg/h).

Surprisingly, two cases were female. The severity of their phenotype and the course of the disease were comparable to those for male and are explained by a selective inactivation of chromosome X.

The medical follow-up of 14 patients revealed that the salt and water losses were resolved between 2 and 18 months: end of indomethacin treatment (6 cases) and/or water and salts supplements. Two cases, with associated disorders died at 1 and 12 months.

Conclusions: We confirmed with our French series of 17 *MAGED2* cases the phenotypic presentation of this transient antenatal Bartters syndrome. This new syndrome has to be considered in the differential diagnosis of Bartters syndrome with the screening of *MAGED2* as part of the molecular diagnosis.

O-69 CALCINEURIN INHIBITOR-INDUCED ENDOTHELIAL CELL INJURY AND DYSFUNCTION - A ROLE FOR COMPLEMENT

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Introduction: Calcineurin inhibitors (CNI) are associated with nephrotoxicity, endothelial cell (EC) dysfunction and thrombotic microangiopathy (TMA). Evolving evidence suggests a central role for complement dysregulation in the pathogenesis of CNI-induced TMA. However, the exact mechanism of CNI-induced complement-mediated injury remains unknown. We hypothesize that CNIs impair complement regulation on EC cell surfaces and induce complement-mediated EC injury.

Material and methods: Complement activation/regulation were assessed by flow cytometry for C3c and surface complement regulators CD46, CD55 and CD59. Complement factor H (CFH) surface binding was assessed by flow cytometry, and its activity assessed by surface CFH cofactor assay. EC cytotoxicity was assessed via LDH assay. EC repair was assessed by scratch wound assay. Blood outgrowth endothelial cells (BOECs) were incubated with cyclosporine (CsA) and subsequently exposed to 50% normal human serum (NHS) or heat inactivated serum (HIS). A previously established method of complement fixation on EC using (blocking) antibody specific to CD59 was utilized.

Results: The sequence of CsA incubation and 50% NHS resulted in a dose- and time-dependent enhancement of EC complement (C3c) deposition and cell death, exacerbated by serum starvation and anti-CD59 antibody alone. An optimal balance of EC survival and CNI effect was obtained with CsA 10 mcg/ml for 24 h. CsA led to upregulation of CD46, CD55 and CD59, and decereased CFH surface cofactor activity. Scratch wound healing over 6 h was significantly impaired by CsA.

Conclusions: CsA causes dose- and time-dependent increase in complement activation on EC with increased cell death and impaired repair. This effect was enhanced by serum starvation and by anti-CD59 antibody. We found that CsA led to decreased surface CFH cofactor activity and upregulation of surface-bound complement regulators CD46, CD55 and CD59. Our findings suggest a role for CsA-induced, complement-mediated EC injury.

O-70 URINARY PROTEOMICS TO DIAGNOSE CHRONIC-ANTIBODY-MEDIATED REJECTION IN PEDIATRIC KIDNEY TRANSPLANTATION

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Introduction: Chronic antibody mediated rejection (cAMR) is the main cause of long-term graft loss and a diagnostic challenge in renal transplantation medicine. Actually it is diagnosed in late stages by detecting donor-specific antibodies (DSA) in the blood in combination with observing typical histomorphological findings in graft biopsy. There is a leak of non-invasive biomarkers for detection of CHR allowing screening and earlier diagnosis.

Material and methods: In a case-control-study, urine samples were analysed with capillary electrophoresis coupled to mass spectrometry of 18 pediatric patients at time of diagnosis of

cAMR, distinguishing from 23 pediatric patients after kidney transplantation without cAMR (no DSA, normal graft biopsy) who were matched vie the Certain-Registry for age, gender, time after transplantation and living donation.
Results: After statistical analysis with the non-parametric Wilcoxon test, 199 potential biomarkers were identified with an AUC ≤ 0.7. These biomarkers were combined in a support vector machine (SVM)-based classifier. After total cross validation, the accuracy for detection cAMR was 90.2%. Sensitivity was 88.9% and specificity 91.3%. Th classifier's accuracy was independent of age, gender and glomerular filtration rate. Most peptides of this cAMP-classifier were fragments of the collagen I alpha chain.
Conclusions: This test should be validated in large cohorts evaluated in longitudinal studies with the aim to identify those patients after kidney transplantation, who would profit from early graft biopsy and intensification of immunosuppression at an earlier stage.

O-71 DOES THE PATHOGENESIS OF ANEMIA DIFFER IN RENAL TRANSPLANT RECIPIENTS VS PATIENTS WITH CHRONIC KIDNEY DISEASE?

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Introduction: Although similar factors play a role in both post-renal transplant anemia (PTA) and anemia in CKD patients, additional mechanisms regarding immunosuppressive medications, antimicrobial drugs, episodes of rejections and altered inflammatory milieu exist in pathogenesis of PTA. The present study aimed at elucidating risk factors of PTA and comparing anemia and inflammation related parameters in RTx and CKD patients.
Material and methods: Of the enrolled 68 participants in our single-centered, cross-sectional study, 48 were RTx, who were transplanted before 18 years of age. The other 20 participants were with CKD and comparable to RTx group in terms of age, gender and eGFR. Data on primary renal disease, mode and duration of dialysis, donor and transplant properties, list of medications and certain laboratory findings were collected from patient records. Serum levels of erythropoietin (EPO), hepcidin-25, and interleukin-6 (IL-6) were measured by enzyme-linked immunosorbent assays. The ratio of EPO to Hgb was calculated to estimate endogenous EPO resistance. Maintenance immunosuppression consisted of standard triple therapy with prednisolone, CNI and purine antagonists in 42 patients. Nine patients were on antiviral treatment**.** Anemia was defined according to the World Health Organization thresholds**.**
Results: The prevalence of anemia was 46% in RTx group and 30% in CKD group (p = 0.285). Renal transplant group had significantly lower median (IQR) of Hgb [12.1 (1.5) vs 13.1 (1.8) g/dl; p = 0.04], higher serum EPO levels [7.66 (2.15) vs 6.46 (4.17) mIU/ml, $p < 0.001$] and higher EPO/Hgb ratios [0.64 (0.19) vs 0.5 (0.37), $p < 0.001$]. Neither hepcidin-25 nor IL-6 levels differed between the two groups. In the multivariate analysis, presence of anemia in RTx group was independently associated with lower levels of eGFR (p = 0.001) and higher ratios of EPO/Hgb (p = 0.014); whereas in CKD group it was independently associated only with lower levels of iron (p = 0.007).
Conclusions: Our findings show that relative EPO resistance plays a causative role in PTA as well as in impaired renal function. Although, the cause of EPO resistance could not be explained in the scope of our research, we postulate that for RTx recipients it might be attributable to immunosuppressive therapy.

O-72 EUROPEAN DEPRIVATION INDEX (EDI), A PROXY FOR SOCIO-ECONOMIC STATUS, IS ASSOCIATED WITH GRAFT OUTCOME IN PEDIATRIC KIDNEY TRANSPLANTATION: DATA FROM THE FRENCH RENAL REGISTRY

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Introduction: Socioeconomic status is an important determinant of health. We aimed to investigate the association between socioeconomic status and graft failure in pediatric kidney transplant recipients.
Material and methods: All pediatric patients listed before 18 years of age who received a first kidney transplant between 2002 and 2014 were included. Data were collected from the French renal replacement therapy registry (REIN). Graft failure was defined as a second transplantation, return to dialysis or death whatever occurred first. An ecological index of social deprivation (European Deprivation Index, EDI) was used as a proxy for family socioeconomic status. Patients were categorized by quintiles of EDI according to the distribution of the French general population.
Results: One thousand fifty kidney transplant recipients (males 59%, median age at transplantation 13.2 years, preemptive transplantation 23%, median dialysis duration 18 months) have been included. After a median follow up of 5.9 years, 211 graft failures have been observed. The most deprived group which belongs to the 5th quintile of the French EDI represented 37% of the sample suggesting that pediatric ESRD patients come from a more socially deprived background than the general population. Five and 10-year graft survival were 85% and 69% respectively in the most deprived group (quintile 5) vs. 90% and 83% respectively in the least deprived group (quintile 1). In a Cox multivariable model adjusted for potential confounders, patients in the most deprived group had almost a two-fold higher hazard of graft failure compared with the least deprived group (adjusted HR 1.96; 95% CI 1.19–3.24).
Conclusions: The results suggest that a lower socioeconomic status is independently associated with poor graft outcome in pediatric kidney transplantation. Specific interventions targeted at low socioeconomic status are needed to reduce these disparities.

O-73 COMBINED LIVER-KIDNEY TRANSPLANTATION IN CHILDREN WITH AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE (ARPKD) – NATIONAL CENTER EXPERIENCE

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Introduction: Autosomal recessive polycystic kidney disease (ARPKD), depending on gene expression, may be presented as the multiorgan disease, with liver involvement. In particular cases combined liver-kidney transplantation (CLKTx) is necessary. Purpose of this presentation was the summary of single center experience with 16 CLKTxs performed in 2000–2015.
Material and methods: Patients and methods: overall 16 patients (8 boys and 8 girls; at the mean age of 13 years) received liver and kidney

transplant due to ARPKD. In 3 cases the renal transplantation (combined with liver) was the second transplantation after loss of the previous (isolated) renal graft. Overall 14 (of 16) patients received monoclonal blocking induction (anti-IL 2Rα moab; daclizumab $n = 13$; basiliximab $n = 1$) and maintenance immunosupression incuded: - Pred + TAC ($n = 9$); − MMF + TAC ($n = 3$) and Pred + MMF + TAC ($n = 2$). Patients with no induction ($n = 2$) were on triple therapy: Pred + MMF + TAC.

Results: Results: There was no episode of acute rejection in renal and a single episode in liver transplants within 5-year follow-up. Renal function was good at two-years post-Tx (Schwartz mean eGFR 72,16 mL/min/1.73 m2); in 3 cases eGFR deteriorated at 3-years post-Tx (mean eGFR 55 mL/min/1.73 m2) and in 5 after 5 years (mean eGFR 53.6 mL/min/1.73 m2); as a result of chronic allograft nephropathy with no signs of antibody-mediated rejection. Overall 1 and – 5 years patient survival was 94% and kidney-liver graft survival was 100%.

Conclusions: Conclusion: - long-term outcome of combined liver-kidney transplantation in children was very satisfactory, with no acute rejection, however chronic calcineurine- inhibitor related nephropathy developed with time in a half of these patients.

O-74 INCREASED CENTRAL BLOOD PRESSURE AND LEFT VENTRICULAR MASS INDEX IN CHILDREN WITH ADPKD

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Introduction: Central blood pressure (cBP) and pulse wave velocity (PWV) have not previously been reported in children with Autosomal dominant polycystic kidney disease (ADPKD). The aim of this study was to measure BP components [peripheral BP (pBP) and cBP], carotid-femoral PWV (PWVcf), indexed left ventricular mass (LVMI) and microalbuminuria in children with ADPKD.

Material and methods: This was a multi-centre prospective observational study of children (aged <18) with a confirmed diagnosis of ADPKD. Comparisons were made against a group of age-matched healthy controls. All children underwent manual pBP, cBP measurement using radial applanation tonometry and PWVcf measured using the SphygmoCor device. Indexed LV mass (LVMI) was measured using standard 2D m-mode echocardiography.

Results: 38 children with ADPKD and 49 healthy controls were recruited from two paediatric nephrology centres (mean age 12.1 years vs. 12.2 years). Children with ADPKD had significantly higher systolic pBP (mean 113 mmHg vs. 104 mmHg, $p < 0.001$) and systolic cBP (mean 96 mmHg vs. 87 mmHg, $p < 0.001$) compared to healthy children. There was no difference in PWVcf between children with ADPKD and healthy children (mean 5.67 m/s vs. 5.57 m/s, $p = 0.67$). Children with ADPKD had a significantly higher LVMI (mean 30.6 $g/m^{2.7}$ vs. 26.2 $g/m^{2.7}$, $p = 0.01$) compared to healthy children. Fifty percent with ADPKD had evidence of microalbuminuria with an abnormal albumin:creatinine ratio.

Conclusions: This study found that children with ADPKD have higher peripheral and central BP than healthy children. We observed no evidence of increased arterial stiffening although children with ADPKD had higher LVMI and microalbuminuria than healthy children. Further research is required to understand the mechanisms of elevated BP in children with ADPKD and its influence on the evolution of target organ damage.

O-75 EVIDENCE FOR BONE AND MINERAL METABOLISM ALTERATIONS IN CHILDREN WITH AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE

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Introduction: Autosomal dominant polycystic kidney disease (ADPKD) is the most common monogenic cause of renal failure. Data from adult ADPKD population show increased fibroblast growth factor 23 (FGF23) levels, resulting in low maximum rates of tubular phosphate reabsorption to GFR (TmP/GFR) and hypophosphatemia. TmP/GFR was however higher than would be expected from the circulating FGF23 level, unrevealing FGF23 resistance, with Klotho deficiency as most probable culprit. As it is unknown whether the same holds true in pediatric ADPKD patients, we prospectively assessed bone metabolism and renal phosphate handling in ADPKD children compared to healthy controls.

Material and methods: Anthropometric data, medical personal/familial history and drug intake were collected at enrollment from all patients and healthy controls. A specific questionnaire on bone health, including previous fractures or bone deformities was performed. Blood and spot urine samples were collected from all participants. In a subgroup, a X-ray of the left hand and a dual-energy X-ray-absorption (DXA) were performed.

Results: We included 92 children with ADPKD and normal renal function (52 males, mean ± SD age: 10.2 ± 5.0 years) and 22 healthy controls (10 males, mean ± SD age: 10.3 ± 4.1). Patients had significantly lower serum phosphate levels compared to healthy controls. Low TmP/GFR was observed in 24% of patients. Serum FGF23, PTH, calcitriol and Klotho levels did not differ between patients and healthy controls. Remarkably, ADPKD patients showed depressed bone alkaline phosphatase levels, suggesting low bone formation.

Conclusions: This is the first report highlighting bone formation anomalies in early ADPKD stages. We demonstrated hypophosphatemia in combination with renal phosphate leak in ADPKD children with normal eGFR with an inappropriately high FGF23 and normal Klotho levels. Further studies are required to elucidate the underlying pathophysiology and to investigate potential clinical consequences.

O-76 AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE (ARPKD) IN THE UK NATIONAL REGISTRY OF RARE KIDNEY DISEASES (RADAR)

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Introduction: Autosomal recessive polycystic kidney disease (ARPKD) is a rare genetic condition that causes cysts to develop in the liver and kidneys. It is usually first diagnosed in infancy and affects approximately 1 in 20,000 live births. It is distributed amongst boys and girls and can cause death in the first month of life. As the condition has multisystem effects, a comprehensive care strategy requires a multidisciplinary team and detailed data collection. This abstract describes data collected by the ARPKD Rare Disease Group via the National Registry of Rare Kidney Diseases (RaDaR) in the UK.

Material and methods: Data is entered retrospectively from the patient's medical records following consent. The dataset is defined by the UK Renal Registry in association with over 20 Rare Disease Groups, made up of

experts in each eligible condition. Data fields include demographics, blood and urine results, medications, transplant and dialysis history, genetics and co-morbidities. The inclusion criteria are open to all ARPKD patients including Congenital Hepatic Fibrosis and Caroli Syndrome with kidney malformation or cyst allowing comparisons between a long-term survivor subset and a cohort that included both neonatal survivors.

Results: 108 ARPKD patients from 31 UK renal units have been consented to date with an age range of 1 week to 65 years. There are 54 (50%) paediatric (under 16) patients with an average age of 8 years and 54 (50%) adult patients with an average age of 39 years. There are 50 females (46%) and 58 males (54%) males.

Conclusions: RaDaR holdsthe largest single cohort of ARPKD patients collected to date in the UK. It provides an important epidemiology data on ARPKD patients which is shared amongst the members of the Rare Disease Group to progress further research into this rare disorder and to develop best practice treatment guidelines and improve the quality of care for these patients.

O-77 BETTER OUTCOME OF HENOCH-SCHÖNLEIN PURPURA NEPHRITIS IN CHILDREN: ANALYSIS FROM THE EUROPEAN HSPN REGISTRY

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Introduction: Henoch-Schönlein Purpura Nephritis (HSPN) progression to ESRD reached 20% in old series. In 2015 ESPN supported the European Registry on biopsy proven HSPN in children.

Material and methods: Aim of the retrospective Registry is the identification of progression risk factors in children diagnosed and followed since 1995.

Results: 203 children from 11 European centers were enrolled (58% M,81% Caucasians). Median age at onset was 8.17 (IQR 6–10.3); dominant extra-renal sign was purpura(86%), associated to abdominal pain(30%) or arthralgias(21%).

Renal involvement appeared 13 days median after purpura (IQR 0–45). Renal biopsy was performed 37 days median after signs of nephritis (IQR 16–95). At time of biopsy 61% had received steroids, 5% steroids and immunosuppressors, 15% RAS–blockers, 32% no-therapy.

Renal histology was supplied by individual Centers according to ISKDC and Oxford MEST: ISKDC I:10.4%, II:36.9%, IIIa:33.2%; IIIb:14.7%, IVa:1.5%, IVb:2.9%, Vb:0.4% and Oxford MEST scores M: M0 44.3%, M1 55.7%; E: E0 58.9%, E1 41.1%; S: S0 78.9%, S1 21.1%; T: T0 89.1%, T1 10.3%, T2 0.9%.

Median follow-up 5.75 years (IQR2.9–8.3). 135/203 (66.5%) reached remission of hematuria at 7.39 months median (IQR 3.27–16.97) and 145/203 (71.4%) of proteinuria at 8.07 months median (IQR 4.83–15.89). One patient (1/203; 0.5%) reached ESRD 15 months after disease onset, none died.

In this cohort histology was not predictive for hematuria or proteinuria remission according to ISKDC classification (ISKDC I + II vs ISKDC III to V; p = ns) or MEST (M0 vs M1, E0 vs E1, S0 vs S1, T0 vs T1–2; all p = ns), however in those not reaching remission a significant proportion of M1 and S1 was observed(Chi-square $p = 0.0006$; Fisher $p = 0.0002$).

Conclusions: The analysis on 203 children enrolled so far indicates a substantially improved prognosis. No renal histology was predictive of remission. Analysis of treatments modalities is expected to enlight the possible role in achieving better outcome.

O-78 RECOVERY OF RENAL FUNCTION IN CHILDREN ON CHRONIC DIALYSIS: A REPORT FROM THE ESPN/ERA-EDTA REGISTRY

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Introduction: Data on recovery of renal function after chronic dialysis at paediatric age are scarce.

We examined the incidence of recovery of renal function and its potential determinants in a large cohort of paediatric dialysis patients in Europe.

Material and methods: Data of 6597 patients commencing dialysis at an age of 15 years or younger between 1990 and 2014 were extracted from the ESPN/ERA-EDTA Registry. Recovery of renal function was defined as discontinuing dialysis for 30 days or more. We used a cumulative incidence competing risk approach and adjusted Cox proportional hazard models to study time to recovery.

Results: After a median of 4.5 (IQR: 1.7–8.7) months on dialysis 149 (2.3%) patients experienced renal function recovery. Sixty percent of the patients recovered their renal function within six months after commencing dialysis, while 15% recovered beyond 12 months on dialysis.

Recovery occurred in 14.6% of ischaemic renal failure, 15.0% of HUS and 11.1% of vasculitis patients. Hence, these patients had an increased likelihood to recover their renal function as compared to CAKUT patients: vasculitis (aHR: 18.64, 95%CI: 8.95–38.80), HUS (aHR: 17.09, 95% CI: 9.90–29.48), and ischaemic renal failure (aHR: 15.43, 95% CI: 7.95–29.86). Younger age (<12 years) when commencing dialysis was also associated with a higher recovery risk.

For 50 patients (35%) renal function recovery was transient, as they had to restart renal replacement therapy after a median recovery period of 19.7 (IQR: 8.1–41.3) months.

Conclusions: We demonstrate a renal function recovery rate of 2.3% in the largest cohort of children on chronic dialysis studied to date. Cause of renal failure and patient age were the main determinants of renal function recovery. Caution is appropriate when planning renal transplantation too soon after dialysis initiation in patients with HUS, vasculitis and ischaemic renal failure.

O-79 NATIONAL VITAMIN D INTOXICATION OUTBREAK AMONG INFANTS DUE TO A MANUFACTURING ERROR OF VITAMIN D3 DROPLETS

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Introduction: Danish Health Authorities (DHA) recommends vitamin D supplementation for children <2y with 10 μg (400 IU)/day. This dose is considered safe and less than recommended in a recent global guideline, why vitamin D intoxication should not be expected. However due to a manufacturing error of D3 droplets a cohort of Danish infants were intoxicated in 2016.
Material and methods: A specific Danish vitamin D3 product was identified as the source of vitamin D3 intoxication of an infant with severe hypercalcemia and unmeasurable high s-25OHD levels. Laboratory analysis showed a concentration of 150 μg/droplet instead of the intended 2 μg/droplet. Infants given the recommended 5 droplets per day unintendedly received 750 μg (30.000 IU)/day. The product was immediately withdrawn. A total of 340 bottles had already been sold. In close cooperation DHA and the relevant national academic societies established a strategy for an urgent national tracing, diagnosis and treatment algorithm for vitamin D intoxication and a patient registry was set up.
Results: Nine days after withdrawal of the product 150 children <2 years at risk of intoxication were identified. Of those 87 children had s-25-hydroxy vitamin D > 150 nmol/L. Serum ionized calcium >1.35 mmol/L was detected in 76 infants, and 18 infants had severe hypercalcemia with ionized calcium of >1.49 mmol/L. Symptoms ranged from non-symptomatic to reduced appetite, vomiting, irritability and failure to thrive. Treatment included hydration and dietary restriction of calcium and vitamin D3 in the milder cases and the more severely affected infants received additional potassium citrate, calcitonin, prednisolone and bisphosphonate. Despite of this severe nephrocalcinosis was seen.
Conclusions: This outbreak illustrates the challenges of categorizing a potentially toxic substance as food supplement instead of registered pharmaceutical. The close cooperation between health authorities, academic societies and the press was crucial in the acute phase identifying and treating all infants at risk.

O-80 USE OF HERBAL AND DIETARY SUPPLEMENTS IN CHILDREN WITH KIDNEY AND URINARY TRACT DISEASE

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Introduction: Use of complementary and alternative medicine in children is as common as in adults. We aimed to determine the frequency of herbal and dietary supplements (HDS) in children with kidney and urinary tract disease, and to define associated factors.
Material and methods: 201 patients (48.3% F; 10.0 ± 3.2 years) with kidney and urinary tract disease in a tertiary center were included; control group consisted of 200 healthy children (10.5 ± 5.1 years). Information was obtained through questionnaires and examination of patient records.
Results: The frequency of HDS use in patient group and control group were 38.9% and 57.0%, respectively ($p < 0.001$). Fish oil was widely used in the population (patient group 16%, control group 38%), when use of fish oil was excluded the frequency of HDS use was comparable (patient group 28.9%, control group 26.0%; p: 0.52). In the patient group the main reason to use HDS was to treat kidney disease (42%); followed to increase immunity and treat upper respiratory tract infections, and to increase appetite. Disease duration, total length of stay in hospital and number of drugs used were increased ($p < 0.05$) in patients who used HDS (fish oil excluded). HDS use was higher among patients with an eGFR < 90 ml/min/1.73m^2 and/or on renal replacement therapy (36.3% vs. 24.0%; p: 0.06). Residential areas and the educational status of the parents had no significant effects. In both patient and control groups, main information sources for HDS were the friends-relatives and the internet; 12% had first acquired information for HDS from a healthcare professional. Among HDS users, 59% do not share with their doctors that they use these products; and 92% are aware of that HDS may have side effects.
Conclusions: Considering possible side effects, the use of HDS should be questioned on routine visits in all patients, with a special focus on the ones with longer disease duration and total length of stay in hospital, and higher number of drugs used.

O-81 IMPROVEMENT OF PATIENT, PARENT / CARER EXPERIENCE THROUGH HIGH QUALITY INFORMATION: INFOKID

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Introduction: In March 2014, the infoKID website was launched providing quality-assured, evidence-based information for parents and carers of children with kidney conditions. Two years on, we undertook an evaluation of the resource to understand the potential for improving patient and family experience and opportunities for further development.
Material and methods: We used Google Analytics to assess 'hits' on the infoKID website in the UK and worldwide. We held focus groups and clinic chats with 53 families and feedback from healthcare professionals to devise questionnaires that were piloted prior to developing the definitive surveys made available online in October 2016.
Results: Between March 2014 and October 2016 there have been a total of 178,469 infoKID sessions, with a month-by-month increase in sessions worldwide (March 2014: 1771 sessions; October 2016: 8764 sessions). Seventy-nine patients, parents and carers (PPC) and 80 HCP participated in the surveys. Only 40% of PPC reported being familiar with the site, compared with 98% of HCP. The majority (86%) of PPC familiar with the site reported preferred times of accessing information at diagnosis and with changes in the condition. PPC familiar with the site reported that the information was easy to understand (93%), trustworthy (100%), increased their knowledge of kidney conditions (90%) and reduced concerns (55%). The majority (90%) of HCP reported signposting families to infoKID, either verbally in clinic (89%), through real-time demonstration of site (40%) and / or by including a link to the site on clinic letters (50%). Interestingly only 5% of PPC reported being made aware of the resource by clinic letters. Fifty-four percent of HCPs reported that infoKID is a recommended resource for teaching and training within their Trust. Areas for development reported by both PPC and HCP included expanding the library of conditions and including patient stories and social fora.
Conclusions: Improved knowledge and reduced concerns reported by PPC indicate that infoKID has the potential to improve patient experience. Healthcare professionals should be aware that optimal timing to signpost families to high quality information appears to be at the time of diagnosis or change in the condition.

O-82 PHARMACOKINETICS OF ENOXAPARIN IN PEDIATRIC RENAL TRANSPLANTATION

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Introduction: Renal allograft vascular thrombosis is a devastating complication and the leading cause of graft loss in the 1st month after kidney transplantation in adults and in the first post-operative year in children. Prophylactic enoxaparin is commonly used in this context, but off-label in children. However, no consensus exists regarding the optimal dosing and dose-adjustment, and practices are highly heterogeneous among centers, and vulnerable to individual physician estimation and empirical practices.
Material and methods: We conducted a retrospective study to describe enoxaparin population pharmacokinetics among renal transplanted children. Anti-Xa levels were measured in 32 pediatric renal transplanted recipients. A total of 444 observations were used for the analysis.
Results: The main results were 1) body-weight but not renal function was the sole covariate influencing clearance(s) and volume(s) of distribution, 2) less than 25% of children achieved the target anti-Xa activity two days after initiation of treatment 3) large inter- and intra-individual variabilities of anti-Xa activity were observed. Based on the final population model, a bayesian-based program has been developed in order to estimate the individual pharmacokinetic parameters thanks to individual anti-Xa activity observation(s) mainly after the 1st and 2nd injections, allowing determining the next enoxaparin dose that will quickly achieve an appropriate antiXa activity (targeting 0.3–0.5UI/mL).
Conclusions: Ultimately, this Bayesian approach should help standardizing and rationalizing practices that remain to date largely heterogeneous.

O-83 LONG-TERM RENAL OUTCOME IN PEDIATRIC GLOMERULONEPHRITIS WITH CRESCENT FORMATION

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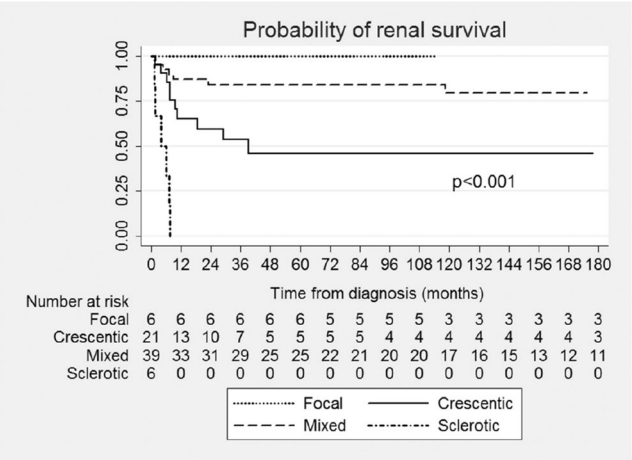
Introduction: Crescent formation, a histologic marker of severe glomerular injury, is associated with unfavorable renal outcome. However, information on prognostic factors and renal outcome of pediatric glomerulonephritis with crescents is limited.
Material and methods: We evaluated 72 children aged ≤18 years and ≥12 months follow-up (44 girls and 28 boys), in whom renal biopsy specimens showed ≥10 glomeruli and ≥10% of crescentic glomeruli, using a histological classification for ANCA-associated glomerulonephritis. Children were classified into four histological classes as six focal (≥50% of unaffected glomeruli); 21 crescentic (≥50% of glomeruli with cellular crescents); 39 mixed (<50% of normal glomeruli, <50% of crescentic glomeruli and <50% of sclerotic glomeruli); and six sclerotic (≥50% of globally sclerotic glomeruli). Time to the endpoint of chronic kidney disease (CKD), defined as ≥3 months of eGFR <60 mL/min/1.73 m^2 or ESRD, was determined. Potential predictive factors for CKD including oliguria, gross hematuria, nephrotic proteinuria, hypertension, baseline eGFR <60 mL/min/1.73 m^2, dialysis at onset, duration before treatment, type of immune deposits, and histological class were evaluated using Cox regression.
Results: At onset, gender, age, oliguria, gross hematuria and duration before treatment were similar among classes. Nephrotic proteinuria, hypertension, eGFR <60 mL/min/1.73 m^2 and dialysis requirement were less common in focal than other classes ($p < 0.05$). Type of immune deposits was different among classes ($p = 0.001$). Focal and mixed classes were observed in patients with immune-complex deposits but not in those with pauci-immune or anti-GBM deposits. Over 629 patient-years of follow-up, CKD occurred in 24 patients (33%). The overall probability of CKD at 1, 5 and 10 years were 25%, 32% and 34%, respectively and differed among classes (Figure). The only factor independently predicting CKD was sclerotic class (hazard ratio 16.70 *vs.*mixed class; 95% CI 3.33–83.70, $p = 0.001$). There was a trend towards higher probability of CKD in crescentic *vs.* mixed class (hazard ratio 3.24; 95% CI 1.97–10.87, $p = 0.06$).



Conclusions: In pediatric glomerulonephritis with crescents, the histological classification applied in this study was a major determinant of CKD. Mixed and crescentic patients had similar outcome which was better than sclerotic patients but worse than focal class. Probability of CKD was substantial and inversely correlated with the number of normal glomeruli.

O-84 MYCOPHENOLIC ACID AREA UNDER THE CURVE IS ASSOCIATED WITH THERAPEUTIC RESPONSE IN PEDIATRIC LUPUS NEPHRITIS

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Introduction: Mycophenolic acid (MPA), the active metabolite of mycophenolate mofetil (MMF), is an effective treatment in lupus nephritis. Therapeutic drug monitoring studies of MMF suggest that area under the concentration-time curve (AUC) values of MPA may be associated with clinical outcome in adults with lupus but data in children is scarce.
Material and methods: In this retrospective study, 27 children with biopsy proven class III-IV-V lupus nephritis were treated with MMF in 2009–2016. AUC of MPA was determined on the basis of sampling times at 20, 60, and 180 min postdose using a Bayesian estimator. In 25 children, AUC was performed within 6 months after kidney biopsy and MMF initiation. Treatment response at 6 months of MMF treatment was defined as follows: normal or improved GFR by 25% compared to baseline, 50% reduction of proteinuria resulting in a level < 0.5 g/day or 50 mg/mmol, no hematuria (red blood cells <10,000/ml or ≤1+ by dipstick).
Results: A total of 62 MPA-AUC were analyzed (median 44 mg.h/L [IQR 33–54]) in 27 patients. Individual dose adaptation was required in 32 cases (52%) to achieve a target AUC of 30–60 mg.h/L. At 6 months, 14/25 patients were defined as responders (56%) with a median AUC value of 49 [40–59] and 11/25 as non-responders (44%) with a median AUC value of 29 [24–38]. Patients with MPA-AUC levels of >45, 30–45, and <30 had response rates of 89% (8/9), 60% (6/10) and 0% (0/6) at 6 months. In a logistic regression model adjusted for age, sex, disease classification and time since MMF, an AUC >45 was significantly associated with therapeutic response (OR 3.9, CI95% 2.4–10.5, $p < 0.03$).

Conclusions: Therapeutic drug monitoring leading to individualized dosing may improve the efficacy of MMF. An AUC of MPA >45 mg.h/L is associated with a better response rate and migt be considered as a target value in pediatric lupus nephritis.

O-85 QUALITY OF LIFE IN IDIOPATHIC NEPHROTIC SYNDROME: COMPARISON BETWEEN ORAL IMMUNOSUPPRESSION AND RITUXIMAB

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Introduction: Severe forms of idiopathic nephrotic syndrome (INS) require immunosuppressive therapy, such as oral treatment (calcineurine inhibitors or mycophenolate mofetil) or rituximab (RTX). Our objective was to compare the quality of life (QOL) between patients treated with oral drugs or RTX.
Material and methods: This prospective multicentric, observational study compared the QOL with a standardized questionnaire (Kindl®) given to children aged from 7 to 17 years old, with a steroid dependent or steroid resistant INS separated in three treatment groups (oral drugs or rituximab or both) in stable remission. The questionnaire was completed during an outpatient consultation and consisted of 30 questions concerning physical and emotional well-being, self-esteem, family, friends, school, and disease resulting in a global score of 0 to 100. A propensity score was used to compensate the absence of randomisation.
Results: 110 patients (72 boys, 38 girls) with a mean age of 11.6 ± 3.1 years were included in three French pediatric nephrology centres. There were 71 patients in the oral drugs group, 27 in the RTX group and 12 in the oral drugs and RTX group. 13.6% of patients had a steroid resistant form of INS (9.9% in the oral drugs group and 18.5% in the RTX group). The number of relapses was 5.8 ± 3.7 (4.7 ± 2.9 in oral drugs group and 8.2 ± 4.1 in RTX group). The score in the whole study population was 74.5 ± 11.03 (74.1 ± 15.6 in the RTX group, 74.9 ± 10.5 in the oral drugs group (p = 0.73). Even after an adjustement using a propensity score there was no statistical difference (p = 0.6).
Conclusions: Quality of life in patients with oral and IV treatment in patients with severe forms of INS is not different. The treatment choice should include the opinion of the patient and his/her family. Neither clinical nor QOL parameters are sufficient to decide for or against RTX treatment.

O-86 ROLE OF VON WILLEBRAND FACTOR (VWF) IN ENDOTHELIAL CELL REPAIR AFTER COMPLEMENT ACTIVATION

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Introduction: Complement dysregulation on endothelial cells causes EC activation/injury and leads to thrombotic microangiopathy. Different from previous concepts, our data demonstrate that complement dysregulation does not result in EC death. The current study focuses on EC complement evasion strategies, especially plasma membrane (PM) repair. We particularly focused on von Willebrand Factor (VWF), a protein stored in EC Weibel-Palade bodies(WPB), which we recently identified as new complement regulator on ECs.
Material and methods: Complement activation on ECs was induced via sensitization on blood outgrowth endothelial cells (BOECs) from healthy controls and patients with von Willebrand disease (VWD) lacking both VWF and their storage WPBs. FM1-43X dye and calcein release were used to determine PM integrity.
Results: Complement activation resulted in PM insertion of C5b-9 pores in control and VWD BOECs resulting in rapid Ca^{2+} influx. In response, VWF was recruited to the EC surface via WPBs merging with the PM and releasing VWF multimers within 30–60 min. Importantly, control but not VWD BOECs resealed their PM within 30 min. In control BOECs known cellular mechanisms for PM repair (endocytosis, lysosomal recruitment) were not critically involved in PM repair.
Conclusions: PM repair is a major strategy of ECs to overcome complement-mediated injury. Our study indicates a new mechanism: Ca^{2+}-dependent VWF recruitment to the EC surface resulting in complement regulation and EC PM repair via WPBs. The understanding of the detailed mechanism warrants further investigation.

P-001 ECULIZUMAB FOR DIARRHEA-ASSOCIATED HEMOLYTIC UREMIC SYNDROME; WHERE DO WE STAND? A REVIEW OF CURRENT EVIDENCE

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Introduction: Background: Diarrhea-associated hemolytic uremic syndrome (D + HUS) is associated with significant mortality and morbidity. Case fatalities are often associated with severe neurological involvement and currently specific treatment is not available. Eculizumab, a terminal complement inhibitor approved for treatment of atypical hemolytic uremic syndrome, has been used in severely affected patients with D + HUS but the efficacy remains undetermined. In addition, increasing evidence shows heightened activity of the complement in this condition.
Material and methods: Methods:We searched the databases Pubmed, Web of Science, Embase and LiLACS for reports describing the outcome of Eculizumab administration in D + HUS. We also selected additional references through bibliographies from identified articles. We restricted the search to articles in English and French with no restrictions concerning time of publication.
Results: We retrieved 9 reports representing 363 cases of D + HUS patients (children and adults) treated with Eculizumab. Reports ranged from case reports to retrospective cohort studies with the largest study population emanating from the 2011 German outbreak. Outcomes were variable but consistently showed dramatic improvements in severe cases with neurological involvement when eculizumab was administered early in the course.
Conclusions: The efficacy of eculizumab in D + HUS cannot be established or disproven based on the limited available data but current data suggest that Eculizumab administration early in the course of severe cases might be efficacious especially in case of neurological involvement.

P-002 ONE-YEAR FOLLOW-UP EVALUATION POORLY PREDICTS LONG-TERM RENAL OUTCOME IN DIARRHEA-ASSOCIATED HEMOLYTIC UREMIC SYNDROME

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Introduction: Diarrhea-associated hemolytic uremic syndrome (D+ HUS) due to Shiga toxin-producing *E. coli* is the main cause of acute kidney injury in young children. Most of patients recover from the acute episode. However, some who apparently fully recover their renal function may develop long-term sequelae. We aim to determine whether renal outcome at one year post HUS predicts long-term renal prognosis in children with D+ HUS.
Material and methods: Retrospective population-based study of children aged <15 years with a diagnosis of D+ HUS hospitalized in South West France region between 1992 and 2012.

Results: During this period, 98 D+ HUS cases were reported in the region. The average annual incidence over the 20-year period was 0.85 cases (IC95% 0.68–1.04) per 100,000 children less than 15 years per year. Of the 96 patients who survived, 42 (44%) of patients had one or more signs of renal impairment at 1 year follow up. Data from 80 patients were collected after a median follow-up of 9.1 (IQR 3.8–12.7) years after HUS onset. Median age of patients at last follow-up was 11.8 years (IQR 7.7–16.6) at which 41 (43%) had renal impairment: 2% hypertension, 10% proteinuria, 29% decreased GFR <90 ml/min/1.73 m^2 (including one kidney transplant). Among the 42 patients with renal abnormalities at 1-year of follow-up, only 22 (52%) still have renal impairment at last follow-up. In some patients, however, renal impairment appeared after an interval of apparent complete recovery. Of the 32 patients without renal impairment at 1-year and available long-term data, 10 (31%) had proteinuria or decreased GFR at last follow-up.
Conclusions: Since renal sequelae may appear at variable time intervals after an acute episode of HUS, it is warranted to follow all patients for a long period in order to early detect signs of CKD and propose measures to slow its progression.

P-003 IS PLASMA EXCHANGE EFFICACIOUS IN DIARRHEA-ASSOCIATED HEMOLYTIC UREMIC SYNDROME? A REVIEW OF CURRENT EVIDENCE

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Introduction: Background: Diarrhea-associated hemolytic uremic syndrome (D + HUS) is associated with significant mortality and morbidity. Case fatalities are often associated with severe neurological involvement in children and advanced age in adults but currently specific treatment is unavailable. Plasma exchange has been used in the deteriorating patient with D + HUS but the efficacy remains uncertain. Plasma exchange could theoretically enable removal of Shigatoxins, pro-inflammatory cytokines and prothrombotic factors thereby limiting disease progression. Aim: To assess the efficacy of plasma exchange in D + HUS based on a review of the literature.
Material and methods: Methods: We searched the databases Pubmed, Web of Science, Embase and LiLACS for reports describing the outcome of plasma exchange in D + HUS.We also selected additional references through bibliographies from identified articles. We restricted the search to articles in English and French to report published after 1990. The decision to exclude reports before 1990 was made in light of difficulties assessing full-text articles which often could not be located online.
Results: We included 16 reports describing outcomes of plasma exchange in patients (children and adults) with D + HUS. Reports ranged from case reports to cohort studies and one case control study with the largest study population emanating from the 2011 German D + HUS outbreak. Outcomes seemed to consistently point to efficacy of early plasma exchange in reducing case fatality rates in patients >60 years. In addition, limited evidence points to efficacy of plasma exchange in improving outcome in children with D + HUS.
Conclusions: Institution of early plasma exchange seems to be justified in D + HUS in patients >60 years while in children and adults <60 years, this should probably be reserved for those at risk for poor outcomes.

P-004 GLYCOPROTEINS FOR SEROLOGY IN STEC-HUS: A CLOSER LOOK TO A NOVEL APPROACH

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Introduction: Differentiation between Shiga toxin producing *Escherichia coli* (STEC) and atypical hemolytic uremic syndrome (HUS) is essential prior to start of treatment, as treatment of atypical HUS (aHUS) comprises one of world's most expensive drugs nowadays: eculizumab. Furthermore, since aHUS is merely a diagnosis per exclusionem, providing proof for the presence of STEC forms the basis for differentiation between aHUS and STEC-HUS. However, the current gold standard (i.e. fecal diagnostics) to demonstrate STEC infection appears far from ideal. Serological detection of antibodies against the lipopolysaccharides (LPS) of STEC by ELISA has improved STEC-HUS diagnostics significantly. Yet, this assay revealed multiple limitations, whereas new techniques show promising results. Here we evaluate the use of serotype specific recombinant glycoproteins as antigens in a novel established glyco-iELISA.
Material and methods: In this retrospective study, all patients (n = 61), who presented with clinical STEC-HUS in the Radboudumc, Nijmegen, Netherlands, between 1990 and 2014 were included. Serological assays using LPS and glyco-iELISA to detect IgM against STEC most common serotype O157 were compared to each other as well as to fecal diagnostics.
Results: With the sole use of fecal diagnostics, 32 (53%) of the patients were diagnosed with an STEC infection. With serological assays, respectively LPS-ELISA and glyco-iELISA, 39 (65%) and 46 (75%) patients were diagnosed with STEC infection. Moreover, fecal diagnostics failed to detect 20 of the 46 patients positive with glyco-iELISA. With LPS-ELISA, 7 patients went undetected when compared to glyco-iELISA. Combining the glyco-iELISA with fecal diagnostics provided evidence of STEC infection in 89% of the patients.
Conclusions: The novel glyco-iELISA is a reliable and accurate serological method to detect STEC-HUS. This together with the stability and the absence of cross-reactivity places the glyco-iELISA ahead of the LPS-ELISA. Furthermore, STEC detection is enhanced with 36% when glyco-iELISA is combined with fecal diagnostics.

P-005 ASSESSMENT OF THE ETIOLOGIES AND RENAL OUTCOMES OF RAPIDLY PROGRESSIVE GLOMERULONEPHRITIS IN PEDIATRIC PATIENTS AT KING ABDULAZIZ UNIVERSITY HOSPITAL

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Introduction: Background Rapidly progressive glomerulonephritis (RPGN), also known as crescentic glomerulonephritis, is an uncommon but serious syndrome in pediatric patients. RPGN is characterized by a rapid deterioration in renal function, and eventually progress to end stage renal disease (ESRD). Etiologies that could lead to RPGN differ regionally; along with early possible intervention, different outcomes are suspected. The aim of this study was to investigate the etiologies and renal outcomes of RPGN in pediatric patients at King Abdulaziz University Hospital (KAUH) in Jeddah.
Material and methods: A retrospective study was conducted of 19 pediatric patients who were diagnosed with RPGN, between 2006 and 2016, at the department of pediatric medicine at KAUH. Relevant data were extracted from medical records. Associations between variables were evaluated using independent t-test, one-way analysis of variance (ANOVA) and chi-squared tests, as appropriate for the dataset.
Results: The majority of patients were males, 13 (68.42%), with a mean (standard deviation) age at diagnosis of 8.52 (3.15) years. The most common underlying etiologies were post-infectious glomerulonephritis (63.2%) and lupus nephritis (21.1%). Macroscopic hematuria and edema were identified in 89.5% of cases, with

macroscopic hematuria being the main presenting symptom in patients with post-infectious glomerulonephritis (P = 0.026). The mean serum creatinine level at presentation was 327.15 μmol/L, decreasing to 217.66 μmol/L at the last follow-up. Serum creatinine levels at the last follow-up were predictive of clinical outcomes (ANOVA, P = 0.019). Thirteen patients exhibited a good clinical prognosis (68.43%), with 6 exhibiting a poor prognosis (31.57%), 4 of whom progressed to ESRD, one experiencing a relapse and one developing chronic kidney disease. Post-infectious glomerulonephritis as the etiology of RPGN was associated with the best clinical outcomes, overall. More than half of the patients received a combination therapy of corticosteroids and immunosuppressive therapy (52.6%), with other patients receiving corticosteroids only. Treatment was implemented early in all patients and continued for 3 months.

Conclusions: Post-infectious glomerulonephritis was the most common etiology of RPGN, with these patients achieving a good clinical prognosis overall. Early identification and treatment of RPGN is important to preserve renal function, which is a key factor for achieving a good prognosis.

P-006 ADAMTS13 ACTIVITY IN CHILDHOOD HUS IN A PEDIATRIC UNIVERSITY HOSPITAL IN EGYPT

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Introduction: Our goal was determining ADAMTS13 activity in children with HUS and correlagting it with clinical manifestations, complications and outcome.

Material and methods: 25 pediatric patients (15 males, 60%) with HUS, attending the Pediatric Nephrology Unit, Cairo University were studied. Fourteen (56%) had history of diarrhea (D + ve) while 11 (44%) were diarrhea negative (D -ve). Their age ranged from 4 months to 18 years (mean 4.63 ± 5.24 years). Detection of ADAMTS13 activity and anti ADAMTS13 autoantibodies was done by ELISA.

Results: Thirteen cases (52%) had normal ADAMTS13 levels and in 12 cases (48%) it was mildly (8 cases, 67%) to moderately (4 cases, 33%) deficient. ADAMTS13 inhibitors were negative in 11 of those 12 cases. One showed borderline positivity. Eight (32%) had associated neurological affection, seven of whom had a normal CT brain scan, and one showed mild brain atrophy. Seven of the 12 ADAMTS13 deficient patients (58.3%) had neurological symptoms, vs. only one of the ADAMTS13 normal ones (p = 0.011). There was a negative correlation between the ADAMTS13 activity and neurological affection (−0.511, p 0.009). Fourteen patients recovered completely, 6 had persistent renal damage or hypertension, 4 died and 1 had a relapse (a D –ve case). Recovery showed a higher incidence among the D + ve cases (64.3%). A favourable outcome was reported in more cases with normal ADAMTS13 activity vs. deficient cases (64.3% vs. 35.7%). All mortalities were among the ADAMTS13 deficient group.

Conclusions: ADAMTS13 activity, even if not below the critical level of <10%, may still have prognostic value as far as outcome and neurologic complications in pediatric HUS cases.

P-007 GENOTYPIC AND PHENOTYPIC ANALYSIS OF PEDIATRIC ATYPICAL HEMOLYTIC UREMIC SYNDROME IN TAIWAN

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Introduction: Clinical manifestation, genetic background, and follow-up studies of pediatric patients with complement-dysregulation hemolytic uremic syndrome (aHUS) in Taiwan are not well studied.

Material and methods: Nine pediatric patients with aHUS belonging to nine unrelated Taiwanese families were enrolled. Genomic DNA was analyzed for *CFH, CFI and MCP genes* by polymerase chain reaction and direct sequencing. The clinical presentations, and laboratory data including serum CFH/I on the first presentation and follow-up outcome were recorded.

Results: The age at onset of nine patients ranged from 2 days to 11 years with male predominant. All patients did not family history of HUS, and one patient had co-existing underlying disease. At presentation, all had hypertension with oliguira, and two of them had bloody diarrhea. Central nervous system and heart were the most common extra-renal involved organs. Infectious triggers were identified in 5 patients, and the time from trigger to presentation ranged from 2 to 10 days. All but one had anemia and thrombocytopenia. All has elevated LDH, which ranged from 468 to 3163 IU/L, and Half of patients had normal C3 and C4 levels. Three patients have abnormal low serum levels of CFH and CFI. Eight patients had either missense mutations or pathogenic SNP on *CFH* gene. All patients underwent plasma therapy initially, but only 3 patients responded to it. Five patients received eculizumab therapy and all respond to it. After mean 2-year-Follow-up, two patients died of sepsis. Two and five of survivals had CNS/heart sequlae and reached to chronic kidney disease, respectively.

Conclusions: aHUS can occur as early as the neonatal period. Viral infection, is a common trigger.Heart and central nervous system are the two most common extra-renal involved organs. Not all patients had abnormal C3/C4 levels. Hot spots of mutation were identified on *CFH* gene. Most patients with their first TMA failed to respond to plasma therapy. Eculizumab is effective and safe therapy in pediatric aHUS patients.

P-008 URINARY BIOMARKERS OF ACUTE KIDNEY INJURY IN VERY LOW BIRTH WEIGHT INFANTS ON INDOMETHACIN FOR PATENT DUCTUS ARTERIOSUS

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Introduction: Indomethacin for closure of a patent ductus arteriosus (PDA) adversely affects renal function in preterm neonates. A serum creatinine (SCr)- or urine output-based diagnosis of acute kidney injury (AKI) as defined by neonatal Kidney Disease Improving Global Outcomes (KDIGO) criteria is methodologically uncertain. We therefore aimed to determine the diagnostic value of novel urinary biomarkers of AKI.

Material and methods: We performed a prospective cohort study in 32 very low birth weight (VLBW) neonates including 14 neonates treated with indomethacin (five doses of 0.2 mg/kg body weight) for PDA on day 2–3 of life and 18 neonates without PDA as control. Urinary neutrophil-gelatinase associated lipocalin (NGAL), kidney injury molecule-1 (KIM-1), calprotectin and the product of tissue inhibitor of metalloproteinase-2 (TIMP-2) and insulin-like growth factor-binding protein 7 (IGFBP7) were measured before, during (at 6, 12 and 36 h) and after (at 84 and 120 h and at day 7, 14 and 28) indomethacin administration.

Results: Indomethacin therapy was associated with higher SCr concentrations at 36, 84 and 120 h compared to controls and with AKI in three (21%) patients. [TIMP-2]•[IGFBP7] was elevated ($P < 0.05$) in the AKI subgroup already at 12 h, in the indomethacin cohort at 36 and 84 h. NGAL and calprotectin were increased ($P < 0.05$) at 6 and 12 h, irrespective of fulfillment of AKI criteria. Urinary KIM-1 was not significantly altered.

Conclusions: While urinary [TIMP-2]•[IGFBP7] proves valuable for the early detection of neonatal KDIGO defined AKI, elevated urinary

biomarkers in indomethacin-treated neonates not fulfilling AKI criteria may indicate subclinical kidney injury.

P-009 AN EXCEPTION FROM THE AHUS THERAPEUTIC GUIDLINES

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Introduction: We report the rare case of a 2 years old girl who was diagnosed with atypical hemolytic uremic syndrome (aHUS), caused by DGKE (diacyl-glycerol-kinase-kappa). At the time of admission, she showed nephroso-nephritic symptoms, paleness with mild jaundice.
Material and methods: Laboratory results showed anemia, thrombocytopenia, increased LDH, Coombs negative hemolysis, and maintained kidney function. Urine analysis showed nephrotic range proteinuria and mild haematuria. Because of the nephroso-nephritic symptoms kidney biopsy was done, the result was secondary thrombotic microangiopathy. Immunologic study proved no abnormalities of alternative Complement route, complement C3 was in normal range, ADAMTS 13 activity was decreased, but not in the range as we see it in thrombotic thrombocytopenic purpura.
Results: After no improvement to parenteral high dose pulse steroid treatment and plasmapheresis, we started eculizumab treatment without any improvement in the patient condition or the laboratory result.
After we ruled out all the other causes of aHUS further genetic testing was done, the result revealed function loss mutation (compound heterozygote) of the DGKE gene.
Conclusions: DGKE plays an important role in the arachidonic acid signaling pathway, by phosphorylating arachidonic acid and inhibiting the activation of protein kinase C, but has no effect on the complement system. If we suspect aHUS with aid of early genetic testing we can avoid the use eculizumab.

P-010 PAEDIATRIC DIALYSIS ACCESS COMPLICATIONS IN THE UK

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Introduction: For children on dialysis, their access device is their lifeline. Existing data on complications are based on historical series or collaborative registries across different healthcare systems. We collated data across one healthcare system in the modern era.
Material and methods: Between January and July 2016, we conducted a six month prospective monthly survey of children established (>3 months) on haemodialysis (HD) or peritoneal dialysis (PD). Data regarding infection and treatment of these events, need for revision of access, and outcomes were collected. Data was collected on a monthly basis from participating centres.
Results: A total of 191 patients from 9 UK centres were included, equating to 787 person/months (HD = 382, PD = 405). Infectious complications were recorded on 54 occasions (peritonitis $n = 33$, bacteraemia on HD $n = 21$, 0.8 per patient/year). Peritonitis resulted in change of catheter in 6/33 (18%), causes included no organism identified ($n = 12$) and coagulase negative staphylococcus ($n = 7$). *Staphylococcus aureus* was the most common cause of HD bacteraemia ($n = 9$). Haemodialysis lines were replaced on 21 occasions, the most common reasons was being dislodged ($n = 12$) or infection ($n = 6$). Alteplase for blocked or poorly flowing lines was used on 13 occasions and was successful on 11 occasions.
Conclusions: In our contemporary study, only a minority of infected dialysis lines resulted in replacement. Interestingly, over 50% of HD line changes were due to accidental displacement. These baseline data will form the basis of future quality improvement projects to minimise preventable loss of dialysis access in children.

P-011 EFFICACY OF TWO VS FOUR DOSES OF RITUXIMAB THERAPY FOR CHILDHOOD NEPHROTIC SYNDROME

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Introduction: Traditionally treatment of steroid dependent and steroid resistant nephrotic syndrome with Rituximab has been four doses one week apart for a total of four doses. We compare the efficacy of two versus for four doses as follows: time to relapse; T & B cell count; recurrence of proteinuria; protein:creatine ratio; requirement of repeat courses.
Material and methods: Twenty eight children between 38 months to 217 months of age received rituximab therapy. We reviewed patient histories, noting time to relapse; T & B cell count; recurrence of proteinuria; protein:creatine ratio; repeat courses.
Results: Eleven children received the traditional course of 4 doses X 1 course, Seven children received 2 doses X 1 course, 2 children received 1 dose X 1 course, 1 child received three doses X 1 course. The remaining seven children received multiple courses of between 1 and 4 doses.
Conclusions: There appeared to be no significant difference in outcomes between the 2 vs 4 dose regimes. We recommend a reduction in rituximab regimes from four to two doses, thus reducing costs, potential adverse reactions to the drug, and inconvenience to children and their families.

P-012 DO SERUM AND URINARY CONCENTRATIONS OF KIDNEY INJURY MOLECULE-1 IN HEALTHY NEWBORNS DEPEND ON BIRTH WEIGHT, GESTATIONAL AGE OR GENDER?

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Introduction: Determination of renal function in neonates can be challenging. Firstly, it is difficult to obtain timed urine samples, and blood sampling requires skills. Secondly, the renal function of normal infants is known to change considerably during the 1st year of age. Thirdly, the renal function of children up to 1 year of age should not be assessed from urine flow rate, as in this age group, glomerular filtration rate (GFR) corrected for body size is not comparable with normal adult values. All this stimulated research on biochemical markers that could be determined in untimed urine samples and predict changes in renal function. The aim of work was to establish the normal levels of serum and urinary kidney injury molecule-1 (KIM-1) in healthy full-term newborns.
Material and methods: Male and female newborns, as well as children in whom the samples were obtained in the first or second day of life, did not differ significantly in terms of their sKIM-1 and uKIM-1 levels. Gestational age correlated inversely with sKIM-1 and positively with uKIM-1.
Results: Male and female newborns, as well as children in whom the samples were obtained in the first or second day of life, did not differ significantly in terms of their sKIM-1 and uKIM-1 levels. Gestational age correlated inversely with sKIM-1 and positively with uKIM-1.

Conclusions: This is the first report of sKIM-1 and uKIM-1 levels in healthy full-term newborns during the first postnatal days. sKIM-1 and uKIM-1 levels correlate with gestational age, but not with birthweight and gender.

P-013 CURRENT TRANSITION POLICY AT EUROPEAN PAEDIATRIC CENTRES AFTER KIDNEY TRANSPLANT

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Introduction: Transition in medical care is a high-risk period in adolescence and young adulthood for patients after kidney transplant (KTx). Some studies report unexpected loss rates in kidney grafts to be as high as 24% to 42% within 3 years after transfer of patients into adult care. In 2011 the International Society of Nephrology (ISN) and the International Paediatric Nephrology Association (IPNA) published a consensus statement on transition. Fifteen components were considered essential. To date, data on transition policy, its application in practice and transfer methods in Europe's countries is scarce.
Material and methods: A responsive online survey was developed and was distributed via ESPN e-mailing list twice to assess the transition-relevant structures from the providers' perspective. Its content was mainly based on the 2011 ISN/IPNA consensus statement and previous research of our workgroup. The survey included multiple-choice and open questions, as well as 5 point Likert scales.
Results: 33 centres of 21 European countries participated in the survey to date. Most of them (85%) are university hospitals. A median number of 3 patients (0–15) is annually transferred to adult based care. In 2 centres children after KTx remain under paediatric care at all ages. 24/31 centres confirmed using an at least unwritten transition procedure (77%), which in most centres is introduced at age 14–16 (21%) or >18 (21%). 14/31 centres confirmed existence of at least one transition coordinator. Transition clinics are offered by 12 centres. Most centres transfer patients primarily to a university hospital (61%). Transfer age is subject of regulation at 15/33 centres. Most centres (48%) transfer patients commonly at age 18–19, 5 centres at age < 18.
Conclusions: The majority of European centres use a transition procedure. However, the implementation of the ISN/IPNA consensus guidelines on transition in clinical routine could be improved at many centers.

P-014 MULTISYSTEM MANIFESTATIONS OF HAEMOLYTIC UREMIC SYNDROME: A PICTORIAL CASE SERIES

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Introduction: We present a comprehensive pictorial case series illustrating the diverse multisystem radiological manifestations of Shiga toxin-producing *Escherichia coli* Haemolytic Uremic Syndrome (STEC-HUS).
Material and methods: STEC-HUS is characterised by haemolytic anaemia, thrombocytopenia and acute renal dysfunction with significant morbidity and mortality. We retrospectively reviewed the imaging findings of 148 cases of STEC-HUS in a national paediatric nephrology centre over a 13 year period.
Results: Renal: Renal ultrasound was performed in 18.2% of cases, of which a hyperechoic renal cortex was the most common finding (19/27). Three cases of renal cortical necrosis and one case of acquired cystic kidney disease were diagnosed on serial ultrasound and MRI imaging. **Gastrointestinal:** Abdominal radiography (AXR) was performed in 14.2% of cases, of which colonic wall thickening was the most common finding (14/21). On abdominal ultrasound colonic thickening (12/27) and free fluid (10/27) were the most common findings. One case of bowel ischaemia was diagnosed on MRI. **Neurological:** Neuroimaging was performed in 8.1% of cases, of which bilateral lentiform nuclei and/or thalamic infarctions were the most common abnormality (5/12). Two patients had multi-territorial infarcts including the first reported case of large vessel arterial thrombosis in HUS on imaging. **Hepatobiliary:** Two cases of hepatomegaly were seen on abdominal ultrasound. On AXR biliary excretion of contrast media due to renal failure was reported in one case. Haemodialysis-related secondary haemochromatosis was diagnosed on follow-up MRI in two cases. **Respiratory:** Chest radiography was performed in 21.6% of cases, of which pulmonary oedema was the most common abnormality (7/32). **Ocular:** Eye ultrasound demonstrated severe vitreous haemorrhages in one case.
Conclusions: This pictorial case series illustrates the imaging findings of common, rare and previously unreported multisystem manifestations of STEC-HUS. Awareness of the role of radiology will aid early diagnosis and subsequent management in this complex disease.

P-015 A NOVEL INTRONIC VARIANT IN MMACHC RESULTING IN ATYPICAL HAEMOLYTIC URAEMIC SYNDROME

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Introduction: Atypical Haemolytic Uraemic syndrome (aHUS) is the triad of microangiopathic haemolytic anaemia, thrombocytopenia and acute kidney injury (AKI). Approximately 50% of cases carry mutations in complement regulators or complement component genes. Secondary causes of aHUS include infections, drugs, and Cobalamin C (CblC) defects.
We present a 4 year old boy with haemolytic anaemia, thrombocytopenia and AKI. At presentation he had features of cardiac dysfunction, transient neurological involvement and renal biopsy demonstrating thrombotic microangiopathy. He initially required peritoneal dialysis (PD) and was commenced on Eculizumab. Two months after starting eculizumab, his neurological defict had resolved and PD was discontinued. Despite adequate complement blockade (absent AH100/CH100), he relapsed during eculizumab treatment with ongoing proteinuria; repeat renal biopsy showed active TMA.
Material and methods: Routine investigations for aHUS were negative, including aHUS related genes and factor H autoantibodies. Initial plasma homocysteine and urinary methylmalonic acid (MMA), in the setting of AKI, were elevated. Subsequent testing demonstrated normal MMA and (spuriously) low homocysteine levels, hence a diagnosis of a CblC defect was considered unlikely. He underwent whole exome sequencing to investigate the cause.
Results: Two variants in *MMACHC* were identified, a previously reported pathogenic variant (c.271DupA) and a second novel intronic variant (c.82-11_8delTTCT). Fibroblast enzyme analysis confirmed the diagnosis of CblC disease. He was treated with hydroxycobalamin and betaine, which allowed his eculizumab to be discontinued. Subsequently, his renal function stabilised and proteinuria has settled.
Conclusions: CblC defects are rare disorders of cobalamin metabolism, secondary to recessive variants in *MMACHC*. The clinical presentation CblC disease is variable; however, aHUS is the most common presentation in children under 6 years. Treatment of CblC disease is with hydroxycobalamin and anhydrous betaine. CblC defects represent a rare, treatable cause of aHUS and should always be considered. Diagnostic sequencing of *MMACHC* should include intronic regions to ensure splice site variants are identified.

P-016 GROSSHEMATURIA IN CHILDREN

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Introduction: Gross hematuria (GH) is rare in children. In a previous retrospective USA study a prevalence of 158 cases of 128.395 accesses to pediatric ER was reported; main causes were documented (26%) or suspicious (23%) UTI.To determine the prevalence and causes of GH in Italian children presenting at pediatric ER we conducted a prospective study.
Material and methods: All children with GH were sent to the nephrologist for full assessment until the final diagnosis; traumatic and surgical causes were excluded. In 17 months 155.833 children were observed; 111.073 had medical problems; 97 of these (0.09%) showed GH.
Results: Data: M/F ratio 1.36; mean age 7.6 ± 4.6 years Main diagnosis 54 (55.6%) Glomerulonephritis (GN) of which 38 post infectious, 8 primary IgAN and 4 secondary to Henoch-Schonlein syndrome, 3 Alport syndromes and 1 SLE nephritis; 17 documented UTI (17%); 18 Idiopathic Hypercalciuria (17%), 3 Hyperuricosuria; 2 Myoglobinuria; 2 HUS and 1 not identified.
Conclusions: In Italy, as well as in the US, prevalence of GH in children is low (0.09%). Compared to US data, causes of GH, in our series, were different: most frequent cause was hematuric GN, mainly in males, meanwhile UTI, in females, represented main cause in USA. Our results are closer to reality because: 1) they derived from a prospective study, 2) the systematic evaluation of children with GH by nephrologists allowed to establish the correct diagnosis in all cases. Only in one case the diagnosis remained uncertain against the 35 cases of "suspicious" UTI and 13 undefined causes of the US series. The true prevalence of GH in our children is probably underestimated for the under-representation of UTI. In fact in Italy the different organization of pediatric health system prevents the trivial causes (i.e. UTI) of GH should come to the ER.

P-017 CALCIUM OXALATE UROLITHIASIS IN CHILDREN: URINARY PROMOTERS/INHIBITORS AND ROLE OF THEIR RATIOS

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Introduction: The role of urine calcium as promoter in calcium oxalate urolithiasis is well established. Seldom used calcium/citrate ratio is acknowledged a as risk factor for calcium /oxalate urolithiasis. Diagnostic criteria for determination of inclination towards idiopathic calcium oxalate (CaOx) urolithiasis based on biochemical urine parameters are not sufficiently well defined in children. The aim of this study was to determine the risk of CaOx urolithiasis in children from concentrations of calcium, oxalate, citrate, glycosaminoglycans in urine and their ratios, all standardized in respect to creatinine.
Material and methods: We collected and analyzed 24-h urine samples of children with CaOx urolithiasis ($n = 61$) and compared with urine samples of matched control group of healthy children ($n = 25$).
Results: The study has showed that all stone formers have higher excretion of calcium (mmol/mmol creatinine), calcium/citrate (mol/mmol) and oxalate/(citrate × glycosaminoglycans) ratio (mol Ox × mol cr)/(mol Cit × g GAGs). ROC analysis of these variables gave criteria (> 0.28; > 1.07; > 0.08 respectively) for distinguishing stone formers from healthy children. Biochemical urine parameters and their ratios (calcium, calcium citrate and oxalate/(citrate × glycosaminoglycans) enable one to discriminate idiopathic calcium oxalate stone formers from healthy children. Oxalate/(citrate × glycosaminoglycans) ratio per se can serve as independent risk for stone formation.
Conclusions: The values of calcium and citrate in clinically and genetically proven idiopathic calcium oxalate urolithiasis makes calcium/citrate ratio useful for diagnostic purposes in such stone formers. Rarely used calcium independent oxalate/(citrate x glycosaminglycans) ratio serves as the second best high specificity marker for idiopathic calcium oxalate urolithiasis. Using biochemical urine parameters and their ratios such as calcium, calcium/citrate and oxalate/(citrate × glycosaminoglycans) enables one to determine diagnostic criteria towards idiopathic calcium oxalate urolithiasis in children.

P-018 COMPARISON BETWEEN NGAL, CYSTATINE C AND FAB URINARY EXCRETION AS A MARKERS OF AN ACUTE KIDNEY INJURY AFTER CONTRAST ADMINISTRATION

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Introduction: Prevention of pediatric contrast nephropathy might be challenging because of the lack of early and widely available biochemical markers to detect acute kidney injury (AKI). RIFLE score has proven its role in epidemiological analyses with less bedsite significance. Newer and more sensitive markers were postulated to increase the rate of early detection and to improve prevention of AKI. We aimed to compare the changes in urinary excretion of neutrophil gelatinase (uNGAL), cystatin C (uCysC) and liver-type fatty acid-binding protein(L-FAB) after administration of contrast media during cardiac catheterisation in children.
Material and methods: The study group consisted of 50 children (35 boys and 15 girls) with congenital heart defects qualified for the scheduled cardiac catheterisation. Median age was 53 months (range 0–18 y). Patients with preexisting renal injury or malformations of the urinary tract were excluded. The study was granted by Polish Mothers Memorial Hospital Reserach Institute (grant 2014/IV/42-GW) Biochemical markers were analysed in absolute values and in relation to urine creatinine. Samples were collected before the procedure and in the 2,6,24 and 48 h after contrast injection.
Results: Significant changes were noticed in the excretion of uCysC, NGAL/Cr and CysC/Cr ratio. NGAL/Cr ratio rose in the 2 h after contrast administration with reduction after 6 h (Me: 9.42 vs 4.91). Significant rise in the urinary CysC and CysC/Cr ratio were noted only after 48 h. L-FAB concentration was below the detection threshold in the majority of patients. Only 21 children had it above the threshold after 24 h. A correlation was detected between contrast media dose and urinary CysC ($R = 0.42$; $p < 0.05$) at the 48 h time point.
Conclusions: Based on the results of the study, we postulate that NGAL and CysC might be considered as promissing urinary markers of contrast induced AKI. NGAL proved to be more sensitive in first hours after invasive procedures.

P-019 CONGENITAL ABNORMALITIES OF KIDNEY AND URINARY TRACT: ANTENATAL AND POSTNATAL DIAGNOSIS

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Introduction: Congenital abnormalities of kidney and urinary tract (CAKUT) are an important cause of childhood morbidity. CAKUT can lead to end-stage renal disease in children and also cause subsequent renal problems in adulthood. Many of congenital abnormalities are detected in the antenatal or immediate postnatal.
The aim of the study was to analyze the occurrence of CAKUT and the usefulness of abdominal ultrasound (USG) screening in the population of 1752 newborns hospitalized in one neonatal center in the years 2014–2015.
Material and methods: The clinical databases concerning newborns and their USG results were analyzed. In 2014 USG was performed only in newborns with medical indications. In 2015 USG was performed also as screening in healthy term newborns.
Results:

Year	2014	2015
Total number of tested newborns	537	1215
Newborns born >37 1/7 weeks of pregnancy	333 (62%)	955 (79%)
Newborns born prematurely 33 0/7–37 0/7	146 (27%)	186 (15%)
Newborns born prematurely 22 0/7–32 6/7	58 (11%)	74 (6%)

There were 187 abnormalities in the low-risk population with normal prenatal screening. In most cases CAKUT were benign in the form of small to medium-degree enlargement of the kidney pelvis. In addition developing ovarian cysts and spleen cysts were diagnosed. However, neuroblastoma in two cases and renal agenesis in two cases were detected. In 71 children prenatal abnormalities were confirmed. In 31 (44%) of those newborns USG showed severe urinary system abnormalities.
In total, CAKUT were observed in 15% of tested newborns. They required intensive treatment or implementation of further diagnostics and follow-up. Such a high percentage of CAKUT may result from screening of newborns whose mothers were hospitalized at the highest referral center.

Conclusions: Postnatal abdominal ultrasound in newborns as a complement to antenatal screening ultrasonography allows detecting additional pathologies that may not produce clinical symptoms in early life.

P-020 AMBULATORY BLOOD PRESSURE MONITORING PARAMETERS IN OBESE AND NON-OBESE HYPERTENSIVE CHILDREN AND ADOLESCENTS

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Introduction: We aim to asses the effect of obesity on ambulatory blood pressure monitoring(ABPM) parameters in officially hypertensive children and adolescents.
Material and methods: Children and adolescents who have BP measurements > 95p on three different occasions in office measurements and referred for ABPM between January 2010 and December 2013 were included into the study. Children with secondary hypertension and those with solitary kidneys were excluded. Patients with a BMI ≥95p were grouped as obese and those with <95p were grouped as non-obese. Age, gender, prematurity, height SDS, urea, serum creatinine, uric acid, left ventricular mass index(LVMI), hypertensive retinopathy(HTRP) findings and proteinuria levels were recorded. As patients were in the different age groups, SDS levels were calculated for APBM measurements including 24 h, daytime and nighttime systolic, diastolic and mean arterial blood pressures (SBP, DBP and MAP, respectively). Laboratory and clinical findings and ABPM values were compared between the two groups.
Results: There were 266 (M/F:148/118) patients with primary hypertension. Of those, 192 were obese and 72 were non-obese; and 24 (7 obese and 17 non-obese) had white coat hypertension. Age, gender, birth weight, urea, serum creatinine levels were similar between the groups. Prematurity, height SDS, uric acid and LVMI were higher in the obese group; however, ratio of hyperuricamia, LVH and HTRP levels were similar between the groups. The rate of WCH is higher in the non-obese group. Twenty-four hour, daytime and nighttime SBP, DBP and MAP levels were significantly higher in the obese group ($p < 0.01$). Systolic dipping is lower in obese patients (p:0.01). When patients with WCH were excluded, the results remained the same.
Conclusions: In officially hypertensive children and adolescents, WCH is more common in non-obese patients and obesity is associated with higher ABPM parameters irrelevant to WCH.

P-021 PREDICTING THE RISK OF POOR OUTCOMES IN CHILDREN WITH POST-DIARRHEAL HEMOLYTIC UREMIC SYNDROME

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Introduction: Aim of study was to determine the variants and frequency of renal-related sequelae of post-diarrheal HUS (HUS D+) in children, to identify risk factors of unfavorable outcomes and their prognostic significance.
Material and methods: The study included 124 patients who underwent HUS D+ in 2005–2014 and at least 1 year of follow-up after onset, as well as those died and reached end-stage renal disease (ESRD) during this period of time. Allocated groups with favorable ($n = 67$, median follow-up: 4.5y) and unfavorable outcomes ($n = 57$, follow-up: 4,5).
Results: The incidence of the renal-related sequelae of HUS D+ was diagnosed in 46% of patients (severe: death and ESRD 5.7%; mild-to-moderate 40.3% of: proteinuria 12.4%, microalbuminuria 15.8%, arterial hypertension 36.8%, reduced GFR 9.4%). The most significant factors for unfavorable outcomes and thresholds of the risk of their development: the presence of anuria (OR 3.84; $p = 0{,}001$), leukocytosis >10.1×10^9/l (OR 3.0; $p < 0.05$), exceeding the upper limit of normal range of alanine aminotransferase (Alt) more than 1.3 times (OR 2.62; $p < 0.05$), need for dialysis (OR 3.6; $p < 0.01$) and mechanical ventilation (MV) (OR 3.92; $p < 0.01$), involvement of the central nervous system (CNS) (OR 6.87; $p < 0.001$). The highest predictive value inherent for the duration of anuria (AUS = 0,73, $p < 0{,}001$) and dialysis (AUS = 0.71, $p < 0.001$), the degree of increase in Alt (AUS = 0.71, $p < 0.001$) and blood leukocytes (AUS = 0.7, $p < 0.001$), less so for the presence of CNS damage (AUS = 0,62, $p < 0{,}05$) and need for MV (AUS = 0.62, $p < 0.05$).
Conclusions: Identification of early predictors of adverse outcome in HUS allows creating the groups of risk that require careful and prolonged follow-up after the acute period of the disease.

P-022 KIDNEYS INVOLVEMENT IN CHILDREN WITH SYSTEMIC DISEASES: CONTRIBUTION OF CARDIOVASCULAR COMORBIDITY

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Introduction: Despite the significant progress achieved in the treatment and outcome of children with renal involvement due to systemic diseases, many questions remain in the early detection of cardiovascular complications (CVC), the leading cause of death in these patients in adulthood. This study is contributed to find out the risk of developing CVC in patients with lupus(LN), IgA Henoch-Schonlein purpura nephritis (HSPN) and ANCA-associated nephritis.
Material and methods: 59 children were enrolled: 27 with LN (2 boys, 7–17 yrs), 27 with HSPN (14 boys, 3–17 yrs) and 5 with ANCA-nephritis (2 boys, 6–17 yrs), all had morphological verification of nephritis. As a control 37 healthy children were examined. Twenty-four hour monitoring of blood pressure(BP), ECHO-CG, carotid intima media thickness(cIMT), left ventricles mass index (LVMI), relative thickness of left ventricles wall (RTLV), body mass index (BMI), serum lipids levels, glucose, uric acide, eGFR and markers of vascular endothelial dysfunction VEGF and TGF1β were measured.
Results: Arterial hypertension(AG) was observed in 43/59 (73%) of children with glomerulopathies: in 23/27 of LN (86%), in 100% of ANCA and 13/27 HSPN (48%), required of an average 3 hypotensive drugs. Dilatations of the LV in 24%, reduced ejection fraction in 2.1% of all patients were seen. LVMI, RTLV, BMI and cIMT were higher compared with healthy ($p < 0.05$). In patients with nephritis serum concentration of VEGF and TGF1β correlated with AG. The mean serum cholesterol level was 5.8 mmol/l in LN, 6.94 in ANCA and 5.16 in HSPN ($p < 0.05$), lipoproteins of low and very low density prevailed. The mean serum glucose level was not significantly higher than in healthy, in contrast to the level of uric acid ($p < 0.05$), especially in ANCA nephritis ($p < 0.01$).
Conclusions: In patients with LN, HSPN and ANCA-associated nephritis abnormalities in serum lipids level were correlated with disease activity and duration, younger age of diagnosis, mean cIMT, eGFR, increased systolic and diastolic BP, BMI, LVMI, RTLV, concentration of VEGF and TGF1β. Such patients are at high risk of early development of cardiovascular complications requiring early correction.

P-023 LOOKS CAN BE DECEIVING: CAN TYPICAL HEMOLYTIC UREMIC SYNDROME BENEFIT FROM ECULIZUMAB?

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Introduction: Hemolytic Uremic Syndrome (HUS) represents one of the major causes of acute renal failure in children. Shiga toxin-producing *Escherichia coli* related forms (STEC-HUS) represent the majority of cases. The aim of this project was to analyze data of the last 7 years concerning Piedmont region, to have an updated screenshot of disease clinical impact and outcomes.
Material and methods: Single-center retrospective study on 45 children with HUS, hospitalized between January 2010 and December 2016 at the Regina Margherita Children's Hospital. The population was divided according to the presence of diarrhea at onset (D+/D-); D+ group was divided in Shiga toxin positive (D + STEC+) and negative (D + STEC-). Genetic analysis was conducted in D + STEC+ with severe course and D- without infection.
Results: 41 patients (91.1%) were D+ and 4 patients (8.9%) D-. 36 patients (87.8%) D+ were positive for STEC. In D- group 2 patients had *Streptococcus pneumoniae*, the others had no history of infection. Five patients of 45 (11%) were treated with eculizumab; 4 of them were D + STEC+ (2 presented prolonged renal damage, the others severe neurological involvement). The 2 patients with renal damage developed cronic kidney disease and they are continuing infusions of eculizumab; patients with neurological involvement presented rapid resolution of symptoms, without sequelae. The D- patient had full recovery. All of treated patients presented genetic polymorphisms in complement alternative pathway (Membrane Cofactor Protein, factor H, factor B, C3), known to be associated with increased risk of HUS.
Conclusions: In STEC-HUS a severe clinical presentation, in particular with serious neurological involvement, may justify the use of Eculizumab while waiting for confirmation of a risk genetic asset.

P-024 GRANULOMATOUS INTERSTITIAL NEPHRITIS PRESENTING AS AN EXTRA-INTESTINAL MANIFESTATION OF CROHN'S DISEASE IN A PAEDIATRIC PATIENT

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Introduction: Renal, predominantly nephrolithiasis and tubulointerstitial nephritis (TIN), manifestations of Crohn's Disease (CD) are well recognised. However, granulomatous interstitial nephritis (GIN) remains a rare association with only 3 cases reported in literature under 18 years of age.
Material and methods: We report a paediatric patient with CD, presenting with renal impairment and concurrent GIN to further guide management of renal involvement in CD.
Results: A 13 year old boy with gastric and ileocolonic CD, diagnosed when 5 years old, received treatment for very difficult colitis with 5-aminosalicylate (5-ASA), methotrexate, infliximab and adalimumab. Renal function was normal and remained so until the age of 12 years. Over the following year, he had a relapsing course of Crohn's colitis, treated with 5-ASA suppositories, which were discontinued, and weekly adalimumab. Renal function (eGFR) declined from 117 to 15 ml/min/1.73 m2 and was treated with high dose steroids. Histology showed TIN and eGFR settled at 30 ml/min/1.73 m2. A further relapse of colitis occurred with deterioration in renal impairment (eGFR 20 ml/min/1.73 m2). Repeat biopsy showed histological evidence of granulomatous disease. High dose and a prolonged course of steroids were used with good effect. Crohn's remission has been achieved over the last 24 months with stabilisation of renal function at 30 ml/min/1.73 m2.
Conclusions: We present a case of difficult Crohn's with renal impairment and histological evidence of granulomatous interstitial nephritis. Whilst 5-ASA treatment is recognised as a causative factor in TIN, the second renal function decline in our patient was not associated with 5-ASA treatment, but occurred concurrently with a relapse of CD. A clinical correlation of renal impairment associated with relapsing CD was noted with stabilisation of renal function with CD remission. To our knowledge, this is the youngest case reported of GIN and supports granulomatous interstitial nephritis as an extra-intestinal manifestation of CD.

P-025 GHRELIN AND LEPTIN LEVELS IN RELATION TO BODY FAT MASS IN CHILDREN WITH CHRONIC KIDNEY DISEASE

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Introduction: The mechanism causing decreased appetite in uremia is not fully understood. Hormonal and metabolic abnormalities, particularly alterations in appetite-regulating hormones have been suggested as causative factors. The aim of the present study was to evaluate serum concentrations of total ghrelin and leptin, and their possible interactions with fat

mass, inflammation, insulin resistance and 25(OH) vitamin D levels in children with chronic kidney disease (CKD).
Material and methods: The study population consisted of 94 patients with CKD [20 patients with CKD stages 2–4, 39 patients on chronic dialysis (22 PD, 17 HD) and 35 renal transplant (RTx) recipients] and 18 healthy controls. Nutritional status was assessed by measuring body mass index (BMI) and multi-frequency bioimpedance analysis (BIA). Fat mass was estimated by the BIA method. Serum total ghrelin, leptin and IL-6 were measured by ELISA method in all patients and controls. High sensitive (hs)-CRP, fasting glucose and insulin, and 25(OH) vitamin D levels were recorded from patients' file. Insulin resistance was estimated by the homeostasis model assessment of insulin resistance (HOMA-IR).
Results: The mean total ghrelin level was significantly higher in patients on dialysis than the controls, CKD patients and RTx recipients ($p < 0.001$ for all). There was no difference considering total ghrelin levels between controls and CKD or RTx recipients. Total ghrelin levels in dialysis patients were negatively correlated with fat mass ($p = 0.002$), fat mass (%)-z score for height-age ($p = 0.004$), BMI-SDS for height-age ($p = 0.015$), albumin ($p = 0.030$) and 25(OH) vitamin D level ($p = 0.036$). Multivariate analysis showed that a lower fat mass was the only independent predictor of increased total ghrelin levels in dialysis patients ($p = 0.015$). Leptin levels were also higher in patient groups compared with healthy controls but the differences were not statistically significant. Higher serum leptin levels in all patients were independently associated with increased fat mass ($p < 0.001$).
Conclusions: Poor nutritional status appears to be the most important cause of high total ghrelin levels in dialysis patients. Leptin is closely associated with increased fat mass in all CKD patients.

P-026 ACUTE KIDNEY INJURY IN CHILDREN AFTER CARDIAC CATHETERISATION

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Introduction: Acute kidney injury (AKI) often occurs in the course of invasive cardiac surgery procedures in children. Invasive cardiology procesures with contrast media administration also might have a negative influence on patients kidney function. The ouf our study was to establish the incidence and recovery rate of AKI after cardiac catheterisation in children with complicated cardiac malformations.
Material and methods: The study was a prospective observational project amongst children with complicated cardiac malformation, who were qualified for diagnostic cardiac catheterisation before cardiac surgery. The study group consisted initially of 72 children aged (0-18y). Eighteen children were excluded because of consent withdrawal. Finally, 50 children (35 M:15F) were analysed. They uderwent standard clinical procedure of cardiac catherisation (11 for diagnostics and 39 for combined diagnostics and intervention). Thirty-two patients had at least on catheterizations before. Eleven children had AKI during previous cardiac surgery procedures. Twenty-six children required chronic diuretic treatment. Patients with preexisting chronic kidney injury or malfomations were excluded. pRIFLE score was applied initially for AKI assessment. Data were gathered before procedure and after 2,6,24 and 48 h. Additionally, serum neutrophil gelatinase (NGAL) concentration was assessed (ELISA). The study was approved by the local EC and all caregivers gave an informed consent. The study was granted by Polish Mothers Memorial Hospital Reserach Institute - internal grant 2014/IV/42-GW.
Results: AKI by pRIFLE criteria was detected in 22 patients (44% - R-17/I-5). It occurred within 24 h of observation in 16 patients, whereas in 6 subjects later between 25 and 48 h post contrast administration. We observed a significant ($p < 0.05$) rise in NGAL concentration in serum after 2 and 6 h (41, and 46 ng/ml) with subsequent decrease within 24 h to 32 ng/ml. Cystatin C rose significantly ($p < 0.05$) after 24 h (816 ± 1139 ng/ml) with decrease after 48 h. Follow-up analysis showed normal renal function in all children. Only 2/16 AKI children had decreased urine output. No significant decrease in cardiac function was noted, however NT-proBNP concentration was elevated before procedure 836 ± 1369 pg/ml.
Conclusions: We showed that post-contrast AKI had high incidence in children with complicated cardiac malformations after cardiac catherisation. Urine output criteria proved little value because of routine diuretic administration in this selected group of patients.

P-027 *Escherichia coli*-ASSOCIATED HEMOLYTIC UREMIC SYNDROME (HUS) AND CHRONIC HEPATOCELLULAR CHOLESTASIS: A REPORT ON 3 PEDIATRIC CASES

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Introduction: Hepatic lesions of shigatoxin-associated HUS (STEC-HUS) are uncommon. Clinical and biological presentations are variable, from minor disturbances of liver function to more severe lesions such as obstruction of biliary ducts or cholangitis.
Material and methods: We report on three observations of STEC-HUS with delayed hepatic involvement, two of them being severe (cirrhosis, waiting list for liver transplantation).
Results: Three children of 2, 6 and 11 months presented STEC-HUS caused respectively by O157H7, O26 and a double contamination at O145 and O80. Extra renal (cardiovascular, neurological and gastro-intestinal) manifestations occurred in the 3 patients. In 2 cases out of 3, there was initially a moderate cytolysis without cholestasis. These 3 children received eculizumab (1 to 3 injections) in addition to symptomatic management (renal replacement therapy and transfusions) due to age and/or severity of the disease.
Approximately 15 days after the onset of the STEC-HUS, all of them developed a severe secondary sclerosing cholangitis while others manifestations improved dramatically. Thorough investigations for underlying causes of liver disease were performed, ruling out other etiologies. Abdominal ultrasounds demonstrated a normal-sized liver with normal parenchymal appearance but a gallbladder with a very thick wall.
Liver biopsy was performed in the 3 patients, all of them displaying a cholangitic reaction with secondary sclerosing cholangitis. There was no histological rationale for post-ischemic chronic lesions or vascular lesions.
Bilirubin levels and GGT slightly improved but 2/3 patients developed cirrhosis and are currently waiting for a liver transplantation (i.e., 8 months after the onset of STEC-HUS), despite a normal renal function (serum creatinine between 19 and 35 μmol/l) and mild proteinuria (treated with ACEi).
Conclusions: These observations of severe hepatic involvement after STEC-HUS treated with eculizumab raise the question of eculizumab hepatotoxicity, in the absence of any rationale for "post-shock" liver lesions, liver microangiopathy or other etiologies for cholestasis.

P-028 HUS IN CHILDREN IN ORENBURG REGION

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Introduction: The aim of the study was to determine clinical manifestations and treatment of children with HUS in Orenburg region.
Material and methods: We examined 51cases of HUS in children in Orenburg region in period 2000–2015 years.
Results: 1 child had atypical HUS. May and June were peak months of disease in year. Girls (53%,*n* = 27) prevailed in sex structure of HUS. Infants and babies (82.3%, *n* = 42) dominated in age structure of HUS. Diarrhea syndrome preceded development of disease in 98% of patients (*n* = 50). Forty percent of them had hemorrhagic colitist (*n* = 20), 60% (*n* = 31) abdominal pain. The degree of renal damage in children with HUS was different: 62.8% of patients had oliguric stage of ARI (*n* = 32), 37.2% of patients had anuria (*n* = 19). Oliguria was stopped for 7 days to more than half of the cases (56.2%, *n* = 18). Duration oliguria to 2 weeks had 8children, and 1 child (3.1%) more than 1 month. Anuria was stopped during second week of ARI in 21% of children (*n* = 4). In 1 child (5.3%) anuria lasted more than 1 month. Patients with HUS had extrarenal syndromes: damage of gastrointestinal tract (60%, *n* = 31), central nervous system, cardiovascular system (33, 3%, *n* = 17), respiratory tract (*n* = 8, 15.7%), hemostasis system (*n* = 19, 37.2%), intoxication syndrome. Pathology of nervous system was presented with violation of consciousness (32.4%, *n* = 16); convulsions (23.5%, *n* = 12), acute cerebrovascular accident (*n* = 3). 76.5% (*n* = 39) of children with HUS had RRT: more than half of patients had hemodialysis, second highest rate of application was combination of hemodialysis and peritoneal dialysis. Two children had only peritoneal dialysis, in 2 cases hemodialysis combined with plasmapheresis. In 12 children (23.5%) ARI was treated without RRT. HUS mortality in the Orenburg region in period 2000–2015 was 6 children (11.8%).
Conclusions: HUS in children in Orenburg region was presented by typical HUS with ARI which was treated by RRT.

P-029 LIFELONG FOLLOW UP POST AKI IN PICU

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Introduction: Historically across the EMEESY network there was no defined pathway for follow up post AKI. In 2014 a new pathway was introduced. Initially this was for all patients receiving RRT for AKI within PICU, for 5 years post AKI. This has now been lengthened to lifelong and has been extended to all stage 3 AKI. Growing evidence suggests there is increased risk of developing CKD post AKI1,2. We also know CKD is a progressive, yet modifiable disease. By offering this lifelong follow up pathway of care, we hope to modify the progression of the disease post AKI.
Material and methods: Patients have been identified for follow up by local link nurses within each centre. This has been aided by the introduction of the new PICANet Renal audit, as each centre within the network is a pilot site for this audit. This follow up pathway involves: • Creatinine check year 1 & 5 • Annual BP check • Annual urine dipstick (+/− protein:creatinine ratio) • Formal GFR 1 year and 5 years • For 5 years this will take place within their local hospital across the network clinics and outcomes collated within the Regional Tertiary Nephrology Centre. After this, if there are no other concerns, the child will be discharged to their GP for ongoing lifelong follow up.
Results: Within the first 3 years of this project 37 patients have been referred for follow up across the EMEESY network. The data and information collated thus far is limited, not allowing us to draw conclusions at this point.
Conclusions: In time through adapting our systems for identification of AKI across the network we hope this pathway will become ingrained in everyday practice. The ultimate aim would be to lessen the severity of CKD post AKI, the lifelong follow up of this cohort of patients will allow this to be assessed.

P-030 SIGNIFICANCE OF EXAMINATION OF CONCENTRATION OF IL-10 IN 24 H URINE FOR EARLY DIAGNOSTIC OF RENAL SCARRING IN PATIENTS WITH VESICOURETERIC REFLUX

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Introduction: The aim of the study was to determine concentration of IL-10 in urine of patients with vesicoureteric reflux (VUR) and reflux nephropathy A (RN A) for early diagnostic of renal scarring.
Material and methods: We examined 60 children with RN A and VUR. All children were comparable on gender and age. All patients underwent ultrasound, X-ray and DMSA scan. We examined concentration of IL-10 in 24 h urine of patients by ELISA. Children were divided into 2 groups: I – with unilateral RN A according to classification of Smellie J. et all, 1975 (*n* = 30); II – with VUR without renal damage (*n* = 30).
Results: We established that data of concentration of IL-10 in 24 h urine of patients with VUR without renal damage was 19.23 ± 0.32 pg/ml. Concentration of IL-10 in 24 h urine of patients with unilateral RN A was 11.98 ± 0,24 pg/ml. The ranges of concentration of IL-10 in 24 h urine of patients with VUR without renal damage were significant different with concentration of IL-10 in 24 h urine of patients with RN A ($p < 0.05$). So, we determined that concentration of IL-10 in 24 h urine decreased in process of renal scarring.
Conclusions: Concentration of IL-10 in 24 h urine can be used for early diagnostic of renal scarring in children with VUR.

P-031 EPIDEMIOLOGICAL AND CLINICAL CHARACTERISTICS OF PATIENTS WITH HEMORRHAGIC FEVER WITH RENAL SYNDROME

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Introduction: Hemorrhagic fever with renal syndrome (HFRS) is a rare viral disease caused by Hanta viruses. It occurs sporadically in Europe. Pathogenic mechanism is based on the increased permeability of the endothelial cells and results in hemorrhagic diathesis. Acute kidney injury is the main clinical manifestation. The aim of our study was to analyze clinical features of HFRS in patients treated in tertiary care center, between 1990 and 2016 year.
Material and methods: We retrieved data of patients diagnosed with HFRS and analyzed their epidemiological characteristics (place of origin, sex, age, serotype of the virus), clinical course of the disease (presentation, duration of symptoms, laboratory findings, stage of acute kidney injury, necessity of dialysis) and the outcome.
Results: 12 patients, 7 from northern Montenegro and eastern Bosnia and Herzegovina and 5 from western Serbia, 8 males and 4 females, at the age 8 to 15 (12.6 ± 2.1) years. Indirect immunofluorescence for Hantaviruses was positive for serotypes Hantaan, Puumala, Seoul and Dobrava/Belgrade in 7 patients and for Hantaan, Seoul and Dobrava/Belgrade in 5 patients. Initial symptoms were fever and abdominal pain in 100%, lumbar pain and myalgia in 91%, vomiting in 73% and headache in 64%. External symptoms of hemorrhagic diathesis were rare (petechia 24%, subconjunctival suffusions 14% and epistaxis in 9%). Arterial hypertension and oliguria was found in 55%, bradycardia not related to hypervolemia 45%, anuria 25%, macroscopic hematuria in 16% of patients. Thrombocytopenia and proteinuria were in 90%, microscopic hematuria in 55%, low C3 in 36%. Serum creatinine level was from 186 to 839 (486 ± 204) μmoll/l. Continuous renal replacement therapy was

performed in 3 patients. Time for the normalization of renal function was 6 to 21 (mean 11.7 ± 4.4) days. Complete restoration of renal function was verified in 86%. All patients survived (100%).
Conclusions: Hemorrhagic fever with renal syndrome in our region had sporadic nature with endemic characteristics. All patients had fever and abdominal pain, half of them arterial hypertension, bradycardia, and oliguria. Dialysis was necessary in one fourth of the patients. Survival was absolute.

P-032 SOLUBLE ADHESION MOLECULES IN CHILDREN WITH TYPICAL HEMOLYTIC-UREMIC SYNDROME (THUS)

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Introduction: The pathogenesis of tHUS depends on the endothelial damage and disfunction caused by shiga-toxin. As a result the expression of endothelium-derived inter-cellular adhesion molecule, vascular cells adhesion molecules and their soluble fractions (sICAM and sVCAM) may change during acute stage of tHUS accordingly to severity of the disease. The aim of the study was to determine the serum levels of sICAM and sVCAM and their prognostic properties in children with tHUS.
Material and methods: We examined 17 children (9 boys) aged 6–128 months (median 33 months) on 2nd-4th day after onset of tHUS, and 10 children (5 boys) of 20–78 months (median 42 months) who recovered from tHUS 6 months up to 5 years before the study. The diagnosis of tHUS was established on the basis of hemolytic anemia, thrombocytopenia, acute renal injury developed after prodromal haemocolitis. Of 17pts with acute tHUS 14 were anuric, 3 had oliguria. Fifteen needed renal replacement therapy. Ten pts. of comparison group were examined in apparently healthy state, with no symptoms of inter-current infection. sICAM and sVCAM in blood serum were determined by an immunoensymatic method on the PC Lab analyzer (Adaltis, Italy) using Bender Medsystems test systems.
Results: In children with acute tHUS sICAM was lower and sVCAM higher than in comparison group (median, Q25-Q75): sICAM 285 (227–297) pg/ml vs 335 (308–408) pg/ml, sVCAM 705 (505–1230) pg/ml vs 355 (325–440) pg/ml respectively. Serum concentration of sVCAM varied widely from 360 to 5200 pg/ml in children with acute tHUS. However, comparing 8 pts. with normal sVCAM (median 482, 372–565 pg/ml) with 9 pts. with high sVCAM values (median1230, 1085–1825 pg/ml) we did not reveal significant differences in initial hemoglobin, platelet count, leukocytes, LDH as well as in the duration of anuria and dialysis, in the serum urea and creatinine at the day of discharge. We did not reveal any significant correlations of above-mentioned data with serum concentrations of sICAM and sVCAM.The direct correlation of sICAM with the duration of thrombocytopenia was revealed ($r = 0{,}63$, $p < 0.05$).
Conclusions: Serum level of sICAM is lower, and sVCAM is higher in children with acute tHUS than in healthy children with the history of tHUS. However the prognostic value of these biomarkers remains unclear.

P-033 ACUTE RENAL FAILURE IN PEDIATRIC PATIENTS WITH SEPSIS

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Introduction: Assessment of acute renal failure (ARF) development, causative factors and outcomes in pediatric patients with sepsis can facilitate prognosis and early recognition of this condition.
Material and methods: Data of 206 pediatric patients (female- 44.7%, male- 55.3%) aged 1 month- 18 years were analysed retrospectively. All patients, treated in the Hospital of Lithuanian University of Health Sciences Kauno klinikos in 2013–2016 because of sepsis, were included in the study. Patients were divided in two groups: I group - sepsis (182), II- sepsis with multiple organ dysfunction (24). Results were compared between groups ($p < 0.05$).
Results: Mean age in groups: **I**- 4.3 ± 0.36, **II**- 4.7 ± 1.20 years 196 (95.1%) patients recovered, 10 (4.9%) - died. All cases of death (10/24) were in group **II** patients ($p < 0.001$), 7 had ARF ($p < 0.001$). ARF was diagnosed in 22 (10.7%) patients: **I**- 5 (2.7%), **II**- 17 (70.8%) ($p < 0.001$). 9 (4.4%) patients needed renal replacement therapy. Most patients who had ARF were 1–4 years old (9–40.9%). No significant difference of outcomes was found between patients who had concomitant diseases (59–28.6%) and those who didn't (147–71.3%) ($p = 0.154$). Mean value of days from the onset of symptoms to hospitalization in groups: **I**- 1.9 ± 2.70; **II**- 0.75 ± 1.03 ($p < 0.05$). Group **II** patients were treated in intensive care unit significantly more often (23–95.8% vs. 66–36.3%) and had longer stays on average (9.62 ± 14.17 vs. 1.3 ± 3.2 days) ($p < 0.001$). *N.meningitidis* (5–20.8%) was the prevalent pathogen in blood cultures in group **II** and among patients with ARF (6–27.3% vs. 8–4.3%) ($p < 0.05$).
Conclusions: 1. Patients with sepsis related multiple organ dysfunction (group II) are diagnosed with ARF significantly more often and have worse overall outcomes. 2. The risk of death in patients with ARF is more prevalent. 3. *N.meningitidis* is the leading causative factor of multiple organ dysfunction including ARF.

P-034 OUR EXPERIENCE IN KIDNEY BIOPSIES DONE BY PEDIATRIC NEPHROLOGIST WITH THE OWN ULTRASOUND GUIDENCE IN CHILDREN

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Introduction: Kidney biopsy (bx) has great impact on diagnosis and management of renal disease. Percutaneous kidney biopsy performed with ultrasound (US) is a safe and widely used tool for kidney bx even in children. In this study we aimed to evaluate effectiveness of our pediatric kidney bx results and complication rate related with procedure and also, compared our results with biopsy standards defined by the British Association of Pediatric Nephrology (BAPN).
Material and methods: The kidney bx performed by nephrologists and nephrology fellows with percutaneous procedure under their own US guidance in Izmir Tepecik Training and Research Hospital between December 2013 and December 2015 were retrospectively evaluated. Age, gender, bx indications, bx type (native/transplant), number of needle penetrations, tissue adequacy, biopsy diagnoses, biopsy complications, first 24 h follow-up were taken from medical records. The bx done by nephrologists and nephrology fellows were evaluated separately. Our results were compared with BAPN standarts.
Results: 91 patients [48 male (52.8%)] were included in the study. The mean age of at the bx was 157.8 ± 58.2 months.132 bx were performed in 91 patients. Twenty-five bx were made by specialists and 107 bx were made by fellows. The most common indication was the protocol bx of the transplanted kidney(51.5%). 92 (69.7%) biopsy procedures were done in first needle puncture.23 of the 25 bx made by the nephrologists and 104 of the 107 bx made by fellows were appropriate to the standards. There were no complication in 108(81.8%),minor complication in 23(17.4%) and only 1 major complication recorded.There was only significant difference seen in minor complications rate among practitioners($p < 0.05$).
Conclusions: Percutaneous kidney bx technique with the guidence of US by pediatric nephrologist can be considered as an easy to perform and safe method for the diagnosis in most of the patients.

P-035 VASCULAR ENDOTHELIAL GROWTH FACTOR IN PAEDIATRIC RENAL TRANSPLANT RECIPIENTS

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Introduction: Vascular endothelial growth factor (VEGF) is an endothelial-specific growth factor that promotes endothelial cell proliferation, differentiation and survival, mediates endothelium-dependent vasodilatation, induces microvascular hyperpermeability and participates in interstitial matrix remodelling. VEGF expression is most prominent in glomerular podocytes and in tubular epithelial cells in the kidney. There remains ongoing work on the role of VEGF in normal renal physiology and in animal models of transplantation where VEGF production may influence susceptibility to acute rejection and chronic allograft damage.
Material and methods: Prospective study measuring VEGF-C by Human VEGF Quantikine ELISA Kit (R&D Systems, Oxon, UK) in 5 paediatric renal transplant recipients (pRTR) at 15 timepoints and compared to 16 age-matched and sex-matched healthy paediatric controls.
Results: 5 pRTR (60% (3) male) aged 4.4–15.3 (median 10.5) years of whom 60% (3) had ESKD due to CAKUT underwent 15 VEGF measurements at 0.2–12.1 (median 2.1) months after first kidney only transplantation (100% (5) living-related and 80% (4) pre-emptive) with eGFR of 52.4–87.5 (median 78.4) mls/min/1.73m^2 at follow-up of 2.3–6.7 (median 6.3) years. Patients underwent 1–2 (median 2) percutaneous renal transplant biopsies showing evidence of chronic changes but there were no biopsy-proven acute rejection episodes. Mean VEGF-C levels were 4.27 +/− 1.04 ng/ml in pRTR and 4.33 +/− 1.47 ng/ml in the healthy controls.
Conclusions: VEGF levels were stable in our pRTR and comparable to healthy controls although this was a stable cohort of living related renal transplant recipients who did not have any biopsy-proven acute rejection episodes, which are normally characterised by an acute inflammatory lymphocytic infiltrate, tubulitis and endothelialitis. VEGF may be important in the generation of this inflammatory response and the general controlled variation in VEGF production may influence susceptibility to acute allograft rejection. This hypothesis needs to be investigated further in pRTR undergoing episodes of acute allograft rejection.

P-036 RISK FACTORS OF LEFT VENTRICULAR MASS INCREASE IN CHILDREN WITH CHRONIC KIDNEY DISEASE

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Introduction: Left ventricular hypertrophy (LVH) is a major risk factor of cardiovascular complications. The study aimed to identify risk factors that contribute to LVH progression in children with chronic kidney disease (CKD) during 1 year observation.
Material and methods: The study was conducted in a group of 48 children (17 girls and 31 boys) with CKD stage 1 to 5. The patients' mean age was 11 years and mean GFR was 32 ml/min/1.73m^2. Serum creatinine, BNP, ADMA and oxyLDL levels were measured. Ambulatory blood pressure measurements (ABPM) were performed. Echocardiographic examinations were carried out with a HP 5500 device. The 95th percentile of LV mass index relative to height age was used to define LVH.
Results: LVH was detected in 22 children. There were no significant differences in biochemical parameters and blood pressure values between children with the progression vs regression of LVMI (change >15%). Twenty-six children without LVH at first observation developed a LVMI increase of >15% during 12-month observation. In this group lower height (124.7 vs 134.4 cm; $p = 0.001$) and significantly higher night blood pressure (BP) values were observed: systolic (100.0 vs. 96.8 mmHg; $p = 0.024$), diastolic BP (57.0 vs. 54.7 mmHg; $p = 0.006$) and mean arterial pressure (73.7 vs. 70.0 mmHg, $p = 0.001$). In addition, higher systolic, diastolic BP and MAP values in 24 h measurements were found.
Conclusions: BNP concentration is not a predictive index of left ventricular hypertrophy. Night blood pressure is a significant risk factor of LVH development in children with chronic kidney disease. Regular ABPM measurements and intensive hypotensive treatment are important elements of cardioprotection.

P-037 THE ROLE OF THE URINARY ANTIMICROBIAL PEPTIDE CATHELICIDIN LEVELS IN EARLY DIAGNOSIS OF URINARY TRACT INFECTION, DIFFERENTIATION OF CYSTITIS-PYELONEPHRITIS AND PREDICTION OF THE RESISTANT MICROORGANISMS

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Introduction: Urinary tract infections(UTI) can lead to renal scarring resulting in hypertension and end-stage renal failure. Therefore,different markers such as antimicrobial peptide cathelicidin are studied for early diagnosis of UTI.
In this study, we investigated the role of urinary cathelicidin levels(UCL) in early diagnosis of UTI,differentiation of cystitis-pyelonephritis and prediction of the resistant microorganisms.
Material and methods: 109 patients(58 female, 23 male) with suspected UTI and 63 healthy children were included in the study.On admission,complete urine analysis and urine cultures of the patients were studied.Demographic data and associated renal diseases were recorded.Fifty eight patients were included in the cystitis subgroup,17 in the pyelonephritis and 34 in the sterile pyuria subgroup.UCL were measured in all patients and control group.
Results: The median(IQR) age of patients was 84(72;1–216) months.UCL levels in the patient and and control groups were 14.99 ± 5.43 and 15.18 ± 4.52 ng/ml respectively,and no significant difference was present.There was no significant difference in UCL of the subgroups compared with each other and the control group($p > 0.05$).In cystitis subgroup,UCL in patients infected with E Coli($n = 43$) were significantly lower than in patients infected with microorganism other than E.coli ($n = 11$)($p = 0.044$).There was no significant relation between antibiotic resistance and UCL($p > 0.05$).Children with accompanying renal disease($n = 8$) in the sterile pyuria subgroup had significantly higher UCL than those without renal disease($n = 26$)($p = 0.033$).
Conclusions: UCL in patients with suspected UTI is not useful in early and differential diagnosis of cystitis and pyelonephritis,and in prediction of antibiotic resistance.However,it may provide early recognition of UTI with microorganisms other than E.coli and may help to decide effective early empirical treatment of these patients.High UCL in children with sterile pyuria and accompanying kidney disease in our study suggest that urinary cathelicidin may be protective against urinary tract infections in this group.

P-038 CONTINUOUS RENAL REPLACEMENT THERAPY IN NEONATES WITH HYPERAMMONEMIA

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Introduction: Continuous renal replacement therapy (CRRT) is one of the useful modalities for the critical care of neonates with hyperammonemia caused by inborn errors of metabolism (IEM) because the medical therapy will not rapidly clear ammonia and hyperammonemia could cause the poor neurologic outcomes. The rapid reduction of ammonia levels through CRRT may decrease neonate mortality and morbidity. However, CRRT in neonates had many limitations such as vascular access, or hypotension. The CRRT effects upon ammonia levels are poor characterized, and the optimal CRRT prescription for neonatal hyperammonemia remains unknown. The aim of this study was to assess the clinical characteristics of neonates with hyperammonemia receiving CRRT and the CRRT effects upon ammonia levels and outcomes.
Material and methods: We retrospectively reviewed the medical records of neonates with hyperammonemia caused by IEM who were admitted to the neonatal intensive care units of Samsung Medical Center, Seoul, Republic of Korea between January 2008 and December 2016 where they underwent at least 24 h of CRRT.
Results: A total of 13 neonates (male to female ratio 2.3:1) were included in this study, of whom 3 (23%) were born prematurely. Birth weight ranged from 1.7 to 3.7 kg. The etiology of IEM included ornithine transcarbamylase deficiency (n = 4), citrullinemia (n = 4), carbamoyl phosphate synthetase deficiency (n = 2), propionic academia (n = 2), and long-chain L-3-Hydroxy acyl-CoA dehydrogenase deficiency (n = 1). The median age at the time of CRRT initiation and the median duration of CRRT was 5 and 4 days, respectively. The mean blood flow rate was 8 ml/kg/min, and the mean dialysis/replacement flow rate (effluent volume) was 3400 mL/h/1.73 m2, which was higher than flows used in neonates with acute kidney injury. The mean ammonia levels at the CRRT initiation was 1197 μmol/L and the time required for half reduction in ammonia levels ranged from 6 to 24 (median 12) hours. The mean ammonia levels after 48 h CRRT was 228 μmol/L. There is no correlation between effluent volume and the time required for half reduction in ammonia levels. Three patients (23%) showed the rebound elevation of ammonia levels after termination of CRRT, and CRRT was restarted during the hospitalization periods. The duration of hospitalization ranged from 4 to 86 (median 37) days, and 4 patients died during follow-up.
Conclusions: Clinically significant ammonia clearance can be achieved within 48 h in neonates with hyperammonemia utilizing CRRT. Our study suggested that a higher flow CRRT might not guarantee the rapid reduction in ammonia levels.

P-039 CARDIAC SURGERY ASSOCIATED ACUTE KIDNEY INJURY

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Introduction: The aim of this study was to estimate the frequency of cardiac surgery-associated acute kidney injury (CS-AKI) according to the predictive Risk-Adjusted Congenital Heart Surgery system (RACHS) in newborns and infants.
Material and methods: 29 (44.62%) neonates and 36 newborns (55.38%) were operated on. AKIN criteria (2007) used to set up the diagnosis of AKI by serum creatinine level. The severity of cardiac surgery was distributed by categories of RACHS system.
Results: 72.1% of neonates and 41.67% of infants developed CS-AKI. Statistically significant differences in the incidence of AKI in newborns operated on with or without cardiopulmonary bypass were not found, but in infants there were differences with regard to using bypass for surgical correction ($p < 0.001$). In the 3rd risk category AKI rate was increased in neonates (45.45%) comparing to infants (10%) ($p = 0.049$). Statistically significant differences in the development of AKI in neonates and infants in categories 4 and 6 RACHS were not found ($p > 0.05$). The high incidence of AKI in 4–6 categories was found both in neonates and infants (88.9% of neonates in 4th and 6th RACHS categories; 70% of infants in 4th category and 100% of 6th category). CS-AKI was established more often in neonates than in infants ($p < 0.05$).
Conclusions: The distribution of patients by RACHS categories allowed to predict the risk and severity of AKI in newborns and infants.

P-040 CORRELATION BETWEEN THE ESTIMATED GLOMERULAR FILTRATION RATE AND CREATININE CLEARANCE IN PAEDIATRIC ONCOLOGY PATIENTS

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Introduction: The use of parametric equations to estimate glomerular filtration (GF) in paediatrics is a trending question. However, there is not enough information referring to paediatric oncology. Aim: To study the potential correlation between the GF estimated by the modified Schwartz (SGFR) and Gao (GGFR) formulas, and measured by creatinine clearance (CrCl) obtained from a 24-h urine sample, all expressed in ml/min/1.73 m2 to see if they can be used interchangeably.
Material and methods: Retrospective study. Charts from 269 children less than 18 years old from the haemato-oncology unit were reviewed. The following data was collected: demographics, anthropometric data, serum creatinine and CrCl. Patients who lacked any of these were excluded. Statistical analysis was done using SPSS.
Results: Sixty-three patients (38 male, 25 female) were included. Mean age was 7.5 (range 0.1–18) years. The most common diagnosis was haematological malignancy (33), central nervous system tumours (15) and Wilms tumour (7). Medium serum creatinine, SGFR, GGFR and CrCl was 0.42 mg/dl (0.12–0.83), 133.36 (58.39–281.88), 107.88 (61.48–128.82) and 152.3 (24.52–475.34), respectively. Data follows a normal distribution (Kolmogorov-Smirnov test). A significant difference, by paired t-test, between SGFR and CrCl was found, with an average difference in their values of 18.93 (95% confidence interval of [4,8–33]) with a correlation coefficient (CC) of 0.6 ($p < 0.09$), with a significant reliability analysis. Similarly, we found a statistically significant difference between GGFR and CrCl with an average difference of 44.4 (95% confidence interval of [27.53–61]), with a CC of 0.3 ($p < 0.02$), with a non-significant reliability analysis.
Conclusions: In oncologic patients, there is significant correlation between CrCl and SGFR and GGFR but the CC is low. SGFR is more accurate than GGFR if CrCl is used as the gold standard. We cannot conclude that SGFR, GGFR and CrCl are equivalent considering that the difference between the mean values is too large and significant.

P-041 SEVERE ACUTE KIDNEY INJURY (AKI) AND RENAL REPLACEMENT THERAPY (RRT) IN CHILDREN WITH ACUTE MENINGOCOCCEMIA: FINDINGS FROM A LARGE COHORT

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Introduction: Compared to other septic conditions, acute meningococcemia is associated with fulminant disease course and high mortality rates. The scarce data about AKI in this population could be explained by increasing immunization and high mortality rates. Lithuania is an endemic zone for serogroup B meningococcal infections and immunization has not been implemented nationwide yet. We aimed to analyze the prevalence and early renal outcomes of children with acute meningococcemia and severe AKI requiring RRT.

Material and methods: A retrospective chart review for the period of January 2009–March 2017 was performed to identify children with laboratory confirmed meningococcal septicaemia treated in our institution.

Results: A total of 161 patients (49.7% boys) with acute meningococcemia were identified. Median age at diagnosis was 2.5 years (range 1 month-17 years). Sixteen patients (9.9%) with fulminant disease course died, typically within the first 24 h since admission. RRT was initiated in four (2.5%) patients (details in Table 1).

Age (y)	Sex	Complications	Time to oliguria (h)	Time to RRT (h)	Time on RRT (days)	RRT employed (duration in days)	Length of stay (days)	Renal function on discharge		
								GFR (ml/min/1.73 m²)	Urinalysis	Hypertension
1.7	M	Septic shock. Acute heart failure. DIC	30	40	10	CCPD (10)	24	>90	N	No
2	M	Septic shock. Acute heart failure. DIC	16	27	12	CCPD (3) -> CVVHDF (3) -> CCPD (6)	33	80	Protein+ Hematuria+	Yes
9	M	Septic shock	10	33	16	CCPD (7) -> CVVHDF (5) -> HD (4)	28	55	N	Yes
1.3	M	Septic shock. Multiorganic failure. DIC	14	46	16	CVVHDF (CARPEDIEM) (2) -> CCPD (14)	47	>90	N	Yes

Conclusions: The prevalence of severe AKI necessitating RRT in our cohort of children with acute meningococcemia was low (2.5%). RRT was typically initiated on day two after disease onset and continued for 10–16 days.

P-042 DISCONTINUOUS ECULIZUMAB THERAPY IN AN ATYPICAL HEMOLYTIC UREMIC SYNDROME WITH COMPLEMENT FACTOR H MUTATION

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Introduction: Management of atypical hemolytic uremic syndrome (aHUS) has been significantly modified by the use of eculizumab, a monoclonal anti-C5-antibody, which blocks the complement terminal alternative pathway activation. First recommandations were to administrate long-term treatment, as relapses of aHUS are frequent, especially in patients with complement factor H (CFH) mutations.

Material and methods: We report an efficient discontinuous eculizumab therapy for management of a seven year's old girl presenting aHUS with CFH mutation.

Results: The first signs of thrombotic microangiopathy (TMA) revealed the pathology at the age of 2 months. After a first line treatment with plasma exchanges, she was therefore treated with perfusions of eculizumab every three and then two weeks since the age of 15 months. We decided to stop eculizumab twice: in 2012 for the appearance of an uncomfortable cough and in 2015 because of the occurrence of unexplained bone lesions. After stopping treatment, she was seen in the clinics once a month with a close monitoring of hematological parameters, proteinuria, hematuria and renal function. She relapsed twice, respectively after 9 and 12 months of treatment interruption, because of an influenza vaccine and a viral infection. At each relapse, she presented with macroscopic hematuria, pallor and icterus. Eculizumab was then given immediately and permited TMA resolution, defined by a normalization of platelet count and improvement of renal function with a $\geq 25\%$ reduction of serum creatinine, within 4 days after eculizumab administration. At last follow-up, renal function was strictly normal and a slight albuminuria only remained, at the same level as before relapses.

Conclusions: We suggest that a discontinuous eculizumab therapy can be considered in every case of aHUS, with a close monitoring of clinical and biological parameters and good information for patients about signs that must alert them.

P-043 CHILDHOOD STEROID-DEPENDENT IDIOPATHIC NEPHROTIC SYNDROME: PREDICTIVE FACTORS FOR THE NEED OF IMMUNOSUPPRESSIVE TREATMENT

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Introduction: Nearly half of the children with idiopathic nephrotic syndrome (SN) become steroid dependent and will later require the use of steroid sparing agents. It appears important to identify as early as possible those children in order to adapt their treatment.
The aim of our study was to analyse the population of children, under 18, diagnosed between 2000 and 2015 with SN to search for predictive criteria of the use of steroid-sparing agents.
Material and methods: In this retrospective study, exclusion criteria were: primary steroid resistance and children free of relapse of proteinuria..
Results: 84 children (54 boys) were included. The mean follow-up duration was 5.5 years (0.75 to 16). Mean age at diagnosis was of 4.6 years old. 65 children (77%) received at least one steroid-sparing agent during their follow-up, with a mean delay of 10 months after diagnosis. In those patients, the first relapse occured earlier when compared with the children who were maintained on steroid alone (4 monts vs 7 months $p < 0.001$). The use of methylprednisolone pulses to obtain a remission, the cumulative dose of steroid treatment and the number of relapses of proteinuria were also signicantly correlated with the use of steroid sparing agents.
Conclusions: We did not find any predictive criteria of the use of steroid-sparing agents at diagnosis in our population of children. Nevertheless, with the steroid regimen used, the time of occurence of the first relapse of proteinuria appears to be a significant criteria for the secondary use of steroid-sparing agent. This data should be taken into account for the choose of the treatment regimen proposed.

P-044 ACUTE KIDNEY INJURY – RARE PRESENTATION OF BURKITT LYMPHOMA

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Introduction: Burkitt lymphoma is a highly aggressive B-cell non-Hodgkins lymphoma. Bilateral renal infiltration by lymphoma cells is an unusual cause of acute renal failure (ARF) and rare manifestation of lymphoma. ARF management is focused on treatment of the malignancy because modern chemotherapy leads to a dramatic normalization of renal function.
Material and methods: We report a 13-year-old male who presented with moderate facial edema and acute renal failure due to massive bilateral kidney infiltration of lymphoid tissue and normal urinalysis.
Results: Laboratory evaluation revealed anaemia, increased serum creatinine level 363 μmol/l, lactate dehydrogenase (LDH) 469 U/L, uric acid 1600 μmol/l, urea 37 mmol/l, impaired renal function GFR < 15 ml/min/1,73 m2. The tumour infiltration comprised 95% of renal parenchyma. Percutaneous renal biopsy was performed. Pathologic examination showed diffuse infiltration of the interstitium with lymphocytes and atypical cells, immunohistochemistry showed infiltrated cells positive for c-MYC translocated gene. The lymphoid cells were positive for CD19 phenotype. The final diagnosis of Burkitt lymphoma/leukemia was made. The patient was treated according to B-NHL 04 protocol including CNS-directed therapy. Six blocks of chemotherapy were released. During the first chemotherapy course the patient showed delayed methotrexate excretion requiring rescue with glucarpidase. Management of AKF involved adequate intravenous hydration, urinary alkalinisation, correction of electrolyte imbalances, forced diuresis. Renal replacement therapy was not necessary. Now the patient is in remission for 16 months. Follow up examination showed normal haematopoiesis and renal function.
Conclusions: Renal failure manifestation of Burkitt lymphoma/leukemia is diagnostically challenging. The presence of renal enlargement, an elevated LDH, uric acid, normal urine test should suggest the diagnosis, but only renal biopsy can provide a definitive diagnosis. Lymphomatous infiltration may respond well to therapy therefore it is highly recommended to use full-dose chemotherapy in Burkitt lymphoma cases presented with severe acute renal injury.

P-045 ECULIZUMAB THERAPY IN A CHILD WITH STREPTOCOCCUS PNEUMONIAE ASSOCIATED HUS, EVIDENCE OF NEURAMINIDASE ACTIVITY AND REDUCED MCP EXPRESSION

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Introduction: Streptococcus pneumoniae associated HUS (SpHUS) is accompanied with invasive pneumococcal diseases. In recent years there has been an increase in the incidence of invasive pneumococcal infections due to vaccination (Prevnar 7,13). We present a case of the girl with necrotising pneumococcal pneumonia, sepsis, HUS symptoms, evidence of neuraminidase activity, reduced MCP expression and significant complement activation.
Material and methods: 5-year-old girl was admitted with quick development of sepsis and MODS. X-ray showed a severe bilateral pneumonia, there were positive a pneumococcal antigen in urine and Str.pneumoniae in the blood culture. Development of septic shock, circulatory failure, acute renal failure, anuria, ARDS, DIC, hepatopathy. Mechanical ventilation and CVVHD were started. Laboratory tests and diagnostic procedures: CRP 200 mg/l, creatinine 172 umol/l, LD 50.4 ukat/l, haptoglobin 0.3 g/l, hemoglobin 79 g/l, leucocytes 0.9, blasts 0.016, schistocytes 26‰. Coombs with mild positivity. Bone marrow biopsy with no signs of malignancy. Significant complement activation (C3 0.10 g/l, C4 < 0.02 g/l). Laboratory evidence of reduced MCP expression and neuraminidase activity. ADAMTS 13: negative, anti-FH antibodies: negative. Treatment: plasmapheresis, bronchoscopy, ATB therapy. Eculizumab administration: 600 mg (day 4), 300 mg (day 11,25 and every other 2 weeks).
Results: Clinical status and pulmonary findings improved, diuresis increased. CVVHD was stopped (day 19). Extubation and weaning (day 21). Renal parameters in normal range (creatinine 20 umol/l), no proteinuria or hematuria, negative HUS activity (PLT 237, schistocytes 0, LDH 6,57). Favorable neurological status. Eculizumab is continued to the exclusion of mutation in complement-regulating genes.
Conclusions: The diagnosis of SpHUS can be difficult for concurrent development of DIC and sepsis. In case of complement activation is necessary to think about the overlap with complement-associated atypical HUS. Mortality rate is reported up to 12%, ESRD 10–16%, CKD gr. I-IV about 16%. Administration of eculizumab in early stages of SpHUS can improve overall prognosis.

P-046 MULTI ORGAN FAILURE WITH SEVERE RHABDOMYOLYIS AFTER CARDIAC TRANSPLANT SUCCESSFULLY TREATED WITH CRRT WITH AN69 HEPARIN ENGRAFTED FILTER OXIRIS

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Introduction: Severe low cardiac output after cardiac surgery may often cause multiorgan ischemic failure (MOF) with rhabdomyolysis, inflammatory response and acute kidney injury increasing dramatically mortality rate. Here we present a case of AKI due to MOF with severe rhabdomyolysis after low cardiac output occurring after heart transplantation in a 14 years-old patient completely recovered with a super high flux CVVHDF with oXiris® filter (Gambro, Sweden) (AN69 permanently heparin grafted designed for specific adsorption of endotoxin and cytokines).
Material and methods: A 14 years-old male born with Hypoplastic Right Heart Syndrome treated with Norwood, bidirectional Glenn, Fontan intervention, aortic, cavo-pulmonary connection, multiple caval angioplasty and stent placements during the first 10 years of life, developed progressive worsening of the right cardiac function until protein wasting entheropathy. Variously treated until high-protein parenteral nutrition.
In december 2016 he received heart transplantation requiring 100% ECMO placement on day 1. Since day 2 he developed progressive oligo-anuria unresponsive to diuretics (10 mg/kg furosemide and 0.5 mg/kg etacrynic acid) and severe hyperkalemia.
Results: CVVHDF was started while on ECMO through a left femoral vein double-lumen catheter (11 F, 15 cm) at Qb = 180 ml/min, Qd = 5000 ml/h and 1000 ml/h prediluition using Prismaflex machine equipped with ST150 filter.
On day 3 Creatine Kinase (CK) and free myoglobin (Myo) dramatically increased showing severe rhabdomyolysis (CK from 1030 UI/L day 1, 150,100 UI/L day 2, to 429,000 UI/L day 3; Myo from 5000 ng/ml to 18,000 ng/ml). The oXiris AN69 filter was maintained for 96 h. We observed a rapid decline of CK to 88,600 UI/L and Myoglobin to 13,725, 14,865, 8063 at 24, 48, 72 and 96 h respectively and a rapid improvement of hemodynamic stability.
The rapid clearance of rhabdomylolysis and inflammatory cascade products allowed a prompt improvement of the cardiac function and exit from ECMO after 5 days.
CVVHDF progressively tapered and stopped after 28 days when diuresis and serum creatinine normalized.
Conclusions: This positive experience with this AN69 heparin grafted filter designed to adsorb cytokines, endotoxin and possibly myoglobin shoud be taken into consideration for severe multiorgan failures with AKI with rhabdomyolysis or septic shock also in children.

P-047 BISMUTH INTOXICATION IN A PREGNANT ADOLESCENT

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Introduction: Bismuth (Bi) based compounds have been commonly used in the industrial, cosmetic and medical applications. To date only a few cases with nephrotoxicity after ingestion of colloidal bismuth have been reported.
Material and methods: We present here a case of an adolescent pregnant girl who developed acute renal failure due to an overdose of colloidal bismuth subcitrate (CBS).
Results: An 16-years-old adolescent, 5 weeks, 4 days pregnant girl was admitted to the emergency department 30 h after taking 20 tablets (300 mg/tablet) of CBS in a suicide attempt. Physical examination upon admission was unremarkable. The laboratory findings and renal imaging findings on admission revealed acute renal failure. She rapidly became oliguric and her serum creatinine level increased in a few hours. Her blood bismuth level was high (102.1 μg/l). She received parenteral chelating agent dimercaprol (14 days) and hemodiafiltration/ high flux hemodialysis daily.The patient recovered clinically and was discharged after 21 days in hospital.The patients gave a birth of a healthy term boy.
Conclusions: Acute toxicity of bismuth may result in nephrotoxicity. The administration of the chelating agent dimercaprol in combination with hemodiafiltration/hemodialysis is an effective treatment.

P-048 A RARE CAUSE OF POST-RENAL ACUTE KIDNEY INJURY BY NEUROBLASTOMA AND A ROLE OF PEDIATRIC NEPHROGIST IN PERCUTANEOUSLY PERFORMED BILATERAL NEPHROSTOMY

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Introduction: Acute kidney injury is mostly caused by pre-renal pathologies, therefore renal and post-renal causes are less common. In this case, we wanted to share our experience about the role of bilaterally performed percutaneous nephrostomy by pediatric nephrologists in management of post-renal AKI secondary to neuroblastoma.
Material and methods: Case report.
Results: 8-month year-old male baby was reffered to our hospital with complaints of watery defecation, vomiting for 3–4 days and refusing to feeding in last 24 h. In physical examination: body weight 8.2 kg(25-50p), height 71 cm (25-50p), body temperature 36.80C, respiratory rate 60/min, heart rate 140/min, blood pressure (87/55 mmHg) (95p:85 mmHg/95p: 53 mmHg), sO2%94, bilateral +2 edema in lower exremities and solid mass palpated in whole abdomen. In laboratuvary: urea 164 mg/dL, creatinine 10.3 mg/dL, potassium 5.54 mmol/L, sodium 132 mmol/L, calsium 10 mg/dL, phosphorus 6.6 mg/dL, uric acid 13.2 mg/dL, hemoglobine 8.8 g/dL, white blood cell 21.000/uL, platellets 492.000 /uL, INR 1.06 (N:0.94–1.28), aPTT 27.5 s, PT 13.5 s, blood pH 7.0, bicorbanoate 3.7 mmol/L, lactat 2.3, base excess −21. In renal ultrasonography, bilateral hydronephrosis and reduced parenchymal thickness was detected. Ureters were covered by soft tissue mass located between superior mesenteric artery and iliac bifurcation in abdominal CT. Double J catheters can not be advace in renal pelvis in cystoscophy. Ultrasound guided bilateral 8F percutaneous nephrostomy catheters were placed by pediatric nephrologist. Hemodiafiltration treatment was commenced on due to hypervolemia and hypertension. In following 48 h, there was no need for dialysis. Neuroblastoma diagnosis was made in histologic specimens. After completion of chemotherapy and surgical course, the patient has normal kidney funcitons and normal ultrasonographic examination.
Conclusions: Intraabdominal masses should be kept in mind in post-renal AKI etiologies. We advocate that ability to perform nephrostomy by pediatric nephrologist has a critical importance in kidney and patient survival.

P-049 RENAL AND SKELETAL TOXICITIES INDUCED BY HUMAN IMMUNODEFICIENCY VIRUS (HIV) THERAPY IN AN ADOLESCENT

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Introduction: We report a rare case of an adolescent who was undergoing antiretroviral (ART) therapy for HIV, presenting with severe hypokalemia and acute renal injury, acquired Fanconi Syndrome with osteomalacia resulting in a previous history of stress fracture as well as possibly acquired diabetes insipidus. Renal prognosis remains guarded on follow up.
Material and methods: Patient, a thin (BMI =16 m2/kg) 18 year-old Indian male was first diagnosed with HIV at 15 years old. He was started on ART with Tenofovir (TDF)/emtricitabine and lopinavir/ritonavir. Eight months later, he was found to have a right 3rd metatarsal fracture after he complained of persistent right foot pain. Reduced bone mineral density, hypophosphatemia, normal serum calcium and 25 hydroxyvitamin D were found. He was given Phosphate and calcium/vitamin D3. He also reported occasional vomiting, nocturia and further weight loss. Soon after, he was admitted with acute onset of vomiting and lethargy without fever.
Results: Investigations showed severe hypokalemia (1.9 mmol/L), severe hypophosphatemia (0.5 mmol/L), renal tubular acidosis, raised serum creatinine (129 μmol/L) but normal urea. Urine output was good and urine SG was low despite persistent vomiting. ECG showed U waves and investigations confirmed Fanconi Syndrome (severe phosphaturia with tubular reabsorption of only 22%, kaliuria, glycosuria and proteinuria). Hypokalemia was corrected emergently. TDF was withdrawn and switched to abacavir/lamivudine and raltegravir. Patient was discharged with oral potassium, phosphate, Shohl's solutions and alfacalcidol. He was successfully treated after 8 months of replacement therapy and gained good weight. A year on, his serum creatinine though within normal range for age, was 50% above his baseline value.
Conclusions: TDF- based ART therapy has reduced significantly HIV associated morbidity and mortality. However ART therapy, especially TDF may induce acute and chronic renal as well as associated skeletal toxicities that need to be pre-emptively monitored and promptly treated, before potentially life-threatening and possibly irreversible damages occur.

P-050 ATYPICAL HEMOLYTIC UREMIC SYNDROME ASSOCIATED WITH BORDETELLA PERTUSSIS INFECTION IN AN INFANT

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Introduction: Hemolytic uremic syndrome (HUS) has been associated with a number of infectious agents.
Material and methods: We report here the case an infant with severe Bordetella pertussis infection who developed atypical HUS.
Results: A 2-month-old preterm boy (34 weeks GA) was admitted for cough and fever. Laboratory investigations showed hyperleukocytosis (60 × 10.9 cells/L) with lymphocytosis, mildly elevated C reactive protein (32 mg/L) and unremarkable remaining blood parameters. The child was not yet immunized and PCR was positive for B. pertussis. He rapidly developed respiratory distress syndrome requiring transient non-invasive ventilation and a macrolide antibiotic. At day 3 after admission, he presented hemolytic anemia with schistocytes, high blood pressure, marked proteinuria and progressive rise of serum creatinine which made us start a diagnostic workup of HUS. Stool culture detected no *E. coli*, and PCR specific for the Shiga toxin genes (stx1, stx2) and the eae gene were negative. Routine complement activation showed no evidence of complement alternative pathway activation (normal C3, C4 and complement factor B levels, transient decreased expression of MCP on peripheral leucocytes, no quantitative deficiency of complement factor H and complement factor I, and no anti-factor H autoantibody). ADAMTS13 activity was within normal range. Severe renal failure led to the initiation of peritoneal dialysis on day 12 after admission. He received eculizumab therapy at days 19, 27, 34 and 48. His renal function gradually improved and dialysis was discontinued at day 37. After 2 months hospitalization the infant was discharged home. Laboratory tests showed normalization of renal function and hematologic values. High blood pressure persisted. Finally, a rare heterozygous complement factor H variant associated with aHUS was found.
Conclusions: Five cases of HUS associated with B. pertussis infection have been previously described (including one with CFH mutation and one with MCP mutation). Our case suggests that pertussis is a trigger of atypical HUS.

P-051 NUTCRACKER SYNDROME (NCS) AND ITS VARIANTS: THREE CASES REPORT

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Introduction: NCS refers to compression of the left renal vein between the aorta and the superior mesenteric artery (anterior NCS) or between the aorta and the vertebral body (posterior NTS), with impaired blood outflow often accompanied by distention of the distal portion of the vein. The most common symptoms are gross hematuria, left abdominal pain, varicoceles, proteinuria. It could be asintomatic too.
Material and methods: Case 1: 4 years old girl, presented with several recurrent episodes of gross hematuria associated with left abdominal pain; renal function, urinary metabolites and urinary tract US were normal. The abdomen magnetic resonance (acronimo in inglese) showed an entrapment of a retro-aortic left renal vein between the aorta and the vertebral column.
Case 2: 11 years old boy, with a history of 4 episodes of gross hematuria associated with lumbar pain; laboratory tests and urinary US were normal. The CT-angiography showed a dilation of initial tract of left renal vein, with a reduced caliber in the portion interposed between abdominal aorta and superior mesenteric artery.
Case 3: 14 years old boy, with mild proteinuria and gross hematuria. Kidney function tests were normal. Abdominal US and CT-angiography showed a dilated left renal vein, with a reduced aorto-mesenteric caliber angle.
Results: Because of the variability of symptoms and absence of consensus on diagnostic criteria, the exact prevalence of NCS is unknown.
Conclusions: NCS represents a cause of non-glomerular gross hematuria always be taken into account, after excluding the most frequent causes, especially if associated with abdominal left side pain, but that may present themselves, as in our case, with asymptomatic proteinuria. In these patients it's essential renal vascular district study to confirm or rule out the suspected diagnosis.

P-052 DEHYDRATION AND ACUTE RENAL FAILURE IN CHILDREN: IS IT A FATALITY?

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Introduction: Dehydration in pediatric population is a frequent symptom especially in developing country.Many complications are observed in moderate to dehydration in children.

Material and methods: This study aimed to describe clinical characteristics, biochemistry features and evolution in hospitalized children with acute renal failure (ARF) complicated dehydration in a pediatric department.
Results: Between 2012 and 2016, 113 patients were admitted in our pediatric department with dehydration, average age was 18 years. Fifteen per cent Presented moderate to severe dehydration. The average age was 12 months. Laboratory tests made in all cases. Only 35 patients had kidney failure at the admission (31%). Intravenous perfusion was the first procedure in 85%. At discharge, only three patients died with ad integrum restitution of creatinine in the others cases (94%).
Conclusions: Children who have suffered ARF from any cause are at risk for late development of kidney disease several years after the initial insult. Continued surveillance will be important to consider many years after the ARF.

P-053 NEPHROTIC SYNDRFOME AND ACUTE KIDNEY INJURY IN A GIRL TREATED WITH LITHIUM CARBONATE

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Introduction: Lithium carbonate has been used for decades in psychiatry for the treatment of depression and bipolar disorder. However, this medicine has several renal side effects. These include nephrogenic diabetes insipidus, chronic tubulointerstitial nephritis, distal renal tubular acidosis and rarely nephrotic syndrome. In addition, lithium intoxication can also lead to acute kidney injury (AKI). Majority of patients recover after lithium treatment discontinuation.
Material and methods: An 18-year-old white Caucasian female with anxiety disorder and depression was referred to our hospital because of nephrotic syndrome and anuric AKI. Current medication included lithium carbonate, amitriptylin, quetiapine and zolpidem. She had been on lithium treatment for 2 years. Her lithium serum levels and renal functions had been repeatedly within the normal range. On initial examination, her blood pressure was 132/75 mmHg, she had significant bilateral periorbital edema and swelling of the hands and legs. The following laboratory values were noted: hemoglobin 12.2 g/dl, leukocytes 17.3×10^9/l, thrombocytes 267×10^9/l, total protein 40 g/l, serum albumin 22.1 g/l, urea 24.2 mmol/l, creatinine 245 μmol/l, sodium 120 mmol/l, potassium 6.7 mmol/l, bicarbonate 23 mmol/l, total cholesterol 9.5 mmol/l, serum lithium 1.88 mmol/l (normal range 0.3–1.3 mmol/l). Urinalysis showed total protein to creatinine spot urine ratio 225.5 mg/mmol without hematuria. Ultrasonographic examination of kidneys showed only mild diffuse parenchymal lesion.
Results: Percutaneous kidney biopsy was performed and revealed minimal change disease. Lithium therapy was discontinued and symptomatic management with fluid restriction, electrolyte and albumin infusions along with diuretics was initiated. Finally, our patient went into full remission in 31 days after lithium treatment discontinuation. At this time lithium was first not detected in serum of our patient.
Conclusions: Lithium administration was a cause of AKI and nephrotic syndrome in our patient. She fully recovered without corticosteroids. Measurement of serum lithium levels was useful in anticipating the time of full remission.

P-054 SEVERE ACUTE POSTINFECTIOUS GLOMERULONEPHRITIS: IS THERE A PLACE FOR ECULIZUMAB?

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Introduction: Postinfectious glomerulonephritis (PIGN) represents the most frequent cause of acute glomerulonephritis in childhood. The outcome is good in the majority of cases but, occasionally, PIGN could lead to ESRD. Activation of classical and alternative complement pathway in PIGN has been clearly proved. Eculizumab could be considered as a possible treatment in severe cases of PIGN.
Material and methods: A 12 years old girl was admitted in ICU for seizures. Clinical examination found a severe hypertension, moderate edema and macroscopic hematuria. Laboratory exams revealed an acute renal failure (SCr 227 μmol/l, urea 23 mmol/l) with a nephrotic range proteinuria (985 mg/mmol); isolated low level of C3 (0.12 g/l). Cerebral MRI revealed lesions corresponding to a posterior reversible encephalopathy syndrome. Diagnosis of PIGN with neurological impairment was made. Supportive care were started and initial evolution was good with an improvement of glomerular filtration rate (GFR).
Results: At day 7, the patient developed an oliguria and SCr level raised up to 400 μmol/l. Kidney biopsy showed 10/18 sclerosed glomeruli. A moderate mesangial hypercellularity was present in the 8 others without any crescent formation. A moderate interstitial inflammation was also seen. Immunofluorescence study revealed a strong C3 and C5b-9 staining in the mesangium and in capillary walls without IgG deposits. sC5b9 was increased (571 ng/ml); C3 Nephritic factor was negative. Treatment with steroids pulse was started without any glomerular filtration rate improvement. According to these results the diagnosis of a rapidly progressive C3 glomerulopathy (C3G) was suggested and, due to the severity of renal failure, eculizumab (ECZ) was quickly started. Renal function dramatically improved after the first infusion. At one month, SCr decreased to 100 μmol/l. Sc5b9 and C3 level returned to normal range. Genetic complement abnormalities implicated in C3G were screened but no mutation was found. At 6 months, another renal biopsy was made revealing a moderate mesangial hypercellularity without C3 deposits in favour of a late PIGN.
Conclusions: Atypical PIGN can be mistaked with a C3G. It has been demonstated that ECZ can be efficient in C3G. But ECZ should be considered as therapy in a severe form PIGN with a pathologic overactivation of complement alternative pathway.

P-055 TUBULOINTERSTITIAL NEPHRITIS AND UVEITIS (TINU) SYNDROME IN A 6 YEAR-OLD MALE PATIENT

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Introduction: Tubulointerstitial nephritis and uveitis (TINU) syndrome is a rare condition which has a combination of ocular inflammation and renal interstitial and tubular inflammation. Less than 300 cases have been reported since 1975, first description of the syndrome. Etiology of this syndrome is poorly understood, though immunologic processes are thought to play a role.
Material and methods: Case presentation.
Results: Six year 2 month-old boy presented with red eyes and blurry vision. There was a history of fever, poor apetite, fatigue one, amoxicillin and nonsteroidal anti-inflammatory drug use a month ago. Patient was referred to our clinic with suspicion of acute kidney injury. Blood urea nitrogen (BUN) and serum creatinine (Cr) levels were increased (23.3 and 2.4 mg/dL, respectively).Tru-cut renal biopsy was performed, and IV pulse methylprednisolone treatment was administered. Lymphocyte predominant leukocytic infiltration, tubulitis, hydrophic degeneration and necrosis in tubule epithelium was reported in the biopsy specimen,

confirming the definitive diagnosis of TINU. Oral methylprednisolone treatment has been continued for a month and gradual decline in both BUN and Cr was documented.
Conclusions: Any patient presenting with uveitis should be considered more carefully not to miss underlying TINU syndrome. Renal biochemical parameters including urinary beta-2 microglobulin could be useful to assess any renal abnormality in patients with uveitis which can be a hint for TINU syndrome.

P-056 DOES THE ULTRASOUND OF THE URINARY TRACT HAVE ITS PLACE IN THE TREATMENT OF EARLY NEONATAL JAUNDICE? NEONATAL BILATERAL ADRENAL HEMORRHAGE: CASE REPORT

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Introduction: Adrenal hemorrhage is a rare clinical entity in the neonatal period, with an incidence of 1.7–2.1 / 1000 births. It is more often diagnosed on the right side whilst bilateral hemorrhage occurs in 10–15% of cases.
Material and methods: Clinical presentation shows a wide range of symptoms, from the signs of adrenal insufficiency to asymptomatic course of illness with incidental finding of changes on tests. Neonatal jaundice due to hemolysis of hemorrhagic content often is an accompanying sign. We present a male neonate born at term, with the early neonatal jaundice of unknown cause and without evidence of perinatal infection. Ultrasound of the urinary tract was made and it was seen a hypoechogenic formation in the upper poles of both kidneys, confirmed with magnetic resonance imaging (MRI) of the abdomen.
Results: Clinical and laboratory test results showed no signs of adrenal insufficiency. There was no confirmation of embryonic tumor or neuroblastoma.
Conclusions: Ultrasound of the urinary tract, as an available and non-invasive test, has its place in the treatment of early neonatal jaundice of unknown cause. With ultrasound monitoring of regression of formation a further invasive treatment and unnecessary laparotomy can be avoided.

P-057 TUBULOINTERSTITIAL NEPHRITIS WITH UVEITIS IN ELEVEN-YEAR-OLD PATIENT

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Introduction: Tubulointerstitial nephritis with uveitis syndrome (TINU) is a rare disorder. The etiology, therapy and outcome of patients with TINU remain unclear.
Material and methods: We review the clinical case of patient with TINU.
Results: An 11-year-old caucasian girl was admitted in hospital with a history of fever, extreme fatigue, weight loss (4 kg/month).At admission she had signs of renal insufficiency (serum creatinine 120 μmol/l, urea 11,6 mmol/l, eGFR 59 ml/min), anemia (hemoglobine 105 g/l), elevated erythrocyte sedimentation rate (65 mm/h) and C-reactive protein (2,5 mg/dl), urinalysis showed low urine density (1003–1005), normoglycemic glucosuria (11 mmol/l), proteinuria (0,27 g/d). ANF, anti-ds-DNA, LE cells, p-ANCA, c-ANCA, anti-cardiolipine-antibodies, PCR for EBV, HSV, CMV in serum, Mantoux test were negative. C3 and C4 serum levels, renal ultrasound, chest X-ray were normal. Diagnosis of acute interstitial nephritis was probable. One month after acute renal injuary the girl presented with recurrent bilateral non granulomatous anterior uveitis, she was treated with topical steroids. A renal biopsy was performed from 6 months after disease onset because of persistent elevated serum creatinine (98–106 μmol/l) and β2-microglobulinuria (2.2 mg/l, $N < 0.3$ mg/l). The biopsy showed significant dense lymphocytes, plasmocytes interstitial infiltration, tubulitis, secondary ischemic glomerular damages, immunofluorescence was negative. The clinical and morphological signs were consistent with TINU. Therapy with prednisone (1 mg/kg/day for 3 months, gradual withdrawal over 3 months) led to remission of uveitis and normalization of serum creatinine (78 μmol/l), urinary β2-microglobuline (0.25 mg/l). To date the duration of remission of TINU is 12 months.
Conclusions: TINU syndrome should be considered in case of "idiopathic" acute interstitial nephritis and in case of bilateral anterior uveitis. The renal biopsy results of our patient from 6 months of disease onset could confirmed that TINU's patients tend to have prolonged clinical course and may need long-term anti-inflammatory treatment. The urinary β2-microglobuline level may reflect the efficacy of therapy of TINU.

P-058 A CASE OF RAPIDLY PROGRESSIVE GLOMERU LONEPHRITIS SUPERIMPOSED ON X-LINKED ALPORT SYNDROME

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Introduction: The outcome if AKI is strictly dependent of preexisting chronic kidney disease and comorbidities.
Material and methods: We report case of rapidly progressive glomerulonephritis in 14 years old boy with X-linked Alport syndrome (AS).
Results: The boy had a history of familial X-linked AS (his elder brother also has morphology of AS with CKD stage 2 and sensoneural deafness stage 3 on third decade), at 1 year he developed hematuria, at 5 years – proteinuria 1.4 g/l. He received nephroprotective and antiproteinuric therapy with fosinopril, losartan and amlodipine, but despite that proteinuria dramatically increased. Since 9 years he had nephrotic range proteinuria. At 14 years next day after vaccination against diphtheria, tetanus and polio he developed nausea, gross hematuria and hypertension and. He was observed and 72 h he became oliguric. Laboratory findings were increasing of serum creatinine to 1087 μmol/l and urea to 31 mmol/l, serum complement was normal, autoantibodies were negative. He was oliguric and needed dialysis (CVVHD) during 16 days. We performed oral steroid therapy 1 mg/kg, daily steroid pulse-therapy 1000 mg and plasmapheresis with almost complete recovery of kidney function, his serum creatinine was 98 μmol/l, eGFR (CKD-EPI) 99 ml/min/1,73m^2. Kidney biopsy showed signs of immunocomplex glomerulonephritis (ICGN) with fibrous and cellular crescents and mesangial granular C3 deposition on immunofluorescence and signs of AS with tubular atrophy and interstitial fibrosis. However within 4 months we observed decline in GFR to 41 ml/min/1,73m^2.
Conclusions: We suggest that preexisting AS can predispose to immunocomplex glomerulonebphritis because of glomerular basement membrane abnormalities. From another hand, despite acute ICGN superimposed on AS may significantly accelerate CKD progression despite efficient treatment with pulses and plasma exchanges.

P-059 ECULIZUMAB TREATMENT IN A SEVERE HEMOLYTIC UREMIC SYNDROME: PEDIATRIC PATIENT

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Introduction: Hemolytic uremic syndrome (HUS) is characterized by anemia, thrombocytopenia and acute kidney injury. Although,

the most of the patients managed by conservatively, small portion of the patients needs further novel treatments. In this case, we want to share our experience about eculizumab treatment in a severe HUS patient.
Material and methods: Case report.
Results: 2 year-old girl was referred to our hospital due to bloody diarrhea, vomiting and pallor for 4 days. In addition, severe anemia and invagination detected in abdominal ultrasound in local hospital. Patient had been received ceftriaxone treatment for 4 days. Reduced urine output and generalized edema developed just 1 day before second admission. In physical examination: body weight 10.6 kg (10-25p), height 83 cm (10-25p), heart rate 104/min, respiratory rate 26/min, blood pressure 88/54 mmHg (95p: 89 mmHg/95.p 57 mmHg), pallor and anasarca edema. In laboratory: hemoglobine 5.9 g/dl, white blood cells 18.000 U/l, platellets 61.000 U/l, creatinine 2.1 mg/dl, urea 133 mg/dl, LDH 3106 mg/dl, uric acid 10.6 mg/dl, normal coagulation parameters. Schistocysts were detected in peripheral blood smear. Reticulocyte count was 8%. Shiga toxin was negative in stool and, ADAMTS 13 level was 50%. Abdominal exploration showed no invagination. Renal replacement treatment was initiated due to decreased urine output and hypertension. Plasmapheresis was started due to severe thrombotic microangiopathy. After 2nd plasmapheresis session, bloody diarrhea recurred and rectal prolapsus developed. Emergently re-abdominal exploration revealed invagination. Eculizumab treatment (300 mg) was initiated due to severe clinical course. Reduction in serum urea, creatinine and regression of hemolysis was achieved in following 48 h after eculizumab treatment. The patient is followed-up without any medication with normal kidney function in out-patient settings.
Conclusions: Generally, HUS has a good prognosis in children. We advocate that promptly initiation of eculizumab treatment has utmost importance in reversing kidney damage in that small portion of HUS patients.

P-060 MULTIPLE HIT HYPOTHESIS: A CASE WITH HERBAL MEDICINE INDUCED ATYPICAL HUS

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Introduction: There are many factors that were reported associations with the Atypical hemolytic uremic syndrome (aHUS) like non-Shiga toxin infectious agents, drugs, cancers, vaccination, transplant, autoimmune disease etc. These data for whether these agents are the primary cause of disease or triggers of an alternative complement system is limited. According to the "multiple-hit" hypothesis, aHUS is a consequence o both genetic predisposition to alternative complement dysregulation as well as the occurrence of events or conditions that may precipitate TMA by activating complement and/or damaging endothelium.
Material and methods: We present a patient who was diagnosed with atypical hemolytic uremic syndrome possibly triggered by over the counter medication, herbs.
Results: 11 years old boy, whose brother died because of renal insufficiency possibly secondary to TMA, admitted to hospital with flank pain. A renal biopsy was done because of the presence of proteinuria and high blood pressure. The complement levels and hematologic parameter were normal but haptoglobulin was low. Biopsy was remarkable for hypertensive nephrosclerosis. He had been following up at the outpatient clinic with ACE inhibitor with normal renal function test and abnormal proteinuria for 5 years until he admitted to hospital with acute renal failure. It was learned that he had been using many herbal drugs for 3 months. On admission, his blood pressure was high and kidney was not working well. In the follow up, his platelet and hemoglobin levels were decreased and LDH levels were increased. With history, he was diagnosed as an aHUS which was trigger by herbals. The patient who is currently on eculizumab needs hemodialysis three times a week.
Conclusions: It is known that atypical hemolytic uremic syndrome results from a defect on the alternative complement system. We think that herbal drugs acting like antigen may trigger alternative pathway of complement especially in patients who have defects in their complement pathway.

P-061 STEROID STARTED FOUR TUBULOINTERSTITIAL NEPHRITIS CASES

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Introduction: Tubulointerstitial nephritis (TIN) is associated with an immune-mediated infiltration of the kidney interstitium by inflammatory cells. Patients often present with nonspecific symptoms, which can lead to delayed diagnosis and treatment of the disease. The causes of acute TIN can be grouped into four broad categories: medications, infections, immunologic diseases or idiopathic processes.
Material and methods: We report four patients with increased creatinine and diagnosed with TIN after biopsy.
Results: CASE 1. A 16-year-old boy presented with a 1-month history of bilateral redness in the eyes and fever. He was diagnosed with uveitis. Laboratory tests showed elevated creatinine levels; 1.7 mg/dL. Renal biopsy findings showed granulomatous interstitial inflammation and tubular atrophy.
CASE 2. A 14-year-girl presented with anorexia. Laboratory tests showed elevated creatinine levels; 1.85 mg/dL. Her eye examination was normal. Renal biopsy findings showed interstitial inflammation and tubular atrophy.
CASE 3. A 15-year-boy presented with a 2-month history of bilateral redness in the eyes. He was diagnosed with uveitis. Laboratory tests showed elevated creatinine levels; 1.3 mg/dL. Renal biopsy findings showed mild chronic parenchymal injury consisted with chronic TIN (interstitial fibrosis and tubular atrophy).
CASE 4. A 17-year-boy presented with bilateral side pain. In his medical history, he had upper respiratory tract infection 1 week ago. Laboratory tests showed elevated creatinine levels; 4 mg/dL. Renal biopsy findings showed interstitial inflammation and tubular atrophy.
Conclusions: TIN should be considered in all patients with or without uveitis, hence steroids should be started to prevent chronic deterioration.

P-062 ACUTE KIDNEY INJURY

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Introduction: The goal: To direct attention on possibility of kidney disease in children who are active in playing sport.In the period of one month, March–April 2016., we had three young men in the age of 17th because of the acute renal insuficiency. All three boys were healthy and were active in sports. One of them was active in athletic, running, and the second and the third boy were regularly in gym. They were taken pills to get a better results.
Material and methods: Methods: We were trying with anamnestic, clinical and laboratory methods to find causes of the acute renal

insuficiency. From the anamneses we got the informations about active playing sports and regularly training in gym. All three of them took pills for achive better results of training. In laboratory results we were interested to see levels of urea and creatinin. We did ultrasound of kidney to all three of them and in two patients biopsy of kidney.
Results: Results: Urea and creatinin were higher in all three patients (urea 7,5–14-21-, creatinin 150–190-330,487–555-625). Ultrasound of kidney in two patients showed kidneys normal position, shapes and sizes, hyperechoes parenchims. Erased borders. The third patient had completely normal ultrasound of kidney. Biopsy of kidney in our two patients proved it was acute tubular necrosis.
Conclusions: Conclusion: All three patients were in our department because they had acute renal insuficiency.. One patient had high level of urea and creatinin so he was on haemodialysis during three days. Two patients also had therapy with bolus of corticosteroids. The third patient only had antibiotic therapy. All three young men had fully recovery of kidney functions. How we in spite of completely anamneses and treatments could not clearly find the cause of acute tubular necrosis, we should consider, like the risk factors, regularly training and pills which they were taken.

P-063 DOES RESOLUTION OF HYDRONEPHROSIS BY ULTRASONOGRAPHY IN DILATING VESICOURETERAL REFLUX ELIMINATES THE RISKS ASSOCIATED WITH REFLUX?

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Introduction: We aimed to evaluate the clinical and radiological course of children with persistent or resolved hydronephrosis associated with vesicoureteral reflux (VUR).
Material and methods: Hospital files of children with primary VUR and collecting system dilatation were retrospectively evaluated. Those with a follow up period >12 months were enrolled. Children were grouped as those with resolution of collecting system dilatation by ultrasonography (Group 1) and with persistent dilatation (Group 2). Both groups were compared for age, gender, follow up period, VUR grade, presence of renal scar, new development of renal scar and urinary tract infection (UTI) recurrence.
Results: 51 patients were enrolled in the study. Group 1 (n = 32) and Group 2 (n = 19) were not different with respect to age (45.2 ± 49.5 vs 39.4 ± 44.3 months), gender (F/M = 16/16 vs 9/10), ≥grade 3 VUR (11/32 vs 10/19), presence of renal scar (20/32 vs 12/19), folow up peiod (51.2 ± 30.9 vs 50.6 ± 31.6 months), breakthrough UTI rate (10/32 vs 5/19) and new scar formation (8/32 vs 4/19).
Conclusions: Resolution of collecting system dilatation by ultrasonography does not eliminate the risk of new UTI and scar formation in children with VUR.

P-064 NEPHROLITHIASIS IN A PORTUGUESE PAEDIATRIC POPULATION

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Introduction: To determine the epidemiology and outcome of a paediatric population presented to a nephrology unit in an urban tertiary centre.
Material and methods: The records of consecutive children (0–18 years) with nephrolithiasis were reviewed (January 2008–December 2016). Clinical features, aetiology, recurrence risk, treatment and outcome were retrospectively evaluated.
Results: We identified 104 cases: nephrolithiasis (n = 69), nephrocalcinosis (n = 24), and both disorders (n = 11). New nephrolithiasis cases (n = 80) increased through the study period from 2.3% to 11.8%. Age at presentation (median 8.6 years) was below 2 years in 21% and 46% were older than 10 years; mean follow-up was 29 months (2–108). Boys predominate (59%). The most common presenting symptom was flank or abdominal pain (44%). The upper urinary tract was most commonly affected (89%). A metabolic abnormality was identified in 51% of cases: hypocitraturia (57%), hypercalcuria (43%), hyperoxaluria (19%), hyperuricosuria (12%), and cystinuria (2%) without age predominance (p = 0,2). Urinary tract infection (UTI) (24%) was the next most significant aetiology. Children below 2 years of age were more likely to have UTI as a cause than the other age groups (p < 0,01). Cases with UTI were more likely to need surgical treatment (p < 0,01). Sixty-three percent of patients were stone free and 24% had recurrence. DMSA scanning was abnormal in 9/20 cases (45%). In a logistic-regression analysis adjusted for age, sex, weight, dietary errors, type of calculi and recurrence of symptoms, a known family history of nephrolithiasis was associated with an increased risk of a metabolic cause for the calculi (odds ratio = 6,86; 95% confidence interval [CI], 1663 to 28,302; p < 0.01).
Conclusions: Nephrolithiasis increased throughout the study period. UTI predominate at younger ages. Metabolic abnormalities usually have an arousing effect in nephrolithiasis and recurrence rates, validating the need for a complete screening in children.

P-065 ARE WHITE CELLS IN CHILDREN'S URINE DIAGNOSTIC OR A DISTRACTION, AND DOES COLLECTION AND CULTURE METHOD MATTER? NEW DATA FROM 4910 ACUTELY UNWELL CHILDREN

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Introduction: To describe the relationship between urine white cell count (WCC) and the microbiological diagnosis of UTI in acutely unwell children, and explore the influence of different methods of urine collection and culture.
Material and methods: Prospective, diagnostic cohort study of acutely unwell children aged 3 months-5 years presenting to UK primary care whose urine samples were collected by clean catch or nappy pad, and cultured by both a Central Laboratory (CL) using spiral-plating, and Local NHS Laboratories (LL) using standard practice. Positive and negative predictive values (PPV and NPV) were calculated for WCC ≥ 100/mm^3 and WCC ≤ 10/mm^3 respectively, comparing laboratories and sampling methods.
Results: Of 4910 samples, 2630 (53.6%) were collected by clean catch and 2198 (44.8%) by nappy pad. 1.9% had UTI according to the CL and 5.3% according to the LL. One

hundred ninety-four samples (4.0%) were positive in the LL but negative in the CL. CL samples were cultured on average 34.1 h later.

The table outlines PPV of WCC ≥ 100/mm^3 and NPV of WCC ≤ 10/mm^3 by laboratory culture and sampling method. Thirty seven of the 92 positive CL cultures (40.2%) had WCC ≤ 10/mm^3.

	CL culture	**LL culture**				
All Samples	Clean Catch	Nappy Pad	All Samples	Clean Catch	Nappy Pad	
PPV of WCC ≥ 100/mm^3	**23.9%** [17.5–31.6%]	**33.3%** [25.0–42.8%]	**0**	**30.3%** [22.7–39.1%]	**38.7%** [29.3–49.0%]	**6.9%** [1.8–23.6%]
NPV of WCC ≤ 10/mm^3	**99.0%** [98.7–99.2%]	**99.3%** [98.9–99.5%]	**98.6%** [98.3–98.9%]	**95.8%** [95.4–96.2%]	**97.7%** [97.2–98.2%]	**93.6%** [92.9–94.2%]

Conclusions: A WCC ≥ 100/mm^3 poorly predicts UTI on culture in acutely unwell young children consulting in the community. However, a WCC ≤ 10/mm^3 has good NPV regardless of collection method (though worse for LL culture, especially nappy pad samples). To discard urines with WCC ≤ 10/mm^3 before culture would miss 40.2% of CL diagnosed UTIs. Alternatively, LL culture of samples with WCC ≤ 10/mm^3 overcalls UTI by identifying 150 (3.1%) positive, which if later re-cultured by the CL's reference standard technique would reduce to 22 (0.4%), potentially minimising unnecessary treatment or investigation.

P-066 RELATION OF RENAL UPTAKE OF 99MTC-DMSA TO PATIENT AGE

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Introduction: 99mTc-DMSA is taken up by renal proximal tubular cells and is used for renal cortical imaging. Need for proximal tubular maturation and timing of cortical scintigraphy for maximum 99mTc-DMSA uptake is not certain. We aimed to find if any relation exists between patient age and renal 99mTc-DMSA uptake.
Material and methods: All 99mTc-DMSA scintigraphy results were retrospectively searched by using hospital imaging database. Patients over 2 years of age at the time of scintigraphy were excluded. Patients with normal scans were identified. Timing of radiopharmaceutical injection and imaging were specified in these patients. Patients with imaging time between 2 to 4 h of injection (n = 438) were enrolled in the study. Patient age during scintigraphy was recorded as weeks or months. Age of premature patients were corrected according to their gestational age. Maximum uptakes of both kidneys on anterior and posterior views were used for calculation of geometric mean by eliminating the negative effect of the differences of the depths of kidneys. Renal uptake was calculated by dividing the maximum uptake values and geometric mean to maximum neighbouring background and maximum liver uptake.
Results: No significant correlation was found between renal 99mTc-DMSA uptake and patient age (r:0.001, correlation coefficient − 0.19).
Conclusions: In the present study, no age limit regarding maximum 99mTc-DMSA uptake was determined. The test could be used in ant age period if it is indicated.

P-067 ASSOCIATION OF VOIDING DYSFUNCTION AND ASTHMA IN CHILDREN

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Introduction: Voiding dysfunction is a common problem in pediatric practice and is defined for children with voiding symptoms or urinary incontinence. Asthma is a chronic recurrent inflammatory disease in which dyspnea and chronic cough are defined to be associated with increase in voiding dysfunction in childhood.
Material and methods: Parents of 252 children between 5 and 15 years age group followed by the Pediatrics outpatient clinic in our hospital were asked to complete questionnares of International Study of Asthma and Allergies in Childhood (ISAAC) for asthma and dysfunctional voiding and Incontinence Scoring System (DVISS). Children with neurological or urological diseases were excluded.
Results: Voiding dysfunction was found in 14.7% of children and according to age groups 81.1% was in 5–10 year age group while 18.9% was in 11–15 years age group (p = 0.001). There was no difference between girls and boys. Voiding dysfunction had a positive correlation with asthma symptoms (p = 0.00). Children with dysfunctional voiding had higher history of wheezing at some point in life (p = 0.009), higher rates of current wheezing (p = 0.002), higher doctor diagnosed asthma (p = 0.02) and higher exercise related wheezing in the last 12 months (p = 0.01).
Conclusions: Asthma and symptoms related with asthma are associated with higher dysfunctional voiding scores in children. It is important to evaluate enuretic children for asthma symptoms in order to improve outcome of both entities and avoid long term morbidities.

P-068 THE ROLE OF FAMILY HISTORY, ASSOCIATED COMORBIDITIES AND BODY MASS INDEX IN PREDICTING RESPONSE TO ENURESIS TREATMENT

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Introduction: The purpose of this study was to evaluate the role of family history of enuresis, associated comorbidities (mental disability, history of head trauma, allergies etc.) and body mass index (BMI) in predicting response to enuresis treatment.

Material and methods: 100 patients with monosymptomatic enuresis were evaluated in this retrospective study. The association between family history of enuresis, associated comorbidities, body mass index and response to enuresis treatment with desmopressin melt oral formulation were analysed with χ^2 statistical test, using computer programme Microsoft Excel statistical tests.
Results: Family history of enuresis was positive in 20 (58.8%) among 34 patients who responded to desmopressin treatment and in 18 (54.5%) patients among 33 non-responders. The difference was not statistically significant.
Among 42 patients who responded to desmopressin treatment, obesity was present in 6 (14.3%), overweight in 8 (19%) and normal weight in 28 (66.7%) patients. Among 38 non-responders, obesity was present in 5 (13.2%), overweight in 3 (7.9%) and normal weight in 30 (78.9%) patients. The difference was not statistically significant.
Among 42 patients who responded to desmopressin treatment, comorbidity was present in 17 patients (40.5%). Among 38 non-responders, comorbidity was present in only 8 patients (21.1%) but the difference did not reach statistical significance.
The group of patients treated with alarm consisted of 23 children which is not enough for a reliable statistical analysis.
All patients had normal urinalysis (5 had mild isolated microhematuria) and normal ultrasound of kidneys and urinary tract (not done in 4).
Conclusions: The study did not prove statistically significant association between family history of enuresis, associated comorbidities or body mass index and response to enuresis treatment with desmopressin.

P-069 HORSESHOE KIDNEY IN CHILDREN: ONE SINGLE-CENTER EXPERIENCE IN TURKEY

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Introduction: To determine the clinical features and outcomes of children with horseshoe kidney. Horseshoe kidney(HSK) is the most frequent renal fusion anomalies that seen in 1/400 incidence. Nephrolitihiasis, recurrent urinary tract infection (UTI), hydronephrosis, malignancy can be seen as complications of HSK.
Material and methods: Medical records of the 26 patients were valuated retrospectively. Age, gender, age at diagnose, clinical presentation (incidentally, antenatally, urinary complaints), follow-up duration, blood pressure, serum creatinine(eGFR), hematuria, proteinuria, UTI, bladder disfunction, ultrasound(US) findings, static cortical scintigraphy(scar, diffential counts), accompaing urinary (VUR, hyronephrosis, obstructive uropathy) and nonurinary anomalies were determined.
Results: There were 26 children (17 girls and 9 boys) aged 72 (1–162) months The mean folow-up duration was 39 (1–132) months. Presenting symptoms were urinary related symptoms in 7 (27%) patients. Ultrasound findings were horseshoe kidney in 12 (46.2%), normal in 5(19.2%), size diffence in size between two kidney in 3 (11.5%), lithiasis in 2 (7.7%), rotation anomaly of he kidney in 1 (3.8%) and hydronephrosis in 1 (3.8%) patient. VUR(Grade 1) was detected only in 1 (3.8%) patients. Five patient (19.2%) had bladder dysfunction and recurrent UTI. Asymmetrical renal cortical function with a relative function difference > 10% between 2 kidneys was found in 9 (34.5%) cases. Hypertension, proteinuria, decrease in eGFR were not detected in any patients.
Conclusions: Most of the patients diagnosed incidentally and ultrasound could detect only half of the patients. Althought our renal complication rates were low, HSKs should be follow-up for renal complications.

P-070 CLINICAL SIGNIFICANCE OF GRADE 4 AND 5 VESICOURETERAL REFLUX WITHOUT RENAL PELVIC DILATATION ON ULTRASONOGRAPHY

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Introduction: We aimed to evaluate the clinical significance (association with urinary tract infection and renal scar) of severe (grade 4–5) vesicoureteral reflux (VUR) in the absence of renal collecting system dilatation.
Material and methods: Hospital files of children with primary grade 4–5 VUR were retrospectively evaluated. Those with a follow up period >12 months were enrolled. Age, gender, presenting complaint, laterality of VUR, presence and grade of hydronephrosis, serum creatinine, proteinuria, hypertension, presence and grade of renal scar by DMSA scintigraphy, need for anti-reflux surgery and follow-up period were recorded. Patients were grouped as those with (Group 1) and (Group 2) without renal pelvis dilatation. Both groups were compared for the variables evaluated.
Results: 46 patients were enrolled in the study. Group 1 (n = 30) and Group 2 (n = 16) were not different with respect to presence of renal scar (19/30 vs 11/16) and need for anti-reflux surgery (16/30 vs 9/16). Similary serum creatinine, proteinuria and hypertension freauency were not different between the groups.
Conclusions: Presence of severe VUR is associated with renal scar and requires anti-reflux surgery even though collecting system is normal by ultrasonography.

P-071 ABSENCE OF LEUKOCYTES IN SEDIMENTS OF URINE SAMPLES WITH POSITIVE LEUKOCYTE ESTERASE TEST: CLINICAL PREDICTIVE VALUE

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Introduction: Urinary tract infection (UTI) is the second most common bacterial infection in children. False negative or positive results may occur in leukocyte esterase (LE) test which is used to determine UTI. In our study, we aimed to investigate the clinical significance of normal urine sediment findings in the presence of positive LE test in children being evaluated for UTI.
Material and methods: A total of 303 patients who were included in this study. Patients were divided into 2 groups: Group 1; 123 patients with a positive leukocyte esterase strip test and no pyuria in urine sediment. Group 2; 164 patients with a positive leukocyte esterase strip test and pyuria in urine sediment. Demographic characteristics, symptoms, physical findings, the presence of renal anomalies, nitrite positivity in urine, urine density, urine culture results, serum creatinine and C-reactive protein levels were recorded.
Results: Group 1 included 123 patients (114 girls, 9 boys) and Group 2 included 164 patients (134 girls, 30 boys). The rate of girls in Group 1 was significantly higher than in Group 2 ($p < 0.001$). Mean age of the girls and boys were 85.2 ± 46.6 and 44.5 ± 41.4 months, respectively. Physical examination revealed vulvovaginitis in 51 (44.7%) girls in Group 1 and in 41 girls (30.6%) in Group 2 ($p = 0.022$).
Conclusions: Almost half of the girls with positive urine LE test and absence of pyuria had vulvovaginits. Thus, physical examination of perineum is important in girls presenting with lower urinary tract symptoms and positive LE test.

P-072 BLADDER DYSFUNCTION REASON OF KIDNEY FAILURE; LEGS LENGTH DISCREPANCY CAN BE AN AWARENESS SYMPTOM OF BLADDER DYSFUNCTION

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Introduction: Stigma of urinary incontinence is still present. Approximately 6% of patients undergoing renal transplant each year have ESRD due to lower urinary tract abnormality.
Orthopedist approach for Lower extremities discrepancy usually is tightly surgical repair, without broader physical examination, which would identify the bladder function problem and preserve kidney function.
Material and methods: A boy 16 years, presented with ESRD Cre: 1080 and Urea:94. Incontinent and constipated, both paretic legs (left shorter with contractures), couldn't walk properly, spinal dysraphism. Leg orthopedic operation three years before presentation. Nurologically FVM gr 3, RTM are lacking. Severe hydronephrotic on ultrasound and with chronic pyelonephritis. Large bladder with 2.5 L urine.
Results: Second day after catheterization developed Hemorrhagic cystitis. Cystrometry: no bladder contraction during feeling. At 500 ml leaked with cough, and on 600 ml leaked continuously. Pressure almost 15 mmH2O. No sensation. Bladder cooling test positive. He felt heat water, had pain on feeling, no contraction. Cold water: during feeling no cold sensation, severe pain and contractions (pressure was 58mmH2O maximal). Not able to present an uroflow curve, micturation: 80 ml with straining, 525 ml residuals. During CIC he was wet even with 150–200 ml urine. MRI: terminal meningocystocele. Sacrum ends at S2 (partial sacrum agenesis), dural sac is bulging downwards and frontally in a few cysts. It is difficult to exactly see where conus ends and if there is a syringohydromyelia.
An indwelling catheter on free drainage is no guarantee of a constantly low intravesical pressure, cause of phasic bladder contractions which occur despite catheter drainage will damage upper urinary tract.
Conclusions: Children with NBD require multidisciplinary team care: pediatricians, neurosurgeon, urologist, nephrologists, orthopedics, allied medical specialists.
Treatment aim: to achieve a low-pressure bladder and prevent posttransplant infection. Options: conservative modalities:clean intermittent catheterization and bladder relaxants. Invasive modalities: bladder augmentation, intestinal conduit, or external sphincterotomy.

P-073 EFFICACY OF 4-MONTH TENS THERAPY FOR OAB - OVERACTIVE BLADDER

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Introduction: Overactive bladder (OAB) is a common type of bladder dysfunction in children. Typical symptoms include urgency, frequent voiding, holding maneuvers and day-time incontinence. ICCS (International Childrens Continence Society) recommends standard urotherapy as first line treatment and either pharmacotherapy or TENS (transcutaneous electrical nerve stimulation) as second line treatment. Single studies have confirmed that TENS may be a valuable method in children, but queries are raised on the length and intensity of TENS therapy necessary to achieve response.
Material and methods: 46 children aged 5–18 years old with OAB who had not responded fully to 4-weeks standard urotherapy were included. Inclusion criteria were typical symptoms of OAB including urgency, frequent voiding, holding maneuvers and day-time incontinence. Children with urogenital malformations, constipation, increased bladder capacity >150% EBC, significant post-void residual and uroflow curve other than tower or bell-shaped were excluded. Duration of TENS therapy was planned for 4 months and was performed at home twice a day (1 h in the morning and 1 h in the evening) with a frequency of 2 Hz. Treatment results were evaluated according to bladder diary, frequency/volume chart and uroflowmetry performed before and after TENS therapy.
Results: A significant decrease in the number of wet-days (−2.78 from 6.74/14 days) (p = 0,0004) and in the number of days with urgency episodes (−2.6 from 6.45/14 days) (p = 0,0015) was observed. 8/34 (25%) children presenting day-time urge-incontinence were full responders, 10/34 (29%) were partial responders and the remaining 16/34 (47%) were non-responders according to ICCS definitions of response to treatment. Bladder capacity remained unchanged before (average MVV 263,5 ml) and after treatment (average MVV 267,1 ml).
Conclusions: TENS is an effective nonpharmacological treatment option for children with overactive bladder who have not responded to standard urotherapy. Four months of TENS therapy is a suboptimal period to reach satisfactory results. It should be offered to children suffering from OAB symptoms as a non-invasive treatment modality alternative to pharmacotherapy.

P-074 ASSESSMENT MARKERS OF SCLEROSIS AND COLLAGENOPATHY IN THE DIAGNOSIS OF REFLUX NEPHROPATHY IN CHILDREN WITH VESICOURETERAL REFLUX

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Introduction: To reveal the informative indicators for the diagnosis of reflux nephropathy (RN) in children with vesicoureteral reflux (VUR) based on the study of the urinary excretion of transforming growth factor (TGF-β1), angiotensin II (Ang II) and markers of collagenopathy (peptide-bound hydroxyproline (PBH) and free hydroxyproline (FH)).
Material and methods: The study group included 71 children with varying degrees of VUR in age from 1 year to 14 years (average age of 5.89 ± 0.45 years), including 52 girls (73.2%, $\chi^2 = 6.72$, $p < 0.05$). All children were divided according to the results of DMSA-scan into 3 groups depending on the degree of renal scars and reflux nephropathy (RN): 9 p. with VUR without renal scars, 17 p. with mild RN (scars 1–3) and 45 p. with severe RN (> 3 scars). Twenty healthy children served as controls, aged 6.24 ± 0.31 year. Urinary excretion and ratios over creatinine of TGF-β1 and Ang II were examined by ELISA.
The level of free and peptide-bound hydroxyproline in the urine was determined by the method of estimation of the density of the red Chromogen, resulting from the oxidation and decarboxylation of hydroxyproline molecules and condensation of oxidation product with paradimethylaminobenzaldehyde.
Results: The level of urinary excretion of AngII and TGF-β1, PBH and FH was significantly higher in all patients with VUR compared to the healthy children. The patients with III-IV degree of RN to have increased urinary excretion of AngII and TGF-β1, PBH and the level was significantly higher than in children with I-II degree of RN ($p < 0.05$), confirming their association with the severity of sclerotic processes in the tubulointersticial tissues. There was a strong direct correlation between the level of excretion of Ang II, PBH, TGF-β1 and the degree of reflux nephropathy ($r = 0.64$, $p < 0.05$) and a moderate inverse correlation

between the FH and the degree of reflux nephropathy ($r = -0.57$, $p < 0.05$).
Conclusions: Established correlation between urinary excretion of AngII and TGF-β1, PBH and FH and the severity of tubulointerstitial damage in children with VUR, evidence of their potential use as diagnostic markers of nephrosclerosis and dynamic observation of patients with vesicoureteral reflux.

P-075 CEFOTAXIME VERSUS PIPERACILLIN-TAZOBACTAM AS EMPIRICAL TREATMENT FOR FEBRILE URINARY TRACT INFECTION IN INFANTS LESS THAN 3 MONTHS OLD

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Introduction: In 2011, American Academy of pediatrics reported clinical practice guideline for the management of the initial urinary tract infection (UIT) in febrile infants more than 2 months old. In 2001, EAU guidelines just suggested subsequent dosage adjustment of antibiotics to compensate for renal function deficit and some contraindicated drugs in young infants with UTI. There was a lack of clinical guideline for UTI under the age of 2 years. This study was conducted to evaluate the efficacy and safety of piperacillin-tazobactam compared with cefotaxime as empirical antibiotics treatment for febrile UTI in infants less than 3 months old.
Material and methods: Infants less than 3 months old who admitted for febrile UTI between Jan 2014 and Feb 2017 were enrolled, and their medical records were retrospectively reviewed. Clinical characteristics and outcomes were compared according to antimicrobial usage.
Results: Eighty three infants (66 boys and 17 girls) were enrolled in this study. Eighteen patients (21.7%) experienced recurrent UTI during the follow-up period. Urine cultures were proven in 76 patients (91.6%), blood cultures were isolated in 9 (10.8%), and ESBL were positive in 13 (15.7%). Thirty five patients (42.2%) were treated with cefotaxime, 39 (47%) were treated with piperacillin-tazobactam, and 8 (9.6%) were treated with switching from a non-carbapenem to a carbapenem. There were no significant differences in clinical characteristics between cefotaxime and piperacillin-tazobactam treatment groups. Four patients among 35 (11.4%) in cefotaxime group and 12 among 39 (30.8%) in piperacillin-tazobactam group had recurrent UTI ($P = 0.052$). Factors associated with recurrence of UTI in infants less than 3 months old were ESBL-producing bacteria in urine culture ($P = 0.026$), bacteremia ($P = 0.009$), and elevated blood urea nitrogen and creatinine ($P = 0.003$, $P = 0.034$, respectively).
Conclusions: Piperacillin-tazobactam antibiotics showed no favorable outcomes in terms of recurrence of UTI as empirical treatment for UTIs in infants less than 3 months old comparing with cefotaxime. Close clinical follow-up monitoring should be maintained to detect early on whether patients with risk factors have recurrent UTI regardless of urinary tract anomalies.

P-076 MULTIDRUG RESISTANT BACTERIA IN CHILDREN WITH URINARY TRACT INFECTIONS AND THEIR MANAGEMENT

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Introduction: In recent years multidrug resistant bacteria has become increasingly recognised as a cause of urinary tract infections (UTI) in children. The aim of our study was to find out the spectrum of bacteria identified in our children with UTI in the last year, including multidrug resistant ones, and to compare it with the study performed in the period from 2011 to 2013. In addition, we were interested in antibiotic treatment.
Material and methods: 103 children, hospitalized at our Nephrology Unit in 2016 with culture confirmed UTI, were included in the study in retrospective manner. Medical documentation was examined and some clinical data, results of urine culture, diagnostics and treatment were collected and compared with the results of similar study performed during the period 2011–2013.
Results: There were 25 boys (24.3%) and 78 girls (75.7%) included in the study. In 9.7% congenital anomaly was present before admission which is less than in the period 2011–2013. In 97% of children US of kidney was performed and in 26.2% further diagnostic procedure has been indicated. *E. coli* was the most common pathogen found in 80.5% of children (before 78.4%), in 47.0% sensitive to all tested antibiotics (before 48.9%). Resistance to ampicillin was present in 48.2%, trimethoprim-sulfamethoxazole in 18.1%, amoxicillin-clavulanate in 9.6%, quinolones in 7.2%, gentamicin in 4.8%, which is similar to the previous study. Multidrug resistant bacteria were found in 4.9% (before 5.5%). Children were treated with gentamicin in 69.9%, amoxicillin-clavulanate in 17.5%, cephalosporins in 3.9%, similar to the period 2011–2013. Reserve antibiotics were prescribed only in 1.9% cases (before 2.3%). Treatment protocol was changed in 9.7% (before 6.4%).
Conclusions: *E. coli* remains the leading uropathogen in our children with UTI, with similar resistant patterns and treatment. Multidrug resistant bacteria have not increased and the prescription of reserved antibiotics was rarely needed.

P-077 THE CHANGING PATTERN OF ANTIBIOTIC RESISTANCE IN URINARY TRACT INFECTIONS

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Introduction: Urinary tract infection is one of the most common bacterial infections in pediatric age group. The resistance profile of uropathogens is critical in treatment planning. The aim of this study was to assess the resistance patterns of bacteria isolated from urinary tract infections to commonly used antimicrobials and evaluate the options for ampirical treatment.
Material and methods: A retrospective cross-sectional study was performed in children diagnosed with either lower urinary tract infection or acute pyelonephritis between January 2016 and June 2016. Among 2000 urinary isolates reviewed 460 patients' culture results were found eligible. Patient demographics (age, sex), infection site (upper/ lower urinary tract), resistance profile of the urine samples and agent used were noted.
Results: Median age of the patients was 44.5 months (min:0.1, max:208) and male to female ratio was 1/3.8. Acute pyelonephritis episodes were observed in 50.7% of the patients. Previous urinary tract infection history was found in 47.2% of the patients. Prophylaxis was used by 19.1% of the patients. The most common causative agent was E.coli (80.4%) followed by K.pneumonia (8.3%) and 22.8% of the strains were producing extended spectrum beta lactamase. Resistance to ampicilline was found as 66.3%, co-trimexasole 41.4%, amoxicilline-clavulonate 35%, cefotaxime 24.6%, nitrofurantoin 16.1%, gentamycine 11.1% and to ertapenem as 1.3% in all isolates.
Conclusions: The study revealed that E.coli was the most commonly isolated pathogen in the sample. Extended spectrum beta lactamase producing bacteria is a major problem which constituted the 22.8% of our isolates. Ampiric antibiotic therapy for urinary tract infection should be switched to proper regimen according to the urine culture results. Having

regional antibiotic resistance patterns in hand would aid in choosing the right ampiric treatment.

P-078 IS URINE INTERCELLULAR ADHESION MOLECULE-1 A MARKER OF RENAL DISORDER IN CHILDREN WITH URETEROPELVIC JUNCTION OBSTRUCTION?

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Introduction: Hydronephrosis (HN) is a common pathology in pediatric nephrology. Ureteropelvic junction obstruction (UPJO) is the most common cause of severe hydronephrosis in children.
The evolution of obstructive nephropathy can be envisioned as an overlapping sequence of cellular events, including tubular dilatation, interstitial inflammation, followed by glomerulotubular injury and progressive interstitial fibrosis. Inflammation is mediated by the up-regulation of chemokines, adhesion molecules (intercellular adhesion molecule-1, vascular cell adhesion molecule-1), monocyte chemotactic protein-1, and osteopontin.
We aimed to investigate whether urine intercellular adhesion molecule-1 (ICAM-1) might serve as a marker of renal disorder in children with hydronephrosis caused by ureteropelvic junction obstruction.
Material and methods: 29 children with severe HN (M- 23; F- 6; mean age 3.8 ± 5.02 years) were compared with 23 participants with mild HN (mean age 6.29 ± 5.04 yrs) and with 19 (mean age 5.28 ± 4.25 yrs) healthy peers age- and sex-matched. ICAM-1 was measured in urine using ELISA method, and was expressed as ng/ mg Cre.
Results: Urine ICAM-1/ uCre levels were significantly higher in HN children than healthy controls ($P < 0.01$), and in severe HN when compared to mild HN ($P < 0.05$). A negative correlation between uICAM-1/ uCre and DRF was found in all studied children ($r = -0.5$, $P < 0.05$).
Conclusions: Present study showed significantly higher uICAM-1/ uCre levels in children with severe HN than with mild HN. Larger studies are invited to confirm the clinical usefulness of ICAM-1 as biomarker in the diagnosis and follow-up of children with obstructive nephropathy. It seemed to us that uICAM-1 could be a good marker of renal disorder, and might have the potential to predict which patients will require surgery.

P-079 IS ROUTINE VOIDING CYSTOURETHROGRAPHY NECESSARY IN CHILDREN WITH MULTICYSTIC DYSPLASTIC KIDNEY DISEASE?

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Introduction: Multicystic dysplastic kidney(MCDK) is the most common cause of cystic kidney diseases in children. Voiding cystourethrography(VCUG) in patients with MCDK was performed routinely to detect vesicoureteral reflux(VUR) which is the most common associated contralateral renal abnormality. However, the routine use of VCUG is currently controversial.
We aimed to evaluate the necessity of routine VCUG in patients with MCDK.
Material and methods: Eighty seven children(27 girls, 60 boys) with MCDK followed-up at our pediatric nephrology outpatient clinic were included in the study. Ultrasonography(USG), ^{Tc99}DMSA scan, ^{Tc99}MAG3 scans and VCUG findings, clinical and laboratory data were retrospectively evaluated.
Results: The mean age of the patients were 9.59 ± 7.1 (2–23) years. Antenatal hydronephrosis or MCDK was diagnosed in 71 (81.6%) patients. Seventy-four patients (%85) were diagnosedMCDK within the first month of life.
Six(6.9%) patients had grade 1–3 and 4(4.6%) had grade 4–5 VUR in contralateral kidney. Bilateral VUR was present in two(2.3%) patients. Voiding cystourethrography was not performed in 14 patients. None of these patients had hydronephrosis and/or urinary tract infection(UTI). At the follow-up, four patients with VUR underwent ureteral reimplantation and endoscopic subureteric injection was performed in four patients.
Nine of VUR patients had UTI and/or hydronephrosis. Three patients with VUR were asymptomatic. Two of them followed-up conservatively and surgical intervention was performed in one patient. Hydronephrosis and/or UTI in patients with VUR were significantly higher than the patients without VUR($p = 0.0001$).
Ureteropelvic junction stenosis(one obstructive and two non-obstructive) of the solitary kidney was found in three patients, they all were detected postnatally. Eighty seven children(27 girls, 60 boys) with MCDK followed-up at our pediatric nephrology outpatient clinic were included in the study. Ultrasonography(USG), ^{Tc99}DMSA scan, ^{Tc99}MAG3 scans and VCUG findings, clinical and laboratory data were retrospectively evaluated.
Conclusions: Routine VCUG in healthy children diagnosed with unilateral MCDK is not necessary in those children without UTI and/or contralateral hydronephrosis.
Families should be informed about the signs and symptoms of urinary tract infection and selective screening for VUR should be considered only for patients with contralateral abnormality and/or patients with UTI.

P-080 MICRoBIOLOGIcAL STRUCTURE OF URINARY TRACT INFECTION IN CHILDREN IN ORENBURG REGION

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Introduction: The aim of the study was to establish micribiological structure of urinary tract infection (UTI) in children in Orenburg region.
Material and methods: We examined 6392 cases of children in period 2006–2016 years who treated with UTI in paediatric clinic №6 in Orenburg region. There were 3451 (53,9%) girls and 2941 (46,1%) boys from 1 till 15 years. All children underwent special microbiological examination of urine including determination of degree of bacteriuria sectoral sowing on blood agar medium and Endo (Feldman J.M. et al., 1984) and quantitative parameters (Grachev N. et al., 1986). Species identification allocated urine's bacteria carried out by conventional methods (Birger M.I., 1982). We determined persistence markers of isolated strains of microorganisms: antilysozyme activity (ALA), antiinterferon activity (AIA) and seroresistance (SR) (Bukharin O.V. et al., 1996).
Results: We established that *E. coli* prevailed in microbiological structure of UTI (78,9%, $n = 5049$). Proportion of other bacteria which were isolated from urine was significantly lower (21,1%, $n = 1343$): Klebsiella (16,8%, $n = 1080$), Proteus (4,3%, $n = 263$). We studied also resistance of bacteria isolated from urine to antibiotics. *Escherichia coli* were resistance to ampicillin (51,5%), amoxicillin (51,5%), 1-st generations of cephalosporins (46%) and penicillins with clavulanic acid (35,5%). Klebsiella were 100% resistant to ampicillin and amoxicillin, penicillins with clavulanic acid (30%), 1-st generations of cephalosporins (56%), furans (40%). Proteus exhibit low sensitivity to furans (resistance 45%), ampicillin and amoxycillin (74%), 1-st generations of cephalosporins (100%), 2-nd generations of cephalosporins (38%), 100% to penicillins with clavulanic acid. We determined that microorganism strains isolated from urine of patients with UTI were characterized by high levels of SR, ALA, AIA.
Conclusions: These data were used in creation of regional register of antimicrobial resistance paediatric patients with UTI. High levels of SR, ALA, AIA show high persistent and pathological potential of urine's bacteria in pathogenesis of UTI.

P-081 FOLLOW-UP OF PATIENTS WITH HYDRONEPRHOSIS; WHEN TO WORRY?

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Introduction: Hydronephrosis is the most common genitourinary system anomaly detected by ultrasound (US) during antenatal period. However, there is no consensus about evaluation and follow-up of these patients in postnatal period.
Material and methods: The data of 203 patients followed with hydronephrosis since 2010 were evaluated retrospectively and the role of postnatal ultrasonography (US) in predicting the final diagnosis and need of surgery was investigated. Patient's the first postnatal, second and the last US, voiding cystourethrography (VCUG) and diuretic renograms (MAG3) were recorded. US findings of different etiologies, anteroposterior diameters (APD) of operated/nonoperated groups are compared.
Results: Regarding the renal pelvic APD in the first postnatal US; mild, moderate and severe HN was detected in 23.1%, 34.4%, 42.4% of them respectively. It was determined that according to the first USG, patients with low grade of hydronephrosis had higher recovery rate and first USG findings gave reliable information about prognosis. The risk of obstruction and surgical intervention increased significantly with the increase in the degree of hydronephrosis in the first USG. Renal pelvic APD of ≤11 mm was found to be an important predictor of complete recovery ($p < 0.001$; AUC: 0.714).
Conclusions: The first USG findings give reliable information about prognosis of patients and guide us to select the appropriate imaging studies and to determine the need for antibiotic prophylaxis at the same time. The risk of obstruction and surgical interventions increased significantly with the higher degrees of hydronephrosis in the first USG of patient.

P-082 THE FREQUENCY OF FAMILIAL CAKUT

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Introduction: Congenital abnormalities of the urinary tract and kidney (CAKUT) affect 3–4% of the population. The prevalence of CAKUT is rather frequent in Turkey due to high rates of consanguineous marriages. In the present study, we aimed to determine the prevalence of CAKUT in asymptomatic first-degree relatives of the patients and to emphasize the importance of genetic inheritance.
Material and methods: All patients followed in the pediatric nephrology outpatient clinic and diagnosed as CAKUT between the years 1998–2016 were evaluated. First-degree relatives with previously undiagnosed CAKUT were invited to take part of this study. Urinary tract ultrasounds were performed by the same radiologist and pedigrees were drawn in the participating families. Patients with incomplete duplex system were not included to the study.
Results: 1228 of 6695 patients (18%) had been diagnosed as CAKUT. A total of 150 CAKUT patients (12%) and their families accepted to participate in the study. Patients with secondary vesicoureteral reflux (VUR) ($n = 3$) and hydronephrosis with extrarenal pelvis ($n = 2$) were excluded; finally 145 patients and their families were enrolled in the study. Urinary malformations of the index cases were VUR ($n = 46$), ureteropelvic junction (UPJ) obstruction ($n = 16$), renal hypodysplasia ($n = 13$), posterior uretral valve (PUV) ($n = 13$), multicystic dysplastic kidney ($n = 8$), renal agenesis ($n = 5$) and other abnormalities ($n = 44$).
Ultrasonographic evaluation was performed on 415 siblings and parents with previously undiagnosed CAKUT. A urinary abnormality was found in 22 first-degree relatives (15%); these were renal agenesis ($n = 7$), renal hypoplasia ($n = 6$), ectopic kidney ($n = 1$), and hydronephrosis ($n = 8$).
Conclusions: The rate of familial CAKUT is 15%. This finding emphasizes the importance of performing urinary tract US in the family members of CAKUT patients. Genetic studies may contribute to understand the pathogenesis of CAKUT.

P-083 RENAL TRACT ABNORMALITIES MISSED IN A HISTORICAL COHORT OF YOUNG OMANI CHILDREN WITH UTI, IF THE NICE AND AAP IMAGING GUIDELINES WERE APPLIED

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Introduction: Urinary tract infection (UTI) in pediatrics, especially acute pyelonephritis (APN), are susceptible to renal scarring that is associated with long-term complications such as hypertension, proteinuria, and reduced renal function. The United Kingdom (UK) presented new guidelines for imaging procedures for UTI but without providing levels of evidence. The American Academy for Pediatrics (AAP) published revised guidelines for children, including levels of evidence grading. We are unsure which guidelines is suitable for Omani children.
Material and methods: A retrospective analysis of children below 14 years with UTI between 1992 and 2010, who underwent full investigation at SQUH according to Royal College of Physician (RCP). Three hundred three children were evaluated for inclusion in the study. Out of them, 298 children had completed the data and the required investigations. We calculated the proportion of abnormalities which would have been missed had the new guidelines from the National Institute for Health and Care Excellence (NICE) in UK or AAP, been used instead.
Results: Out of all patients, 49 (16%) male, 249 (84%) female, 191 (64%) had recurrent UTI. On the other hand, atypical UTI was present in 116 (39%) patients. E coli in the urine was found in 206 (69%) of the patients, while the other 92 (31%) had non E coli organisms. Hydronephrosis was the most prevalent US finding (in 49 patients).

Percentage of abnormalities potentially missed.

	NICE	*AAP*
HYDRONEPHROSIS	*14%*	*0%*
VESICO-URETERIC REFLUX	*74%*	*85%*
RENAL SCARRING	*33%*	*42%*
DECREASE RENAL UPTAKE	*38%*	*40%*

Conclusions: The prevalence of renal tract abnormalities missed by the new guidelines is high. They should be used with full awareness of their limitations and this should be carefully balanced against using the prior guidelines, that advocate more use of US, VCUG, and DMSA scintigraphy, which are costly, time-consuming, sometimes unpleasant, and associated with radiation exposure as well as prolonged clinic follow-up visits.

P-084 EARLY ULTRASOUND DETECTION OF CONGENITAL ANOMALIES OF THE KIDNEY AND URINARY TRACT

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Introduction: Abdominal ultrasound is currently the primary method of assessing organs inside of the abdomen. In the literature there is no explicit opinion on the need of performing abdominal ultrasound as a

screening test in children. Because many disorders of small children are asymptomatic, the early detection of abdominal abnormalities and the implementation of appropriate treatment in the early stages of the disease may be a chance to avoid serious complications.
The main objective of the study was to demonstrate the potential usefulness of abdominal ultrasound in children as a screening test for the early detection of congenital defects in the abdominal cavity including particularly the urinary tract.
Material and methods: The study included 500 children (252 girls and 248 boys) aged from 1 month to 7 years, who never had ultrasound examination of the abdomen before. All the children had no clinical symptoms.
Results: In the whole group 44.4% were infants, children over 1 year constituted 55.6%. Abnormalities were observed in 13.6% of the evaluated children. Congenital malformations of the kidneys and urinary tract were identified in 5.8% (29/500) patients. These abnormalities accounted for 42.6% (29/68) of all identified anomalies. Significant pelvic dilatation were found in 51.7% (15/29) of children with congenital malformations of the kidneys and urinary tract. Duplicated collecting system with hydronephrosis were found in 6/29 patients (20.7%). In 4/29 patients (13.8%) ultrasound showed enlarged ureter. 2/29 children (6.7%) were diagnosed with asymmetric kidneys. Ureterocele and renal agenesis were identified in single patients.
Conclusions: 1. Abdominal ultrasound is effective for early detection of renal and urinary tract anomalies. 2. In spite of the current prenatal ultrasound anomaly screening program infants are still diagnosed with congenital anomalies of the kidneys and urinary tract which haven't been found in utero.

P-085 RISK FACTORS FOR RENAL SCARRING IN CHILDREN WITH MYELOMENINGOCELE

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Introduction: Most children with myelomeningocele have neurogenic bladder while bladder dysfunction predisposes patients to urinary tract infection (UTI), renal scarring and renal failure. We aimed to evaluate risk factors for renal scarring in these patients.
Material and methods: Fifty-three children with neurogenic bladder due to myelomeningocele (28 male; mean presentation age 18 ± 19 months; current age 7.0 ± 3.6 years), followed-up at least one year after urodynamic testing were enrolled in this single center study. Anthropometric indices, spinal lesion levels, shunt status, ambulatory ability, episodes of UTI, ultrasonographic and urodynamic findings, presence of vesicoureteral reflux (VUR), eGFR and serum cystatin-C levels were recorded. Forty-seven patients (89%) had been performing clean intermittent catheterization (CIC). Patients applying at least ≥75% of CIC suggestions were defined as "compliant". Low bladder capacity was defined as bladder capacity <65% of expected volume by urodynamic testing. Renal scarring was diagnosed by most recent DMSA scans.
Results: The mean follow-up period was 66 ± 34 months. Twenty-four patients (45%) had VP shunt. Spinal lesion levels were as follows: lumbosacral region in 28, lumbar in 13, sacral in 7 and thoracolumbar in 5. DMSA scintigraphy revealed renal scarring in 9 (17%) patients, which was not associated with gender, age of CIC initiation or current age, level of spinal lesion, ambulatory disability or none of urodynamic parameters except low bladder capacity. Significant risk factors for renal scarring are shown in Table 1. There was no difference between eGFR values of the patients with or without scarring, whereas serum cystatin C levels were significantly higher in patients with scarring (0.80 ± 0.20 vs 0.63 ± 0.09, $p = 0.03$).

Table I: Significant risk factors for renal scarring

	All patients $n = 53$	Scarring (+) $n = 9$	Scarring(−) $n = 44$	P
No. of UTI between 0 and 2 years	0.9 ± 1.4	3.0 ± 1.9	0.6 ± 1.0	0.007
UTI episodes > 3, *n (%)*	8 (15%)	4 (44%)	4 (9%)	0.008
VUR (+), *n (%)*	9 (17%)	5 (56%)	4 (9%)	0.001
CIC compliance, *n (%)*	38 (71%)	4 (44%)	34 (90%)	0.002
Low Bladder Capacity, *n (%)*	10 (19%)	5 (56%)	5 (11%)	0.007

Conclusions: CIC compliance and avoidance of UTI may preventrenal scarring in patients with myelomeningocele.

P-086 CONGENITAL ANOMALIES OF THE KIDNEY AND URINARY TRACT (CAKUT) - PRE AND POSTNATAL DIAGNOSIS

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Introduction: Congenital anomalies of the kidney and urinary tract (CAKUT) constitute 20 to 30% of all malformations identified in the prenatal period, with the overall rate of 0.3 to 1.6 per 1000 newborns. CAKUT are the most common cause of chronic kidney disease and end-stage renal disease in the pediatric population. The goal of prenatal screening is to identify pathologic conditions that would require postnatal therapy in order to prevent or delay these complications.
This study aims to determine the incidence rate of CAKUT in newborn and their clinical and imagiologic outcomes.
Material and methods: Retrospective study of medical reports of infants born at Centro Hospitalar Póvoa de Varzim/Vila do Conde, Portugal from 2001 to 2016. Hydronephrosis was defined an anteroposterior diameter of the renal pelvis (APD) ≥ 5 mm and has been classified in mild (5–9 mm), moderate (≥10–14 mm) or severe (≥15 mm). Size and structure of the kidney, dilatation of calices or ureters, and bladder morphology were also considered.
Results: In accordance with the hospitals CAKUT's protocol, 885 children were studied. The incidence was 4.8%, of which 70% were male. Hydronephrosis was diagnosed by prenatal ultrasound in 96% (49% bilateral). In 37% there was no postnatal ultrasound hydronephrosis confirmation. The postnatal diagnosis included: transient hydronephrosis (43%), ureteropelvic junction (5%), vesicoureteral reflux (3.3%), duplex collecting system (3.2%), renal agenesis (1.8%), megaureter (1.1%) and multicystic displastic kidney (0.6%). Surgery was performed in 3.2% of patients. The mean follow up duration was 10 months (1 month-16 years).
Conclusions: Correct identification and adequate follow up of patients at risk is a challenge. Although most of the hydronephrosis detected in the prenatal period do not correspond to significant nephro-urological pathology, early ultrasound diagnosis is important in the detection of severe malformations and prevention of complications.

P-087 FACTORS THAT INFLUENCE DIPSTICKS SENSITIVITY IN URINARY TRACT INFECTION DIAGNOSIS

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Introduction: The authors aimed to analyze the influence of age, sex, uropathy, vesicoureteral reflux (VUR), fever and prophylaxis on dipsticks sensitivity in urinary tract infection (UTI) diagnosis.
Material and methods: Retrospective analysis of positive urine culture results from children and adolescents aged <18 years, during the year of 2015, in a tertiary hospital. Children submitted to renal transplant, with asymptomatic UTI or neuropathic bladder were excluded from the analysis. Statistical comparisons between categorical variables were performed by chi-square tests.
Results: A total of 219 positive urine cultures were analyzed, corresponding to 174 patients (68.4% female) with a median age of 3 years. The most common pathogens identified were *E. coli* (69.4%), *P. mirabilis* (13.7%), E. faecalis (5%) and K. pneumonia (4.6%). Altered dipsticks test results corresponded to urine sediment alterations in 92.5% (vs. 7.5%, $p < 0.001$).Regarding age distribution, dipsticks test alterations were most frequent in children aged >3 years (13.4% vs.38.1% vs.48.5%, in <3 months, 3 months-3 years, >3 years, $p = 0.011$). With regards to the sex distribution, there was a significant rate of alterations in the dipsticks test for males (74.4%) and females (88.7%), although higher in the female sex (76.1% vs. 23.9%, $p = 0.026$). Dipsticks sensitivity was higher for *E. coli* UTI compared to other agents (73.9% vs. 26.1%, $p = 0.002$). The presence of uropathy, VUR, antibiotic prophylaxis or the presence of fever had no impact on sensitivity of dipsticks test.
Conclusions: Rapid urine tests, such as dipsticks and urine sediment analysis are frequently used to guide early diagnosis and treatment of UTI, being often performed at that same time. In the present study, the authors conclude that dipsticks is equivalent to urine sediment when dealing with a suspected UTI, allowing for a rapid and cheap approach. Of all factors analyzed only age and sex had an impact on dipsticks test sensitivity.

P-088 ROLE OF PIC CYSTOGRAPHY FOR EVALUATING VUR IN CHILDREN WITH RECURRENT URINARY TRACT INFECTION

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Introduction: Voiding cystourethrography (VCUG) is considered as the standard evaluation method for identifying vesicoureteral reflux (VUR) in children with urinary tract infections (UTI). The common pediatric problem is the management of children who have normal VCUG finding with recurrent UTI. PIC cystography is a new modality that has been shown to demonstrate ureteral incompetence in pediatric patients who have normal standard reflux studies. In this study, we evaluated the utility of PIC cystography in detecting VUR in patients with recurrent UTIs that standard VCUG failed to reveal.
Material and methods: A retrospective analysis was conducted on 14 patients who were examined with PIC cystography between 2014 and 2016 with a diagnosis of recurrent UTI. All patients had at least one previous negative VCUG. The collected data included patient age, sex, individual ultrasonography (US), DMSA findings, PIC reports and outcome.
Results: A total of 14 patients were included in the study, of which 3 (21.4%) were male and 11 (78.6%) were female. The mean age of the study group was 9.07 ± 3.92 years. The 8 (57.1%) patient had normal US, 5 patients (35.7%) had bilateral scar detected by DMSA. Twelve patients (85.7%) were shown to have VUR on PIC. The relationship between having scar on DMSA and reflux on PIC was evaluated and no statistical difference was found.
Conclusions: According to our results, PIC cystogram appears to be a clinically relevant test in patients who have frequent UTIs with negative VUR on standard imaging modalities. In this sense, we believe that providing prospective randomized study with PIC cystogram is essential to optimize the algorithm for the treatment of children with febrile UTIs to show the hidden VUR.

P-089 FACTORS THAT INFLUENCE ANTIBIOTIC RESISTANCE OF *E. coli* URINARY TRACT INFECTIONS

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Introduction: The authors aimed to analyze the influence of age, presence of uropathy, vesicoureteral reflux (VUR), antibiotic prophylaxis and previous urinary tract infection (UTI) on antibiotic resistance of *E. coli* UTI.
Material and methods: Retrospective analysis of positive urine culture results from children and adolescents aged <18 years in a tertiary hospital in 2015. Children submitted to renal transplant, with asymptomatic UTI or neuropathic bladder were excluded from the analysis. Statistical comparisons between categorical variables were performed by chi-square tests.
Results: A total of 219 positive urine cultures were analyzed, corresponding to 174 patients (68.4% female) with a median age of 3 years. The most common pathogens identified were *E. coli* (69.4%), *P. mirabilis* (13.7%), E. faecalis (5%) and *K. pneumoniae* (4.6%). Considering only the group with *E. coli* infections ($n = 94$), 40.4% of children presented an uropathy, being VUR (21.6%) and primary hydronephrosis (18.6%) the most common. *E. coli* antibiotic resistance was higher for ampicillin (56%), fosfomycin (33.3%), trimethoprim-sulfamethoxazole (30.8%), nitrofurantoin (23%), amoxicillin/clavulanic acid (20.2%) and cefuroxime (6.6%). Regarding age distribution, amoxicillin/clavulanic acid resistance was higher among children aged 3mo-3 yr., and >3 yr. (51.6% vs. 45.2%, $p = 0.034$), while trimethoprim-sulfamethoxazole resistance was highest in children >3 yr. (62.2%, $p = 0.048$). Resistance to trimethoprim-sulfamethoxazole was higher in children with uropathy (57.7% vs. 42.3%, $p = 0.035$) and lower in those under prophylaxis (25% vs. 75%, $p = 0.001$). Children under prophylaxis had a low resistance to cefuroxime (13.3% vs. 2.2%, $p = 0.024$). The presence of uropathy, VUR, prophylaxis and a previous UTI had no impact on ampicillin and amoxicillin/clavulanic acid *E. coli* resistance.
Conclusions: Amoxicillin/clavulanic acid and second generation cephalosporins are the drugs of choice in many centers. As none of the factors analyzed in the present study seemed to have an impact on the sensitivity of *E. coli* UTI to these group of drugs, the authors conclude that these agents remain an adequate choice.

P-090 ANTENATAL RENAL PELVIS DILATATION: DETERMINING RISK AND PROGNOSTIC FACTORS WITH POSTNATAL SIGNIFICANCE: UPDATE 2009–2013

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Introduction: As awareness and techniques in antenatal screening advance, detection of foetal anomalies is increasing. Little evidence has been accumulated to determine the importance of antenatal detection of renal pelvis dilatation (RPD) and its association with pathology postnatally. We retrospectively audited all infants born from 2009 to 2013 with RPD and followed their postnatal course in an aim to determine critical RPD dimensions and the importance of associated risk factors.
Material and methods: We collected data retrospectively on all patients identified by the obstetric ultrasound department as having RPD antenatally in a tertiary neonatal unit with approx. 6500 births a year. The data was compared to and reflected upon the RPD guideline used across Scotland for identifying infants with significant RPD antenatally and their postnatal management. The data was statistically analysed to determine the risk factors and prognostic factors in outcome of RPD detected antenatally.
Results: In five years 107 patients were identified as having RPD. Seventy-seven patients were male and 30 female (2.6:1). Seventy-three identified as high risk and 34 low risk. Of the high risk patients 53 continued to be high risk, with 18 requiring surgery and 8 had UTI. Those that became low risk did not require surgery or UTI. One that did not attend the follow up scan required surgery and had a UTI. Of the low risk patients 11 became high risk with 4 requiring surgery and 5 having breakthrough UTI. Those that continued to be low risk did not require surgery with one UTI. One that did not attend follow up had 1 UTI. In total 24 required surgery, 11 female, 13 male. Thirty-seven percent of females required surgery compared to 17% of males referred. The females had 1 or two risk factors each, however of the boys 4 had one risk factor and 2 had no risk factors, 1 of which did not have high risk RPD dimensions. Fifteen patients had a breakthrough UTI despite trimethoprim prophylaxis. One patient had a UTI not on prophylaxis, however this infant had RPD >15 mm. Fifty-nine referred to nephrology.
Conclusions: Risk factors and RPD >15 mm were the strongest indicator for development of significant RPD requiring surgery or resulting in symptomatic UTI. High risk patients who became low risk at second remained at risk of UTI and surgery. Low risk patients who became high risk became at risk of surgery and UTI. Those who continued to be low risk did not require surgery. Incidence of breakthrough UTI whilst on prophylaxis reflects the need for a high index of suspicion of UTI in a child with RPD. The guideline reflects infants who are at risk of ongoing renal and urology problems postnatally and highlights the importance of follow up in all patients.

P-091 DIAGNOSIS, MANAGEMENT AND OUTCOME OF POSTERIOR URETHRAL VALVES IN CHILDREN AT A TERTIARY CENTRE IN SOUTH AFRICA

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Introduction: One of the most common causes of congenital abnormalities of the kidneys and urinary tract is posterior urethral valves (PUV). This can easily be diagnosed by third trimester scan which leads to better prognosis. However, in resource-limited environments, the diagnosis is often missed or made late upon presentation with life-threatening clinical features. The study aimed to review the age at diagnosis, management and outcomes of PUV in children in a tertiary hospital in South Africa.
Material and methods: All files of children who were referred to the Paediatric Nephrology Unit at Steve Biko Academic Hospital in Pretoria, South Africa, from January 2000 to December 2015 were retrospectively reviewed. Data extracted included: antenatal diagnosis of PUV, age at diagnosis postnatally, presence of acute kidney injury at diagnosis, management, complications and outcomes i.e. chronic kidney disease (CKD) or death.
Results: A total of 62 boys with a mean and median age at diagnosis of 21.7 and 3.5 months respectively, had PUV. Micturating cystourethrograms were done in 81% of patients. Sonar reports showed that 18% of patients had renal dysplasia. Primary valve ablation was conducted in 36% of patients while diversions (vesicostomies, nephrostomies and ureterostomies) were done in 50% of patients. The commonest presenting clinical features were urinary retention 28/62 (45%), failure to thrive 15/62 (24%), urinary tract infections 6/62 (10%). Fifty-eight percent presented with associated acute kidney injury. 18/52 (36%) had CKD, 16/62 (26%) had hypertension and 6/16 (37%) had proteinuria at follow-up. Only 6/60 (10%) were diagnosed antenatally.
Conclusions: A deliberate policy to have third trimester antenatal scans may improve outcome of affected children.

P-092 BLADDER DIVERTICULA CAUSED BY OCCIPITAL HORN SYNDROME-A CASE-REPORT

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Introduction: Occipital horn syndrome is an X-linked recessive connective tissue disorder caused by a deficiency in the transport of copper. The disorder is considered a milder variant of Menkes disease, associated with mutations in the ATP7A gene. Children may present with features such as intractable diarrhea, bladder diverticula or recurrent UTI. Motor development is delayed due to muscle hypotonia. Inguinal hernia is common. Arterial aneurysms have also been described. Deformations in the skeleton are present. Diagnosis is based on the clinical features and confirmed by identification of a mutation in the ATP7A gene. Radiography shows characteristic occipital horns which are symmetric exostoses protruding from the occipital bone.
Material and methods: We present a 6,5 year old boy with Occipital horn syndrome who has a large bladder diverticula.
Results: A 14 month-old boy was addmited with macrohaematuria and recurrent UTI. He presented at birth with poor muscle tone and fracture of the occipital bone. The patient was hypotrofic and hypotonic with delayed psyhomotoric development. His ultrasound examination revealed massive bladder diverticula, which was confirmd by VCUG. The urodynamics was normal. In laboratory findings low cooper and ceruloplasmin were found. Genetic analysis showed mutation of ATP7A gene (duplication in exons 11 and 12) and diagnosis of Occipital horn syndrom was made.
During the follow-up the diverticula did not empty propertly and boy suffered numerous UTI despite prophylactic antibiotic. At the beginning clean intermitent cateterisation was helpfull. But as the infection continued and new diverticula appeared, he underwent cystostoma and now he has been free of infection for a six months.
Conclusions: Large bladder diverticula can be the part of underlying connective tissue disorder. Our case showed the boy with large bladder diverticula in a rare Occipital horn syndrom caused by a deficiency in the transport of copper, associated with mutations in the ATP7A gene.

P-093 PREVENTION OF RELAPSE OF UTI IN CHILDREN

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Introduction: The aim of the study was to evaluate the effectiveness of antirelapse therapy of UTI in children.
Material and methods: We examined 236 children aged from 1 year to 17 years with UTI. Patients underwent DMSA scan and US examination, microbiological investigations of urine, renal function on the sample Rehberg translated at the Schwartz formula. Examined patients were divided in 2 groups:
I – children who received Canephron N (n = 117);
II – children who didn't receive Canephron N (n = 119).
Definition of indicators was carried out in dynamics of 1 year from the start of antirelapse therapy of UTI with antimicrobial and fitodrag Canephron N. The dose was adjusted to patients individually depending on age, weight of the child.
Results: The main performance indicators of antirelapse therapy of UTI with antimicrobial and fitodrag Canephron N was decrease on 27% ($p < 0.05$) frequency and duration of UTI in patients who received Canephron N (n = 117).
Conclusions: Antirelapse therapy of UTI with antimicrobial and fitodrag Canephron N as a measure of prevention of progression of UTI is effective. Evaluating the effectiveness of antirelapse therapy of UTI with antimicrobial and fitodrag Canephron N justifies the use of these groups of drugs as prevention of progression of UTI in children.

P-094 MATRIX METALLOPROTEINASES IN INFANTS WITH URETEROPELVIC JUNCTION OBSTRUCTION

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Introduction: The management of patients with ureteropelvic junction obstruction (UPJO) remains a controversial issue. The objective is the preservation of renal function through early selection of patients who will require surgical intervention as opposed to the others who will be monitored systematically by various imaging methods. Aim of the present study was to measure Matrix Metalloproteinases (MMPs) levels in UPJO patients who were planned to undergo surgery using standard criteria and thus clarify if MMPs levels could serve as potential biomarkers of obstruction in hydronephrosis.
Material and methods: Serum samples of infants with UPJO diagnosis who were planned to undergo surgery were compared to 17 serum samples of healthy age matched controls. MMP2 and MMP9 were quantified using immunoenzymatic (ELISA) assay.
Results: 17 infants with UPJO diagnosis, 14 males and three females, median age 1.5 months (min: one month, max: 20 months), mean anteroposterior diameter in the hydronephrotic kidney 23.4 mm (min: 19 mm, max: 50 mm) and T ½ > 20 min in the MAG3 renogramm were recruited. MMP9 levels were significantly decreased in serum samples of UPJO patients (418.53 ng/ml +/− 256.78 ng/ml) vs control levels (572 ng/ml +/− 226.96 ng/ml) (p = 0.037). MMP2 values were higher in UPJO patients (502.2 ng/ml +/− 64.2 ng/ml) vs controls (471.42 ng/ml +/− 123.35 ng/ml). However, the difference was not statistically significant (p = 0.206).
Conclusions: This study presents decreased concentrations of MMP9 in the serum of patients with obstructive hydronephrosis but the results should be tested in larger population samples and even be evaluated simultaneously with urine samples in order to delineate MMPs ability to serve as obstruction biomarkers.

P-095 GIANT HYDRONEPHROSIS DUE TO URETEROPELVIC JUNCTION OBSTRUCTION: A CASE REPORT

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Introduction: Ureteropelvic junction obstruction (UPJO) is the most common cause of prenatal hydronephrosis and has an incidence of 1 in 1000–1500 newborns. Hydronephrosis caused by UPJ does not always progress, but may increased rapidly and without warning in adult life. Giant hydronephrosis is defined as a hydronephrotic kidney containing more than 1 lt of fluid and the cause is usually due to a delay in diagnosis and treatment.
Material and methods: We will present a 16 years old girl who was admitted with complain of flank pain and diagnosed as giant hydronephrosis required surgery.
Results: A 16-year-old girl, who was attended to another center with flank pain, was admitted our hospital with seriously increased right kidney size measuring 220 mm in diameter with thinning of the renal parenchyma and nephrolithiasis. Computerized abdominal tomography was remarkable for UPJO with a lesion in a multicystic structure extending to the pelvis grim in the size of 25x13x15 cm. Her biochemistry was normal in terms of kidney functions. Markedly decreased renal uptake in right kidney (% 20) was detected by Tc-99 m. Pyeloplasty was done. Then, her flank pain was gradually decreased.
Conclusions: Giant hydronephrosis may be detected at an advanced age with nonspecific complaints such as flank pain. In general, nephrectomy is performed if there is non-functioning kidney. Our patient underwent pyeloplasty because of having functioning kidney. Giant hydronephrosis should be kept in mind in children having chronic flank pain even in teenagers.

P-096 WHAT MIGHT HAPPEN TO CHILDREN WITH NEUROGENIC BLADDER DYSFUNCTION IF THEIR PARENTS NEGLECT NEPHRO-UROLOGICAL FOLLOW-UP?

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Introduction: Myelomeningocele is a congenital defect leading to serious sequels for various organs and systems and therefore medical care with participation of various specialists is necessary.
Aim: Descriptive clinical study of children whose parents neglected nephro-urological follow-up. This illustrative presentation aims to emphasize the role of pediatric nephrologist and urologist in a long term care of dysraphic patients.
Material and methods: From the clinical database of myelomeningocele the patients who had ceased their regular follow-up and in whom serious urological complication occurred were identified. Their medical files were analyzed in details including initial urological management and the results of diagnostic imaging and functional studies.
Results: Seven children (3 girls, 4 boys) with neurogenic bladder dysfunction were included into this study. They had been lost for follow-up review for 4–13 years. During this period the parents / guardians had not been following the principles of nephrological /urological management. Two of them were re-admitted on emergency basis due to ventriculoperitoneal shunt dysfunction. One girl was admitted to intensive care station with sudden incidence of cardiorespiratory insufficiency in the course of undetected chronic kidney failure. Two children were referred by a local pediatricians because of significant kidney dilatation detected on USG scan and recurrent urinary tract infection. The last two patients neglected the regular medical care following

bladder augmentation. In both of them large bladder calculi were found. In all but two patients a marked deterioration of kidney function was noted.
Conclusions: All parents / guardians, family doctors and pediatricians should be aware that regular and long term nephro-urological follow up is necessary in children with neurogenic bladder dysfunction, even when urinary symptoms are absent or vague.

P-097 CONGENITAL VESICOVAGINAL FISTULA IN 8-MONTHS OLD BABY OF A DIABETIC MOTHER- CLINICAL CASE REPORT

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Introduction: Vesicovaginal fistula in childhood usually occurs following penetrating trauma, foreign bodies and genitourinary surgery. However, a congenital vesicovaginal fistula is a very rare entity.
Material and methods: Herein we report a 8-months old girl, born from second pregnancy of a diabetic mother after Cesarean section. After the delivery multiple skeletal, facial and renal anomalies were found plus hydrocephalia. Because recurrent febrile urinary tract infections (UTI) from her first month of age she was referred for further investigations.There was no history of surgical procedures, foreign body or trauma in the genitourinary tract which might have created a vesicovaginal fistula.
Results: From the physical status she was with epicant, short palpebral fissures, low set protrudent dysplastic ears, horizontally placed proximal flank of the lower limb, short lower legs (rhiso-and mesomelia) and club feet. She was with signs of UTI. From the US- hydronephrosis with hydrocalycosis of the left kidney. Excretory urography showed a hydronephrotic left kidney and contrast material drained into the vagina. Cystography revealed a vesicovaginal fistula without vesico-ureteral reflux. Gynecologyst found out inperforated hymen, with normal uterus and filled with fluid vagina. MLPA scrinning for microdeletion syndromes, subtelomer deletions and duplications was negative. Now she is on antibiotic prophylaxis and waiting for surgical procedure.
Conclusions: Vesicovaginal fistula is a rare congenital anomaly in childhood. In our case we report a baby of a diabetic mother, which does not have any laboratory findings of diabetes, admitted after reccurent febrile urinary tract infections. Early diagnostis and immediate surgical treatment could prevent the further renal damage.

P-098 CATHETER-ASSOCIATED BACTERIURIA IN CHILDREN WITH NEUROGENIC BLADDER DUE TO SPINAL DYSRAPHISM

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Introduction: One of the most efficient methods of preventing renal damage due to neuromuscular dysfunction of bladder in patients with spinal dysraphism is a clean intermittent catheterisation (CIC). However, this treatment causes the emergence of catheter-associated bacteriuria (CAB) and / or urinary tract infection (UTI) that both can be called as a catheter-associated urinary tract infection (Catheter-associated Urinary Tract Infections / CAUTI).
Material and methods: All patients who were admitted to Zhytomyr Regional Children's Clinical Hospital with the neurogenic bladder and whom the CIC was administered, were subjected to the mandatory microbiological examination of urine. In case of therapeutic failure of the conservative treatment and the progressive renal damage, surgical treatment has been employed.
Results: The most frequent etiological agents of the CAUTIs were the following microorganisms: *Escherichia coli, Enterococcus faecalis, Pseudomonas aeruginosa, Klebsiella pneumonia, Proteus spp.* and others. It was found that the percentage of the UTIs caused by *P. mirabilis, P. vulgaris* and *Enterococcus faecalis* tripled for the last three years, and the infections caused by *E. coli* increased by 30% as well in patients with the CIC. In contrast, the growth of *Kl. pneumoniae* decreased by 2.5 times and the *Ps. aeruginosa* growth decreased by 30% as the cause of the UTI in the patients who underwent surgical treatment.
Conclusions: The adequate choice of urinary diversion in paediatric patients with neuromuscular dysfunction of the bladder due to spinal dysraphism can reduce the bacterial contamination of urine and prevent the irreversible renal damage.

P-099 THE ASSOCIATION BETWEEN VITAMIN D STATUS, ANTIMICROBIAL PEPTIDES AND URINARY TRACT INFECTION IN SMALL CHILDREN

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Introduction: Vitamin D stimulates production of cathelicidin and β-defensin-2, endogenous antimicrobial peptides, expressed in the urinary tract. Both peptides are active against most common uropathogens. We sought to study vitamin D, cathelicidin and β-defensin-2 levels in children with urinary tract infection (UTI) and healthy controls.
Material and methods: The study is a cross-sectional study of 77 children under 2 years of age with UTI, and of a control group of 46 healthy children. Serum vitamin D (25-OH cholecalciferol) levels were measured by direct competitive electro-chemiluminescence immunoassay (ECLIA), and plasma cathelicidin and β-defensin-2 concentrations were analyzed by ELISA. Samples were taken two months after the UTI in the study group and during a random hospital visit in the control group.
Results: The mean ± SD serum vitamin D level in the UTI group was 80.8 ± 21.2 nmol/l *vs.* 101.1 ± 33 nmol/l in the control group, which is a significant difference ($p = 0.0003$). Children with UTI also had significantly lower plasma cathelicidin and β-defensin-2 levels (medians 33.6 ng/ml and 208.1 pg/ml, respectively) as compared with healthy children (medians 53.8 ng/ml and 394.0 pg/ml, respectively, $p < 0.0001$ for both cathelicidin and β-defensin-2).
Conclusions: Low serum vitamin D levels and low levels of antimicrobial peptides cathelicidin and β-defensin-2 in plasma are significantly associated with urinary tract infection in small children. Vitamin D deficiency may prove to be a risk factor for urinary tract infection. Moreover, supplementation with vitamin D may become a new prophylactic strategy for recurrent UTIs.

P-100 DEMOGRAPHIC AND CLINICAL CHARACTERISTICS OF PATIENTS WITH AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE: A SINGLE CENTER EXPERIENCE

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Introduction: Autosomal dominant polycystic kidney disease (ADPKD) is the most common hereditary kidney disease, with a prevalance of 1:500 to

1/1000. ADPKD is genetically heterogeneous: the genes involved are PKD1 and PKD2.The aim of this study was to investigate patients with ADPKD.
Material and methods: We evaluated every child with ADPKD diagnosed between October 2002 and January 2017. The diagnosis was based on familiy history and ultrasound confirmation of cysts. The investigated demographic and clinical characteristics were gender, age at diagnosis, mode of presentation, parental inheritance pattern, renal function, and the existence of hypertension, hematuria, proteinuria, and pyuria. Follow up of patients were also noted.
Results: A total of 61 patients with ADPKD were analyzed; 36(59%) patients were female and 25(41%) were male. The mean age at onset and admission(± standard deviation) of the patients was 96.98 ± 53.60 (range: 0–176, median: 102.5) and 108.77 ± 53.07(range: 3–187, median:113) months respectively. The mean follow-up time was 24.59 ± 28.59(range: 3–153, median: 14) months. Nine patients(15%) were diagnosed in the first year of life. Family history of ADPKD was known at presentation in all patients. The most common presentations leading to diagnosis of ADPKD were positive family history for 24 patients(39%), abdomainal pain or mass for 15 patients(25%), other causes for 12 patients(20%) and antenatal detection of cysts with USG for 4 patients(7%). Bilateral renal findings were present in 43(70%), HT in 6 patients(10%). Ninety-eight percent of the patients had normal renal function. Only one patient was followed as chronic kidney disease. Thirteen patients(21%) had persistent proteinuria, 10(16%) had pyuria, and 2(3%) had microscopic hematuria. One patient had hepatic cysts, and none of them had splenic or pancreatic cysts.
Conclusions: The results of the present study are better than most other series. The majority of children with ADPKD in this study were found to have a good prognosis.

P-101 CONGENITAL ANOMALIES OF THE KIDNEY AND URINARY TRACT (CAKUT) IN THE ETIOLOGICAL STRUCTURE CHRONIC KIDNEY DISEASE (CKD) IN CHILDREN AND ADOLESCENTS

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Introduction: To study the frequency CAKUT in the etiological structure CKD by children and adolescents.
Material and methods: This study includes 80 children with CKD, 34 children of them had CAKUT in age 9.4 ± 5.4 years. Out of 34 children - 22 boys (64.4%) and 12 girls (35.6%).
Results: It is established that in the etiological structure of CKD in 80 children and adolescents is dominated by congenital and hereditary kidney diseases in 75% cases. CAKUT was diagnosed in 24 (70.5%) patients in perinatal and infantile period. From 34 children with CAKUT: 18 patients (53%) had vesicoureteral reflux (VUR) (single or double direction) stage IV- V, 1 patient (3%) had bladder extrophy, 7 patients (20.5%) had hydronephrosis stage III-IV, 8 patients (23.5%) had reflux-nephropathy. Patients with VUR had posteror urethral valve, bladder dysfunction, ureterohydronephrosis. Reflux-nephropathy stage C, D is manifested by proteinuria, hypertension, renal scarring. Urinary tract infectionhad 26 (76.4%) patients. Children with CAKUT had eGFR 28.6 ± 22.3 ml/ min on 1.73 m^2. Out of 34 children with CKD was diagnosted stage V in 11 (32.4%) patients, stage IV in 13 (38.2%) patients, stage III in 3 (14.7%) patients, stage II in 4 (11.5%) patients, stage I in 1 (2.9%) patients.
Conclusions: It is established that in the etiological structure of in 80 children and adolescents with CKD dominated by congenital and hereditary kidney diseases (75%), among which prevails CAKUT (56.5%).

P-102 CORRELATION OF ULTRASONOGRAPHICAL FINDINGS OF HYDRONEPHROSIS/ATROPHY WITH 99MTC-DMSA IN CHILDHOOD: A SINGLE CENTRE EXPERIENCE FROM TURKEY

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Introduction: We aimed to assess the association of abnormalities (hydronephrosis and/or atrophy) detected on renal ultrasound and DMSA scan with the presence of vesicoureteric reflux on micturiting cystourethrography(MCU) to find out new perspectives.
Material and methods: We retrospectively reviewed the DMSA findings and medical records of pediatric patients with hydronephrosis and/or atrophy who were at follow-up between January 2013 and December 2016 in our center which is located in the south-east region of Turkey.
Results: Among 148 pediatric patients (M/F = 60/88), 66 had hydronephosis, 72 had atrophy, and 10 patients had both. The mean age of the children was 56.7 ± 6.1 months (range 3–194 months). MCUG study detected VUR in 66 patients. Patients with atrophy were significantly older than patients with hydronephrosis (77.8 ± 58.6 vs 39.3 ± 38.9 months, $p = 0.002$). Only 19.4% of the our patients with atrophy had VUR. The rate of VUR was higher in the high-grade group than the mild-to-moderate grade group, although the difference was not statistically significant (80% vs 61%, $p = 0.199$). Patients wiht high grade hydronephrosis had more severe DMSA findings (73% vs 39%). On the other side, 79% of the patients with high grade VUR had severe DMSA findings. A total of 10 patients had both atropy and hydronephrosis all affecting the left side. Six of them had VUR. Severe DMSA findings were more likely in toddlers (age 24–72 months) (48%).This finding was abruptly lowered after 72 months of age.
Conclusions: Ultrasonographic findings of atrophy should also be evaluated for vascular renal pathologies in additon to VUR evaluation. In cases of left side renal pathologies of hydronephrosis and or atrophy the patients should closely monitored and furter evaluated even there is no urinary tract infection. Lastly, DMSA may not be necessary in cases with high grade hydronephrosis before MCU.

P-103 ONE-YEAR FOLLOW-UP RESULTS OF INFANTS WITH ANTENATAL HYDRONEPHROSIS

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Introduction: The infants who had diagnosed antenatal hydronephrosis are followed up by pediatric nephrology department according to the Turkish Pediatric Nephrology Society CAKUT (Congenital Anomalies of Kidney and Urinary Tract) Guidelines. The guideline is summarized as Figure 1.
Material and methods:
Results: Ninety-three infants (64 males,0.83 years, range 0.1–2.1 years) were followed up. According to 3-4th days and 4-6th week ultrasonography results 5 (5.4%) patients were normal, 70 (75.3%) patients were mild, 14 (15.1%) patients were moderate and 4(4.3%) patients were severe hydronephrosis. Voiding cystourethrogram (VCUG) was performed in 10 (10.7%) patients, 2 of them had unilateral grade 4 vesico-ureteric reflux (VUR). MAG3 scan was performed in 8 (8.6%) patients, 2 of them had ureteropelvic junction obstruction, 1 of them had pyeloplasty. After one year follow-up of 88 infants, 47(53.4%) patients were normal, 23(26.1%) patients were mild, 11(12.5%) patients were moderate and 7(8%) patients were severe hydronephrosis. Three cases (3.4%) progressed from moderate to severe. Six patients who had performed both VCUG and MAG3 have no pathologic finding and have no clinic disease such as urinary tract infection or failure to thrive.
Conclusions: Regular follow-up of antenatal hydronephrosis cases; while preventing unnecessary invasive testing of transient hydronephrosis, provides early intervention in cases of high risk for the urinary system anomaly.

P-104 PROPER VOIDING OBSERVATION WE CAN EXCLUDE A LOT OF EXPENSIVE AND TIME CONSUMING EXAMINATIONS

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Introduction: In children with voiding dysfunction improper bladder emptying contributes to recurrent urinary tract infections and progressive renal scarring.
Material and methods: Case presentation of a adolescent girl with Recurrent urinary tract infections. On examination: dysraphismus with spinal nevus. No walking or other neurological deficits.
Ultrasound: kidneys normal, bladder large and large amount of residuals, which can't be emptied after double voiding. Urine: pathologic usually, and urine pH is 7.0, even without UTI; Labs: normal values.
Results: Uroflow: On presentation: intermittent curve, >800 ml urine, >200 ml residuals, voiding time > 40s, max flow 30 ml/s;.
Lumbosacral MRI normal.
Cystometry: increased compliance 655 ml, no involuntary contractions, stress test negative. Voiding phase: low bladder contractions, max flow 14 ml, intermittent curve, and with residuals 50 ml.
Voiding Observation: first observations were not done properly, what made us to go further with examinations. And after the examinations we did observation again at the clinic and resulted: voiding around 4 L of urine and drinks large amount > 5 L. Voiding frequency was normal, no wetting incidents. By thirst and dessmopresin test, which resulted good kidney concentration ability, we excluded insipid diabetes. Conclusion was: Polyuria after polidypsia.
We recommended limiting liquid intake and regular bladder emptying every 4–6 h, with double voiding and with regular bowel regime. And the results were good so far, no infections, urine pH <6, and no residuals after 1 year fellow up. Bladder wall was thinner, also.
Conclusions: Increased residual urine on post-void ultrasound increases the risk of UTI recurrence in children with voiding dysfunction. This residual can be as a consequence of increased intake, also. Because of increased voiding frequency needs attempts to avoid voiding voluntary. This continuously increase bladder capacity.
Thus, voiding and defecation history, physical examination, voiding frequency charts and defecation diaries are all essential parts of urodynamics and urologic examination.

P-105 SINGLE-POINT IOHEXOL PLASMA CLEARANCE IN CHILDREN: VALIDATION OF MULTIPLE FORMULAS AND SAMPLING TIMES

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Introduction: The non-ionic agent iohexol is increasingly seen as the marker of choice for glomerular filtration rate (GFR) measurement. Since estimated GFR (eGFR) has low accuracy in children and limitation of number of blood draws to a minimum is especially relevant in children, we performed a study to evaluate different methods and find the optimal sampling point for calculating measured GFR (mGFR) based on iohexol clearance with a blood sample drawn at only one time-point (GFR1p).
Material and methods: 96 children with chronic kidney disease (CKD), median age 9.2 years, range 3 months to 17.5 years, were examined using iohexol plasma clearance and blood sampling at seven time points within five hours (GFR7p) as the reference method. Median GFR7p was 65.9 mL/min/1.73 m2, range 6.3–153 mL/min/1.73 m2. The performances of six different formulas (Fleming-2005, Ham-1991a, Ham-1991b, Stake-1991, Groth-1984, Jacobsson-1983) were validated against the reference.
Results: GFR1p calculated according to the formula of Fleming with sampling at 3 h, had the best performance with 80% within +/−10% of the reference (P10). The Fleming formula gave GFR1p with significantly better P10 ($p < 0.05$) than all other tested formulas with sampling at 2 h, 3 h and 3.5 h. With sampling at 4 h both the Fleming formula and the Jacobsen formula performed significantly better than the other formulas with P10 of 73 and 70%, respectively. At 5 h the formula of Jacobsen had the best performance with P10 of 74%, significantly better than all other tested formulas.
Conclusions: The Fleming formula with sampling at 3 h is recommended when GFR1p is used in children with CKD.

P-106 AUTOMATED HEIGHT INDEPENDANT ESTIMATED GFR REPORTING LEADS TO EARLIER DETECTION OF CHILDREN WITH CKD

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Introduction: Automated estimated glomerular filtration rate(eGFR) reporting is established adult practice and improves detection of chronic kidney disease (CKD). eGFR formula in children commonly require height which is not routinely available when measuring creatinine. We have validated a height independent eGFR formula (Table 1) and report changes in referrals before and after introduction of automated reporting.

Table 1; Assessment of our formulua and comparison to other height independent eGFR formulal

	Mean difference	St Dev	R (Correlation)	% Diff <20%	% Diff <30%
Pottel	27.1	45.8	0.62	47%	62%
Lund-Malmo (Revised)	18.7	23.9	0.57	60%	75%
NCH-BCCH2	14.6	20.0	0.71	71%	83%
EPI-CKD	Previously reported adult formula	83%			
MDRD	Previously reported adult formula	83%			

Material and methods: We reviewed our new referrals for 6 months prior to introduction of automated eGFR reporting in children aged 2 to 17 years of age. The case notes of all patients referred because of a reduced estimated GFR were reviewed and the outcome determined.
Results: In the 6 months prior to reporting no patients were referred beacuse of a reduced estimated GFR.
In the 6 months following reporting 9 patients were referred with an eGFR less than 90 ml/min per 1.73m^2. Three patients had an ultrasound which showed a small kidney, one of whom also had proteinuria. The remaining 6 patients had no proteinuria, no hypertension and normal ultrasound when done. All patients with abnormal ultrasounds had an eGFR <80 ml/min per 1.73m^2.
Conclusions: Automated reporting of estimated GFR in children is feasible and increases early detection of children with CKD. If the estimated GFR is greater than 80 ml/min per 1.73m^2 and there are no other risk factors for CKD then ultrasound is not required.

P-107 EARLY PROTEINURIA LOWERING BY ACE INHIBITION IS PREDICTIVE OF RENAL SURVIVAL IN CHILDREN WITH CHRONIC KIDNEY DISEASE

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Introduction: The degree of proteinuria predicts progression of renal failure in adults and children with chronic kidney disease (CKD). While lowering of proteinuria by various interventions has been demonstrated to be nephroprotective in adults with proteinuric nephropathies, pediatric data on the relationship of pharmacotherapeutic proteinuria lowering and long-term renal survival is scarce. Here we have revisited the ESCAPE Trial to investigate a potential quantitative association of the initial antiproteinuric effect of standardized ACE inhibition with subsequent renal disease progression in children with CKD.
Material and methods: All children were started on a fixed dose of ramipril (6 mg/m^2/day) and randomized to aim for conventional or intensified blood pressure control. The initial log-transformed change in proteinuria was assessed from baseline to first measurement after starting ramipril (at 2.6 ± 1.4 months). Cox proportional hazard models were used to estimate the association between initial proteinuria change and risk of reaching the renal endpoint (composite of 50% decline in eGFR or progression to end-stage-renal disease), adjusted for age, gender, CKD diagnosis, baseline proteinuria, blood pressure, eGFR and change in blood pressure.
Results: Of 285 eligible patients (59% male, age 11.5–3.9 years), 85 reached the endpoint within 5 years of follow-up. Proteinuria was reduced following start of ramipril treatment by a median of 40% (interquartile range 8–64%). As compared to a reduction in proteinuria of less than 30%, a 30–60% reduction accounted for a HR of 0.65 (CI 0.38–1.12) and reduction of more than 60% gave a HR of 0.42 (0.22–0.79). This association was independent of eGFR (HR 0.93, CI 0.91–0.94), proteinuria (HR 1.24, CI 1.13–1.38) and blood pressure (HR 1.21, CI 1.03–1.42).
Conclusions: The early antiproteinuric effect of ACE inhibition is independently predictive for long-term preservation of renal function in children with CKD. This finding suggests that proteinuria lowering is an important target in the management of pediatric CKD.

P-108 CHANGES IN URINARY ANGIOTENSINOGEN AFTER TREATMENT WITH RENIN-ANGIOTENSIN SYSTEM BLOCKADE IN PEDIATRIC CHRONIC KIDNEY DISEASE PATIENTS WITH LOW BIRTH WEIGHT

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Introduction: Children born with low birth weight (LBW) have higher risk of developing chronic kidney disease (CKD) because of a low number of nephrons. Some develop proteinuria and decreased renal function as early as childhood. However, no effective therapy has been established to suppress the progression of CKD in children with LBW. Renin-angiotensin system (RAS) activation plays a critical role in the development of hypertension and CKD, and our previous work demonstrated that urinary angiotensinogen (uAGT) is a useful marker of intrarenal RAS activation. The aim of this study was to assess whether treatment with RAS blockade is beneficial for suppressing the progression of CKD in children with LBW using uAGT as a surrogate marker.
Material and methods: We recruited 11 children with LBW who were started on RAS blockade with Candesartan between April 2013 and August 2016 to treat hypertension or proteinuria. The mean birth weight was 797.8 ± 282.6 g, and the mean age at evaluation was 13.8 ± 3.5 years. We compared uAGT, blood pressure, urinary protein, serum electrolytes, and renal function before and after treatment with Candesartan.
Results: After treatment with Candesartan for 20.7 ± 13.0 months, the uAGT to urinary creatinine ratio was significantly decreased (56.9 ± 41.9 vs. 8.2 ± 5.5 μg/g, p = 0.003). Urinary protein was also significantly decreased (p = 0.003), although there were no significant changes in blood pressure, serum sodium and potassium, or estimated glomerular filtration rate based on serum creatinine.
Conclusions: These data indicate that treatment with Candesartan suppresses the activation of the intrarenal RAS and reduces proteinuria, which may slow the progression of CKD in children with LBW. Further studies with a larger number of patients and longer observation of renal function are required.

P-109 FIVE YEAR RENAL OUTCOME IN PATIENTS WITH POSTERIOR URETHRAL VALVES

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Introduction: Posterior urethral valves (PUV) are a congenital anomaly causing obstructive uropathy. We describe our cohort, their presentation and the renal status at 5 year of age.
Material and methods: We retrospectively reviewed the medical notes of all boys with PUV born from 2005 to 2011 and followed-up at our institution. We collected data on presentation and renal function at age 5 years. Values are shown as median (range). T test and Chi Square tests were used for statistical analysis.
Results: 94 patients were identified, 44(46.8%) had an antenatal diagnosis. Those diagnosed postnatally had age at presentation 2 (0–106) months. Twelve children presented at age > 2 years. The most common postnatal presentation was sepsis and renal impairment in neonates and recurrent urinary tract infection in older children. Serum Creatinine at presentation was 140umol/L (20–515) in the prenatal group and 55 umol/L (20–557) in the postnatal (p = 0.0034). Two patients (with multiple associated pathologies) died of reasons not related to PUV, at 1 and 3 years, respectively. The first patient died of severe sepsis and acute cerebral ischemia and the second patient of central nervous system vasculitis on the background of progressive steno-occlusive cerebrovascular disease. Estimated GFR at age 5 years was available for 75 patients. Before the age of 5 years, six patients had renal transplant and 1 was started on peritoneal dialysis. Renal function was worse

in the group with prenatal diagnosis ($p = 0.056$). Table shows the patients divided per their CKD stage I-V and V + RRT (Renal Replacement Therapy).

CKD	ANTENATAL $N = 32$	POST NATAL $N = 50$	TOTAL $N = 82$
I	8 (25%)	17(34%)	25(30%)
II	10(31%)	18(36%)	28(34%)
III	5(16%)	8(16%)	13(16%)
IV	3(9%)	2(4%)	5(6%)
V	2 (6%)	2(4%)	4(5%)
V + RRT	4(12.5%)	3(6%)	7 (8.5%)

Conclusions: In our cohort, the majority had normal or mildly reduced renal function at age of 5 years with worse renal function in those diagnosed prenatally perhaps reflecting more evident disease prenatally.

P-110 APOL1 RISK VARIANTS GENOTYPING AND THE ASSOCIATION WITH THE EARLY KIDNEY DAMAGE IN CHILDREN FROM THE DEMOCRATIC REPUBLIC OF CONGO (DRC)

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Introduction: There are evolving epidemiological and biological data to support an association between the gene encoding apolipoprotein-L1 (APOL1) and progressive chronic kidney disease (CKD) among African-Americans. In Africa, data related to the geographical distribution of *APOL1* genetic risk variants G1 and G2 are limited, and there is no reliable data from Democratic Republic of Congo (DRC). We aimed to determine the frequencies of *APOL1* risk variants in a large population from Central Africa and to assess the association with the early kidney damage in children.
Material and methods: A total of 465 participants from four large districts in Kinshasa were enrolled. *APOL1* high-risk genotype was defined by the presence of 2 high-risk variants (G1/G1, G2/G2, G1/G2) and low risk genotype if 0 or 1 risk variants were present. Albumin-to-creatinine ratio (ACR) was assessed in a fresh morning urine sample in children only, and elevated ACR was defined as ACR > 30 mg/g.
Results: From 465 subjects enrolled, 453 were successfully genotyped, of whom 388 children and 65 adults. *APOL1* sequence analysis revealed 201 (44%) participants carrying at least one *APOL1* risk variant, 36 (8%) 2 risk variants. Concerning the frequency of *APOL1* risk alleles, 14% of all chromosomes carried G1 whilst 13% carried G2. Thus, the burden of *APOL1* risk allele was 27%. Of 388 children, 39 (10%) had elevated ACR. Compared to those carrying low-risk genotype, children with *APOL1* high-risk genotype had a higher prevalence of microalbuminuria (OR 1.48, 95% CI 0.41–4.84).
Conclusions: *APOL1* risk variants are common in DRC. However, the high-risk genotype did not show a strong statistical association with the early kidney damage in a general population of children.

P-111 GFR-ESTIMATION BY SERUM CREATININE DURING GLUCOCORTICOSTEROID THERAPY

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Introduction: While glucocorticosteroids (GCS) are widely used in patients with kidney disease, little is known about their effect on serum creatinine, the most commonly used endogenous marker of kidney function.
Material and methods: We assessed the effect of GCS on the relationship between estimated GFR using the Schwartz equation (eGFR) and measured GFR using a single injection inulin clearance (Cin) in children both in a paired analysis and a cross-sectional study. Primary outcome parameter was the difference between eGFR and Cin (DGFR). Paired analysis was done in 22 patients during and without GCS treatment (median GFR 114 ml/min/1.73 m2, median prednisone dose 35.5 mg/m2/d). In a cross sectional analysis in 50 patients, 31 of which received GCS (median dose of 12 mg/m2/d) a dose-dependent effect was explored using univariate linear regression of various variables including GCS dosage, with DGFR as dependent variable.
Results: The paired analysis showed no significant difference in DGFR with or without GCS (−23 [SD 53] vs. -9 [SD 41] ml/min/1.73 m2, $p = 0.203$). Linear regression analysis showed a significant correlation between age and DGFR, while GCS dose was not related to DGFR.
Conclusions: GCS use does not hamper the use of creatinine as a marker for kidney function.

P-112 SLEEP DISORDERS IN CHILDREN WITH CHRONIC KIDNEY DISEASE

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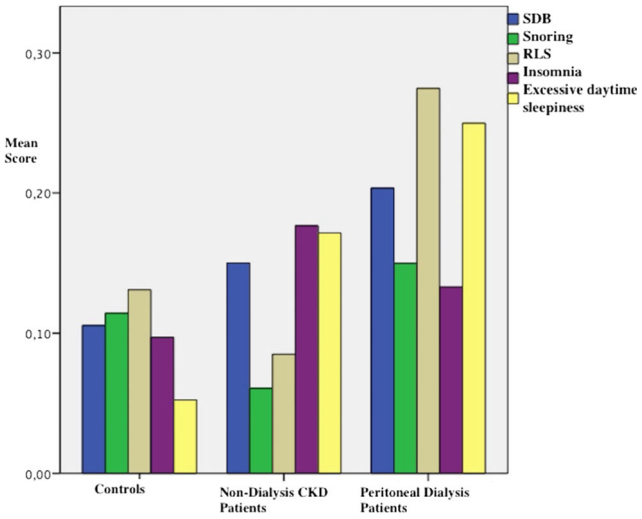
Introduction: Chronic Kidney Disease (CKD) in children has been associated with sleep disorders. However, only a few papers have been published on this is subject in pediatric patients. The aim of present study was assess the presence of sleep disorders in pediatric CKD patients.
Material and methods: The present study evaluated children and adolescents with CKD and healthy children and adolescents aged 5–19 years. Parents completed the Pediatric Sleep Questionnaire from November 2015 to October2016. Scores greater than 0.33 in the subscales of sleep disorders were considered to be diagnostic.
Results: The study population included 46 pediatric patients with CKD (10 dialysis depedent, all in peritoneal dialysis) and 68 controls. Children with CKD had higher scores in the subscale of sleep disordered breathing (SDB), but the incidence of SDB was not statistically different to that of the control group. Pediatric patients with CKD had higher frequency of insomnia (20% vs. 3% controls, $P < 0.01$), excessive daytime sleepiness (26.7% vs. 1.5% in controls, $P < 0.001$) and nocturnal awakenings (21.4% vs. 1.5% in controls, $P < 0.001$). Among patients with CKD, dialysis dependent patients presented higher scores in restless legs syndrome scale compared to patients with CKD non-dialysis dependent ($P < 0.05$), but not in other scales' scores (Figure 1).



Conclusions: Children with CKD have sleep disorders more frequently than healthy children. Therefore, clinicians caring for these children should always bear this in mind, in order to improve quality of medical care.

P-113 METASTATIC PULMONARY CALCIFICATION (MPC) IN A CHILD WITH CHRONIC KIDNEY DISEASE (CKD) IN RENAL REPLACEMENT THERAPY

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Introduction: Different pulmonary or systemic conditions are associated with pulmonary calcifications, also described as calcium salt storages in lungs. A well accepted classification distinguishes metastatic calcifications where calcium storage occurs in a previously normal lung, from dystrophic calcifications, in which calcium is accumulated in a damaged lung. Though MPC is a quite rare condition, adult patients with CKD in hemodialysis (HD) often show this pulmonary pattern due to chronic calcium-phosphate imbalance.

Material and methods: We report a case of a 12-year-old female with previous history of CKD in chronic PD treatment for 7 years and then in chronic HD due to peritoneal membrane failure. She was admitted to our Pediatric Nephrology Unit for urgent kidney transplantation without any symptoms. Routine preliminary investigations revealed a remarkable calcium-phosphate imbalance with a high level of intact parathyroid hormone (1326 ng/L) and high levels of calcium and phosphate as seen in secondary hyperparathyroidsm. The inflammatory markers were not meaningful.

Results: The chest X-ray showed an unexpected right upper lobe opacity. Previous Chest X-rays were negative. Because of the suspect of an infection, transplantation was not performed and investigations were started. Pulmonary infectious disease, arteriovenous malformations and granulomatous disorders were ruled out. To clarify the clinical picture a CT pulmonary angiogram was performed. During the precontrastography stage, an extensive calcification at the upper right lobe was revealed. The final diagnose was made after lung needle-biopsy, which demonstrated a limphomonocitary inflammation with diffuse alveolar calcium stones, referable to a pulmonary lithiasis.

Conclusions: Although MPC is reported in pediatric patients with CKD, this is a rare complication, especially in the last years with the global improvement in CKD-MBD therapies. However, MCP must be considered in the differential diagnosis of unexplained lung opacities in children with CKD, mainly in the presence of secondary hyperparathyroidsm and strong clinical-radiological dissociation.

P-114 EFFECT OF HYPERPARATHYROIDISM ON RESPONSE TO ERYTHROPOIETIN IN CHILDREN WITH CRF

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Introduction: The response to recombinant human erythropoietin (rHuEPO),50 unit/kg twice weekly was studied prospectively in 35 children and adolescents with end- stage renal failure who were either transfusion dependent or had hematocrits(HCT) <25%.

Material and methods: rHuEPO was given to 22 haemodialysis (HD) patients and 13 patients on conservative treatment,with mean age (10.84 ± 4.08) years,25 males and 10 females with mean HCT(26.75 ± 4.70) Blood pressure, haematocrit, iron-indices, serum potassium, calcium,phosphorus, alkaline phosphatase,urea nitrogen and intact parathyroid hormone (iPTH) were monitored serially.

Results: Serum aluminum was measured randomly in 6 patients, results were normal ranging from 12 to 22 μg/l.When serum ferritin was <100 ng/ml during therapy,they received iron supplementation.According to the response, patients were divided into 2 groups,the non –responders group with HCT < 27,mean age (9.97 ± 3.55) years, with mean iPTH (669.9 ± 461.77) pg/ml and group of responders with HCT > 27 with mean age (11.66 ± 4.32) years, with mean iPTH (261.19 ± 233.17) pg/ml,15 HD patients never reached target HCT at this dose versus 2 patients on conservative treatment; By comparison between both groups as regards laboratory values, it shows reticulocytic count,iPTH and serum ferritin were significantly higher in the non- responders(NR) versus the responders(R) with p values ($p = 0.04$,$p = 0.006$,$p = 0.04$) respectively,while serum calcium,albumin were significantly lower between NR versus R with p-values ($p = 0.007$,$p = 0.003$) respectively.Inspite that iron, TIBC,% transferin saturation and KT/V shows non significant difference between NR and R group, also caloric intake as % of recommended daily allowance shows non significant difference between both groups.However intact parathormone levels were significantly higher before and after 16 weeks of erythropoetin (EPO) therapy in the NR versus R groups, $P < 0.05$ versus $p = 0/006$ respectively.

Conclusions: So we conclude that in the absence of other well-known response–limiting factors, the erythropoetic response to erythropoetin therapy depends largely on the extent of secondary hyperparathyroidism.

P-115 CARDIAC FGF23 IS STIMULATED BY RAAS ACTIVATION AND INDUCES A PRO-FIBROTIC CROSSTALK BETWEEN CARDIAC MYOCYTES AND FIBROBLASTS

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Introduction: FGF23 is discussed as a new biomarker associated with cardiac hypertrophy and mortality in patients with CKD, heart failure, and cardiogenic shock that promotes diastolic dysfunction, congestive heart failure, arrhythmia, and sudden death.Since we previously demonstrated

that FGF23 is expressed by cardiac myocytes, enhanced in CKD, and causes cardiac hypertrophy via activation of FGFR4/calcineurin/NFAT signaling, we aimed to investigate whether induction of cardiac FGF23 associates with myocardial fibrosis in uremia and directly promotes pro-fibrotic crosstalk of cardiac myocytes and fibroblasts *in vitro*.
Material and methods: We conducted a retrospective case-control study including 24 myocardial autopsy samples from CKD patients and investigated cardiac fibrosis by histological quantification of fibrillar collagens and fibrosis RT^2 profiler PCR array analyzes. Data were correlated with clinical parameters, cardiac FGF23, and klotho levels. The specific impact of FGF23-mediated induction of cardiac fibrosis was further evaluated in isolated cardiac fibroblasts and myocytes.
Results: Accumulation of fibrillar collagens was increased in myocardial tissue of CKD patients and correlated with duration of dialysis, klotho deficiency, and enhanced angiotensinogen expression. TGF-β and its related TGF-β receptor/Smad complexes, extracellular matric remodeling enzymes, as well as pro-fibrotic growth factors were significantly upregulated in myocardial tissue of dialysis patients. In cultured cardiac fibroblasts, FGF23 stimulated pro-fibrotic TGF-β receptor/Smad complexes and collagen synthesis, whereas treatment of isolated cardiac myocytes with FGF23 resulted in enhanced collagen remodeling, induction of pro-survival pathways, pro-inflammatory and pro-hypertrophic genes. Angiotensin II and aldosterone, as components of the renin-angiotensin-aldosterone system (RAAS), strongly induced FGF23 in cardiac myocytes and FGF23 further stimulated angiotensinogen expression in both cardiac cell types.
Conclusions: Cardiac FGF23 is stimulated by RAAS components in cardiac myocytes to directly promote fibrotic and hypertrophic response. Secreted by cardiac myocytes, FGF23 induces pro-fibrotic pathways and collagen synthesis in cardiac fibroblasts in a paracrine fashion, suggesting that FGF23 impact on both pathological cardiac remodeling processes.

P-116 IMPACT OF FGF23 EXCESS AND KLOTHO DEFICIENCY ON CARDIAC REMODELING: LESSONS FROM TWO MOUSE MODELS

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Introduction: Clinical and experimental studies associate FGF23 excess, high phosphate and PTH levels, and deficiency of active vitamin D (1,25D) and klotho with the development of cardiovascular events, e.g. endothelial dysfunction, left ventricular hypertrophy (LVH), and myocardial fibrosis. However, 1,25D and klotho ameliorate myocardial hypertrophy *in vivo*, and klotho suppresses cardiac fibroblast activation and collagen synthesis, and protects against FGF23-mediated oxidative stress *in vitro*.
Material and methods: We investigated the cardiac phenotype in two mouse models of FGF23 excess and klotho deficiency, i) klotho hypomorphic ($Kl^{-/-}$) mice presenting with high plasma levels of phosphate, 1,25D, and FGF23, as well as suppressed PTH, and ii) *Hyp* mice presenting with elevated FGF23 and PTH, but low phosphate and 1,25D levels in parallel with reduced renal klotho expression.
Results: In both mouse models relative heart weight and cross-sectional area of individual cardiac myocytes were larger than in respective wild-type controls. In $Kl^{-/-}$ mice, cardiac Fgf23, Fgfr4 and calcineurin/NFAT signaling as well as pro-hypertrophic genes *Rcan1*, *BNP*, *ANP*, *bMHC* were clearly induced. Investigation of transcription factors and fibrosis-related molecules characteristic for pathological cardiac remodeling processes demonstrated enhanced expression of Cebpb and Gata4, as well as *Tgfb1*, *collagen I*, and *Mmp2* in $Kl^{-/-}$ mice. In contrast, *Hyp* mice showed significantly enhanced cardiac *Fgf23* mRNA levels and high intact cardiac Fgf23 protein, but the induction of Fgfr4/calcineurin/NFAT pathway, *BNP*, *ANP* and *bMHC* expression, and stimulation of pro-fibrotic factors was absent when compared to respective wild-type controls.
Conclusions: Our data suggest that despite of high circulating and cardiac FGF23 levels and enhanced PTH, as well as reduced renal klotho and 1,25D synthesis, *Hyp* mice appear to be protected against the development of cardiac pathology possibly at least partly due to hypophosphatemia. In contrast, $Kl^{-/-}$ mice present with strong induction of cardiac hypertrophy and fibrosis despite high 1,25D plasma levels.

P-117 CHRONIC KIDNEY DISEASE IN NEONATES

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Introduction: Chronic kidney disease (CKD) requiring renal replacement therapy (RRT) is rare in neonates. RRT in neonates is expensive, time consuming and difficult. Very little data about neonatal CKD is available from the developing countries.
Material and methods: All neonates with evidence with CKD from 2005 to 2015 were reviewed. Follow up serum creatinine was recorded every six months.
Results: Total of 181 children presentedwith CKD. Their mean age at presentation was 11.1 days (95% CI 9.5–12.8) and the mean creatinine was 106.5 uml/l (95% CI 91.3–121.7).
Congenital anomalies of the kidneys and urinary tract (CAKUT) was the underlying cause in 84.5%. Mortality was high particularly in the first 6 months (10%) and reached 16% by 4 years of follow up. Younger age at presentation, male sex and hypertension were associated with higher mortality. On the last follow up 42 (41%) children had hypertension and 27 (26.5%) had significant proteinuria. Five children had received dialysis in the neonatal period and another 6 were started dialysis later on.
Conclusions: Management of advanced CKD in neonates is challenging particularly in developing countries.

P-118 PREVALENCE OF FAILURE TO THRIVE IN IRANIAN CHILDREN WITH CHRONIC KIDNEY DISEASE

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Introduction: Malnutrition and inflammation are considered as risk factors of morbidity, hospitalization, and mortality in chronic kidney disease children (CKD). The aim of this study was to find the prevalence and severity of failure to thrive (FTT) in moderate to severe CKD children.
Material and methods: This was a cross sectional study of 84 children (30 Female, 54 male) aged 2–16 years old with CKD from June 2014 to June 2015. The inclusion criteria were eGFR less than 90 ml/min/1.73m^2, being healthy in previous month of visit, having no other chronic diseases except CKD. Anthropometric data including the body mass index, height, weight, mid upper arm circumference were collected. Protein wasting energy was scored and the severity of failure to thrive estimated by Gomez and Jelliffe classifications. *P*-values less than 0.05 were significant.

Results: Glomerulopathy and hereditary /tubulopathy constituted the main causes of underlying disease. About 79% of CKD children had FTT and this rate increased with declining of renal function (p-value < 0.05). Using modified PWE 65.5% identified to have score > =2 and it was more frequent in eGFR less than 30 (P > 0.05). A quarter of patients with FTT classified as no PWE and vice versa.

Table 2- The severity of Failure to Thrive and Protein Wasting Energy in different classes of eGFR

Classifications ml/min/1.73m²		FTT No N=18	FTT Mild N=16	FTT Moderate N=29	FTT Severe N=21	P-value
eGFR	<15	5 (20%)	12(28.6%)	12(28.6%)	13(31%)	0.014
	15-30	4(21%)	3(16%)	10(52.6%)	2(10.5%)	
	30-60	4(25%)	1(6%)	6(38%)	5(31%)	
	>60	5(71)	-	1(14)	1(14)	
Classifications ml/min/1.73m²		PWE No N=29	PWE Minimal N=30	PWE Standard N=8	PWE Modified N=17	P-value
eGFR	<15	14 (33%)	17(40.5%)	2(5%)	9(21%)	NS
	15-30	5(26%)	8(42%)	3(16%)	3(16%)	
	30-60	7(44%)	5(31%)	2(12.5%)	2(12.5%)	
	>60	3(43)	-	1(14)	3(43)	

Conclusions: The majority of children with moderate to severe chronic kidney disease had failure to thrive and protein wasting energy. There was no correlation between inflammatory markers and the severity of CKD or the presence of failure to thrive.

P-119 AN OPEN-LABEL, SINGLE-DOSE STUDY TO EVALUATE THE SAFETY, TOLERABILITY, PHARMACOKINETICS, AND PHARMACODYNAMICS OF CINACALCET IN PEDIATRIC SUBJECTS AGED 28 DAYS TO < 6 YEARS WITH CHRONIC KIDNEY DISEASE RECEIVING DIALYSIS

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Introduction: We evaluated the safety, tolerability, pharmaco kinetics(PK), and pharmacodynamics(PD) of a single dose of cinacalcet in children with secondary hyperparathyroidism(sHPT) receiving dialysis.

Material and methods: Twelve subjects (28 days to <6 years old) received a single 0.25 mg/kg dose of cinacalcet. The dose was administered orally (syringe) or by nasogastric or gastric (NG/G) tube. Subjects were randomized 1:1 to two parathyroid hormone [PTH] and serum calcium sampling sequences post-dose: (1) 2, 8, and 48 h; or (2) 2, 12, and 48 h. Subjects were followed for 72 h after dosing for the study assessments.

Results: Cinacalcet median t_{max} was 1 h (range: 0.50 to 4.0 h). Plasma cinacalcet mean (SD) C_{max} and AUC_{last} were 2.83(1.98)ng/mL and 11.8(8.74)hr.•ng/mL, respectively. The mean (SD) half-life ($t_{1/2,z}$) was 3.70(2.57)hours. Subject age, weight, body surface area, and BMI did not have an effect on the PK of cinacalcet. Reductions in serum PTH concentrations from baseline were observed at 2 h and 8 h post-dose (median: −10.8% and −29.6%, respectively), and returned to near baseline levels at 72 h (−5.4%). These reductions correlated with changes in plasma cinacalcet concentrations. Safety findings were similar to safety profile observed in adults.

Conclusions: A single dose of cinacalcet was well-tolerated in pediatric patients. Cinacalcet was rapidly absorbed and eliminated with no notable effects of age, weight, body surface area, and BMI on cinacalcet PK. Discernable reductions in serum iPTH were observed up to 8 h after a single 0.25 mg/kg dose of cinacalcet and returned to baseline by 72 h. Results from this study indicate that a 0.25 mg/kg dose of cinacalcet is deemed an appropriate safe starting dose in children 28 days to <6 years of age.

P-120 IS NT-PROBNP A RELIABLE MARKER FOR BODY FLUID STATUS IN CHILDREN WITH CHRONIC KIDNEY DISEASE?

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Introduction: The aim of the study was to evaluate the body fluid content with calculations of NT-proBNP and bioimpedance spectroscopy in children undergoing hemodialysis (HD) and peritoneal dialysis (PD), and in children with chronic kidney disease (CKD) with no need of renal replacement therapy, and to investigate the association between cardiovasculary changes and NT-proBNP.

Material and methods: The study included 65 children: 10 predialysis patients with CKD (mean age: 10.50 ± 2.27 years), 13 patients undergoing HD (mean age: 11.92 ± 3.13 years), 12 patients undergoing PD (mean age: 11.42 ± 3.18), and 30 healthy controls (mean age: 10.11 ± 3.74).

Results: There was no statistical difference between the groups considering age, sex, and body surface area (BSA). NT-proBNP levels of HD and PD patients were statistically higher compared with the control group (p < 0.001, p < 0.001), there was no difference between pre-dialysis patients and the control group. OH, Rel OH, E/I levels of HD and PD patients significantly increased compared with the pre-dialysis and control group. Left atrium diameter, early dialostic flow velocity/late diastolic flow velocity (E/A) in HD and PD patients, and left ventricle mass index in HD, PD and predialysis patients were significantly higher compared with the control group. A positive correlation between NT-proBNP and OH, Rel OH, E/I, left atrium diameter, left ventricle mass index and a negative correlation with E/A was detected.

Conclusions: NT-proBNP and body fluid load was found to be increased in dialysis patients and these pararmeters were associated with left ventricle systolic and diastolic function parameters. Therefore, we believe that the routine use of NT-proBNP in children with chronic kidney failure has importance for close follow-up and prognosis of patients.

P-121 AN EXPLORATION OF THE ASSOCIATION BETWEEN URAEMIC TOXIN CONCENTRATIONS AND QUALITY OF LIFE IN CHILDREN WITH CHRONIC KIDNEY DISEASE

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Introduction: As kidney function deteriorates, uraemic toxins accumulate and contribute to the clinical picture of children with chronic kidney disease (CKD). These children are reporting poorer overall quality of life (QoL) and poorer physical, school, emotional, and social functioning. The aim of this study was to explore the association between levels of uraemic toxins and QoL.
Material and methods: In 23 children (11.0[6.9;14.6]years, 61%boys) with non-dialysis CKD stage 1–5, plasma concentrations of small solutes (asymmetric dimethyl arginine, symmetric dimethyl arginine, creatinine), middle molecules (β2microglobuline, complement factor D), and protein-bound solutes [p-cresylglucuronide, hippuric acid (HA), indole acetic acid (IAA), indoxyl sulfate, p-cresylsulfate, and 3-carboxy-4-methyl-5-propyl-furanpropionic acid (CMPF)] were measured. Their parents were asked to fill in the general (PedsQLTM 4.0 Generic Core: total score, physical & education subscale) and disease-specific QoL questionnaire (PedsQLTM End Stage Renal Disease (ESRD): disease & fatigue subscale). Lasso regression was used as an explorative method to select a set of predictive uraemic toxins (when $\beta \neq 0$) in models for the PedsQL questionnaires.
Results: The mean estimated GFR was 50.4 [31.2;74.5]mL/min/1.73 m2. CMPF was found to predict the total PedsQL ($\beta = -0.34$) and physical PedsQL subscales score ($\beta = -1.19$). Besides CMPF, IAA was predominant in the prediction of the total PedsQL and physical PedsQL subscale score(respectively $\beta = -0.84$ and $\beta = -1.26$); and HA in the education PesdsQL subscale ($\beta = -0.31$). The disease subscale of the PedsQL ESRD questionnaire was predominantly predicted by HA ($\beta = -1.18$) and IAA ($\beta = -2.30$). Using this model, creatinine was for none of the questionnaires selected as a possible predictor.
Conclusions: This model selected CMPF, IAA and HA as promising predictors for the hard endpoint QoL in children with CKD. Moreover, creatinine was not selected as a possible predictor for any of the QoL measures. A more extensive longitudinal study is necessary to strengthen our findings about the impact of uraemic toxins on the QoL in children with CKD.

P-122 EVALUATION OF WAVE PULSE VELOCITY IN CHILDREN AND ADOLESCENTS WITH CHRONIC KIDNEY DISEASE

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Introduction: Cardiovascular disease remains the most common cause of mortality in chronic kidney disease (CKD). Arterial stiffening measured by wave pulse velocity (PWV) predicts cardiovascular events and mortality in adults. Defining arterial stiffness may help to determine the cardiovascular risk.
Objectives: Investigate wave pulse velocity among children and adolescents with CKD.
Material and methods: In this cross-sectional study 57 patients (61.4% male), age 6.2–17.5 years, 44 with non-dialysis CKD and 13 on chronic dialysis were included in the analysis. The WPV was measured with an oscillometric device with inbuilt ARCSolver-algorithm (estimated by using the brachial waveform) and compared with previously established percentiles for PWV.
Results: The prevalence of elevated WPV was 21.1%. In multivariate logistic regression, it was noted a higher risk of elevated WPV in patients on chronic dialysis when compared with non-dialysis CKD patients (PRadj = 4.31, 95%CI:1.26–14.83, $p = 0.020$). Hypertensive patients (stage 2) have a higher risk of elevated WPV when compared with normotensives (PRadj = 3.11, 95%CI:1.17–8.24, $p = 0.022$) as the patients younger than 12 years comparing with the older patients (PRadj = 3.41, 95%CI:1.25–9.29, $p = 0.017$).
Conclusions: The findings suggest that lower ages, dialysis and hypertension in children and adolescents are independently associated with the increase of WPV. Further researches are needed to clarify these relations with cardiovascular complications in children and adolescents with CKD.

P-123 URIC ACID, HYPERTENSION AND CAROTID INTIMA-MEDIA THICKNESS IN CHILDREN WITH CHRONIC KIDNEY DISEASE

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Introduction: Markers of cardiovascular disease have been associated with increased morbidity and mortality in patients with chronic kidney disease (CDK). Serum uric acid (UA) is related to hypertension and increase the cardiovascular risk. These surrogate markers have been commonly used to study the evolution of cardiovascular disease in children with CKD. The measurement of the intima-media thickness (IMT) has become an additional tool for the early detection of arterial injury and evaluation of cardiovascular risk in these patients. **Objective:** Evaluate the serum uric acid level, hypertension, the use of allopurinol and its relationship with IMT in patients with CKD.
Material and methods: This cross sectional study included 55 patients (60% males), aged 11.7 years (6.2–17.4 years), 43 with non dialysis treatment and 12 on chronic dialysis. Serum levels of UA were obtained for all patients. Hypertension was defined according to the Fourth Report of Blood Pressure in Children as BP > 95th percentile or when the patient received anti-hypertensive medications. The IMT were evaluated by ultrasonography by the same examiner and compared with established percentiles for IMT according to gender and height.
Results: We found that 25 (45.45%) patients had elevated serum UA, 23 (41.8%) were on allopurinol treatment, 18 (32.7%) were hypertensives and in this group 94% had IMT altered. In multivariate logistic regression there was a higher chance of elevated IMT in patients who were not on allopurinol treatment (PR = 1.32; 95% CI: 1.01; 1.74; $p = 0.047$) and hypertensive patients (PR = 1.28; 95% CI: 1.04; 1.58; $p = 0.023$).
Conclusions: Ours findings suggest that no allopurinol treatment and hypertension were independently associated with increased of IMT. Further studies are needed to elucidated these relations. Close monitoring of blood pressure, treatment of hypertension, monitoring of UA and its treatment with allopurinol are important prevention of cardiovascular disease progression.

P-124 ARE 1–25 VITAMIN D MEASUREMENT USEFUL FOR MANAGING CHILDREN WITH NEPHROLITHIASIS AND/OR NEPHROCALCINOSIS?

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Introduction: The measurement of calcitriol, the active form of vitamin D, can be expensive and time-consuming, and there is no clear evidence of its interest in the management of patients with nephrolithiasis (NL) and/or nephrocalcinosis (NC). The aim of this study was to study our

current practice of 1–25 vitamin D3 (1–25-D) assessment in order to evaluate its interest in the initial diagnosis and follow-up of children with NL/NC.

Material and methods: We retrospectively studied all pediatric patients having undergone at least one 1–25-D measurement in our pediatric nephrology unit between December 2014 and November 2015. Clinical, biological and radiological (ultrasounds) data were recorded.

Results: A total of 264 measurements of 1–25-D levels were performed in 200 patients (age range 1 month to 18 years). The primary renal diseases were the following: 39% NL, 18% NC, 14% transplantation, 5% nephrotic syndrome, 3% neonatal hypercalcemia, 3% hypercalcemia, 2% rickets, 1% tubulopathy, and 15% miscellaneous.

In the 113 patients with NC or NL (mean age 6.4 ± 5.5 years, 73 boys), the etiology of NC/NL were the following: 27% unexplained despite extensive investigations, 15% neonatal hypercalcemia, 11% hypercalciuria without hypercalcemia, 10% urinary malformations, 9% infections, 6% nutritional mistake, 5% hypervitaminosis D, 4% hypercalcemia hypercalciuria, 4% tubulopathy, 3% hyperoxaluria, 6% miscellaneous.

1–25-D levels were found inappropriately high in 39 (35%) patients and further led to the diagnosis of CYP24A1 mutation in 3 patients and of vitamin D hypersensitivity without any identified mutation in 3 patients. Moreover, it modified the clinical management in 54 (47%) patients.

Conclusions: Assessment of 1–25D levels modifies clinical management in 47% of patients, mainly by allowing an adaptation of native vitamin D supplementation. In 5% of pediatric patients with NC and/or NL, it also clearly improves the diagnostic strategy. It therefore seems useful in the evaluation of NC/NL in children.

P-125 PROGNOSTIC VALUE OF SERIAL KIDNEY BIOPSIES IN HENOCH-SCHöNLEIN NEPHRITIS PATIENTS - COMPARISON OF TWO CLASSIFICATIONS

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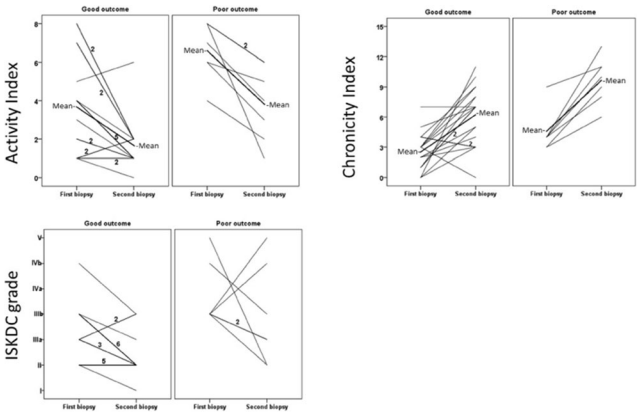
Introduction: Prediction of clinical outcome in Henoch-Schönlein nephritis (HSN) patients is difficult. Histological findings of sequential kidney biopsies from HSN patients were classified using the ISKDC (International Study of Kidney Disease in Children) classification and a novel semiquantitative classification (SQC) and their prognostic value on patient outcomes were compared.

Material and methods: Sequential kidney biopsies from 26 HSN patients were re-evaluated using the ISKDC classification and the SQC. SQC scores renal findings (glomerular, tubular, interstitial and vascular) and gives a total biopsy score as well as activity and chronicity indices. The biopsy findings based on the two classification systems were compared to the clinical findings at the last control visit. Nineteen (73%) patients had no signs of renal disease or minor urinary abnormalities (good outcome) and seven (27%) had active renal disease or chronic kidney disease (poor outcome). Median follow-up time was 8.5 years and time elapsed between the biopsies was 2.1 years.

Results: The patients with poor outcome had significantly higher activity and chronicity indices than patients with good outcome in both the primary kidney biopsy as well as the follow-up biopsy. Activity index increased in three cases between the two biopsies and stayed the same or decreased in 23. The respective figures for chronicity index were 22 and 4. ISKDC grading worsened in 4 and improved or stayed stable in 22. Changes between biopsies in the activity index, chronicity index and ISKDC grading occurred similarly in both outcome groups (Figure).



Conclusions: The present findings support our previous findings suggesting that SQC is superior to ISKDC classification in predicting renal outcome in HSN patients. Activity scores decreased and chronicity scores increased between primary and follow-up biopsy despite patient outcome. The control biopsy does not seem to be indicated routinely, but needs to be decided individually.

P-126 VASCULAR ENDOTHELIAL DYSFUNCTION IN RENAL REPLACEMENT THERAPY MODALITIES

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Introduction: Vascular endothelial dysfunction (VED) is an important cause of cardiovascular morbidity and mortality in end-stage renal disease (ESRD). Endothelin-1 (ET-1) and nitric oxide (NO) are vasoactive substances which was affected in ESRD. The aim of the study was to compare serum ET-1 and NO levels in renal transplant recipients (RTx group) and patients receiving hemodialysis (HD group), online-hemodiafiltration (HDF group), peritoneal dialysis (PD group).

Material and methods: Fourty-one patients and 25 healthy children were enrolled in the study. Serum ET-1 and NO levels were measured by ELISA in all patients and controls. Intradialytic symptoms and ambulatory blood pressure monitoring was evaluated in HD and HDF groups.

Results: Rtx group had the lowest level of serum ET-1 and NO although the difference did not reach statistical significance. Median serum ET-1 level were not significantly different between the HD and HDF group (589.944 ng/l and 593.717 ng/l; respectively, $p > 0.05$). Also, these levels were not different between the PD, RTx and HD groups (546.343 ng/l, 343.555 and 589.944 ng/l; respectively, $p > 0.05$). Median ET-1 level were significantly lower in RTx group than the HDF group (343.555 ng/l and 593.717 ng/l; respectively, $p = 0.025$). The median serum NO level was not different between the HD, HDF, PD and RTx

groups (590.237 μmol/l, 563.084 μmol/l, 582.433 μmol/l, 438.268 μmol/l; respectively, $p > 0.05$).
There was no difference between the HD and HDF groups in terms of hypertension, hyperparathyroidism, anemia, metabolic acidosis, hyperlipidemia, inter-dialytic weight gains and Kt/v ($p > 0.05$). When each patient groups were compared with the control group separately NO and ET-1 levels were higher in patient groups ($p = 0.0001$). Median NO level was 56.212 μmol/l, ET-1 level was 31.827 ng/l in the control group.
Conclusions: Our results suggest that ESRD causes VED. No difference between ESRD treatment modalities (HD, HDF, PD) in terms of ET-1 and NO indicates that VED continues in all treatment modalities. Renal transplantation is superior to other treatment modalities.

P-127 RENAL AMYLOIDOSIS IS STILL A HEALTH PROBLEM IN ARMENIA – EFFECT OF COLCHICINE

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Introduction: Familial Mediterranean fever (FMF) is a serious health problem in Armenian children. Although amyloidosis – a potentially fatal complication of FMF – can largely be prevented by colchicine administration, we are still confronted with renal amyloidosis. The aim was to analyse the demography and reasons for amyloid nephropathy in pediatric patients and the late effects of colchicine.
Material and methods: The National Pediatric Center for FMF (NPC FMF) was established in 1998 to allow early diagnosis, treatment, follow-up and wide dissemination of information on FMF. Since 2003 NPC FMF has provided colchicine at no charge to over 3000 children below the age of 18 years. Diagnosis of FMF is based on Tel-Hashomer criteria and molecular genetic analysis (since 1998). Amyloid nephropathy was confirmed by renal biopsy (Congo Red). All pediatric renal biopsies since 1993 ($n = 307$) are included. The data 1993–2004 (group 1; $n = 155$) and 2005–2016 (group 2; $n = 152$) have been analyzed separately. Colchicine was administered to all patients with renal amyloidosis.
Results: Amyloid nephropathy was diagnosed in 38 pts. (24.5%) in group 1 and in 22 (14.5%) in group 2 ($p < 0.05$). In the second group diagnosis of FMF had been missed in 18 patients, one was noncompliant and in three colchicine was not sufficiently effective. On admission six had proteinuria, 14 were nephrotic and two had CKD (stage 3). Late administration of colchicine could not reverse the course.
Conclusions: The National long-term program on early diagnosis and regular colchicine treatment of FMF in children in Armenia is only partly effective in prevention of renal amyloidosis and requires additional efforts. Colchicine is not able to reverse advanced amyloid nephropathy.

P-128 INTERRELATION OF ERYTHROPOIETIN (EPO), HYPOXIA INDUCIBLE FACTOR-1α (HIF-1α) AND THE GLOMERULAR FILTRATION RATE (GFR) BY ANEMIA IN CHILDREN WITH CHRONIC KIDNEY DISEASE (CKD)

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Introduction: To evaluate the interrelation of EPO, HIF-1α and the GFR in children with anemia in CKD.
Material and methods: We analyzed levels of EPO and HIF-1α in 80 patients with anemia in CKD: 39 boys (48, 75%) and 41 girls (51, 25%). Three groups of patients: I (32) predialysis without therapy, II (18) predialysis with therapy, III (30) dialysis (21-HD, 9-PD).
Results: 80 patients with anemia in CKD32 patients (I Group) with CKD had mean GFR 42.89 ± 25,2 ml/min/1.73 m^2; Hb level 103.2 ± 8.5 g/dl, EPO level was 28.1 ± 20.6 mIU/ml; HIF-1α levels 0,163 ± 0,4 pg/ml. Eighteen patients (II Group) had mean GFR 31.5 ± 26.4 ml/min/1.73 m^2; Hb level 100 ± 10 g/dl, EPO level was 118 ± 25.4 mIU/ml; HIF-1α levels 0.12 ± 0.1 pg/ml. Thirty patients (III Group) had mean GFR 13.1 ± 6,2 ml/min/1.73 m^2; Hb level 85 ± 14,4 g/dl, EPO level was 34.3 ± 43,8 mIU/ml; HIF-1α levels 0.09 ± 0,07 pg/ml. EPO level in children II Group was higher, compared with I and III Group ($p < 0.05$). HIF-1α correlated with GFR by18 patients II Group ($R = 0,589$, $p < 0.05$), by 32 patients I group ($R = 0, 42$), by 30 patients III group ($R = 0,087$).
Conclusions: We identified the evolution in indicators HIF-1α in children with anemia in CKD. In parallel with a decrease GFR, the tightness of the connection between HIF-1α and GFR decreases.

P-129 LIFE THREATENING ARRHYTHMIA AND NON-CONVULSIVE STATUS RELATED TO CEFEPIM IN A PATIENT WITH CHRONIC RENAL FAILURE

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Introduction: Cephalosporins, particularly cefepime, exert neurotoxic side effects that can lead to status epilepticus. These neurotoxic side effects include myoclonus, dystonic movements, tremor, asterixis, seizure, status epilepticus, encephalopathy, and sometimes coma. Status epilepticus, particularly nonconvulsive status epilepticus (NCSE), is a well-known but unusual complication in patients with altered renal function who were receiving treatment with intravenous cephalosporins, especially cefepime. Cardiotoxicity due to cephalosporins is not reported yet.
Material and methods: Here we report a chronic peritoneal dialysis patient who received cefepime for peritonitis.
Results: He developed tremor and asterixis on the 6 th day of treatment. His electroencephalography revealed status epilepticus and considered to be in a state of non-convulsive status. Electrocardiography revealed frequent ventricular extrasistoles, while his echocardiographic examination revealed slight ventricular hypertrophy. Cefepime was discontinued immediately. His neurological symptoms subsided in 24 h while arrhythmia subsided in a week.
Conclusions: Nonconvulsive status epilepticus was observed between 2 and 8 days (average of 5.6 days) after initiation of cephalosporins. Cephalosporins are epileptogenic drugs, especially when used in excessive doses or when renal function is impaired. Critically ill patients with chronic kidney disease are particularly susceptible to cefepime neurotoxicity. However, there is not any reported case related to cardiotoxicogenic or arrhythmogenic affect of cefepime in literature. Cefepime should be used carefully in patients with renal failure. Its neurotoxic and arrhythmogenic affect should be kept in mind.

P-130 OPTIMISING FLUID INTAKE HELPS IN THE MANAGEMENT OF VOMITING IN INFANTS AND CHILDREN WITH CHRONIC KIDNEY DISEASE: A CASE SERIES

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Introduction: It is well recognised that vomiting frequently occurs in infants and children with chronic kidney disease (CKD). Multiple factors

have been proposed to explain its' occurrence including gastroesophageal reflux and delayed gastric emptying.
Material and methods: We retrospectively report 3 patients with CKD who were receiving enteral feeds and experienced issues with vomiting.
Results: Patient 1: 28 month old girl with CKD following placental abruption. Vomiting daily for the previous 5 months despite domperidone. Care transferred to our unit with plan for fundoplication. On presentation 110mls/kg/d total water from gastrostomy feeds and oral water. Initially feeds changed to ¼ strength and total water (all sources) increased to 145mls/kg/d. Vomiting settled and feeds increase to full strength over 2 week period. Patient 2: 3 day old boy with PUV and left multicystic dysplastic kidneys. Feeds provided 180mls/kg/d water and family advised to offer water between feeds. Two weeks later started vomiting, had outgrown 180mls/kg/d target and parents admitted difficulty giving additional water. Additional water added to feeds to achieve 180mls/kg/d and vomiting settled. No anti-reflux medications required. Patient 3: 1 month old girl with dysplastic kidneys tolerated continuous feeds (155mls/kg/d fluid) but would vomit when bolus feeds were trialled. Half strength continuous feeds with increasing water to 170mls/kg/d were not tolerated. Patient given IV fluids and domperidone over a weekend period then successfully regraded to continuous feeds of 170mls/kg/d.
Conclusions: Optimising fluid intake as water either added to feeds and/or given separately helps in the management of vomiting in infants and children with CKD. Further work is required to investigate this relationship.

P-131 PAEDIATRIC CHRONIC KIDNEY DISEASE – IDENTIFYING THOSE AT RISK FROM MALNUTRITION

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Introduction: Identifying children with Chronic Kidney Disease (CKD) who are at increased risk from malnutrition is difficult due heterogeneity of populations. There is no formalised, agreed process whereby children are determined to be at nutritional risk.
Material and methods: Children aged 3 and 18 years with CKD (stages 2 to 5D) were screened for risk of malnutrition using nutrition screening tools: Screening Tool for the Assessment of Malnutrition in Paediatrics (STAMP); Screening Tool for Risk On Nutritional status and Growth (STRONGkids); Simple Paediatric Nutrition Risk Score (PNRS); and Paediatric Yorkhill Malnutrition Score (PYMS). For comparison, the degree of malnutrition was assessed by anthropometry alone using World Health Organization International Classification of Diseases (ICD-10) criteria.
Results: 60 children with CKD were recruited; 46 of whom were conservatively managed, 10 had previous undergone renal transplantation, and 5 were receiving dialysis (4 haemodialysis, 1 peritoneal dialysis). Mean values (with standard deviations) of age, height SDS, weight SDS and BMI SDS for the cohort were 10.7 years(±4.0), −1.19(±1.53),-0.42(±1.76), and 0.46(±1.36), respectively. Ten children(17%) had weight SDS < −2. 1 child(1.7%) was under-weight for height(BMI SDS < −2). Seventeen children(28%) were short-for-age(height SDS < −2). There was poor concordance between ICD-10 anthropometric definitions of malnutrition risk and all screening tools. Additionally, there was poor inter-tool agreement; with no 2 tools showing a Cohen's kappa value of greater than 0.2.
Conclusions: Although discrepancy is expected, to be a clinically useful screening tool, they must be able to identify those at risk, and not miss individuals that require assessment / intervention. Currently used screening tools are not adequate for stratifying nutritional risk, and standardised nutritional assessment is needed, although inadequate resources for this are in place. Attention must be given to a identifying those at risk from nutritional inadequacy; including adiposity. It therefore seems essential to utilise a specialist paediatric renal dietitian.

P-132 CO-EXISTANCE OF INTERRUPTED INFERIOR VENA CAVA SYNDROME AND AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE: CASE REPORT

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Introduction: Interrupted Inferior Vena Cava Syndrome (IVCS) is a rare vascular anomaly. Although it usually presents with other congenital anomalies, there is no report about its co-existing with autosomal recessive polycystic kidney disease (ARPKD).
Material and methods: Here we present a 4-month-old girl with ARPKD and IVCS.
Results: Four month-old female patient presented to our clinic due to vomiting which started in the neonatal period but intensified gradually. History of small for gestational age and consanguineous marriage were noted. Growth retardation, tachycardia (140/min), 2/6 systolic murmur, severe hypertension and end-stage renal disease were detected in her physical and laboratory examination.
Interrupted inferior vena cava, which continued as a dilated azygos vein to the superior vena cava was detected on abdominal ultrasonography. Left isomerism and polysplenia were found as the component of IVCS. The liver parenchymal echogenicity had risen in accordance with congenital hepatic fibrosis. Bilateral grade 2 hyperechogenicity and multiple ubiquitous millimetric anechoic cysts were seen in renal parenchyma. Echocardiography revealed left ventricle wall and intraventricular septum hypertrophy as a result of chronic hypertension. Cranial ultrasound was normal. Peritoneal dialysis and the relevant medical therapy for end stage renal disease and hypertension has been arranged.
Conclusions: Interrupted inferior Vena Cava Syndrome may be co-exist with ARPKD. Patients with ARPKD should be evaluated in terms of other congenital anomalies.

P-133 ASSESSMENT OF LABORATORY ABNORMALITIES IN RENAL OSTEODYSTROPHY AND ITS RELATION WITH THE STAGE OF CHRONIC KIDNEY DISEASE

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Introduction: The final result of chronic kidney disease (CKD) without considering the type of kidney disease is progression to kidney failure and its complications such as renal osteodystrophy, cardiovascular, endocrine, neurologic, acid-base disorders, electrolyte disorders and anemia. Mineral bone disease could be assessed with laboratory abnormalities and radiologic findings. The purpose of this study was assesssing the laboratory abnormalities in renal osteodystrophy and finding a relation with the stage of CKD.
Material and methods: A cross-sectional study was performed on 104 children between 1 month to 17 years old who were admitted to the nephrology ward of Mofid hospital. They were evaluated by history taking, physical examination, blood and urine analysis, ultrasound and echocardiography. According to the glomerular filtration and KDOQI instruction, patients were divided into 5 groups: results were expressed using SPSS statistics and $p < 0.05$ was considered statistically significant.
Results: Out of 104 cases, 65 patients (62.5%) were male and 39 (37.5%) were female. The age range was 1 month to 17 years old and the mean age

was 7.15. Four patients (3.8%) were at stage 1, 14 patients (13.5%) were at stage 2, 15 patients(14.4%) at stage 3 and 71 patients (68.3%) at stages 4 and 5. The most common etiology of CKD was neurogenic bladder with 19 patients (18.2%),then glomerular diseases such as nephrotic syndrome and lupus with 15 patients(14.2%), obstructive uropathy with 12 patients (11.5%), reflux nephropathy and atypical HUS each with 10 patients (9.6%), polycystic kidney disease with 5 patients (4.8%) and other etiologies (like malignancies, nephronophthisis and Bardet- Beidl sundrome) formed 12.5%.Twenty patients (19.9%) had unknown etiology. Eighty two patients (78.8%) had anemia, 30 patients (28.8%) had hypocalcemia, 30 patients (28.8%) had hyperphosphstemia, 75 patients (72.1%) had metabolic acidosis and 34 patients (32.6%) had high alkaline phosphatase. Seventy two patients (69.2%) were insufficient or defficient for 25 (OH) vitamine D. Twelve patients (11.5%) were treated with hemodialysis, 30 patients(28.8%) with peritoneal dialysis and 7 patients (6.7%) underwent renal transplantation.A significant correlation between uric acid and 25 (OH) vitamine D level and CKD stages ($P = 0.018$) and between 25(OH) vitamine D and PTH level ($P = 0.016$) was found.
Conclusions: The prevalance of CKD complications in children is high and assessment of patients in early stages for early treatment is advised.

P-134 CAUSES OF CHRONIC RENAL INSUFFICIENCY IN ALBANIAN CHILDREN

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Introduction: There is an increased incidence of congenital and hereditary diseases causing chronic renal failure in the pediatric age-group. To determine the major causes, clinical expression, course, and outcomes of CKD in Albanian children we conducted a prospective study from January 2015 to January 2017 in the pediatric nephrology department at the UHC "Mother Tereza" in Tirana.
Material and methods: This prospective study included all children (up to 15 years old) with the diagnosis of CRF who presented to the department of pediatric nephrology at the University Hospital Centre of Tirana during the period January 2015 to January 2017. The parameters studied included: gender, age, place of residence, age at the first complaint, age when the diagnosis of CRF was made, age at which the patient reached ESRD (if applicable), family history of similar kidney diseases, consanguinity, cause of CRF, associated malformations, co-morbidity {recurrent urinary tract infections (UTI), hypertension and its response to therapy.
Results: Forty-eight patients with varying degrees of renal impairment were involved in the analysis. A total of 27 children (56%) had obstructive nephropathy (ON) as the cause of chronic renal insufficiency and 21 children (44%) had non-obstructive nephropathy (Non-ON). Neurogenic bladder was the commonest cause of ON, seen in 13 patients (27%), nephrolithiasis was seen in 9 patients (19%), urethral stenosis in three (6%), Uretro-Pelvie Junction (UPJ) stenosis in one (2%), and posterior urethral valves in one case (2%).
Conclusions: In our study obstructive nephropathy has been shown to be the most important cause of CKD. Whether this is due to a true higher prevalence of some causes of obstructive nephropathy or an insufficient sample size, can only be elucidated with further studies involving larger number of patients.

P-135 RENAL REPLACEMENT THERAPY FOR SEVERE HYPEROSMOLALITY AND AKI

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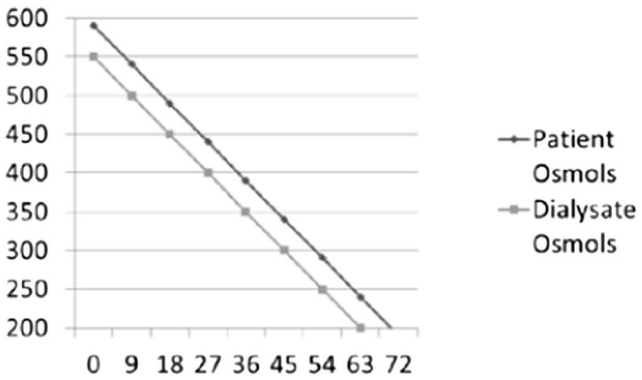
Introduction: Management of AKI & hyperosmolality using conventional renal replacement methods places patient at higher risk of rapid osmolar shifting that leads to major neurological consequences. CRRT provides the ability to control rate of reduction in osmolality by allowing the adjustment of dialysate solution and narrowing osmolar gap between the patient and dialysate. Further, "inefficient" solute clearance will less the rate of pH and osmolar changes over time.
Material and methods: Case Report.
Results: A 16-kg male child known case of Central Diabetes Insipidus presented unconscious and anuric with septic shock, anemic (Hb 4.8 g/l), AKI (BUN 427 mg/dl, Creatinine 7.6 mg/dl), severe hypernatremia (Na 216 mmol/l), and a PH of 7.0. Measured osmolality was 593 osmols/l. Patient was resuscitated, incubated and shifted to PICU. "Inefficient" CVVHD was begun at 8 mls/kg/h (in order to slowly improve the pH) with the use of PrismaSate® with an additional 80 meq/l of NaCl added to give total Na of 200 meq/L of PrismaSate® resulting in a dialysate bath of 550 osmols/l. Patient osmols were recalculated at 3 h increments and additional Na in the dialysate was decreased as needed. Based upon patient osmolar changes, additional sodium was adjusted until normal osmols were obtained (Figure). Over 72 h the child had gradual drop of sodium till reaching 170 mmol/l then CVVHD was stopped and patient was shifted to medical treatment of hypernatremia. Over time patient had recovery of osmols, PH, renal and neurological function and continued on medical management.



Conclusions: To our best knowledge, this is the first case in literature to have such presentation and manage by this way. The patient presented with severe hyperosmolality and significant metabolic acidosis. A rapid correction of either of these conditions places him at risk for herniation and pontine demyelination. Utilizing a slow approach to osmolar and pH corrections is recommended in the literature to avoid these risky complications. Standard dialysis dosing of 35 mls/kg/h or 2000 mls/m2/h will result in significant solute clearance. By making the CVVHD prescription inefficient, one can then do a slow correction of the metabolic acidosis and with manipulation of the sodium bath of the dialysate one can narrow the osmolar gap between the patient and dialysate allowing for slow and continuous correction of the osmolality.

P-136 A RARE INHERITED DISEASE IN TWO TWIN SIBLINGS

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Introduction: Objective: To identify the molecular disease cause in two twin siblings with nephrotic syndrome and dysmorphic features, we performed mutational analysis.

Material and methods: Through a collaborative work, the two cases were studied with regards to their phenotype in the pediatric nephrology unit at the faculty of medicine, university of Alexandria, and mutation analysis was performed in the Hildebrandt lab at BostonsChildren Hospital, Harvard MedicalSchool. To identify the causative mutation in these two siblings born of consanguineous union, we combined whole exome sequencing with homozygosity mapping.
Results: The two twin siblings, the outcome of consanguineous marriage,were a boy and a girl, who presented with short stature, dysmorphic facial features, skeletal anomalies,and hypothyroidism. Furthermore, the boy had congenital heart disease, and he presented with nephrotic syndrome, that rapidly progressed to end stage kidney disease, while the girl only showed nephrotic range proteinuria without renal impairment. Whole exome sequencing revealed a homozygous mutation in the gene *COG1* that segregated with the affected status in this family. The mutated allele is likely deleterious as it has never been reported in healthy control databases (i.e. ExAC or gnomAD) and affects a well-conserved splice site. Mutations in the gene *COG1* have been previously described in two unrelated families as monogenic causes of cerebrocostomandibular-like syndrome (MIM# 611209). However, renal involvement has not been described yet in patients with *COG1* mutations.
Conclusions: By whole exome and homozygosity mapping, we identified a rare genetic syndrome as the molecular disease cause in these siblings. We expand the phenotypic spectrum of *COG1* mutations by describing renal involvement for the first time in this monogenic disease.

P-137 A VERY UNCOMMON CAUSE OF ACUTE KIDNEY INJURY IN INFANCY

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Introduction: *Severe Combined Immuno-Deficiency* (SCID) is rarely associated with non-infectious/non-toxic acute kidney injury (AKI).
Material and methods: We report herein a unique cause of AKI related to lymphoproliferation, revealing a SCID.
Results: A previously healthy four month-old boy was admitted for AKI (creatinine 363 μM), hypertension, and edema, in the setting of a rhinovirus-positive bronchiolitis. He exhibited hyperechoic enlarged kidneys, nephrotic syndrome, anemia (8.8 g/dL), thrombocytopenia (87,000/mm3), elevated LDH, 1.7% schistocytes, and reticulocytes 135,000/mm3. He was started on peritoneal dialysis. An atypical HUS was suspected and eculizumab was given. Despite rapid hematologic parameter normalization, renal function did not improved. Kidney biopsy, performed after the fifth dose of eculizumab, revealed acute tubulointerstitial nephritis with lymphocyte and macrophage infiltration, inflammatory cells within the glomerular capillaries, with no evidence of thrombotic microangiopathy. Eculizumab was withdrawn, and three methylprednisolone pulses followed by oral prednisone were given without any improvement. Due to persistent lymphopenia and agammaglobulinemia, immune-deficiency was suspected and confirmed by immunophenotyping showing low T-cells (900 CD3/μl-93% γδ-T-cells), B-cells (70/μl) and NK-cells (10/μl) compatible with the diagnosis of SCID. Surprisingly, γδ-T cells were from maternal origin. Immunoscope showed oligoclonal profile of T-cells. Immunologic analyzes revealed a*n IL-2R common-γ chain* defect causing X-linked SCID, and the diagnosis of materno-foetal graft-versus-host disease (MF-GVHD) caused by intrauterine materno-fetal transfusion was almost indisputable. He underwent successful geno-identical stem-cell transplantation at 7 months of age, followed by cadaveric kidney transplantation at the age of 3.4 years.
Conclusions: Placenta barrier allows bidirectional passage of nucleated cells during pregnancy. While healthy infants reject these cells, profound immunodeficiency related to SCID allows persistence of maternal T-cells in newborns and peripheral expansion possibly driven by microorganisms. Maternal T-cell engraftment may be responsible for MF-GVHD mainly of skin, liver or digestive tract. We report herein a highly singular case of isolated nephritis related to presumed MF-GVHD.

P-138 A LYSIS CRISIS

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Introduction: We report a diagnostic dilemma in a child who presented with acute kidney injury and unusual clinical features.
Material and methods: Clinical history, repeated clinical assessment,urine dipstick, microscopy, haematology, biochemical tests, histopathology and flow cytometry will be presented.
Results: 16 year old boy with no significant past medical problems presented with history of passing cola coloured urine and abdominal pain. Urine dipstick showed haematuria and proteinuria but there was no erythrocyturia on microscopy. Blood picture was consistent with intravascular haemolysis with thrombocytopenia. There was rapid progression of the acute kidney injury with peak creatinine of 698 umol/l. Urine output was well preserved and there was no evidence of fluid overload. Kidney biopsy was performed in view of rapid deterioration of renal function. Autoimmune screen was negative. Histopathology was consistent with acute tubular injury. The striking feature was significant deposition of haemosiderin in the tubules with no evidence of glomerulonephritis. Flow cytometry confirmed the diagnosis of paroxysmal nocturnal haemoglobinuria. He was treated with eculizumab and responded well to treatment.
Conclusions: Paroxysmal nocturnal haemoglobinuria can present as rapidly progressive acute kidney injury. Careful interpretation of urine dipstick results in the context of absence of red cells on microscopy is an important diagnostic clue to diagnose haemoglobinuria. Treatment with eculizumab led to dramatic recovery.

P-139 GENETIC PREDISPOSITION TO INFECTION IN A CASE OF ATYPICAL HEMOLYTIC UREMIC SYNDROME (AHUS)

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Introduction: Hemolytic uremic syndrome (HUS) is a major cause of renal failure in childhood. Most cases are caused by infection with Shiga-toxin producing *Escherichia coli* (STEC). In 5–10% of cases, HUS is not preceded by the STEC infection and is considered atypical (aHUS). These cases are strongly associated with genetic defects leading to dysregulation of the complement system. Often aHUS is triggered by a non-STEC

infection, however, genetic predisposition to such infections in aHUS has not yet been studied. Here we present thorough complement analysis of a 2 months old patient in whom aHUS episode coincided with Bordetella pertussis infection (whooping cough), Klebsiella oxytoca sepsis and Moraxella catarrhalis pneumonia.
Material and methods: In vitro kinetics of complement activation products (C3bc and TCC) in serum were quantified using ELISA. DNA analysis was performed by Sanger sequencing. Recombinant vitronectin variants were produced in HEK293T cells, purified and used in hemolytic assay with sheep erythrocytes and purified C5b-6, C7, C8 and C9 complement proteins.
Results: The in vitro complement activation kinetics were compared in patient serum and normal human serum (NHS). In patient serum C3 activation rate (expressed as generation of C3bc) was comparable to the rate in NHS, but the rate of TCC generation was slower. Genetic analysis of TCC components and TCC inhibitors revealed a rare heterozygous variant p.Arg229Cys in vitronectin. Prediction software (SIFT, PolyPhen-2) indicated this change as pathogenic. In vitro experiments using recombinant vitronectin variants have shown that this mutation enhances complement inhibition at TCC level.
Conclusions: Our work indicates that not only genetic changes leading to uncontrolled complement activation but also these increasing vulnerability to infections contribute to aHUS.

P-140 THROMBOTIC MICROANGIOPATHY IN THE FRAME OF SYSTEMIC DISEASES: FOUR CASES FROM PEDIATRIC CLINICAL PRACTICE

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Introduction: Thrombotic microangiopathies result from the interaction between predisposing factors and triggering events.
Material and methods: We present four pediatric cases with unusual forms of thrombotic microangiopathy in the frame of systemic diseases.
Results: Case 1: 5 years old, acute pancreatitis with complement activation and HUS-triad (hemolytic anemia, thrombocytopenia and acute renal failure). Early treatment with Eculizumab results in rapid hematological and renal remission. MRI shows anatomic variant (pancreas divisum) associated to high risk of spontaneous pancreatitis. Recurrence of pancreatitis under treatment with Eculizumab, with no signs of TMA. Genetic analysis shows risk haplotypes in CHF, MCP and CFHR3-CFHR1 deletion. Pancreatitis triggered aHUS or vice versa?
Case 2: 2 years old, acute myeloid leukemia M5, after haploidentical BMT, complete chimerism. After BMT develops transfusion-dependent anemia, malignant hypertension and proteinuria. Transitory stabilization after suspension of cyclosporine. Progressive disease with two-fold increase of creatinine, renal biopsy shows TMA. Under treatment with Eculizumab normalization of blood pressure and renal function, no proteinuria. Hemoglobin recovery, no need for transfusions. Material for genetic testing from saliva (blood cells from donor), result pending.
Case 3: 15 years old, previously healthy, presents with cholestasis and HUS-triad. Ocular evaluation shows Keyser-Fleischer Ring. Copper in urine and low ceruloplasmin confirm diagnosis of Wilson's disease. Hemolytic anemia only better under treatment with Trientine. Progressive recovery of hemoglobin and renal function. Marked complement activation, CFB-Antibodies and CFHR5 variant.
Case 4: 10 years old, Wilson's disease. Three years after liver transplantation presents HUS-triad in the frame of viral infection. Recovery after suspension of treatment with Tacrolimus and Everolimus. Carrier of CHR1 and 3 homozygous deletions.
Conclusions: This cases picture the relevance of considering TMA in non-aHUS related diseases in which complement activation and endothelial damage may be trigger or consequence of complement activation. The relevance of complement gene polymorphisms as TMA predisposing condition is not yet well understood.

P-141 ATYPICAL HAEMOLYTIC UREMIC SYNDROME CASE REPORT TRIGGERED WITH E.COLI 0119 K69 INFECTION

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Introduction: To describe a case who presented with atypical HUS and carries a homozygous CFHR1/CFHR3 deletions. She was found to have E coli 0119 k69 infection as well. We assume that E coli infection has triggered HUS in our case.
Material and methods: Case report.
Results: 1 year old girl presented with fever, non bloody diarrhea for 5 days. She has been anuric for couple of days before presenting to Hospital. Her initial investigations revealed acute renal failure with microangiopathic haemolysis. Platelets has been normal all the way through her illness. Her creatinine was 4.9 mg/dl (0.2–0.4), urea was 178 mg/dl (12–40), uric acid 15.4 mg/dl (2–6.2). high retic count at 3.13% (0.8–2). Normal electrolytes. Her parents are first degree cousins with no family history of renal diseases. LDH 5229 u/l (0–850). Blood film was consistent with HUS. She has received 1 dose of Eculizumab on her second day of admission, she has needed 6 haemodialysis sessions over 10 days as she has remained anuric for 10 days. She has also required blood transfusion. Her urine output has improved and she was discharged home after 3 weeks. She was not given any further doses of Eculizumab. Her initial total complement activity CH50 was low at presentation, however, her repeated level at 3 monthly basis has all been normal along with normalCFH level. Full genetic testing for atypical HUS has revealed that she carries a homozygous CFHR1/CFHR3 deletions. She has remained well with normal renal function, she was kept on low dose of ACE inhibitor due to low grade proteinuria.
Conclusions: To our knowledge, this is the first case in literature to have this combination. It also raises the question of the benefit of giving Eculizumab for life in this very rare case. After discussing with the family, we keep repeating CH50 as a marker of complement activity. Further studies needed to answer this question.

P-142 THROMBOTIC MICROANGIOPATHY AND BREASTFEEDING. WHERE IS THE LINK?

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Introduction: Thrombotic microangiopathy (TMA) is exceptional in infants and is mostly linked to thrombotic thrombocytopenic purpura via ADAMTS13 gene mutations or to atypical hemolytic uremic syndrome. However, vitamin B12 disorders can also be associated with TMA. We report a case of TMA in a 7-month-old patient due to severe vitamin B12 deficiency.
Material and methods: A 7-month-old boy presented to our emergency department with marked pallor and severe asthenia. Laboratory tests revealed a severe anemia (haemoglobin 30 g/L), thrombopenia (26×10^9/ml) and 20% schistocytes. Initial investigations revealed also leukopenia, lymphopenia and megaloblastosis. Renal function was normal. Test for Shiga toxin producing *Escherichia coli* was negative, there was no complement activation and ADAMTS13 activity was normal.
Results: He was rapidly treated by fresh frozen plasma infusion and a high dose of vitamin B12 (5000 μg IM) at the time of admission,

which allowed stopping the hemolysis within 12 h. Further metabolic investigations revealed a markedly elevated urinary methylmalonic acid (MMA) at 1245 μmol/mmol creatinine (< 10 μmol/mmol creatinine). Vitamin B12 level was very high (>1500 pmol/l) and plasma homocysteine very low (< 3 μmol). Unfortunately, these tests were performed after plasma infusion and B12 injection. Five days after the beginning of the vitamin B12 treatment, MMA level was normal (2 μmol/mmol). Moreover, the patient received exclusively breastfeeding; his mother has a severe vitamin B12 deficiency (< 50 pmol/l) and we noted a progressive lethargy and a poor weigh gain since he was 4 months old, suggesting a vitamin B12 deficiency rather than a disorder in the cobalamin metabolism.
Conclusions: Vitamin B12 deficiency can lead to severe TMA, even in infants. TMA is probably linked to hyperhomocysteinemia which is known to cause endothelial dysfunction.

P-143 ZEBRAS CAN KILL YOU- THE ONE ZEBRA YOU SHOULD NEVER FORGET ABOUT DURING HAEMODIALYSIS

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Introduction: Haemodialysis (HD) is a challenging treatment especially within the paediatric population. Though life saving, it is not without risk to the patient. Acute haemolysis associated with haemodialysis (HD) is rare, but potentially life threatening complication which requires prompt recognition by the clinician. We describe the first paediatric patient with this sequelae, a 13 year old girl with a background of HIV nephropathy, well established on HD post failure of her renal transplant.
Material and methods: Approximately 90 min into a routine haemodialysis session via an AV fistula on a Gambro™ AK200 highflux dialyser, the patient developed facial flushing, whole body rash, diffuse abdominal pain and significant hypertension (>190 mmHg). Haemodialysis was ceased and the patient treated for a presumed severe allergic reaction. She remained symptomatic for the next 24 h. Unfortunately her post dialysis bloods were unable to be analysed due to 'gross haemolysis'.
The following morning, she recommenced haemodialysis on a Fresenius™ 5008 using a highflux dialyser. Post connection, a blood leak alarm was triggered. A second Fresenius™ machine was prepared with a midflux dialyser. Again, a blood leak alarm was triggered. A third circuit was prepared on the Gambro™AK200.
Results: At this point we became strongly suspicious of a severe haemolytic reaction. Bloods revealed an acute drop in haemoglobin and haematocrit, grossly raised LDH and bilirubin and positive DAT. The patient tolerated the dialysis session but was not 'washed back'. Dialysate fluid was sent for chemical, bacterial toxin and copper analysis. Tubing and dialysers were returned to the respective companies for analysis. All results returned negative. By elimination, we deduce the most likely cause of haemolysis to be mechanical from the AV fistula in combination with intermittent HIV or EBV viraemia. The patient fully recovered within 72 h.
Conclusions: Acute haemolysis is a rare but life threatening complication associated with haemodialysis. Prompt recognition and appropriate management is vital.

P-144 NEONATAL ONSET OF ATYPICAL HEMOLITIC UREMIC SYNDROME (AHUS) TREATED WITH THERAPEUTIC PLASMA EXCHANGE (TPE) USING CARPEDIEM MACHINE

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Introduction: aHUS is a potentially life-threatening rare disease characterized by the triad:Coombs negative microangiopathic anemia,thrombocytopenia and acute kidney injury(AKI).It can be sporadic or familial,and its often associated with genetic complement abnormalities/anti-complement factor-H antibodies.
Material and methods: A 42-day-old male was referred to our center because of AKI,anemia,and thrombocytopenia.He was a term neonate(bw.2765 g)with an uneventful course of pregnancy.On the 2nd day of life,the patient experienced abdominal distension,oliguria,anemia and thrombocytopenia.An abdominal X-ray showed a necrotizing enterocolitis,but,despite the treatment he became hypotensive,with a remarkable increase in inflammatory markers,and finally reached a 30%fluid overload(3490 g).At 6 weeks of life,he was transferred to our unit.Anemia,thrombocytopenia and AKI suggested a thrombotic microangiopathy(TMA).The blood tests showed a consumption of haptoglobin,increase in LDH,schistocytes with negative Coombs test and a reduction of C3 with normal fibrinogen and coagulation tests.Potential causes of TMA were investigated and the overall picture was then suggestive for a complement-mediated aHUS.Due to the abdominal condition and the infectious risk,no complement-blocker was used.
Results: TPE was then initiated using the CARPEDIEM machine and the Plasmart05 filter,with fresh frozen plasma(FFP)as replacement fluid and an exchange volume of 200 ml.The circuit was primed with 50 mL of 4%albumin.The blood pump rate was set at 10 mL/min,with an exchange rate of 1 mL/min and a total session time of 3.5 h.He received 5 consecutive daily sessions of TPE,without any clinical/technical problem.The first 2 sessions of TPE were followed by a CVVH.Afterwards,an increase in urinary output was observed,with restoration of normal fluid status,renal function and blood counts.At 2 months of age,after complete resolution of the abdominal and infectious condition,Eculizumab was started.Molecular analysis resulted as negative,but an increase in the amount of C5b9 on human microvascular endothelial cells was found during the acute phase.
Conclusions: Few reports have described the experience of using Eculizumab,and still less TPE in neonates,with aHUS.We would like to underline the efficacy of TPE with CARPEDIEM when Eculizumab is contraindicated.

P-145 EXPECTING THE (UN)EXPECTED - NEPHROTIC SYNDROME IN A CHILD WITH A HISTORY OF ACUTE MYELOID LEUKEMIA

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Introduction: Both nephrotic syndrome and acute myeloid leukemia (AML) are rare diseases in childhood. In most children the cause of nephrotic syndrome is minimal change nephropathy, however nephrotic syndrome is also a recognized manifestation of neoplastic diseases. Nephrotic syndrome associated with malignancy can appear prior to

onset, concurrently, or after diagnosis of malignancy. Although rare, the association of nephrotic syndrome and AML has been reported previously. In this report, we describe a patient with a history of AML who developed nephrotic syndrome.
Material and methods: A 5-year-old boy presented at our hospital with edema, abdominal distension and persistent proteinuria. He had a history of AML at 13 months of age, for which he was successfully treated with chemotherapy conform the Dutch-Belgian pediatric AML protocol and so far remained in complete remission. Initial laboratory evaluation in combination with edema showed the triad of nephrotic syndrome. In order to exclude recurrence of AML or other malignancies an invasive and extensive work-up was done prior to the start of nephrotic syndrome treatment to avoid masking malignancy and thereby a delay in final diagnosis.
Results: Complete blood count and white blood cell differential showed no abnormalities. Bone marrow biopsy and lumbar puncture indicated no signs of AML recurrence. PET-CT scan excluded other malignant causes. Renal biopsy was consistent with minimal change nephropathy. After excluding malignancy as secondary cause of nephrotic syndrome, standard oral prednisolone treatment was started and the patient went into remission within 3 weeks. At follow-up 1.5 years later, our patient is doing well, has remained in remission and shows no signs of recurrence of AML.
Conclusions: We report a child with two unrelated rare diseases in childhood. It is important to rule out various causes of nephrotic syndrome, especially in children with a history of malignancy, but it may turn out to be an idiopathic nephrotic syndrome.

P-146 THE TORTOISE AND THE HARE- A CASE STUDY OF HOW NXSTAGETM REDUCES THE RISK OF DIALSYS DISEQUILIBRIUM SYNDROME IN A HIGH RISK PAEDIATRIC PATIENT

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Introduction: Dialysis disequilibrium syndrome is a potentially life threatening complication usually associated with the commencement of haemodialysis (HD). However, paediatric patients and those with pre-existing CNS lesions remain at risk. We report the case of a dialysis dependent ten year old girl with a background of Spina Bifida and VP shunt who experienced headaches and generalized tonic-clonic seizures when dialysing using a conventional GambroTM AK200 dialysis machine.
Material and methods: The patient generally experienced symptoms after >3 h of dialysis. Seizures required rescue therapy and as a result, she was commenced on regular anticonvulsant medication before each HD session. Prior to starting dialysis, there was no history of headaches or seizures. We propose that this patient's symptoms were a result of disequilibrium syndrome. Various alterations to the patient's HD regimen to reduce the risk of disequilibrium were employed including shorter, more frequent dialysis sessions, cooling on the machine, a reduction in blood flow rate (and thus reduced rate of clearance of uraemic toxins including urea) as well as using smaller dialysers. Eventually the patient's self-reported and witnessed symptoms were successfully minimised by moving to the NxstageTM system with equitable clearance. The patient continued on NxstageTM HD for seven months until she was successfully transplanted. Managing intra-dialytic and post dialysis symptoms was vital in our patient for whom peritoneal dialysis would have likely failed for a multitude of reasons.
Results: We speculate that a combination of cooling and a slower rate of purification with the NxStage dialysis system was responsible for the improved tolerance to dialysis treatments compared with the GambroTM dialysis system. Thus remained true when the dialyser size was increased to improve the total treatment single pool Kt/V.
Conclusions: HD using the NxstageTM dialysis system appears to improve the tolerance to dialysis treatments in patients prone to intradialytic symptoms and dialysis disequilibrium syndrome.

P-147 AN UNUSUAL CASE OF ADIPSIC DIABETES INSIPIDUS

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Introduction: Objectives. To characterize hypernatremia in a suspicion of Rapid onset Obesity with Hypothalamic dysfunction, Hypoventilation and Autonomic dysregulation, Neuro-Endocrine Tumors (ROHHAD-NET) syndrome and to suggest a treatment.
Material and methods: We describe the case of a 6 year-old obese boy (60 kg, 135 cm) with a suspicion of ROHHAD-NET syndrome and a severe and unexplained hypernatremia.
Results: At admission, our patient presented with hypernatremia (154 mmol/L), a plasmatic hyperosmolarity of 332 mosmol/L and a hypotonic polyuria (207 mosmol/l) which was suspicious for diabetes insipidus (DI). After a water deprivation test, natremia increased to 160 mmol/l with a plasmatic osmolarity of 346 mosmol/L and, surprisingly, a urinary osmolarity of 535 mosmol/L, suggesting remaining endogenous vasopressin secretion. Plasmatic vasopressin values (1.47 pg/ml - 1.76 pg/ml) were low and inappropriate for natremia which ranged between 152 and 160 mmol/L, strengthening the diagnosis of partial central DI.
The absence of thirst despite severe hypernatremia suggests a specific type of DI known as adipsic DI. The hypothalamic dysfunction has been described in ROHHAD-NET syndrome. Our patient seems to have characteristics of both entities.
Desmopressin treatment resulted in a decrease of natremia to 146 mmol/L and urinary osmolarity increased to 822 mosmol/L. No genetic mutations in genes linked to monogenic obesity and pituitary deficiency including PCSK1 were identified.
Conclusions: Hypernatremia seems to be a component of ROHHAD-NET syndrome, even if the pathophysiological mechanism has not yet been identified. We report the case of a boy with a condition compatible with ROHHAD-NET syndrome and central partial adipsic DI. Oral desmopressin treatment and free water access improve natremia and potential neurologic damage. Careful attention should be paid concerning the narrow gap between benefits and potential risks of desmopressin treatment and free water intake in such conditions.

P-148 NPHS2-ASSOCIATED STEROID-RESISTANT NEPHROTIC SYNDROME IN A BOY WITH STEROID SULFATASE DEFICIENCY-X-LINKED RECESSIVE ICHTHYOSIS

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Introduction: Steroid-resistant nephrotic syndrome (SRNS) is a genetically heterogeneous glomerulopathy progressed to end-stage of renal disease (ESRD) during childhood or adolescence. X-linked ichthyosis is a rare inherited disorder due to steroid sulfatase deficiency (MIM #308100). Very few cases of SRNS associated with X-linked recessive ichthyosis were described, but molecular genetic basis of SRNS was not studied yet.
Material and methods: We report on a 11-year-old boy presenting with SRNS since age of 7 years with an underlying ichthyotic skin since birth.
Results: The boy with SRNS and early-onset ichthyosis was referred to our clinic for further evaluation. On admission he had proteinuria (1.2 g/

d), hypoalbuminemia (27 g/L) without oedema, normal blood pressure (110/60 mmHg) and kidney function (eGFR 176.5 mL/min/1.73 m^2). The boy had also severe hyperkeratosis. Renal ultrasound showed enlarged kidneys with bilateral diffuse hyperechogenicity of parenchyma. Kidney histopathology revealed focal segmental glomerulosclerosis with moderate interstitial fibrosis and tubular atrophy. Diffuse foot process effacement in podocytes was described by electron microscopy. Targeted next-generation sequencing covering 68 genes implicated in SRNS identified known heterozygous compound mutations c.890C > T (p.Ala297Val, rs199506378) in exon 8 and c.686G > A (p.Arg229Gln, rs61747728) in exon 5 in the *NPHS2* gene, associated with of autosomal recessive SRNS, type 2 (MIM #600995). Copy number variation analysis revealed novel hemizygousdeletion in the *STS* gene on chromosome Xp22.31 (6527321–8,088,112)×0 (Z-score < −3.2), confirming the diagnosis of X-linked recessive ichthyosis (MIM #308100). Therapy with ACE inhibitors was administered to the patient to prevent potential progression to ESRD. After 2 years of follow-up the boy had the same extent of proteinuria (1.5 g/d), and his renal function remains unimpaired (eGFR 123.5 ml/min/1.73 m^2).

Conclusions: We presented the case of *NPHS2*-associated SRNS in a boy with steroid sulfatase deficiency-X-linked ichthyosis caused by *STS* mutation. Digenic inheritance of *NPHS2* and *STS* mutations in this case might have modifier effects with unclear molecular pathogenicity causing an uncommon clinical phenotype.

P-149 ANOTHER ATYPICAL CASE OF ACUTE KIDNEY INJURY-OR NOT?

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Introduction: A 12-year-old boy presented at the pediatric department with persistent non bilious vomiting since 3 days and anuria.

Material and methods: There was no diarrhea nor other symptoms. His medical history was negative besides episodes of vomiting during periods of stress from the age of 7 years. At physical examination moderate dehydration was seen.

Results: His laboratory results showed metabolic alkalosis and elevated serum creatinine (1.7 mg/dl) and BUN (123 mg/dl) with hyperphosphataemia and hyperparathyroidism. Urinalysis was normal. Ultrasound showed enlarged kidneys with increased cortical echogenicity. After IV rehydration he recovered with normalization of diuresis and cessation of vomiting. Hyperphospataemia and hyperparathyroidism together typically are more associated with CKD than AKI and the ultrasound image was atypical for acute tubular necrosis. A renal biopsy was performed which showed normal glomeruli but tubules filled with amorphous material consistent with the microscopic presentation of Hyperoxaluria.

Conclusions: He was suspected of primary Hyperoxaluria and treatment was commenced. Genetic testing for primary Hyperoxaluria was negative. We noticed that his growth chart showed stunted growth from the age of 9 years and he confirmed to having episodes of vomiting almost every night for the past 2 years. A hypotonic duodenography showed subobstruction at the duodeno-jejunal junction which was confirmed at gastroscopy where a complete torsion of the distal duodenum and stomach traction was seen. On laparascopy a malrotation with chronic volvulus without vascular compromise were seen and corrected. Hyperoxaluria was thought to be secondary to the chronic intermittent intestinal obstruction with malabsorption. Enteric hyperoxaluria is responsible for 5% of cases of hyperoxaluria and is seen secondary to (fat) malabsorption with increased enteric oxalate absorption. It is usually secondary to conditions such as intestinal surgery, bacterial overgrowth syndrome and Inflammatory Bowel Disease. After surgery his growth improved with cessation of vomiting and serum creatinine settling around 0.9 mg/dl (CKD stage 2).

P-150 REFRACTORY HYPERTENSION AND ARTERIAL STIFFNESS IN A TODDLER WITH NEUROFIBROMATOSIS TYPE 1

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Introduction: Arterial hypertension due to renal artery stenosis or mid-aortic syndrome is a common finding in patients with Neurofibromatosis (NF) type 1 (20%). Arterial stiffness is used to describe the elasticity of the arteries, it reflects the procedure of aging on the vascular system and is associated with increased cardiovascular risk, chronic renal disease and arterial hypertension. It can be assessed non-invasively through the determination of the carotid femoral pulse wave velocity (cf-PWV). The purpose of this case is to describe the correlation between arterial stiffness and arterial hypertension as documented with office Blood Pressure (BP) levels and ambulatory BP levels.

Material and methods: We report a case of refractory hypertension accompanied with increased arterial stiffness in a 4 year-old girl with NF type 1 and mid-aortic syndrome. She presented initially with office BP levels below the 90th percentile but with a routine ambulatory blood pressure monitoring (ABPM) with a nondipping profile. The patient underwent percutaneous transluminal angioplasty (PTA). Office BP presented a decrease at 4 months after PTA. In constrast, ambulatory BP levels increased and were accompanied by an increased in cf.-PWV. After 8 months renal function was improved, the hypertention persisted, but there was a significant reduction of ambulatory BP levels, which agrees with the reduction in cf.-PWV values, despite sustained office BP elevation.

Results: Masked hypertension may be present and a nondipping profile may enable the early diagnosis of arterial hypertension. There seems to be a superiority of ABPM in the assessment and follow up of hypertension and a better correlation to arterial stiffness.

Conclusions: Renovascular hypertension may have adverse effect on arterial stiffness. Regular assessment of BP using ABPM may enhance the medical care of patients with NF type 1 and guide their further Management.

P-151 BK-VIRUS NEPHRITIS IN PEDIATRIC PATIENT AFTER HEMATOPOIETIC STEM CELL TRANSPLANTATION: SERIAL OF CASES

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Introduction: Renal dysfunction in hematopoietic stem cell transplantation (HSCT) recipient traditionally has been attributed to use of nephrotoxic drug. BK-virus nephritis (BKVN) can remain undiagnosed in these patients due to lack of screening for BK-infection and to its nonspecific clinical picture.

Material and methods: The cases of BKVN after HSCT were reviwed.

Results: Pt 1. Six-year-old girl with multilinear myelodysplasia presented with the rising of serum creatinine (from 58 to 120 μmol/l) on 13 mo after HSCT. One month before renal dysfunction she had pulmonitis of unknown etiology. The girl had stem cell transplant's hypofunction and

secondary immunodeficiency and was treating with Methotrexate (0.6 mg/kg/week), Cyclophosphamide (200 mg/m^2/week) because of chronic graft-versus-host disease (GVHD). Urinalysis and renal US were normal. BK-virus PCR analysis revealed 6.7×10^6 copies/ml in urine. A renal biopsy showed tubulointerstitial injuary with many tubular intranuclear inclusions and positive immunostainig for SV-40. Immunosuppression was discontinued, therapy with IVIG 1 g/kg, Cidofovir 0.5 mg/kg/week, Leflunomide 10 mg/day was started. Despite BK eradication there was no improvement in renal function after 2 mo of treatment. To date she has stable renal insufficiency: Cr_s = 128 μmol/l, eGFR =26 ml/min.

Pt 2. Eight months after HSCT due to refractory T-cell acute lymphoblastic leukemia ten-year-old boy had an episode of encephalitis with extrapontial myelinosis (etiology?) and serum creatinine level increasing (from 38 to 180 μmol/l). He received Prednisone 2 mg/kg/day and Prograf 0,015 mg/kg/day because of GVHD. Urine BK-virus titer was 2×10^6 copies/ml. A renal biopsy revealed tubulointerstitial nephritis with positive immunostainig for SV-40. Immunosuppression was discontinued, therapy with IVIG 1 g/kg, Leflunomide 10 mg/day, Ciprofloxacin 20 mg/kg/day was started. During 2 years of observation the boy had stable renal dysfunction: eGFR =60 ml/min.

Pt 3. The 22-year boy with acute myeloblastic leukemia (M1/M2) presented with renal impairment: Cr_s rose from 54 to 202 μmol/l 12 months after of HSCT. He was on treatment with Prednisone 0.5 mg/kg/day, CyclosporineA 200 mg/day and Bortezomib (Velcade) 0.4 mg/kg/week due to skin form of GVHD. The boy had secondary immunodeficiency and recurrent CMV-virus infection. The renal dysfunction was preceded by hemorrhagic cystitis. Urine BK-virus titer was 12×10^3 copies/ml, the renal biopsy confirmed BKVN. Therapy with IVIG 1 g/kg, Cidofovir 0.5 mg/kg/week, Leflunomide 10 mg/day and Ciprofloxacin 250 mg/day was started. There was no recovery of renal function; the boy had stable eGFR about 30–35 ml/min during 2 years of observation.

Conclusions: We suppose that the actual incidence of BKVN after HSCT is much higher than what is commonly thought. Our clinical cases demonstrate that BK-virus screening and renal biopsy would be performed in all non-renal transplant and immunosuppressed patients with unexplained rising of serum creatinine. These patients may benefit from early detection and treatment of BKVN and modification of immunosuppressive therapy.

P-152 URETHRAL DUPLICATION IN A FEMALE CHILD

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Introduction: Urethral duplication in girls is a very rare congenital pathology. Successful surgical reconstructive treatment heavily depends on appropriate clinical examination and evaluation of anatomy.

Material and methods: We describe a case of successful treatment of type II urethral duplication according to Stephens in a 2.5-year-old girl.

Results: Several fever episodes in infancy, recurrent urinary tract infections (UTI) since the age of 1 year. Daytime incontinence from 2 years, following complete disability to control urination. A spherical formation in the region of the labia, collapsing after outflow of urine. Referred to the The Hospital of Lithuanian University of Health Sciences Kauno klinikos at 2.5 years

Examination: height - 25 ‰, weight - 3 ‰, deep eye sockets, wide nasal bridge, broad shoulders, large abdominal girth, hypertrophic clitoris. XX karyotype, normal kidney function. UTI (Enteroccocus faecalis) was diagnosed.

Kidney ultrasonography: bilateral ureterohydronehrosis, megacystis. Post void residual - 115 mL.

MAG-3: bilateral obstructive curves with slow excretion after furosemide infusion (accumulation of radioisotope media in diluted ureters). Right kidney function - 71% of total kidney function, left kidney - 29%.

Cystograms - left kidney V° VUR.

Gynecologist, endocrinologist: clitoral hypertrophy.

Cystoscopy: large trabeculated urinary bladder, ureter foramina not open, two urethrae - one opening into the clitoris, the other opening normally into the perineum.

Surgery was performed: 0.5-cm fragment of urethra removed in the area of the clitoris, clitoroplasty, Foley catheter inserted. Microscopy analysis of the segment: visible cavernous bodies making up the clitoris, urethra lined with urethral epithelium.

The Foley catheter was removed after 4 days, urinary bladder catheterization every 3 h due to valvelike bladder. The patient was no longer incontinent, ultrasonography revealed subsiding urostasis.

Conclusions: Urethral duplication is a rare pathology in girls, requiring thorough physical examination and testing.

P-153 GLOMERULOPATHY WITH FIBRONECTIN DEPOSITS: A RARE BUT POSSIBLE CAUSE OF PROTEINURIA IN PAEDIATRIC AGE

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Introduction: Glomerulopathy with fibronectin deposits (GFND) is an extremely rare autosomal dominant disease with age-related penetrance, characterized by proteinuria, microscopiche ematuria, hypertension and massive fibronectin deposits in the mesangiumand subendothelial space, leading to end-stage renal failure. It usually develops in adulthood. Only two cases with childhood onset have been described in literature so far.

Material and methods: We report the case of a two year-old male child who presented with mild proteinuria, normal kidney function and mild increase of creatine kinase and lactated hydrogenase plasmatic levels. The physical and neurological examination showed short stature, rhizomelia and speech delay.

Results: Given the presence of persistent proteinuria, a targeted resequencing analysis of 27 genes related to nephrotic syndrome was performed and a novel missense variant, c.341G > T (p.Arg114Leu) in the FN1 gene, associated with GFND, was found. The pathogenicity of this variant is supported by its absence in any public database and in silico predictions with a DANN Score of 0.9987 (Varsome). However, a kidney biopsy is still required to confirm the diagnosis by showing the presence of fibronectin deposits.

Conclusions: GFND should also be considered in the differential diagnosis of persistent proteinuria in pediatrics age, even if it is rarely described in childhood. Accordingly, molecular analysis of FN1 gene should be included in the genetic work-up of persistent proteinuria.

P-154 FAMILIAL HYPOMAGNESEMIC HYPERCALCEMIC NEPHROCALCINOSIS IN A PATIENT WITH DIFFUSE MEDULLARY NEPHROCALCINOSIS

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Introduction: Renal tubuler disorders are a group of diseases that can occur with different clinical findings.Familial hypomagnesemia with hypercalciuria and nephrocalcinosis is a rare autosomal recessive renal disease characterized by tubular disorders. It is caused by mutations in the tight junction structural proteins claudin-16 or claudin-19, which are encoded by the CLDN16 and CLDN19 genes, respectively. Patients exhibit excessive wasting of calcium and magnesium, nephrocalcinosis, chronic kidney disease, and early progression to end-stage renal failure during infancy.
Material and methods: A six year-old boy presented with fever and abdominal pain. In his past medical history, he had recurrent urinary tract infections, kidney stones and chronic renal failure. There was no consaguinity between his mother and father. There were a large number of kidney stones in his family. Increased serum creatinine and decreased magnesium levels were detected in blood biochemistry.The urinal ultrasonography showed diffuse medullary nephrocalcinosis and multiple stones in both kidneys and ureter. The right ureteral stone causing complete obstruction was removed. Hypercalciuria, hypermagnesuria, and hypocitraturia were detected in his urinary examinations. Eye examination was normal.
Results: C113Y mutation was detected in the CLDN-16 gene analysis sent by FHNCC. Supportive treatments and potassium citrate was started.
Conclusions: Serum and urine magnesium examinations must be performed in children who are admitted with nephrocalcinosis and / or nephrolithiasis in childhood. In chronic renal diseases with unknown etiology, which are associated with nephrocalcinosis and nephrolitiasis, FHNCC should come to mind.

P-155 HEPATIC MANIFESTATIONS AT THE ONSET OF SYSTEMIC LUPUS ERYTHEMATOSUS: CASE REPORT

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Introduction: Multi-organ involvement is the hallmark of systemic lupus erythematosus (SLE). Clinically significant hepatic disease is generally regarded as unusual in SLE, but studies showed that hepatic disease may be more common in SLE that was initially thought. Hepatic disease is not a significant cause of morbidity and mortality, but subclinical liver involvement is common. We aimed to determine if whether to classify our patient as having a primary liver disease with associated autoimmune features or having liver disease as a manifestation of SLE.
Material and methods: We investigated a 14 years old boy who presented with one-year history of elevated serum aminotransferase and complaints of daily fever in the last month, polyarthralgia, swelling of the right knee, hepatosplenomegaly. Family history was negative for autoimmune or inherited liver disease, including Autoimmune hepatitis (AIH) and SLE.
Results: SLE was confirmed by 4 clinical and 3 immunological criteria (according to SLICC Classification Criteria). Liver biopsy showed chronic hepatitis with discreet inflammatory activity, no fibrosis, no interface hepatitis, no fatty infiltration, so no specific elements were identified. Renal biopsy revealed class IIIC kidney disease.
Conclusions: The difference between the hepatic involvement in SLE and AIH has not been clearly defined due to similarities in the clinical and biochemical features. Lupus hepatitis is a SLE-related liver dysfunction and it has been described as hypertransaminasemia owing to the fluctuations in the levels of alanine transaminase that are consistent with the activity of SLE. We consider that our patient has SLE and lupus hepatitis. Children with liver dysfunction and SLE should be investigated for Autoimmune Hepatitis (AIH) as these two entities can occur together and their complications are different. AIH may lead to end-stage liver disease, while SLE may result in end-stage renal disease. Liver biopsy might be necessary to establish the diagnostic in SLE patients with persistent increase of liver enzymes.

P-156 A COLLAPSING GLOMERULOPATHY IN A CHILD WITH BARDET-BIEDL SYNDROME

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Introduction: Bardet-Biedl syndrome (BBS) is a multisystem disorder due to a ciliopathy with autosomal recessive inheritance. The cardinal features are pigmentary retinopathy, central obesity, postaxial polydactyly, mental retardation, hypogonadism and renal abnormalities.Renal involvement with progressive deterioration in renal function was recognized as a major cause of morbidity and mortality. Different renal histological pictures were reported in BBS including tubulo-interstitial lesions, focal segmental glomerulo- sclerosis and renal cysts. To our knowledge this is the first reported case of collapsing glomerulopathy with progressive deterioration in renal function in a child with BBS.
Material and methods: A 9 year old girl with BBS presented with nephrotic range proteinuria and advanced chronic kidney disease. Her serum creatinine was 169 μmol/l (serum creatinine value recorded 6 months back was 79 μmol/l) giving estimated glomerular filtration rate of 23mls/min/1.73m^2 and urine protein/creatinine ratio was 467 mg/mmol. Serum albumin was 27 g/l and serum cholesterol was high.Examination revealed minimal oedema with elevated blood pressure (146/102 mmHg).
USS revealed normal renal sizes with markedly increased cortical echogenicity with loss of cortico-medullary demarcation. Her renal biopsy showed only 1/23 of viable glomeruli and 22/23 glomeruli with tuft collapse and peri-glomerular fibrosis. She was treated with ACE inhibitors and calcium channel blockers.
Results: After 6 months of treatment her urine protein/creatinine ratio has reduced to 87 mg/mmol. Last recorded blood pressure was 112/76 mmHg. But serum creatinine has increased to 184 μmol/l.
Conclusions: Children with BBS are at higher risk of renal impairment. They can present with nephrotic range proteinuria and ACE inhibitors are effective in decelerating the progression. Collapsing glomerulopathy is a rare histological sub type in BBS which could be associated with adverse prognosis.

P-157 A CONGENITAL DISEASE MIMICKING CONGENITAL NEPHROTIC SYNDROME

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Introduction: Primary lymphatic dysplasia (PLD) is a hereditary developmental abnormality of the lymphatic system, resulting in primary lymphedema. Some of the patients have generalized edema and hydrops fetalis.
We report a case of a female patient with PLD with systemic involvement presented as nephrotic syndrome.
Material and methods:
Results: A three months old girl referred with generalized edema with severe abdominal distension and hypoalbuminemia. She was born to consanguineous parents at 34 weeks of gestation due to hydrops fetalis diagnosed at 3rd trimester. The patient was treated in neonatal intensive care unit for two months because of respiratory distress and sepsis. Physical examination showed abdominal distension due to ascites fluid,

generalized edema including severe genital edema. Laboratory investigation revealed: serum albumin: 2.9 mg/dl, 24 h urine protein: 15 mg/m^2/h. Renal functions, serum electrolytes, complement 3 and antinuclear antibody were normal. Metabolic screening tests and viral serology including TORCH, hepatitis B and C virus were negative, ascites fluid was serous. During the follow-up, serum albumin level of the patient was remained between 2.6–2.9 g/dl and she did not need regular albumin infusion. Based on clinical and laboratory findings, lymphatic edema was considered in differential diagnosis. Lymphoscintigraphy confirmed diagnosis and revealed primary lymphatic dysplasia.
Figure 1. Lymphoscintigraphy of the patients
Conclusions: Primary lymphatic dysplasia should be kept in mind in the differential diagnosis of the patients presented as nephrotic syndrome without massive proteinuria and mild, stabile hypoalbuminemia. Generalized edema and hydrops fetalis may develop in these patients. Lymphoscintigraphy is widely considered the main investigative tool for establishing the diagnosis of PLD.

P-158 INFANTILE NEPHOCALCINOSIS (NC): SEEK AND YOU SHALL FIND!

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Introduction: NC in children recognizes many causes, both endogenous and exogenous /iatrogenic nature (eg. Vit D intoxication, diuretics). Among the endogenous early onset causes there are inborn errors of metabolism including primary hyperoxaluria (PH).
Material and methods: We describe the case of a two months and half infant with bilateral renal medullary hyperechogenicity we found occasionally during hips ultrasonography.
Results: At the beginning, the patient showed good clinical conditions. The most common causes of nephrocalcinosis, including tubulopathy were excluded since we found good renal function, negative inflammatory markers, normal electrolytes, vitamin D and PTH. Urinary electrolytes-metabolites, proteinuria and amminoaciduria were normal too. Renal ultrasonography confirmed the presence of renal medullary hyperechogenicity. Computed tomography angiography showed multiple calicealhyperdense images. In order to find inborn errors of metabolism we dosed urinary organic acids which documented an increased level of oxalic acid and glycerine. In the suspect of type 2 PH, molecular analysis was carried out and confirmed the diagnosis. A hydropinic therapy was started continuing follow-up. Subsequent ultrasound evaluations documented the evolution towards a clear picture of nephrolithiasis. Actually the patient presents normal kidney function with regular height-weight growth.
Conclusions: Type 2 PH is an extremely rare cause of nephrocalcinosis not so different from type 1; for that reason molecular analysis assumes a discriminating role. The importance of an early diagnosis derives from the possibility to start an early hemodialysis treatment that, in some cases, may prevent the rapid evolution of the pathology towards the widespread nephrolithiasis characterized by terminal renal insufficiency, systemic oxalosis and multiorgan damage.

P-159 DOES THE ULTRASOUND OF THE URINARY TRACT HAVE ITS PLACE IN THE TREATMENT OF EARLY NEONATAL JAUNDICE?NEONATAL BILATERAL ADRENAL HEMORRHAGE:CASE REPORT

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Introduction: Adrenal hemorrhage is a rare clinical entity in the neonatal period, with an incidence of 1.7–2.1 / 1000 births. It is more often diagnosed on the right side whilst bilateral hemorrhage occurs in 10–15% of cases.
Material and methods: Clinical presentation shows a wide range of symptoms, from the signs of adrenal insufficiency to asymptomatic course of illness with incidental finding of changes on tests. Neonatal jaundice due to hemolysis of hemorrhagic content often is an accompanying sign. We present a male neonate born at term, with the early neonatal jaundice of unknown cause and without evidence of perinatal infection. Ultrasound of the urinary tract was made and it was seen a hypoechogenic formation in the upper poles of both kidneys, confirmed with magnetic resonance imaging (MRI) of the abdomen.
Results: Clinical and laboratory test results showed no signs of adrenal insufficiency. There was no confirmation of embryonic tumor or neuroblastoma.
Conclusions: Ultrasound of the urinary tract, as an available and non-invasive test, has its place in the treatment of early neonatal jaundice of unknown cause. With ultrasound monitoring of regression of formation a further invasive treatment and unnecessary laparotomy can be avoided.

P-160 UNEXPLAINED THROMBOCYTOPENIA AS THE FIRST MANIFESTATION OF A SYSTEMIC DISEASE – A CASE REPORT

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Introduction: Immune thrombocytopenia (ITP) can be triggered by a viral infection or another immunologic mechanism. The management and study of this pathology might reveal an association with several underlying systemic diseases.
Material and methods: NA.
Results: A previously healthy, nine-year old boy, son of a non-consanguineous healthy couple, presented to the emergency department with intermittent fever over the course of seven days, petechial rash and bruises on his legs. The platelet count was 63,000/μL, with normal hemoglobin and leucocyte count. Investigation showed frankly positive direct antiglobulin test, elevated erythrocyte sedimentation rate and positive antinuclear antibodies (ANA), lupus anticoagulant test, anticardiolipin antibodies and anti-beta2-glycoprotein-I. These tests continued to be positive 12 weeks after the presentation. During follow-up, he continued to present sporadic but spontaneously resolving petechial rash. Four years after presentation, he was found to have cutaneous vasculitis on his hand and a positive anti-double-stranded deoxyribonucleic acid antibody, thus meeting enough criteria for the diagnosis of systemic lupus eritematosus (SLE) with antiphospholipid syndrome. Treatment with hydroxychloroquine (6 mg/kg/day) and aspirin was initiated. Two years after SLE diagnosis, he was started on oral prednisone due to the finding of active urinalysis and elevated serum creatinine (1.07 mg/dL). These parameters normalized and no renal flare occurred since then. Renal biopsy revealed mesangial proliferative lupus nephritis (class II). Recently on ophthalmologic examination a retinopathy in bull's eye was observed and hydroxychloroquine was timely interrupted.
Conclusions: In 15% of cases, ITP might be the first manifestation of SLE, antedating this diagnosis. The authors emphasize the importance of monitoring closely children with positive ANA and ITP, in order to allow the early identification of SLE manifestations. Bull's eye retinopathy is rare

and its risk increases with high cumulative dosis of hydroxychloroquine. Nonetheless, in this patient case, it is not totally clear if it represents another manifestation of SLE rather than an iatrogenic consequence.

P-161 FAMILIAL LABIAL ADHESION

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Introduction: Labial adhesion, also known as labial fusion, labial synechiae or labial agglutination, is the partial or total fusion of labia minora in prepubertal girls, most commonly in 3 months - 3 years age group. The condition may be asymptomatic or result in urinary tract infection (UTI), postvoid dripping or obstruction. Although the etiology is not clear, vulvar irritation, poor hygiene or excessive cleaning may be responsible. Familial labial adhesion has not yet been reported in the literature.
Material and methods:
Case report: We present a three year old girl evaluated for recurrent UTIs and found to have labial fusion that did not disappear with topical estrogen. Surgical separation was needed after which UTIs stopped. Her 16 months old sister also had labial fusion without any symptoms that disappeared with topical estrogen. We do not know if this is a coincidence or not.
Results:
Discussion: Maternal estrogen insufficiency was previously believed to cause neonatal labial adhesions which disappear in puberty. However estrogen levels of children with and without labial adhesions were compared in a study demonstrating no difference, so hypoestrogenic theory is no longer accepted. Familial labial adhesion cannot be thought as an analogous of androgen insensitivity syndrome seen in familial hipospadias. As labial adhesions disappear with intense estrogen treatment, it may be speculated that there may be estrogen insufficiency or estrogen insensitivity in local tissues of these children. Androgen insensitivity is defined for testosterone but there is not a study concerning estrogen. Association of labial adhesion and familial phimosis that is not physiological is also not defined in literature too. As the presented sisters do not have a brother, we are not able to query about this. The mother of these children may had similar complaints in childhood but this is not stated by her. As a result, labial adhesion in two sisters has brought in mind a possible familial predisposition of the condition.
Conclusions:

P-162 CARBOPLATIN DOSING IN CHILDREN USING ESTIMATE GLOMERULAR FILTRATION RATE: EQUATION MATTERS

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Introduction: Renal function-based carboplatin dosing results in more consistent drug exposure than flat dosing. We aimed to validate the Newell dosing equation using estimated glomerular filtration rate (GFR) and study which renal function marker most accurately predicts carboplatin clearance in children.
Material and methods: In 30 children with a wide spectrum of solid tumours, 78 carboplatin clearance values were obtained from individual fits using NONMEM. Observed carboplatin clearance was compared with predicted clearance calculated according to the Newell dosing equation using three different GFR estimates, one creatinine-based (eGFR-Schwartz), one cystatin C-based (eGFR-CKiD1) and one based on creatinine and cystatin C (eGFR-CKiD2). Bias and precision of the predictions was examined.
Results: Both CKiD equations were accurate with a bias of 1.7 (95% CI -1.7 to 5.1) and −3.3 (95% CI -7.0 to 0.35) ml/min for respectively eGFR-CKiD1 and CKiD2, whereas the bias of eGFR-Schwartz significantly differed from zero (−16.2; 95% CI -21.5 to −10.9 mL/min). eGFR-CKiD1 gave the lowest bias and imprecision, the other two eGFR equations showed overprediction of carboplatin clearance as reflected by negative bias and higher mean prediction error values. The proportion of variance in observed clearance that can be explained by the predicted clearance was lowest for Schwartz (R^2 = 0.58), the explained variance was 0.65 for both CKiD equations.
Conclusions: The two cystatin C-based CKiD equations outperform the widely used creatinine-based Schwartz equation in predicting carboplatin clearance. We recommend the use of estimated GFR based on cystatin C for carboplatin dosing in children unless a gold standard GFR measurement is available.

P-163 STEROID TREATMENT REDUCTION IN RELAPSING CHILDHOOD NEPHROTIC SYNDROME: A NEW NATION-WIDE RANDOMIZED CONTROLLED TRIAL IN THE NETHERLANDS - THE RESTERN STUDY

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Introduction: Nephrotic syndrome is the most common glomerular disorder in childhood. Corticosteroids are the first-line treatment for nephrotic syndrome in children as over 80–90% of patients achieves complete remission after prednisolone treatment. Yet, over 80% experience one or more relapses, necessitating repeated courses of corticosteroid therapy. This exposes patients to severe side effects and long term complications. No randomized controlled trials are available to determine the optimal corticosteroid treatment of an infrequent relapse of nephrotic syndrome. Recent studies show that treatment schedules for the first episode can safely be reduced. The hypothesis of the REducing STEroids in Relapsing Nephrotic syndrome (RESTERN) study is that a 4-week reduction of alternate day steroids is effective and safe, reduces steroid exposure by 35% on average, and is therefore preferable.
Material and methods: The RESTERN study is a nation-wide, double-blind, randomized, placebo controlled, noninferiority intervention study. Children aged 1–18 years with a relapse of steroid sensitive nephrotic syndrome (n = 144) are randomly assigned to either the current standard therapy in the Netherlands (prednisolone daily until remission, than 6 weeks on alternate days) or a reduced prednisolone schedule (prednisolone daily until remission, than 2 weeks on alternate days, followed by 4 weeks of placebo on alternate days).
Results: The primary endpoint of the RESTERN study is the time to the first relapse. The secondary end points are the number of relapses, progression to frequent relapsing or steroid dependent nephrotic syndrome and the cumulative dosage of prednisolone during the study period of 48 months follow-up.
Conclusions: The results of the RESTERN study may provide evidence-based recommendations for national and international guidelines to treat children with relapsing nephrotic syndrome. If corticosteroid exposure could be reduced, this would reduce toxicity of prednisolone and thereby decrease the side effects and long-term complications associated with corticosteroid therapy in children with relapsing nephrotic syndrome.

P-164 ULTRASONOGRAPHIC ASSESSMENT OF KIDNEY LENGTH IN HEALTHY FULL-TERMED TWIN INFANTS COMPARED WITH SINGLETON INFANTS

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Introduction: To compare kidney lengths, birth lengths and weights of healthy full-termed twins with singleton infants.
Material and methods: Utrasonographic measurements of kidney lengths were performed on 1073 on time-born singleton infants (2146 healthy kidneys, 549 boys, 524 girls) (group A) and 87 twin infants (174 healthy kidneys, 46 boys, 41 girls) (TIG). Birth lengths and weights were measured in 309 singleton infants (167 boys, 142 girls) (group B) and in TIG and compared. Birth lengths were analyzed by T-test, while birth weights by Mann-Whitney U test. The average, minimum and maximum values (5% and 95% CI) and standard deviations were calculated by statistical program SPSS 16.
Results: 4 different nomograms of kidney lengths in singletons and twins were presented according to the gender and side. Left and right kidneys were significantly longer in males than in females ($p < 0.05$) between the 2nd-6th months of life, but not between 7th–12th months of life ($p > 0.05$). Left kidneys were significantly longer than right kidneys in both sexes in the first year of life ($p < 0.05$). No significant difference in the kidney length existed between TIG and group A. Arithmetic mean birth length was 2 cm higher in males from group B than in TIG ($t = 6.4$; p<0,001), while it was 2.4 cm higher in females in group B than in TIG ($t = 6.4$; p<0,001). Median birth weight was 620 g higher in males from group B than in TIG (z = 6.9; p<0,001), while it was 800 g higher in females from group B than in TIG (z = 7.3; p<0,001).
Conclusions: We present four nomograms of kidney size in the first year of life. Although the birth lengths and weights were significantly lower in twins than in singletons, kidney lengths were equal in both groups. Therefore, the same nomograms are useful for measurements of kidney length in both groups.

P-165 THE ROLE OF NEU1 GENE İN THE ETIOPATHOGENESİS OF HENOCH SCHÖNLEİN VASCULİTİS AND ITS RELATİONSHİP WİTH RENAL İNVOLVEMENT

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Introduction: Henoch-Schönlein Vaskulitis (HSV) is a small vessel vasculitis which is seen common in pediatric population.The incidence is 14–18 / 100.000 children per year. Renal involvement is the determinant factor in prognosis. Because it is known that neurominidase 1 (NEU1) gene, which is cellular lysosomal sialidase and responsible for the sialization of IgA molecule and the defects in the sialization steps cause IgA accumulation in the vascular wall, we investigated expression of NEU1 gene in our HSV-diagnosed patients and tried to clarify its role in the disease formation.
Material and methods: Fifty patients with HSV renal involvement and 50 healthy control groups were included in this study. Patients were evaluated for demographics and NEU1 gene mutation. NEU1 gene analysis was performed by the PCR method.
Results: Forty-nine comma 4 % of the patients were male and 50.6% were female. Seventy comma one percents' had renal involvement at different grades. The average age of the patients was 10.21 (± 3.95). The 40 % of control group was male and 60% was female. The average age of the control group was 11.24 (± 4.16). At the time of application, 41.4% of the patients had abdominal pain, 95.4% had rash, 2.3% had edema and 31% had arthritis / arthralgia. Patients were most frequently diagnosed during the autumn season with %31. Patients' 21.8% have passed the URI in the last two weeks. Presence of hypertension at the time of diagnosis was 6.9%. Kidney involvement was detected in 60.9% of patients, GIS involvement in 16.1% and scrotal involvement in 1 person. None of the patients had any abnormality in renal function tests and coagulation tests. Significant proteinuria was present in 66.7% of the patients and hematuria was detected in 73.6% of the patients. Skin biopsy was performed and all of the patients were found compatible with leukocytoclastic vasculitis. Renal biopsy was performed eleven patients and all of them were detected compatible with IgA nephropathy. When the biochemical laboratory parameters was evaluated between the patients groups and control group, no significant difference was found. The urine findings of the patients with renal involvement was significantly different from those without renal involvement ($p < 0.001$). No mutations were detected in the NEU1 gene in all HSV kidney-affected patients and the control group.
Conclusions: Neu1 gene mutation was not detected in Henoch-Schönlein vasculitis patients with renal involvement. In this study, there was no link between HSV kidney involvement and the NEU1 gene, which is a lysosomal sialidase and involved in the sialization of IgA.

P-166 HAEMODIAFILTRATION (HDF) IS ASSOCIATED WITH SUPERIOR FLUID CONTROL AND REDUCED CARDIOVASCULAR RISK PROFILE COMPARED TO CONVENTIONAL HAEMODIALYSIS (HD) – DATA FROM THE HDF VS HD (3H) STUDY

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Introduction: Fluid overload, hypertension and cardiovascular disease are common in children on dialysis. In adults, HDF is shown to reduce cardiovascular mortality, but causes for this are not clear and data in children are scarce.
Material and methods: We analysed the baseline data in prevalent dialysis patients from the HDF vs HD (3H) study to assess fluid status, BP and cardiovascular measures on HD and HDF.

Results: Of the 179 children (from 28 centres in 10 European countries) in the HDF vs HD study 69 were prevalent dialysis patients, 35 on HD and 34 on HDF, and are further described here. There was no difference between HD and HDF groups in age, gender, underlying renal disease, dialysis vintage (median 3.4 months), type of vascular access (AVF in 34 vs 29% on HD and HDF), blood flow or presence of residual renal function. On bioimpedance spectroscopy children on HDF were less likely to have fluid overload compared to HD (Rel OH 4.2 vs 9.8%; $p = 0.016$), and had lower 24 h mean arterial pressure (MAP, 93 vs 87.5 mmHg; $p = 0.04$). Although there was no difference in interdialytic weight gain, children on HDF required fewer rescue sessions (4.1% HDF vs 19.9% HD; $p = 0.008$). The height-adjusted pulse wave velocity-SDS was lower in HDF vs HD (0.09 vs 1.33; $p < 0.0001$) and correlated positively with MAP, ultrafiltration and the blood flow rate. HDF patients had a lower left ventricular mass index (30.7 vs 43.5; $p = 0.001$). Incident dialysis patients from the same centre did not demonstrate a difference in Rel OH status or MAP between HD and HDF groups, suggesting that centre bias is unlikely and dialysis modality significantly influences the fluid status.
Conclusions: Children on HDF have improved fluid control and cardiovascular measures compared to those on HD, and this effect is seen even with a short dialysis vintage of 3.4 months.

P-167 IMPROVED LEFT VENTRICULAR STRUCTURE AND FUNCTION IN CHILDREN ON CHRONIC HAEMODIALYSIS: A LONGITUDINAL STUDY

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Introduction: Our aim in this study was to examine longitudinal changes in left ventricular (LV) structure and function and to evaluate factors associating with LV adaptation in children on chronic haemodialysis.
Material and methods: Retrospective longitudinal study in a paediatric dialysis centre over past 4 years. Children on chronic haemodialysis with ≥2 m-mode 2D echocardiograms and tissue doppler studies were included. Indexed LV mass (LVM) in $g/m^{2.7}$, geometry and LV function were compared at baseline (dialysis start) with follow-up studies at least 6 months following commencement. Left ventricular hypertrophy (LVH) was defined as indexed LVM > 51 $g/m^{2.7}$. We estimated fluid volume status using averaged interdialytic weight changes over 4 weeks prior to echocardiography, and blood pressure (BP) was analysed using 24-h ambulatory monitoring and routine aneroid BP measurements pre- and post-dialysis. Stepwise multiple regression analysis was performed to assess factors associating with LVM index change.
Results: 24 of 32 children <18 years were included ($n = 6 < 5$ years) with last follow-up scan performed following median dialysis duration of 19 months (range 6–64). The prevalence of LVH at baseline was 45.8% (12.5% concentric, 33.3% eccentric) compared with 20.8% (no concentric, 20.8% eccentric) on follow-up, with reduction in mean indexed LVM from 52.3 $g/m^{2.7}$ to 39.4 $g/m^{2.7}$ ($p = 0.001$); similar changes were observed for LV mass-for-height z-scores (0.71 vs. -0.43; $p = 0.001$). Mean fractional shortening changed from 37% to 38% ($p = 0.51$) and mean E/E' improved from 10.9 to 9.1 ($p = 0.04$). Multiple regression analysis identified improved systolic BP control and younger age at dialysis commencement as independent predictors for indexed LVM change ($p = 0.017$ and 0.042 respectively).
Conclusions: We report improvement in LV structure and function in children despite being on chronic intermittent haemodialysis. Encouragingly, these findings suggest that cardiovascular health in this population does not always deteriorate but can be stabilised and indeed improved with good BP management over time.

P-168 VALUE OF ACCELERATED PERITONEAL EXAMINATION TIME IN PEDIATRIC NOCTURNAL INTERMITTENT PERITONEAL DIALYSIS

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Introduction: Pediatric peritoneal dialysis patients depend on optimal dwell time for ultrafiltration and clearance. In clinical practice, most of the pediatric dialysis patients lack optimal dwell time resulting in suboptimal ultrafiltration. However, it is possible to lineate appropriate dwell time from the APEX (Accelerated Peritoneal Examination time), derived from a standardized PET (peritoneal equilibration test). The value of APEX is well recognized and appreciated in adapted peritoneal dialysis (combination sequences of short and long dwells within one peritoneal dialysis session). However, in nocturnal intermittent peritoneal dialysis (NIPD) it may be a potential parameter in maximizing the ultrafiltration.
Material and methods: It is a retrospective cohort study, where peritoneal dialysis details were derived from the digital memory card which is incorporated in the dialysis machine (Fresenius cycler-sleep safe (V2.2X). Both paper chart review (for patient data) and electronic chart review (for lab data) were assessed.
Results: Out of 15 patients enrolled in our study, mean ultrafiltration significantly improved ($p < 0.01$) after calculated APEX (Table 1). The mean (SD) were 189.4 ± 44.7. Moreover, mean ultrafiltration remarkably improved ($p = 0.006$) both in low/low-average and high/high-average peritoneal transporters. In relation to clearance, Kt/V did not change ($p = 0.16$) before and after APEX (Table 2). The mean (SD) were $n = 15$, 2.1 ± 0.3. whereas in relation to creatinine clearance, significant improvement ($p = 0.04$) noted in our study (Table 2). The mean (SD) were $n = 15$, 46.8 ± 7.
Table 1: Relationship between mean ultrafiltration, Kt/v and CrCl before and after APEX

($N = 15$)	Mean ± sd	p-value
Mean ultrafiltration after APEX	189.4 ± 44.7	<0.001
Mean ultrafiltration before APEX	140.5 ± 47.1	
KT/V (after APEX)	2.1 ± 0.3	.16
KT/V (Before APEX)	2.2 ± 0.4	
CrCl (After APEX)	46.8 ± 7.0	0.04
CrCl (Before APEX)	40.5 ± 7.7	

Kt/v- measures a change in the concentration of urea; CrCl -creatinine clearance; APEX -Accelerated Peritoneal Examination time; SD -Standard deviation).
Conclusions: APEX time in NIPD can be essential in maximizing the ultrafiltration and CrCl especially in patients with low and low-average transporters. Also, APEX gives a valuable support to peritoneal high and high average transporters in optimizing the ultrafiltration.

P-169 AMBULATORY HYPERTENSION AND LEFT VENTRICULAR HYPERTROPHY IN DIALYZED CHILDREN: DOES DIALYSIS MODALITY MATTER?

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Introduction: To compare 24-h blood pressure and left ventricular mass in children on different peritoneal and extracorporeal dialysis modalities.
Material and methods: Prospective registry study of 579 paediatric patients undergoing chronic dialysis (121 CAPD, 327 Automated (A)PD, 91 HD, 40 HDF) in whom 1277 ABPM profiles were performed. Nine hundred five echocardiography studies were available in 451 patients. Generalized linear modeling was applied to identify factors affecting 24 h–MAP and left ventricular mass index (LVMI).
Results: At study entry, 24 h–MAP SDS differed significantly between HD (2.9 ± 3.7 SDS), HDF (1.3 ± 1.9), APD (2.1 ± 3.6) and CAPD patients (0.9 ± 3.7). Differences were explained by variation in daytime MAP. Nighttime MAP was more markedly elevated than daytime MAP (night: 2.1 ± 2.5 vs. day: 1.3 ± 3.1 SDS) and not different across treatment modalities. Nocturnal dipping was reduced (<10%) in 49% and even reversed in another 13.4% of patients, without differences between the treatment groups. Median (iqr) LVMI was lower in HDF (40.6 (20.3) g/m$^{2.16}$) than in all other treatment groups (medians 43.5–45.9 g/m$^{2.16}$; $p = 0.022$).
In the multivariable longitudinal analysis, 24 h–MAP increased with age (0.5 ± 0.1 mmHg/year; $p < .001$), decreased by 5.0 ± 0.9 mmHg per L/m^2 daily urine output ($p < .001$) and was 5.2 ± 1.9 mmHg higher in HD than in CAPD patients ($p = 0.007$). 24-MAP did not change with time on dialysis. Higher MAP was associated with the use of Ca-channel blockers (+4.6 ± 1.1 mmHg, $p < .001$) and beta-blockers (+2.7 ± 1.2 mmHg, $p = 0.027$). Nocturnal dipping was greater in patients with preserved urine output and higher with APD and HDF than with CAPD. LVMI increased with 24 h–MAP (+0.32 ± 0.04 g/m$^{2.16}$ per mmHg, $p < .001$), BMI (+3.1 ± 0.7 g/m$^{2.16}$ per SD, $p < .001$) and diminishing urine output (−3.2 ± 1.6 g/m$^{2.16}$ per L/m^2/d, $p = 0.049$), and was higher in CAPD than in APD (−10 ± 2.5 mmHg, $p < .001$), HD (−9.2 ± 3.5, $p = 0.008$) and HDF (−12.9 ± 4.2, $p = 0.002$).
Conclusions: After multivariate adjustment HD associates with higher 24 h–MAP, and CAPD with lower nocturnal dipping and higher LV mass.

P-170 DEVELOPING A TRIGGER TOOL TO MONITOR ADVERSE EVENTS DURING HAEMODIALYSIS IN CHILDREN: A PILOT PROJECT

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Introduction: Haemodialysis trigger tools (HTT) have been proposed to enable regular monitoring of adverse events in adults during haemodialysis. There is currently no specific HTT available for children. We describe results of a pilot project to monitor adverse events during haemodialysis following development of a paediatric HTT.
Material and methods: Prospective data collection was performed using a broadly applicable 'per-dialysis session' tool including 53 triggers across 6 domains. Each trigger was additionally evaluated for level of physical harm; allocation to harm categories was subsequently reviewed jointly by two authors to minimise subjectivity. Trained haemodialysis nurses completed the HTT at the end of designated dialysis sessions in <5 min.
Results: The HTT was completed for 91 haemodialysis sessions of 17 children over an eight-week period, in which 139 triggers were identified. The 5 most frequent triggers included: failure to have nursing safety huddle during session ($n = 23$), need for additional fluid removal ($n = 19$), need for tissue plasminogen activator infusion ($n = 13$), need for line reversal and problems with dialysis machine failure ($n = 8$ each) and delayed reporting of abnormal laboratory results ($n = 7$). Sixty-three percent of triggers were categorised to have a potential to cause temporary harm and required intervention and others categorised to have no potential for harm. For an individual patient, the need for additional fluid removal was the most frequently cited trigger for potential harm. There were no triggers categorised to have potential to cause permanent harm.
Conclusions: This pilot study provides evidence of the risks inherent to paediatric haemodialysis provision and the value of regular monitoring of adverse events using a paediatric HTT. Further cycles of modifications to the HTT with re-testing and continuing team education are ongoing. We would propose the use of paediatric HTT to be included as part of standard care by centres providing haemodialysis to children in the future.

P-171 AUTOLOGOUS ARTERIOVENOUS FISTULAS USING MICROSURGERY TECHNIQUES IN CHILDREN WEIGHTING ≤ 20 KG: A SINGLE CENTER STUDY

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Introduction: This study aims to describe a single-center experience regarding the efficiency and longevity of arteriovenous fistulas (AVF) created in children weighting ≤20 kg for haemodialysis (HD).
Material and methods: We collected data of AVF created using microsurgery techniques between 1988 and 2015. Early failure was defined as the inability to use the AVF even once. Primary patency was defined as the interval time from VA placement until any intervention designed to maintain or reestablish patency and secondary patency as the interval time from VA placement until VA failure.
Results: 48 AVF (35 distal, 13 proximal) were created in 41 children with a median weight of 13.5 kg (range 5.5 to 20). Small age and body weight at AVF creation were associated to the need for a second AVF ($p = 0.046$ and $p = 0.019$ respectively). Early failure was observed in 6 (12.5%) AVF due to 4 access thromboses and 2 no-maturations. Median time to first utilization in HD was 22 weeks. Cumulative primary patency rates (± standard error) at 1 and 2 years were 54.2% (± 7.2) and 39.6% (± 7.1) respectively. Secondary patency rates (± standard error) at 1, 2, 3, 4 and 5 years were 85.4% (± 5.1), 83.3% (± 5.4), 70.5% (± 6.6) and 64.1% (± 7) and 57.7% (± 7.2) respectively. The average number of interventions performed per initially functional AVF was 1.36 (range 0–5). One third of thrombosis after AVF utilization were observed at kidney transplantation peri-operative time.
Conclusions: Arteriovenous Fistulas are feasible in younger children with an early failure rate of 12.5%. Time to first utilization is longer than in older children but secondary patency is excellent. Thrombosis rate is considerably high during transplantation surgery.

P-172 DEFINING OPTIMAL WEIGHT IN CHILDREN WITH CHRONIC KIDNEY DISEASE AND ON DIALYSIS: A COMPARISON OF TECHNIQUES

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Introduction: Fluid balance is pivotal in the management of children with chronic kidney disease (CKD) and on dialysis. Although many techniques are available to assess fluid status, there are few studies in children.
Material and methods: We performed a longitudinal study in 30 CKD children and 13 age-matched healthy controls (71 measurements) to determine a correlation between optimal weight by bioimpedance spectroscopy (Wt-BIS) and clinical assessment (Wt-CA). The accuracy of Wt-BIS (relative overhydration [Rel OH]) was compared against indicators of fluid status such as peripheral blood pressure, central blood pressure, N terminal pro-brain natriuretic peptide (NT-proBNP) levels and cardiovascular end-points namely pulse wave velocity (PWV) Z-score for age, left ventricular hypertrophy end diastolic distance (LVEDd) and left ventricular mass index (LVMI).
Results: There was poor agreement between Wt-CA and Wt-BIS among children on dialysis when compared to CKD5 or control subjects ($p = 0.01$). We developed a modified chart to plot Rel OH against systolic blood pressure (SBP) Z- score for the appropriate representation of volume status and blood pressure in children. A quarter showed SBP above 90th percentile but not with concurrent overhydration. Rel OH correlated with peripheral pulse pressure ($p = 0.03$; $R = 0.3$), higher NT-proBNP ($p = 0.02$; $R = 0.33$) and LVEDd ($p = 0.05$;$R = 0.38$). Central aortic mean and pulse pressure did not correlate with Rel OH but significantly associated with LVEDd. ($p = 0.03$;$R = 0.47$ and $p = 0.01$;$R = 0.50$ respectively). Systolic BP correlated with PWV Z-score ($p = 0.04$). Forty percent of children on HD and 30% on PD had increased LVMI.
Conclusions: We report a marked discrepancy between BP and hydration status in children on dialysis, suggesting that an objective method for the assessment of hydration status of children on dialysis is necessary.

P-173 AGREEMENT OF FLUID ASSESSMENT MEASURED USING PRE-DIALYSIS BIOIMPEDANCE SPECTROSCOPY VERSUS CLINICAL JUDGEMENT IN CHILDREN ON CHRONIC HAEMODIALYSIS

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Introduction: Body composition monitoring using a multifrequency bioimpedance (BCM-BIS) device has been shown to be a useful tool in the assessment of dry weight in adult haemodialysis (HD) patients. There is limited data regarding its clinical utility in children.
Aim: Our aim was to investigate the agreement of fluid assessment measured pre-dialysis using BCM-BIS with clinical judgement in children on chronic haemodialysis.
Material and methods: Estimated dry weight (EDW) was calculated in all patients using a combination of clinical examination and observations during dialysis and history of symptoms and signs during and/or post dialysis of muscle cramps, dizziness, hypotension or hypertension. As per clinical judgement, the fluid volume to be removed at each dialysis session was calculated as the difference between pre-dialysis weight and EDW ($FA_{clinical}$). We analysed agreement of fluid assessment using BCM-BIS (FA_{bcm}) with $FA_{clinical}$ when the two were <0.5 l; 0.5-1 l; and >1 l with one another. We further analysed fluid assessment as measured by FA_{bcm} with $FA_{clinical}$ as a percent change (gain or loss) of EDW.
Results: Single centre, retrospective review including 8 children with 58 BCM-BIS measurements. We excluded children younger than 5 years. Mean age was 10.2 years (range 5-17 yrs) including 5 boys. Fluid assessment following pre-dialysis measurement by FA_{bcm} was <0.5 l in 31 (53%), between 0.5-1 l in 11 (19%) and >1 l in 16 (28%) when compared with $FA_{clinical}$; with no significant correlation between FA_{bcm} and $FA_{clinical}$ ($p = 0.12$). Comparison of FA_{bcm} with $FA_{clinical}$ as a percent change of EDW suggests that although the vast majority of FA_{bcm} measurements were similar with $FA_{clinical}$ there was a significant difference in the absolute volumes suggested by FA_{bcm} when compared with $FA_{clinical}$ (figure).
Conclusions: There is poor agreement between fluid assessment performed using bioimpedance and clinical evaluation with significant differences in the estimated fluid gain or loss by bioimpedance in children on chronic haemodialysis.

P-174 IMPACT OF RECOMBINANT HUMAN ERYTHROPOIETIN TREATMENT ON LEFT VENTRICULAR MASS AND CARDIAC FUNCTION IN PATIENTS WITH END STAGE RENAL DISEASE ON HAEMODIALYSIS

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Introduction: Objective:of this work was to demonstrate the effect of rHu EPO therapy on LVH and LV systolic function in patients with end stage kidney disease.
Material and methods: Thirty two patients were enrolled in this study, 14 females and 18 males. Their age ranged from 5 to 17 years along with 15 age and sex matched healthy subjects as controls. The inclusion criteria were; the presence of renal anemia, adequate serum iron status with serum ferritin level of 100 ng/ml or more and a transferrin saturation of >20%, normotension or controlled hypertension and no history of valid heart disease or other systemic illness. We analyzed the laboratory and echocardiographic data before starting EPO treatment and after treatment in period of follow up ranged between 4 and 9 months with a mean of 5.8+/−1.5 months.
Results: Hb level increased from 8.5+/−1.87 to 9.3+/−1.7 g/dl, Hct level increased from 25.78+/−6.59% to 28.88+/−5.5%, LVMI showd reduction from 108.8+/−41.97 to 97.13+/−43.9 g/m2, SV decreased from 59.58+/−21.17 to 53.9+/−18.49 ml and finally CO decreased from 5.74+/−2.2 to 5+/−1.5 L/min. No significant change was detected regarding the HR, EDV, &ESV. LV systolic function was normal at the start of the work and remained so in the follow up examination.
Conclusions: We concluded that in patients with ESRD on chronic hemodialysis, LVH regression can be obtained after partial correction of anemia with rHu EPO which can be also associated with reduction of the high CO encountered in these cases. Weather this regression would improve outcome in haemodialysis patients remain to be established.

P-175 TREATMENT OF SEVERE SEPSIS USING POLYMYXIN-B DIRECT HEMOPERFUSION (PMX-B DHP) IN TWO PEDIATRIC PATIENTS

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Introduction: PMX-B DHP is an extracorporeal technique used in gram negative(G-)sepsis,based upon a selective adsorption of circulating endotoxins from the blood,improving hemodynamic,organ dysfunction and 28-days mortality.
Material and methods: We report two cases of pediatric patients treated with PMX-B DHP in a pediatric intensive care unit(PICU)setting.

Results: Case 1:2 years old male underwent 2nd liver transplantation.In 5th post-operative day(POD)the patient developed an increase in inflammatory markers,stage 2 AKI,respiratory failure and severe hypotension,unresponsive to fluid resuscitation and vasoactive drugs.Clinical state was strongly suggestive for septic shock.Microbiological samples were collected and broad spectrum antibiotic therapy was started,but with lack of efficacy.Ematic endotoxin activity(EEA)resulted 0.67 suggesting a G- sepsis.Considering the clinical picture and antibiotic resistant sepsis,PMX-B DHP was started,with a dialysis equipment using a cartridge PMX-20R,designed for adult patients.DHP was carried out with a 4% albumin priming,Qb 70 ml/min and heparin as anticoagulation.Five daily consecutive sessions(3 h each)was performed without any complications,followed by a remarkable clinical/laboratoristic improvement and EEA of 0.4.In 23rd POD children was discharged from PICU. Case 2:3 years old male with congenital heart disease.During the corrective surgical intervention the patient experienced a serious hypotension,subsequent prerenal AKI and fluid overload with need for RRT.In the 7th POD,during the admission in PICU,a G-septic shock with a EEA of 0.71 was diagnosed.Broad spectrum antibiotic therapy was started but without any efficacy,then six daily consecutive DHP was performed using PMX-20R cartridge with a Qb 60 ml/min,priming with 4% albumin and anticoagulation with heparin.The DHP was well tolerated with an improvement in the clinical conditions and a gradual weaning from vasoactive drugs and EEA of 0.4.Unfortunately the patient died after 25 days because of cardiac failure.
Conclusions: Nowadays,the use of extracorporeal methods based on selective adsorption are getting more and more employed in the treatment of septic shock in adult patients.Our case series seems to demonstrate the safety and efficacy of PMX-B DHP also in critically ill children.

P-176 MAINTENANCE REGIMEN OF INTRAVENOUS IRON SUPPLEMENTATION PREVENTS ANEMIA IN PEDIATRIC HEMODIALYSIS PATIENTS

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Introduction: Anemia in pediatric chronic hemodialysis patients is usually undertreated. Iron supplementation and recombinant human erythropoietin (rHuEPO) have been recommended for improving anemia in hemodialysis patients. However, there are limited data on the optimal response to achieve sustained normal hemoglobin (Hb) level using maintenance regimen of intravenous iron supplementation in children.
Material and methods: In this retrospective cohort study, data of two groups were retrieved from the medical records of pediatric patients on regular hemodialysis. Patients in both groups had normal hemoglobin and iron values (transferrin saturation and ferritin serum level). Group 1 (n = 47) were patients receiving two-weekly 2 mg/kgBW/dose of intravenous iron sucrose for 2 doses without adjusted dose; while group 2 (n = 27) did not receive any iron supplementation. Exclusion criteria included hemolytic anemia, bleeding manifestations, hemoglobinopathies, receiving red blood cell or whole blood transfusion, severely malnourished, evidence of active inflammation or infection and incomplete medical record. Primary outcomes were the percentage of patients with reduced Hb and TSAT levels. Secondary outcome was side effects of intravenous iron supplementation.
Results: We retrieved data of 74 children from the medical records. No difference was found in clinical characteristics and mean values of hemoglobin, ferritin and TSAT between both groups. There was a significant difference ($p < 0.001$) on the percentage of patients with reduced Hb and TSAT levels between both groups. No clinical side effects was observed; however, iron overload occurred in 6/47 (12.77%) patients receiving intravenous iron.
Conclusions: Maintenance regimen of intravenous iron supplementation should be considered to achieve sustained hemoglobin and iron levels in pediatric hemodialysis patients and it consequently may prevent anemia. The regimen is safe as no clinical side effects were found; however, iron overload may occur.

P-177 SURGICAL OUTCOME OF PERITONEAL DIALYSIS CATHETER INSERTION IN PEDIATRIC PATIENTS: EXPERIENCE IN IRAN

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Introduction: Early and late surgical complications of peritoneal dialysis (PD) come along with devastating morbidity and eventually increase the rate of mortality. The aim of our study was to evaluate the changes in the rate of surgical complications and the outcome of PD catheter in a tertiary center in Iran.
Material and methods: This is a retrospective cohort study conducted between 1993 and 2012. Inclusion criteria were all children aged <=14 years with chronic kidney disease who underwent PD. Patients with acute peritoneal dialysis or follow up less than six months were excluded. eGRF was calculated using Schwartz formula. The surgical complications including catheter malfunction, leak, Dacron sheet extrusion, and hernia were considered. Catheter survival, rate of catheter changes, and rate of peritonitis calculated in two time period. P-value less than 0.05 treated statistically significant.
Results: During a 19 year interval, 86 PD catheters were inserted in 50 patients, with a median (range) age of 22.5 (1–192) months. The most common underlying diseases were CAKUT. Median eGFR at the time of operation was 7.8 (4–31.4 ml/min/1.73m^2). Catheters were inserted laparoscopicaly in 4.6%. Among surgical complications, 39% of patients developed hernia in median of five months after surgery, in addition catheter malfunction, dislocation, adhesion, or cuff extrusion developed in 22% of cases. The most common reasons for removal were catheter related (outflow failure, adhesion, cuff extrusion) (21%) and infection (peritonitis, tunnel infection) (17.4%). Reoperation for catheter related complication was required in 21 patients (42%). However, the number and the cause of catheter exchange and the outcome of patients were not statistically significant in two time periods; The rate of outflow failure (77% vs. 25%), peritonitis rate (1 per 7.5 vs. 56.9 patient- months) and catheter reinsertion rate (1 per 30.8 vs. 63.7 patient-months) improved significantly from the time period before 2005and afterward. The median (range) follow up of patients was 29 months (6–126 months). Almost 20% transplanted, −26% were still on CAPD, 6% switched to hemodialysis, renal function recovered in 10%,, and38% died.
Conclusions: This study shows that although improvement in our technique has been accomplished and complications related to technique of insertion are declining; management and care of the catheter in order to reduce peritonitis is still lacking and more should be done to educate nurses and parents and care givers in this regard.

P-178 FINANCIAL BARRIERS PREVENT IMPLEMENTATION OF INTENSIFIED HEMODIALYSIS - SURVEY ANALYSIS OF THE INTERNATIONAL PEDIATRIC DIALYSIS NETWORK

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Introduction: In pediatric patients on conventional hemodialysis (HD), morbidity is high and quality of life is poor. To overcome these shortcomings of conventional dialysis, intensified HD programs have been developed. The feasibility and outcome in children have been reported from few pediatric dialysis centers only.
Material and methods: An online survey was carried out in all 221 pediatric dialysis centers participating in the International Pediatric Dialysis Network (IPDN) to assess, the attitude of pediatric nephrologists towards intensified HD, penetrance into clinical practice and respective barriers.
Results: 134 (61%) of the centers replied to the questionnaire. Sixty-nine percent of the pediatric nephrologists recognize sufficient evidence in favor of intensified HD independent from the fact whether they offer intensified HD or not. Fifty percent consider daily nocturnal HD, 21% short daily and 10% intermittent nocturnal HD to offer the best overall patient outcome. Only 2% consider conventional HD to provide the best patient outcome. Eighty percent work in centers where new ideas are greeted, and institutional barriers are few. Fifty-seven percent of pediatric nephrologists always try to convince patients and care givers to apply the best available dialysis modality. Thirty-eight percent of the respondent pediatric nephrologists realize intensified HD in a subgroup of patients, mainly short daily HD. Thirty-six percent of these centers offer home HD. The most important organizational barriers to expand intensified HD programs were lack of adequate funding (66%) and lack of stuff (63%), whereas lack of expertise (21%) and of motivation (14%) were reported infrequently.
Conclusions: The majority of nephrologists consider intensified HD as the best HD treatment for children, but a minority only applies it to some of their patients. Inappropriate funding represents the most important barrier for implementation of intensified HD into clinical practice.

P-179 ROLE OF BIOELECTRICAL IMPEDANCE ANALYSIS IN NUTRITIONAL EVALUATION OF CHILDREN ON HAEMODIALYSIS: A SINGLE CENTRE STUDY

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Introduction: This cohort aims to study the usefulness of bioelectrical impedance analysis (BIA) for body muscle mass assessment in children on haemodialysis (HD).
Material and methods: We have undertaken a retrospective study from January 2013 to December 2016 on the results of lean tissue mass index (LTI) measured by bioelectrical impedance analysis (BIA) in all children aged ≥5 years old with HD duration ≥6 months. Annual average LTI, normalized protein catabolized rate (nPCR), body mass index (BMI), diuresis, haemoglobin and serum ferritin, albumin, bicarbonate, 25-hydroxyvitamin D levels were calculated for each child.
Results: 16 children (9 boys, 7 girls) aged from 5 to 17 years old were included. The median study period per child was 2 years (range 1–4). BMI z -score was <−1.5 in 6 patients. Total median LTI change of all patients was +5.07%. Total average LTI of each patient was not influenced by sex, age or diuresis status. A significant linear correlation was observed between BMI level and LTI and between BMI z-score and LTI ($r = 0.593$, $p < 0.001$ and $r = 0.673$, $p < 0.001$). We noticed that only 1 child presented a BMI z-score fall below −1.5 during the study period and was the only one who presented an LTI decrease more than 2%. A significant linear correlation was observed between nPCR level and LTI ($r = 0.35$, $p = 0.031$). Annual changes of nPCR level was related to those of LTI (Rho = 0.465, $p = 0.029$) and its annual positive or negative changes were related to similar LTI changes ($p = 0.075$). 25-hydoxyvitamin D level was the only biochemical parameter significantly correlated with LTI ($r = 0.331$, $p = 0.042$).
Conclusions: Bioelectrical impedance analysis seems to be a valuable and reliable tool for body muscle mass assessment and follow-up in children on HD. 25-hydroxyvitamin D plays probably a beneficial role on body muscle mass.

P-180 OUTPATIENT NEPHROTIC SYNDROME WITH PHYSICIAN AND SPECIALIZED NURSE

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Introduction: Objectives 1. Improving care for children with nephrotic syndrome by introducing the specialized nurse at the outpatient clinic. 2. Developing written information regarding the care for children with nephrotic syndrome. 3. Stimulating self-management for the patients and their care-givers.
Material and methods: Method • Children suffering steroid sensitive nephrotic syndrome have their consult in the morning at the same outpatient clinic. The patients are alternately seen by a dedicated pediatric nephrologist and one of the two specialized nurses, supervised by the pediatric nephrologist. By doing this, we aimed more time for consultation. Furthermore the different caretakers would guarantee a more multidisciplinary approach. • All patients have received a letter with information before their first visit, regarding the new organization of our care. • To measure the quality of care we performed a Dutch validated survey among all children visiting our outpatient clinic with nephrotic syndrome over 12 years of age and all parents. The same questionnaire was repeated 1 year after introducing the specialized nurse care. • Literature search was performed, but revealed only scarce data on this subject.
Results: Results At baseline 32 parents and 25 kids returned a completed survey. After 1 year we received surveys of 36 parents and 33 kids. Results were analyzed and details will be presented. Noticeably more contacts by mail or phone with parents were recorded.
Conclusions: Conclusions • Parents know to find us for questions and they contact us easier. • The same faces at the outpatient clinic. • Written information regarding nephrotic syndrome is completed. • Preliminary results surveys: score after 1 year 3.3 points higher. Further analysis will follow.

P-181 PEDIATRIC DIALYSIS PATIENTS IN A SOUTHERNE CITY OF ALGERIA

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Introduction: End stage renal disease (ESRD) in pediatric population is a major challenge. In Algeria, the number of children reaching ESRD increases anuually. The epidemiological studies of the pediatric ESRD in Algeria are few. The objectives of this study are: Determine the epidemiological characteristics of dialyzed children and Analyze the results of pediatric dialysis.
Material and methods: In this retrospective study, we included all patients under the age of 19 years at the time of the ESRD, living in Ghardaïa, purified at least 03 months by hemodialysis (HD) or peritoneal dialysis (DP) during the period of ten (10) years: 01/01/2005 to 31/12/2014. Information was collected from the medical files, interrogation of patients and their parents.
Results: During the period of study, twenty-five (25) children under the age of 19 years have reached the ESRD and have been dialyzed.The average age was 12 years (1–19), sex ratio (M / F) was 0.9. The calculated annual incidence of ESRD in the pediatric population in Ghardaïa was: 15.28 pmarp / yr. (Per million age related population). The prevalence is: 73.36 pmarp. The frequency was high for patients between 10 and 14 years of age (44%). Glomerular nephropathies remain the leading cause of pediatric ESRD in our study (36%). The cortico-resistant nephrotic syndrome (6 cases) is the chief of wire but renal biopsy was rarely practiced (2/6) likewise for the genetic study (2/6). Congenital malformations of the kidneys and the excretory pathwurinary tract (CAKUT) are frequent,

dominated by obstructive uropathies (neurological bladder) followed by vesico-ureteral reflux.In 20% of the cases, the etiology was not found.
Dialysis was in most cases urgent (68%), anemia was predominantly present at the time of dialysis (88%) and transfusion was necessary in 64% of cases. Hemodialysis is the first treatment method for incident (76%) and prevalent (70%) patients in our series.
A very high mortality rate (20%) was founded mainly due to dialysis insufficiency, a very low school enrollment (40%) and a significant retardation of growth (60%). none of our patients was regularly followed in pediatrics during years of dialysis and none of patients benefited from treatment with growth hormone.
The transplant rate (4%) is well below the national average (20%), Only 1 patient has been transplanted, obstacles to kidney transplantation are numerous, mainly the absence of donor (58%).
Conclusions: Our study is the first work on ESRD of the child in southern Algeria; we have highlighted the following problems:- Late diagnosis of kidney disease- Absence of targeted screening programs- Lack of coordination among practitioners- lack urological surgery for complex CAKUT- Absence of genetic diagnosis in (CRNS, Oxalose, Nephronophtisis ... ..) A comprehensive management of the dialyzed child should enable them to achieve acceptable growth, good schooling and quality of life, require a good training of health care workers and close collaboration between the different treatment practitioners.

P-182 PEDIATRIC CONTINUOUS RENAL REPLACEMENT THERAPY- A REPORT OF SEVEN YEARS, THREE CENTERS

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Introduction: Continuous renal replacement therapies (CRRT) either as continuous venovenous hemofiltration(CVVH) or hemodia filtration(CVVHD) are used frequently in critically ill children. Many clinical variables and technical issues are known to affect the result. The factors that could be modified to increase the success of renal replacement are sought. As a contribution, we present the data on 104 patients who underwent CRRT within a seven years period.
Material and methods:A hundred and four patients admitted between 2009 and 2016 were included in the study. The demographic information, admittance PRISM (Pediatric Risk of Mortality) scores, indication for CRRT, presence of fluid overload, CRRT modality, durations of CRRT and PICU stay were compared between survivors and nonsurvivors.
Results: The overall rate of survival was 51%. Patients with fluid overload had significantly increased rate of death, CRRT duration and PICU stay. Multiorgan dysfunction syndrome as the indication for CRRT, was significantly related with decreased survival when compared to acute renal failure and acute attacks of metabolic diseases. CRRT modality was not different between survivors and nonsurvivors. SMR (Standardized mortality ratio) of the group was calculated to be 0.8.
Conclusions: CRRT in critically ill patients is successful in achieving the primary targets of therapy. It has a positive effect on expected mortality in high risk PICU patients. To affect the outcome, follow up should be focused on starting therapy in early stages of fluid overload. Prospective studies defining relative importance of risk factors causing mortality can assist in building up guidelines to affect the outcome.

P-183 IMPACT OF HEMOGLOBIN VARIABILITY ON OUTCOME PARAMETERS IN PEDIATRIC DIALYSIS PATIENTS

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Introduction: During treatment with ESA, level of hemoglobin (Hb) usually fluctuates; this phenomenon is known as "hemoglobin cycling (HC)" and there is some debate about whether or not this may lead to increased morbidity/mortality in adults. It was aimed to evaluate the impact of HC on patient-important outcome parameters including left ventricular hypertrophy (LVH) and inflammation in pediatric dialysis patients.
Material and methods: Records of patients followed-up in nine centers (2008–2013) were retrospectively reviewed. Biochemical parameters, complete blood count, CRP, echocardiographic data, monthly-Hb and albumin levels for the last one year were collected, where available. More than 1 g/dL decrease or increase in Hb level was considered as HC. Patients were divided into two groups according to 12-month Hb-trajectory as rare cycling (RC)(≤ 3) and freuquent cycling (FC)(≥4 fluctuation) as well as three groups based on time-averaged-Hb levels; <10, 10–11 and >11 g/dl.
Results: 245 dialysis patients aged 12.3 ± 5.1 (range: 0.5–21) years were enrolled in this study. Fifty percent of the patients had 1–3 cycling, 82% had 1–5, only 3% had no cycling. There were no differences between HC groups with respect to age, primary renal disease, dialysis modality, having anemia and hospitalization rate, while RC patients had higher urine output ($p < 0.01$) and higher CRP levels ($p < 0.001$). Echocardiographic data were available in 137 patients. Although LV mass index (LVMI) was higher in RC than FC group (65 ± 37 vs 52 ± 23 g/m$^{2.7}$, $p = 0.056$), prevalence of LVH was not different between groups. Time-averaged-Hb levels were inversely correlated with ESA requirement ($r = -0.497$), mean arterial pressure ($r = -0.213$), LVMI ($r = -0.471$) and CRP ($r = -0.443$), but positively with urine output ($r = 0.296$) and albumin levels ($r = 0.275$). Patients with time-averaged-Hb levels < 10 g/dl had an increased risk of LVH and inflammation.
Conclusions: Hb cycling is common among dialysis patients.Severity of anemia rather than its cycling has more significant impact on the prevalence of LVH and on inflammatory state.

P-184 MORTALITY RATE IN CHILDREN WITH ESRD DUE TO NEUROGENIC BLADDER IN POLISH PEDIATRIC RENAL REPLACEMENT THERAPY REGISTRY

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Introduction: There is scarce information in the medical literature on mortality in subjects with ESRD due to neurogenic bladder (NB) though it has been suggested that the long term survival of this group is inferior to that of children with other diagnoses. The aim of the study was to assess survival of children diagnosed with congenital neurogenic bladder in a cohort of patients treated with renal replacement therapy in Poland.
Material and methods: A retrospective longitudinal analysis involving 1136 pediatric subjects receiving renal replacement therapy in period 2000–2015 was conducted. Cox proportional hazard method was used to analyse risk of death in children with neurogenic bladder compared to those with other etiologies.
Results: 58 children (42 girls, 16 boys) with neurogenic bladder were identified. Hemodialysis was the dominant mode of initial therapy. Seven deaths were reported in this subgroup during 6659 patient-years of follow-up. After adjusting for age, the risk of death among patients with neurogenic bladder was significantly higher than that of all other causes: hazard ratio − 2.95 (95% confidence interval 1.33–6.56). Subjects with neurogenic bladder were older at start of RRT compared to other children (13.85 years vs. 10.13 years, $p < 0.001$). Average age at death was higher than in the remaining group 15.2 ± 6.9 year vs. 10.1 ±. 7.5 year. The main causes of death were cardiovascular events followed by infections.
Conclusions: 1. Children with neurogenic bladder have three time higher mortality on RRT compared to children with other etiologies of ESRD. 2. Age for both start of RRT and at death is higher compared to other children. 3. The main causes of death (cardiovascular and infections) are similar to that observed in the total cohort of children on RRT.

P-185 ULTRASOUND DILUTION AND THERMODILUTION VERSUS COLOR DOPPLER ULTRASOUND FOR ARTERIOVENOUS FISTULA ASSESSMENT IN HEMODIALYZED CHILDREN

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Introduction: This study aims to compare ultrasound dilution (UD) and thermodilution with Color Doppler ultrasound (CDU), which is the most accurate method for AVF assessment in children on HD.
Material and methods: All patients were dialyzed with the Fresenius 5008 HD machine. UD was performed using the Transonic device. The two methods were realised during the first 90 min of the same HD session and their results were compared to those acquired from the CDU performed in the same period in a non-HD weekly day.
Results: Seventeen simultaneous measurements of AVF flow rate and recirculation with UD and thermodilution method and 17 CDU were realized in 16 patients with a median age of 14.5 and a median weight of 38.4 kg. The median AVF flow rate was 752 ml/min (range 387–2520) corrected to 1292 ml/min per 1.73m^2 (range 614–3893) with thermodilution, 810 ml/min (range 350–3250) corrected to 966 ml/min per 1.73 m^2 (range 513–3675) with UD method and 550 ml/min (range 310–2300) corrected to 750 ml/min per 1.73 m^2 (range 471–2601) according to the measurement of the CUD. A significant linear correlation was observed between AVF measured with UD method and CUD ($r^2 = 0.786$, $p < 0.001$) and between AVF measured with thermodilution and CUD ($r^2 = 0.232$, $p = 0.05$). AVF flow rate was higher with UD in comparison to CDU in 15 out of 17 cases and with thermodilution in comparison to CDU in 15 out of 17 cases as well. Whereas recirculation in all AVF was 0% and <15% with UD and thermodilution respectively, 8 significant stenosis were observed in CDU.
Conclusions: Recirculation is not an accurate method for early screening of significant AVF stenosis in children on HD. UD seems more reliable in comparison to Thermodilution for AVF flow rate assessment. Both methods tend to overestimate AVF flow rate when compared to CDU.

P-186 ASSISTED HOME HAEMODIALYSIS

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Introduction: In the UK 13 haemodialysis (HD) centres serve the paediatric population. Consequently some children travel large distances for their dialysis but at a significant cost to their schooling, psychosocial health and family dynamics. Home HD offers an obvious solution but it is not suitable for every family.
Material and methods:
Objective: We report the case of a 8 year old girl with complex medical needs whose HD was carefully transitioned to East Anglia Childrens hospice close to her home.
Results: Patient K with VACTERL has a tracheostomy, colostomy, poor vascular access and renal failure. She travelled up to 3 h each way for in-centre HD and had repeated, frequent admissions to hospital with acute, often infective, respiratory symptoms.. Peritoneal dialysis was not an option. Home HD was medically optimal but not possible at home. Therefore we approached the hospice familiar to the family that was providing respite care and proposed a package of care that eventually transitioned Patient K's dialysis care to them. They agreed in principle. Funding was secured from the local council. The NxStage ™ dialysis system was chosen as it required no water conversion and was mobile. A robust 3 month training programme was designed and executed for 4 hospice nurses to learn to provide HD in the hospice 4 times/week. A risk assessment and symptoms care plan was written for the patient's specific needs. We secured the hospice local medical support from her local General Paediatric consultant and a honorary contract was put in place which enabled the hospice nurses to dialyse patient K at the local hospital during admissions.
The impact has been extremely positive. Patient K is free of dietary and fluid restrictions and is growing. She has made excellent progress at school with improved social skills with children her own age and for the first time mum has worked. The change in the quality of life for Patient K has been dramatic.
Conclusions: We present the first reported case of assisted home HD that has been successful in improving a child's health outcomes and rehabilitating them back into school, social and family life. Mum- "She sings on the way to the hospice and says it's where she goes for a holiday –it's such a relief for me".

P-187 CREATING A SAFE ENVIRONMENT FOR YOUNG ADULTS TO ACHIEVE INDEPENDENT HOME HAEMODIALYSIS

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Introduction:
Objective: Some young adults may be suitable for home haemodialysis (HD) but lack of supervision or support may prevent this becoming a reality.
Material and methods:
Method: We describe our enhanced safety approach which enabled a 17 year old with a arterio-venous fistula to transition from in-centre HD to home HD, independently.
Results: When planning the possibility of a young adult dialysing independently at home a number of unique safety issues need to be considered

and mitigated against in addition to addressing common concerns such as non-adherence to treatment. We individualised the training programme, modifying the standard teaching to incorporate additional safety measures focusing on a heightened awareness and earlier recognition of potential complications. Technology such as medical alarm necklaces and watches, alarm amplifiers and blood leak detectors alerted the young adult or the local medical emergency teams of impending risks or complications. The speed and clarity by which the young adult communicated with the community medical support teams was improved utilising WhatsApp® and smart phones.

After a stable and adverse incident free six months of treatment the young adult transitioned to a nocturnal home programme.

Conclusions: A young adult can haemodialyse at home, independently provided strategies are put in place to ensure the safety of the patient in the community.

P-188 PERITONEAL DIALYSIS IN VERY LOW BIRTH WEIGHT NEONATES

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Introduction: The aim of this retrospective study is to evaluate clinical characteristics and outcomes of the very low birth weight (VLBW) neonates with acute kidney injury (AKI) treated with peritoneal dialysis (PD).

Material and methods: A retrospective study has included 10 VLBW neonates treated with PD. Intravenous (IV) cannula and umbilical venous catheter were used for the peritoneal access.

Results: Mean age in the moment of starting PD was 14.9 ± 9.3 days. Mean body weight (BW) was 825 ± 215 g. The average gestational age was 26.3 ± 1.1 weeks. The average duration of dialysis was 20.5 ± 14.7 h. The average ultrafiltration was 7.7 ± 4.2 ml/kg/h. In the moment of starting the PD, the average BW was 302 ± 317 g (22 ± 13%) higher than at birth. The main cause of AKI was sepsis (n = 8/10). The dialysate leak was registered in two patients, one patient had peritonitis and the other one had a blockade of PD catheter. Six patients have died during the PD (severe sepsis), one has died due to hypoxic encephalopathy and coma and two patients have survived. One patient (with hypoxic encephalopathy and coma) has died 10 days after PD was stopped due to sepsis. The overall mortality was 80%.

Conclusions: Acute PD is still an appropriate treatment choice for VLBW neonates with AKI. In VLBW neonates, PD can be performed by an improvised PD system and catheters.

P-189 MUTATION OF THE EYA1 GENE IN A PATIENT WITH BRANCHIO-OTO-RENAL SYNDROME

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Introduction: Branchio-oto-renal (BOR) syndrome is an autosomal dominant disorder characterized by the coexistence of branchial cysts or stulae, external ear malformation with pre-auricular pits or tags, hearing impairment and renal malformations. However, the presence of the main features varies in affected families.

Material and methods: 0.

Results: Here, we present a 16-year- old boy admitted to the Department of Nephrology at the Pediatric Clinic, University Clinical Center of Kosovo, Pristina, Republic of Kosovo because of severe renal in- suf ciency diagnosed 6 years ago, which progressed to end-stage renal failure. Clinical examination on readmis- sion showed a pale, lethargic and edematous child, with auricular deformity, pre-auricular tags and pits as well as bilateral branchial stulae. Laboratory tests revealed high blood urea nitrogen (BUN) 15.96 mmol/L and serum creatinine 633.0 mol/L; low glomerular ltration rate (GFR) 12 mL/min./ 1.73 m2 and massive proteinuria 4+. Abdominal ultrasound showed bilateral kidney hypoplasia. A novel mutation of the *EYA1* gene was con rmed. Daily hemodialysis is continuing until renal transplantation is done.

Conclusions: This case is presented to increase awareness among general practitioners to consider BOR syndrome or other renal abnormalities in patients with branchial stula and/ or external ear anomalies or similar ndings in other fam- ily members.

P-190 IMPAIRED SYSTOLIC AND DIASTOLIC LEFT VENTRICULAR (LV) FUNCTION IN CHILDREN WITH CHRONIC KIDNEY DISEASE (CKD): RESULTS FROM THE CARDIOVASCULAR COMORBIDITY IN CHILDREN WITH CKD (4C) STUDY

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Introduction: Tissue doppler velocities are sensitive markers of LV function. The aim of this work was to analyze tissue doppler velocities in a large cohort of children with CKD and to assess risk factors for LV dysfunction.

Material and methods: A standardized echocardiographic examination was performed in 128 patients of the 4C Study aged 6–18 years with eGFR 10–60 ml/min/1.73 m^2. Tissue doppler measurements included early (E') and late (A') diastolic and systolic (S′) velocity at the mitral and septal annulus of the left ventricle. Measured values were normalized to z scores. Predictors of E'/A', E/E', S′ and LVMI were assessed by multiple linear regression analysis.

Results: Tissue doppler diastolic E' velocity was reduced and A' increased at the mitral and septal annulus, resulting in a reduced E' to A' ratio (z-score − 0.14, $p < .0001$), indicating diastolic dysfunction. Diastolic function (E'/A') was positively correlated with midwall fractional shortening ($r = 0.23$, $p < 0.01$). Reduced diastolic function was independently associated with declining renal function ($p = 0.005$), increased systolic blood pressure ($p = 0.045$) and pulse wave velocity ($p = 0.07$). LV filling pressure E/E' was increased (z-score 0.65,

$p < .0001$) and inversely correlated with the E'/A' ratio. Patients treated by RAS antagonists had significantly lower E/E'. Systolic tissue doppler velocities were significantly decreased (z-score − 0.24, $p = 0.001$) and inversely correlated to LV filling pressure ($r = -0.40$, $p < .0001$). The LVMI was not associated to systolic or diastolic tissue doppler velocities.
Conclusions: eGFR, systolic blood pressure and the type of antihypertensive medications are significant predictors of diastolic function in children with CKD. Tissue doppler velocities are independent of LV mass and provide sensitive information about early LV dysfunction in this population.

P-191 ELEVATED TIME-VARYING MYOCARDIAL WALL STRESS IN CHILDREN WITH HYPERTENSION

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Introduction: Myocardial wall stress (MWS) has been shown in adults with hypertension to be a primary determinant of ventricular remodeling, the changes in left ventricular (LV) geometry and structure. Studies investigating time-varying arterial load and instantaneous LV geometry as determinants of wall stress in children with hypertension have not been performed.
Material and methods: Transthoracic echocardiographic imaging of the left ventricle was performed and endocardial and epicardial volumes obtained from Tomtec wall tracking analysis. Carotid tonometry during systole was used to estimate LV pressure and was calibrated by mean and diastolic blood pressure (BP). MWS was calculated from LV volume and pressure measurements. Eighty children (41 boys) aged 13.0 ± 3.0 (mean ± SD) years (29 normotension and 51 hypertension) were studied.
Results: Children with hypertension were older (Hypertension VS normotensive: 14.1 ± 2.8 VS 11.0 ± 3.3 years, $p < 0.001$) and have higher body mass index (BMI) (22.3 ± 4.6 VS 19.4 ± 4.3 kg/m^2, $p = 0.007$), systolic (132 ± 19 VS 103 ± 11 mmHg, $p < 0.001$) and diastolic BP (72 ± 16 VS 57 ± 11 mmHg, $p < 0.001$) than controls. LVM (120.0 ± 37 VS 79.0 ± 29.5 g, $p < 0.001$), LVM index (33.6 ± 9.6 VS 28.1 ± 5.1 g/m$^{2.7}$, $p = 0.006$) and relative wall thickness (RWT) (0.39 ± 0.07 VS 0.32 ± 0.05, $p < 0.001$) were higher in children with hypertension than in those with normotension. Peak stress (Normotensive VS Hypertension: 338.8-18.5 VS 397.5-14.3 kdyne/cm^2, $p < 0.001$) and mean stress (323.6-14.5 VS 411.7-13.8 kdyne/cm^2, $p < 0.001$) was significantly elevated following adjustment for age, BMI and LVM index This relation disappeared after adjustment for blood pressure indices.
Conclusions: In children with hypertension, despite evidence of LV geometry alteration, MWS remained elevated and was explained by high blood pressure alone. We would postulate that improved blood pressure control may improve LV remodeling.

P-192 MYOCARDIAL DEFORMATION MEASURED BY 3D SPECKLE TRACKING IN CHILDREN AND ADOLESCENTS WITH PRIMARY SYSTEMIC ARTERIAL HYPERTENSION

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Introduction: Systemic arterial hypertension predisposes children to cardiovascular risk in childhood and adult life. Despite extensive study of left ventricular hypertrophy, detailed 3D strain analysis of cardiac function in hypertensive children has not been reported. The aim of this study was to evaluate left ventricular mechanics (strain, twist and torsion) in young patients with hypertension compared to a healthy control group and assess factors associated with functional measurements.
Material and methods: Patients aged less than 18 years with a diagnosis of primary arterial hypertension were included in the hypertension group and compared to healthy children. Standard 2D measurements were made using M-mode as well as 3D assessment of ventricular volumes and deformation, using speckle tracking echocardiography.
Results: 63 patients (26 hypertension and 37 healthy controls) were enrolled (mean age 14.3 years and 11.4 years, 54% male and 41% male respectively). There was no difference in left ventricular volumes and ejection fraction between the groups. Myocardial deformation was significantly reduced in those with hypertension compared to controls. For hypertension and controls respectively global longitudinal strain was −15.1 ± 2.3 vs −18.5 ± 1.9 ($p < 0.0001$), global circumferential strain −15.2 ± 3 vs −19.9 ± 3.1 (<0.0001), global radial strain +44.0 ± 11.3 vs 63.4 ± 10.5 ($p < 0.0001$) and global 3D strain −26.1 ± 3.8 vs −31.5 ± 3.8 ($p < 0.0001$). Basal clockwise rotation, apical counterclockwise rotation, twist and torsion were not significantly different. Following multivariate regression analyses blood pressure, body mass index and left ventricular mass maintained a significant relationship with measures of left ventricular strain.
Conclusions: Despite maintaining a normal ejection fraction, children with hypertension had significantly lower global longitudinal, circumferential, radial and 3D strain than healthy children. Whether reduced strain might predict future cardiovascular risk merits further longitudinal study.

P-193 IMPACT OF SALT AND POTASSIUM INTAKE ON BLOOD PRESSURE IN OBESE CHILDREN

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Introduction: Obesity and high salt intake are the two most important modifiable risk factors for hypertension. Higher salt intake has been associated with higher systolic blood pressure (BP) in obese children, but not in non-obese children. The impact of dietary salt intake on BP is affected by consumption of potassium. The urine Na/K ratio is a stronger correlate of BP than either sodium or potassium alone. The aim of this study was to determine if there was a difference in salt and potassium intake between obese children with or without hypertension.
Material and methods: A total of 34 obese children (22 male) aged 12.6 ± 2.8 (6.9–17.2) years were evaluated. Anthropometric measurements and 24-h ambulatory BP monitoring were performed. Estimated salt intake and potassium consumption were determined by 24-h urinary sodium and potassium excretion, respectively. Children with hypertension were compared with normotensive children with respect to gender, age, BMI z-score, urine Na,K, Na/K ratio and estimated daily salt intake. In addition, correlation of urine sodium and urine sodium/potassium to casual and ambulatory BP values were evaluated.
Results: The average estimated salt intake was 9.6 ± 4.3 g/day and it was not different between the hypertensive and normotensive children. Salt intake exceeded the upper limit of the US Dietary Reference Intake in 82% of children. Hypertensive and normotensive children were not different with respect to age, gender, BMI z-score, urine Na, K and Na/K ratio. Estimated salt intake and urine Na/K level were not correlated with ambulatory BP levels. However, office systolic BP measurements were correlated with estimated salt intake ($r = 0.431$, $p = 0.011$).
Conclusions: Our results demonstrate an extremely high salt intake among obese Turkish children. Although hypertensive and normotensive obese children had similar salt and potassium intake, higher salt intake was associated with higher casual systolic BP. Thus, dietary salt reduction interventions along with obesity control programs should be implemented as early as possible.

P-194 BLOOD PRESSURE AND ARTERIAL STIFFNESS IN PEDIATRIC PATIENTS WITH SICKLE/BETA-THALASSEMIA

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Introduction: Blood pressure (BP) in patients with sickle cell disease (SCD) has been reported to be lower than in the general population, but increased risk of cardiovascular disease. The aim of the present study was to investigate the prevalence of BP phenotypes and possible differences in arterial stiffness in pediatric patients with sickle/beta-thalassemia compared with matched controls.
Material and methods: We included in the study 16 pediatric S/b-thal patients and 16 controls matched for age and sex. Controls were otherwise healthy children and adolescents visiting our hypertension center for suspected hypertension. All patients underwent ambulatory BP monitoring, measurement of carotid-femoral pulse wave velocity (cf-PWV). Office hypertension was defined as office BP levels ≥95th percentile for age, gender and sex. Ambulatory hypertension was defined as daytime and/or nighttime BP greater than the 95th percentile for sex and height.
Results: Mean age of the study population was 13.30 ± 4.63 years (34.4% boys). Despite lower office systolic BP levels (115.43 ± 10.03 vs. 123.37 ± 11.92 mmHg, S/b-thal vs. controls, $P = 0.05$), there was no statistical significant difference in 24 h, daytime and nighttime BP. Twenty five % of the S/b-thal patients and 43.8% of the controls presented office hypertension (P = NS), while 18.8% of the S/b-thal patients and 25% of the controls presented hypertension by ambulatory BP levels (P = NS). All S/b-thal patients with office hypertension presented normal ambulatory BP values (white-coat hypertension). None of the S/b-thal patients had daytime hypertension, while all 18.8% presented nighttime hypertension with office normotension <90th percentile (masked hypertension). S/b-thal patients and controls presented equal prevalence of masked hypertension (19%). S/b-thal patients presented also similar levels of cf.-PWV with controls (7.1 ± 1.25 vs. 7.25 ± 1.43 m/s, P = NS) and an 18.8% of the patients presented cf.-PWV levels above the 95th pc for age and sex.
Conclusions: Children and adolescents with S/b-thal present similar prevalence of BP phenotypes and levels of cf.-PWV with pediatric population referred for suspected hypertension. A significant number of children and adolescents with S/b-thal may have nighttime hypertension despite normal office BP levels.

P-195 DO CHILDREN WITH HYPERTENSION AND SNORING HAVE SEVERE OSAS?

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Introduction: Blood pressure elevation is an OSAS-associated morbidity which is apparent mostly in children with apnea-hypopnea index (AHI) > 5 episodes/h (moderate-to-severe OSAS). We aimed to evaluate systolic or diastolic hypertension (≥95th percentile for age, gender and height; Pediatrics 2004;114:555) as predictors of AHI > 5 in children with snoring.
Material and methods: Retrospective cohort of children aged ≥5 years with snoring and adenotonsillar hypertrophy and/or obesity who were referred for polysomnography (PSG) over 10 years. Blood pressure (BP) was measured X3 in the morning after polysomnography and percentiles were calculated for average systolic and diastolic BP. Logistic regression was applied to assess the association of systolic or diastolic hypertension with AHI > 5 adjusted for body mass index z-score and age.
Results: Data of 598 children with snoring (median age 6.6 years;range 5–15.1;26.8% obese) were analyzed. Prevalence of systolic or diastolic hypertension was 9.4% and 8.4%, respectively and frequency of AHI > 5 was 16.7%. Systolic hypertension was a significant predictor of moderate-to-severe OSAS (OR 2.1; 95% CI 1.1–4.1;$P = 0.02$), but diastolic hypertension was not (OR 1.2; 0.6–2.5; $P > 0.05$). The odds of AHI > 5 prior to considering systolic hypertension was 0.20 and after considering its presence increased to 0.40 (Bayes theorem), i.e. for every 6 children with snoring undergoing PSG, 1 had AHI > 5, while for every 4 children with systolic hypertension and snoring tested, 1 was found to have AHI > 5.
Conclusions: In the context of systolic hypertension and snoring in an otherwise healthy child, referral for PSG to rule out moderate-to-severe OSAS seems to be clinically appropriate.

P-196 AMBULATORY ARTERIAL STIFFNESS INDEX (AASI), A SURROGATE MARKER OF ARTERIAL STIFFNESS, IS INCREASED IN OBESE CHILDREN

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Introduction: Increased arterial stiffness is an important risk factor for cardiovascular disease. One way to measure arterial stiffness is the ambulatory arterial stiffness index (AASI), which is the relationship between diastolic and systolic ambulatory blood pressure (BP) over 24 h. In this prospective study, we studied the difference in AASI between obese and lean children.
Material and methods: AASI was calculated from ABPM in 53 obese children (33 girls) and compared with age- and gender-matched 42 healthy subjects (20 girls). AASI was obtained by performing a linear regression analysis of diastolic BP over systolic BP and subtracting the regression slope from 1. Hypertension was defined according to the criteria of American Heart Association. To evaluate inflammation, the blood level of high sensitive C-reactive protein (hsCRP) was measured.
Results: The mean age was 10.6 ± 2.83 years in obese children and 11.3 ± 3.17 years in healthy subjects. Hypertension was determined in three (5.6%) obese children. AASI was significantly increased in obese children compared to healthy subjects (median, IQR: 0.43, (0.13–0.73) versus 0.28, (−0.05–0.85), $p < 0.001$). Heart rate was also higher in the obese group (mean ± SD: 88.2 ± 7.5 versus 83.2 ± 8.4, $p = 0.002$) but pulse pressure and blood pressure values were similar. In a univariate analysis, AASI was independently correlated with indexed casual systolic blood pressure (cSBP, $p = 0.026$), nighttime SBP-standard deviation score ($p = 0.005$), systolic ($p < 0.001$) and diastolic ($p = 0.022$) nocturnal dipping, and hsCRP ($p = 0.02$). In a multivariate analysis, AASI was independently predicted by indexed cSBP ($p = 0.005$) and systolic nocturnal dipping ($p = 0.010$).
Conclusions: This study confirms that AASI and heart rate increased in obese children. AASI calculation is a useful, cheap, and an easy method to evaluate arterial stiffness. Early detection of increased arterial stiffness can help clinicians come up with preventive measures in the management of their patients.

P-197 NOVEL HYPERTENSION DIAGNOSTIC SCORE IN CHILDREN

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Introduction: Accidental discovery of high blood pressure in children usually requires unnecessary consultation, admission and investigation. Most of accidently measured high blood pressure in the clinics or emergency is falsely labelled as "hypertension". Reactive or "white coat hypertension" is the most common cause of that medical staff and family worry. Ambulatory blood pressure monitoring is complex and expensive method to confirm this phenomenon. We invented a novel valid and reliable diagnostic score to use mainly for the newly discovered high blood pressure. This score is considered a simple, effective and rapid tool to avoid the unnecessary work-up and to objectively decide the next action.
Material and methods: The diagnostic score is composed of 10 items with grades for each item 1, 2 or 3. The total minimum score is 10 and the maximum is 30. Score of 12 or less exclude the hypertension and requires no action needed. Score of 25 or more diagnose hypertension and requires consultation, work-up and treatment. Score from 13 to 24 needs monitoring (score 20–24) or reassessment after 48 h (score 13–19). The score is used prospectively in 30 children on their first consultation and or referral for high blood pressure reading (considering hypertension) and then validated to the final diagnosis to confirm or to rule out pediatric hypertension and its management.
Results: The majority of the children (56.6%) have transient or false hypertension and they scored less than 13 with a mean10.9 ± 1. Only 16.6% scored 25 or above (mean 26.4 ± 1.3) and those are considered as hypertensive by the scoring system. 26.6% scored between 13 and 24; the majority of those (75%) are between 13 and 18 and 25% are between 19 and 22. Data is revalidated within 7 days to evaluate and compare the score to the final confirmation or exclusion of hypertension. Results showed 100% of the children scored 25 or above were diagnosed as hypertensive 80% started on medication. Eighty-eight percent of the children scored 12 or less (and one extra child was missed) have normal blood pressure started from the second or third day and confirmed on follow up outpatient clinic visit. 37.5% of the remaining group (score 13–24) had persistently high blood pressure on monitoring and follow up.
Conclusions: The novel pediatric systemic hypertension diagnostic score showed a significant accuracy validity and reliability for the diagnosis and for the recommended further action. Because it is simple, cheap, fast, accurate and reliable, we recommend its mass use in practice at least as a screening tool to select the children require further work-up or management. Larger multicenter study is needed to give more evidence for this new scoring system.

P-198 EVALUATION OF CHANGES IN MYOCARDIAL MECHANICS IN CHILDREN DURING TREATMENT OF MALIGNANT HYPERTENSION

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Introduction: The aims of this study were to evaluate the influence of malignant hypertension (MHT) on left ventricular mass and mechanics using advanced echocardiographic techniques to quantify changes in left ventricular (LV) adaptation following management of hypertension.
Material and methods: Children with MHT ≤16 years of age were identified. Left ventricular assessment was performed retrospectively using M-mode and two-dimensional echocardiography (2DE), in addition to 2D and 3D speckle tracking echocardiography (STE). Hypertension was defined according to the Fourth Report of the National Blood Pressure Education Program. LV mass (LVM) was calculated by Devereux formula and indexed to height ($g/m^{2.7}$). Left ventricular hypertrophy (LVH) was defined as indexed LVM (LVMI) for height z-scores > 1.64SD.
Results: 37 patients (age 9 ± 6, years) with mean glomerular filtration rate (82.11 ± 34.9 ml/min/1.73m^2) and mean SBP z-scores (6.25 ± 2.82), showed abnormal LVM and mechanics at presentation. The mean LVMI z-score was 2.1 ± 2.4, with 22 patients (62%) exhibiting LVH at presentation. There were significant changes for 2DSTE longitudinal strain (LS) (−14.82 ± 4.2 vs.-20.74 ± 2.8, %; $p < 0.001$) and circumferential strain (CS) (−13.74 ± 5.5 vs.-20.65 ± 5.2, %; $p < 0.001$) between baseline and last visit. Similarly, significant changes were observed in 3DSTE LS (p 0.002), CS (p0.020) and radial strain (RS) (p.0.004). LVMI z-scores showed significant reduction (2.1 ± 2.4 vs.01 ± 2.1; $p < 0.001$) over time. These changes though were not related to extent of reduction in the blood pressure despite relatively strong positive association ($r^2 = .6$; p0.65). Adjustment for potential confounders (heart rate and type of antihypertensive medication) has changed the strength of association, but did not reach statistical significance.
Conclusions: Abnormal indices of LV mass and mechanics are evident in children with MHT with changes reversible on management of blood pressure. It is possible that other factors such as class of antihypertensive agent have an impact on LVM and deformation beyond reduction of blood pressure alone.

P-199 COMPARISON OF PULSE WAVE VELOCITY OSCILLOMETRIC MEASUREMENT WITH APPLANATION TONOMETRY IN CHILDREN AND ADOLESCENTS

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Introduction: Pulse wave velocity (PWV) is a well-recognized marker of arterial stiffness. Although the clinical value in children is not yet established its use is increasing in children and adolescents with cardiovascular risk factors. The gold-standard technique is tonometry, but this technique can be challenging, especially when used on children. The purpose of this study was to validate PWV assessment with novel oscillometric device (SphygmoCor XCEL) for use in children and adolescents.
Material and methods: Children and adolescents aged 5–20 years were recruited subsequently. Carotid-femoral PWV (PWVton) was measured by applanation tonometry with the "classic" Sphygmocor device and by SphygmoCor XCEL device (PWVosc). Regression analysis and Bland-Altman plots were used for comparison of the tonometer- to oscillometric-based method. ARTERY Society guidelines criteria were used to assess the performance of the oscillometric device.
Results: Sixty-eight children and adolescents with mean age 11.5 ± 3.6 years, 32 (47.1%) male were included in the analysis. Mean pulse transit time was 81.48 ± 12.55 s by the tonometric method, and 81.63 ± 12.24 s by the oscillometric method (P = NS). Mean PWVton was 4.85 ± 0.81 m/s and mean PWVosc 4.75 ± 0.81 m/s. The mean difference between the two devices was 0.09 ± 0.47 m/s (P = NS) and the accuracy of the oscillometric device was rated "excellent" according to the ARTERY Society guidelines (mean difference less than 0.5 m/s, SD of difference less than 0.8 m/s). Bland-Altman analysis showed good agreement with LoA ranging from −0.83 to 1.01.
Conclusions: The new oscillometric SphygmoCor XCEL device provides equivalent results for PWV values to those obtained by tonometry in children and adolescents. Thus, the SphygmoCor XCEL device is appropriate for assessing PWV in studies in the pediatric population.

P-200 THE ROLE OF SUBENDOCARDIAL VIABILITY RATIO AND EJECTION DURATION IN CARDIOVASCULAR RISK DETERMINATION IN CHILDREN

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Introduction: Subendocardial viability ratio (SEVR) and ejection duration (ED), parameters of pulse wave velocity, have been proposed as important factors of cardiovascular risk determination in adults. The aim of our pilot study was to investigate their role in children and adolescents with cardiovascular risk factors - hypertension, obesity and hypercholesterolemia.
Material and methods: 176 children and adolescents have been included in the study and divided in four groups: 31 children and adolescents with hypertension, 36 with overweight, 49 with hypertension and overweight and 70 with hypercholesterolaemia. They were compared to a control group of 50 healthy individuals. In each patient blood pressure, anthropometrical parameters, and pulse wave analysis (PWA) measurements using applanation tonometry technique were performed and calculations made, including SEVR and ER.
Results: The results show a statistically significant difference in ED ($p = 0.013$), but not in SEVR ($p = 0.074$) in hypercholesterolemia group in comparison to control group. In other research groups, compared to control group, no statistically significant differences in both parameters have been found. In all study groups, SEVR correlated significantly with age and heart rate as well as with central mean pressure. In addition, the correlation between ED and both heart rate and age has also been comfirmed.
Conclusions: In our pilot study the important role of SEVR and ED in early cardiovascular risk determination in children has not been confirmed. However, some results do indicate a potential role of both, at least in hypercholesterolemia, and should be further investigated.

P-201 LOWERING OF DIETARY FRUCTOSE LOAD MAY LEAD TO AN IMPROVEMENT OF ARTERIAL STIFFNESS IN HYPERTENSIVE AND PREHYPERTENSIVE CHILDREN

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Introduction: Non-farmacological treatments of hypertension include lowering the salt load in the diet, consuming more fruits and vegetables, and less saturated fats. Recent research has shown that consuming sweet beverages could elevate blood pressure. It may be postulated that fructose might increase BP by influencing the uric acid genaration pathway. The aim of this study was to assess the influence of decreasing dietary fructose on arterial stiffness.
Material and methods: This study was a prospective clinical analysis of the influence of decreasing the daily fructose load. Forty-one individuals aged 12–18 with elevated blood pressure (70% with hypertension, 30% with pre-hypertension) participated in this study. A questionnaire was used to assess the day-to-day diet. Blood pressure measurements and applanation tonometry were performed to assess arterial stiffness. A dietitian instucted participants to lower their fructose intake by 10%. After 6 weeks of a low-fructose diet, the same measurements were repeated. Patient adherence was validated by assessing the urine uric acid-to-creatine ratio (uUA/Cr). The study was sponsored by the Polish Mother's Memorial Hospital—Research Institute—Young Researcher Grant no 2014/V/8-MN.
Results: After 6 weeks of a low-fructose diet, no changes in casual systolic and diastolic blood pressure were recorded. However, a significant decrease in pulse wave velocity was observed (6 m/s vs 5.6 m/s, $p = 0{,}004$). Aortic systolic blood pressure was also lower (103 vs 100 mmHg, $p = 0.02$), with no changes in diastolic blood pressure. Augmentation index adjusted for heart rate tended to be lower after decreasing diatary fructose (5.0 vs 2,0, $p = 0.07$). There was a significant fall in the uUA/Cr after applying the diet (0.4 vs 0.07 mg/mg, $p < 0{,}001$) which confirmed the participants adherence to the protocol.
Conclusions: Decreasing dietary fructose may improve arterial stiffness without affecting blood pressure. Our data suggests that a low-fructose diet could be a beneficial component of a non-pharmacological treatment in children with hypertension.

P-202 EVOLUATION OF TREATMENT RESISTANT-HYPERTENSIVE CHILDREN WITH RENAL MAGNETIC RESONANCE ANGIOGRAPHY

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Introduction: The incidence of accessory renal artery (ARA) varies between 4%–27% in general population. Although most of the cases with renal artery stenosis (RAS) are due to renal vascular involvement of systemic disorders, intrinsic renal vascular abnormalities such as accessory RAS also account an important percentage of renovascular hypertension (RVH) in childhood. The aim of this study was to investigate renovascular abnormalities with renal magnetic resonance angiography (MRA) in hypertensive children whose blood pressure was difficult to control with medical treatment.
Material and methods: Clinical data and MRA findings of 49 treatment resistant hypertensive patients who were followed in our institution between 2015 and 2017 were analyzed retrospectively.
Results: Forty nine patients diagnosed with essential hypertension were enrolled in this study. The ambulatory blood pressure measurements (ABPM) of all had revealed increased blood pressure load despite of medical treatment with one antihypertensive drug. Mean systolic blood pressure load was 44.37 ± 7,89 and diastolic blood pressure load was 21.08 ± 3,41 and the mean systolic and diastolic blood pressures in patients with RVA were 139.6 ± 3,2 and 82.7 ± 2,8 respectively, despite of anti-hypertensive medication. The mean age of patients with RVA was 13.6 ± 0,4 years old and 10 of 18 patients were male (55%). Eighteen of them (36.7%) were found to have renal vascular abnormalities (RVA) consistent with accessory renal artery, 7 were left, 7 were right and 4 were both sided. Plasma renin levels of 12 patients (66%) were within the normal limits. Renal arterial doppler ultrasound was normal in 10 patients (56%).
Conclusions: Presence of accessory renal artery is a common finding in hypertensive children and MR angiographic investigation should be kept in mind even in the presence of normal plasma renin levels and renal doppler US.

P-203 AMBULATORY BLOOD PRESSURE MONITORING IN PATIENTS WITH MULTICYSTIC DYSPLASTIC KIDNEY

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Introduction: The prevalence of hypertension in children with multicystic dysplastic kidney (MCDK) varies between 0 to 8% based on casual blood pressure (BP) measurements. However; there is limited data on the prevalence of hypertension evaluated by ambulatory blood pressure monitoring (ABPM) in these patients and the rate is up to 20%. The aims of the present study were to evaluate prevalence of and risk factors for ambulatory hypertension in children with MCDK.
Material and methods: This cross-sectional single center study enrolled 31 children (16 male, aged 5–17 years) with MCDK and age, gender and BMI similar 20 healthy children. All subjects were evaluated by casual and ambulatory BP measurements. Ambulatory BP findings were classified according to the updated American Heart Association recommendations in children. Plasma renin activity (PRA) was measured in all patients and controls. Renal ultrasound examinations were performed in the patient group. Total renal volume was calculated and adjusted to height (defined as renal volume index).
Results: There were no differences in SD scores of ABPM values between the patient and the control groups. Three patients (9.6%) were diagnosed as hypertensive based on ABPM; two were classified as masked hypertension and one as sustained hypertension. In addition, two patients had white coat hypertension (WCH) and another two had prehypertension. In the control group, only one patient had WCH.
There was no difference in PRA levels between the patient and control groups. Five patients (16%) had accompanying urological abnormalities in the contralateral kidney and 26 (84%) showed compensatory hypertrophy. None of the three hypertensive patients had contralateral urological abnormality; while only one did not reveal compensatory hypertrophy. There was no significant relationship between renal volume index and ABPM values.
Conclusions: The mechanism(s) of hypertension in patients with MCDK is obscure. Further studies with larger sample sizes are needed to clarify pathogenesis of hypertension in these patients.

P-204 OUTCOMES OF COMBINED MEDICAL AND SURGICAL TREATMENT FOR PEDIATRIC RENOVASCULAR HYPERTENSION

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Introduction: Renovascular disease is an uncommon, but important cause of hypertension in children. In the case of resistant hypertension (failure of medical therapy despite full dose of ≥3 drugs including diuretic), surgical techniques can improve control of blood pressure (BP).
Material and methods: This is a retrospective review of patients who underwent combined medical and surgical in our clinic for renovascular hypertension (RVH) between 1999 and 2016.
Results: A total of 9 children (7 boys, 3 months-14 years, median age 5.5 years) were underwent the complex of clinical, laboratory and instrumental investigations. All of them had normal glomerular filtration rate, different degree of microalbuminuria (30 mg/g > microalbumin/creatinine < 150 mg/g) and second organ damage (heart – 5/9, kidneys −6/9 and eyes – 6/9). The BP level was above the 95th centile for age and height in all children (up to 180/120 mmHg). In clinic everyone were treated for resistant hypertension by different combined therapy. Seven children had renal artery stenosis (3 – unilateral, 4 - bilateral), 1 - had mid-aortic syndrome, 1 – had aneurysm of renal arterial. There was no children with vasculitis or Takayasu arteritis. The angiography was performed in all children, with separate detection of plasma renin activity in renal veins (in all children it was extremely high – 10-20 normative values, in the case of bilateral lesion – it was more than in 1.5 times higher than in the side of stenosis), also 3/9 patient – had underwent to the percutaneous transluminal renal angioplasty during angiography, but it was unsuccessful. All patients received renovascular surgery on the renal arteries: 4 – unilateral, 2 - bilateral autologous surgery, 1 – reimplantation of renal artery and 1 - aortic reconstruction with a synthetic graft. One 3 year old boy with bilateral stenosis had undergo autologous surgery only in one side, because of severe total condition. Fibromuscular dysplasia was the most common morphological diagnosis (6/9). Post-operative complications were hemorrhage (1/9), that indicated repeated surgery (without nephrectomy) and 1/9 had failed hemodynamics, which required resuscitation (was successful). There were no peri- or postoperative deaths. BP was improved in all 9 children and of those 5 (4 - with unilateral, 1 – with bilateral RAS) were cured (havent antyhypertensive therapy in a 1-year after the operation).
Conclusions: All children are needed to measurement the BP, because moderate or even severe elevated blood pressure (for example, as a result of RVH) often is unspecific or have no symptoms. If the case of RVH - medical treatment must undergo immediately, but if it is not enough - surgery should not be delayed. In our exclusively pediatric population angioplasty safely improved blood pressure control in all of patients, more than half of which are cured.

P-205 EXECUTIVE FUNCTION PERFORMANCE IN HYPERTENSIVE CHILDREN

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Introduction: Young hypertensive adults have been reported to have worse performance on neurocognitive testing compared with normotensives. The objective of the present study was to assess the potential early effects of hypertension on the brain in children by evaluating neurocognitive test performance.
Material and methods: We evaluated executive function in hypertensive children compared to normotensive ones in a cross-sectional study. All children and adolescents included in the study underwent ambulatory blood pressure monitoring. Hypertension was defined as daytime and/or nighttime BP greater than the 95th percentile for sex and height. BP index was calculated as mean BP divided by the 95th BP pc specific for each child. To evaluate executive function parents completed the Behavior Rating Inventory of Executive Function (BRIEF), a rating scale that evaluates different aspects of executive function behaviors in the home environment during child's everyday life.
Results: The study population included 38 children and adolescents, mean age 12.18 ± 3.26, 47.4% boys. Fifty % of the children had hypertension (88% secondary causes). Hypertensive children compared to normotensives had higher T scores (52.00 ± 9.42 vs. 39.94 ± 11.63, $p < 0.05$) and percentiles (60.82 ± 25.31 vs. 30.17 ± 25.98, $p < 0.001$) in the clinical scale of organization of materials. The statistically significant differences persisted after adjustment for age, sex, and e-GFR. When daytime and nighttime hypertension were examined separately children with nighttime, but not those with daytime hypertension presented significantly higher values in T scores and percentiles for organization of materials. BP elevation expressed by BP index presented significant correlations with BRIEF scales after adjustment for e-GFR, hemoglobin levels, age and sex. Systolic daytime and nighttime BP index correlated with monitor percentile ($r = 0.72$, $p < 0.01$, and $r = 0.66$, $P < 0.05$, respectively), while diastolic daytime BP index correlated with both monitor percentile and organization of materials percentile ($r = 0.64$, $p < 0.05$ and $r = 0.63$, $p < 0.05$, respectively).
Conclusions: BRIEF scores were higher in the hypertensive children suggesting worse executive function compared with the normotensives

subjects. Correlations of BRIEF scales with BP measures occurred even within the normal limits of the rating scale implying that early effect of BP on the brain may occur within the normal range of the neurocognitive measures.

P-206 PREVALENCE AND AETIOLOGY OF HYPERTENSION IN DANISH CHILDREN

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Introduction: 1) To determine prevalence of hypertension in children <16 years of age in Denmark. 2) To study aetiology of the hypertension and complication at start of pharmacological treatment for hypertension.
Material and methods: Using nationwide health registries with ICD-10 codes from hospitalized and out-patient-clinic patients, we extracted data on all hypertensive children seen from 30th April 2014 to 1st May 2015. The prevalence of hypertension in patients <16 years in Denmark was calculated. Medical records of all hypertensive children from Central and Eastern Denmark, corresponding to approximately 55% of Danish population, were reviewed and patients with antihypertensive pharmacological treatment were included.
Results: The prevalence of hypertension in children <16 years in Denmark is 0.03% (330/1.030.766).
The prevalence of pharmacologically treated hypertension in children was 0.02% (126/553.784). Of the pharmacologically treated, 82% (103/126) had secondary hypertension. The most common cause was of renal aetiology ($n = 68$). Among the renal patients, 68% (46/68) had decreased glomerular filtration rate and 73% (46/63) had proteinuria. Baseline evaluation of target organ damage was done in most hypertensive children with primary or non-renal aetiologies ($n = 58$), echocardiography in 98% (57/58) of cases and retinal exam in 90% (52/58) of cases. Significantly fewer patients with renal aetiology received echocardiography in 63% (43/68) of cases, ($p < 0.001$) and retinal exam in 60% (41/68) of cases ($p < 0.001$). Among the examined patients, hypertensive retinopathy was found in 23% of patients and septum hypertrophy in 34% of patients irrespective of cause.
Conclusions: Considering the low prevalence of hypertension in children <16 years in Denmark, Danish national guidelines for blood pressure screening in children should be revised. Furthermore, considering the large proportion of target organ damage efforts should be done to ensure that hypertensive children undergo thorough evaluation for target organ damage prior to initiation of pharmacological management.

P-207 ARTERIAL HYPERTENSION IN CHILDREN AFTER DIARRHEA-ASSOCIATED HEMOLYTIC UREAMIC SYNDROM

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Introduction: Arterial hypertension (AH) is one of the most common complications after diarrhea-associated hemolytic uremic syndrome (HUS).
Aim of study was to determine the incidence and characteristics of arterial hypertension in children after HUS, to assess circadian rhythm and blood pressure (BP) variability.
Material and methods: The study included 59 children, aged 7.5 ± 1,9 years and follow-up of HUS 5.12 ± 2,07 years. All patients underwent daily blood pressure monitoring (ABPM), determined daily urine losses of protein and albumin.
Results: AH was diagnosed in 16 (27.1%) children (9 with antihypertensive therapy and 7 without it). According ABPM established the prevalence of different forms of AH in children after HUS: prehypertension in 10.2% of patients, white coat hypertension in 13.6% and masked AH in 6.8%. AH was detected with high frequency as those who received dialysis in the acute period of HUS, so without it (29.6% and 20%, respectively). High blood pressure was defined mainly at night due to systolic BP with high load and insufficient night dipping. In some patients obesity worsened the systolic hypertension, which persisted throughout the day. Microalbuminuria was found in 50% of children with hypertension and was closely correlated with the severity of renal damage in the acute period of HUS ($r_s = 0.4$; $p < 0.05$) and AH ($r_s = 0,41$, $p < 0.05$).
Conclusions: Considering the predominantly nocturnal AH in children after HUS, 24 h monitoring of blood pressure is given a key role in its diagnosis. The detection of pathological microalbuminuria is the indication for carrying out ABPM, even if normal office blood pressure.

P-208 A RARE ENDOCRINE CAUSE OF HYPERTENSION: APPARENT MINERALOCORTICOID ACCESS SYNDROME

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Introduction: Apparent mineralocorticoid excess syndrome(AME) is an autosomal recessive disorder characterized by hypertension,hypokalemia,low plasma renin levels and hypoaldosteronism due to deficiency of 11-β-OH-steroid dehydrogenase type 2 enzyme(11 beta-HSD2),which metabolizes cortisol to cortisone.It is one of the rare causes of childhood hypertension.We herein present two siblings who were previously misdiagnosed as Bartter syndrome and then diagnosed with AME syndrome to emphasize the importance of blood gas and serum electrolytes in differential diagnosis of hypertension.
Material and methods: The 14-month-old girl was presented with polydipsia,poliuria,and fatigue.She had a history of having a brother who was being followed with the diagnosis of Bartter syndrome.Physical examination findings were normal except for her weight which was below 3rd percentile.Laboratory findings indicated hypokalemic metabolic alkalosis,hypostenuria and hypercalciuria.Urine output was high as 25-30 cm^3/kg per hour.Although the diagnosis of Bartter syndrome was suspected based on those clinical and laboratory findings,low plasma renin activity (<0.1 ng/mL/h) and high blood pressure levels during the follow up excluded Bartter syndrome.Cortisol to cortisone ratio was found to be high in 24-h urine,indicating AME syndrome.Her brother was also using antihypertensive therapy and he had stage 2 hypertensive retinopathy and left ventricular hypertrophy.He was evaluated for AME syndrome;he had a low plasma renin activity and high 24-h urinary cortisol to cortisone ratio.His renal ultrasound showed bilaterally medullary nephrocalcinosis but the female case had normal renal ultrasound.
Results: Both siblings were diagnosed as AME syndrome and the diagnosis was confirmed by steroid metabolites and homozygote mutations in 11 beta-HSD2 gene.They are being followed in a good condition under the therapy with oral potassium chloride, spironolacton and hydrochlorotiazide treatments.

Conclusions: AME syndrome should be kept in mind in patients with hypokalemic metabolic alkalosis and hypertension.

P-209 ACCURACY OF CASUAL CLINIC BLOOD PRESSURE MEASUREMENT AS A DIAGNOSTIC TEST FOR BLOOD HYPERTENSION IN CHILDREN WITH CHRONIC KIDNEY DISEASES

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Introduction: The ambulatory blood pressure monitoring (ABPM) is not obligatory test for diagnosis of blood hypertension (BH) in children with chronic kidney diseases (CKD). But it know that about 1/3 of children with CKD has masked hypertension which associates with left ventricular hypertrophy. The aim was to determine the accuracy of casual clinic blood pressure measurement (CBPM) for diagnosis of blood pressure hypertension in children with CKD.
Material and methods: Demographic, clinical, laboratory date including CBPM and ABPM were obtained in 359 children with CKD (mean age 12.68 ± 3.13 years; F/M = 1.03; mean eGFR = 84.18 ± 29,6 ml/min/ 1,73m^2). Normal casual blood pressure (BP) defined as systolic blood pressure (SBP) and diastolic blood pressure (DBP) < 90th percentile for gender, age, height. Ambulatory BH was defiened as mean wake and/or sleep SBP and/or DBP levels ≥95th percentile for gender, age, height. White coat hypertension (WCH) was defined as casual clinic SBP and/or DBP ≥95th percentile for gender, age and height but mean wake and sleep SBP and DBP <95th percentile for gender, age and height. Masked hypertension (MH) was defined as normal casual clinic SBP and DBP and mean wake and/or sleep SBP and/or DBP ≥ 95th percentile for gender, age and height.
Results: BH was revealed in 100 pts. (q = 0.28) and in 199 pts. (q = 0.55) by CBPM and ABMP, respectively. The 18 children (q = 0.05) had WCH; 99 pts. (q = 0.27) had MH. The most pts. with MH (n = 56; q = 0.57) had isolate sleep systolic and/or diastolic BH; 4 pts. (q = 0.04) had isolate wake systolic BH; whole day systolic and diastolic BH was diagnosed in 8 (q = 0.08) and 10 (q = 0.1) children, respectively; whole day systolic-diastolic BH was detected in 21 pts. (q = 0.21). Ambulatory BH was revealed in about 1/5 of children with normal casual BP and in more than 2/5 of pts. with casual BP = 90–95‰ (tabl.1). There was high incidence of overdiagnosis of BH by CBPM. The sensitivity/specificity of CBPM for the diagnosis of ambulatory systolic and diastolic BH were 0.6/0,82 and 0.5/0.78, respectively with positive predict value 0.5 for systolic BH and 0.42 for diastolic BH.
Table 1. Prevalence of systolic and diastolic ambulatory blood pressure hypertension by casual clinic blood pressure (CBP) percentile

	CBP < 90‰	90‰ ≤ CBP < 95‰	CBP ≥ 95‰
Systolic wake BH	0.10	0.25	0.5
Systolic sleep BH	0.26	0.45	0.6
Diastolic wake BH	0.11	0.55	0.44
Diastolic sleep BH	0.21	0.41	0.89

Conclusions: The sensitivity/specificity of CBPM for the diagnosis ambulatory BH is suboptimal. We believe that CBPM is not considered adequate for use as single diagnostic test for hypertension in children with CKD.

P-210 VALIDATION OF THE SPHYGMOCOR XCEL DEVICE FOR CENTRAL PRESSURE MEASUREMENT IN CHILDREN AND ADOLESCENTS

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Introduction: Central systolic blood pressure (BP) has been increasing used in adults and seems to be superior to peripheral BP to predict cardiovascular events. The gold-standard technique is tonometry, but this technique can be challenging, especially when used on children. The purpose of this study was to validate central systolic BP assessment with novel oscillometric device (SphygmoCor XCEL) for use in children.
Material and methods: Children and adolescents aged 5–20 years were recruited subsequently. Central systolic BP (cSPton) was measured by applanation tonometry with the "convetional" Sphygmocor device and by SphygmoCor XCEL device (cSPosc). For each patient, the average of the three recordings taken with each device was calculated. Bland-Altman plots were generated for comparison of the tonometer- to oscillometric-based method. The ANSI/AAMI/ISO for Standardization 2013 validation criteria were used to assess the accuracy of agreement between devices.
Results: Five participants were excluded from the analysis due to low quality of recordings, four with tonometric technique and one with both devices. The remaining 67 participants had mean age 11.5 ± 3.7 years, were 31 (46.3%) male and had mean peripheral systolic, and diastolic BP, 121.42 ± 12.64,and 72.69 ± 10.38 mmHg, respectively. cSPosc was strongly correlated with cSPton (R2 = 0.87, P < 0.001). Mean cSPton was 103.23 ± 9.43 mmHg and mean cSPosc 103.54 ± 8.87 mmHg. The mean difference between the two devices was −0.30 ± 3.34 mmHg and fulfilled the AAMI criterion 1 (difference < 5.0 ± 8.0 mmHg). The estimated s.d. (inter-subject variability) was 2.17 mmHg. Bland-Altman analysis showed good agreement with LoA −6.24 to 6.84.
Conclusions: The new oscillometric SphygmoCor XCEL device provides equivalent results for cSP values to those obtained by tonometry in children and adolescents. Thus, the SphygmoCor XCEL device is appropriate for assessing cSP in clinical studies in the pediatric population.

P-211 FIBROMUSCULAR DYSPLASIA – A RARE CAUSE OF RENOVASCULAR HYPERTENSION

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Introduction: Fibromuscular dysplasia is a non-atherosclerotic, non-inflammatory disease that most commonly affects the renal and internal carotid arteries. In patients affected by FMD, renal artery aneurysms (RAAs) may occur with a prevalence as high as 10%.
Material and methods:
Results: We present the case of a11-year-old Caucasian boy who was admitted with complaints of hypertension and was using propranolol and enalapril. He had no known pathological conditions in medical history and family background. He had no inflammatory syndrome and normal blood tests. No changes were found concerning the plasma cortisol and thyroid hormones levels. The urine analysis revealed no signs of proteins, cellular elements. ABPM revealed diastolic nondipper hypertension and hypertensive drug dosage were titrated accordingly. The initial performed abdominal and renal doppler ultrasonographic examination did not show any renal or adrenal pathology.The diagnosis was established incidentally due to an imagistic exam performed for other reasons. There were no end organ damage of heart and eye during four year follow up. A digital subtraction angiography confirmed the left saccular renal artery aneurysms (RAA) and stenosis before the bifurcation and distal left renal artery dysplasia. At follow-up, our patient had percutaneous transluminal renal stent implantation as a definitive treatment.
Conclusions: In patients with uncontrolled hypertension we should suspect renovascular reasons and examination with CT or MRI angiography reveal the pathology.

P-212 ORGAN INVOLVEMENTS IN CHILDREN WITH HENOCH-SCHONLEIN PURPURA AND AFFECTING FACTORS ON KIDNEY INVOLVEMENT

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Introduction: IgA vaskulitis is the most common vasculitis seen in childhood. Though kidney involvement is a major factor affecting prognosis, every organ on body might be affected. The aim of this study is to determine the organ involvement and clinics, to identify the risk factors that increase the nephritis and to determine the long-term prognosis of patients with IgA vasculitis.
Material and methods: 415 IgA vasculitis patients who were followed in 1990 and 2016 in Erciyes University Pediatric Nephrology and Rheumatology Department were included into the study. Patients files were reviewed retrospectively. The patients presenting semptoms, organ involvement, treatment and long-term prognosis were determined.
Results: In our study, 415 patients diagnosed with IgA vasculitis (HSP) were evaluated. Of the patients, 173 (41.7%) were female and 242 (58.3%) were male. The mean reference age was 8.3 ± 3.1 years. Skin involvement was found 100%, joint involvement was 77.1%, GIS involvement was 58.6%, renal involvement was 38.3%, scalp edema was 10.1%, scrotal involvement was 5.3% and central nervous system involvement was 1.6%. Pulmonary involvement was detected in a child. One patient died from pulmonary involvement and one patient died from infectiuos complication possibly due to Eculizumab. When risk factors for nephiritis were assessed, it was found that gastrointestinal involvement was more frequent in those with nephritis ($p = 0.01$). Age, diastolic blood pressure and GIS involvement were correlated with renal involvement, and GIS involvement and diastolic blood pressure are found to be risk factors for renal involvement.
Conclusions: Age at diagnosis, elevated blood pressure and GIS involvement are important findings in predicting the development of nephritis in patients. Patients with GIS involvement should be carefully monitored for kidney involvement.

P-213 LOCAL VS. REGIONAL PATHWAY AND COST-IMPLICATIONS IN THE RENAL MONITORING AND MANAGEMENT OF HENOCH-SCHONLEIN PURPURA

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Introduction: Henoch-Schönlein purpura (HSP) is an IgA mediated commonest systemic vasculitis with risk of long-term renal involvement. The regional pathway recommends a six-month nurse led follow-up with stratification of children into either Standard pathway(SP) or Proteinuria pathway(PP) at one week after presentation with PP cohort at increased risk of renal involvement. Our objectives were to compare our local practice management with regional pathway and study its cost implications.
Material and methods: A retrospective audit involving 50 consecutive children (33♂;17♀) diagnosed with HSP from 2011 to 14 formed the study cohort. The mean age at presentation was four years and one child was followed-up at a different organisation. At presentation blood pressure(BP) and urine analysis(UA) were undertaken in all 50 patients. Twenty-nine patients had normal UA and BP, 16 had abnormal UA and 4 had hypertension. All 49 patients were evaluated at one week and subclassified into SP (47) and PP (2), but additional 5 children in SP developed proteinuria during the study course.
Results: SP cohort had far more than recommended number of health professional(HP) reviews, UA and BP but at random/variable frequency. Majority of PP cohort missed the key intensive scheduled reviews. UA and BP was done at majority of the reviews but none had Primary investigations (urea and electrolytes, urine microscopy and urine protein creatinine ratio) potentially missing out on early identification of renal involvement. None of the seven patients in Proteinuria pathway developed any long-term renal sequelae. Altogether there were at least 171 HP reviews, and 15 inpatient admissions which were unwarranted and far more frequent UA monitoring. This created more anxiety/inconvenience amongst families and stretched HP resources.
Conclusions: We introduced a modified local nurse-led community pathway to standardize care, improve clinical care and cost-efficiency without compromising safety. Adherence to this would have resulted in estimated cost-savings of £48,000.

P-214 FIVE CASES OF GROUP A STREPTOCOCCUS – ASSOCIATED C3 GLOMERULONEPHRITIS: PATHOLOGICAL FINDINGS AND RESPONSE TO TREATMENT

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Introduction: To clarify the hypothesis that group A streptococcus (GAS) infection could trigger C3 glomerulonephritis (C3GN).
Material and methods: We retrospectively reviewed patients in whom C3GN was diagnosed at our institution between 2014 and 2016 based on the findings of a renal biopsy and the 2013 C3 glomerulopathy consensus report. We then performed immunofluorescent staining of nephritis-associated plasmin receptor (NAPlr) antigen, previously described as a nephritic antigen causing poststreptococcal acute glomerulonephritis, in order to extract patients among this population who were positive for NAPlr staining (Oda et al., J Am Soc Nephrol 2005).
Results: Five patients with C3GN (3 boys; mean age: 11.2 years) were enrolled. The average duration of the follow-up was 23.2 months (range: 19–34). Urinalysis showed proteinuria and hematuria in all the patients. However, none had nephrotic syndrome. The renal function of all the patients was normal, but the serum C3 levels were below the normal range (12–26 mg/dL; normal range 80–140 mg/dL) despite normal serum C4. The serum titer of antistreptolysin O antibody (ASO) was elevated (315–527 IU/mL; normal range < 240 IU/mL). GAS infection in three patients was confirmed by pharyngeal cultures. The patients' renal pathological findings were consistent with typical C3GN. Furthermore, the glomeruli of all the patients showed positive staining for NAPlr. Three patients were treated with two courses of intravenous methylprednisolone pulse therapy (MPT) followed by oral prednisolone. The other two were given lisinopril. At the last visit, the proteinuria in the two patients treated with MPT showed improvement. However, with the exception of one patient treated with MPT, four of the patients showed persistent hypocomplementemia.
Conclusions: Together with the elevated ASO, NAPlr staining of the glomeruli suggested an association between GAS infection and C3GN

among a number of the patients. As with GAS-independent C3GN cases, those associated with GAS should be followed up over the long-term.

P-215 NEW COMBINATION THERAPY WITH PREDNISOLONE (PSL), MIZORIBINE AND LISINOPRIL FOR SEVERE CHILDHOOD IGA NEPHROPATHY (IGAN)

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Introduction: Angiotensin converting enzyme inhibitors such as lisinopril has been widely used for childhood IgAN since 2000s. To determine the benefits and harms of the treatment with the combination of PSL, mizoribine and lisinopril (new therapy) compared with the combination of PSL, mizoribine, warfarin and dipyridamole (previous therapy) for severe childhood IgAN.
Material and methods: Retrospective analyses of 34 consecutive biopsy-proven severe IgAN children from August 1998 to November 2016 to compare clinicopathological findings between the 11 children treated with new therapy and 23 cases with previous therapy.
Results: There was no significant difference in onset mode and clinical findings at the start of treatment. As to the pathological findings by Oxford classification, there were significant differences in ratio of M1 (64 vs. 96%, $p = .03$) and C1 + C2 (100 vs 70%, $p = .04$). During the observation period, proteinuria remission was found in all cases with new therapy and 20 cases (87%) with previous therapy. The Kaplan-Meier analysis of proteinuria remission showed that patients with new therapy achieved significantly faster proteinuria remission than those with previous therapy (2.8 M vs. 7.3 M, $p = .005$).
Conclusions: We confirmed the usefulness of new therapy for severe childhood IgAN. Long term outcome will be needed for further investigation.

P-216 RESPONSE TO RITUXIMAB OF NEPHROTIC SYNDROME IN A CHILD WITH IPEX SYNDROME

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Introduction: Immunodysregulation polyendocrinopathy enteropathy X-linked syndrome (IPEX) is a rare genetically determined disorder, caused by a mutation of the gene encoding FOXP3. This causes dysfunction of the regulatory T cells leading to a variety of autoimmunological disorders including: diabetes mellitus, autoimmune enteropathy and hypothyroidism. Among 160 published cases only 12 demonstrated glomerular involvement. Treatment involves bone marrow transplantation (BMT). The report presents a subject with IPEX syndrome who manifested with steroid resistant nephrotic syndrome.
Material and methods: A 12 months old child with neonatal diabetes mellitus and hypothyroidism was admitted to hospital due to onset of proteinuria and oedema. Recurrent eczema and diarrhea had been noted in the previous months. Laboratory results revealed nephrotic range proteinuria, hypoalbuminemia, haematuria, anaemia and thrombocytopenia with normal kidney function. All immunological markers tested including ANA, ANCA, anty GBM antibodies, C3, C4, CH50 were normal. Regulatory T cell count was within reference range. Standard steroid therapy was prescribed and due to lack of remission after 4 weeks, cyclosporine was introduced. Due to onset of neurological symptoms cyclosporine had to be withdrawn.
Results: Genetical testing confirmed the diagnosis of IPEX revealing a deletion in the terminal part of exon of the gene encoding FOXP3 (NM_014009). In preparation for BMT intensified immunosuppression (rituximab and sirolimus) was administered to alleviate symptoms of all immunological disorders. After 2 months of therapy (before BMT) complete remission of nephrotic syndrome was achieved. Extrarenal manifestations of immunological involvement (seizures, eczema, diarrhea) markedly improved.
Conclusions: Steroid resistant nephrotic syndrome concurrent with extrarenal immunological symptoms requires genetic diagnosis of IPEX, a rare congenital abnormality of Treg lymphocytes. Prompt remission of proteinuria can be achieved with Rituximab and sirolimus.

P-217 TGFB1 -509C/T POLYMORPHISM IN CONGENITAL UROPATHIES

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Introduction: In human promoter region of TGFB1 has been described a polymorphism -509C/T (rs1800469) that influences the cytokine production. The aim of the study was to explore the functional significance of TGFB1–509C/T polymorphism for cytokine serum level and genetic predisposition to congenital anomalies of the kidney and urinary tract (CAKUT) among Bulgarian children.
Material and methods: The study includes 97 CAKUT patients and 104 controls from Bulgaria. Patients group consist of hypo/dysplasia ($n = 20$); obstructive uropathies ($n = 36$) and VUR ($n = 41$). The TGFB -509C/T polymorphism was determined using the PCR–restriction fragment length polymorphism method. The quantity of TGFβ1 in sera was detected by ELISA.
Results: Genotype distribution of TGFB1–509C/T polymorphism between total group of CAKUT patients and controls was approximately equal (2 = 0.571; $p = 0.75$). However, there was a tendency for higher frequency of CC-genotype in VUR cases compared to controls (39% vs. 27%; OR = 1.737). In addition, VUR patients with CC-genotype showed significantly higher serum TGFβ1 than TT-genotype (1659.6 ± 203 vs. 1377.5 ± 163 pg/ml; $p = 0.008$). In contrast, CC-genotype was less frequent among patients with hypo/dysplasia than controls (10% vs. 27%; OR = 0.302). The serum TGFβ1 among patients with hypo/dysplasia across the different genotypes were similar. There was a significant difference in serum TGFβ1 between genotypes among patients with obstructive uropathies. CC and CT genotypes were associated with significantly increased TGFβ1 (1498 ± 253 pg/ml and 1535 ± 156 pg/ml) than TT-genotype (1198.8 ± 217 pg/ml with $p = 0.045$ and $p = 0.002$, respectively).
Conclusions: Our data demonstrate that −509*CC genotype in TGFB1 gene is associated with higher cytokine serum level in VUR and

obstructive uropathies and might be involved in pathogenesis of these congenital uropathies.

P-218 POSSIBLE ROLE OF COMPLEMENT CASCADE IN HSPN/IGAN; A CASE REPORT

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Introduction: Henoch-Schonlein purpura (HSP) is the most common vasculitis in children and skin, joints, gastrointestinal tract and kidneys are involved. Renal involvement, range from asymptomatic microscopic hematuria to severe renal disease, is the main origin of the resulting chronic disease. It is believed that HSPN is an immune complex nephritis and the activation of the complement system appears to play a major role in the pathophysiology.
Material and methods: In this case report, we discuss a patient with HSPN who had a complement system defect and did not give complete response to Eculizumab.
Results: The nine years old girl presented with typical clinical picture of HSPN verified with kidney biopsy. Her complements levels were normal. She was given prednisolone, cyclosporine, AZA, cylophosphamide, PLEX and RTX. However, she did not give reasonable response to immunosuppressive medications. Repeated kidney biopsy showed sclerosis in 17 of 26 glomeruli and cellular crescent in the rest of the glomeruli. So, we decided to start eculizumab with an experience coming from IgA nephritis. Initially, she gave a partial response to Eculizumab. Finally, ESRD was developed. Homozygous mutation in Factor H gene was detected in a study of a complement system. While she was on peritoneal dialysis, she experienced with osteomyelitis on the left foot. She died possibly because of complication of osteomyelitis.
Conclusions: We detected a mutation in factor H gene showing the activation of the complement system appears to play a major role in etiopathogenesis of HSPN. We think Eculizumab may be a rescue treatment option in children who are unresponsive to conventional treatment if it can be given early period of disease.

P-219 RESPONSE OF A CHILD WITH FAMILIAL DYSREGULATION OF THE COMPLEMENT SYSTEM TO ANTI-C5 ANTIBODY

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Introduction: C3 glomerulopathy is a new entity of a heterogeneous group of glomerular diseases associated with acquired or genetic abnormalities of complement alternative pathway (AP) components. It is characterized by predominant C3 deposits in the mesangium and along the glomerular basement membrane (GBM). Based on Electron Microscopy, C3 Nephritis was previously classified as Type III Membranoproliferative Glomerulonephritis. Advances in the understanding of processes involved in the pathogenesis through immunoflouresence classified MPGN as being mediated by immune complexes or by complement dysregulation that leads to persistent activation of the complement AP. As well Classification that is based upon the pathogenic process helps to direct the clinical evaluation and to provide disease-specific treatments. C3 glomerulopathy may present with haematuria, proteinuria or renal failure.
Material and methods: We report the clinical progress of a child with a progressive glomerulonephritis through her first decade of life, whose renal biopsy confirmed Proliferative glomerulonephritis with endocapillary and Membranoproliferative patterns, Mesangial and subendothelial deposits with occasional subepithelial deposits on electron microscopy. Immunoflouresence profile was consistent with C3 related glomerulonephritis. Assessment of the complement panel revealed low C3, CFB & CFH levelS and increased activity of the alternative complement system. Genetic study revealed a familial homozygous miss ensue mutation of CFH.
Results: The optimal treatment of C3 glomerulopathy remains unknown. It is currently based on the use of angiotensin-converting-enzyme inhibitors (ACEI) and angiotensin II-receptor blockers (ARB), sometimes associated with immunosuppressive therapy. Plasma infusion and plasma exchange therapy was used by different authors. We also report patient's response to the humanized anti-C5 antibody.
Conclusions: A renal biopsy report that denotes C3 glomerulopathy should prompt investigation of the complement system. Identifying patients with dysregulation of the complement system is mandatory as they may benefit from therapeutic use of the complement inhibitor.

P-220 LONG FOLLOW-UP OF IGA NEPHROPATHY WITH SEVERE DEBUT IN CHILDHOOD

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Introduction: We try to know the prognosis in adulthood of IgA nephropathy (IgAN) with onset in infancy with severe nephritic or nephrotic syndrome.
Material and methods: Retrospective study. Inclusion criteria: patients with diagnosis of IgAN in renal biopsy report. We recorded age at biopsy, criteria to carry it out, number of glomeruli, pathology report and glomerular filtration rate (GFR), presence of hypertension (HTA), proteinuria and treatments, at debut, at one year of follow-up, at the time of transition to adults and at the last review.
Results: 14 patients with a mean age at onset of 10 years. Percutaneous biopsies were performed with radiological control except one. The average number of glomeruli was 16. Light microscopy: mesangial hypercellularity (11), cellular crescent (3), relevant interstitial fibrosis with tubular atrophy (3). In Immunofluorescence was detected IgA (14); C3 (11) and C4d (2/2). Initial treatment: ACEI or ARAII in all patients, we associated mycophenolate in 2; prednisone, and cyclophosphamide bolus in 2; prednisone and azathioprine in one. One year after debut: 2 patients had nephrotic proteinuria, 1 more than 1 g and 1 lower. One had HTA. GFR was less than 90 ml/min/1.73 m2 in a patient. The average pediatrics follow-up was 71 months. In the transition to adults: 2 had microalbuminuria; 2 proteinuria > 1 g. Two patients had HTA. 1 patient had been transplanted, 1 had CKD stage 2 DOQI and the others normal GFR. In the last review, in adult units, no more patient had CKD; 2 had nephrotic proteinuria. The remaining patients received ACEI and had proteinuria <1 g or were untreated.
Conclusions: The chances of preserving a normal GFR of patient with IgAN, despite debut with a severe nephrotic or nephritic syndrome in childhood, are high. Although proteinuria is considered one of the most important prognostic factors, in our series, patients with high proteinuria maintained normal renal function.

P-221 ANCA-ASSOCIATED VASCULITIS WITH RENAL INVOLVEMENT IN CHILDREN: CLINICAL CHARACTERISTIC AND OUTCOME

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Introduction: ANCA-associated glomerulonephritis(GN) is a rare disorder in children, presentation and outcomes vary significantly among patients. Renal involvement contributes significantly to the morbidity with high numbers of patients progressing to end-stage kidney disease.
Material and methods: We review 5 patients (2boys, 6-17 years, mediana 8.0) with ANCA-positive GN from Belarus National Center for pediatric nephrology patients. Caspase-1, TNFα, IL1β, BAFF, RANTES, VEGF, TGF1β, IgG ANCA (PR3, MPO, BPI, elastase, lysozyme, lactoferrin, cathepsinG), 24 h monitoring of blood pressure(BP), ECHO-CG, carotid intima media thickness(cIMT), left ventricles mass index (LVMI), relative thickness of left ventricles wall (RTLV), body mass index (BMI), serum lipids, glucose, uric acide levels, eGFR were measured.
Results: The median time from presentation until first clinical or serologic signs was 12 months (6-29mo). The main organs involved were: renal in 5(100%); respiratory tract 5 (100%); joints 2(40%); skin rash 3(60%); eyes 2(40%). PR3 and MPO-ANCA, caspase-1, TNFα, IL1β, BAFF, RANTES, VEGF, TGF1β levels were positively correlated with disease activity and organ involvement. Hypertension was observed in 100%, dilatations of LV in 40%, reduced ejection fraction in 20% of patients. LVMI, RTLV, BMI and cIMT were higher compared with healthy($p < 0.05$). The mean glucose, uric acid and cholesterol level was significantly higher than in healthy($p < 0.05$), lipoproteins of low and very low density prevailed.
Conclusions: 1 patient was treated with plasmapheresis and prednisolone pulses at presentation, required dialysis. He died after 12 months from the manifestation of the disease due to infection. Induction therapy in others included cyclophosphamide pulses and steroids, maintenance azathioprine and steroids. Cardiovascular, metabolic and immune abnormalities were correlated with disease activity and duration. Such patients are at high risk of early development of cardiovascular complications requiring early correction.

P-222 PEDIATRIC RAPIDLY PROGRESSIVE GLOMERU LONEPHRITIS: 11 YEARS EXPERIENCE

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Introduction: In this study, we aimed to evaluate etiology, clinicopathological features and outcome of rapidly progressive glomerulonephritis (RPGN) in children followed at pediatric nephrology unit, Alexandria University over 11 years (2004–2014).
Material and methods: We investigated 41 children (22 girls and 19 boys, mean age 8.3 ± 3.3 years) with crescentic glomerulonephritis (CrGN) retrospectively, and compared them to another 25 cases (16 girls, 9 boys) who had RPGN (glomerulonephritis with rapidly progressive renal failure) but with less than 20% crescentic glomeruli in renal biopsy (non-crescentic RPGN).
Results: Among CrGN group, type II (immune complex) accounted for 78% of cases and type III (pauci-immune) accounted for the other 22% who were chiefly ANCA-negative (88.9%). When comparing crescentic and non-crescentic RPGN, children with CrGN were more hypertensive ($P = 0.015$), more oligo-anuric ($P = 0.05$), they had lower hemoglobin ($p = 0.002$), and were more proteinuric ($p = 0.027$).There was no significant difference between them regarding serum creatinine ($p = 0.746$) nor dialysis dependency at presentation ($p = 0.152$).Glomerular sclerosis ($p = 0.001$), tubular atrophy ($p = 0.001$) and interstitial inflammatory infiltrate ($p = 0.023$) were more frequent in biopsies of crescentic than non-crescentic group. At latest follow-up; 17.3% of CrGN cohort had passed away, 23.1% had achieved complete remission and around one fifth had developed end-stage kidney disease. Crescentic lupus nephritis showed the highest risk of disease progression. Worst outcome of CrGN was associated elder age ($p = 0.027$), presence of hypertensive emergencies at presentation ($p = 0.037$), and evidence of chronic histologic changes i.e. sclerosis ($p = 0.007$), interstitial fibrosis ($p = 0.003$), tubular atrophy ($p = 0.027$) and fibrous crescents ($p = 0.002$), but not with the percentage of crescents per se ($p = 0.754$).
Conclusions: In context of RPGN, no laboratory investigation could neither be relied upon in prediction of severity of pathology, nor replace renal biopsy while taking a therapeutic decision. Additionally, the percentage of crescents beyond 20% did not prove to be a useful outcome indicator.

P-223 RECURRENT HENOCH-SCHOENLEIN PURPURA WITH SEVERE COMPLICATIONS: A CASE REPORT

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Introduction: Henoch-Schoenlein purpura (HSP) is the most common vasculitis in childhood. The main symptom is a palpable purpura, but inflammation can also affect joints and multiple organs, especially the gastrointestinal tract and the kidneys. Mostly the disease is self-limiting and has a good prognosis. In complicated cases, treatment is often guided by the degree of renal involvement.
Material and methods: We report on a 3-year-old girl who presented with palpable purpura and abdominal colicky pain. Due to severe enterocolitis, therapy with prednisolone was started. But despite treatment recurrent episodes with acute abdomen including intraperitoneal bleeding and ileus occurred. She developed a nephritic-nephrotic syndrome (macrohematuria, arterial hypertension, protein/creatinine up to 24 g/g and azotemia with blood urea up to 30 mmol/l) which was treated with mycophenolate initially and due to inadequate response, cyclosporine was added. Four weeks later, complex focal seizures occurred and EEG and cMRT showed severe pathological findings. After a skin biopsy proved IgA-vasculitis, therapy with methylprednisolone-pulse combined with rituximab treatment achieved a remission. In the next 4 months, she claimed about abdominal distension, intermittent abdominal pain and petechial lesions recurred. A further progress could be stopped by increasing the dose of prednisolone and continuing combined immunosuppressive therapy with cyclosporine and mycophenolate. However, 6 weeks later she developed a severe relapse with acute abdominal pain and nephrotic-nephritic syndrome. Despite monitoring showed complete CD20 depletion in peripheral blood, rituximab treatment in combination with methylprednisolone pulses were again successful. Immunosuppression was continued with daily prednisolone, cyclosporine, mycophenolate and low dose methylprednisolone 200 mg/m^2 every 2nd-4th week. After 6 months during an upper airway infection, there was another episode with acute

abdominal pain and nephritic syndrome but without purpura. We decided to stop the regular methylprednisolone infusions and continue rituximab treatment every 3–4 months despite no detectable CD20 in peripheral blood. Tragically, her father suffered a fulminant type of IgA nephritis with rapid progression to end-stage renal disease 3 years ago.
Results:
Conclusions: HSP is mainly a self-limiting disease. Genetic predispositions can increase the likelihood of developing severe complications and aggressive immunosuppressive therapy is required irrespective by the severity of renal involvement.

P-224 CLINICAL AND IMMUNOPATHOLOGICAL STUDY OF GLOMERULOPATHIES WITH DOMINANT MESANGIAL IMMUNOGLOBULIN M EXPRESSION IN BELARUSIAN CHILDREN

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Introduction: IgM nephropathy (IgMN) is a primary glomerulopathy characterised by diffuse mesangial immunoglobulin M (IgM) deposits. IgMN has been proposed as variant of glomerulopathy between minimal change disease and focal segmental glomerulosclerosis. It usually presents with steroid resistant or dependent nephrotic syndrome.
Material and methods: As material for research have served 8 kidneys biopsies patients with diffuse mesangial IgM positive on immunohistochemistry, whether there was single or dominant positivity, executed on the basis of the Republic Center of Pediatric Nephrology and Renal Replacement Therapy in Minsk/Belarus between January 2013 and March 2017. All the cases included were classified as primary glomerulopathy.
Results: The age of patients ranged from 3 year to 16 years; there were 4 boys and 4 girls. The initial manifestation was nephrotic syndrome in 5 of 8 children, of whom 2 were steroid resistant, three were steroid dependent. Three of nephrotic patients also presented high blood pressure level and haematuria. One child had nephrotic proteinuria with microhaematuria, two children were haematuric with non-nephrotic proteinuria. The most frequent glomerular morphological finding was diffuse mesangial hypercellularity: in 6 of 8 patients. In two cases, focal and segmental glomerulosclerosis was found. Tubulointerstitial changes (atrophy of the tubular epithelium, interstitial fibrosis or inflammation) were found in 5 of 8 cases. All biopsies had diffuse mesangial dominant positivity for IgM and in five cases there were also focal and segmental deposits of the immunoglobulin A, G and/or C3, C1q complement fractions.
Conclusions: The clinical manifestations and morphological findings in pediatric patients with IgMN is highly variable. The long-term prognosis of these patients and their response to steroid therapy are still unknown. Therefore, this question demands the further studying.

P-225 MISDIAGNOSE OF SYSTEMIC LUPUS ERYTHEMATOSUS AFTER CHILDHOOD THROMBOCYTOPENIA, RESULTED WITH CHOREA MANIFESTATION DURING PREGNANCY-TREATED SUCCESSFULLY WITH NO APPARENT ADVERSE EFFECTS ON FETAL GROWTH

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Introduction: Idiopathic Thrombocytopenia is associated with LES, but ANA and LEcells may not be positive during early childhood. Alike, Chorea gravidarum, can be caused by other systemic collagen diseases, as (SLE).
Material and methods: We report a case of chorea gravidarum caused by SLE. This case during early childhood by age of 8, presented with Idiopatic Trombocytopenia but ANA and LE cells, were negative at presentation. Bone biopsy resulted: immune trombocitopenia. Treated by Prednisone tbl.4 mg. By age of 16teen she had hand-palm pain and was diagnosed as Arthritis and no specific treatment. Her first pregnancy with no problems.Second pregnancy age 25, had Blood pressure PA: 140/90 mmHg. By 25 gestational weeks she presented with uncontrolled choreiform movements of the hands, body and head, diagnosed Chorea gravidarum.Treated with Haldol tbl. 10 weeks.
Results: Because of a positive autoimmune history, she was examined for LES and resulted with active disease and all Antibodies positive Anti DNA IgG, Anti ds DNA; C3, C4, CH50: decreased); CCP: neg;. Brain MRI (twice): acute ischemic lesions-microinfacts, also Glial zones corresponds to lacunar chronic ischemic lesions of vascular ethiology, suspicious as thrombotic complications. Abdominal Ultrasound findings within normal. Proteinuria: 500 mg/24 h. Fetal growth on ultrasonography was normal. A female infant, normal weight 38 gw.
Conclusions: Misdiagnose of LES at time and as a consequence, a new trigger initiated symptoms of active Cerebral LES manifestation with Chorea. Even during pregnancy is treatable and with no consequences to fetal growth and pregnancy outcomes. Criteria for thrombocytopenia fellow up should be set, to prevent complication of LES.

P-226 HENOCH-SCHÖNLEİN PURPURA WITH POLYNEU ROPATHY A CASE REPORT

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Introduction: Henoch-Schönlein purpura (HSP) is the most common vasculitic disease of childhood. Palpable purpura on the skin, joint, gastrointestinal tract and renal involvement are common symptoms of HSP. Central or peripheral nervous system involvement is rare. In this study, we present a case applied to our clinic with stepping gait and polyneuropathy.
Material and methods: Case report.
Results: An 8-year-old male patient first applied red-colored rashes on his legs, swellings and pain in ankle which starts 10 days before. He used only anti-inflammatory treatment. Five days after gait disturbance was occurred. On physical examination, both lower extremities were palpable purpura sharply raised especially more on extansor side of the extremities. In neurological examination, deep tendon reflexes were normal in lower extremities, muscular strength was decreased. His feet could not dorsofleksion and he had steppage gait. Other systems were normal. Laboratory investigation showed normal levels of B12, proteinuria (19 mg/m2/d) and microscopic hematuria. The lumbosacral and cranial MR was normal. Electromyography (EMG) had

an affect on the peroneal nerve bilateraly. We started IV methylprednisolone (500 mg/day) treatment for 3 consecutive days and continued with oral prednisolone (2 mg/kg/day). Also he was startted to the physical therapy program. The walk of the patient was significantly improved rapidly.
Conclusions: Neurological involvement in HSP can be seen very rare in childhood and good response can be obtained with steroid treatment.

P-227 IS THE INCIDENCE OF POSTINFECTIOUS GLOMERULONEPHRITIS INCREASING?: CASES WITH POSTINFECTIOUS GLOMERULONEPHRITIS FROM A SINGLE CENTER IN NOVEMBER-DECEMBER 2016

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Introduction: There is a certain increase in the number of patients with postinfectious glomerulonephritis (PIGN) in department of pediatric nephrology at Cukurova University during November–December 2016.
Material and methods: Clinical, laboratory and follow up results of the 13 patients that have PIGN are studied. Pediatricians diagnostic approaches and the reasons for referral were evaluated in patients with clinical symptoms of PIGN.
Results: The patients are diagnosed PIGN in the presence of haematuria, proteinuria, evidence of recent streptococcal infection, low serum C3 levels with normalization on 8 weeks follow up. Thirteen patients (5 male, 8 female) are conducted into the study. Mean age was 9 (3–15). All of patients have had recent infection. The symptoms of patients at the time of admission were oedema (70%), macroscopic haematuria (23%), hypertension (15.3%) and respiratory distress (15.3%). Seven of the patients (53%) had blood pressure higher than 95.percetile for age, gender and hight. Nonnephrotic proteinuria was detected in 11 patients. All patients had low C3 levels. Two of the patients had cardiac systolic dysfunction, pulmonary edema, and a referral to our hospital. There was improvement in clinical findings with fluid restriction and diuretic therapy in patients follow-ups. Because of nephrotic proteinuria and acute kidney injury, kidney biopsy was done to one of the patients and diagnosed with diffuse proliferative glomerulonephritis.
Conclusions: PIGN is observed with higher incidence in the developing countries. In these patients with different clinical presentation, it was aimed to draw attention of pediatricians.

P-228 IGA NEPHROPATHY IN CHILDREN: A SINGLE CENTER EXPERIENCE

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Introduction: Immunoglobulin A nephropathy (IgAN) is the most common biopsy-proven primary glomerulonephritis in pediatric population. The clinical presentation of the disease in children varies from microscopic hematuria to end-stage kidney disease. The aim of this study was to evaluate the clinical features, treatments, and follow-up results of the children with IgAN.
Material and methods: This is a retrospective study. Patients who had a histopathologically proven diagnosis of IgAN during the period of 2000–2017 were included in the study.
Results: A total of 30 patients with IgAN were analyzed; 17 (56.7%) patients were male and 13 (43.3%) were female. The mean age at onset and admission (± standard deviation) of patients with IgAN was 123.53 ± 50.64 (range:31–237, median: 112.5) and 132.27 ± 50.32 (range:34–240, median: 129.5) months respectively. Kidney biopsy was performed approximately 16 ± 22.12 months after onset of the disease. The mean follow-up time was 44.53 ± 38.81 (range:6–150, median: 34.5) months. Nineteen (63.3%) patients presented with recurrent macroscopic hematuria, five (16.7%) with microscopic hematuria ± proteinuria, five (16.7%) with nephritic syndrome, and one (3.3%) with proteinuria. IgA co-deposition with C3 (n:16, 53.3%) was the most common finding in the IF study. At present, 17 (56.7%) patients have minor urinary abnormalities, three (10%) have active renal disease while 10 (33.3%) patients are followed with normal renal and urinary findings. None of the patients developed renal failure. Hypertension was present in only 2 (6.7%) patients.
Conclusions: The majority of children with IgAN in this study were admitted with recurrent macroscopic hematuria and most of them had a good prognosis. We think that favorable prognosis may be possible with early diagnosis of IgAN in children.

P-229 TREATMENT FOR INDUCTION OF REMISSION IN LUPUS, WHAT TO CHOOSE?

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Introduction: We aimed to evaluate the treatment regimens efficacy for induction in systemic lupus erythematosus (SLE) patients admitted in our referral center.
Material and methods: We performed a longitudinal retrospective study on 18 patients diagnosed with SLE between 2008 and 2017. The inclusion criteria were a diagnosis of SLE, clinical, laboratory and treatment data available until remission or 12 months since diagnosis date for those who did not achieve remission. We evaluated all patients and determined the Systemic Lupus Erythematosus Disease Activity Index 2000(SLEDAI2K). A decrease of 4 points or more of SLEDAI2K was used to assess the presence of the remission. We also calculated the SLEDAI2K-variance as the difference between SLEDAI2K at diagnosis and at the end of induction.
Results: The median age at diagnosis was 14.08 years [12.81;15.77], with female predominance 88.88%. Corticoid (CT) only, associations CYC with CT and MM with CT was administered to 6, 10 and 2 patients, respectively. Sixteen patients achieved remission and the median induction time was 3.5 months [2;6.5]. The median SLEDAI2K at baseline was 20[10;27.25]. The median SLEDAI2K-variance was 10.5[5.75;22] (see Table I). Univariate logistic regression showed no statistically significant differences between treatment regimens and induction with a p-value of 0.73.
Conclusions: With respect to ALMS Trial, our study confirmed that choosing a treatment regimen does not influence induction. We note an improvement of SLEDAI2K when MM is associated with CT vs. when CYC is associated with CT vs. CT only for induction. We admit as limitations of the study the number of patients and that disease activity at diagnosis may bias the selection of agents to treat.

Table I. Lot characteristics depending on treatment regimen

	Total	CT	CT + CYC	CT + MM	*P* value
N	*18*	*6 (33.33%)*	*10 (55.55%)*	*2 (11.11%)*	–
Male	*2 (11.11%)*	*2 (33.33%)*	*0 (0%)*	*0 (0%)*	*0.105*
Age (years)	*14.08 [12.81; 15.77]*	*15.08 [13.31; 16.83]*	*13.66 [12.36; 14.83]*	*14.04 [13.33; 14.75]*	*0.468*
Baseline SLEDAI 2 K	*20 [10; 27.25]*	*10 [7.7; 14.8]*	*21 [17.6; 26.4]*	*35 [20; 50]*	*0.105*
Achieving remission	*16 (88.88%)*	*6 (100%)*	*8 (80%)*	*2 (100%)*	*0.406*
Time to remission (months)	*3.5 [2; 6.5]*	*4 [2; 6.2]*	*2.5 [2; 4.6]*	*3 [2;4]*	*0.636*
SLEDAI 2 K variance	*10.5 [5.75; 22]*	*6 [4.4; 11.3]*	*12 [7.6; 22]*	*23.5 [11; 36]*	*0.22*

P-230 IDIOPATHIC MEMBRANOUS NEPHROPATHY IN CHILDHOOD

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Introduction: To know the clinical manifestations, treatment and prognosis of membranous nephropathy (MN) in childhood.
Material and methods: Multicentric, retrospective study (1980–2016). Inclusion criteria: children under 16 years old with diagnosis of MN in renal biopsy report. Exclusion criteria: secondary MN (hepatitis B, C, LES, etc.…) We recorded clinical manifestations at debut, age at biopsy time, pathology report and treatment. We assessed clinical situation at last review.
Results: 13 patients. Mean age at onset 9.8 years (range 6–15). Nine girls and 4 boys. At debut: eight nephrotic syndrome (NS) with microscopic hematuria (2 hypertension, 1 of them with renal failure (RF). Three nephrotic proteinuria (NP), 2 of them with macroscopic hematuria. Two non-NP. Findings in light microscopy were MN type I in 2, type II in 5 and type III in 2. Immunofluorescence detected IgG deposits in 11/13 with C3 in 10/13. In four, there were slight deposits of IgA (2) and IgM (2) and C1q (2) with chains lambda y kappa (1). Treatment included ACEI or ARA-II in 9, with prednisone 6. One was treated with acetylsalicylic acid. Nine received prednisone (6 NS and 3 NP). Two children with RF were treated with IECA, prednisone and cyclophosphamide. At mean 9 years of follow-up (1-11 years): patients with NS: three complete remission; two proteinuria >1 g; two proteinuria <1 g (1 with RF and hypertension normalized blood pressure and glomerular filtration) and one are in chronic RF G3b stage without proteinuria. Patients with NP: two complete remission and 1 proteinuria <1 g. Patients with proteinuria <1 g: 1 developed NP and 1 proteinuria <1 g with chronic RF G2 stage.
Conclusions: In childhood, the most typical presentation of idiopathic MN is nephrotic syndrome. The prognosis is not so bad as referred in adults and the chances of recovery renal involvement are high despite severe debut.

P-231 EARLY CARDIOVASCULAR MANIFESTATIONS IN CHILDREN AND ADOLESCENTS WITH AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE: A MONOCENTRIC STUDY

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Introduction: This study aims to describe the cardiovascular manifestations in children and adolescents with Autosomal Dominant Polycystic Kidney Disease (ADPKD) and detect their relation to kidney injury and type of gene mutation.
Material and methods: Twenty patients aged from 7 to 19 years old were included. Cardiovascular evaluation involved measurement of Blood Pressure (BP), Indexed Left Ventricular Mass (LVMI), Pulse Wave Velocity (PWV) and carotid Intima-Media Thickness (cIMT). Patients were classified according to percentile reference values of these parameters in healthy children. The 95th percentile was the highest level of normal values. Prehypertension was defined as average systolic BP or diastolic BP levels greater than or equal to the 90th percentile, but less than the 95th percentile. Mutation in Polycystic Kidney Disease 1 (PKD1) gene was available in 16 patients.
Results: Hypertension, large LVMI, high PWV and increased cIMT were observed in 4 (20%), 2 (10%), 4 (20%), and 8 (40%) patients respectively. Hypertension was not correlated to either high PWV or increased cIMT. Linear correlation was observed between LVMI and PWV ($r^2 = 0.272$, $p = 0.018$) and also between LVMI and cIMT ($r^2 = 0.223$, $p = 0.035$). The median age of patients with high PWV, increased cIMT, BP > 90th percentile and large LVMI was 9.5, 13, 13 and 18 years old. Patients with increased cIMT and BP > 90th percentile presented higher levels of urine microalbumin/creatinine ratio ($p = 0.041$, $p = 0.022$ respectively). Finally, hypertension was more frequent in patients without missense mutation ($p = 0.044$).
Conclusions: High PWV and increased cIMT indicating arterial stiffness and hypertrophic vasculopathy may be present in children with ADPKD regardless BP status, and their levels are significantly related to LVMI. Vascular injury seems to precede the development of hypertension and left ventricular hypertrophy. Increased cIMT could be indicative of low grade albumin excretion. Missense mutation of PKD1 is probably associated with a better cardiovascular prognosis.

P-232 FIRST SUCCESSFUL CONCEPTION INDUCED BY A MALE CYSTINOSIS PATIENT

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Introduction: Cystinosis is a rare autosomal recessive lysosomal storage disease characterized by multi-organ cystine accumulation, generalized proximal tubular (renal Fanconi syndrome) and progressive glomerular dysfunction, followed by various extra-renal organ manifestations. Azoospermia of unknown origin is the main cause of infertility in all male cystinosis patients. Although spermatogenesis has shown to be intact at the testicular level in some patients, no male cystinosis patient has been reported yet to have successfully induced conception.
Material and methods: A 27-year old male infantile nephropathic cystinosis patient, transplanted at the age of 11 with a properly functioning kidney graft, was diagnosed with azoospermia of unknown origin. For realizing the wish of the patient and his wife of having children, a percutaneous epididymal sperm aspiration (PESA) procedure was performed, revealing the bilateral presence of mature spermatozoa. After a third ICSI attempt, six out of eight obtained oocytes could be fertilized and two weeks after double embryo transfer, a pregnancy test resulted positive. A dichorial diamniotic (DCDA) twin was identified by ultrasound at seven weeks of gestation, and at 36 weeks and six days the twin was born via an uncomplicated caesarean section in an apparently healthy status and having normal birthweights.
Results: We present the first successful conception ever reported, induced by a male renal transplant infantile nephropathic cystinosis patient through percutaneous epididymal sperm aspiration (PESA) followed by intracytoplasmatic sperm injection (ICSI).
Conclusions: Our observation opens a new perspective in life for many male cystinosis patients whom nowadays have become adults, by showing that despite azoospermia fathering a child can be realized. Notably, we demonstrated that sperm of epididymal origin in selected male cystinosis patients can be viable for inducing successful conception. In addition, our findings raise questions about the possibility of sperm cryopreservation at a young age in these patients.

P-233 CYSTEAMINE AFFECTS NOT ONLY OSTEOCLASTIC ACTIVITY BUT ALSO OSTEOBLASTIC DIFFERENTIATION

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Introduction: The cornerstone therapy of nephropathic cystinosis (e.g., cysteamine) was reported to induce toxicity (bony, cutaneous, vascular, neurologic and muscular). There are no experimental data to explain these clinical symptoms. The aim of this study was to evaluate the effect of cysteamine on bone cells.
Material and methods: Osteoclasts were differentiated from circulating monocytes isolated from blood, in patients with nephropathic cystinosis (n = 7) and age- and gender-matched healthy controls (n = 7).Cells were treated with increasing doses of cysteamine (50-200 μM/L) during the differentiation process or after a normal differentiation process to evaluate the impact of cysteamine on differentiation and resorption, respectively. Osteoclast differentiation and activity were evaluated by tartrate resistant acid phosphatase (TRAP) staining and bone resorption assays. Osteoblasts were differentiated from murine mesenchymal stem cells (MSC). Differentiation was evaluated trough (Osteocalcin,Osterix,Collagen 1 time course expression), alkaline phosphatase and Von Kossa staining. A cysteamine (10–50–100-200 μM/L) dose response was performed on bone cells.Cell toxicity and proliferation tests using LDH levels measurements in the culture media and BrdU assay.
Results: Cysteamine has no impact on osteoclastic differentiation, but inhibits osteoclastic resorption in all patients and all controls.Cysteamine stimulates osteoblastic differentiation and maturation (by both qRT-PCR and quantitative mineralization assessment) when low concentrations of cysteamine (50 μM/L) were used; however, this effect was no longer observed at higher concentrations (100–200 μM/L).A cytotoxic effect of cysteamine is ruled out since LDH levels in the medium was not modified at the different experimental conditions, however BrDU assays show a decreased proliferation at high doses of cysteamine (200 μM).
Conclusions: In vitro low doses of cysteamine impair osteoclastic resorption and stimulate osteoblastic differentiation and mineralization. However when used at higher doses, an inhibitory effect on osteoblastic proliferation is observed. These results could explain, at least partly, the bone phenotype observed in patients receiving high doses of cysteamine.

P-234 RENAL EVALUATION OF PAEDIATRIC PATIENTS WITH TUBEROUS SCLEROSIS COMPLEX (TSC) IN A NATIONAL TSC MULTIDISCIPLINARY CLINIC

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Introduction: Tuberous Sclerosis Complex (TSC) is a dominantly inherited multisystem disorder characterised by development of hamartomas (benign lesions) in several organs. Previous studies have shown that renal manifestations are common in TSC and also the most frequent cause of TSC-related death in affected individuals. With the recent discovery of targeted therapy with mTOR inhibitors, early recognition of renal involvement allowing early surveillance and subsequent prompt therapy is now more important than ever.
Material and methods: To review the current renal evaluation of paediatric patients attending the TSC MDT clinic data were collected from electronic medical records pertaining to molecular diagnosis, renal surveillance and presence or otherwise of sequelae affecting the renal or other systems.
Results: Data were obtained for 40 children (23 girls, 17 boys). Current age ranged from 23 weeks to 18 years (mean 8.7 years). Twenty nine (72.5%) patients were diagnosed at <1 year of age. Genetic data were available for 29 patients, 15 had a confirmed TSC1 mutation and 14 a confirmed TSC2, including one with a contiguous PKD1 deletion. Details of abdominal imaging were available in 37/40 (92.5%). There was wide variability in timing of interval scanning and modality. However 34/37(92%) had had abdominal imaging within the last 3 years with 8/40 having had abdominal MRI. Thirteen patients had documented renal lesions (seven with AML lesions, five cystic lesions, one with both) on follow up imaging. Four patients with renal lesions have been commenced on mTOR inhibitors, one having previously had embolization. Presence of renal lesions was broadly associated with a more severe phenotype affecting other systems.
Conclusions: Tuberous Sclerosis is a clinically heterogeneous disorder with manifestations ranging in severity and in the case of renal sequelae age of onset. With the advent of molecular targeted therapy this population has demonstrated the requirement for early and regular surveillance.

P-235 PATIENTS WITH NEPHROPATHIC CYSTINOSIS DISPLAY CORTICAL BONE IMPAIRMENT

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Introduction: As patients receive cysteamine and improve survival, bone impairment appears to be a novel complication of nephropathic cystinosis. Even though the exact underlying pathophysiology is unclear, six hypotheses are discussed: copper deficiency, bone consequences of severe hypophosphatemic rickets during infancy, cysteamine toxicity, abnormal thyroid metabolism, chronic hypoparathyroidism and/or direct bone effect of the *CTNS* mutation. The objective of this study was to evaluate bone status in the French Crystobs study.
Material and methods: In addition to clinical data, bone status was evaluated with biomarkers (copper, ALP, PTH, 25-D, 1–25D), DXA (spine) and High Resolution peripheral Quantitative Computed Tomography (HR-pQCT) at the tibia and radius. Results were compared to age- and gender-matched healthy controls (1:2 basis) from the local reference cohorts (OFELY in women, STRAMBO in men, and VITADOS in teenagers). Results are presented as median (range), and non-parametric Mann-Whitney tests were performed.
Results: At a median age of 22.5 (10.2–34.6), 10 patients with nephropathic cystinosis were included (2 supportive therapies, 2 hemodialysis, 6 renal transplantation; 7 females; 2 diabetes, 0 tobacco exposure, 6 patients with a past/ongoing rhGH therapy): 7 out of 10 patients complained of a bone symptom (past of fracture, bone deformations and/or bone pains). Biochemicals and spine DXA did not show any significant abnormalities. Using tibial HR-pQCT, significant decrease in cortical area (81(53–120) *vs* 116(48–141) mm^2, $p < 0.05$) and cortical thickness (850(520–1100) *vs* 1200(480–1680) μm, $p < 0.05$), with a trend towards decreased total volumetric bone mineral density, were observed in cystinosis patients in comparison to controls. There were no differences for trabecular parameters.
Conclusions: Bone impairment (rather cortical than trabecular) in nephropathic cystinosis is a reality: 70% of patients in this pilot study displayed significant bone symptoms, during teenage or young adulthood. This new complication should be known by physicians because of the potential dramatic impact on quality of life.

P-236 INHERITED URINARY TRACT DISORDERS DIAGNOSED DUE TO NEONATAL ULTRASONOGRAPHIC SCREENING

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Introduction: Neonatal abdominal ultrasonography (US) is an easy and safe method of diagnosis of congenital urinary tract abnormalities. In the era of antenatal examinations, it is not routinely recommended. The aim of the study was to estimate the frequency and type of CAKUT and other urinary tract disorders found in ultrasonographic screening performed in regional neonatal unit.
Material and methods: During last 3 years (March 2014–March 2017) 941 out of 1153 neonates born in our hospital (87%) had US performed by the same radiologist or nephrologist. Children with abnormal urinary tract (renal pelvis A-P diameter > 5 mm, dilated ureter, abnormal volume and/or echogenicity of the kidney, renal cysts) were followed up in nephrological outpatient till the final diagnosis. US was repeated in 4–6 weeks and then every 3 months and other imaging techniques (voiding cystourethrography, radioisotopes and intravenous pyelography) ordered when needed.
Results: 165 neonates were transferred to the clinic for abnormalities found in the first US. Among them 18 finally presented with various kinds of CAKUT: 5 –hydronephrosis (1 – bilateral), 5 – megaureter (2 – bilateral), 2 – hypoplastic kidney, 2 – posterior urethral valves (PUV), 2 – multicystic dysplastic kidney, single cases of segmental renal dysplasia and horshoe kidney with hydronephrosis and 1 with ADPKD (2.0% of all neonates included into the study and 11.5% of those with abnormal postnatal US). Only 5 of these children (26.3%) had any data about abnormal prenatal USG, neither ADPKD nor PUV patients among them.
Conclusions: Neonatal US is a key procedure in early diagnosis of CAKUT and other structural abnormalities of urinary tract even in the cases with normal intrauterine US. For it is safe, easy and cheap one could introduce US screening into routine examination of neonates, especially in regions with poor standard of prenatal care.

P-237 IN VITRO SPLICING ASSAYS TO DETECT INTRONIC PATHOGENIC VARIANTS IN INHERITED KIDNEY DISEASES

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Introduction: So far, many intronic variants have been detected in genes responsible for inherited kidney diseases.To test the pathogenic nature of these intronic variants, transcript analysis is necessary to see splicing abnormalities caused by these variants. However, mRNA is not usually available and in those cases, it's very difficult to prove these variants as pathogenic and therefore, an alternative strategy should be developed. In this report, we determined the pathogenic nature by both transcript analysis extracted from patients' samples and *in vitro* assays using minigenes in addition to the *in silico* analysis using Human Splicing Finder for two patients with inherited kidney diseases.
Material and methods: Case 1 is a 35 year-old-man who was pathologically diagnosed with glomerulopathy with fibronectin deposits. A novel variant of c.5888 -2A > G (IVS37-2A > G) was detected in *FN1* gene by genome DNA sequencing analysis. RT-PCR using mRNA extracted from urine sediments and minigene assays were conducted.Case 2 is a 16 year-old-male who was clinically diagnosed with Lowe syndrome. A variant of c.940-11G > A was detected in *OCRL* gene (IVS11-11G > A) by genome DNA sequencing. RT-PCR extracted from peripheral blood leukocytes and minigene assay was conducted.
Results: Case 1. Twelve base pair (bp) deletion from the first nucleotide of exon 37 was detected by RT-PCR analysis in both patient's sample and minigene assay. Case 2. Nine bp insertion between exon 10 and 11 was detected by both patient's sample and minigene assay. These splicing abnormalities were exactly predicted by *in silico* splicing assay.
Conclusions: We detected the same splicing defects in both patients' derived cells and minigene assay shown to be two cases. *In silico* analysis also predicted the same results. From these results, the assays were as proof of the pathogenic nature of intronic variants.

P-238 MAFB MUTATION IN END STAGE RENAL DISEASE

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Introduction: MAFB mutation causes multicentric carpotarsal syndrome, with severe skeletal deformations and nephropathy. We report a 14 years old patient who was hospitalized to our clinic because of end stage renal disease (eGFR 10,1 ml/min/1,73 m2), and hypertension.
Material and methods: The patient history was negative, the physical examination did not show any significant findings. Because of the unknown cause of the ESRD kidney biopsy was performed, and we have collected samples for genetic testing. The result of the kidney biopsy was focal, but dominantly diffuse glomelural sclerosis, with the fusion of the podocyte foot process, and chronic tubolointersticial nephritis.

Results: Genetic testing (new generation sequencing) has found a mutation in the gene which is encoding MAFB (vmaf avian musculoaponeurotic fibrosarcoma homolog B) protein. The mutation involved the DNA binding site of the protein which was not published in the literature. Imaging studies revealed that, our patient did not have any bone deformities, or carpotarsal osteolysis.
Conclusions: This is the first published case when MAFB mutation did not cause complete multicentric carpotarsal syndrome, just severe nephropathy.

P-239 A NOVEL HETEROZYGOUS DE NOVO 7BP FRAMESHIFT DELETION IN PBX1 CAUSING BILATERAL DYSPLASTIC KIDNEYS AND HYPOPLASTIC CLAVICLES IDENTIFIED BY WHOLE-EXOME SEQUENCING

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Introduction: Congenital anomalies of the kidney and urinary tract (CAKUT) represent the primary cause of chronic kidney disease in children. Many genes have been attributed to the genesis of this disorder. Recently, haploinsufficiency in *PBX1* caused by microdeletions has been shown to result in bilateral renal hypoplasia and other organ malformations. Here, we report on a 14 year old male patient with congenital bilateral dysplastic kidneys and hypoplastic clavicles with a novel *PBX1* variant.
Material and methods: Presuming a syndromic origin, we performed SNP array analysis to scan for large copy number variations (CNVs) followed by whole-exome sequencing (WES). Sanger sequencing was done to confirm the variant's *de novo* status.
Results: SNP array analysis did not reveal any microdeletions or –duplications larger than 50 or 100 kb, respectively. WES identified a novel 7 bp frameshift deletion in *PBX1* (c.413_419del, p.Gly138Valfs*40) resulting in a loss of function. This variant cannot be found in 60.000 individuals of the Exome Aggregation Consortium (ExAC) browser. Furthermore, the ExAC database does not list any frameshift or nonsense variants in *PBX1* indicating that this gene is intolerant to a loss of function. The *de novo* status could be confirmed by Sanger sequencing.
Conclusions: We report on a 14 year old male patient with congenital bilateral dysplastic kidneys and hypoplastic clavicles. By WES, we identified a novel *de novo* 7 bp frameshift deletion in *PBX1*. Our findings expand the spectrum of causative mutations in *PBX1*-related CAKUT as small deletions have not been described as pathogenic yet. In this case, WES proved to be the apt technique to detect the variant responsible for the patient's phenotype, as a multitude of genes is involved in CAKUT and other techniques of whole-DNA analysis like SNP array analysis do not have the diagnostic accuracy to identify small deletions.

P-240 WAYS TO DEFINE DEFECTS OF TUBULAR EPITHELIAL CELL FUNCTION IN POLYCYSTIC KIDNEY DISEASE (PKD): COMPARISON OF CELLS FROM ARPKD AND NPH PATIENTS

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Introduction: PKD-related cellular defects are best understood genetically or in epithelial (cell) models analysing cell proliferation and signalling. Use of primary patient-derived renal epithelial cells provides a unique opportunity to study and quantify cell properties in epithelial function-related assays. Apart from the analysis of differences due to specific mutation, it appears interesting to correlate the severity of the renal disease i.e. GFR or organ size / morphology with alterations in epithelial cell function determined *ex vivo*.
Material and methods: We culture urine-derived renal epithelial cells (URECs) of patients with genetically confirmed causes of PKD, autosomal recessive polycystic kidney disease (ARPKD) and nephronophtisis (NPH), and respective controls. Populations of primary cells obtained within 14 days of culture are being characterized by different criteria and tested in 3D cell culture conditions. Using micro-patterned adhesion chips, defined surface shape, size, and extracellular matrix coating allow quantitative analysis of epithelial spheroid formation, a measure of the cells' potential to perform epithelial morphogenesis.
Results: URECs from two cohorts of patients (5–6 each) with ARPKD and NPH (mostly NPHP-1 mutation), respectively, are compared in cell culture to generate quantitative characteristics that can be correlated to clinical parameters and progress of PKD. Cells are tested with respect to their proliferation rates, formation of cell-cell junctions in monolayer, induced formation of cilia, and their capacity to build spheroids both in matrigel and when challenged by defined adhesion on chips allowing high content analysis of spheroid properties.
Conclusions: Detection of genotype and / or disease state specific renal epithelial cell properties will provide a better understanding of the mechanism and progression of the disease process in renal epithelium and may provide options for testing of pharmaceutical intervention.

P-241 ROLE OF GENETIC INVESTIGATIONS IN DIAGNOSING CHILDHOOD CKD: A POPULATION SURVEY

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Introduction: Accurate diagnosis of primary renal disease may be difficult, particularly for late presenters but is vitally important to determine disease progression and recurrence risk following transplantation. This retrospective population study aims to review current use of genetic investigations, and assess the utility of genetic techniques to achieve a diagnosis for children with significant CKD in the future.
Material and methods: All children ≤18 years with CKD stage ≥3A in the North East of England and North Cumbria were included. Clinical information and previous investigations were reviewed using hospital records and the regional genetics database.
Results: 68 children with CKD ≥3A were identified in our region, and 33/68 (49%) have had genetic investigations performed. A diagnostic genetic abnormality was identified in 14/33 (42%) of those investigated. Patients were more likely to have undergone genetic investigations if they had associated extra-renal problems. No association was found between investigation and patient age or CKD severity. Diagnostic genetic mutations were most commonly found in children in the syndromic non-CAKUT (congenital abnormality of the kidney and urinary tract) group. A diagnostic genetic change was present in 8/20 (40%) of patients with a condition not identifiable on ultrasound scan, and a combination

of ultrasound and genetics revealed a diagnosis in 56/68 (82%) children.

Diagnosis	Number	Genetic Investigated	%	Diagnostic mutations	%
CAKUT Isolated	24	4	17	0	0
CAKUT plus extra-renal diagnoses	16	7	44	1	6
Non-CAKUT Isolated	7	5	71	2	29
Non-CAKUT plus extra-renal diagnoses	19	15	79	11	58
Unknown	2	2	100	0	0
Total	**68**	**33**		**14**	

Conclusions: The combined use of genetic investigations and ultrasound scan has a high diagnostic yield in children with CKD. As whole exome sequencing becomes cheaper, utilizing this as an early non-invasive investigation in children with CKD appears inevitable but will manifest new challenges.

P-242 A VARIETY OF PHENOTYPES REFLECTED BY GENOTYPES AND LAMININ β2 EXPRESSION ON GLOMERULUS IN PIERSON SYNDROME

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Introduction: Null variants in LAMB2 cause Pierson syndrome (PS), a severe congenital nephrotic syndrome with ocular and neurologic defects, and show completely negative expression of laminin β2 on glomerular basement membrane (GBM). Conversely, it has been reported that some cases with LAMB2 non-truncating variants tend to show milder phenotypes. Here we report 3 cases with atypically milder phenotypes including 2 siblings and 1 case with typical PS whose LAMB2 variants were detected by next generation sequencing (NGS). We identified biomolecular mechanisms for leading to milder phenotypes in PS.
Material and methods: Cases 1–3 are milder cases and have not developed end-stage renal disease (ESRD), ocular abnormalities nor psychomotor retardation despite they are 4 years, 1 year and 2 years old, respectively. Case 4 showed the typical clinical signs for PS with development of ESRD at 3 months old. All 4 cases were identified *LAMB2* pathogenic variants. Molecular study was conducted to clarify the backgrounds that lead to the clinical differences in those cases.
Results: Cases 1–2 were possessing a frame shift (c.225delC) and a missense (c.2095G > C, p.Gly699Arg) variant. Case 3 was possessing a frame shift (c.5073_5076dupCCAG) and a splice site variant (c.3797 + 5G > A). Case 4 had a homozygous missense variant (c.482 T > C, p.Leu161Pro), which is located in laminin N-terminal domain: an important site for interaction of laminin and lead to severe phenotype. Cases 1–3 showed laminin β2 expression on GBM although Case 4 exhibited completely negative. The transcript analysis for Case 3 revealed that the splice site variant resulted in the generation of 4 different splicing patterns including 1 normal transcript which lead to the mild phenotype.
Conclusions: We clarified the genetic and biomolecular backgrounds for atypically milder phenotypes in PS. These results can explain a variety of phenotypes in PS and NGS can reveal genetic backgrounds in atypical cases with inherited kidney diseases.

P-243 THE MULTIDISCIPLINARY PAEDIATRIC RENAL GENETICS CLINIC: A MODEL FOR MAINSTREAMING GENETICS IN MEDICINE

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Introduction: Our understanding of the genetic factors involved in many conditions is continually evolving, and the availability of genomic testing is rapidly expanding, creating a necessity for new services, new ways of working, and modified professional roles in order to maximise the benefit to patients and families. The multidisciplinary clinic is one model to achieve this and integrate genetics into mainstream medicine.
Material and methods: In order to evaluate processes and outcomes, as well as inform future practice, a retrospective clinical audit of the first 14 months of clinic operation was conducted.
Results: The joint Royal Children's Hospital and Victorian Clinical Genetics Services Renal Genetics Clinic was established in February 2016, with the aim of improving diagnosis rates and informing management in patients with suspected genetic renal disease. The clinic team includes a nephrologist, clinical geneticist, genetic counsellor, and administrative support, with each member of the team contributing unique expertise and benefitting from opportunities to cross-train and up-skill. Patients typically attended for one or more of: clinical diagnostic assessment, genetic counselling, genetic/genomic testing, and/or research recruitment; and benefitted from multidisciplinary case review prior to, during, and after clinic appointments. Outcome data for 34 patients is presented, including instances of genetic diagnosis leading to altered management.
Conclusions: Results of the clinical audit provide insight into the first paediatric multidisciplinary renal genetics clinics in Australasia. Outcome data has demonstrated benefits for patient care, as well as opportunities for biological relatives to access genetics services. Anecdotal evidence supports professional satisfaction with the service model and professional development opportunities by participating staff.

P-244 THE PREVALENCE AND INCIDENCE OF ATYPICAL HEMOLYTIC UREMIC SYNDROME IN IRAN- A SYSTEMATIC REVIEW AND META-ANALYSIS

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Introduction: Hemolytic Uremic Syndrome (HUS), being more prevalent in infants and children, is recognized by the triad of acute kidney injury, microangiopathic hemolytic anemia, and thrombocytopenia. The aim of this study is to find the incidence and prevalence of HUS, the etiology, clinical presentation, and the outcome of Iranian patients.
Material and methods: We explored the following search engines: PubMed, EMBASE, OVID, SCOPUS, Web of Sciences, Google Scholar, Google, health.barakatkns.com, IranMedex, MagIran, SID, dociran, PDFiran, and ganj.irandoc were used. Besides that, all online university databases for thesis, abstract books of local or international congresses, between January 1985 and January 2016 searched. We did hand-searching to identify pertinent cross references. Quality assessed by three reviewers using STORB checklist and the risk of bias evaluated. Chi-squared and I-squared statistic tests were applied to weigh heterogeneity between the studies in effect measures. Point prevalence, proportion, and incidence are calculated. The result would be expressed as 95% confidence intervals.
Results: A total of 25 articles and two abstract of congress containing 6728 patients met all the inclusion criteria and were eligible for the final

analysis. Considering 1258 patients with hemolytic uremic syndrome, the incidence was 15.92 pmp and annual incidence was 0.4 pmp per year. In children less than 15 years, the incidence was 72.38 pmp and annual incidence was 1.85 pmp/year. Atypical HUS was identified in 601 patients (18%) between 1976 to 2015. The incidence was 34.57 pmp and the annual incidence was 0.88 pmp/ year of children aged less than 15 years old. The mean annual incidence rate increased from 0.12 in time period before 2000 and gradually increased to 0.74 in 2010 afterward (p-value < 0.001).. It contributed to 12.98% of acute kidney injury and 5.48% (95%CI: 3.5–7.9) of CKD and end stage renal disease .
Conclusions: The rate of disnosis of HUS especially atypical was incereasing during the previousfour decades.AHUS consisdt significant number of AKI and ESRD. the result about mortality rate of aHUS was inconclusiive Further prospective study is required.

P-245 IMPACT OF GENETIC MODIFIERS IN FEMALE PATIENTS WITH ALPORT SYNDROME

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Introduction: Alport syndrome (AS) is caused by mutations in the genes COL4A3, COL4A4, and COL4A5. In approximately 85% of patients with AS a X-linked inheritance of mutations in the COL4A5 is found. Therefore, mostly males are affected. In male patients a distinct genotype-phenotype correlation could be detected. In female patients with a heterozygous mutation, a variable intrafamilial and interfamilial penetrance exists, resulting in a broad spectrum of clinical symptoms ranging from mild microhematuria to severe AS. In these patients genotype-phenotype correlation is less-well described so far. The type of mutation, genetic modifiers or the degree of mosaicism following lyonization of the X chromosome are discussed as possible causes for the phenotypic variability. Up to now only little and inconclusive information concerning this topic can be found in the literature.
Material and methods: The main focus of this upcoming study is the identification of factors influencing and explaining the phenotypic variability in female patients with X-linked AS. To address this issue the following steps are planned: I. Female patients with different clinical phenotypes and a heterozygous mutation in COL4A5 will be included in this study. II. X-inactivation in peripheral blood cells as well as in renal progenitor cells/podocytes (urine; kidney tissue if available) will be investigated. III. By using whole exome sequencing the study cohort will be examined in further genes for genetic modifiers. IV. The results of the molecular examinations will be correlated to the clinical phenotype.
Results: –/–.
Conclusions: By identifying risk factors (e.g. genetic modifiers, skewed X-inactivation) in female patients, female relatives with a high risk for developing severe AS can hopefully be identified at an early stage of disease in the future and thus be treated in good time in order to slow down the progression of disease and to avoid dialysis and kidney transplantation. Detailed information: julia.hoefele@tum.de

P-246 THE NEUROLOGIC MANIFESTATIONS OF ATYPICAL HEMOLYTIC UREMIC SYNDROME IN CHILDREN

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Introduction: Atypical hemolytic uremic syndrome (aHUS) is a rare heterogeneous disorder which is defined by the association of hemolytic anemia, thrombocytopenia and acute renal failure. Uncontrolled complement activation results in systemic complement-mediated thrombotic microangiopathy and subsequent renal and extra-renal organ damage. The information in the literature about extra-renal involvement including neurologic system is rare. So we aimed to determine clinical, laboratory and imaging characteristics of aHUS patients with neurologic manifestations.
Material and methods: The demographics, clinical, laboratory and imaging characteristics of aHUS patients with neurologic involvement among 170 patients in the national Turkish aHUS Registry were recorded.
Results: The study included 45 patients with neurologic manifestations, of which 12 had other extra-renal system involvement. The neurological system manifestations in decreasing order were seizure, encephalopathy, hemiparesis, loss of vision, unconsciousness, headache and hallucination. Cranial MRI, which was performed in 33 patients, showed that 17 had abnormal findings and revealed increased signal intensity and/or limited diffusion ($n = 8$), signal changes secondary to hypertension ($n = 3$), increased intensity and hypoxic changes ($n = 1$), and focal changes ($n = 5$). The four of 45 patients (8.9%) died. The eculizimab was given to 33 (73.3%) patients. The ratio of having normal serum creatinine level and the rate of hematologic remission were higher and having renal replacament at discharge is lower in patients treated with eculizumab than patients not treated with eculizumab.
Conclusions: The neurologic involvement is the most common extra-renal manifestation of aHUS and it is a major cause of morbidity and mortality. If eculizumab is available, it can be first-line and life-saving treatment for neurologic involvement in children with aHUS resulting in rapid normalisation of renal and hematological findings. In addition, the present findings may increase the awareness of clinicians about neurologic manifestations and lead to earlier diagnosis and treatment of patients with such manifestations.

P-247 CAREGIVERS' ATTITUDE TOWARDS FAMILY PLANNING AND TIMING OF DIAGNOSIS IN AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE

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Introduction: Several aspects in the management of Autosomal Dominant Polycystic Kidney Disease (ADPKD) are still controversial, including family planning and testing for disease presence in at-risk individuals. However, the attitudes of caregivers and the underlying arguments towards these topics have never been studied.
Material and methods: We performed an online survey aiming to assess the attitudes of European pediatric and adult nephrologists, as well as clinical geneticists, regarding these issues.

Results: A total of 410 caregivers (53% male, mean (SD) age of 48 (10) years) responded, including 216 pediatric nephrologists, 151 adult nephrologists, and 43 clinical geneticists. While the 3 groups agreed to encourage clinical testing in asymptomatic ADPKD minors and adults, only geneticists would recommend genetic testing in asymptomatic at-risk adults ($P < 0.001$). Statistically significant disagreement between disciplines was observed regarding the ethical justification of prenatal genetic diagnosis, termination of pregnancy and pre-implantation genetic diagnosis (PGD) for ADPKD. Particularly, PGD is ethically justified according to geneticists (4.48 (1.63)), whereas pediatric (3.08 (1.78); $P < 0.001$) and adult nephrologists (3.66 (1.88); $P < 0.05$) appeared less convinced by the option of PGD.
Conclusions: Our survey suggests that most caregivers support clinical testing of at-risk minors and adults in ADPKD families. However, there is no agreement for genetic testing in asymptomatic offspring and for family planning, including PGD. The present data highlight the need for a consensus among caregivers to avoid conflicting information in the management of ADPKD families.

P-248 CLINICAL AND GENETIC HETEROGENEITY OF EARLY-ONSET NEPHROCALCINOSIS IN CHILDREN

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Introduction: Nephrocalcinosis (NC) due to inborn metabolic diseases is a very rare condition associated with progression to CKD during childhood. In recent years, multiple monogenic causes of NC have been identified. However, the prevalence of each monogenic gene in children with NC has not yet been studied. The arm of the study was to identify the structure and prevalence of monogenic causes of NC in pediatric cohort.
Material and methods: 35 children (29 M/6F) with NC were examined. The median age of patients was 5.0 (IQR: 1.0; 8.0) years. We examined of blood electrolytes levels; urinary excretion of Ca, P, urate (Ur), oxalate. Ultrasound grades (G) of NC were as follows: G1 - mild echogenicity around medullary pyramid borders; G2 - moderate echogenicity around and inside pyramids; and G3 severe echogenicity of entire pyramids. Genetic analysis was performed in all children using by direct Sanger sequencing in 60%, next generation sequencing in 37.1%, and fluorescence in situ hybridization in 2.9%.
Results: We identified 36 causative mutations of 13 inherited kidney diseases with NC, including 13 (36.1%) novel mutations. Most of them was detected in patients with HHRH (38%) (Table). Children with NC had hypercalciuria (51.4%), hypercalcemia (20%), reduction phosphorus reabsorption with phosphaturia (25.7%), hyperoxaluria (14.3%), increased urinary Ur excretion (8.3%). Changes of acid-base composition were identified in 20% of cases (acidosis 17.1%, alkalosis 2.9%). Hypercalciuria was found in 100% of children with Dent disease 1, HHRH, Lowe syndrome, FHHNC, dRTA, BS3 and in 25% cases with IC1. Hypercalcemia was revealed in 100% patients with IN2, Williams syndrome, 75% children with IC1 and 40% children with HHRH. Hyperoxaluria and increased Ur excretion had 100% patient with PH1 and LNs, respectively. NC G1 was found in 35.3% of cases, NC G2 in 50% and NC G3 in 14.7% patients. Renal function was normal (CKD1) in 48.6% of children. CKD2 had 37.1% and CKD3–14.3% of children. Patients with NC G2–3 had CKD2–3 more frequently compared with children with NC G1: 83% vs. 47% ($p = 0.03$), RR = 3.17 (95% CI:1.03–9.7).

Phenotype	Patients, % (n)	Gene	Mutation
Dent disease 1	20.0 (7)	*CLCN5*	c.731C > T c.1909C > T* c.842C > T c.1497C > T c.2119C > T
Primary hyperoxaluria, type 1 (PH1)	14.3 (5)	*AGXT*	c.121G > A c. 1020A > G c.508G > A c.33_34insC
Hypophosphatemic rickets with hypercalciuria (HHRH)	14.3 (5)	*SLC34A3*	c.1453C > T* c.846G > A c.1094-3C > T* c.1382G > A* c.586G > T* c.1018A > T* c.1058G > T c.1394C > T*
Infantile hypercalcemia 1 (IH1)	11.4 (4)	*CYP24A1*	c.1186C > T c.428_430delAAG c.475C > T c.442 T > C
Lowe syndrome	8.5 (3)	*OCRL*	c.1351G > A c.1847G > C*
Lesch–Nyhan syndrome (LNs)	8.5 (3)	*HPRT1*	c.359 T > G* c.599G > A c312C > G*
Cystinosis nephropathic (CN)	5.7 (2)	*CTNS*	57 kb del c.681G > A c. 433 C > T
Infantile hypercalcemia 2 (IN2)	2.8 (1)	*SLC34A1*	c.1438_1440delTTC* c.464 T > C
Distal renal tubular acidosis (dRTA), AD	2.8 (1)	*SLC4A1*	c.1765C > T
dRTA with deafness	2.8 (1)	*ATP6V1B1*	c.172A > C*
Familial hypomagnesaemia with hypercalciuria and nephrocalcinosis (FHHNC)	2.8 (1)	*CLDN16*	c.453G > T
Bartter syndrome, t ype 3 (BS3)	2.8 (1)	*CLCNKB*	c.1972G > C*
Williams syndrome	2.8 (1)	*ELN, LIMK1, RFC2*	D7S613, D7S1870, D7SELN1, D7SELN CA

*novel mutation
Conclusions: The present study demonstrated the clinical and genetic background of children with early-onset NC. Dent disease 1 was the most prevalent inherited kidney disease among pediatric cohortwithNC.Hypercalciuria and hypercalcemia were the most common causes of NC. Increase in severity of NC leads to progression of inherited kidney disease with decline of kidney function.

P-249 THE NEW MUTATION IN PKD2 FOR EARLY ONSET OF ADPKD

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Introduction: Only 2% of patients with Autosomal Dominant Polycystic Kidney Disease (ADPKD) are presented with early clinical manifestations, before 15 years of age. Out of that number, patients rarely present symptoms in perinatal period, which is clinically indistinguishable from Autosomal Recessive Polycystic Kidney Disease. PKD2 gene on chromosome 4q21 is responsible for 15%–20% of all ADPKD and generally has milder clinical presentation and prognosis than ADPKD caused by PKD1 gene. The huge clinical variability of ADPKD has been described within the same family.
Material and methods: We ultrasonographically disclosed the female index case with early onset (EO) form of ADPKD whose clinical presentation started from the birth. During her pregnancy in 29th year of life, gynecologist noticed olygohydramnios and polycystic kidneys in fetus. The male premature newborn, was born cyanotic, with big belly and died after 1 h. Pathologist diagnosed enlarged cystic kidneys and hypoplastic lungs. Indexs mother had ADPKD and has been transplanted in her 40-es. Mothers sister has only one asymptomatic kidney cyst, while their parents were healthy.
Results: Genetic analysis showed pathogenic heterozygous PKD2:c.2118 + 1G > A variant in intron 10 (NGS) which has not been previously described in other patients listed in NHLBI Exome Variant database or the ExAc database.
Conclusions: Despite the claims that mutations in PKD2 have a mild clinical presentation, our patient developed EOADPKD and gave birth to seriously ill son, affected by EOADPKD with fatal outcome. At the other hand, some other family members have not been affected or only minimally affected. This newly described mutation of PKD2 gene might be responsible for EOADPKD. In that sense, it would be useful to collect as much patients with EOADPKD as possible and make their genetic analysis in order to check whether they are caused by the same or similar mutations.

P-250 MUTATION SPECTRUM IN RUSSIAN CHILDREN WITH ATYPICAL HEMOLYTIC UREMIC SYNDROME

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Introduction: Atypical hemolytic uremic (aHUS) is an ultra-rare mostly genetic complement system disorder leading to thrombotic microangiopathy (TMA) and acute renal injury. It may have a relapsing course leading to end stage renal disease. A variable spectrum of mutations reported in Europe and North America. Treatment with Eculizumab (anti C5 antibody) is promising but needs further studies. This study is the first to investigate molecular genetics of aHUS in Russia.
Material and methods: Seventy one children aged 0.5–17 years were included. Diagnosis of aHUS was established by carefull exclusion of other TMA forms. Medical records and blood samples were collected during 3 years term. We investigated *CFH, CFI, CFB, MCP* and *THBD* genes mutations by new generation sequencing and *CFHR1/CFHR3* deletions by MPLA technique. The case reports of children with aHUS were analyzed to find a possible relation of clinical and laboratory data during the onset and follow up to genetic data and Eculizumab efficacy.
Results: Mutations were found in 46.5% of children and 7% had polymorphisms that may be assotiated to the disease. The numbers of mutations was: *CFHR1/CFHR3* - in 12 children (16.9%), *CFH* - 9 (12.7%), *CFB* - 7 (11.3%) и *MCP* - 3 (8.5%), *CFI* - 1 (1.4%) and *THBD* - 1 (1.4%). We did not find any significant clinical difference at onset time between children with or without mutations. At follow up mutation carriers relapsed more often (57.6% vs 31.6%). ESRD within one year after the onset was reached in 41%. Treatment with Eculizumab was performed in 20 children with mutations and 19 without with equal efficacy. Only in 4 children without mutations it was discontinued without recurrence ot TMA, 3 children with mutations relapsed.
Conclusions: The prevalence of gene mutations in Russian children with aHUS is compatible with worldwide data. The genetic data may help to predict a disease course and duration of Eculizumab treatment. Supported with Russian Science Fund grant 14–15-00994.

P-251 GASTROINTESTINAL INVOLVEMENT IN ATYPICAL HEMOLYTIC UREMIC SYNDROME

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Introduction: Atypical hemolytic uremic syndrome (aHUS) is a thrombotic microangiopathy with systemic future and it has a poor prognosis. The extrarenal involvements in aHUS are being increasingly characterized in the literature and occur up to half of all patients. In this study we aimed to investigate the clinical and genetic features and treatment of pediatric aHUS patients with gastrointestinal involvement.
Material and methods: Clinical features of aHUS patients were obtained from the Turkish national aHUS registry.
Results: This study included 170 children from the national aHUS Registry. The number of the patients with extraranal manifestations are 68 (23 male and 45 female). The most common type of extra-renal involvement of aHUS was neurological system involvement (n = 45 [66.2%]), followed by gastrointestinal (n = 19 [27.9%]), cardiovascular (n = 10 [14.7%]), and respiratory (n = 10 [14.7%]) involvement.Mean age was 7.9 years (2.5–14.7) in patients with gastrointestinal manifestations. Among the gastrointestinal manifestations, the most common clinical features were gastrointestinal hemorrahage in 4 patients, pancreatitis in 3 patients, non- bloody diarrhoea at presentation in 3 patients, epigastric pain in 3 patients, elevated liver enzyes in 2 patients, persistent vomiting in 2 patients, invagination in 1 patient, ischemic hepatitis in 1 patient and cholelithiasis in 1 patients. Of these 19 patients, 13 patients had other system involvement including neurological, cardiovascular and respiratory systems. Five patients with multisystem involvement died during the fallow-up. The genetic analyses revealed Factor H mutation in two patients and C3 mutation in one patient. The complement mutation analysis is under investigation in other patients.The patients were treated with eculizumab, with or without plasmapheresis/plasma infusion.
Conclusions: In this study we demonstrated a rare manifestation of pediatric aHUS patients with gastrointestinal involvement. The results of this registry can give important clues about the rare manifestations of the disease and this can improve the diagnosis, management and prognosis of aHUS patients.

P-252 RENAL INVOLVEMENT IN GLYCOGEN STORAGE DISEASE TYPE I: A CROSS SECTIONAL STUDY

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Introduction: Glycogen storage disease type I (GSDI) is a rare autosomal recessive disease characterized by accumulation of glycogen and fat

in liver and kidneys leading to chronic kidney disease. Glomerular hyperfiltration is the first stage of renal dysfunction but tubular abnormalities due to lipid and glycogen accumulation have also been recently described with the apparition of cysts development. This study describes renal dysfunction in GSDI.
Material and methods: We studied retrospectively 21 patients with GSDI (median age 22 years [11–62]) followed between 1976 and 2016. Renal function was measured with inulin clearance and magnetic resonance imaging (MRI) was performed in all patients.
Results: 85% of patients had a dietary treatment but only 45% had optimal metabolic control according to the ESGSD I criteria's. Median GFR was 151 ml/min/1.73m^2 (range 97–209), 75% had glomerular hyperfiltration. No patient had chronic kidney disease but 30% had microalbuminuria. Thirty-five percent had metabolic acidosis; 36% had hypercalciuria, none of them had renal calculi. We found 31% of patients with nephromegaly, 15% with renal cysts. MRI can not detect overt glycogen/lipid deposition. No correlation was found between the GFR, albuminuria and metabolic control.
Conclusions: Hyperfiltration and tubular dysfunction was present as previously reported whereas nephromegaly and renal cysts was found in a minority of patients. Renal disease remains a major complication of GSDI. GSDI patient could benefit of a regular tubular evaluation as well as kidney morphological assessment by echography and/or adequate MRI sequencing.

P-253 CLINICAL AND PROGNOSTIC FEATURES OF AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY WITH A CHILDHOOD DEBUT

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Introduction: Autosomal dominant polycystic kidney disease (ADPKD) is the most common hereditary renal disease. The description of the molecular basis of ADPKD in the last decade has led to the development of promising therapies in adults. Early diagnosis in pediatric patients allows for optimal anticipate therapeutics that could alter the natural history of disease. We review the morbidity of pediatrics patients with ADPKD and we try to identify predictors of renal deterioration.
Material and methods: A retrospective descriptive study of patients diagnosed with ADPKD in the pediatric age, followed-up in a pediatric nephrology service between 1993 and 2016. Statistical analyses were performed to evaluate long-term renal dysfunction.
Results: A total of 25 patients (15 W/10 M) were assessed with a median age of 5.4 years at diagnosis (<1 months-13 years). Only one patient was prenatally diagnosed. All but one had family history of ADPKD, in that case, a mutation in PKD1 gene was found. The patients were studied from 1.4 to 20.9 years after diagnosis and all of them maintained normal glomerular filtration rate. Hypertension was presented in 16% and proteinuria was found in 8%. At ultrasound the 78.3% of kidneys had normal volume, while 21.7% had nephromegaly. As complications, 12% had suffered urinary infection, 8% urolithiasis and 8% episodes of abdominal pain. None had hepatic cysts. There was a significant relationship between nephromegaly and hypertension. (P = 0.021; Fisher exact test). Significant correlation was also detected between diagnosis at age 5 or older and renal damage (P = 0.037; Fisher exact test). No correlation was found between proteinuria, hypertension, urinary infection, urolithiasis and episode of abdominal pain and age.
Conclusions: Children with ADPKD maintain glomerular filtration rate during infancy. The prevalence of urinary infections, urolithiasis, proteinuria and hypertension in children with ADPKD is greater than in the general population. Ultrasound nephromegaly is associated with hypertension.

P-254 DIAGNOSIS OF ALPORT SYNDROME AND THIN BASEMENT MEMBRANE NEPHROPATHY IN CHILDHOOD

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Introduction: Alport syndrome (AS) and thin basement membrane nephropathy (TBMN) are difficult to differenciate glomerulopathies form genetical origin, with no predictors of long term outcome.
The aim of the study was to analyze clinical data regarding kidney histopathology in children with AS and TBMN.
Material and methods: The study group consisted of 53 children (27 girls) aged 9.5 yeras (median) treated for AS (25 patients) or TBMN (28 patients) in Department of Pediatric Nephrology, University Childrens' Hospital of Cracow between years 1988–2015. Kidney biopsy result was corretated to clinical and laboratory data in both groups using Statistica12(Statsoft).
Results: Hematuria was diagnosed in all children at age of 1–17 (medin 6 years), proteinuria coexisted in 14 patients with AS (56%). In 4 boys with AS hypoacusis was found. Follow up time before kidney biopsy was significant longer in TBMN 38 vs.11 months in AS (p = 0.01). In 11 (39%) of TBMN patients mesangial proliferation was observed. Among AS patients in 4 cases (16%) no kidney pathology was found. Multiple regression analysis pointed irregular basement membrane structure as predictor for proteinuria (R^2 = 0,56,p = 0,011). Therapie with ACEI was introdused in 14 patients with proteinuria, prednisone was used in 8 patients, ciclosporine A in 7, methylprednisolone pulses in 2. Two boys progessed to end stage renal failure befrore age 18 years. Two patients with TBMN developed proteinuria in later follow-up.
Conclusions: Proteinuria and irregularity of basement membrane in children with AS and TBMN are predictors of poor prognosis. Hematuria and thinning of basal membrane do not ensure benign follow-up. Early kidney biopsy in young children with isolated hematuria seems to be not useful. Intorduction of genetical testing is needed for early diagnosig instead.

P-255 COMPLEMENT-AHUS/PTT IN ARGENTINA, SIX YEARS EXPERIENCE OF GENETIC INTERNATIONAL LABORATORY

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Introduction: Describe the results found in 6 years of genetic studies of Argentine patients with aHUS/PTT clinic by an international laboratory (SECUGEN S.L., Spain).
Material and methods: Analysis of aHUS in 30 patients by Sanger and next generation sequencing (NGS) technology in SECUGEN S.L.-España from January 2011 until April 2017. Sanger technology: DNA sequencing: Exons and promoter region of *CFH, MCP, CFI, CFB, C3* and *THBD* genes. NGS technology: Amplification of the region of interest with Ampliseq technologies and sequencing by Ion Torrent and Nextera capture of the region of interest with Illumina (MySeq). The genes studied by NGS are *CFH, CFI, MCP (CD46), CFB, C3, THBD, DGKE, CFHR1, CFHR2, CFHR3, CFHR4, CFHR5, CFP* and *ADAMTS13*. Additionally, the

promoter region of *CFH* and promoter region and 3'UTR of *MCP (CD46)* genes were included. Analysis of sequencing and description of changes compared to the DNA reference in databases.

Results: Of the 29 patients studied, 15 (51.72%) were male and 14 (46.42%) women. 10 (34.48%) of them were children and 19 (65.51%) adults. In 18 patients (62%), 10 adults and 8 children, a total of 23 mutations were found: related or possibly related to the disease in 11, and in the other 12 were variants of uncertain significance (VUS), benign alterations or non pathogenic mutation. The mutations are described in Table 1.

Gen	Mutation
ADAMTS13	c.3074C > T; p.Ser1025Leu
ADAMTS13	c.3541G > A; p.Gly1181Arg
C3	c.193G > T; p.Lys65Gln
C3	c.4851-1G > A
C3	c.485C > A; p.Thr162Lys
CD46	Del_ex1-ex12
CD46	c.963delC, p.Tyr321*
CD46	c.535G > C; p.Glu179Gln
CD46	c.38C > T; p.Ser13Phe
CFB	c.1-12C > A
CFB	c.1051 T > C; p.Ser351Pro
CFH	c.1646G > C, p.Gly549Ala
CFH	c.3628C > T; p.Arg1210Cys
CFH	c.3572C > T; p.Ser1191Leu
CFH	c.3572C > T; p.Ser1191Leu
CFH	c.3590 T > C; p.Val1197Ala
CFH	c.2258G > T; p.Cys753Phe
CFH	p.Ser1191Leu
CFH	p.Val1197Ala
CFHR2	c.212C > T; p.Thr71Met
CFHR5	C.2542_266dupAGGAATGTGTTCCTT; p.Ser88_PheinsLeuGlyMetCysSer
CFI	c.502A > G; p.Arg168Gly
CFI	c.1071 T > G;p.Ile357Met

Conclusions: We found mutations or genetic variants in 62% of the cases studied. We found mutations or genetic variants in the following genes: *CFH, MCP (CD46), C3, ADAMTS13, CFI, CFB, CFHR5* and *CFHR2*. Mutations in *CFH* and *MCP (CD46)* occurred more frequently representing 52.17% of cases. In 3 patients combined forms were found: *MCP (CD46) + CFH* in 2 cases and other *ADAMST13 + CFI*. CD46 mutations were found only in children.

P-256 AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE (ARPKD) IN THE UK NATIONAL REGISTRY OF RARE KIDNEY DISEASES (RADAR)

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Introduction: Autosomal recessive polycystic kidney disease (ARPKD) is a rare genetic condition that causes cysts to develop in the liver and kidneys. This abstract describes data collected by the ARPKD Rare Disease Group via the National Registry of Rare Kidney Diseases (RaDaR) in the UK. ARPKD rare disease group uses RaDaR to provide patients information and to build a communication network for those affected by the condition. The group supports patients by offering family information days where latest advancements in treatment are discussed.

Material and methods: Data is entered retrospectively from the patient's medical records following consent. The dataset is defined by the ARPKD Rare Disease Group. Data fields include demographics, blood and urine results, medications, transplant and dialysis history, genetics and co-morbidities. The inclusion criteria are open to all ARPKD patients including Congenital Hepatic Fibrosis and Caroli Syndrome with renal involvement.

Results: 108 ARPKD patients from 31 UK renal units have been consented to date with an age range of 1 week to 65 years. There are 54 (50%) paediatric (under 16) patients with an average age of 8 years and 54 (50%) adult patients with an average age of 39 years. There are 50 females (46%) and 58 males (54%) males. The first paediatric patient was recruited in October 2012 and the first adult patient in August 2013.

Conclusions: RaDaR holds the largest single cohort of ARPKD patients collected to date in the UK. It provides an important epidemiology data on ARPKD including the progression of the condition. The registry provides a ready cohort for research into this condition: from observational to interventional studies.

P-257 IDIOPATHIC INFANTILE HYPERCALCEMIA DIAGNOSED IN ADOLESCENCE

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Introduction: Idiopathic infantile hypercalcemia (IIH) usually manifests in infancy with severe clinical features- failure to thrive, poor appetite, vomiting, dehydration and seizures. The majority of cases are due to mutations in *CYP24A1* gene which encodes 24 hydroxylase, an enzyme involved in degradation pathway of the vitamin D metabolism. Herein we present an adolescent girl with incidental finding of bilateral nephrocalcinosis who was found to carry typical Central European CYP24A1 mutation (E143del).

Material and methods: Standard clinical and biochemical evaluation, ultrasound examination of the kidneys amd molecular genetic study of nephrolithiasis/nephrocalcinosis genes.

Results: This 12 year old female came to our attention due to incidental finding of bilateral medullary nephrocalcinosis. The physical examination including growth parameters was unremarkable. Laboratory examination revealed mild hypomagnesemia (0.5–0.6 mmol/l), normocalcemic hypercalciuria, suppressed PTH 6.9 pg/ml (normal 10–65) and incomplete distal renal tubular acidosis. Mutational analysis of the *CLDN16, CLDN19, AE1, ATP6V1B1* and *ATP6V0A4* genes was negative. At the age of 16 with targeted next generation sequencing homozygous mutation in *CYP24A1* gene (E143del) was detected establishing the diagnosis of idiopathic infantile hypercalcemia.

Conclusions: Our patient had unusual clinical phenotype without hypercalcemic crisis in infancy and laboratory findings suggesting familial hypomagnesemia with hypecalciuria and nephrocalcinosis. The correct diagnosis was established owing to next generation sequencing of the panel of the nephrolithiasis/nephrocalcinosis genes. The genetic diagnosis had important implication for further management of this girl (avoiding vitamin D supplements and sunlight exposure).

P-258 DETECTION OF GENE KAL1 MUTATIONS IN CHILDREN WITH UNILATERAL KIDNEY AGENESIS

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Introduction: The purpose of thiw study is gene testing to detect mutations responsible for X-linked form of Kallmann Syndrome (KS), gene KAL1, in children with unilateral renal agenesis (URA) who have no family history of URA, to determine the frequency of gene mutations KAL1 in URA.
Material and methods: The study included 42 children up to 15 years (groupA:12 children, groupB: 30 children) with confirmed congenital URA. Medical history, family tree and physical examination, focusing on the detection of possible KS indicators to patients and their families. Follow the molecular control of gene KAL1 by PCR and cycle sequencing. Seventy-five percent of patients were male and 25% female. In 58% the diagnosis was prenatal URA, while 42% of them stated cystic formations which disappeared after birth.
Results: Of the 15 patients who completed to date genetic testing, we found KAL1 mutation in a patient 12 years old, who showed anosmia and insilateral cryptorchidism.
Conclusions: Thre finding mutations KAL1 gene in children with URA may reaffirms anosmins-1 (the KAL1 gene product engagement) in organogenesis of the kidney and it allows early diagnosis of KS. The etiologic diagnosis of URA should be offered to patients who can be benefited from early diagnosis of KS and treatment.

P-259 OVERVIEW OF RARE RENAL DISEASES AT A PAEDIATRIC RENAL CENTRE THROUGH THE NATIONAL REGISTRY OF RARE KIDNEY DISEASES (RADAR) IN THE UNITED KINGDOM

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Introduction: Rare kidney diseases need better understanding through collection of better clinical data. Therefore our centre participates in RaDaR, a UK Renal Association initiative designed to gather information. RaDaR recruitment began in 2010 and covers more than 40 conditions. There are around 10,000 recruits from 78 renal adult & paediatric units in the UK. Our Paediatric Renal Centre is the leading recruiting hospital in the UK.
Material and methods: The RaDaR dataset is defined by the UK Renal Registry in association with the Rare Disease Groups, made up of experts in each eligible condition. Data fields include demographics, blood and urine results, medications, transplant and dialysis history, genetics and comorbidities. Data is entered retrospectively from the patient's medical records following consent.
Results: 319 patients have been consented at BCH. The age range is from birth to 16 years with mean of 4.9 years with male to female ratio of 55%:45%. The most common condition is Idiopathic Nephrotic Syndrome (n = 128; 39%), followed by Alport Syndrome (n = 33; 10%), ARPKD (n = 25; 8%), Hyperoxaluria (n = 24; 7%) and STEC HUS (n = 22; 7%). The other conditions with numbers of patients recruited so far include: ADPKD (n = 14), aHUS (n = 13); Cystinosis (n = 9); Cystinuria (n = 3); Dent & Lowe (n = 7); HNF1b (n = 6); Hypokalaemic Alkalosis (n = 8); MPGN (n = 14) and Vasculitis (n = 7).
Conclusions: RaDaR provides important epidemiology data based on the UK population which is shared amongst the renal team to develop further research into rare kidney diseases and improve the quality of care. It also gives an opportunity to define the best treatment practices across the country.

P-260 FIRST RESULTS OF SYSTEMATIC GENETIC TESTING FOR ALPORT SYNDROME AND THIN GLOMERULAR BASEMENT MEMBRANE NEPHROPATHY IN CROATIA

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Introduction: The aim is to present the initial results of first systematic genetic analysis of patients with Alport syndrome (AS) and thin glomerular basement membrane nephropathy (TBMN) in Croatia as a part of a project titled "Genotype–phenotype correlation in Alport syndrome (AS) and thin glomerular basement membrane nephropathy (TBMN)" funded by Croatian Science Foundation. Our project is multidisciplinary, nationwide, collaborative research in which seven leading Croatian nephrology, paediatric nephrology and nephropathology institutions participate.
Material and methods: We have identified 255 AS and TBMN patients (age range 2–85) by search of patients' records and data registries. From all identified patients, we have collected clinical data regarding family status, gender and age, kidney function (haematuria, proteinuria, chronic renal failure or ESRD), hearing loss and ocular lesions. In all patients kidney biopsy was performed and histological diagnosis of AS or TBMN was made. Genetic counselling for first 11 selected patients (pilot study) was performed in three Croatian hospitals where patients were selected for mutation screening in the COL4A3, COL4A4 and COL4A5 genes by next generation sequencing (NGS).
Results: First selected 11 patients (4 males, 7 females, age 3–47) belonged to 7 unrelated families. Four families had a mutation in the COL4A5 gene, one in COL4A3 gene and one had digenic COL4A3 and COL4A4 mutations. In one patient mutation was not found. We found six new mutations and one previously reported mutation. Affected individuals had a wide range of phenotypes from a non-progressive isolated microhematuria to ESRD in the fourth or fifth decade of life.
Conclusions: Results of our pilot study show significance of systematic analysis of AS and TBMN patients in Croatia. We found 6 new mutations in 7 tested families. Our ultimate goal is to investigate Croatian AS and TBMN patients from clinical, histological and genetic aspect with creation of nationwide patient registry.

P-261 NEPHROPATHIC CYSTINOSIS- THE FIRST COMPREHENSIVE REPORT FROM POLAND.

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Introduction: Nephropathic cystinosis (NC) is a rare, inherited storage disease caused by impaired lysosomal transport of cystine. Until recently the Polish NC patients were not totally registered, they had poor access to

specific diagnostics and causative therapy. Therefore the aim of the study was to evaluate the incidence and medical status of all known living patients with NC in Poland.
Material and methods: Retrospective analysis based on medical records.
Results: The study comprised of 13 patients aged 6.1–36 (median 26) years. In 12 and 1 of them infantile and juvenile form of NC were diagnosed, respectively. The median age at diagnosis and the median follow-up were 2 and 25 years, respectively. The final diagnosis was made mostly on the basis of clinical picture. Recently all but 2 patients underwent molecular testing which showed different mutations in the *CTNS* gene, but surprisingly a deletion of ~57 kb was found only in 2 (18%) cases. Nine patients developed ESRF at the median age of 10 years followed by kidney transplantation (KTx). Six of them have still functioning first graft- median time 15.5 years and the remaining 3 were re-transplanted. Although most of them were treated with oral cysteamine and symptomatically, the specific therapy was delayed, limited in time and dose and/or not correctly monitored. All of them have characteristic NC extrarenal complications. In contrast, the 2 youngest patients at the age of 6.1 and 8.6 years who are treated according to recommendations show normal growth and renal function. Unfortunately NC patients have still limited access to cysteamine eye drops and suffer from ocular problems.
Conclusions: The incidence of NC in Poland seems to be much lower than in Western European countries. Due to recent regulations in health care system, the situation of patients with NC has been improved. The early start of specific and symptomatic treatment of NC are crucial for the outcome.

P-262 PRIMARY COENZYME Q10 DEFICIENCY-6 (COQ10D6): CASE REPORT

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Introduction: Steroid resistant nephrotic syndrome (SRNS) is an important health problem that causes end stage renal disease (ESRD). Genetic mutation can be detected in nearly 25% of the patients with SRNS. Primary coenzyme Q10 deficiency-6 (COQ10D6) is an autosomal recessively inherited disorder due to *COQ6* mutation. The main clinical manifestations are infantile progressive nephrotic syndrome leading to ESRD and sensorineural deafness.
Material and methods: Herein, we report a girl diagnosed SRNS and sensorineural deafness with a COQ6 mutation.
Results: A 9 year old girl was admitted to our hospital for genetic evaluation due to SRNS. She was diagnosed with SRNS at the age of seven and audiologic work-up revealed bilateral sensorineural deafness. Renal biopsy specimen demonstrated focal segmental glomerulosclerosis. Cyclosporine was administered after the renal biopsy. There was no response to cyclosporine therapy. Cyclosporine was discontinued and mycophenolate mofetil was added to the therapy. Despite immunosuppressive therapy, serum creatinine was increased and hemodialysis was indicated one year after disease onset. Living-donor kidney transplantation was performed at the eighth month of hemodialysis. Genomic DNA material, isolated from peripheral blood sample, analyzed by Next Generation Sequencing technology by Ion Torrent, with in-house designed panel-gene kit, embracing 30 nephrotic syndrome genes, covering all the coding regions, exon-intron boundaries and un translated regions (UTR), incorporating 272.26 kb region in 995 amplicons with coverage of 98.59%. Bioinformatic analysis revealed homozygous c.1058C > A (p.A353D, rs 397,514,479) mutation in exon nine of *COQ6* gene, which was further confirmed by Sanger sequencing.
Conclusions: *COQ6* mutations should be consider in patients with SRNS and sensorineural hearing loss and COQ10 replacement therapy should be initiated, as primer COQ10 deficiency is considered to be the only known treatable mitochondrial disease.

P-263 PATIENT WITH LATE-ONSET OF RENAL FAILURE, HYPERCALCIURIA,NEPHROCALCINOSIS AND OCULAR SYMPTOMS: A CASE REPORT

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Introduction: Familial hypomagnesaemia with hypercalciuria and nephrocalcinosis (FHHNC) is a rare autosomal recessively inherited disease characterized by excessive wasting of renal tubular magnesium and calcium, bilateral nephrocalcinosis and progressive renal failure. FHHNC is associated with mutations in CLDN16 and CLDN19.
Material and methods: We describe a case of a well grown 11 year old boy with night blindness since childhood, born to consanguineous parents from rural South India. He has an 18 year old sister with night blindness who is otherwise asymptomatic. He presented with a 3 week history of anasarca and was noted to have CKD stage-V with signs of severe acidosis, hypertension and fluid overload. At a local hospital, investigations revealed bilateral obstructive renal calculi for which bilateral DJ stents were inserted and he was given 3 sessions of haemodialysis. On admission to our unit, he had the following laboratory findings:Creatinine-11.3 mg/dl, BUN-77 mg/dl, PTH – 207.6 pg/ml and Bicarbonate – 9.1 mmol/lt. Due to renal failure, he had hypocalcaemia-8.3 mg/dl and hypermagnesaemia- 3.1 mg/dl. A 24 h urine oxalate/creatinine ratio was normal; however he had hypercalciuria (0.23 mg/mg). Renal ultrasound showed bilateral slightly small sized kidneys, Grade III renal parenchymal changes, medullary nephrocalcinosis and non-obstructive calculi. ECHO was normal. Ophthalmological examination done in view of night blindness revealed features of retinitis pigmentosa. Blood tests for the mutations in the CLDN16 and CLDN19 genes have been sent and results are awaited.
Results: In addition to his renal condition, he had a challenging behavior of being withdrawn and being non-compliant with medications. Involvement of child psychiatry team led to better compliance and he has been successfully established on a thrice weekly haemodialysis program, with a view to undergo a renal transplant in the near future.
Conclusions: FHHNC should be suspected in patients who present with hypercalciuria, nephrocalcinosis and renal failure along with ocular abnormalities. Addressing psychosocial factors is important to achieve better outcomes.

P-264 NEUROLOGIC INVOLVEMENT IN SCHIMKE DISEASE

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Introduction: Neurological symptoms may be related to abnormalities in the structure or function of cortical neurons resulting from SMARCAL1 gene expression disorders (ischemic lesions, edema?).
Material and methods: Authors describe the case of currently 11-year old girl with Schimke immuno-osseous dysplasia with confirmed

SMARCAL 1 gene mutation. Nephrotic range proteinuria with FSGS has been detected at the age of 4.5 y. Immunosuppression was ineffective and she started peritoneal dialysis at age of 8 y. Apart from kidney disease with severe hypertension, hypothyroidism, hypostature with osseous deformations, recurrent pericarditis and immunodeficiency the girl presents also recurrent neurological symptoms.
Results: In February 2017 she rapidly deteriorated. In a few hours the child became sleepy, without logical contact, anxious, with pointless movements, and progressively confused. Neurological examination confirmed stiffness of the neck, lack of verbal communication. Right-side pyramid syndrome (Lovett III) was identified. Taking into account the clinical course and data on neurological symptoms in the Schimke syndrome available in the literature, the association of current neurological disorders with direct CNS infection was less likely. In the medical history, the girl had previous headache (beginning before 2013), diplopia episode with symptoms of dysarthria (2014, cerebrospinal fluid test - normal result), symptoms of right-sided paresis (2016). The onset of current neurological symptoms was associated with dehydration and electrolyte abnormalities (hyponatremia Na:126 mmol/l). On MRI examination smoothing of cerebral gyri of the left brain hemisphere with thickening of the cortex of the frontal, parietal and temporal lobes and right frontal area, narrowing of the subarachnoid space were found. The EEG record revealed numerous generalized, left side localized delta waves. The consultant neurologist recommended the introduction of valproic acid.
Conclusions: Observed neurological disorders including partial seizure episodes are probably of paroxysmal nature and are possibly associated with previous disturbances of consciousness and focal deficits. The direct cause of recurrent abnormalities is still unknown.

P-265 ATYPICAL HYPOTONIA-CYSTINURIA A NEW CASE: GENOTYPE-PHENOTYPE AND DESCRIPTION

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Introduction: "Hypotonia-cystinuria syndrome" (HCS) and "**2p21 deletion syndrome"** are **two** recessive contiguous gene deletion syndromes associated with cystinuria. The deletions differ in size and number of genes involved. **HCS** is characterized by hypotonia, failure to thrive, severe growth retardation, growth hormone deficiency, characteristic facial dysmorphy and cystinuria. **2p21 deletion syndrome** presents with HCS features in addition to mental retardation and respiratory chain complex deficiency. In **HCS**, *SLC3A1* and *PREPL* genes are disrupted, while in **2p21 deletion syndrome**, two additional genes (C2orf34 and PM1B) are deleted. Mutations in *SLC3A1* cause cystinuria. The extended phenotypes are attributed to *PREPL* and C2orf34 PM1B deletions. HCS is described in 17 families, 2p21 syndrome only in one Bedouin family. An intermediate phenotype, resulting from deletion of *SLC3A1, PREPL* and *C2orf34* has been reported twice and known as **"atypical HCS".** We describe a new case of **"atypical HCS"** and his genotype-phenotype correlation.
Material and methods: We report the case of a 4-years-old girl from Libya with consanguineous parents. She presented generalized hypotonia and failure to thrive and developed severe growth and developmental delay. She showed cranial dysmorphy with pale scale, long face, bitemporal narrowing, frontal bossing, turned up nose, tented upper lip and slight ptosis. Brain MRI was normal, renal ultrasound revealed nephrocalcinosis and corraliform lithiasis. Urine amino acid chromatography confirmed cystinuria. Blood analysis showed low IGF-1 and elevated lactates. Because of this suggestive phenotype we explore a genomic imbalance by array-cgh in chromosome 2p21.
Results: Array-CGH (180,000-oligonucleotide microarray) evidenced **homozygote** microdeletion of 250 kb in chromosome 2p21 (chr2:44,507,915-44757213pb) including partially genes *SLC3A1* (exons 3–6) and *KAMCAT* (exons 1–2) entirely *PREPL*. Each parents showed the same deletion in the heterozygous state.
Conclusions: In contiguous deletion syndromes, characterization of the different deletions, is crucial to precise genotype-phenotype correlation and understand the role of each gene.

P-266 FABRY DISEASE: A RARE CAUSE OF PROTEINURIA IN A MALE ADOLESCENT

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Introduction: Fabry disease is a lysosomal storage disease caused by the mutations of the enzyme α-galactosidase A and is inherited as X-linked trait. The patients develop multiple organ dysfunctions because of the deposition of glycosphyngolipids, mostly globotriaosylceramide, at several tissues.
Material and methods: Here, we report a case presenting with proteinuria accompanied with hearing loss, who was diagnosed as Fabry disease after the electron microscopic findings of a renal biopsy specimen.
Results: An 18 year old male was referred with proteinuria found during routine examination. He denied edema or macroscopic hematuria. His past medical history revealed that he had bilateral sensorineural hearing loss for two years. Family history revealed that his parents had had consanguineous marriage and his grandmother had died of chronic renal failure at her 50's. Blood pressure and systemic examination was normal except a systolic murmur at mesocardiac region. On urinalysis, 3+ proteinuria was detected, no erythrocytes were seen. Daily urine protein excretion was 1245 mg/d; serum albumin was 4.3 g/dL. An echocardiography yielded 2. degree mitral regurgitation. Fundoscopic exam was normal. A renal biopsy was performed, showing proliferation of visceral epithelial cells with foam cells on light microscopy; immunofluorescent examination was normal. However, on electron microscopy, lamellar lipid inclusion bodies in podocyte cytoplasm, so-called zebra bodies, were seen, which is characteristic for Fabry disease. Genetic analysis confirmed the diagnosis, showing a missense mutation (p.R342Q) in GLA gene. Also, blood Lyso-GL-3 (globotriaosylsphingosine) level was high (81.8 ng/ml, >3.5). Later on, an enzyme replacement therapy was initiated.
Conclusions: We conclude that all males presenting with proteinuria with unknown etiology should be evaluated for Fabry disease.

P-267 NOVEL MUTATION IN THE NUP160 GENE IN THREE SAUDI PATIENTS WITH FOCAL SEGMENTAL GLOMERULOSCLEROSIS: A CASE SERIES

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Introduction: Focal Segmental Glomerulosclerosis (FSGS) is a genetically heterogeneous disorder with variable clinical presentation and course. Mutation in the NUP160 gene has not been described in early onset steroid-resistant nephrotic syndrome and FSGS so far. We described the clinical courses of three Saudi patients with early onset FSGS with novel mutation in this gene.
Material and methods: We described three siblings of consangious parents.The first patient was eight years old girl who presented with

hypertension and proteinuria without oedema and found to have urea 34 mmol/l and creatinine 412 micromol/l, her kidney biopsy showed sclerosed glomeruli and severe interstial fibrosis. She required peritoneal dialysis within three months and then she received a living-related kidney transplantation. Her brother was nine years old who had a similar presentation with hypertension and proteinuria, found to have urea 36 mmol/l and creatinine 1064 micromol/l and his kidney biopsy showed severe glomerulosclerosis and interstial fibrosis; he was maintained on chronic hemodialysis. The other brother is six years old and was found to have a 2+ protein in urine dipstick screening and urine protein/creatinine of 119 mg/mmol with normal blood pressure, renal profile and serum Albumin. We did a kidney biopsy for him and it showed FSGS. We sent genetic testings for them.
Results: Whole exome sequencing revealed the homozygous splice site mutation c.1179 + 5G > A in intron 8 of the NUP160 gene, which was validated by Sanger sequencing. Both parents were heterozygous carriers. NUP160 is part of the NUP107–160 subcomplex of the nuclear pore complex. Mutations in NUP107 and other nucleoporins, but not NUP160, have been described in FSGS. To our knowledge, this aberration has never been described in the literature.It seems likely this homozygous mutation explained the clinical phenotype of the three siblings.
Conclusions: Our work expands the mutational spectrum of early onset FSGS.

P-268 GENETICS IS RESPONSIBLE FOR ALL – HYPOSPADIAS

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Introduction: Hypospadias represents a rather frequently observed congenital disorder of the male external genitalia. It is an inborn developmental defect of the urethra, which may be associated with other congenital developmental disorders. Hypospadias are divided according to meatal location to anterior, middle, posterior, and hypospadias with chordee. The condition has a prevalence of 4–43/10.000 born children, and is most frequently observed in white population, less often in black population, and rarely in Asians and Hispanics.Many cases are of unknown etiology; the condition is most often caused by interaction of genes and multifactorial causes.
Material and methods: Within the common genetic practice, we analyse karyotype in boys with hypospadias, no other gene panel associated with hypospadias is assessed routinely in boys with hypospadias in the Czech Republic. Nevertheless, other genes associated with hypospadias have been analysed in clinical trials performed abroad (WT I, SF I, BMP4, HOXA4 and others).Another proof of gene involvement may be deduced from known syndromes usually associated with hypospadias (WTI gene mutations are associated with Denys-Drash and Frasier syndromes), further also ZEB2- associated with Mowat-Wilson Syndrome, with more than 50% of boys suffering from hypospadias.
Results: We have examined a boy with hypospadias at our nephrology clinic. Assessment of family history showed the presence of hypospadias in male members of the family – father's siblings, the boy's father, and also mother's brother suffered from hypospadias.Seven per cent of hypospadias cases are familial; the risk of hypospadias development in brothers is 9–17%. Due to the occurrence of hypospadias in the family, genetic assessment of the whole family has been performed, with a family tree.
Conclusions: We have concluded that the hypospadias in our male paediatric patient is genetically conditioned, with autozomal dominant inheritance pattern.

P-269 ADPKD MUTATION IN A SMALL ISLAND POPULATION

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Introduction: Autosomal dominant polycystic kidney disease (ADPKD) is caused by mutations in *PKD1* or *PKD2*. *PKD1* mutations account for 85% of cases and cause more severe disease with earlier onset of end-stage. ADPKD accounts for 10% of end-stage renal disease in Maltese adults who reach this stage generally between 40 and 50 years The mutation in this population has not yet been described.
Material and methods: A young child referred with renal cysts on ultrasound and a strong family history of ADPKD was identified, and blood and saliva samples were obtained from the family with written informed consent. High throughput sequencing (HTS) was used to explore the entire *PKD1* and *PKD2* coding regions extending up to 50 bp into the introns in each direction. SureSelect^{XT} Target Enrichment capture of 2.6 Mb of the genome, including *PKD1* and *PKD2* was followed by sequencing on Illumina HiSeq4000. HTS data was mapped to GRCh37 as paired-end libraries using NextGENe software. A BED file was used to ensure mapping to *PKD1* and *PKD2* excluding their pseudogenes. To remove potential pseudogene contamination of the data, the mutation list was filtered against an in-house database of 90 control HTS datasets and by pairwise blast of *PKD1* and relevant pseudogenes.
Results: A heterozygous novel mutation in exon 15 of *PKD1*, c. 4305C > G, p.Y1435X, was identified in the proband and the affected child but in none of the control datasets. The novel stop codon is in the 6th consecutive extracellular PKD domain resulting in a truncated protein lacking a number of domains, including all the transmembrane domains.
Conclusions: We report a novel, pathogenic, nonsense mutation in *PKD1* in a Maltese family.

P-270 A VARIANT MUTATION ON THE PKHD1 GENE IN A PORTUGUESE CHILD

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Introduction: Autosomal recessive polycystic kidney disease (ARPKD) represents an important cause of paediatric end stage kidney disease. To date, polycystic kidney and hepatic disease 1 (PKHD1) is the only identified causative gene for ARPKD, encoding the amino acid fibrocystin.
Material and methods: NA.
Results: We report the case of a female infant with a prenatal diagnosis of severe oligoamnios and mild urinary tract dilation (8 mm) at 28 weeks of gestation. She was born at 34 weeks by caesarean section and required non-invasive ventilation for 3 days because of pulmonary distress and cyanosis. A renal ultrasound (US) scan performed at the 3rd day of life revealed bilateral enlarged hyperechogenic kidneys and small cysts at the renal medulla. Serum creatinine levels, glomerular filtration rate and liver panel were normal. The liver and spleen appeared normal in a posterior US. An abdominal US was performed on both parents and demonstrated no cystic change within the kidneys. At 9 months of age she developed hypertension and started enalapril 0.32 mg/kg/day. In the absence of liver disease or hepatic fibrosis, the molecular genetic testing of the PKHD1 gene was performed. In our patient, a previously reported pathogenic missense mutation, c.664 A > G, was identified. A second, previously reported mutation in one portuguese not related with our patient, c.2280-1G > A, was also identified. This mutation affects the splice site and is most likely pathogenic. Genetic testing for PKHD1 mutations on the parents is ongoing. At clinical follow-up, at 3 years of age, she maintains a severe hypertension controlled with amlodipine, lisinopril and propranolol.
Conclusions: Although the phenotypic presentation of ARPKD is variable, liver involvement is always present, leading to congenital hepatic fibrosis and dilatation of the intrahepatic ducts. In this particular case, the child never presented hepatic involvement, so the genetic study was important to confirm the diagnostic suspicion.

P-271 CLINICAL AND LABORATORY MARKERS VIOLATION FIBRYLOHENESIS IN INFLAMMATORY KIDNEY DISEASES IN CHILDREN FROM ECOLOGICALLY UNFAVORABLE REGIONS

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Introduction: to increase the effectiveness of treatment of children with pyelonephritis associated with undifferentiated connective tissue dysplasia in conditions of ecological trouble, and by improving personalization schemes preventive measures, developed on the basis of knowledge of molecular genetic and biochemical markers of the severity of its course. To determine the prevalence and clinical manifestations of undifferentiated connective tissue dysplasia (UCTD) in patients with pyelonephritis who live in conditions of environmental pollution. Determine the level of serum oxyproline content free oxyproline, creatinine and glycosaminoglycans hour urine as markers of severity at UCTD pyelonephritis in children.
Material and methods: The work was conducted at the first pediatric ward and ent circuit LvivReginal Clinical Children Hospital "OKHMATDYT" and during expeditions in ecologically unfavorable regions of Lviv in 2 stages: Stage I - the definition UCTD frequency in children with renal pathology; Stage II - children with pyelonephritis with polluted regions were divided into 2 groups study on 30 people: children with pyelonephritis and UCTD (30 persons) and pyelonephritis without UCTD (30 children), depending on the anthropometric indexes - body mass index and index Varga. In the control group included 30 healthy children. Children held oxyproline laboratory determination of levels of serum content free oxyproline, creatinine and glycosaminoglycans hour urine as markers of severity at UCTD pyelonephritis in children from ecologically disadvantaged regions.
Results: the prevalence of phenotypic traits UCTD such as posture, hiperroztyazhymist skin, hypermobility of joints, congenital anomalies development hipertelorizm eyes, emotional lability, cardialgia, sore joints and spine in patients with renal pathology in polluted regions of Lviv is 82.0% in children with UCTD level oxyproline excretion in the urine (+++) are directly proportional relationship to the level of the free fraction oxyproline serum (47.14 ± 29.03 mg / dL in children with UCTD; 40.08 ± 24, 09 mmol / l in children without UCTD 17.65 ± 21.15 and in healthy). Analysis of serum creatinine hour urine of children with contaminated salts of heavy metals and titanium parts showed its reduction in 80.0% of the surveyed children UCTD in no UCTD 25.0% and 15.0% of healthy patients, that almost every patient with UCTD he was slightly below the reference values of the patients, indicating the beginning of the process of glomerular sclerosis nephron in these children. In children who are products of anthropogenic load Cement and heavy metal salts hlikozoaminohlikaniv excretion was increased in 65.0% of children 6–8 years old (223.17 ± 11.64 Ed. TSPH / 1 g creatinine), 78.0% children 9–12 years (254.5 ± 23.15 Ed. TSPH / 1 g creatinine) and in 92.0% of children ages 13–17 (223.45 ± 19.20 Ed. TSPH / 1 g creatinine) children with UCTD when these values in children without NDST: in 61.0% of children 6–8 years old (221.67 ± 10.54 Ed. TSPH / 1 g creatinine), 75.0% of children ages 9–12 (260.9 ± 20.08 Ed. TSPH / 1 g creatinine) and 88.0% of children ages 13–17 (229.25 ± 18.40 Ed. TSPH / 1 g creatinine) and in healthy d dren increased excretion of glycosaminoglycans (82 ± 36.75 Ed. TSPH / 1 g creatinine) was diagnosed in only 3.0–5.0%, indicating a marked xenobiotics toxic damage of kidney tissue with involvement in the pathological process of glomerular apparatus which reduces functionality biotransformation of xenobiotics in the third phase, the phase of output and leads to the accumulation of toxic products of previous stages of detoxification in the body.
Conclusions: Children with pyelonephritis from polluted areas are often recorded and phenotypic features UCTD determined by elevated levels of free oxyproline in blood and excretion in the urine oxyproline. In violation of the functional state of the parenchyma renal ekonefropathie associated with the violation fibrylohenesis points decrease creatinine excretion and increased hlikozoaminohlikaniv in daily urine, which are more pronounced namely in children UCTD compared with these children, where clinical and laboratory signs UCTD found.
Iskiv Mariana – M.D., PhD student at Institute of Hereditary Pathology of National Academy of Medical Science of Ukraine, Age: 28 years. The age of the presenting author should be indicated as well (below 35).
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Abstract Topics: Tubular Disorders, Inherited renal diseases, Bladder dysfunction & CAKUT &Urinary tract infection.

P-272 MAKING A RAPID GENETIC DIAGNOSIS IN A NEONATE WITH ANURIC RENAL FAILURE

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Introduction: To rapidly make a diagnosis can be of paramount importance in the rare case of neonatal end-stage kidney disease.
Material and methods: We present a girl born at 39 + 1 weeks of gestation to maternal primigravida without consanguinity. The antenatal ultrasounds were normal, including third trimester ultrasounds. She had a birth weight of 2.65 kg. At 21 days of age she presented with lethargy, poor feeding and anuria with the presumed diagnosis of sepsis. She had been breastfeeding well with an admission weight of 3.14 kg. She was intubated and mechanically ventilated and transferred to PICU where she was found to be anuric and hypertensive. Continuous veno-venous hemofiltration (CVVH) was started on day 22 of life and switched to peritoneal dialysis (PD) on day 25. Since no diagnosis had been made, blood was collected for DNA analysis in the patient and both parents as part of the rapid paediatric sequencing (RaPS) research project.
Results: On admission on PICU her plasma creatinine was 504 μmol/L, urea 30.6 mmol/L and serum albumin 27 g/L. Renal ultrasound showed bilateral large and echobright kidneys, with reduced corticomedullary differentiation and multiple small cysts. Plasma oxalate was measured before dialysis initiation and primary hyperoxaluria was excluded. Given the early onset of renal failure, a genetic cause was suspected and she was recruited to the RaPS project. This showed a de novo heterozygous mutation in the WT1 gene and the diagnosis of Denys Drash syndrome was made.
Conclusions: In rare and life-threatening conditions, it can be of major importance to have a quick diagnosis for clinical decision making and family counselling. New techniques like high-throughput DNA sequencing technologies can help to rapidly establish a correct diagnosis.

P-273 NEPHROPATIC CYSTINOSIS – CASE REPORT

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Introduction: Cystinosis is a rare autosomal recessive lysosomal storage disorder characterized by excessive accumulation of cystine within the

lysosome. Cystinosis is caused by mutations in the lysosomal cystine transporter, cystinosin (CTNS). Leucocyte cystine measurement is the cornerstone for both diagnosis and therapeutic monitoring of the disease.
Material and methods: The infantile nephropathic form is the most frequent (95%) and the most severe type of cystinosis. The renal phenotype consists of renal Fanconi syndrome, and a consecutively progressive loss of glomerular function leading to end-stage renal failure.Symptomatic treatment and the specific cystine-depleting therapy represent the mainstay of cystinosis treatment.
Results: We report a first case of nephropathic cystinosis in Slovakia presenting at age 9-months with complete Fanconi syndrome. Major symptoms included failure to thrive, polyuria, polydipsia, episodes of severe dehydration and electrolyte imbalance, vomiting and constipation. Renal Fanconi syndrome, characterized by excessive urinary loss of amino acids, glucose, sodium, potassium and tubular proteinuria has been detected. Nephropathic cystinosis was confirmed by the high concentration of cystine in leukocytes (2.1 nmol/mg protein cystin). The treatment with cysteamin was started at the age of 15- months. The cystine levels significantly decreased to 0.55 nmol/mg protein cystin. In addition, supplementation of potassium, bicarbonate and vitamin D were given. Excessive polyuria was reduced by indomethacin in dose of 2 mg/day. Cystine crystals in the cornea were detected at age of 25- months and Cystagon eye drops were started. Molecular analysis of the CTNS gene is pending.
Conclusions: Cystinosis is the major cause of inherited Fanconi syndrome, and should be suspected in young children with failure to thrive and signs of renal proximal tubular damage.

P-274 PKD1 MUTATIONS IN SISTERS WITH SUSPECTED ARPKD

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Introduction: Two sisters, one with enlarged bright kidneys and one with polyhydramnion and kidney cysts on prenatal ultrasound, went through their newborn period without any kidney complications. The youngest sister was born with oesophagus atresia. They have non-consanguineous parents; their mother had used carbamazepine during the pregnancies due to epilepsy. Examination of the parents showed that the mother had two kidney cysts at the age of thirty. Further investigations have revealed short processus on C3-C6 and rudimentary ribs on C7 in both girls.
Material and methods: In 2006 both sisters had negative genetic examination of ARPKD concerning PKHD1-mutations using dinucleotide Standard Tandem Repeat (STR). The conclusion was based on inheritance of different haplotype from their father. Due to next-generation sequencing (NGS) the possibilities in genetic evaluation have improved. A new blood sample was sent to Germany for analysis in 2016. The laboratory (Bioscientia Human Genetics) used Roche/NimbleGen sequence capture technology on an Illumina HiSeq 1500 system to enrich coding exons of known or potential ciliopathy genes.
Results: Both sisters were found to have heterozygote mutations in c.3490 > A (p.Gly1164Arg) and c.5830 > A (p.Gly1944Arg) on exon 15 in the PKD1 gene. The first mutation is a missense variant and is not previously described; the second mutation is a rare mutation according to The ADPKD Mutation Database and is classified as of clinically intermediate significance.
Conclusions: The missense mutations in c.3490 > A (p.Gly1164Arg) probably have clinical significance in PKD1. It is uncertain if the two heterozygote mutations can explain the skeletal abnormalities, however, a connection with bone defects has been detected in PKD1 mutant mice. These girls now adolescents, are asymptomatic and have normal kidney function. We propose that the aforementioned heterozygote mutations in the PKD1 gene might lead to similar development of disease as in ADPKD. The segregation pattern in this family will be looked further into.

P-275 THREE NOVEL PATHOGENIC MUTATIONS WITH PHENOTYPIC ATYPICAL HUS: SUBSEQUENT WITHDRAWAL OF ECULIZUMAB THERAPY

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Introduction: Haemolytic Uraemic Syndrome (HUS) is most commonly associated with Shiga Toxin producing *Escherichia coli* (STEC). Increasingly non-STEC (atypical) HUS is recognised to be associated with complement dysregulation with specific gene defects identified. Eculizumab a humanised monoclonal antibody binds to complement protein C5, blocking cleavage and preventing production of the terminal complement components C5a and the membrane attack complex (MAC) C5b-9a and is a proven successful treatment in atypical HUS at a cost of €300,000 per annum with little published evidence on the duration of therapy or the ability to withdraw therapy in those responding to therapy. We present a 3.5 year old boy who presented with acute haemolysis following a mild viral infection with no history of diarrhoea. Genetic analysis revealed three mutations that have not been previously reported as pathogenic.
Material and methods: On presentation of a clinical picture of aHUS, supportive therapy was commenced with complete investigation of alternative aetiologies. Stool and blood culture were negative, as was *E.Coli* serology. ADAMTS 13 activity was normal. The pattern of thrombotic microangiopathy was typical, with ongoing haemolysis causing decrements in haemoglobin and platelets despite replacement. Eculizumab was commenced within 48 h of presentation. There was prompt improvement in clinical and biochemical markers. Renal function rapidly normalised, and blood product replacement was not required four days after the first dose administration.
Results: Subsequent genetic analysis demonstrated heterozygosity for two CFI variants c.1402 > G p.(IIe468Val) and c.1642G > C p.(Glu548Gln) and homozygosity for a CD46 variant c.100G > p.(Ala34Thr). These variants were all suggested to be rare benign polymorphisms. He received maintenance therapy with eculizumab for two years with no recurrence of haemolysis or development of further systemic disease. Parental choice was for a trial of withdrawal of Eculizumab which was supported by the nephrology team and Eculizumab was withdrawn as per guidance on rarerenal.org.
Conclusions: The genetic mutations described in our patient are not previously associated with the development of atypical HUS. Of interest as previously described (Loirat C et al. Pediatr Nephrol 2016;31:15) isolated CFI mutations and mutations in CD46 as observed in our patient are associated with an improved prognosis in contrast to CFH mutations.

P-276 PSEUDOHYPOALDOSTERONISM TYPE 1 – A CASE REPORT

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Introduction: Pseudohypoaldosteronism type 1 (PHA1) is due to aldosterone resistance, causing impaired sodium reabsorption and potassium excretion in presence of mineralocorticoid receptor or epithelial sodium channel mutation. Secondary forms exist in infants with urinary tract malformation and/or infection.
Material and methods: -.
Results: A 7-week-old infant (birth weight 4280 g) presented with a failure to thrive and atonic vomiting present since second week of life. Several milk formulas where changed without success and the child was admitted to hospital dehydrated, with an estimated weight loss of 5–10%. Labs revealed hyponatremia (125 mmol/l), hyperkalemia (6,9 mmol/l), metabolic acidosis (7.33; bicarbs 14.6 mmol/l) and hypercalcemia (3.0 mmol/l). Neonatal screening for congenital adrenal hyperplasia was

negative. Parenteral rehydration with 0.9% saline was started. Hormonal profile showed normal 17-hydroxyprogesterone (1,8 ng/ml), ACTH (47 pg/ml), cortisol (280 nmol/l) and high aldosterone (3900 pg/ml, repeated sample 3320 pg/ml). Blood pressure was normal. The diagnosis of pseudohypoaldosteronism type 1 was made and the child was put on sodium chloride supplements.

Conclusions: Pseudohypoaldosteronism type 1 presents as failure to thrive, dehydration and typical electrolyte disturbance (hyponatremia, hyperkalemia and metabolic acidosis and/or hypercalcemia) in presence of resistance to aldosterone.

P-277 DELAYED DIAGNOSIS OF PRIMARY HYPEROXALURIA TYPE 1 – A CASE RAPORT OF TWO FEMALE SIBLINGS

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Introduction: Hyperoxaluria - increased urinary excretion of oxalate exceeding 0,5 mmol (45 mg)/1,73 m2/24 h. The disease spectrum extends from kidney stones, nephrocalcinosis to end stage renal disease. There are three types of primary hyperoxaluria - different in severity and genetic cause. Primary hyperoxaluria type 1 (PH1) is the most common and severe (80% of the cases), autosomal recessive disease, caused by defect in the Vitamin B6 dependent hepatic peroxisomal enzyme, alanine glyoxylate aminotransferase (AGT). It results in increased synthesis and subsequent urinary excretion of oxalate and deposition of insoluble calcium oxalate in, among others, kidneys and urinary tract. The *AGXT* gene is located on chromosome 2q37.3. Currently more than 150 mutation have been identified. PH1 patients with inadequate response to conservative treatment, require kidney or combined liver-kidney transplantation.

Material and methods: We present a case of two female siblings 15 and 17- year old, referred to our Department due to nephrolithiasis, with a diagnosis of medullary sponge kidney. Method. Retrospective analysis of Medical Records, taking into account also results of genetic testing.

Results: Ultrasound revealed nephrocalcinosis grade III and nephrolithiasis in both sisters. Serum creatinine concentration were within normal limits, eGFR were over 60 ml/min/1,73m^2. Twenty-four hour urine collections found low calcium and strongly elevated oxalate excretion (>1,0 mmol/1,73m^2 per day). Raising suspicion of PH we performed genetic testing. They confirmed autosomal-recessive primary hyperoxaluria type I. Both sisters carry two AGXT mutations [c603C > A; c942 + 1G > T] that in combination (in compound-heterozygous state) are causative for their disease. Patients started treatment with high doses of pyridoxamine and citric acid with good tolerance. We observed no further loss of eGFR and decreased excretion of oxalate.

Conclusions: Diagnosis of PH should be made as early as possible to slow progression of the disease. Each case of nephrocalcinosis with hyperoxaluria should be carefully examined in terms of PH.

P-278 CHARACTERIZATION OF POTENTIAL RISK MARKERS OF THE RENAL ARPKD PHENOTYPE

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Introduction: Autosomal recessive polycystic kidney disease (ARPKD) is among the most severe pediatric renal diseases. In contrast to the more common autosomal dominant polycystic kidney disease (ADPKD) clinical or biochemical risk markers are not established for the clinically highly variable ARPKD and defining primary end points for clinical interventional trials remains challenging. We have therefore recently established an international ARPKD registry study to gather longitudinal clinical data of a deeply-phenotyped cohort of ARPKD patients to lay the foundation for the development of clinical risk profiles.

Material and methods: Clinical courses of ARPKD patients are documented in the international, multicenter, observational study ARegPKD using a web-based approach. The renal phenotype is characterized e.g. by data regarding kidney function and sonographic morphology as well as the symptomatic treatment options.

Results: Since start of recruitment in 2013 more than 400 patients from 17 countries have been included with a median follow-up time of 3.4 years (0.1 to 29.3 years). Renal symptoms vary substantially, but hyperechogenic kidneys with decreased or abrogated cortex-medulla differentiation and cysts are observed throughout all age groups. Kidney pole-to-pole-length and total kidney volume, as determined by sonography, increase with age. Interestingly however, height-adjusted pole-to-pole-length decreases with age, while the ratio of kidney volume to body height remains relatively stable. Renal function shows variable courses depending on the age of presentation. Furthermore, the data suggests an association of larger kidneys with reduced kidney function. Renal

survival after 10 years is about 85%, after 20 years about 60%. There is no gender-specific effect.
Conclusions: ARegPKD represents the largest international ARPKD cohort with detailed and longitudinal clinical follow-up data. We describe the renal phenotype in detail and present first data on associations that may become valuable as risk markers in daily clinical life. Data evaluation for assessment of further clinical and prognostic markers is ongoing

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P-279 SEVERE RENAL PHENOTYPE IN A PATIENT WITH BOTH TUBEROUS SCLEROSIS AND AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE

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Introduction: We describe the phenotype of a child presenting with a heterozygous deletion involving two renal associated genes; TSC2 (associated with tuberous sclerosis complex) and PKD1 (associated with autosomal dominant polycystic kidney disease (ADPKD), resulting in abnormalities of kidneys, heart, brain and development.
Material and methods: Case report.
Results: We describe a 5 year old Caucasian male who is the child of non consanguineous parents who presented at 8 weeks of age with tachycardia secondary to multiple cardiac rhabdomyomas. Further imaging revealed bilateral, multiple, simple renal cysts and a cranial ultrasound showed subependymal nodules and cortical tubers consistent with a diagnosis of tuberous sclerosis. Chromosomal microarray detected a complex rearrangement at 16 p13.3 defined as a 13 kilobase deletion of TSC2 extending into the nearby PKD1 gene as well as a duplicated segment of the PKD1. This is known as TSC2/PKD1 contiguous gene syndrome (CGS). He began to experience frequent infantile spasms at 7 months of age which were refractory to treatment. Simultaneously he developed hypertension and was admitted to PICU with a hypertensive crisis. A fusiform aneurysm of the intracranial portion of the right internal carotid artery has been present since the age of 3 years. His growth plots greater than the 99th centile for all parameters, but all investigations for an overgrowth syndrome have been negative, systemic overgrowth has not previously been reported in these patients. He is also dysmorphic, has characteristic facial angiofibromas and marked developmental delay.
Conclusions: A deletion of contiguous genes affecting the renal system can cause a particularly severe and multisystem phenotype with earlier age of onset of renal sequelae than in typical TSC or ADPKD. Whilst mTOR inhibition therapy is established in patients with TSC it's effect in patients with CGS is unproven and requires further evaluation.

P-280 THROMBOEMBOLISM IN CHILDREN WITH CONGENITAL NEPHROTIC SYNDROME – LESSONS FROM AN ESPN SURVEY

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Introduction: Thromboembolism is a serious and potentially fatal complication in children with congenital nephrotic syndrome (CNS).
Material and methods: We conducted a 6-year survey across members of the ESPN Dialysis Working Group to evaluate thrombosis risk, treatment and outcome in children with CNS across Europe.
Results: Eighty-two children (52% male) were included from 19 tertiary nephrology units in 11 European countries. Median age at presentation with CNS was 9.5 (IQR 0–164) days with S-albumin 11 (4–29) g/L and S-creatinine 27 (2–480) μmol/L. Prophylactic antithrombotic medications were given in 47 (57%); Warfarin being the most common (51%). Ten (12%) patients developed thrombosis: 5 not on prophylaxis (14.3%) versus 5 on prophylaxis (10.6%) ($p = 0.618$); At thrombosis median age was 1.5 (0.7–5.5) months and median S-albumin 15 (10–22) g/L. Nine patients received regular albumin infusions with diuretic treatment and 5 were on ACE inhibitors. No patient had a history of preceding dehydration. Nine patients had central lines in situ, and one patient was on haemodialysis at the time of thrombosis. In 5 thrombus formation was at the site of the central line (infection related in one) and 4 needed line removal. Symptoms were catheter malfunction in 3, asymmetric extremity symptoms in 2, convulsion in 1 and fever with malaise in 1. Three were asymptomatic and diagnosed by routine heart ultrasound. All patients received therapeutic anticoagulation (heparin in 7 or warfarin in 3) and one additional thrombolysis. At final follow-up (median 22 months), complete resolution of thrombus was achieved in 4, partial in 2. One patient had ongoing convulsions and 2 died (17 days and 7.5 months after thrombosis).
Conclusions: In infants with CNS the risk of thrombotic episodes is high and the benefit of prophylaxis needs to be considered. Treating clinicians need a high level of vigilance, particularly in infants with central lines.

P-281 LONG TERM RESULTS OF RITUXIMAB IN A TERTIARY REFERRAL CENTER FOR DIFFICULT-TO-TREAT NEPHROTIC SYNDROME

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Introduction: Rituximab (RTX) has been proposed as an alternative treatment for steroid dependent nephrotic syndrome (SDNS), frequently relapsing nephrotic syndrome (FRNS) and steroid resistant nephrotic syndrome (SRNS).
Material and methods: We evaluated the data of SDNS, FRNS and SRNS patients under RTX treatment. Initial RTX course consisted of 2–4 weekly infusions at the dose of 375 mg/m2.
Results: A total amount of 38 patients (20 girls, 18 boys) were included in the study. The median age of NS diagnosis was 4.3 years (IQR, 1.8–12.1 years). Renal biopsy performed to all patients before RTX and revealed focal segmental glomerulosclerosis in 20, minimal change disease in 13 and other diagnoses in 5 patients. A total of 21 patients were categorized as SRNS, three patients as FRNS and 14 patients as SDNS according to steroid response. All SRNS patients were also resistant to calcineurin inhibitors (CNI). Median duration between nephrotic syndrome and initial RTX dose was 3.7 years (IQR; 1.7–8.4). The mean age of RTX treatment was 11.7 ± 4.9 years. Transitory side effects were observed in two patients (throat soarness, erythematous rash; respectively). Mean duration of follow-up after RTX treatment was 2.7 ± 1.6 years. Nineteen patients received regular maintenance treatment every 6–9 months. At last visit, six (28.5%) out of 21 SRNS patients were in remission. In three out of these six SRNS patients with remission, steroids and CNIs could be also stopped. Two (66.6%) out of three FRNS were relapse free after RTX treatment at last visit. In SDNS group, steroids or CNIs could be stopped in 11 out of 14 patients (78.6%). In SDNS group, five patients received regular RTX treatment every 6–9 months. In SDNS group, mean steroid dose at last visit was lower than before RTX treatment ($p = 0.008$).

Conclusions: Rituximab seems to be effective and safe treatment option for difficult-to treat nephrotic syndrome especially for SDNS and FRNS patient groups.

P-282 LONG-TERM EFFECT OF RITUXIMAB TREATMENT: TWO SIDES OF THE SAME COIN

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Introduction: Rituximab (RTX), an anti-CD20 antibody, is effective in pediatric idiopathic nephrotic syndrome (INS). However, memory B cell subsets remain significantly reduced with long-term immunological impairment.
Material and methods: B cell subsets were assessed by flow cytometry in 26 steroid-dependent INS children, with a minimum of 5 years after the 1st RTX infusion. For some patients levels of total immunoglobulins and of specific immunoglobulins to Measles, Tetanus, and Hepatitis B were assessed. At baseline, all patients were in remission on prednisone and 1 or 2 steroid-sparing agents (calcineurin inhibitors and mycophenolate mofetil). An initial dose of RTX (375 mg/m^2), repeated at 7 days in case of incomplete depletion of B cells, was administered. All patients tapered concomitant immunosuppression.
Results: Seven patients maintained complete remission during the entire follow-up. All other patients relapsed. Ten patients were retreated with RTX and 2 patients maintained complete remission after the 2nd RTX infusion whereas the others relapsed. Survival analysis showed a significantly delayed time to relapse ($p = 0.015$) comparing 2nd to 1st RTX infusion. Three patients were retreated with a 3rd RTX infusion and relapsed. At last follow-up, immunosuppression was significantly reduced compared to baseline (0.7 ± 1 vs 2.7 ± 0.5, $p < 0.001$) and all patients were in remission. Total CD19$^+$ cells were recovered, but 21 patients still showed reduced switched memory B cells (<1.1% of total lymphocytes). Nine/20 patients showed reduced levels of IgG (<700 mg/dl) and 6/20 hypogammaglobulinemia (<480 mg/dl). Interestingly, low levels of IgG and/or IgA were observed in 3/5 non-relapsers after just one RTX infusion. Regarding vaccine response, we observed that only 4/19 patients resulted positive for anti-Hepatitis B IgG, 6/14 for anti-Measles IgG, and 3/11 for anti-Tetanus IgG.
Conclusions: Although RTX treatment is an effective therapy for pediatric INS, it may have long-lasting effects on their immune memory also following a single RTX infusion.

P-283 PREDNISOLONE TREATMENT OF RELAPSES OF STEROID SENSITIVE NEPHROTIC SYNDROME MAY RESULT IN LOSS OF BRAIN GREY AND WHITE MATTER VOLUME

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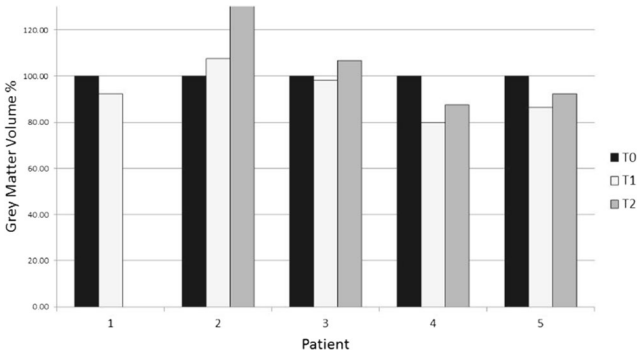
Introduction: Children with steroid sensitive nephrotic syndrome (SSNS) may receive multiple courses of high dose prednisolone for treatment of relapses. Behavioural change is a frequent complication. This study aimed to undertake multi-parametric brain magnetic resonance imaging prior to, during and following completion of high dose prednisolone. Analysis included both anatomical structural assessment as well as cerebral perfusion at all three time points.
Material and methods: Three boys and two girls aged 7-12y with relapsing SSNS (1–11 relapses) and no prior history of exposure to drugs other than prednisolone underwent brain imaging at the time of relapse, prior to commencement of high dose prednisolone (T0). Imaging was repeated on D21 of high dose prednisolone (T1) and again once prednisolone had been discontinued (T2). Imaging was performed on a 3 T Philips Achieva scanner. We acquired a whole brain T1 volume 0.9mm^3 MPRAGE for anatomical segmentation (FSL, Oxford, UK). In addition we assessed cerebral perfusion using an arterial spin labelling technique analysed using in house MATLAB routines (www.mathworks.com).
Results: Only one child failed to complete all scan time-points. Four children showed a reduction in total brain volume at T1 with recovery to baseline in only one of these children by T2. Whilst there was loss of both grey and white matter the main driver for this volume loss was grey matter reduction (Figure 1).



In three children there was a corresponding reduction in cerebral perfusion at T1 which only recovered to baseline in one child.
Conclusions: High dose prednisolone for treatment of SSNS relapses may result in permanent reduction in both grey and white matter volumes and cerebral perfusion. Further work is ongoing to confirm and further refine these observations.

P-285 CLINICAL SIGNIFICANCE OF EXTENDED- SPECTRUM BETA-LACTAMASE PRODUCING BACTERIA IN CHILDHOOD FIRST FEBRILE URINARY TRACT INFECTION AND ITS DIFFERENCES BETWEEN AGE GROUPS

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Introduction: Extended-spectrum beta-lactamase(ESBL) producing bacteria induced urinary tract infection(UTI) is increasing in frequency and resistant to most of penicillins and cephalosporins, need more potent antibiotic such as carbapenem. Previous results of ESBL(+)UTI in children were different in severity and outcomes, and there was no report of ESBL(+)UTI comparing between age groups. The aim of this study were to evaluate clinical significance of ESBL(+)UTI under 5-year-old children for selecting proper antibiotics and finding out prognostic factors of outcome, and also comparing differences between age groups.
Material and methods: We retrospectively studied 288 patients with first febrile UTI under 5-years-old children. Patients were divided into ESBL(+)UTI and ESBL(−)UTI. Clinical characteristics and outcome were compared, and also young infants group(onset age < 3 months) were compared with older age group.
Results: The mean age of patients was 6 months and M:F ratio was 7:3. The incidence of ESBL(+)UTI were 11%. ESBL(+)UTI had more pre-onset admission history($p = 0.02$) and recurrenec of UTI($p = 0.045$). In antimicrobial susceptibility test(AST), 3rd cephalosporin were all resistant in ESBL(+)UTI, but 98% responded clinically. Results of susceptibility were 100% for

amikacin and 81% for gentamycin. In young infant group(<3 months), ESBL(+)UTI were 13% in incidence, had more pre-onset hospitalization history (p = 0.002), prenatal hydronephrosis(p = 0.015), higher CRP(p = 0.04) and recurrence of UTI(p = 0.02) than older age group.
Conclusions: ESBL(+) UTI need more attention because of high recurrence rate. Infants(< 3 months) with pre-onset history of admission had more severe infection and recurrence rate, so we should select antibiotics carefully. The 3rd cephalosporins showed resistance in AST, but can be used as first-line empirical antibiotics because of its high clinical response rate. In ESBL(+) UTI resistant to 3rd cephalosporine, we can consider aminoglycoside as a second-line antibiotics before start carbapenem.

P-286 URINARY POTASSIUM TO URINARY POTASSIUM PLUS SODIUM RATIO CAN ACCURATELY IDENTIFY HYPOVOLAEMIA IN NEPHROTIC SYNDROME

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Introduction: Background: Acute Kidney injury (AKI) as a complication of idiopathic nephrotic syndrome is uncommon. However, evidence exists pointing to a decrease of GFR in a subgroup of nephrotic children, likely secondary to hypovolaemia. Aim: To validate the use of urinary potassium to the sum of urinary potassium plus sodium ratio (UK/UK + UNa) as an indicator of hypovolaemia in nephrotic syndrome enabling detection of those patients who will benefit from administration of albumin.
Material and methods: Methods: We prospectively studied 44 nephrotic children before and after administration of a water loading test and compared different parameters to a control group (36 children). Renal perfusion and glomerular permeability were assessed by measuring clearance of para-aminohippurate and inulin. Vaso-active hormones (plasma renin activity, aldosteron) and urinary sodium and potassium were also measured.
Results: Results: Subjects were grouped into low, normal, and high GFR groups based on reference values obtained from the control group. In the low GFR group hypovolaemia was seen as demonstrated by significantly lower renal plasma flow (p = 0.01), filtration fraction (p = 0.01) and higher UK/UK + UNa (p = 0.03) ratio. In addition, non significant higher plasma renin activity (p = 0.11) and aldosteron (p = 0.09) were also seen in the low GFR group compared to high GFR and normal GFR groups.
Conclusions: Conclusion: A subgroup of patients in nephrotic syndrome has a decrease in glomerular filtration, apparently related to functional hypovolaemia which can be detected by an elevated urinary potassium to urinary potassium plus sodium ratio (> 0.5–0.6). In the clinical setting, this group will most likely benefit from albumin administration.

P-287 MUTATIONS IN LAMB2 ARE ASSOCIATED WITH FSGS AND SEPTO-OPTIC DYSPLASIA

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Introduction: Mutations in LAMB2, the gene encoding laminin-β2, a multidomain protein are associated with Pierson Syndrome, an autosomal recessive disorder characterized by congenital nephrotic syndrome, ocular abnormalities including microcoria and neurodevelopmental delay. Very few patients survive to adolescence, with only 3 reported to date. We present an 11 year old male born to a non-consanguineous pedigree, who presented at 5 years of age with steroid resistant nephrotic syndrome and focal segmental glomerulosclerosis together with poor vision, growth hormone deficiency and seizures. MRI of his brain showed a global lack of white matter, a small anterior pituitary and bilateral hypoplastic optic nerves, features which are reminiscent of septo-optic dysplasia (SOD). His current estimated eGFR is 59mls/min/1.73 m2 with urine albumin/creatinine ratio measuring 834 mg/mmol with his medications consisting of angiotensin receptor blockade. SOD is characterized by the presence of two or more of the following: hypopituitarism with isolated or combined hormone deficiencies, optic nerve hypoplasia and/or midline brain defects including absence of the corpus callosum and septum pellucidum. The pathogenesis of SOD is multifactorial arising from either environmental causes, viral infections and alcohols. A hereditary aetiology has also been described with recessive and dominant cases reported and several genes implicated including HESX1 which plays a key role in pituitary morphogenesis. We describe a known SRNS disease gene implicated in SOD phenotype presenting with childhood onset FSGS.
Material and methods: Genetic analysis was undertaken using whole exome sequencing with validation by Sanger sequencing. Dual immunofluorescent microscopy was employed to validate loss of function and expression in patient biopsies relative to controls.
Results: Using whole exome sequencing, we identified compound heterozygous missense mutations in LAMB2 [c.737G > A p.Arg246Gln, c.3982G > A p.Gly1328Ser]. Dual immunofluorescent histochemistry revealed reduced glomerular laminin-β2 expression compared to control biopsies [time zero renal transplant]. Interestingly, laminin β2 is expressed during murine anterior pituitary morphogenesis and analysis of murine Lamb2 mutant pituitary morphogenesis is currently underway.
Conclusions: Our case raises the possibility that mutations in LAMB2 may be associated with SOD-related phenotypes. We propose that patients presenting with genetically undefined SOD should be screened for albuminuria.

P-288 CONGENITAL NEPHROTIC SYNDROME: A NOVEL NON-PATHOGENIC INTRAGENIC DELETION OF WT1 WITH DEVELOPMENT OF WILMS TUMOUR: C.[643-6C > A];[(=)]

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Introduction: Mutations in the WT1 gene are associated with Congenital Nephrotic Syndrome (CNS) and Wilms Tumour. We present a female infant who presented at age 4 months markedly oedematous, hypoalbuminaemic and with nephrotic range proteinuria. Immediate initial management consisted of haemofiltration and albumin replacement with subsequent therapy consisting of regular albumin infusions and iatrogenic reduction in GFR with ACE inhibition and NSAID therapy.
Material and methods: A diagnosis of CNS was made clinically with an initial PCR of >7000 mg/mmol in the presence of oedema and hypoalbuminemia. Genetic analysis did not identify a known causal genotype, however a likely non pathogenic Wt1 mutation class 2 was identified c.[643-6C > A];[(=)]. The mutation was rediscussed with the primary analyst and local geneticist due to concerns of risk of developing Wilms Tumour and the need for subsequent USS monitoring. On rediscussion it was still felt to be likely non-pathogenic as no previous intragenic deletions of Wt1 have previously been found to be pathogenic.
Results: At 11 months of age she underwent bilateral nephrectomy due to ongoing hypertension despite maximal medical management resulting in

PRES. At resection a left renal mass was noted arising from the anterior surface of the medial aspect of the left kidney. Pathology revealed an incompletely excised stage III Wilms Tumour with capsular spread and spread to the perinephric tract. There was no metastatic spread on full body imaging and she underwent an 11 day course of radiotherapy and 28 weeks of chemotherapy of Vincrsitine, Actinomycin and Doxorubicin.
Conclusions: The identification of pathogenic mutations in the Wt1 gene are crucial in identifying patients at risk of developing Wilms Tumour with requirement of ongoing ultrasound surveillance. We report a case of congenital nephrotic syndrome and Wilms Tumour with mutation c.[643-6C > A];[(=)] that phenotypically may be significant in our patient and we are not aware of intragenic deletions reported as pathogenic elsewhere in the literature.

P-289 VITAMIN D DEFICIENCY – RISK FACTOR FOR GROWTH RETARDATION IN CHILDREN WITH IDIOPATHIC NEPHROTIC SYNDROME?

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Introduction: The children with idiopathic nephrotic syndrome /INS/ are at increased risk of Vitamin D /VitD/ deficiency due to long-term steroid treatment, protein-calorie malnutrition and urinary loss of VitD-binding protein. It is known that high-dose steroid therapy impairs normal growth. Is the VitD deficiency also a risk factor for growth retardation?
Aim: To determine the VitD status in children with steroid sensitive INS and to analyze the correlation between VitD deficiency and growth retardation.
Material and methods: We report observation in 32 children −14 girls and 18 boys (age 8.24 ± 4.2 yr) with INS and normal renal function. Height was expressed as SD scors /Ht_{SDS}/, using references for Bulgarian healthy children and WHO growth standarts. Serum levels of 25(OH)D and PTH were assayed. Bone age was assessed according to Greulich&Pyle.
Results: We found short stature (mean Ht_{SDS} − 2.3) in 9.38%ofallpatients. In 18.75% growth retardation (mean Ht_{SDS} − 1.4) and in 71.87% of the children normal height for age were established/$p = 0.0001$/. The children with severe VitD deficiency showed significant growth impairments (22.2% - short stature; 44.4% - growth retardation), than these with VitD insufficiency (16.67% - growth retardation) and normal VitD status (9.1% - short stature) /$p < 0.05$/. Retardation of bone age in seven nephrotic children was established. The serum levels of 25(OH)D showed a significant negative correlation with alterations in the height /$p = 0{,}009$;$r = -0.41$/. We found no relationship between VitD level and bone age/$p > 0.05$/.
Conclusions: The results of our study show that children with INS andVitD deficiency are at high risk of growth retardation. VitD supplementation is required in conditions with insufficiency or deficiency in order to prevent growth impairment.

P-290 PUROMYCIN AMINONUCLEOSIDE INDUCES PODOCYTE APOPTOSIS BY ENDOPLASMIC RETICULUM STRESS

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Introduction: Puromycin aminonucleoside (PAN) is known to be a podocytotoxin, therefore, PAN-induced nephrosis is a widely studied animal model of human idiopathic nephrotic syndrome. Endoplasmic reticulum (ER) stress is the common findings under various pathogenic microenvironments, contributing to the progression of various podocyte diseases. Abnormal protein accumulation associated with ER stress in the ER of podocytes produces structural and functional damage in the cells, which in turn leads to podocyte apoptosis and severe proteinuria. In the present study, we investigated the effect of PAN on ER stress and apoptosis in in vitro podocytes.
Material and methods: We cultured rat and mouse podocytes and treated with various concentrations of PAN and evaluated ER stress markers by western blotting and apoptosis by FACS and TUNEL assays.
Results: PAN treatment increased ER stress markers, such as, ATF6 and caspase-12 at 12 and 24 h, in a dose-dependent manner, which were improved by chemical chaperones, such as, sodium 4-phenylbutyric acid (PBA) and TUDCA. PAN also induced podocyte apoptosis significantly in concentration- and time-dependent manners in FACS and TUNEL assays, which were improved by Nox4 siRNA, ATF6 siRNA, and chemical chaperones. LY294002, a PI3-kinase inhibitor, exaggerated ER stress and apoptosis significantly. Therefore, PAN induced ER stress, thereafter, increased oxidative stress, subsequently induced podocyte apoptosis by the inhibition of PI3-kinase signaling.
Conclusions: Our studies suggest that PAN could induce podocyte ER stress of mainly ATF6 and caspase-12 pathways, which would contribute to the development of podocyte apoptosis by oxidative stress and inhibiting PI3-kinase survival signaling.

P-291 SURVEILLANCE BIOPSIES OF TACROLIMUS EFFECT IN CHILDHOOD NEPHROTIC SYNDROME

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Introduction: Tacrolimus is increasingly advocated as a steroid sparing agent in the management of nephrotic syndrome. As concerns exist about long term nephrotoxicity, interval renal biopsies have been advocated to detect early changes. Associations between histological changes and patient factors are not well established.
Material and methods: Single centre review of surveillance renal histology of children who received tacrolimus to treat nephrotic syndrome between 2002 and 2015. For consistency a single histologist independently re-analysed all histology.
Results: 26 children (16 male, 1 non-caucasian, 25 minimal change, 1 FSGS) commenced tacrolimus at median age 4.9 years (range 1.4–8.9). 7/26 (27%) of first biopsy taken after median duration 4.7 years (range 1.9 to 6.9) demonstrated features of calcineurin inhibitor (CNI) nephrotoxicity. Toxicity was associated with higher mean 12-h trough serum tacrolimus levels: 0/11 showed toxicity with levels o 4.36 to 5.54 μg/L vs. 7/15 (47%) with levels 5.69 to 7.36 μg/L ($p < 0.0001$). No association was found with number of relapses, gender or duration of time on tacrolimus. Nephrotoxicity was mainly globally sclerosis or minimal chronic tubular changes. 10/19 patients without initial CNI toxicity underwent second biopsy at a median time of 5.0 (range, 1.8–6.7) years later. One developed toxicity. Estimated glomerular filtration rate in those who developed toxicity was normal. We attempted tacrolimus weaning in 20 patients: 11 patients relapsed, 4 within 2 months, further 5 within 1 year after dose reduction.
Conclusions: Significant number of children on tacrolimus showed histological nephrotoxicity after a short duration of therapy. In this small single centre experience, toxicity correlated with tacrolimus level rather than duration. A high number relapsed shortly following dose reduction. We suggest maintaining tacrolimus levels ≤5.5 μg/L to minimise nephrotoxicity.

P-292 RITUXIMAB TREATMENT FOR REFRACTORY STEROID-DEPENDENT NEPHROTIC SYNDROME (SDNS) IN CHILDREN: A SINGLE-CENTER STUDY

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Introduction: We evaluated the efficacy and safety of RTX in children with refractory SDNS treated in our center
Material and methods: Sixteen patients who were included in prospective single-center study remained dependent on high doses of prednisolone - the median 0.42 mg/kg/day, revealed a high incidence of relapses NS - the median 2.6 times/years, despite the use of different immunosuppressants, and had clinical signs of severe steroid toxicity. At the beginning of RTX therapy, the median age of the patients was 11.5 years. During the first course of RTX, one to four infusions were administered, the majority of patients (69%) received two infusions.
Results: Within 6 and 12 months after an initial treatment of RTX, 15 (94%) of 16 patients showed no relapses of NS. Steroids has been discontinued in 50% of patients after 6 months and in 64% at 12 months, in other children the dose of prednisolone was significantly reduced to 0.09 mg/kg/day (range 0.04–0,25) (p = 0.043) and 0,04 mg/kg/day (range 0.02–0,16) (p = 0.011), respectively. Calcineurin inhibitors have been discontinued in half of the patients, six months after the initial course. Eight patients had a long-term follow-up period of 2 to 7.5 years (median 2.8 years), long-term remission of NS from one to three years without prednisolone is maintained in three (37%), the others were on therapy of low-dose prednisone (the median 0.05 mg/kg/day, range 0.02–0.08) with significant reduction in the frequency of relapses – the median 0.1 times/year, p = 0.012. Cyclosporine have been discontinued in all patients. Two (22.2%) of the sixteen patients developed agranulocytosis 4 months after the initial and repeated courses of RTX. In half, hypogammaglobulinemia was detected. One had self limited pulmonary tuberculosis.
Conclusions: Rituximab shows a significant reduction in the frequency of relapses, as well as a significant steroid-sparing and cyclosporine-sparing effect at refractory SDNS. Carefull follow-up is mandatory.

P-293 DISSEMINATED BARTONELLA HENSELAE INFECTION IN A CHILD RECEIVING MYCOPHENOLATE MOFETIL FOR IDIOPATHIC NEPHROTIC SYNDROME

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Introduction: Mycophenolate mofetil (MMF) is an efficient steroid-sparing drug in steroid-dependent idiopathic nephrotic syndrome (SDNS). Infectious complications have been rarely reported in patients receiving only MMF.
Material and methods: We report on a 7-year old girl receiving MMF for SDNS and developing disseminated *Bartonella Henselae* infection.
Results: The patient was receiving MMF for one year when she presented fever. The daily dose of MMF was 920 mg/m^2; it had been decreased from 1220 to 920 mg/m^2 11 months before the onset of symptoms because of a MMF area under the curve of 114 mg.h/L. After 5 days of fever, clinical examination showed an isolated 15 mm-diameter pre-ear tragus lymph node. Blood formula was normal, whilst serum ALAT and gammaGT were increased (318 and 72 IU/L, respectively), serum protein C reactive subnormal (9.7 mg/L), blood EBV and CMV viral load and *Bartonella Henselae* serology were negative. Abdominal ultrasonography showed multiple abdominal lymph nodes (diameter, 10 to 22 mm) at liver and spleen hilums, with one liver and multiple spleen hypodense nodules. Bone marrow aspiration was normal. PCR for *Bartonella Henselae* was positive on surgical biopsy of the cervical lymph node performed after 13 days of fever. The child had a 5 year-old cat at home and played with kittens during the last weeks. MMF was withdrawn and prednisolone restarted at 10 mg every other day. The child received clarithromycin and amikacin for 10 days, and then clarithromycin for 4.5 months. Fever disappeared within 4 days and abdominal lymph nodes after 3 months.
Conclusions: Disseminated *Bartonella Henselae* infection has been described in patients receiving a combination of immunosuppressive agents after transplantation. This case illustrates that it can also be observed in a patient receiving only MMF. Thus, direct contact between such patients and cats (especially kitten) should be avoided, and MMF exposure should be minimized if possible.

P-294 DOES C.3979 G > A/P.VAL1327MET VARIANT OF COL4A4 HAS ANY PATHOGENIC EFFECT IN TURKISH PATIENTS WITH ALPORT SYNDROME?

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Introduction: Alport syndrome is an inherited glomerular disease caused by mutations in COL4A3,COL4A4 or COL4A5 encoding the type IV collagen α3,α4, and α5 chains.The relationship between the various genotypes and phenotypes in AS has not been fully elucidated. In this study, we report underlying mutations in a family with Alport syndrome.
Material and methods: We had analyzed four siblings(three girls,one boy) presented with hematuria and/or proteinuria. DNA isolation was performed using Qiamp kit from peripheral blood samples and COL4A3,COL4A4 and COL4A5 genes were analyzed with Next Generation Sequencing Multiplicom MASTR(Niel,Belgium) kit with 139 amplicons and sequenced at MISEQ platform.
Results: Two sister and brother presented with microscopic hematuria and one girl with proteinuria. Renal functions were normal in all patients and father of the siblings has microscopic hematuria. The mutation detected at COL4A3 was; c.3644_3646delGAGinsAAA/ p.Arg1215_Gly1216delinsGlnArg and was reported as "pathogenic" with a frame change and possible structural abnormality. In addition, the heterozygous variant identified at COL4A4 was c.3979G > A/p.Val1327Met which was reported as having conflicting pathogenicity due to its unknown effect on the glomerular basal membrane. The pathogenic mutation of COL4A3 was present in three sisters but absent in brother. The heterozygous variant detected at COL4A4 was present in brother and one sister who also had pathogenic COL4A3 mutation.Only the sister who was carrying pathogenic COL4A3 mutation along with the heterozygous variant,has moderate proteinuria in the family.
Conclusions: The detection of COL4A4 c.3979G > A/p.Val1327Met heterozygote mutation,which was previously reported as nonpathogenic variant, in a child with hematuria and Alport syndrome phenotype suggests that this mutation may be pathogenic, leading to mild form of the disease alone. In addition, coexistence of this mutation with pathogenic COL4A3 c.3644_3646delGAGinsAAA/p.Arg1215_Gly1216delinsGlnArg mutation may lead more severe forms of the disease with proteinuria. Molecular analyses are important to identify new mutations to clarify their clinical importance and to assess the prognosis of the disease.

P-295 COQ2 MUTATION, AN INHERITED MITOCHON DRIOPATHY, IN FAMILIAL NEPHROTIC SYNDROME

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Introduction: The majority of steroid-resistant nephrotic syndrome (SRSN) is genetically determined. Recessive coenzyme Q2 (COQ2) mutations have recently been identified. Most affected individuals presented with neurological and muscular symptoms, whereas nephrotic syndrome (NS) has been reported very rarely. We here present siblings with SRNS. Genetic analysis performed in one of them revealed pathogenic compound heterozygous COQ2 mutations.
Material and methods: Laboratory investigations were performed in Yerevan and Zurich, renal biopsy was evaluated in Zurich (Switzerland) and molecular genetics studied in Marburg and Ingelheim (Germany).
Results: A girl aged 17 months of Armenian origin was admitted with severe NS, microhematuria and oliguria. No known consanguinity. Treatment with prednisolone for 8 weeks followed by cyclosporine A and ACE inhibitors was not effective. Two months after admission right-side hemi-myoclonus and hydrocephalus documented by MRI were diagnosed. NS persisted, renal function deteriorated (serum creatinine at the age of 2 y 3 months was 463 μmol/l) leading to death. No renal biopsy or genetic analysis could be done. Two years later her younger brother aged 18 months was admitted with NS, microhematuria and oliguria. No extrarenal abnormalities. Renal biopsy revealed FSGS (NOS, not otherwise specified) and mild irregularities of the glomerular basement membrane; no abnormal mitochondria. Genetic analysis showed compound heterozygous mutations in COQ2 (maternally, COQ2 p.Leu340Val; paternally, COQ2 p.Arg173Leu). Treatment with ubiquinone (300 mg/d) introduced at CKD stage3 did not prevent progression to stage 5 at age 26 months.
Conclusions: Isolated SRNS due to COQ2 mutations is extremely rare. Treatment with ubiquinone (coenzyme Q10) may reverse the NS. Genetic analysis in familial cases with early renal manifestations is therefore of utmost importance. This is a good example of cooperation of different laboratories and centres in Europe.

P-296 TRENDS IN HISTOPATHOLOGICAL PATTERN AMONG CHILDREN WITH IDIOPATHIC NEPHROTIC SYNDROME

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Introduction: Recent studies found that there is increasing incidence of focal segmental glomerulosclerosis (FSGS) in children with idiopathic nephrotic syndrome (INS). We aimed at study of the histopathological pattern in children attending the Alexandria University Childrens Hospital.
Material and methods: Review of the files of 356 child with INS from the year 1992 to 2001 (group A) and comparing their data with those of 185 child from the year 2010 to 2014 (group B).
Results: The mean age was 9.72 ± 4.7 years and age at onset of the disease was 4.97 ± 2.97 years for group A, while the mean age was 7.35 ± 3.02 years and age at onset of the disease was 4.46 ± 2.57 years for group B. Renal biopsy was performed in 63 patients in group A and 34 patients in group B because of steroid resistance or frequent relapse. Steroid responsive patients were considered to have minimal change nephrotic syndrome (MCNS). The incidence regarding mesangioproliferative glomerulonephritis (MesPGN), FSGS, Membranoproliferative glomerulonephritis (MPGN) and MCNS was 7.8%, 2.3%, 4.3% and 77.9% respectively in group A, while it was 10.1%, 6.8%, 3.4% and 79.7% respectively in group B.
Conclusions: MCNS is still the predominant histopathological type. Although there was almost a threefold increase in the incidence of FSGS, MesPGN was the next coming histopathological type after MCNS.

P-297 A BOY WITH CONGENITAL NEPHROTIC SYNDROME ASSOCIATED WITH MICROCEPHALY AND NEUROLOGICAL ABNORMALITIES

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Introduction: Congenital nephrotic syndrome (CNS) is defined as a nephrotic syndrome occurring before 3 months of age. As many as 85% of the cases have a genetic basis for the renal disease and a poor outcome. There are other sydromes where CNS is associated with microcephaly and neurologic symptoms, for example Galloway-Mowat Syndrome, Arthrogryposis-renal dysfunction-cholestasis (ARC) syndrome, progressive encephalopathy-hypsarytmia-optic atrophy (PEHO) syndrome, primary coenzyme Q10 deficiency (COQ2 gene mutation) or carbohydrate-deficient glycoprotein (CDG) syndrome. The pontocerebellar hypoplasia (PCH) is characterised by prenatal development of an abnormally small cerebellum and brain stem, which is caused either by specific gene mutations or it is secondary associated to chromosomal abnormalities, neurodegenerative or neurometabolic disorders.
Material and methods: A hypotrofic newborn with perinatal asfyxia, hypotonia, hyporeflexia, microcephaly and facial stigmatisation manifested with CNS in 45. day of life without any reaction to corticotherapy. Later the oedema disappeared, the level of albumine was more than 25 g/l but there was still nephrotic proteinuria. Brain MRI showed the pontocerebellar hypoplasia.
Results: Genetic testing for the most frequent causes of CNS (NPHS1, NPHS2 and WT1 gene) was negative. Regarding the clinical findings, COQ2 mutation, CDG syndrome and the mutations of genes for pontocerebellar hypoplasia were considered. The screening tests for COQ2 mutation and CDG syndrome proved negative. However, mutation in EXOSC3 gen which causes type 1B of pontocerebellar hypoplasia was confirmed. The boy died at the age of 5 months due to cardiopulmonary arrest during respiratory infection.
Conclusions: This case report shows the coincidence of CNS with PCH type 1B. The most frequent mutations causing CNS were excluded. COQ2 mutation and CDG syndrom as potential causes of CNS with neurologic abnormalities were excluded also.

P-298 FROM UGANDA TO ITALY: A CASE OF NEPHROTIC SYNDROME SECONDARY TO PLASMODIUM INFECTION. QUARTAN MALARIA NEPHROPATHY AND KIDNEY FAILURE

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Introduction: Malaria (M), the first parasitic infection, is sometimes associated with nephrotic syndrome (NS) in tropical areas. Kidney involvement during quartan malaria is represented by immune-complex mediated glomerulonephritis (GN). Generally NS develops several weeks after onset of quartan fever and its clinical course proceeds slowly to end-stage kidney disease (ESKD) even after eradication of the infection.
Material and methods: A 17-year-old Ugandan boy was transferred in Italy, as part of a humanitarian project, to treat ESKD with onset 13 months before as NS. In Uganda, the patient received steroids and immunosuppressive therapy with poor results and some dialysis sessions. On admission in our department, clinical examination showed a pale, febrile and edematous boy with hypophonesis in the right hemithorax and a malodorous purulent secretion from exit site of central venous catheter.
Results: Laboratory tests revealed massive proteinuria (11.9 g/24 h) and laboratory findings of kidney failure: serum creatinine 4.0 mg/dl and glomerular filtration rate 21 mL/min./1.73m^2. There was also leukocytosis (WBC 26.1 × 10^3/μL) and anemia (Hb 8.44 g/dl) and high serum c-reactive protein (213 mg/dl). The peripheral blood smear showed rare ring-form trophozoites and gametocytes of Plasmodium spp. Abdominal ultrasound showed small and hyperechoic kidneys; chest CT scan showed copious left pleural effusion with partial lung collapse. Renal biopsy revealed chronic proliferative GN with capillary wall thickening producing a double contour, segmental sclerosis and tubular atrophy. On immunofluorescence, granular deposits of IgA and C3 were observed on mesangium. The patient was treated with atovaquone/proguanil for 3 days; pleural effusion required the placement of pleural drainage for 3 weeks. Actually he is inserted in a dialysis/kidney transplant.
Conclusions: This case highlights the importance of obtaining remote travel histories from immigrants presenting with nephrotic syndrome especially for the current immigration crisis in Europe. Malaria has low prevalences or is slightly known in our continent and requires more medical attention by European doctors.

P-299 COLLAPSING FOCAL SEGMENTAL GLOMERU LOSCLEROSIS IN THE PAEDIATRIC COHORT: A SINGLE CENTERS EXPERIENCE

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Introduction: The collapsing variant of focal segmental glomerulosclerosis (FSGS) has been associated with poor treatment response with rapid progression to renal failure. There is relative paucity of knowledge on the clinical outcome of idiopathic collapsing FSGS in the paediatric population. We describe the units experience with 4 cases of idiopathic collapsing FSGS in the past 6 years.
Material and methods: The case records of 4 paediatric patients with histological diagnosis of collapsing FSGS were reviewed with regards to patient demographics, clinical presentation, treatment, clinical course and eventual renal outcome. The primary endpoint for this descriptive review was the progression to end stage renal failure (ESRF).
Results: The 4 patients were predominantly males (3:1). Age of presentation varies from 2.3–12.3 years. The degree of proteinuria at presentation ranges from 2.90 g/L to massive proteinuria (13.9–20.7 g/L) and did not have a correlation to renal outcome. At the time of the review, 3 patients had developed ESRF. All patients had a full virology and immunological work-up done and results returned negative. All were started on prednisolone therapy. Two were late non-responders while the remaining two patients demonstrated steroid resistance right from the start. One patient demonstrated extensive tubulointerstitial injury with more than 50% global sclerosis on the initial renal biopsy performed three weeks into the presentation and developed rapid progression to ESRF. All patients were treated with prednisolone in combination with other immunosuppressants (cyclosporine, mycophenolate mofetil or rituximab). Two patients achieved partial remission. The remaining two who presented with elevated serum creatinine at initial diagnosis remained refractory to therapy and progress to ESRF within 3 years from initial presentation (0.08–2.25 years).
Conclusions: An elevated initial serum creatinine, lack of remission and tubulointerstitial involvement have been identified as poor prognostic factors in previous studies. The same observations were made in this case series with a male predominance.

P-300 TACROLIMUS IN THE TREATMENT OF MYH9-RELATED DISORDERS: CASE REPORT

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Introduction: MYH9-related disorders (MYH9RD) belong to a group of inherited diseases caused by mutations of MYH9 gene, which encodes non-muscle myosin heavy chain IIA. The aim of the study is to report a patient with MYH9RD with progressive proteinuria (PU) treated with combination therapy of ACE inhibitors (ACEi), angiotensin receptor blockers (ARB) and Tacrolimus (TAC).
Material and methods: We report a girl with macrothrombocytopenia, Döhle like bodies in leukocytes, deafness, cataracts, hypertension, hematuria and continually increasing PU since 4 years of age reaching nephrotic level with generalized edemas at the age of 15 years. DNA analysis showed 7 known MYH9RD polymorphisms.
Results: Despite therapy with ACEi (Ramipril 0.1 mg/kg/d) and ARB (Losartan 1.0 mg/kg/d) since the age of 10 years, PU has been worsening to nephrotic range (5.4 g/24 h). The therapy with TAC has been started (TAC through level 2.8–4.0 μg/l) and has led to partial remission. After 3 month of combination therapy with TAC, ACEi and ARB, the ACEi was reduced (0.05 mg/kg/d) due to hypotension. This has led to a relapse of PU (6 g/24 h), which prompted re-increase of ACEi to the initial dosing (0.1 mg/kg/d); this combination therapy has then improved PU to 2.1 ± 0.59 g/24 h. The slopes of PU were significantly different when analyzed before TAC, on TAC + ACEi + ARB before PU relapse and on TAC + ACEi + ARB after PU relapse (+0.03,-0.74, −0.54 g/24 h/month; respectively, $p = 0.019$) suggesting a favorable effect of combination therapy with TAC and higher dose of ACEi and ARB. At the same time periods, the slopes of eGFR were not significantly different (−0.001, 0.037, 0.026 ml/s/1.73 m2/month; respectively, $p = 0.75$).
Conclusions: The combination therapy with ACEi, ARB and TAC may lead to a significant improvement of proteinuria while preserving renal function in children with MYH9RD. We hypothesize that the therapy is effective not only via its hemodynamic changes, but also due to the TAC interaction with synaptopodin.

P-301 UNEXPECTEDLY HYPOTHYROIDISM POST BILATERAL NEPHRECTOMIES IN A PATIENT WITH CONGENITAL NEPHROTIC SYNDROME

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Introduction: To describe the case of an infant with congenital nephrotic syndrome who was unexpectedly found to have congenital hypothyroidism after thyroxine supplements were stopped post bilateral nephrectomy.
Material and methods: Case report.

Results: An infant with Finnish Type congenital nephrotic syndrome (CNS) was started on thyroxine after having biochemical hypothyroidism detected following routine investigations for CNS. His CNS was managed conservatively with regular albumin infusions and IV furosemide to reduce the oedema, anticoagulation, immunoglobin infusions, antibiotic prophylaxis, ace inhibitors and later indomethocin to reduce his GFR. He went on to have bilateral nephrectomise by 2 years and 10 months of age. His hypothyroidism was expected to resolve following the reduction in urinary leak of thyroid binding proteins after bilateral nephrectomise, however he became severely biochemically hypothyroid (TSH >500 mU/L, fT4 < 5.1 pmol/L) after stopping the levothyroxine and he was later confirmed to have thyroid dysgenesis. Patients with nephrotic syndrome (NS) can become hypothyroid due to urinary losses of thyroid binding proteins and our patient was mistakenly thought to be hypothyroid for this reason. He was quickly restarted on thyroxine and his thyroid function tests stabilised. At the age of 4 years he went on to have a renal transplantation and remains well on his thyroxine replacement.
Conclusions: This case illustrates the importance of monitoring thyroid function post nephrectomies to ensure there are no other unusual causes for their hypothyroidism.

P-302 GIRLS HAVE HIGHER RISK FOR PRIMARY STEROID RESISTANT NEPHROTIC SYNDORME - NEPHROSIS ONLINE STUDY

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Introduction: The annual incidence of idiopathic nephrotic syndrome (INS) in Europe ranges from 2 to 7 per 100,000 children**.** Male predominance among young children and rare onset after 10 years age has been historically reported. Almost 80% of children achieve remission following steroid treatment**.** The aim of the presented study was to assess the contemporary demographics and initial response to steroids in a Caucasian cohort of INS.
Material and methods: Data of 500 Caucasian children with 1st episode of NS was collected prospectively from 17 Pediatric Nephrology centers through a web based platform – Nephrosis Online (NOL). Standard steroid therapy was administered to 479 children with INS and steroid-responsiveness was assessed according to KDIGO Guidelines.
Results: Mean age at onset was 5.14 years +/− 3.7 years, median age: 3,87 years Nearly equal distribution between genders was noted (M:269,F:210 = 1.28:1). 429/479 children (89.5%) responded to steroids (SSNS). 50/479 (10.4%) were steroidresistant (SRNS) after 8wks of therapy. Mean age at onset of SSNS was 4.8 ± 3.4 years, with majority (91%) younger than 10 years Only a slight predominance of boys was observed (M:250 vs. F:179 = 1.4:1). Among SSNS children girls were slightly older (mean age for M: 4.6 vs. F: 5.2 years, $p = 0.039$).Mean age at onset of SRNS was significantly higher (6.9 ± 4.8 years), median age: 6.15 years 70.% were younger than 10 years Predominance of girls was present in the SRNS subgroup (M:19, F:31 = 1:1.63). Age at onset of SRNS was independent of sex (F: 7.1 years vs. M:7.2 years).
Conclusions: In contrast to previous publications an almost equal distribution of both sexes is observed in a cohort of Caucasian children with INS. In this cohort female sex is associated with an increased risk of initial steroid resistance.

P-303 GROWTH PATTERN OF CHILDREN WITH IDIOPATHIC NEPHROTIC SYNDROME (INS): A STUDY OF 41 PATIENTS

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Introduction:
Objective: To evaluate the growth pattern in steroid responsive nephrotic syndrome (SRNS) Tunisian children.
Material and methods: We realized a retrospective analysis of the growth pattern of the children with SRNS followed in the department of pediatrics at the University hospital of sahloul (Sousse, Tunisia) during period of 16 years.
Results: Forty-one patients were included, 29 boys and 12 girls. The mean age at onset was 4.3 ± 2.3 years. According to patients initial response to steroids there were 16 cases of SRNS with infrequent relapses, 5 frequently relapsing SRNS and 20 cases of steroid dependent nephrotic syndrome. The threshold of steroid dependence was equal to 39 ± 20.6. mg/m^2. The mean number of relapses was 4.3 ± 4,06. The use of immunosuppressive medications was necessary in 14 cases (34.1%), after an average time of 36.28 ± 31.35 months after the disease onset. A steroid sparing agent was introduced because of a slowing down in the growth speed in 5 cases. At the beginning of follow up (first year), the mean height standard deviation (SD) was equal to 0.03 ± 1.38 SD, the growth speed was of 5.9 ± 3.2 cm/year. We noticed a slowing down from the 5th year of follow up, to end to a mean height SD equal to – 1.5 ± 2.35 SD with a growth speed equal to 3.5 ± 2.38 cm/year in the 10 th year. The mean follow up duration was 5.1 ± 3 years.
Conclusions: Growth impairment in SRNS is a severe complication that needs an early detection and an early use of steroid sparing agents.

P-304 EFFICIENCY OF CYTOSTATIC THERAPY FOR RELAPSING AND FREQUENTLY RELAPSING STEROID DEPENDENT NEPHROTIC SYNDROME (SDNS) IN CHILDREN

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Introduction: The aim of the study was to assess the efficiency of cytostatic therapy in children with SDNS.
Material and methods: Out of total 168 children with initial Steroid-Senstive NS, 70 (41.7%) became relapsing and frequently relapsing with steroid dependence and toxicity. The mean age of SSNS diagnosis was 3.6 ± 0.7 years, with a male/female ratio of 2:1. We analysed frequency of relapses in children with SDNS during 2 years after cytostatic therapy.
Results: All 70 children with SDNS had increased specific IgE to alimentary allergens (100%), 47 (67.1%) - to dust allergens, grasses and trees pollen allergens, 41 (58.6%) children had clinical manifistations of allergy. Out of 70 children with SDNS, 54 were treated with steroid-sparing agents: 28 - Clorambucil 0.15–0,3 mg/kg/day during 2–3 months, 17 - Cyclosporin A (CsA) 2.5–5 mg/kg/day (including 3 after Clorambucil) and 21 - Mycophenolate mofetil (MMF) (including 3 after Clorambucil and CsA; 6 after CsA; 3 after Clorambucil) during 3–6 months. Out of 28 children treated with Clorambucil, 17 (60.7%) had relapses; out of 17 children treated with CsA, 15 (88.2%) had relapses; out of 21 children treated with MMF, 10 (47.6%) had relapses during 2 years after cytostatic therapy. Out of 54 children 24 (44.4%) had no relapses of SDNS during 2 years after cytostatic therapy.

Conclusions: Children with increased specific IgE to allergens and clinical manifistations of allergy had subsequent relapses of SDNS after cytostatic therapy with Clorambucil, CsA, MMF.

P-305 TOXOPLASMOSIS AND INFNATILE NEPHROTIC SYNDROME

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Introduction: Toxoplasmosis is a cosmopolitan parasitic zoonosis caused by an intracellular protozoan, Toxoplasma (T.) gondii. Usually benign, but potentially severe to the fetus and the immunocompromised subject. In the immunocompromised patient, toxoplasmosis is a severe infection with severe consequences. This is most often the reactivation of an old toxoplasmosis. It can be disseminated or localized.
Material and methods: We report the case of a child followed for transfusional homozygous B-thalassemia, who presented at the age of 03 a nephrotic syndrome with febrile convulsions following the prescription of bolus of solumédrol,
Results: The exploration revealed a positive toxoplasmic serology Confirmed by a PCR positive in the blood, concluded to a disseminated reactivation of a toxoplasmosis; treated by antibiotics.
Conclusions: Disseminated toxoplasmosis is fatal without treatment. The fever is constant, isolated at first. Secondarily, an alteration of general condition and visceral involvement Multiple. Among the localized forms, toxoplasmosis Cerebral is the most frequent.

P-306 EFFICACY OF RITUXIMAB IN A CASE OF PRIMARY MEMBRANOUS NEPHROPATHY

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Introduction: Objectives: To present the efficacy of Rituximab in a case of primary membranous nephropathy, refractory to usual treatment protocols.
Material and methods: Methods: Use of Rituximab in a single dose of 375 mg/m2.
Results: Results: A 12 years - old girl, presented with severe edema, proteinuria (10 g/m2/24 h), hypoalbuminemia (albumin 1.53 g/dl) and hyperlipidemia, with normal renal function. Nephrotic syndrome revealed steroid resistance and renal biopsy showed membranous nephropathy (MN). Investigation for an underlying systemic disease or infection was negative. Treatment with cyclosporine in a dose of 4 mg/kg/day in combination with prednisone on alternate days and lisinopril for 12 weeks failed as nephrotic range proteinuria persisted and the patient continued to be dependent to human albumin infusions. Therefore, alteration of treatment approach was decided into a combination of cyclophosphamide, prednisone on alternate day and lisinopril for 9 weeks with no clinical or laboratory response. After an overall period of treatment for 28 weeks the patient still presented heavy proteinuria and hypoalbuminemia and we decided to use rituximab. Rituximab was given to a single dose of 375 mg/m2 in combination with cyclosporine, lisinopril and prednisone on alternate day. One week later the number of CD20 B lymphocytes was reduced to zero. Proteinuria gradually decreased and complete remission was achieved in a period of 2 months after rituximab infusion. Today 12 months after the single rituximab infusion the number of CD 20 B lymphocytes remains zero and the patient is still in complete remission.
Conclusions: In our case Rituximab was more effective to other treatment protocols to achieve initial remission. However it is still not clear if a combination of cyclosporine and minor steroids dose is a sufficient treatment to keep patient clear of proteinuria or the increase of B-cells will be accompanied by relapse of proteinuria indicating that patient is "rituximab depended".

P-307 PATTERNS OF STEROID SENSITIVE NEPHROTIC SYNDROME (SSNS) IN CHILDREN IN TUNISIA CHILDREN: A SINGLE CENTER EXPERIENCE

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Introduction:
Objective: To describe the clinical patterns and outcome of .steroid sensitive syndrome in Tunisian children.
Material and methods: We retrospectively investigated clinical characteristics of children diagnosed with SSNS in the department of pediatrics in the university hospital of Sahloul –Sousse (Tunisia) over a period of 16 years.
Results: We collected 41 cases (29 boys and 12 girls). A family history of consanguinity and nephrotic syndrome were found respectively in 24.4% and 14.6%. The mean age at onset was of 4.3 ± 2.3 years. The mean time to remission was 19.2 ± 10.1 days. There were 16 cases of SSNS with infrequent relapses, 5 frequently relapsing SSNS and 20 cases of steroid dependent nephrotic syndrome. The mean time to first relapse was 7.2 ± 9.7 months, with an average number of relapses of 4.3 ± 4. No predictive factor of relapse was found. Fourteen patients required immunosuppressive agents 36.28 ± 31.3 months after the first outbreak. Mycophenolate (MMF). and cyclophosphamide, were used in 7 cases each. Five out of seven patients on MMF achieved complete remission with relapses, and 2 achieved complete remission without relapses, whereas patients on cyclophosphamide experienced therapeutic failure in 3 cases, complete remission with relapses in 2 cases and partial remission in 2 cases. The mean follow up duration was 5.1 ± 3 years. Steroid related complications were present in 10 patients: hypertension in 2 cases, a cataract in one case, osteoporosis in one case, skin stretch marks in one case and growth retardation in 5 cases.
Conclusions: Short as well as long term outcomes of SSNS are mainly related to steroid response and cumulative dose.

P-308 EXPERIENCE OF SPONTANEOUS BACTERIAL PERITONITIS IN CHILDREN WITH NEPHROTIC SYNDROME

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Introduction: Spontaneous bacterial peritonitis (SBP) is defined as a bacterial ascitic fluid infection without intra-abdominal surgically treatable source. The incidence of peritonitis has been reported that 1.4–16% in children with NS. We aim to report four cases of primary peritonitis in children with NS and describe the clinical and laboratory findings.
Material and methods: Total of 4 children, diagnosed with peritonitis, secondary to NS, in Izmir Tepecik Research Hospital between March 2014 and November 2016 were analyzed retrospectively in this study. Epidemiological, clinical, and laboratory characteristics of patients were also evaluated.
Results: All patients were male and median age was 7. Peritonitis and NS were seen together at all of the patients but none of them have presented with peritonitis symptoms of the first episode of NS. Two patients were steroid dependent, and others were steroid sensitive. All patients presented with abdominal pain and tenderness. Only one patient showed

vomiting and fever was monitored in two patients. During the admission period, severe hypoalbuminemia and proteinuria was determined in all of cases. In three cases, diagnostic paracentesis could not be undergone because their parents didn't allow for this process. In one case, diagnostic paracentesis performed. However, fluid culture was founded to be sterile. In one patient, *Streptococcus pneumonia* was isolated in blood culture. We treated patients empirically with ceftazidime and ceftriaxone. All patients showed regression in findings of peritonitis on the second and/or third day of the treatment.

Conclusions: In conclusion, nephrotic patients presented with abdominal pain and tenderness should be considered as peritonitis.

P-309 COMPARISON OF PAIN EXPERIENCED DURING URINE COLLECTION WITH TRANSURETHRAL CATHETERIZATION OR URINE BAG COLLECTION IN NON-TOILET-TRAINED CHILDREN: A NON-INFERIORITY STUDY

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Introduction: Urine collection from non-toilet-trained children is a daily challenge in pediatric emergency departments to diagnose urinary tract infections (UTI). Guidelines recommend to use transurethral catheterization (TUC) to obtain uncontaminated urine samples. As TUC is often considered as a painful procedure, non-invasive urine bag collection (UBC) remains widely used despite its high contamination risk.The aim of this study was to demonstrate that TUC is not significantly more painful than urine bag removal.

Material and methods: A prospective, observational, monocentric, non-inferiority study was conducted. Children aged 0 to 3 years, non-toilet-trained and presenting at the pediatric emergency department with a suspicion of UTI were eligible. Pain and experience of the procedure during transurethral catheterization or urine bag removal were assessed independently by nurses and parents using a questionnaire. The primary outcome was the mean pain score as assessed by the nurse using a visual analog scale (VAS, range of score: 0–10). A non-inferiority margin of 2 points on the VAS was chosen.

Results: One hundred patients were included (50 per group). Mean age was 10.2 months ±8.2. The mean VAS score assessed by nurses was 1.9 ± 2.1 in the TUC group and 0.9 ± 1.5 in the UBC group. The difference of VAS between the groups was 1.0 point with an upper limit of the unilateral 95% confidence interval of 1.6. The pain assessed by nurses during catheterization was not significantly superior to the pain during urine bag removal. In the TUC group 84.0% of parents and 77.6% of nurses reported that the procedure went well or very well.

Conclusions: Transurethral catheterization is not more painful than bag's withdrawal, with a good parental and nurses' experience of the procedure. This should encourage health professionals to follow the actual guidelines recommending to use TUC whose bacteriological superiority is well established.

P-310 FACTORS INFLUENCING DAYTIME AND COMBINED INCONTINENCE IN CHILDREN WITH CEREBRAL PALSY

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Introduction: Incontinence is a major issue in children with cerebral palsy (CP) with important effects on quality of life, financial costs and risk for upper urinary tract dysfunction. This study aims to identify which factors increase the risk of daytime or combined urinary incontinence (UI) in children with CP.

Material and methods: A cross-sectional case-control study was conducted including children with CP with or without UI. Explanatory variables were subdivided in three clusters, namely demographic and general medical data, CP classification and bladder and bowel dysfunction. Data was obtained using uroflowmetry with EMG testing, a questionnaire and bladder diaries. Univariate and multivariate analysis was performed for variables and clusters respectively. A final associative logistic model including all clusters was developed.

Results: The study included 34 incontinent children and 45 continent children. UI was associated with intellectual disability (OR 7.69), swallowing problems (OR 15.11), the use of external aids (OR 27.50) and the use of laxatives (OR 13.31). UI was positively associated with dyskinesia (OR 5.67) or combined spasticity and dystonia (OR 4.78), bilateral involvement (OR 4.25), gross motor function classification system level IV (OR 10.63) and V (OR 34.00) and severe impairment in manual (OR 24.27) or communication skills (OR 14.38). Lower maximum voided volume (OR 0.97) and oral fluid intake (OR 0.96) influenced UI negatively. Pathological uroflow curves were not significantly associated with incontinence. The final model defined functional impairment, intellectual disability and oral fluid intake as predictive factors for UI.

Conclusions: Risk analysis revealed functional impairment, intellectual disability and fluid intake as important factors influencing continence in a child with CP. Clinicians and caregivers should be alert to both bladder dysfunction and additional risk factors and adapt environment and treatment strategies to minimize the impact of these factors and enhance the probability of achieving continence.

P-311 SHOULD VITAMIN D PROPHYLAXIS BE CEASED IN CHILDREN WITH UROLITHIASIS?

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Introduction: Vitamin D prophylaxis, 400 units/day, is recommended for all infants, rather up to 18 years old. This prophylaxis is usually ceased in children with urolithiasis, although there is no evidence. The aim of the study was to determine whether the cessation of vitamin D prophylaxis effects on stone size in children under age of 3 with urolithiasis.

Material and methods: Vitamin D prophylaxis was ceased during the first 3 months of the study and was administered again during the following 3 months. The number and the size of the stones were evaluated by ultrasound at the end of both periods with and without vitamin D prophylaxis.

Results: Study group consisted of 29 children (12 F, 17 M). Mean age was 15 ± 12 months (0–36 months). Diameter of the kidney stones were 1.2–11 mm and 18 patients had multiple stones at the time of diagnosis. Kidney stone size was decreased in 16 (55.2%) patients, increased in 6 (20.6%), not changed in 7 (24.2%) at the end of the period without vitamin D prophylaxis. The number of stones decreased in 13 (44.8%) children, increased in 1 (3.5%), not changed in 15 (51.7%). The patients received 400 IU per day vitamin D during the following 3 months. Kidney stone size was decreased in 16 (55.2%) patients, increased in 5 (17.2%), not changed in 8 (27.6%) at the end of the period with vitamin D prophylaxis. The number of stones decreased in 11 (38%) children, increased in 2 (6.8%), not changed in 16 (55.2%). There was no significant difference between the periods of with and without vitamin D in terms of the number and size of kidney stones ($p > 0.05$).

Conclusions: Our results suggest that cessation of vitamin D prophylaxis is not necessary for infants with urolithiasis.

P-312 A RETROSPECTIVE CASE SERIES 1996–2014 OF SIMPLE RENAL CYSTS IN CHILDREN IN A TERTIARY PAEDIATRIC CENTRE

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Introduction: Simple renal cysts are increasingly identified on imaging. We describe their natural history in this case series and propose a clinical and imaging follow up regimen.
Material and methods: We performed an ethically approved, retrospective case series of children who had one or two simple renal cysts identified on ultrasound (USS) from 1996 to 2014 and who had at least one subsequent USS in our institution. Cases were identified electronically, USS reports were manually read and clinical data extracted from electronic records. Exclusion criteria included; multicystic dysplastic kidney, autosomal dominant polycystic kidney disease (ADPKD), cystic dysplastic kidneys, more than two cysts, hydronephrosis, tuberous sclerosis and kidney transplant.
Results: 88 patients were identified, 35 (39.8%) were male. Median age at first USS 8 years (range 10 days-21 years). 79 (89.8%) patients presented with 1 cyst, 9 (10.2%) patients with 2 cysts. Overall median follow up was 2.6 years (36 days-11.8 years). Cysts resolved (definition; not seen on 2 sequential USS) in 10 patients (11.4%). A further 10 patients (11.4%) had no cyst on 1 repeat USS, but had no subsequent scans. Ten patients (11.4%) developed further cysts, median time 3.2 years (range 35-days-5 years). Two patients (2.2%) were diagnosed with ADPKD. Nine patients (10%) developed morphological changes; 7 septations, 1 haemorrhage, 1 complex. Median time 1.5 years (range 50 days to 4.8 years). Three patients developed calcification, median time 1.3 years (range 1–1.3 years). Two patients (2%) were diagnosed with hypertension. Twenty-three patients (26.1%) had an albumin creatinine ratio (ACR) available. Median ACR 3.25 mg/mmol (range 0.3–118.5). Four patients' (4%) ACR >30 mg/mmol.
Conclusions: Simple renal cysts spontaneously resolve in 10–20% of cases. A proportion develop further cysts, morphological change, hypertension and proteinuria. Twenty-five patients (28.4%) developed one or more complications. We recommend at least 3 ultrasound scans and 5 year follow up with regular blood pressure and ACR measurements.

P-313 IMBALANCE IN URINARY PROTEOGLYCANS AND THE INSULIN GROWTH FACTOR AXIS IN CHILDREN WITH NEPHROLITHIASIS

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Introduction: In vitro experiments have shown that exposure of renal epithelial cells to calcium oxalate crystals leads to synthesis of proteins involved in extracellular matrix (ECM) production. Based on the role of ECM in the Randall plaque formation, we aimed to identify and quantify ECM proteins in the urine of children with nephrolithiasis (RS) using a proteomic approach.
Material and methods: Prospective, controlled, pilot study of pooled urine from RS ($N = 30$, 24 females, mean age 12.95 ± 4.03 years) versus age- and gender-matched healthy controls (HC), using mass spectrometry. Relative protein abundance was estimated using spectral counting. The criteria for protein selection were: 1) ≥5 spectral counts; 2) ≥2-fold difference in spectral counts; and 3) ≤0.05 p-value for the Fisher's Test. Results were confirmed by ELISA.
Results: We found 36 (15.7%) ECM proteins out of 229 that met the above criteria. Significant differences between RS and HC were found among two proteoglycans and four insulin growth factor (IGF) proteins (Table). Significant increase in the urinary excretion of IGFBP4 in RS (37.00 ± 37.68 ng/mg creatinine) versus HC (19.04 ± 20.32 ng/mg creatinine) was confirmed by ELISA ($p = 0.049$). Statistically significant correlation was found between urinary IGFBP4 concentration and 24-h urinary calcium excretion ($p < 0.001$).

Accession Number	Protein	Assigned peptides	Ratio (Patient/ Control)
PRG4	Proteoglycan 4	38 (5)	7.6*
SDC1	Heparan sulfate proteoglycan (Syndecan 1)	5 (50)	0.1*
IGF2R	Cation-independent mannose-6-phosphate receptor	2 (13)	0.15*
IGFBP1	Insulin-like growth factor-binding protein 1	8 (0)	Unique
IGFBP4	Insulin-like growth factor-binding protein 4	17 (4)	4.2*
IGFBP6	Insulin-like growth factor-binding protein 6	49 (2)	24.5*

*$P < 0.01$.
Conclusions: Alteration in proteoglycans and the IGF axis appears to have a significant role in the mechanism of nephrolithiasis, likely by modulating ECM biosynthesis. Further understanding of their roles in nephrolithiasis may aid in generation of novel therapeutic approaches.

P-314 NEPHROCALCINOSIS IN A PORTUGUESE PAEDIATRIC POPULATION

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Introduction: To review the data of children with nephrocalcinosis (NC) concerning aetiology, clinical manifestations, growth and renal function at presentation and outcomes of a paediatric population presented to a nephrology unit in an urban tertiary centre.
Material and methods: The records of consecutive children (0–18 years) with NC were reviewed (January 2008–December 2016). Clinical features, aetiology, treatment and outcomes were retrospectively evaluated.
Results: Thirty five cases of NC were identified, 24 of which alone and 11 associated with nephrolithiasis. The group was constituted mostly of girls (54%). Age at presentation (median 5,7 years) was below 2 years of age in 40%, 31% had between 3 and 9 years and 29% were older than 10 years; mean follow-up was 4,4 years (1–9). The most common presenting symptom was failure to thrive in the first year of life (43%) and flank or abdominal pain (20%); in 17% NC was an incidental finding and in 6% of patients had a systemic syndromatic disease. Renal function at diagnosis was normal in all children. The most frequent causes of NC were hereditary tubulopathies (26%), prematurity and diuretic use (20%) and iatrogenic, namely vitamin D treatment (9%) (one case with hipophosphatemic rickets); in 6 cases there was no identifiable cause. In a logistic regression analysis, sex, age of presentation and familiar history

of NC showed no correlation with NC alone, NC associated with nephrolithiasis or genetic/metabolic aetiology.
Conclusions: Despite the small sample, in this study, we confirmed that genetic and/or metabolic disorders are the main cause for NC and urolithiasis. Associated symptoms and comorbidities, such as growth retardation, intestinal absorption, or bone demineralization, should be evaluated for diagnostic and therapeutic purposes. Preterm infants are a special risk population with high incidence of NC.

P-315 DIFFUSION WEIGHTED MAGNETIC RESONANCE IMAGING IS MORE SENSITIVE THAN DMSA SCINTIGRAPHY IN ACUTE PYELONEPHRITIS IN CHILDREN: A PROSPECTIVE STUDY

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Introduction: To compare static renal scintigraphy (SRS) using ^{99m}Tc-dimercaptosuccinic acid (DMSA) with diffusion-weighted magnetic resonance imaging (DWI-MRI) to detect inflammatory changes in the renal parenchyma of patients with acute pyelonephritis.
Material and methods: Thirthy one (30 girls) aged 3–18 years with acute pyelonephritis were included.. Both SRS (^{99m}Tc-DMSA) and DWI-MRI of the kidneys were performed within five days of diagnosis in all patients to confirm the presence of inflammatory lesions. The DWI-MRI examination was performed without contrast agent or general anesthesia.
Results: DWI-MRI confirmed inflammatory infiltration of the kidney parenchyma in all patients (100%); the impairment was always unilateral. SRS confirmed inflammation in only 22 children (71%). Control examinations were performed in 31 patients after six months with both methods. Scarring was confirmed by DWI-MRI in five and SRS in five patients each.
Conclusions: DWI-MRI imaging had higher sensitivity during the acute stage of acute pyelonephritis and identified inflammatory lesions in the renal parenchyma better than SRS. Moreover, DWI-MRI also provided more precise information about the extent of kidney damage.

P-316 RENAL CYSTS IN THE CONTEXT OF POLYMAL FORMATIVE SYNDROMES: RENAL EVOLUTION

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Introduction: Numerous polymalformative syndromes associate renal cysts but there is no proved evidence about the role of these cysts in the renal prognosis so far. The objective of this study is to review the long-term renal function in patients with syndromes and renal cysts.
Material and methods: A retrospective descriptive study of patients diagnosed with polymalformative syndromes and renal cysts between 2004 and 2016.
Results: We include 14 patients (7 males) with a median age of 9.6 years at the beginning of the study. Eleven patients have a specific syndromic diagnosis: 3 Tuberous sclerosis, 2 Beckwith-Wiedemann syndrome, 2 oro-facial-digital syndrome, 2 Townes-Brocks syndrome, 1 COACH syndrome and 1 Dent syndrome with interventricular communication. The three remaining patients had a polymalformative condition without evidence of specific diagnosis. Both kidney affected were found in 71% of analyzed population, while the rest presented unilateral renal cystic disease. NIne (64%) patients had normal kidneys volumen, 3 (21%) had nephromegaly and 2 (14%) had a reduced kidney size (both present Townes-Brocks syndrome). Three patients (21%) progressed to chronic kidney disease (CKD): 1 patient with oro-facial-digital syndrome at 15 years of age, 1 patient with Townes-Brocks syndrome presented at 1.5 years old CKD associated with proteinuria and 1 patient with COACH syndrome presented at 2 years old CKD with proteinuria and hypertension. A patient with tuberous sclerosis presented *isolated hypertension* with two antihypertensive drugs treatment. The child with Dent syndrome had *isolated proteinuria*. Only one patient with Townes-Brocks syndrome had recurrent urinary infections.
Conclusions: The presence of renal cysts associated with polymalformative syndrome determines a high risk of CKD. There is variability in the age at the onset of *CKD* in patients with the same *syndrome. If* renal involvement is not included in the typical *features* of the *syndrome*, the presence of renal cysts does not indicate a worse prognosis.

P-317 CHARACTERISTICS OF INFANT UROLITHIASIS: A SINGLE CENTER EXPERIENCE IN WESTERN TURKEY

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Introduction: We evaluated the clinical, radiological, metabolic factors and course of UL in Turkish infants. We evaluated the clinical, radiological, metabolic factors and course of UL in Turkish infants.
Material and methods: Medical records of the infants between 1 and 12 months of age with a diagnosis of UL were reviewed retrospectively for gender, gestational age, age at diagnosis, presenting symptoms, past medical history, parental consanguinity, family history of UL, urinary tract abnormalities, urinary tract infections, localization-size-number of stones, course of stones, treatment modality of UL (medical vs surgical) and follow up duration.
Results: Patients were grouped as those with regressing/disappearing or stable stone disease (Group 1) and those with increasing stone size or requiring surgery (Group 2). There were 82 infants with UL with a mean follow-up duration of 29.8 ± 22.8 months (range 7 to 120 months). Two thirds of the patients presented with symptoms/signs not directly related to urinary system. Predisposing factors included metabolic abnormalities in 31 (38%) patients and urinary tract malformations in 11 (13%). Group 1 (n = 55) and Group 2 (n = 27) were not different with respect to the study parameters except higher rate of microlithiasis in Group 1 (49% vs 22%).
Conclusions: In conclusion, as most infants with UL are diagnosed incidentally, awareness of UL in this age group is important for early detection and appropriate management. Every infant with urolithiasis should be evaluated for risk factors as almost half of the patients had predisposing factors. Clinical course is better in infants with microlithiasis.

P-318 PRENATAL ALCOHOL EXPOSURE AFFECTS RENAL FUNCTION IN SCHOOLCHILDREN

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Introduction: Prenatal ethanol exposure was shown to reduce nephron endowment in animal models but the effect of alcohol consumption during human pregnancy on postnatal kidney function has not been explored. We assessed in a prospective longitudinal study a potential association of maternal alcohol consumption during pregnancy with the renal function of the offspring, taking into account potential confounders such as intrauterine growth and the children's current nutritional status.
Material and methods: A random sample of 1093 children from a population-based birth cohort was evaluated. Mother's alcohol consumption during pregnancy was self-reported at baseline. Anthropometrics and estimated glomerular filtration rate (eGFR) were assessed at 7 years of age. The association of gestational alcohol exposure with renal function in childhood was assessed by multiple linear regression analysis, adjusting for gender, current age, birth weight adequacy for gestational age, and maternal age, years of school education, pre-pregnancy nutritional status and smoking during pregnancy.
Results: Thirteen percent of mothers consumed alcohol during pregnancy. At 7 years of age, eGFR was significantly lower in children with prenatal alcohol exposure (134 ± 17 vs.138 ± 16 mL/min/1.73 m^2, $p = 0.014$). The effect was dose dependent and most marked in overweight and obese children, among whom adjusted eGFR was −6.6 (−12.0 to −1.1) ml/min/1.73 m^2 in children exposed to ≤40 g alcohol per week and −11.1 (−21.3 to −1.2) ml/min/1.73 m^2 in those exposed to >40 g per week compared to no consumption (p for trend = 0.002).
Conclusions: Prenatal alcohol exposure has a dose-dependent adverse effect on renal function at school age in overweight and obese children.

P-319 RENAL COMPLICATIONS OF SEASONAL INFLUENZA VIRUS INFECTIONS

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Introduction: Renal complications of seasonal influenza A virus are uncommon and have been reported primarily as single cases or small series of patients. The aim of this study was to determine the incidence of renal complications of seasonal influenza infections in children.
Material and methods: Influenza test findings in nasopharyngeal swabs, which were performed between September 2015 and March 2016 at the virology laboratory of our institution,by determining influenza antigens or using pcr method, were interpreted. The children having positive influenza test results were included in the study. The medical records of these children were evaluated retrospectively.
Results: A total of 568 patients with positive influenza test results have been included in the study. Median age was 54.5 (1–216) months. One hundred fifty-nine patients (%28.0) had an underlying chronic disease (renal, cardiac, pulmonary, haematological, oncological, endocrine, neurological, rheumatological, immunological, metabolic or genetic disease). 41 (7.2%) of them had renal complications.3 patients had acute renal failure, 2 had acute glomerulonephritis. Thirty had proteinuria, 2 had isolated hematuria, and 4 had both hematuria and proteinuria. Median age of the patients with renal complications were 83.9 (7–216) months. Renal complication rate of children with underlying chronic medical condition was higher than that of patients without underlying chronic medical condition (13.8% vs 4.6%, $p = 0.000$, OR: 3.29)] 9 patients have been followed up in intensive care unit; 2 of them had proteinuria as renal complication; one died following sepsis.
Conclusions: Although most of influenza infections are selflimited, especially patients with underlying diseases are at increased risk for renal complications, which should be considered during seasonal epidemics of influenza infections.

P-320 UROLITHIASIS IN INFANTS; IS TREATMENT NECESSARY?

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Introduction: There is little information about treatment strategy of infantile urolithiasis. The main treatment in infantile stone disease is hydration and urine alkalinization with potassium citrate. The aim of this study is to investigate the clinical significance of medical treatment in infants with millimeter-size stones in the urinary system.
Material and methods: The cases of 197 infants (86 girls, 111 boys), who were referred to our department between 2014 and 2016 with urolithiasis (stone size ≤5 mm), were evaluated for metabolic risk factors, and response to medical treatment.
Results: The mean age at diagnosis was 5.2 months (14 days–12 months). The major clinical symptoms were restlessness in 92 children (46.7%) and asymptomatic in 49(24.9%). One hundred sixty six infants (84.3%) had multiple stone in urinary system. Hypercalciuria and hypocitraturia were detected in 17.8 and 15.3%, respectively. Stones measuring ≤3 mm (microlithiasis) were found in 141 infants (71.6%), while 56 infants (28.4%) had stones 3–5 mm. One hundred thirty one infants (66.5%) were able to use treatment regularly and 66 (33.5%) were unable to use treatment. Response to treatment was achieved in 77% of the treated infants. Spontaneous stone resolution was detected 59.1% of the patients who did not use the treatment. The rate of spontaneous stone resolution was higher in infants with microlithiasis ($p < 0.05$).
Conclusions: This is the first study to investigate the clinical consequences of urinary alkalinization in infantile urolithiasis. We conclude that infants with microlithiasis can be followed clinically without treatment because they have a high spontaneous stone resolution rates, and effective treatment with urinary alkalinization can be performed in infants with larger size urinary tract stones.

P-321 IGA NEPHROPATHY AND IGG4 RELATED ORBİTAL DİSEASE: A CHİLD CASE

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Introduction: IgG4-related disease (IgG4-RD) is recognized systemic autoimmune disorder characterized by high levels of serum IgG4 and

infiltration of IgG4-positive plasma cells in multiple organs. Co-existence between IgG4-RD and IgA nephropathy (IgAN) is very rare.Orbital involvement in IgG4-RD includes lacrimal glands, extra-ocular muscles, trigeminal nerve and other parts of the orbit.Herein we presenta child with IgAN and IgG4-RD with orbital findings.
Material and methods: CASE REPORT. A 10 year old female was admitted with fever, macroscopic hematuria (4 weeks),proteinuria (15 mg/m2/h)and decreasing GFR (59 ml/sc/1.73 m2).NormalC3,C4,ANA,Anti-ds-DNAand elevated serum IgA (284 mg/dl) were found in laboratory.Renal biopsy was done for prolonged hematuria and showed positive segmental granular IgA+, IgM++ staining and minimal mesenchymal cell proliferation,fibro-necrotizing and segmental sclerosing (5/25 glomerulus) and focal chronic tubulointerstitial inflammation.Improvement in renal function, macroscopic hematuria and proteinuria were observed in 2 weeks.The case was diagnosed as IgAN and ramipril was started.Two years later,patient admitted again with swelling redness, ptosis on left eye lid.MRI revealed a supero temporal orbital mass (29x11x31mm) extending to lacrimal gland.Incisional biopsy of the mass revealed lympho-plasmocytic infiltration and fibrolipomatosis.With the existing findings IgG4-RD was suspected and elevated serum IgG (1910 mg/dl), IgG4 (137 mg dl),acute phase reactants supported this diagnosis.Proteinuria (4.5 mg/mg creatinine)was also present.
Results: Systemic steroid (40 mg/day) and mycophenolate mofetil (500 mg/day) were started.The orbital mass regressed rapidly,elevated acute phase reactants returned to normal,but proteinuria resisted.
Conclusions: IgG4-RD is rare in adults,and very rare in childhood.To the best of our knowledge this is the first report on coexisting IgAN and IgG4-RD in childhood.

P-322 PERSISTENT ISOLATED MIROHEMATURIA IN CHILDREN: IS RENAL BIOPSY REALY UNNECESSARY?

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Introduction: Isolated microhematuria (IMH) is frequently encountered in pediatric practice. No definite conclusions have been reached about the natural history of patients with IMH. There is controversy too, whether besides other examinations there is need of performing renal biopsy in children with persistent IMH. The aim of the study was to evaluate the contribution of renal biopsy to the diagnosis of the diasease in IMH and prognosis of such patients.
Material and methods: Renal biopsy was performed in 92 children with IMH (46 boys and 46 girls with mean age of 9.44 and 9.74 years respectively) in whom urological abnormalities, hypercalciuria, systemic diseases, coagulopathy or overt family history of renal disease were excluded. The mean duration of IMH prior to biopsy was 2 years and 5 months. Biopsy speciments were examined by light (LM), immunofluorescent (IF) and electron microscopy (EM). After biopsy the patients were followed-up for 3–13 years.
Results: Seventy-seven (83.6%) children had pathological histologic finding. The most common were EM changes consistent with Alport syndrome found in 27 (35.0%) cases, followed with IgA nephropathy in 18 (23.4%) cases and changes consistent with acute postinfectious glomerulonephritis in resolution in 15 (19.5%) cases. Diffuse thinning of GBM was found in 13 (16.9%) cases, membranoproliferative glomerulonephritis in 3 (3.9%) and fibrillary glomerulonephritis in 1 (1.3%) case. On follow-up, in 7 of 27 children with EM changes consistent with Alport syndrome appeared proteinuria and in 2 perceptive hearing impairment. Further surveillance is needed to confirm the significance of EM findings in others.
Conclusions: In children with IMH of proven glomerular origin renal bioipsy is justified and specimen should always be analyzed by light, immunofluorescent and electron microscopy.

P-323 IGA NEPHROPATHY IN CHILDREN IN THE CZECH REGISTRY OF RENAL BIOPSIES (CRRB)

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Introduction: To analyze cases of IgA nephropathy (IgAGN) in the CRRB. The CRRB includes data from 2197 renal biopsies (RBs), performed between 1994 and 2012, in children and adolescents aged ≤18 years. CRRB currently contains data of native kidneys in the Czech Republic of 11 paediatric centers performing practically all RBs in children.
Material and methods: These data are already available for 540 RBs conducted in the 2007–2012 period.
Results: IgAGN, is the most common diagnosis, was found in 92 (17%) RBs. The mean age of IgAGN patients (73.9% of boys) was 13.7 ± 3.7 years (2.5 months–18 yrs), 2% were <5 years, 11.9% were 5–10 years, 30.8% were 10–15 years, and 55.2% were ≤18 years old. At the time of RB, 59.7% of children with IgAGN had microscopic hematuria and 36% gross hematuria. Proteinuria 1–3 g/24 h was present in 48.9% of children and nephrotic proteinuria in 13.1%, with 1.2% of children meeting the criteria of nephrotic syndrome. Most RBs were classified as IgAGN (67.6%), with only a minority further characterized as: mesangioproliferative GN in 19.7%, with membranoproliferative GN features in 1.2%, with crescents in 1.8%, with sclerotization in 4.3%, and as a combination of several of the above types in 5.3%. At the time of RB in the 93 children with IgAGN 18.5% had hypertension (HT), with 15.2% of these treated for HT. Those with HT had lower glomerular filtration rate (GFR) (Schwartz formula, 1.47 ± 0.72 vs. normotensive 1.62 ± 0.52 ml/s/1.73 m2; $p < 0.05$) and significantly higher proteinuria (89.92 ± 113.52 vs. normotensive 33.13 ± 50.46 mg/m2/h; $p < 0.01$).
Conclusions: IgAGN is the most common diagnosis in the CRRB. HT is associated with known risk factors for progression of biopsy-proven GN such as GFR or proteinuria. CRRB provides us with important information about the epidemiology of glomerulonephritis in our region, and represents a basis for cooperation in this field.

P-324 IMPROVED SLEEP QUALITY FOLLOWING ADENOTONSILLECTOMY IS ASSOCIATED WITH ENURESIS RESOLUTION IN CHILDREN WITH SLEEP-DISORDERED BREATHING

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Introduction: We have previously reported that adenotonsillectomy (TA) leads to complete resolution of nocturnal enuresis (NE) in about 50% of children with sleep-disordered breathing (SDB), but the mechanism is not entirely clear. In this study we assessed the effect of TA on sleep quality, night time urinary volume (NUV) and secretion of antidiuretic hormone (ADH) and brain natriuretic peptide (BNP) in children with NE and SDB.

Material and methods: Prospective pilot study of 41 children 5–18 years of age diagnosed with SDB (snoring, $N = 17$, and obstructive sleep apnea syndrome, $N = 24$) on polysomnography, and monosymptomatic primary NE requiring TA for upper airway obstruction release. Arousal score, nocturia, NUV, and plasma levels of ADH and BNP were measured pre and 1 month post-surgery.

Results: Decrease in arousal score and plasma BNP level, and increase in plasma ADH level were seen in all patients post-surgery. However, mixed ANOVA showed that responders (dry) had significantly more improvement than non-responders (wet) in the quality of sleep (Table). Following TA, nearly all dry children reported nocturia and significant decrease in BNP levels ($P = 0.017$) without significant change in their NUV.

	Dry ($N = 20$)	**Wet ($N = 21$)**	***P*-value**		
	Pre-T&A	Post-T&A	Pre-T&A	Post-T&A	
Heavy sleeper Yes (%)	10 (50%)	2 (11.8%)	13 (27.8%)	16 (88.9%)	0.020
Snoring Yes (%)	20 (100%)	3 (15.8%)	20 (100%)	7 (35.0%)	0.001
Arousal score	4.92 ± 0.35	2.38 ± 0.29	4.92 ± 0.34	3.57 ± 0.28	0.015
NUV (ml)	279.56 ± 128.83	340.13 ± 190.36	385.05 ± 151.56	352.65 ± 161.26	0.072
ADH (pg/ml)	7.28 ± 6.51	8.00 ± 8.48	4.55 ± 6.36	6.59 ± 6.30	0.297
BNP (ng/ml)	26.49 ± 28.83	16.48 ± 30.87	38.89 ± 32.49	28.33 ± 26.11	0.017

Conclusions: Conclusions: Change in children's quality of sleep and arousal score was associated with NE resolution post-TA. Improvement in sleep quality appears to be responsible for the effect of TA on NE in children with SDB.

P-325 HUS INDUCED CARDIAC FAILURE IS REVERSIBLE USING CARDIOPULMONARY BYPASS AS RESCUE

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Introduction: The extra-renal involvement in hemolytic uremic syndrome (HUS) includes gastrointestinal, pancreatic, hepatic, neurological and cardiac symptoms. The mortality in HUS patients of 3–5% is primarily attributed to complications related to CNS and the heart. In the case presented here, we illustrate that severe cardiac involvement in a patient with HUS is potentially reversible using cardiopulmonary bypass as rescue.
Material and methods: Data are collected from the boy's medical journal.
Results: A 12 years old boy was diagnosed with EHEC induced HUS related to *E. coli type 2A*. The patient developed anuria, hypertension of 150/105 mmHg, and had neurological symptoms with lethargy, confusion and later a tonic-clonic seizure successfully treated with midazolam. Blood samples showed renal insufficiency with a creatinine of 3.98 mg/dL, thrombocytopenia of $47*10^9$/L, LDH of 3620 IU/L, low haptoglobin <20 mg/dL, anemia of 10.0 g/dL and schistocytes were seen on blood smears. Peritoneal dialysis was initiated without complications. Serum potassium was normal. At day 3, the patient had cardiac arrest twice. Troponin-T, creatine kinase, and creatine kinase-MB were significantly increased. The second episode of cardiac arrest was irreversible to advanced CPR and a cardiopulmonary bypass circuit was established to provide cardiac output. Declining cardiac pump function to a near non-contractile state with an ejection fraction below 10% was observed by echocardiography. This persisted during the following days. However, after seven days on the cardiopulmonary bypass circuit, the myocardium slowly recovered function. Three days later, the cardiopulmonary bypass was successfully discontinued and echocardiography showed near-normal ejection fraction and ECG showed sinus rhythm.
Conclusions: Fatal outcome may be the result of severe cardiac involvement in HUS patients. The present case illustrates the need for intensive supportive care including the use of cardiopulmonary bypass as the cardiac symptoms in HUS patients may be reversible.

P-326 THE EUROPEAN REFERENCE NETWORK FOR RARE KIDNEY DISEASES (ERKNET)

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Introduction:
ERKNet is one of 24 European Reference Networks recently initiated to improve the clinical care of patients with rare and complex diseases throughout the EU.
Material and methods:
The Network is comprised of 38 centers (17 pediatric, 9 adult, 12 combined nephrology units) from 12 European countries which demonstrated, in a stringent accreditation process, a minimum numbers of patients treated per disease group, adequate equipment and infrastructures, and provision of care by multidisciplinary staff according to guidelines and SOPs.
Results:
ERKNet comprises 10 expert workgroups taking care of >40,000 patients with immune ($n = 9900$) and hereditary mediated glomerulopathies ($n = 3300$), hereditary tubulopathies ($n = 2800$), metabolic and stone forming disorders ($n = 1300$), autosomal-dominant renal dysplasias ($n = 7700$), CAKUT and ciliopathies ($n = 7000$), obstructive uropathies ($n = 4600$), thrombotic microangiopathies ($n = 900$), pediatric CKD and dialysis ($n = 3500$), and pediatric transplantation ($n = 2300$). In the reference centers specialized care is provided by a median of 7 nephrology consultants, 4 nephrology trainees, 5 outpatient nurses, 2 dieticians, 1 psychologist, 1 social worker, 1 clinical geneticist and 2 study nurses per center. A median of 250 ABPM profiles, 30 pediatric and 110 adult kidney biopsies per center are performed annually. Genetic diagnostics is performed in most or all patients with suspected hereditary glomerulopathies, tubulopathies, metabolic nephropathies and aHUS in >90% of the centers. The Network plans to: establish a virtual consultation system for challenging cases across the EU; harmonize and optimize quality of care by Network-wide implementation of clinical guidelines and monitoring and benchmarking of core quality and outcome indicators; foster education and

training by CME, Webinar and staff exchange programmes; provide an online repository of patient information; and facilitate clinical research by documenting available patients in a central registry.

Conclusions:

ERKNet will initiate multifaceted activities to improve the access of patients with rare kidney diseases to high-quality healthcare and optimize treatment outcomes throughout Europe.

P-327 AUTOMATED ESTIMATED GFR REPORTING CAN REDUCE THE NEED FOR MEASURED GFR IN ONCOLOGY PATIENTS

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Introduction: Automated estimated glomerular filtration rate(eGFR) reporting is established adult practice and improves detection of chronic kidney disease (CKD). eGFR formula in children commonly require height which is not routinely available when measuring creatinine. We have previously assessed a height independent eGFR (Table 1) and are using this in clinical practice to automatically report eGFR. We reviewed our data to establish whether it could be used to identify patients who did not need a measured GFR (mGFR) in order to determine dosing of chemotherapy."

Table 1; Assessment of our formula and comparison to other height independent eGFR formula

	Mean difference	St Dev	R (Correlation)	% Diff <20%	% Diff <30%
Pottel	27.1	45.8	0.62	47%	62%
Lund-Malmo (Revised)	18.7	23.9	0.57	60%	75%
NCH – BCCH2	14.6	20.0	0.71	71%	83%
EPI-CKD	Previously reported adult formula	83%			
MDRD	Previously reported adult formula	81%			

Material and methods: Data from patients who had been used to locally optimise and validate a height independant formula were assessed to determine cut-offs which would identify those with a mGFR >60 ml/min per 1.73m^{2}and those with a mGFR >80 ml/min per 1.73m^2.

Results: 263 patients (56% male, mean age 9.4 years, range 5 months to 17.9 years) were included in our data analysis. One hundred forty-four patients had an eGFR >90 ml/min per 1.73m^2 of whom 99.3% had a mGFR >60 ml/min per 1.73m^2. Seventy-nine patients had an eGFR >100 ml/min per 1.73m^2, all of whom had a mGFR >80 ml/min per 1.73m^2.

Conclusions: Estimated GFR reporting can accurately identify children with a measured GFR greater than 60 ml/min per 1.73m^2 and greater than 80 ml/min per 1.73m^2. This could reduce the number of children requiring a measured GFR prior to being treated with some chemotherapy agents.

P-328 THE CORRELATION BETWEEN MONOSYMPTOMATIC ENURESIS AND ALLERGIC DISEASES IN CHILDREN

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Introduction:

Aim: The aim is to investigate the correlation between monosymptomatic enuresis (MSE) and allergic diseases (asthma, allergic rhinitis, eczema, and food allergy) in pediatric patients.

Material and methods: The study was conducted on 50 pediatric patients with a MSE clinic who were ≥7 years old and applied to two tertiary health institutions between November 2015 and June 2016. Fifty healthy children in the similar age group, who applied to pediatric outpatient clinics for various reasons, were included as the control group. A questionnaire questioning the presence of food allergy and enuresis in the family and also including the questions of International Study of Asthma and Allergies in Childhood (ISAAC) was distributed to the parents of the children included in the study.

Results: It was found that 52% of 100 children participating in the study were boys and 48% were girls and their mean age was 10.8 ± 2.8 years. While allergic diseases accompanied 34% of the cases with enuresis, this rate was found as 12% in the control group ($p < 0.01$). It was determined that the family history in terms of enuresis and atopy was at a higher rate in the study group (40% and 26%, respectively) and at a lower rate in the control group (2% and 6%, respectively) ($p < 0.01$).

Conclusions: It was observed that allergic diseases were more frequent in the cases with MSE at a statistically significant level compared to the group without enuresis.

P-329 HEMOLYTIC UREMIC SYNDROME OUTBREAK IN ISTANBUL-TURKEY IN 2015; OUTCOME AND ECULIZUMAB EXPERIENCE

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Introduction: An outbreak of hemolytic uremic syndrome (HUS) occured in Istanbul, Turkey between August–December 2015. Epidemiological, demographic and clinical characteristics of the patients and Eculizumab (ECU) treatment from 8 Pediatric Nephrology departments were presented.

Material and methods: Thirty-one children (20 girls, 11 boys) mean age 5.4 ± 4.6 (0.9–14.2) years were included. Demographic, clinical, laboratory data, intensive care unit (ICU) stay, dialysis, blood product and ECU treatments and final renal-hematologic status were recorded retrospectively.

Results: Most of the patients were from the same town [12 patients (38%)]. Stool samples were cultured from 22 (70%) patients and 9 (40%) were positive (5 Enteroaggregative, 4 Enterohemorrhagic *E. coli*). One patient was diarrhea negative but stool was positive for Enteroaggregative *E. coli*. Seven (22%) patients had neurologic manifestation. Nineteen patients had dialysis. Nine patients were treated with ECU with the indications of prolonged anuria, severe hematologic involvement and extra-renal complications. When we compare ECU (+) and ECU (−) dialysis patients, maximum LDH value and duration of ICU were higher ($p = 0.028$, $p = 0.021$, respectively), red blood cell infusion was lower ($p = 0.05$) in ECU (+) dialysis patients (Table-1). There was no difference between groups for proteinuria, hypertension or eGFR in the last control. Mean duration of follow up was 8 ± 3.5 months ($n = 25$). Complete renal recovery was achieved in 70% of the patients, hypertension persisted in two cases (8%), proteinuria persisted in 10 cases (40%), none of the patients were on dialysis at the last control. One patient with multi-organ failure and sepsis had died.

Table-1: Comparison of dialysis patients treated with and without Eculizumab.

	ECU(−) ($n = 11$)	ECU (+) ($n = 8$)	p value
C3, mg/dl	85 ± 15	84 ± 20	NS
Max creatinine, mg/dl	4.7 ± 3.5	4 ± 2	NS
Min Hb, g/dl	5.8 ± 0.9	5.7 ± 0.9	NS
Min platelet, /mm3	38 ± 24	36 ± 15	NS
Max LDH, IU/L	2487 ± 809	4736 ± 3529	0,010
ICU stay, day	7.57 ± 3.3	26 ± 19	0,021
Hospital stay, day	22.2 ± 10.5	36 ± 21	NS
Oligoanuria, day	11.5 ± 6.5	14.4 ± 6.8	NS
Total ES	3 ± 1.8	4.7 ± 3.1	NS
Total TS	0.36 ± 0.5	2.7 ± 3.7	NS
PLT > 150,000/mm3	8.7 ± 4.0	14 ± 3.8	0,024
Hb > 2 g/dl	20.3 ± 20.4	17 ± 7.8	NS
GFR > 15 ml/min/1.73 m2	14.3 ± 11	11.8 ± 7.3	NS

ECU: Eculizumab, ICU: Intensive care unit, ES: erythrocyte transfusion, TS: thrombocyte transfusion, PLT: platelet, Hb: hemoglobin, GFR: glomerular filtration rate.

Conclusions: This outbreak has occurred around 2015 autumn when increased cattle circulation had occurred due to the religious festival called Kurban which could be the source of the *E. coli*. In this outbreak ECU treatment was given to the severe cases. ECU treatment did not change the renal survival but decreased the need for transfusions in the acute phase of the disease.

P-330 CARGLUMIC ACID MAY PREVENT THE NEED OF INVASIVE PROCEDURES IN PATIENTS WITH HYPERAMMONAEMIA DUE TO BRANCHED-CHAIN ORGANIC ACIDAEMIAS

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Introduction: Hyperammonaemia in branched-chain organic acidaemias (BCHOA) is caused by the blocking an activity of the first urea cycle's enzyme. Reducing hyperammonaemia by using extra corporal elimination (EE), along side with natrium benzoate and symptomatic treatment was a standard management of severe metabolic decompensation, especially during the first manifestation of neonatal BCHOA. Nowadays there is a new therapeutic option in a form of carglumic acid, a drug registered for the treatment of hyperammonaemia in N-acetylglutamatsyntase deficiency (NAGS) and in the organic acidurias.

Material and methods: We report the single-centre experiences with treatment of acute hyperammonaemia by carglumic acid in comparison to EE in cohort of 6 children with BCHOA. Two neonates with propionic acidaemia (PA) presented with acute hyperammonaemia above 500 μmol/L in the first week of life were treated with carglumic acid in addition to carnitine, protein-restricted diet and high energy load. Concentration of ammonia significantly reduced after one dose of carglumic acid 100 mg/kg in the first 24 h of treatment and maintained in normal range during the next week. Seven years old girl suffered by the neonatal form of PA, with repeated attacks of hyperammonaemia crises was admitted in severe metabolic acidosis, ketosis and hyperglycaemia. Carglumic acid 200 mg was given because of rapid raise of ammonia (over 4 h with 200 μmol/L) in addition to carnitin, insulin and bicarbonate.

Results: The efficacy of ammonium removal was compared with group of 3 cases treated by peritoneal dialysis.

Conclusions: Carglumic acid reduces hyperammonaemia as effectively as extra corporal detoxication and has fewer side effects. Our data strongly support current recommendations to use carglumic acid immediately after raising a suspicion of organic aciduria caused by metabolic disorders of branched amino acids even instead of EE.

P-331 MAY DECREASED COMPLEMENT 4 BE A PREDICTİVE FACTOR OF DECREASED GLOMERULAR FILTRATION RATE IN CHILDREN WİTH ACUTE POSTSTREPTOCOCCAL GLOMERULONEPHRITIS?

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Introduction: Acute poststreptococcal glomerulonephritis (APSGN) is the most common postinfectious glomerulonephritis in childhood. It was aimed to determine the possible risk factor(s), responsible for decreased glomerular filtration rate (GFR) in APSGN.

Material and methods: The data of patients followed up with the diagnosis of APSGN in the Pediatric Nephrology Clinic of Gaziantep University Hospital between October 2014 and October 2016 were evaluated retrospectively.

Results: Total number of subjects was 75 (E/K: 42/33) with the mean age of 8.20 ± 3.25 years. Most common presentations were edema (86.7%), macroscopic hematuria (82.7%), proteinuria (77.3%), and hypertension (73.3%). In laboratory examination, 28 children (37.3%) had hypoalbuminemia, 20 (26.7%) with increased C reaktive protein (CRP), while 74 (98.7%) and 12 (16%) had decreased C3 and C4, respectively. The number of children with GFR value <90 ml/min/per 1.73/m^2 was 22 (29.3%). The risk of decreased GFR was found significantly higher in patients with increased CRP (p:0.001, OR:3.58), hypoalbuminemia (p:0.006, OR:4.83), and decreased C4 (p:0.010, OR:11.53). Additionally, leukocyte count, neutrophil count, and neutrophil lymphocyte ratio (NLR) were significantly higher (p:0.02, p:0.006, p:0.004, respectively) in patients with low GFR.

Conclusions: Although the prognosis of APSGN in children is good, severe systemic complications and renal failure may develop in the follow-up period. We suggest that decreased C4, presence of hypoalbuminemia, increased inflammatory markers (CRP, number of leukocyte and NLR) might be possible risk factors for degree of renal involvement.

P-332 THE ROLE OF ADAMTS13 DEFICIENCY IN SHIGA-TOXIN ASSOCIATED AND ATYPICAL HEMOLYTIC UREMIC SYNDROME IN CHILDREN

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Introduction: Moderate deficiency of ADAMTS13 may occur in other microangiopathic syndromes, because this enzyme limits the growth of thrombi in microcirculation. We conducted a comparative evaluation of ADAMTS13 activity in patients with STEC-HUS and atypical HUS (aHUS).
Material and methods: A total of 64 patients were diagnosed with STEC-HUS (mean age 2.65 ± 2 years) and 41 patients with aHUS (mean age 5.2 ± 4 years). The activity of ADAMTS13 was determined by the FRET method (fluorescence resonance energy transfer) using the fluorogenic substrate FRETS-VWF73 (PeptaNova GmbH, Germany) and expressed as a percentage (%). The interval of activity of ADAMTS13 in healthy patients was 80–122%.
Results: The activity of ADAMTS13 in children with STEC-HUS was 60.2 ± 18.7% (23–100%), in patients with atypical HUS - 74.5 ± 20.2% (37–131%). STEC-HUS in all cases was associated with acute intestinal infection. The manifestation of aHUS in 29% was associated with acute renal injury, 22% with diarrhea, 10% with vaccination, and 12% with no trigger. The severity of anemia (65.2 ± 12.7 vs 62 ± 13 g/l), thrombocytopenia (60 ± 35.5 vs 56 ± 29 × 109 / L) did not differ in groups, but the average values of azotemia, lactate dehydrogenase and D-dimer were in 1.5 times higher in patients with STEC-HUS (462.6 ± 202 vs 300 ± 211.5 μmol/L, 3864.6 ± 2983 vs 2580 ± 1831 U/L, 3554.5 ± 2495 vs 2050 ± 1268). Clinically, diarrhea (100 vs 22%, $p < 0.0001$), fever (79.8 vs 53.6%, $p < 0.005$), anuria (90.6 vs 29%, $p < 0.0001$) was significantly more frequent in STEC-HUS. Also, patients with STEC-HUS were needed dialysis in 1.5 times, and in artificial lung ventilation -2 times more often (93.7%vs63%, 20%vs7%) compared with patients with aHUS. Central nervous system lesions (seizures, coma) almost 2 times more often detected in children with STEC-HUS (40vs24.3%). Cardiovascular complications developed more often in patients with aHUS (78vs21%, $p < 0.0001$).
Conclusions: Moderate deficiency of ADAMTS13 activity is more common in patients with STEC-HUS than with aHUS (78.1%vs66%). The severity of the HUS correlates with the activity of ADAMTS13. Decreasing of ADAMTS13 activity in patients with STEC-HUS may be associated with its consumption caused by shiga toxin endothelial dysfunction.

P-333 DELAYED EFFECTS OF ECULIZUMAB THERAPY

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Introduction: A 14-year-old boy was admitted to our hospital after he had bloody diarrhea, weakness, nausea and abdominal pain. Laboratory test results indicated a thrombotic microangiopathy (MTA).
Material and methods: We documented low hemoglobin and platelet level, high LDH and Creatinine serum (Cs) levels and the presence of schistocytes in peripheral blood smear. ADAMTS 13 activity was normal and direct Coombs test was negative. Our patient was initially treated with plasma exchnge (PE) and hemodialysis. Before to begin PE a complement dysregulation test for mutation on CFH, CFI, MCP, CFB, CF3 and for anti CFH antibodies was conducted. Stool culture for STEC was negative. Few days after the onset of MTA the diagnosis of atpical hemoltic uremic syndrome (a-HUS) was made, a mutation on MCP was documented, and the patient was switched to eculizumab, after antibiotc prophylaxis and antimeningococcal vaccination was done.
Results: After first dose of eculizumab was done a complete normalization of hematological values of LDH, platelet and hemoglobin in a few days was observed. We stopped PE and continued hemodialysis for 3 months for the persistence of renal failure. During this period the patient continued eculizumab (1200 mg) therapy every two week, as maintenance regimen. Six months later the first dose of eculizumab Cs levels was 3.3 mg/dl. One year later Cs levels was 2.5 mg/dl. So we dosed CH50 activity <10% and decided to increase interval between doses to 21 days. Two years later MTA Cs levels was 1.9 mg/dl. Actually, after 3 year of eculizumab therapy, Cs levels is 1.4 mg/dl, no haematuria and proteinuria was revealed and no hematological activation of MTA was documented.
Conclusions: We report 3 - year safety and efficacy of eculizumab in a boy with a-HUS caused by a MCP mutation. In contrast, to other case reports, we documented a delayed effect of eculizumab regardig kidney function, that in our experience was indipendent of the dose and the interval of the dose we used.

P-334 DIAGNOSTIC DILEMMA: RENAL SIMPLE CYST AND CALYCEAL DIVERTICULUM

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Introduction: Renal calyceal diverticulum is a cystic formation related to the pelvicalyceal system which is rarely diagnosed in childhood age group and can be confused with other cystic diseases of the kidney. Due to the relationship with the pelvicalyceal system, it can cause clinical symptoms such as infection, stone formation and hematuria.
Material and methods: The children with a diagnosis simple renal cysts and with detailed ultrasonographic findings and advanced imaging results of the renal cyst were evaluated retrospectively according the their age, sex, presenting symptoms, simple renal cyst features (size, location) and advanced imaging results.
Results: A total of 22 cases (11 girls) were evaluated. Mean ages were 10.5 ± 3.9 (5–18) years. Simple renal cyst was detected in 13 left, 7 right kidney and two both of the kidneys. The mean size of the cysts were 21 ± 11.5 (10–58) mm. The location of the cysts were 11 corticomedullary, 4 cortical, 6 medullary, and 1 parapelvic. All of the cases had been evaluated with MR urography as advanced imaging. MR urographic results showed that totally 7 calyceal diverticulum (31%) in all of the patients . The other 15 simple renal cysts diagnosis were confirmed as renal cyst. The presenting symptoms of cases with calyceal diverticulum were; 3 with abdominal pain, 2 with microscopic hematuria, 2 with urinary tract infection. Other cases were asymptomatic. Ultrasonographic cyst sizes of patients with diverticulum detected by MR urography were 20.8 ± 6.1 mm (10–30) in average and there was no

significant difference in terms of cyst size. None of the cases of cayceal diverticulum require surgical intervention.
Conclusions: It should be kept in mind that some of the patients with simple renal cysts may be calyceal diveritculum. MR urography can be used safely with advanced imaging techniques in differential diagnosis.

P-335 BABY PORT: WHY NOT?

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Introduction: Port is a totally implantable system which allows a reliable long-term vascular access. It respects the body integrity and causes fewer local and/or systemic complications in respect of tunneled venous catheters. Port implantation procedure requires a special practice for obtaining insertion techniques skill, specific know-how to manage medical staff, and care-givers compliance. Despite of this advantages, the insertion of Baby Port devices is not yet not a widespread procedure.Baby Ports and VADs small-sized availability prompts the authors to investigate prcaticalutilization of these devices, verifying their performance in chronic patients requiring venous accesses for treatment.
Material and methods: In a period of 18 monthseight male children aged between 4 months and 8 yearswere enrolled. The youngest one was affected by Congenital Nephrotic Syndrome finnish type (CNS) Six requiredi.v. discontinuous substitutive therapy; two required daily Total Parenteral Nutrition (TPN).
Results: Baby Ports implant was successfully performed in all. In the child with CNS, Baby Port, used for the daily albumin i.v. infusion, had a permanence lasting 13 months without serious adverse events.Primary complications were not observed. Secondary complications were more frequent during the period of care givers training and reduced after completing learning period.The complications rate was 1.5 sepsis/ 1000 catheters day,and 2.0 reversible clotting occlusion / 1000 catheters day; this rate was as high as the number of care givers/units involved.
Conclusions: Baby Port is a real reliable device that can be used also for pt. who need daily therapies. Important to highlight its use is possible only with an appropriate training of care-givers.

P-336 A RARE CAUSE OF CHRONIC RENAL DISEASE: CONORENAL SYNDROM

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Introduction: Conorenal syndrome (Mainzer-Saldino Syndrome) was reported with cone-shaped epiphyseal plates, retinitis pigmentosa, ataxia and nephronophthisis leading to chronic renal damage in the later period. In this report, a case with the diagnosis of conorenal syndrome was reported.
Material and methods: A 5 year old girl admitted to our hospital with diffuse edema on the eyelids and lower extremities, bilateral basiller crackles and mild ascites. She had growth retardation. Laboratory tests at admission revealed metabolic acidosis and azotemia. The GFR calculated using the Schwartz formula was 8.59 mL/min/1.73 m2. The ultrasound scan showed that both kidneys were small for her age and increase at bilateral renal echogenicty. She had accepted Stage 5 chronic kidney disease and got involved peritoneal dialysis program. Examinations carried for the etiology of renal failure; there wasn't vesicoureteral reflu or different urological pathologies. Ophthalmologic examination revealed bilateral diffuse choroidal and retinal atrophy, retinitis pigmentosa. During the fallow up period, noticed on traces striking asymmetry in her fingers. When the upper and lower extremity radiographs obtained in, detected symmetrical shortening of the ulna in both forearms, shortening all the phalanx and dilatation of proximal metaphyseal joints. The biopsy required to show the nephronophthisis in the kidney, while directing it to Senior-Loken Syndrome, could not be done due to end-stage renal disease. In genetic screening, the previously reported p.G212R homozygote mutation associated with the disease was detected in the IFT 140 gene.
Results: Following a 3-month peritoneal dialysis program, the patient underwent kidney transplantation from her mother.
Conclusions: Detection of primary disease in patients with end-stage renal insufficiency can be difficult due to the reduced probability of biopsy. However, in the studies to be done for etiology, detection of retinitis pigmentosa findings and detection of cone shaped fingers should suggest conorenal syndrome.

P-337 GELFOAM BIOPSY TRACT EMBOLIZATION IS SAFE AND MINIMIZES COMPLICATIONS AFTER RENAL BIOPSY

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Introduction: The kidney is the organ with the highest post-biopsy clinically-relevant bleeding risk, estimated at 1.5–10% in pediatric patients. Gelfoam slurry tract embolization may improve post-biopsy outcomes.
Material and methods: Retrospective review of 13 biopsies performed in 12 patients. A fully-automatic core-biopsy needle was used coaxially in all patients, (19/20-gauge in one patient, 17/18-gauge in the rest). Gelfoam slurry was performed by agitating gelfoam pledgets with normal saline until achieving slurry consistency, and injected at the completion of biopsy. All biopsies were completed with real-time pathologic verification using 4 or less needle passes. All patients were admitted overnight with ultrasound follow-up the next day.
Results: Biopsy and gelfoam administration was technically successful during all biopsy sessions. One hematoma (1/13 biopsies, 7.9%) occurred in a patient with acute renal failure (Cr 2.3) increased from 3 mm to 12 mm on post-procedure day 1 but was clinically insignificant and required no treatment. One episode of hematuria (mild to moderate severity, 1/13, 7.9%) occurred in a patient with acute renal failure (Cr. 2.7) but was self-limited and required no treatment. Hemoglobin levels remained stable after the procedure (pre versus post-biopsy hemoglobin 12.4 g/dL versus 11.6 g/dL, within the range of error of the laboratory machine, $p = 0.0173$ paired student t-test). Creatinine levels (available at both time points for 10 biopsies) did not show a statistically significant increase after biopsy (0.82 pre versus 0.9 post, $p = 0.2576$ paired student t-test).
Conclusions: In this small pilot series, there were no clinically significant complications (complications that require treatment or an unplanned elevation in level of care) in the cohort of patients who received gelfoam slurry embolization. Furthermore, creatinine levels remained at safe levels in the immediate term (any longer term follow-up of creatinine would be confounded by treatment response for the renal failure).

P-338 EFFICACY OF TREATMENT WITH ECULIZUMAB IN CHILDREN WITH AHUS

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Introduction: Atypical hemolytic uremic syndrome (aHUS) is an orphan disease with severe relapsing course originating from complement system dysregulation with uncontrolled activation of alternative pathway leading to the production of membrane-attacking complex and consequent damage of endothelium again manifesting with thrombotic microangiopathy (TMA) and renal failure. Treatment with Eculizumab so far is an only pathogenically aimed approach to aHUS.
Material and methods: Medical records from 39 hospitals countrywide were analyzed. Molecular genetic analysis was performed using NGS method for *CFH, CFI, CFB, MCP,* and *THBD* genes, and MLPA for deletion at *CFHR3/R1* locus.
Results: We have selected and analyzed data from 71 children with aHUS. Eculizumab was used in 39 children including 20 with and 19 without mutations. Median time of treatment initiation was 4.5 and 2.0 months after the onset correspondingly. In most of the cases at that time the result of genetic study was not yet available. About 50% in each group elevated their GFR 25% and more within 6 months. Eight patients of 9 in the mutation group and 7 of 9 in non-mutation discontinued dialysis. Even if Eculizumab was started after 6 or more months after it onset, 7 patients with mutations markedly (>25%) improved their GFR. None of untreated patients after 6 months on dyalisis (5 children) stopped dialysis. Side effects as allergic angioedema in 2, dilating cardiomyopathy in 1 and non-viral hepatic disorder in 1 were reported.
Conclusions: Eculizumab efficacy was compatible in patients with or without mutations, both improved renal function at least 25%. Treatment with Eculizumab despite needs a further study is likely to significantly improve prognosis even if it started months later.

P-339 AN UNUSUAL CAUSE OF MACROSCOPIC HEMATURIA: ANTERIOR - POSTERIOR NUTCRACKER SYNDROME AND LEFT RENAL VEIN DUPLICATION

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Introduction: Nutcracker syndrome (NCS) is a rare reason of hematuria. It has two anatomical appearances as anterior and posterior. Here, anterior-posterior NCS and left renal vein duplication (LRV) in a 15-year-old adolescent who applied due to complaint of macroscopic hematuria was reported.
Material and methods: Case: A 15-year-old female patient applied with the complaint of red colored urination for the last 2 days. There was no macroscopic hematuria at the time of application. She had no pain, urinary burning, high fever or vomiting accompanied with this complaint. No trauma was specified. Physical examination was normal. In the urinary examination, density was 1010, ph was 5.0, and erythrocyte was +2 positive and in her microscopic examination there were 15–20 erythrocytes in every area. There was no proteinuria. Urinary calcium creatinine ratio, complete blood count, and renal function tests performed for hematuria etiology were found to be normal. In renal ultrasonography, kidneys were at normal size and echogenicity for her age. Hydronephrosis, stone and renal mass were excluded. Complement tests and antinuclear antibody (ANA) were negative. In renal doppler examination, accessory renal vein on the left and also left renal vein with a retroaortic localization drew attention. In computed tomography angiography examination, it was assessed double renal vein that more anterior one among the renal veins was compressed between SMA and aorta and the other one was compressed between the abdominal aorta and vertebral column. (Figure 1) Thus the patient was diagnosed with anterior posterior NCS with LRV duplication. She was followed for hematuria and proteinuria.
Results:
Conclusions: Occurrence of both of anterior and posterior NCS is a quite exceptional situation. NCS should be kept in mind in the differential diagnosis of hematuria. Also, LRV variations should be remembered in the differential diagnosis of hematuria and venous anatomy should be shown by advanced imaging methods.

P-340 RENAL CYSTS IN CHILDREN – OWN OBSERVATIONS

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Introduction: Renal cysts are mainly detected during ultrasound. They may be congenital (associated with genetic disorders or not) or acquired.
Material and methods: The aim of the study was the retrospective analysis of patients hospitalized in two-years period (2015–2016) with suspicion/diagnosis of renal cysts.
Results: In the analyzed time 1604 patients were hospitalized, among them 49 children (aged from 1 month to 17 years) met the inclusion criteria. In 30 cases, pateints were admitted to the clinic for the first time, in other cases - again. Among children hospitalized for the first time, there were 14 infants (mean age 4.2 months). Positive family history was noted in 2 children, renal cystic disease were suspected prenatally in 5 cases. Final diagnosis was: autosomal recessive polycystic kidney disease (ARPKD) in 3 children, autosomal dominant polycystic kidney disease (ADPKD) in 1, multicystic dysplastic kidney (MCDK) in 4, connected with ectopia of left kidney in 1, bilateral MCDK in 1, isolated renal cyst in 2, renal hypodysplasia with cysts in 2 children. In 16 older children (mean age 10.1) diagnosed for the first time due to hypertension or abnormality in abdominal USG, ADPKD was diagnosed in 11 children, right MCDK in 4 and left MCDK in 1. In 19 patients hospitalized again (mean age 9.5) the diagnosis was: ADPKD in 4, ARPKD in 6, MCDK right in 4, MCDK left in 3, ADTKD in 1 and medullary sponge kidney in 1. In cases of 23 (47%) described children, chronic kidney disease (CKD) in different stages was diagnosed, in 3 patients kidney transplantation was performed. Acquired cysts were not found.
Conclusions: Renal cysts are not a common cause of hospitalization (3.1% of our patients), but they require early diagnosis and monitoring of patients due to the risk of CKD development in some cases.

P-341 THE COMPLICATIONS OF PERCUTANEOUS KIDNEY BIOPSY: ONE CENTER EXPERIENCE

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Introduction: Percutaneous Kidney Biopsy (PKB) has remained a gold standard procedure for obtaining further information about the diagnostic, prognostic and therapeutic processes of kidney diseases. Although PKB is a safe and useful procedure in terms of the diagnosis in nephrology practice, some complications still exist due to invasive nature of procedure. In this study, we evaluated the complications and identify factors possibly affecting bleeding complications associated with PKB.

Material and methods: This retrospective study includes patients who underwent PKB procedure at the Department of Pediatric Nephrology, Kayseri, Turkey from 2003 to 2016. We analyzed 397 biopsy reports, 93 of them who underwent kidney biopsy between 2011 and 2016 were included into study the further evaluation.
Results: 41 girls and 52 boys with regular long-term follow were included into the study. The mean age was 10.0 ± 4, 3 years and 12, 0 ± 4, 4 at the time of biopsy and last follow-up, respectively. We detected obesity in 10 children, failure to thrive in 3 children, short stature in 13 children, hypertension in 9 children, prolonged PT and APTT required fresh frozen plasma prior to procedure in 3 children, and nephromegaly in 27 children. Seventeen children had gross hematuria and 59 patients had microscopic hematuria prior to procedure, then 9 new patients experienced with gross hematuria after the procedure. Renal ultrasound showed perinephric hematoma in 15 patients and arteriovenous fistula in 4 patients after the procedure. The occlusion of the fistulae was performed in 1 of them. There is no relationship between bleeding complications and Hb value, obesity, blood pressure, acute kidney damage (GFR < 60), hypoalbuminemia, PT, APTT.
Conclusions: Our study showed that PKB is a useful technique with usually minor and rarely life life-threatening complications like arteriovenous fistula requiring renal angiogram and closing with coil embolization.

P-342 CALCIUM-SENSING RECEPTOR GENE POLYMORPHISMS AND IDIOPATHIC HYPERCALCIURIA IN CHILDREN

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Introduction: Idiopathic hypercalciuria is the most common identifiable cause of calcium kidney stone disease in children. Polymorphisms in several genes have been associated with a genetic susceptibility for the disease. Calcium-sensing receptor (CaSR) gene is involved in the normal balance among renal calcium, phosphate, protons, and water excretion. The aim of the present study was the investigation of the CaSR gene polymorphisms in children with idiopathic hypercalciuria.
Material and methods: The polymorphisms A986S, R990G and Q1011E (exon 7) of CaSR gene were studied in 27 children with idiopathic hypercalciuria and 67 normal children age and gender matched to the patients. The diagnosis of hypercalciuria was based on the urine calcium excretion value of >4 mg/Kg/24 h in two consecutive 24-h urine collections. Children with secondary hypercalciuria were excluded from the study. Genomic DNA was extracted from the whole blood sample. Polymerase chain reaction (PCR) and sequencing analysis of amplified segments were performed.
Results: The frequency of the R990G and Q1011E in the patients did not differ significantly from that of controls (11.1% versus 1.5% and 7.4% versus 10.5% respectively). The A986S polymorphism in either the heterozygosity or the homozygosity state (genotype GT or TT) of the CaSR gene was found to be in a significantly higher rate in the patients (59.2%) compared to the controls (26.8%), $p < 0.05\%$. The genotype GT was found in 55.5% of the patients and 25.3% of the controls, and the genotype TT in 3.7% and 1.5% respectively.
Conclusions: The polymorphism A986S of the CaSR gene was found to be associated with idiopathic hypercalciuria in the study population, indicating a genetic predisposition of the disease. Studies in a higher number of patients are necessary to reach unequivocal conclusions.

P-343 OXIDATIVE STRESS AND RENAL TUBULAR DYSFUNCTION IN PEDIATRIC NEPHROLITHIASIS: PROTEOMIC EVIDENCE

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Introduction: Using a proteomic approach, we assessed the differences in the urinary proteins between children with renal stones (RS), and healthy controls (HC), with particular attention to the proteins involved in oxidative stress and tubular injury.
Material and methods: Quantitative proteomic comparison of pooled urine from RS ($N = 30$, 24 females, mean age 12.95 ± 4.03 years) versus age- and gender-matched HC ($N = 30$), using liquid chromatography-mass spectrometry (LC-MS/MS). Relative protein abundance was estimated using spectral counting. Proteins of interest were selected using the following criteria: 1) ≥5 spectral counts; 2) ≥2-fold difference in spectral counts; and 3) ≤0.05 *p*-value for the Fisher's Exact Test. Cytoscape was used to investigate the key nodes of unique proteins.
Results: Of the 1813 proteins identified, 230 met the above criteria, with 163 proteins up-regulated in RS group and 67 up-regulated in HC. Function analysis revealed 23 inflammatory proteins, 8 proteins involved in oxidative stress, and 4 involved in tubular injury. Of those, NADPH-oxidase, a major source of reactive oxygen species, was only found in RS group, while Glutathione S-transferase, an important antioxidant enzyme, was more abundant in controls (Table). Protein-protein interaction modeling of selected proteins identified syndecan-1 as the key node.

Accession Number	Protein	No. of assigned peptides*	No. of unique peptides*	Ratio (patient/control)
BLVRB	Flavin reductase (NADPH)	5 (0)	4(0)	Unique to stone
GSTA2	Glutathione S-transferase A2	19 (38)	8 (10)	0.59
B2MG	Beta 2-microglobulin	156 (27)	8 (8)	4.89
RET4	Retinol-binding protein 4	520 (70)	22 (14)	6.54
LYSC	Lysozyme C	109 (19)	14 (8)	4.68
CYTC	Cystatin C	146 (34)	14 (11)	3.87

*Values presented in patients and controls (brackets)

Conclusions: We provide proteomic evidence of oxidative stress, inflammation, and tubular injury in children with renal stones. We speculate that inflammation and changes in the oxidant-antioxidant balance may cause tubular damage in these patients. Targeting these proteins may have therapeutic benefits.

P-344 ATYPICAL HEMOLYTIC URAEMIC SYNDROME: 10 YEARS SINGLE CENTRE EXPERIENCE

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Introduction: Hemolytic uremic syndrome (HUS) is defined by the triad: microangiopathic hemolytic anemia, thrombocytopenia, and acute kidney injury. Atypical HUS (aHUS) is usually caused by disorders in complement alternative pathway regulation or may be secondary due to *Streptococcus pneumoniae* or other causes. aHUS represents 5–10% of HUS in children. The disease has a poor prognosis - 50-60% progress to ESRD.
Material and methods: We present a single centre experience of aHUS.
Results: 7 patients (2 girls, 5 boys) were treated in The Hospital of Lithuanian University of Health Sciences Kauno klinikos during the period of 2007–2016. The mean age at the onset of the disease – 6.0 (2.5–13) years. Anti-CFH was diagnosed for two patients. Four patients had negative genetic test results. Genetic tests were not performed for 2 patients. Plasma therapy as initial treatment was prescribed for 5 patient, one received *Eculizumab* and 1 patient was treated for sepsis. Supportive treatment was prescribed for 3 patients: 2 received *Eculizumab*, one - *Mycophenolate Mofetil with Ig-Immunoglobulin G*. Four (57%) children developed Chronic Kidney Disease (CKD). One of them was transplanted. Children treated with *Eculizubam* had good outcomes: one has normal renal function, the other improved renal function (chronic dialysis was stopped and the child has CKD st. 3).
Conclusions: 7 children diagnosed aHUS during the period of 10 years.
1. Four (57%) children developed CKD. One of them was transplanted.
2. Causes of aHUS are different thats why clinical features and outcomes are different.
3. Treatment with *Eculizumab* significantly improved outcome of the disease.

P-345 A SIMPLIFIED APPROACH TO THE CHILD WITH HEMATURIA

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Introduction: Hematuria is a frequent reason for consultation of pediatric nephrologists. However, hematuria in children is usually a less frequent complaint for consulting community medical settings. Furthermore, many cases of hematuria are tansient and do not recur. This frequently results in diagnostic dilemmas, as unnecessary investigations and treatment should be avoided. However, children suffering any serious renal or urologic disease, should urgently be referred to a pediatric subspecialist. In this abstract, we present our simplified approach to the child with hematuria.
Material and methods: By means of a literature review and a review of charts of patients who were referred to pediatric nephrology clinic, we developed a structured and simplified approach to the child with hematuria.
Results: A thorough history and complete physical examination should be the first step in all children evaluated for hematuria. However, children with any indication of traumatic hematuria, or signs of severy systemic illnesses such as sepsis, should urgently be treated according to applicable protocols such as APLS guidelines. In selected cases, different urinalyses are usually a rational next step in the diagnostic process. Other tests, such as radiology and lab tests, should be performed with clear indications. An adequate follow-up is mandatory for all children presenting with hematuria.
Conclusions: The diagnostic work-up of hematuria can be simplified using a structured approach.

P-346 PARTICULAR QUALITIES OF RENAL FILTRATION FUNCTION IN CHILDREN AND ADOLESCENTS WITH OBESITY

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Introduction: The objective of this study was to determine the frequency of renal malfunctions in children and adolescents with obesity.
Material and methods: Glomerular filtration rate (GFR) was estimated in 76 patients with obesity, age range 5 to 17 years. Male/female - 42/34. Forty one patients had II, 31 - III, 4 - IV grade obesity. GFR was calculated by using Schwartz formula.
Results: In 11 patients (14.5%) GFR was increased more than 130 ml/min/1.73 m^2, in 9 (11.8%) - moderately decreased <90 ml/min/1.73 m^2, but >60 ml/min/1.73 m^2. The variation of GFR males had twice frequently (65%) than females. The age of the patients with decreased GFR was significantly higher (14.9 ± 0.9 years), than in patients with increased GFR (11.0 ± 0.9 years, $P < 0.01$). The elevation of GFR more often demonstrated patients with the second grade obesity (63.6%). Decreasing of GFR was not depended by the grade of obesity.
Conclusions: In children and adolescents with obesity (more frequently in males) the variation of glomerular filtration rate can be noted. Obviously, initially the increasing of GFR provocated by superfluous intake of protein. With the increasing of age and obesity rate decreasing of GFR can be noted more frequently.

P-347 HOSPITALIZATION BURDEN IN CHILDREN WITH CHRONIC KIDNEY DISEASE (CKD): FINDINGS FROM THE 4C STUDY

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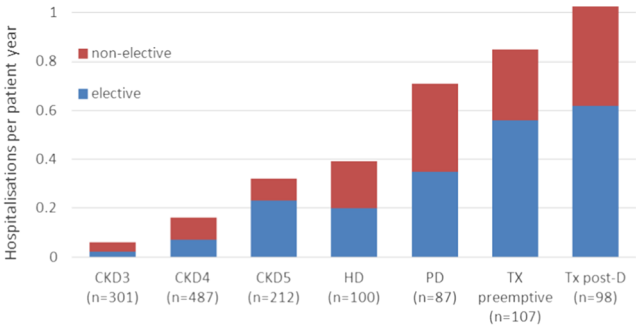
Introduction: Hospitalizations are associated with significant psychosocial and economic burden. Whereas studies in adults have demonstrated an association between advancing CKD and increased risk of hospitalizations, information about the pediatric CKD population is scarce. We

analyzed hospitalization burden in the 4C Study, a large European pediatric CKD cohort.
Material and methods: Patients aged 6–17 years with CKD stage 3–5 or renal replacement therapy (RRT) followed at 55 centers in 14 countries with at least two study visits were included in the analysis. Data about CKD stage, RRT and hospitalizations were collected 6-monthly. Negative binomial regression model was used for multivariable analysis.
Results: A total of 662 patients were followed for a total of 2718 patient-years (PY). Overall and non-elective hospitalization rates increased with advancing CKD and start of RRT (Figure). The same trend was observed for time spent in hospital (0.6, 0.75, 1.2, 6.1 and 9.9 days per PY in patients with CKD3, CKD4, CKD5, dialysis and renal transplant (Tx) recipients, respectively). In the multivariable analysis, RRT (IRR 6.64 and 5.3 for Tx; IRR 3.78 and 3.78 for dialysis, all $p < 0.0001$; reference - CKD), presence of comorbidity (IRR 1.5 and 1.53, $p = 0.001$) and younger age (IRR 0.92 and 0.93 per 1-year increase, $p < 0.0001$) but not renal diagnosis ($p = 0.59$) were associated with higher overall and non-elective hospitalization rates, respectively. RRT (IRR 9.9 for Tx and IRR 7.43 for dialysis, both $p < 0.0001$; reference - CKD) and younger age (IRR 0.92 per 1-year increase, $p = 0.002$) were also associated with higher number of days spent in hospital per PY.



Conclusions: Advancing CKD, younger age and presence of comorbidities are associated with increased hospitalization burden in children with CKD.

P-348 #KIDNEYSCHOOL – A NOVEL, INTERACTIVE, TRAINEE LED, VIDEO-CONFERENCING TEACHING PROGRAMME

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Introduction: #kidneyschool is an informal, interactive and engaging method of peer led education established by Paediatric Nephrology trainees in the UK. #Kidneyschool builds upon Telehealth which is an innovative and rapidly expanding industry worldwide. It is used as a tool to link patients with medical specialists, in education and to overcome the barriers of distance. In our survey 60% of UK trainees indicated the need for more dedicated teaching. Paediatric Nephrology trainees are geographically dispersed across the UK; teleconferencing offers a unique opportunity to remove the barriers to education such as travel distance, expense and time, whilst allowing for co-ordination around busy schedules. We aimed to increase the amount of dedicated educational time for Paediatric Nephrology trainees and build a strong peer network amongst trainees that can be translated into collaborative working across Paediatric Nephrology as consultants.
Material and methods: We designed and co-ordinated a trainee run, interactive, monthly video-conferencing facility that allows shared learning in a more informal, but still productive environment. #kidneyschool uses the multi-media conferencing facility Zoom, this allows presentations, PDF files or webpages to be viewed and users to interact with these. Teaching is based around interesting cases, which is important as evidence shows that learning and discussion focused around a "real-life" work based problem, is more effective than didactic teaching.
Results: Trainees are exposed to varied case-loads depending on local patient populations and this forum allows trainees to share expertise on the wide-ranging spectrum of patients seen across the UK. We also facilitate trainees fulfilling their RCPCH GRID curriculums. Documents are stored on Dropbox and provide trainees a resource to refer to or for those who could not attend.
Conclusions: We present our feedback from #kidneyschool and demonstration of a session. We believe this trainee led innovation provides an excellent platform for teaching and education whilst building a successful peer network.

P-349 A HUMAN PROXIMAL TUBULAR EPITHELIAL CELL MODEL TO EXPLORE A KNOWLEDGE GAP ON NEONATAL DRUG DISPOSITION

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Introduction: Finding the right drug-dosage for neonates is still a medical challenge. Up to now, neonatal doses are extrapolated from adult and children doses. However, there are differences between neonatal and adult kidney physiology that should be put into consideration, especially when it comes to active drug metabolism. Studying renal drug clearances in neonates is limited by the lack of reliable human cell models. Our aim was to illustrate the feasibility to develop an *in vitro* model for neonatal proximal tubule epithelial cells (nPTECs) for studying renal drug clearances at this age.
Material and methods: nPTECs were isolated from urine samples of neonates of different gestational age (GA) and conditionally immortalized using a temperature sensitive SV40T antigen and human telomerase hTERT. The cell clones were characterized on gene expression level for PTECs markers such as P-glycoprotein (P-gp), aquaporin1 (AQP1), and organic cation transport protein 2 (OCT2). In addition, protein expression and functional assessment were performed for P-gp and OCT2.
Results: We established 101 clonal cell lines of cinPTECs derived from neonatal urine. Gene expression analysis confirmed the expression of the PTECs (P-gp, AQP1, and OCT2), similar to the expression in the adult control ciPTECs. P-gp was expressed in cinPTECs from the different gestational ages and exhibited similar functionality as the adult derived ciPTECs. In contrast, OCT2 functionality was significantly lower in the cinPTECs cell lines compared to the adult ciPTECs.
Conclusions: We demonstrate the feasibility of culturing cinPTECs expressing mature ciPTECs markers with high efficiency out of the urine samples of neonates. The cell model presented here can serve as a valuable tool to study proximal tubule physiology and pharmacology in new-borns.

P-350 STRESS, ANXIETY AND DEPRESSION IN PARENTS OF CHILDREN WITH CHRONIC KIDNEY DISEASE

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Introduction: The quality of life in children with chronic kidney disease (CKD) is impaired. However, little is known about the impact on the family. The aim of this study was to explore the psychological wellbeing in this parent group with special emphasis on socio-demographic variables, parental stress, anxiety and depression.
Material and methods: Thirty-six parents (27 mothers) of children with CKD completed a socio-demographic questionnaire, 2 questionnaires assessing parental stress (Parenting Stress Index - Short Form (PSI-SF) and Pediatric Inventory for Parents (PIP)), and the Hospital Anxiety and Depression Scale (HADS) measuring symptoms of anxiety and depression.
Results: The socio-demographic questionnaire revealed that half of the parents (18/36) perceives a deterioration of their own health since the CKD diagnosis of their child. In 17/36 families, at least one of the parents reduced work activities due to the CKD diagnosis. Regarding general parental stress (PSI-SF), parents generated scores within the normal range (59 ± 23; normal range 43–61). The disease related parental stress measurement (PIP) showed the highest stress level on the domain 'emotional distress' (42 ± 13) and equal stress levels (22) on the 3 other domains 'communication', 'medical care', and 'role function'. Parents reported mild symptoms of anxiety (8 ± 4) but normal symptoms of depression (5 ± 4). Exploring the relation between all measurements of parenting stress, anxiety and depression, overall significant correlations could be found (correlation coefficients between $r = 0.51$ and $r = 0.69$; $p < 0.01$).
Conclusions: CKD in children does not only affect the child itself. Parents of CKD patients report more health problems since the diagnosis of their child, and a suggestive presence of anxiety problems can be seen. Normal but rather high mean levels of parental stress can be found. As the impact of CKD goes beyond the child and affects the entire family, a multidisciplinary family-based therapy is recommended.

P-351 PSYCHOLOGICAL WELLBEING OF CHILDREN WITH CHRONIC KIDNEY DISEASE

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Introduction: Chronic kidney disease (CKD) influences the psychological wellbeing of children. The aim of this study was to explore quality of life (QoL), psychological problems, attention and executive functioning in this patient group.
Material and methods: Thirty-six parents completed questionnaires regarding the psychological wellbeing of their child with CKD (0–18 years), 26 children were also evaluated by their teacher or caregiver of the daycare center. Twenty-four children with CKD and older than 8 years (age 13 ± 3; 17 boys) completed questionnaires to evaluate their own psychological wellbeing. This multi-informant assessment explores QoL (PedsQL™ 4.0 Generic Core), psychological problems [Child Behavior Checklist (CBCL) / Teacher Report Form (TRF) / Youth Self Report (YSR)], attention [Disruptive Behavior Disorder Rating Scale (DBDRS)), and executive functioning (Behavior Rating Inventory of Executive Function (BRIEF)].
Results: Parents and patients both significantly ($p < 0.001$) reported a lower QoL (69 ± 19 and 71 ± 14, respectively) compared to control parents and healthy children (88 ± 11 and 83 ± 11, respectively). Regarding the CBCL/TRF/YSR, 15/36 parents scored their child in the (sub)clinical range of internalizing problems compared to only 6/20 children and 7/26 teachers. Externalizing problems in the children seems to be less present: 7/36 parents, 1/20 child and 6/26 teachers generated scores in the (sub)clinical range. (Sub)clinical scores for attentional functioning of children were found in 2/19 parents and 1/18 teachers. The executive functioning of children was considered as (sub)clinical by 3/26 parents and 3/20 teachers.
Conclusions: The quality of life in children with CKD is impaired. Almost half of the parents are concerned regarding possible internalizing problems of their child. A multidisciplinary family-based therapy is therefore recommended.

P-352 PARENTAL EXPERIENCE OF PAEDIATRIC KIDNEY TRANSPLANTATION

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Introduction: Kidney transplantation is life changing not only for the individual receiving the transplant but for the whole family/support network. We were keen to explore the parental experience of pre and post kidney transplantation preparation and education at our hospital. With the aim of exploring parental experiences and views to improve the care provided to our children and their families.
Material and methods: Parents of children receiving a kidney transplant in the previous 6 to 24 months were invited to a 90 min focus group. The group was facilitated by clinical psychology and a renal registrar and explored parents' experiences of pre and post-transplant preparation and support. With parental consent audio-recordings were made and later transcribed. The data was then analysed using thematic analysis.[1]
Results: 7 out of 24 families attended. Thematic analysis identified overarching themes reflecting the data collected. Many families described positive experiences of feeling supported and well informed and viewed the team as caring and professional in their approach. Transplant was described by many as transformative for child and family. Challenges were raised for some, such as the demands of hospital visits and understanding of chronicity of condition. Sibling and family peer support were highlighted by some as areas for improvement.
Conclusions: The data has highlighted positive aspects of care and areas for further improvement. On-going thematic analysis will further refine the overarching themes.

P-353 OVERWEIGHT AND OBESITY ASSOCIATED WITH RENAL PROBLEMS IN CHILDREN

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Introduction: The effect of overweight especially central obesity on the kidney is well studied in adults and still evolving. In children the obesity related glomerulopathy (ORG) is not fully understood in children. On the other hand, many congenital and hereditary conditions are associated with obesity are there but not gathered yet in any single study. Obesity and overweight are frequently induced by the interventions needed in pediatric nephrology as well. The purpose of this work is divided to two portions; the first is to review the studies included the effect of obesity on the renal diseases in children. The second portion is to gather all the congenital, hereditary and acquired renal conditions associated with overweight and obesity in children in one systematic review.
Material and methods: The obesity related glomerulopathy including the effect of proteinuria, hypertension, nephrolithiasis and kidney cancer were thoroughly reviewed. Extensive search in OMIM; the Online Mendelian Inheritance in Man came up with only seven obesity syndromes associated with renal disorders. The acquired obesity induced by the renal conditions intervention in children was categorized to

malnutrition, drug-induced, peritoneal dialysis and issues related to hemodialysis and kidney transplantation.
Results: The obesity related glomerulopathy is not studied well in children although obese children usually share the same squeals of ORG like adults. The most common of the seven obesity syndromes associated with renal disorders is Bardet–Biedl syndrome. Unless a pediatric dietician with experience in kidney problem is actively involved in the management of kidney diseases, many children will develop malnutrition towards overweight than underweight in developed countries. Glucocorticoid is the most common drug-induced obesity in pediatric nephrology intervention. Peritoneal dialysis and "Let them eat" is the beneficial strategy for hemodialysis. Obesity is a known complication post kidney transplantation and no consensus or guideline is present for the obese kidney donors for children yet.
Conclusions: Obesity and overweight in children are horribly increasing every year worldwide. Obesity related glomerulopathy is not studied well in children. Few genetic errors are associated with combined obesity and renal disorders. Pediatric nephrology intervention is frequently associated with overweight in children if it is not actively supervised with the expert pediatric nephrology dietician.

P-354 VITAMIN B6 CONCENTRATIONS IN PAEDIATRIC DIALYSIS PATIENTS

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Introduction: Studies in adult dialysis patients have shown vitamin B6 deficiency is common. Vitamin B6 levels are not routinely tested in paediatric dialysis patients and recent data is lacking in this patient group. K/DOQI guidelines recommend that children with CKD on dialysis receive a water-soluble vitamin supplement. This study investigated vitamin B6 concentrations in our paediatric dialysis patients.
Material and methods: Retrospective review, including all children on maintenance dialysis (peritoneal dialysis, PD; home haemodialysis, HHD; intermittent haemodialysis, IHD), for vitamin B6 concentrations over a recent 24 month period.
Results: 18 children (8 HD, 7 PD, 3 HHD) with mean ± SD age 8.4 ± 6.3 years (15 boys). There were 14 white British, 3 Black and 1 Asian ethnicity. Vitamin B6 concentrations were within range (35.2–110.1 nmol/L) in 44% with the remainder above normal range (130–404.1 nmol/L). Eleven children were receiving enteral feeds, 8 had high vitamin B6 levels of which 6 were feed dependent (feeds provided 0.08–1.04 mg vitamin B6/d) (40–148% DRV). Half of these children were meeting their DRV for vitamin B6 from feeds alone. Four of the feed dependent children received a supplement containing vitamin B6 (1-11 mg vitamin B6/d) (1–12 times the DRV). Seven children were not tube fed, 2 had high vitamin B6 levels, 1 was receiving a supplement containing vitamin B6 (0.25 mg daily; 21% DRV).
Conclusions: Children in our dialysis group, who were not receiving a tube feed had normal/high vitamin B6. The need for supplementation in children who are tube feed dependent needs to be reviewed as this is contributing to high vitamin B6 levels. Half of these children were already meeting their DRV for vitamin B6 from feed alone. Routine vitamin B6 supplementation is currently not indicated in our paediatric dialysis patients.

P-355 AN ASSOCIATION BETWEEN URINARY METABOLIC DISTURBANCES AND INFANT FEEDING IN CHILDREN WITH MALABSORPTION SYNDROME

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Introduction: Big affinity of functioning of plasmatic membranes of a kidney and bowel at transport of various elements, including oxalates and uric acid is noted. So, the aim of our study is investigate urinary metabolic disturbances at children with a malabsorption syndrome depending on character of food on the first year of life.
Material and methods: 92 children with syndrome of malabsorption who are earlier not examined and not the treated concerning changes from an urinary system aged from 2 months up to 6 years have been examined. From them - 42 (45.6%) patients with a celiac disease, 50 (54.4%) - with chronic enteritis. 51 (65.2%) of the patient had clinical manifestation of the main disease, 41 (34.8%) had a phase of incomplete clinical laboratory remission. The daily excretion of oxalates, uric acid, creatinine was investigated, character of food on 1 year of life was analyzed.
Results: Direct correlation between metabolic disturbances in relatives and hyperoxaluria in the child is revealed (r = 0.42; p = 0.01). It is received that children early separated from a breast had incresed risk of hyperoxaluriya by 1.4 times (p =0.003). Children who separated from a breast up to 6 months had excretion of oxalates of urine 2.90 ± 0.24 and after 6 months — 2.33 ± 0.21 mg/kg/days. Children who received meat up to 6 months had a statistically significantly higher daily excretion of uric acid (2.91 ± 0.49 and 1.78 ± 0.16 respectively, t = 2.605; p = 0.01), and uric acid/creatinine and calcium/creatinine index too.
Conclusions: Character of food on 1 year of life has significant effect on a functional condition of kidneys. The early end of the breastfeeding and start of a meat feeding up has adverse effect.

P-356 OBTAINING GROWTH DATA USING HOSPITAL INFORMATION SYSTEMS

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Introduction: CKD is associated with significant morbidity, and mortality. A poor nutritional state can adversely affect its progression, and be reflected in anthropometric measures. Such measurements are poorly characterised outside those with the most severe disease, despite data being recorded and essential to identifying early changes to improve clinical outcome. The aim was to explore the feasibility of producing a report on the prevalence of under and over nutrition in children with CKD. The objectives to use the electronic systems to obtain data and explore relationships with kidney function.
Material and methods: Children were identified using clinic codes. The first appointment recorded for weight, height, BMI and creatinine was used. Data was electronically transferred onto a specifically designed database and used to generate z scores and estimated glomerular filtration rate (eGFR). The prevalence of under and over nutrition was estimated, and the relationship with kidney function explored.
Results: 819 children were included. Weight and height were lower in the end stage programme (CKD stage 4–5) compared to general nephrology clinics (CKD2–3) (p = <0.001). Stunting was more pronounced in the transplant clinics, and under-nutrition in the low clearance clinic. Stunting, under-nutrition and over-nutrition were found across all clinics. As the severity of disease worsened, weight and height decreased (p = <0.001) adjusted for age and group. The younger children had lower weight and height sds (p = <0.001).
Conclusions: Poor nutrition (under and over) starts early in disease and becomes worse as disease progresses. Electronic systems can be used to identify those at risk from poor nutritional status and to characterize a clinical population, and provide larger cohorts have manual reviews. The ability to access this data annually has the potential to be a powerful tool to early identification of those that may not be thought to be at risk and to target preventative clinical care as well as end stage disease.

P-357 HEALTH-RELATED QUALITY OF LIFE IN CHILDREN WITH CHRONIC KIDNEY DISEASE

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Introduction: Health-related quality of life (HRQoL) measures a patient's broader health status.
Material and methods: Families of children with Chronic Kidney Disease (CKD) were asked to complete the self-reporting and parent-proxy components of the Paediatric Quality of life Inventory (version 4.0 – English (U.K.)).
Results: 60 families that were recruited (46 conservatively managed). Overall scores from this cohort were lower than the previously published control data, but conservatively managed patient scores were similar to healthy controls, and higher than previously published cohorts from Brazil, Canada and the USA. There was no correlation with estimated glomerular filtration rate (eGFR) or duration of illness and any domain score. Disparity was found in the emotional domain between self-reported and parent-proxy questionnaires ($p = 0.005$), but on subgroup analysis by age-group, only remained in those aged 5–7 years ($p = 0.046$). Analysis of only those receiving conservative management demonstrated a similar pattern. Ten children had received functioning renal transplant. Comparison between those that had a renal transplant and those that did not, did not demonstrate any difference in scores in the self-rated questionnaire, but demonstrated lower parent-proxy scores in those with a graft.
Conclusions: These UK data were not concordant with previously published PedsQL scores in conservatively managed cohorts. The reasons for this need exploration, but may be due to populations receiving different healthcare services, and/or the small, relatively homogenous population of this cohort. Parents tended to score their child lower than the child themselves in the emotional domain. This data contradicts previous literature that suggests that discordance should increase with the age. HRQoL tools have the potential to be used to evaluate services, as screening tools, and to identify areas to be explored during consultation. HRQoL assessment should be considered for introduction into clinical care for ongoing holistic care of children with chronic illnesses, including CKD.

P-358 HEALTH-RELATED QUALITY OF LIFE IN CHILDREN WITH CHRONIC KIDNEY DISEASE, COMPARISONS BETWEEN PARENT AND CHILD REPORTS

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Introduction: We aimed to assess health related quality of life (HRQoL) in children with chronic kidney disease (CKD) or a kidney transplant (CKD-T) from the perspective of their parents (proxy ratings). Additionally, the parental life satisfaction was evaluated as well as its possible associations with HRQoL of their children.
Material and methods: Sixty parents to children with CKD stage 3–5 or CKD-T participated. HRQoL in children was assessed by parent proxy versions of the generic instruments Kidscreen-27 and Disabkids-37. Parents own life satisfaction was measured by self-reported LiSat-11questionnaire.
Results: In most areas parent proxy ratings were significantly lower than ratings by the children themselves. Female sex and older age were associated with lower HRQoL. Compared with proxy ratings by parents to children in the general population, proxy ratings of HRQoL in children with CKD and CKD-T were significantly lower in the domains Physical Well-being, Psychological Well-being, Social inclusion, Social exclusion, and in overall score. Compared with general population, parents in the study rated their own life satisfaction lower in the domains Life as a whole, Leisure and Contacts. Mothers' life satisfaction were lower than fathers' in domains Life as a whole and Leisure.
Conclusions: The agreement between parent and child reports of HRQoL was generally poor. Parent ratings of HRQoL in children with CKD seemed to correlate with parents own life satisfaction. The differences between parent and child ratings should be considered in clinical practice.

P-359 PATIENTS EXPERINCE OF TRANSPLANT

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Introduction: As Clinical Psychologists who have recently joined the paediatric renal team at the Royal Hospital for Children we want to get a clearer understanding of the patients journey through their kidney transplant. We are currently sending questionnaires, devised by the authors to all children and their families who have undergone a kidney transplant in the last 5 year (approx. $n = 60$). We are looking at their experiences pre, during and post transplant in relation to the psychological support they received and require. Drawing on themes from the questionnaires we then plan to interview 3–4 families about their experiences.
Material and methods: Questionnaire devised by authors and interviews with 3–4 families using interpretative phenomenological analysis.
Results: Not known yet.
Conclusions: We hope that the findings will enable us to revise the support Clinical Psychology offer to children and families undergoing kidney transplants at the RHC in Glasgow.

P-360 ANALYSIS OF CILIATED PODOCYTES DURING NORMAL HUMAN GLOMERULAR DEVELOPMENT AND IN NEPHROTIC KIDNEYS

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Introduction: Primary cilia are non-motile sensory organelles involved in the control of different cellular processes, including cell differentiation. The role of primary cilia has been implicated in numerous cystic kidney diseases. By now, primary cilia have been detected on the surfaces of tubular cells and collecting ducts in developing and postnatal human kidneys. In the present study we analyze appearance and percentage of primary cilia on the surfaces of developing human healthy and nephrotic podocytes.
Material and methods: Kidney tissue were dissected from human conceptuses 8th–38th postovulatory week old, postnatal healthy and nephrotic syndrome of the Finnish type (CNF) kidneys. They were stained by double immunofluorescent technique, using α-tubulin as primary cilia marker and DAPI as nuclear marker. Cilia were counted on cell surfaces and their percentage was calculated

using Kruskal–Wallis test, followed by Dunns post-hoc test. Ultrastructural characteristics of podocytes were analyzed by transmission electron microscopy.
Results: Between the 8th and 10th developmental week, population of metanephric cup cells contained 24% of ciliated cells, immature glomeruli 47% and Bownam's capsule cells (parietal podocytes) 26%. Percentage of ciliated podocytes decreased to 10% in the 38th week and then to 3% in postnatal period. Number of ciliated cells in Bowman's capsule first increased to 32% by the 22nd developmental week, and postnatally decreased to 16%. Compared to healthy kidneys, CNF podocytes showed increased number of ciliated podocytes (12%) and Bowman's capsule cells (19%).
Conclusions: Prospective human podocytes contain primary cilia already at the metanephric cup stage. During development, number of ciliated podocytes decreases in accordance with podocyte differentiation, while Bowman's capsule cells retain higher percentage of ciliated cell, thus indicating their potential in podocyte regeneration. Compared to healthy kidneys, both cell populations of CNF kidneys display higher percentage of primary cilia, pointing to their incomplete maturation.

P-361 EXPRESSION OF CONNEXINS CX 43 AND CX45 IN DEVELOPING HUMAN KIDNEYS, POSTNATAL HEALTHY KIDNEYS AND IN CONGENITAL NEPHROTIC SYNDROME OF THE FINNISH TYPE (CNF)

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Introduction: Connexins are transmembrane proteins that either form gap junctions, enabling cell-to cell communications, or form unpaired channels that allow communication between the cell and extracellular environment. In animals, Cx43 and Cx45 are found in renal vasculature, while Cx45 in vascular muscle wall and in glomeruli, including mesangial cells and podocytes. Cx43 expression decreases during epithelial – to mesenchymal transition, while increases in chronic renal diseases. Here, we investigate changes in expression and distribution of Cx43 and Cx45 during human nephrogenesis, in healthy and nephrotic kidneys.
Material and methods: Human kidney tissue was dissected from conceptuses 8th to 38th postovulatory weeks old, from postnatal healthy kidneys as well as from the CNF kidneys. Double immunofluorescence technique was used to detect expression and localization of Cx43 and Cx45, combined with DAPI nuclear stain. Their distribution and intensity of expression was presented semiquantitatively.
Results: In the metanephric cup stage moderate expression of Cx45 characterized all cup cells; weak Cx43 expression characterized mesenchymal cells corresponding to developing vasculature. During progression of development, Cx45 expression increased in all glomerular cells (visceral and parietal podocytes and mesangial cells), while Cx43 expression was restricted to vascular (endothelial) cells and parietal podocytes. Postnatally, Cx45 was observed throughout glomeruli; Cx43 was observed in vascular cells and parietal podocytes. Stronger and changed expression of Cx45 and Cx43 characterizes CNF glomeruli. Co-expression of Cx43/Cx45 was observed in some glomerular cells and parietal podocytes.
Conclusions: During human kidney development Cx43 predominantly appears in the endothelium of blood vessels indicating its role in vasculogenesis. Cx45 is expressed during the whole nephrogenesis, pointing to importance of cell-to-cell communication in differentiation of podocytes and mesangial cells. Changed expression of Cx45/Cx43 in CNF kidneys imply interference of defective cells signaling with chronic kidney disease.

P-362 THE STAGES OF MORPHOLOGICAL CHANGES AT THE DEVELOPMENT OF NEPHROSCLEROSIS IN CHILDREN WITH OBSTRUCTIVE UROPATHY

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Introduction: Despite the variety of pathogenetic mechanisms of damage of renal tissue, the consequences of all these processes lead up to the formation of nephrosclerosis with the development of renal failure. The purpose of the study is to develop an algorithm for early diagnosis of the extent of kidney damage based on data of correlation analysis of morphometric changes.
Material and methods: Material for the research included 29 specimens of renal tissue of children. Seventeen of them were obtained using in vivo biopsies and 12 autopsies from children suffering from kidney diseases at the age from 3 months to 16 years. All the children suffered from obstructive uropathy, the duration of illness in 70% of them were more than a year. At the statistical processing of the received materials the package of applied programs Statistica 10.0 in Windows 2010 are used. To study the correlation between pairs of symptoms using the Pearsons correlation (r) was performed.
Results: When analyzing the results of a study of the morphological samples, the criteria for changing the tissue structure of a particular area are determined, reflecting the degree and severity of the renal parenchyma lesions. Early indicators of renal parenchyma damage include: the presence of degeneration and/or tubular atrophy, lymphohistiocytic infiltration, angiomatosis of blood vessels. To more pronounced changes in the kidney tissue, to the early changes included: thinning of the parenchyma and thickening of the blood vessels walls. And the most severe pathological changes is the progression of atrophy, sclerosis of tubules, glomeruli, renal vessels with impaired cortical and medulla differentiation of the kidney substance. Statistical analysis are revealed correlations of different strength and direction of the morphometric criteria of nephrosclerosis with the degree of development of degenerative changes of glomeruli, tubules, vessels with outcome in sclerosis. Comparing the data of the statistical study of correlation and morphometric changes, there are 3 successive stages of development of nephrosclerosis.
Conclusions: Statistics in medicine is one of the tools for the evaluation of experimental data and clinical observations. On the basis of statistical analysis the classification of nephrosclerosis stages are proposed with the indication the extent and depth of the renal lesions at the histomorphological study. The statistically significant criteria nephrosclerotic changes in the kidneys are allocated.

P-363 RABBIT ANTI-HUMAN THYMOCYTE IMMUNO GLOBULIN FOR RESCUE TREATMENT OF CHRONIC ANTIBODY-MEDIATED REJECTION AFTER PEDIATRIC KIDNEY TRANSPLANTATION

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Introduction: Chronic antibody-mediated rejection (cAMR) is the leading cause of late kidney graft loss but current therapies are often ineffective. Rabbit anti-human thymocyte immunoglobulin (rATG) may be helpful but its use is virtually undocumented.
Material and methods: Nine pediatric kidney transplant patients with cAMR were treated with rATG (1.5 mg/kg × 5 days) at our center after non-response to pulsed prednisolone, intravenous immunoglobulin, rituximab and increased immunosuppressive intensity (including switch to belatacept in some cases), with or without bortezomib.
Results: The median time from diagnosis to cAMR was 179 days. rATG was started 5–741 days after diagnosis. Median estimated GFR increased from 40 mL/min/1.73m^2 when rATG was started to 62 mL/min/1.73m^{2}nine months later ($p = 0.039$). Four patients showed substantially higher eGFR after nine months and two patients showed a small improvement; eGFR continued to decline in three patients after starting rATG. No grafts were lost during follow-up. At last follow-up DSA were no longer detectable, or median fluorescence intensity had decreased, in 7/8 patients for whom data were available. No adverse events with a suspected relation to rATG, including allergic reactions, leukocytopenia or infections, were observed in any patient.
Conclusions: In conclusion, in this small series of patients rATG appears a promising treatment for unresponsive cAMR. Further evaluation, including earlier introduction of rATG, is warranted.

P-364 BLOOD PRESSURE MONITORING AND TREATMENT POST PAEDIATRIC RENAL TRANSPLANTATION: A LONGITUDINAL STUDY OF PAEDIATRIC PATIENTS IN SCOTLAND

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Introduction: Hypertension is a recognised complication of renal transplantation and is common in patients with end stage renal failure (ESRF). Renal transplantation is recognised to produce significant survival benefits in patients with ESRF. Hypertension in the ESRF population is a known risk factor for the development of cardiovascular disease and is associated with poorer graft function.
Material and methods: A retrospective review was undertaken of all paediatric patients receiving a renal transplant in Scotland from January 2001 to July 2014. During this period 119 patients were identified, aged 2.4 years to 20.3 years at time of transplantation. Data gathered included demographics, diagnoses, blood results, blood pressure and BMI (body mass index) over time.
Results: 23 patients had pre-emptive transplants, 34 were on haemodialysis and 62 peritoneal dialysis prior to transplantation. On long term follow up 4 patients were deceased, 2 had moved out of area, 108 were in transplant clinic follow up and 5 were on haemodialysis. Our results demonstrate that post-transplant 15–39% of our patients had systolic blood pressures (SBP) of over 95th Centile during the 5 year follow up period, with a peak at 30 days post transplantation. During this period there were 17.6% to 31.0% of patients recorded to be receiving antihypertensive medication. The estimated glomerular filtration rate (eGFR) at the different time points was recorded and whether antihypertensives were prescribed at these time points. Results showed that patients receiving antihypertensives tended to have higher eGFR, however this was not statistically significant.
Conclusions: Our data demonstrates a peak of SPB 30 days post transplant which is in keeping with previous data. However overall our study population demonstrated a much lower level of hypertension than previous studies, which suggest 60–90% of patients are hypertensive after renal transplant. Interestingly we found that patients on antihypertensive treatment tended to have longer time with eGFR greater than 30 ml/min/1.73 m2, than those not receiving antihypertensives. This result was unexpected as we would expect those with hypertension to have poorer renal function. This positive association with antihypertensives and increased GFR may be a surrogate marker for compliance. An alternative explanation may be that we need to be more aggressive with hypertension management post transplantation aiming for a lower SBP.

P-365 PREDICTORS OF POST-TRANSPLANT GROWTH IN PREPUBERTAL CHILDREN WITH END-STAGE CKD

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Introduction: Disproportionate short stature is a frequent complication of CKD. Here we evaluated post-transplant growth during childhood in renal allograft recipients who were enrolled in the CKD Growth and Development observational cohort study.
Material and methods: Linear growth (height, leg length, and sitting height) was prospectively investigated in a cohort 207 renal transplant recipients aged 2–12 years with a mean follow-up of 4.3 years. Predictors of growth outcome after transplantation were evaluated by the use of linear mixed-effect models.
Results: Pre-transplant mean SD scores (SDS) for all linear body dimensions were reduced (height, −2.32 SDS; sitting height, −1.49 SDS; leg length, −2.43 SDS), and sitting height index was increased (1.33 SDS) compared to healthy children (each $p < 0.001$), indicating disproportionate short stature. After transplantation mean standardized height continuously increased until the age of 12 years (−1.38 SDS, $p < 0.01$ vs. pre-transplant). Leg length was the anthropometric parameter with the greatest plasticity after transplantation. The preferential stimulation of leg growth resulted in normalization of sitting height index by puberty in most patients. Beside living-related transplantation, use of growth hormone in the pre-transplant period, steroid exposure, and hemoglobin levels were identified as potentially modifiable significant predictors of post-transplant growth. Growth hormone treatment before transplantation showed the highest impact on stature and sitting height, whereas transplantation at a young age (< 5 years) showed the strongest association with leg growth and consequently the degree of body disproportion.
Conclusions: Renal transplantation during childhood results in normalization of height in the vast majority of patients. Use of growth hormone in the pre-transplant period, living-related transplantation, transplantation at a young age, reduction of steroid exposure, and better control of anemia are promising measures further improving growth outcome.

P-366 POST-KIDNEY TRANSPLANT ANNUAL PROTOCOL BIOPSIES FOR FIVE YEARS IN CHILDREN

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Introduction: Protocol biopsy (PB) is an appropriate method for the early diagnosis of graft disorders. We aimed to evaluate the effectiveness and safety of annual BPs.

Material and methods: Biopsies perfomed annually for 5 years in kidney transplant recipient children between October 2007–April 2017 were evaluated retrospectively. Gender, transplantation age, donor types, mismatches, treatment modalities, follow-up durations were recorded. Indication biopsies(IB), inappropriate samples(IS,<7 glomerulus) and unaccesible materials(UM) were excluded. All biopsies were scored (0–3) for peritubular capillaritis, glomerulitis, C4d staining, interstitial inflammation, tubulitis, intimal arteritis, vascular intimal fibrosis and arterial hyalinosis. Sum of the scores were notes as "Banff score(BS)". Complications, interventions, concurrent serum creatinine, glomerular filtration rates(GFR) and proteinuria levels were recorded and their relation with BS was evaluated.
Results: A total of 205 biopsies were performed in 42 patients (M/F: 22/20) between above-mentioned dates. When IBs(n:45), ISs(N:26) and UMs(n: 16) were excluded, there were 118 PBs (29 implantation, 89 follow-up). Forty of the patients received basiliximab for induction and 38 received steroid + tacrolimus + mycophenolate for initial maintenance therapy. The number of follow-up biopsies at 5th-year,4th-year,3rd-year,2nd-year and 1st-year were 7,16,16,20 and 30, respectively. Abnormalities were found in 10(%8.5) of the PBs (bordeline: 6, subclinical rejection: 4) and an intervention was made in 4 of them. Negative correlations between GFR and BS were significant in 4th and 5th year biopsies (p: 0.003,r: −0.685;p:0.004,r:-0.912,respectively). The rates of macroscopic hematuria (%10.4), perinefric hematoma (%4.3), arteriovenous fistula AVF(%1.7), transfusion(non) and radiological intervention (%0.8) were acceptable. There is no patient lost, 1 graft lost due to nonadherence and 2 patients with serum creatinine > 2.5 mg/dL due to chronic allograft nephropathy all through the follow-up duration.
Conclusions: The treatment protocol and follow-up with annual protocol biopsies seems efficient and safe in children. Better results may be attained when all our patients complete the five-year period.

P-367 OUR EXPERIENCE IN EN-BLOC KIDNEY TRANSPLANTATION INCLUDING PARTIAL BLADDER PATCH TECHNIC PATIENTS, INTO CHILDREN RECIPIENTS

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Introduction: The concept of en-bloc kidney transplantation was born due to high discard rate of young donor patients kidneys (5-year-old,<15 kg in weight, 6 cm in kidney diameters).Despite improved outcomes are reported in pediatric kidney transplantation, literature is still limited about en-bloc kidney transplantation with/without partial bladder segment.
Material and methods: This retrospective study was conducted in 4 children who underwent en-bloc kidney transplantation with/without partial bladder segment between 2005 and 2016. Baseline donor, patient and transplant features and also, protocol biopsy results were collected.
Results: There are totally 71 pediatric kidney transplantation was performed (en-bloc kidney transplantation in 4 patients (5.6%)). The mean donor age and weight are 8 months and 9.5 kg, respectively. Recipient number 4 is in the 3rd month of follow-up. The mean age at the transplantation and body weight were 8 years and 20.9 kg,respectively. En-bloc kidney transplantation was applied with partial bladder segment in two. There was only one renal artery thrombosis due to organ displacement required unilateral donor nephrectomy (recipient 4). Five years of follow-up serum creatinine, eGFR and urinary protein excretion of 3 patients were in normal range. There were no biopsy proven rejection or chronic allograft nephropathy detected in protocol biopsies. Also, growth of these graft pairs were detected in routine ultrasonographic imaging.
Conclusions: In the setting of inadequate donor pool, for maximize of organ usage en-bloc kidney transplantation seems suitable approachement. The outcomes of en-bloc kidney transplantation in adults are same as an ideal kidney transplantation. To delinate effectiveness of en-bloc kidney transplantation into children further studies are needed.

P-368 RELATIONSHIP BETWEEN RECIPIENT'S AGE AT TRANSPLANT AND CHANGE OF ESTIMATED GLOMERULAR FILTRATION RATE AFTER PEDIATRIC KIDNEY TRANSPLANTATION

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Introduction: In Japan, generally donors for pediatric kidney transplantation are adults, and have mismatch of body size with the recipients. However, the impact of recipient's age on transplant outcomes remains unclear. We investigated short-term outcome of estimated glomerular filtration rate (e GFR) in children after kidney transplantation based on their age at transplant.
Material and methods: We retrospectively reviewed the charts of pediatric living donor kidney transplant recipients who were transplanted adult kidney in our department from March 2009 to October 2014. Forty two recipients were followed over two years. These recipients were divided into 3 groups based on the age at transplant: Low (2–5 years, $N = 22$), median (6–10 years, $N = 19$), and high (11–15 years, $N = 18$) age group. We compared the median eGFR of each age group at 1 month (P1), 4 months (P2), 12 months (P3), and 24 months (P4) after transplantation. The mean age and eGFR of donors at transplant were 41.5 ± 7.9 years and 79.7 ± 15.2 ml/min/1.73 m2, respectively.
Results: eGFR was calculated using a creatinine-based equation for Japanese children and adolescents aged 2–18 years. The changes of eGFR (ml/min/1.73 m2) in each age group were shown as follows:
Low (P1, P2, P3, P4): 92.3 ± 18.9, 78.1 ± 18.6, 69.7 ± 16.8, 68.1 ± 17.3.
Median: 84.0 ± 16.5, 72.7 ± 13.9, 67.0 ± 13.0, 64.6 ± 12.5.
High: 77.0 ± 18.7, 67.7 ± 16.1, 64.2 ± 16.0, 64.1 ± 14.4.
The changes of eGFR between points were examined by paired *t*-test.
Low: P1-P2 (P<0.01), P2-P3 (P<0.01), P3-P4 ($P = 0.51$).
Median: P1-P2 (P<0.01), P2-P3 (P<0.01), P3-P4 ($P = 0.38$).
High: P1-P2(P<0.01), P2-P3($P = 0.125$), P3-P4($P = 0.97$).
Conclusions: In the first two years after pediatric kidney transplantation, eGFR of the recipients deteriorated over time regardless of age. Furthermore, the slope of eGFR decline tended to be larger in younger age group and earlier period of post-transplant.

P-369 MULTI-DRUG RESISTANT CMV VIREMIA SECONDARY TO A RARE VIRAL MUTATION IN PAEDIATRIC KIDNEY TRANSPLANT RECIPIENT SUCCESSFULLY TREATED WITH ANTI-PROLIFERATIVE CESSATION AND IMMUNOGLOBULINS

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Introduction: International Transplant Society for CMV Consensus Group advises ganciclovir, valganciclovir and foscarnet as treatment for CMV in transplant recipients. Viral DNA polymerase resistance mutations are rare but may be associated with cross resistance. We report a rare CMV mutation associated with resistance to all available medications in a CMV seronegative recipient treated with immunosuppression dose reduction and IVIG.

Material and methods: Retrospective data analysis from medical records.
Results: Sixteen year old girl with ESRD due to Alport Syndrome received deceased donor kidney transplant (mismatch 120, CMV Donor+/Recipient-, EBV Donor−/Recipient+). She received basilixumab on induction and her maintanance immunosuppression included tacrolimus, azathioprine and prednisolone. As per our centre protocol, she was started on valganciclovir prophylaxis for 90 days. Two months post-transplant, whilst on valganciclovir prophylaxis, she developed CMV viremia. Viral loads continued to rise (highest log 5.41) despite the treatment dose intravenous ganciclovir. She was found to have a rare CMV UL 54 mutation (POL gene deletion 981/982), associated with multidrug resistance. She had no evidence of CMV disease and stable graft function, and was managed with cessation of azathioprine and intravenous immunoglobulins (IVIG). CMV IgM was positive at 4 months, with IgG seroconversion demonstrated at 9 months post transplant with CMV PCR not detected from month 9 onwards. She developed borderline T cell mediated rejection successfully treated with high dose of oral prednisolone. Eighteen months post transplant, patient is well with stable graft function (estimated GFR 69 ml/min/1.73 m2) on CNI inhibitor and daily prednisolone only. Donor specific antibodies are not detected. To date, cytotoxic T cell response to CMV is negative but CMV remains undetectable.
Conclusions: We report a case of multidrug resistant CMV viremia in a transplant recipient with primary CMV infection, managed with IVIG and cessation of anti-proliferative agent only with excellent patient outcome including preservation of good graft function.

P-370 MEDICATION NON-ADHERENCE IS A LEADING MODIFIABLE CAUSE FOR RENAL GRAFT LOSS IN CHILDHOOD – A LARGE CENTRE STUDY

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Introduction: Despite continued advances in transplantation, it has been observed that the rate of renal graft failure is disproportionately high amongst the adolescent population. This study looked into why grafts fail in children.
Material and methods: Retrospective observational study in a single paediatric transplant centre between 2003 and 2016. All patients transplanted during study period and those transplanted previously and followed up during the study period were included.
Results: During the study period, 171 paediatric kidney transplants were performed. Median follow up was 8 years (IQR 10 years). Fifteen grafts failed before adulthood. Graft loss was caused by recurrent acute rejections following medication non-adherence in four patients (27%) and chronic antibody mediated rejection (CAMR) in five patients (33%). The mean age at time of transplant was 6.4 years and average age at the time of graft loss 15.7 years (range 2–17.9). Living and deceased donors were evenly distributed and well matched. HLA antibodies were detected in 70%. There was no statistically significant difference in graft longevity between the CAMR group and the medication non-adherence group ($p = 0.07$). The medication non-adherence group were 12–17 years old at the time of graft failure. All were well matched (MM 110/111) with graft lifespan 21–120 months. All had DSA, multiple episodes of rejection (Banff 2b and 4a), with an average of five biopsy proven episodes each and low/undetectable CNI levels.
Conclusions: Medication non-adherence was a significant contributor to poor transplant outcomes in the adolescent population. We propose a multidisciplinary staged adherence pathway to improve graft outcomes for paediatric recipients. This encompasses early identification of vulnerable patients, enrolment into a hospital passport program overseen by play specialists, MDT meetings, adherence workshops and early psychology and psychiatry referral pathways for vulnerable patients.

P-371 RITUXIMAB AND PLASMA EXCHANGE DOES NOT PREVENT DISEASE RECURRENCE IN HIGH RISK FSGS FOLLOWING LIVING DONOR TRANSPLANTATION

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Introduction: For patients with end stage renal failure (ESRF) secondary to focal segmental glomerulosclerosis (FSGS), disease recurrence (DR) following renal transplantation (RT) is a significant concern. Patients with rapidly progressive primary disease, negative genetic screening and those with previous graft loss secondary to DR are most at risk. There is no consensus regarding the prevention and management of DR.
Material and methods: Four children with high risk FSGS, including 2 with previous graft loss due to DR, underwent living donor RT. All were managed with a uniform protocol of a single dose of rituximab (375 mg/m2) 4 weeks prior to RT and 4 sessions of plasma exchange (PE) over the week prior to RT. DR was defined as urine protein:creatinine ratio > 200 mg/mmol on 2 consecutive days.
Results: One child with previous graft loss (Pt 1) had immediate DR and received PE for 5 months post-transplant. Another had immediate DR as well as impaired graft function and following a partial response to PE, developed graft failure at 6 months. The third, who had previous graft loss (Pt 3) had DR at day 4 which responded to 5 sessions of PE. The fourth child had a comparatively late DR at 1 month and has continued on weekly PE 7 months.

Pt no	1	2	3	4
Sex	M	F	F	M
Immunosuppression	Ba/T/MMF	Ba/T/MMF	Ba/T/MMF/P	Ba/T/MMF
Age at Tx	16	14	12	6
Previous Tx	Y	N	Y	N
Time to recurrence	3 days	3 days	4 days	30 days
No of PE sessions	31	52	5	20
Duration of FU	24mo	11mo	8mo	7mo
Last urine PCR	164	585	22	43
Current eGFR	54	<15	138	90
On ACEi/ARB	Y	Y	N	N

Conclusions: In this small cohort of patients with high risk FSGS, rituximab and PE pre-RT, did not reduce the risk of DR. However, in children with DR, including 2 patients with previous graft loss, the disease responded well to PE. We are encouraged by our data to continue advocating the use of living donors for this difficult group of patients.

P-372 PRESERVATION OF RENAL FUNCTION WITH EVEROLIMUS PLUS REDUCED-DOSE TACROLIMUS AND CORTICOSTEROID WITHDRAWAL-BASED REGIMEN IN DE NOVO PEDIATRIC RENAL TRANSPLANT RECIPIENTS: 12-MONTH RESULTS FROM THE CRADLE STUDY

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Introduction: CRADLE (NCT01544491) study evaluated the efficacy and safety of everolimus with reduced-dose tacrolimus (EVR + rTAC) and early corticosteroid (CS) withdrawal regimen in pediatric renal transplant recipients (pRTxR). Here, we report the 12-month (M) renal function outcomes from the study.

Material and methods: In this 12 M core + 24 M follow-up, phase III, multicenter, open-label study, pRTxR (≥1- < 18 years) on mycophenolate mofetil (MMF) + standard-dose TAC (sTAC) + CS were randomized (1:1) at 4–6 weeks post-Tx to EVR + rTAC with CS withdrawal at 6 M (EVR C_0:3-8 ng/mL; TAC C_0:randomization [RND]-M3, 4-6 ng/mL; from M4, 2-4 ng/mL); or sTAC + MMF + CS (TAC C_0:RND-M3, 7-10 ng/mL; from M4, 5-8 ng/mL). Co-primary objectives were to evaluate renal function (eGFR; updated Schwartz) and composite efficacy failure (biopsy-proven acute rejection [BPAR], graft loss, or death) at M12. Other objectives included overall safety.

Results: Of 106 (EVR + rTAC, *N* = 52 and MMF + sTAC, *N* = 54) patients randomized, 65.4% in EVR + rTAC arm and 87.0% in MMF + sTAC arm completed study treatment. M12 adherence to TAC target C_0 was 50% in EVR + rTAC and 51.1% in MMF + sTAC arms; 38.9% patients in EVR + rTAC arm were above TAC target C_0 and 26.7% in MMF + sTAC arm were below target C_0 at M12. Mean eGFR was comparable between arms (**Table 1**). From RND to M12, mean eGFR numerically increased in EVR + rTAC arm, whereas it decreased in MMF + sTAC arm. Rates of BPAR were comparable between arms (**Table 2**) with 100% renal graft and patient survival. Overall safety including renal adverse events (AE; EVR + rTAC, 15.4% vs MMF + sTAC, 18.5%) were similar between arms. AE leading to study drug discontinuation were higher in EVR + rTAC vs MMF + sTAC arm. Most patients (EVR + rTAC, 72.5% vs MMF + sTAC, 78.3%) had mildly increased urinary protein:creatinine ratio at M12. One patient in EVR + rTAC arm discontinued study treatment due to proteinuria.

Conclusions: Despite poor adherence to TAC C_0, EVR + rTAC and CS withdrawal regimen preserved renal function while maintaining efficacy and safety in pRTxR up to 12 M post-Tx.

Table 1: Summary of renal function

	RND	Month 12			
	EVR + rTAC *N* = 52	MMF + sTAC *N* = 54	EVR + rTAC *N* = 52	MMF + sTAC *N* = 54	
Renal function, FAS					
eGFR (updated Schwartz formula), mL/min/1.73 m^2	*n* = 33	*n* = 32	*n* = 44	*n* = 48	Treatment difference (80% CI), *P* value*
eGFR, mean ± SE	69.4 ± 3.9	84.8 ± 7.5	76.2 ± 4.1	72.5 ± 3.6	3.8 ± 5.4 (−3.3, 10.8) 0.4899[a]
eGFR, median [range]	66.0 [32–112]	75.5 [37–264]	75.5 [31–147]	66.5 [23–136]	
	–	–	***n* = 37**	***n* = 44**	
Change from RND, mean ± SD	–	–	3.0 ± 18.7	−5.0 ± 32.8	
eGFR, (updated Schwartz formula) by CKD stage, mL/min/1.73 m^2, n(%)†	***n* = 33**	***n* = 32**	***n* = 44**	***n* = 48**	ND
< 15 (end stage)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	
15- < 30 (severe)	0 (0.0)	0 (0.0)	0 (0.0)	1 (2.1)	
30- < 60 (moderate)	13 (39.4)	6 (18.8)	15 (34.1)	14 (29.2)	
60- < 90 (mild)	13 (39.4)	14 (43.8)	16 (36.4)	22 (45.8)	
≥ 90 (normal)	7 (21.2)	12 (37.5)	13 (29.5)	11 (22.9)	
Urinary protein:creatinine ratio (mg/mmol), n (%)†	***n* = 30**	***n* = 34**	***n* = 40**	***n* = 46**	ND
< 3.39 (normal)	0 (0.0)	0 (0.0)	0 (0.0)	0 (0.0)	
3.4–33.9 (mild)	17 (56.7)	20 (58.8)	29 (72.5)	36 (78.3)	
34–339 (sub-nephrotic)	12 (40.0)	12 (35.3)	11 (27.5)	10 (21.7)	
> 340 (nephrotic)	1 (3.3)	2 (5.9)	0 (0.0)	0 (0.0)	
Incidence of CAN, n (%)	3 (5.8)	3 (5.6)	11 (21.2)	8 (14.8)	ND

*Two-sided *P* value; [a]t-test, †corresponds to SEP

CAN, chronic allograft nephropathy: CKD, chronic kidney disease; CI, confidence interval; eGFR, estimated glomerular filtration rate; EVR, everolimus; FAS, full analysis set; MMF, mycophenolate mofetil; N, total number of patients in either arm; n, total number of poatients vailable for 12-month analysis; ND, not determined; RND, randomization; rTAC, reduced-exposure TAC; SE, standard error; SEP, study endpoint; sTAC, standard-dose TAC; TAC, tacrolimus

Table 2: Summary of composite efficacy and safety outcomes at month 12

Efficacy failure, FAS [n (%)]	EVR + rTAC *N* = 52	MMF + sTAC *N* = 54	Treatment difference (80% CI)	*P* value*
Composite efficacy failure (BPAR, graft loss, or death)	5 (9.6)	3 (5.6)	4.4 (−2.6, 11.4)	0.4166[a]
BPAR	5 (9.6)	3 (5.6)	4.4 (−2.6, 11.4)	
Graft loss	0 (0.0)	0 (0.0)	0.0 (0.0, 0.0)	
Death	0 (0.0)	0 (0.0)	0.0 (0.0, 0.0)	
Safety, SAS n (%)			**Risk ratio (80% CI)**	
AE/infection	51 (98.1)	50 (92.6)	1.1 (1.0, 1.1)	0.3634[b]
Any serious infection AE/infection	34 (65.4)	31 (57.4)	ND	0.3992[c]
AE leading to study drug discontinuation	13 (25.0)	6 (11.1)	ND	0.0624[c]
Renal AE				
Increased blood creatinine	8 (15.4)	10 (18.5)	0.83 (0.48, 1.45)	

* Two-sided *P* value; [a]Z-test; [b]Fisher's test; [c]Chi square test

AE, adverse events; BPAR, biops-proven acute rejection; CI, confidence interval; EVR, everolimus; FAS, full analysis set; MMF, mycophenolate mofeftil; N, total number of patients in either arm; ND, not determined; rTAC, reduced-dose TAC; SAS, safety analysis set; sTAC, standard-dose TAC; TAC, tacrolimus.

P-373 KIDNEY RE-TRANSPLANTATION DURING CHILDHOOD-FEASIBILITY AND OUTCOMES

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Introduction: Kidney transplantation (Tx) has been increasing in small children. This may lead to more children needing 2nd Tx during childhood. This study looked into the characteristics of renal transplant recipients who underwent more than 1 Tx during childhood.
Material and methods: Single centre retrospective analysis of all paediatric kidney transplants during 2003–2015.
Results: One hundred and seventy one transplants were performed, nine of which were re-transplants (5.3%). At last follow up (median 8 years; IQR 10 years), there was no difference in graft survival for children with a single Tx compared with those re-transplanted ($p = 0.255$). Of the re-transplanted patients, eight had two and one had three transplants (80% deceased donors at 1st Tx; 50% at 2nd Tx). Recurrent acute rejections caused graft failure in four patients, 2 had chronic AMR, 2 thrombosis and one FSGS recurrence. All patients were CMV and EBV naive at 1st Tx. Two patients have previously undergone Tx onto aorta/IVC and had a re-transplant to the same vessels. Five were HLA sensitized, one highly (cRF > 85%). The difference in graft survival at 3 year follow-up between first and second transplant was not significant (98% and 88% respectively; $p = 0.2$). Three patients lost 2nd graft before adulthood due to chronic AMR and BKVAN, one of which received 3rd Tx at the age of 8 years.
Conclusions: The re-transplantation rate is low in this cohort. Despite surgical and immunological challenges kidney re-transplantation in childhood is feasible and with good outcomes; this should be accounted for when consenting and determining the management for young recipients.

P-374 IMPROVED RENAL ALLOGRAFT SURVIVAL FOR PRE-EMPTIVE PAEDIATRIC RENAL TRANSPLANT RECIPIENTS

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Introduction: The aim of this study was to investigate whether being on dialysis (peritoneal dialysis or haemodialysis) at the time of renal transplantation affected patient or renal allograft survival in paediatric renal transplant recipients (pRTR).
Material and methods: Data were obtained from the UK Transplant Registry (NHS Blood and Transplant) on all children (aged <18 years) who received a kidney only transplant between 1 January 2000 and 31 December 2015. Baseline demographic data were collected, including dialysis modality at the time of renal transplantation (none vs peritoneal dialysis vs haemodialysis). Kaplan-Meier estimates of 1 and 5-year patient and renal allograft survival were calculated, as well as Cox regression modelling accounting for donor type.
Results: 2038 pRTR were analysed: 607 (30%) were pre-emptively transplanted, 789 (39%) and 642 (32%) were on peritoneal dialysis and haemodialysis, respectively at the time of transplantation. Five-year renal allograft survival was significantly better in the pre-emptively transplanted group (90.6%) compared to those on peritoneal dialysis and haemodialysis (86.4% and 85.7% respectively; $p = 0.02$). There was no significant difference in 5-year patient survival or in 1-year patient or renal allograft survival across the groups. After accounting for donor type, we found a significantly lower hazard of 5-year renal allograft failure in pre-emptively transplanted children (HR 0.742, $p = 0.05$).
Conclusions: Children who are pre-emptively transplanted have improved 5-year renal allograft survival, compared to children on haemodialysis or peritoneal dialysis at the time of transplantation. This provides further evidence to support attempts to achieve pre-emptive renal transplantation wherever possible in children and avoid dialysis.

P-375 CONTINUOUS MONITORING OF KIDNEY TRANSPLANT PERFUSION WITH NEAR INFRARED SPECTROSCOPY

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Introduction: Current reliance on clinical/ laboratory parameters and doppler ultrasound imaging to detect renal allograft thrombosis results in significant delays in recognizing vascular complications that account for up to 35% of early graft losses in children. Near infrared spectroscopy (NIRS) is a real time, non-invasive technique for monitoring oxygenation percutaneously. Validation of NIRS in real time monitoring of renal allograft perfusion by direct comparison to doppler ultrasound has not been reported. The aim of this pilot study is to determine whether tissue oxygenation indices from NIRS can reliably assess blood flow in established renal transplants.
Material and methods: Paediatric renal transplant recipients (pRTRs) aged 1–18 years with stable allograft function were eligible. Participants underwent routine outpatient renal transplant ultrasound assessment of perfusion as per protocol including resistive index (RI) calculations at upper, and lower poles. NIRS data (Tissue Oxygenation Index- TOI) were recorded for 2 min with two NIRS sensors placed on the skin over the transplant poles as previously marked from ultrasound.
Results: Thirteen pRTRs with median age 16 years (range 5.3 to 17.8) and median allograft age 44.7 months (range 6.4 to 148.7) were recruited. Fifty- 4 % were male and 69% living donor pRTRs. NIRS monitoring was well tolerated by all participants with a 96–100% rate of valid measurements. Significant negative correlations were observed between TOI recorded with NIRS, and RI from doppler ultrasound for both upper and lower poles ($r = -0.7$ for both, $p = 0.01$ and 0.02 respectively). TOI measured with NIRS was a significant predictor of doppler ultrasound RI in adjusted linear regression models for upper and lower pole ($p = 0.01$ and 0.02 respectively).
Conclusions: NIRS indices correlate well with doppler ultrasound graft perfusion parameters in established pRTRs. Further studies are warranted to extend NIRS use for continuous real-time monitoring of early post-transplant vascular complications.

P-376 CLINICAL RISK STRATIFICATION OF PAEDIATRIC RENAL TRANSPLANT RECIPIENTS USING C1Q AND C3D FIXING OF DE NOVO DONOR SPECIFIC ANTIBODIES

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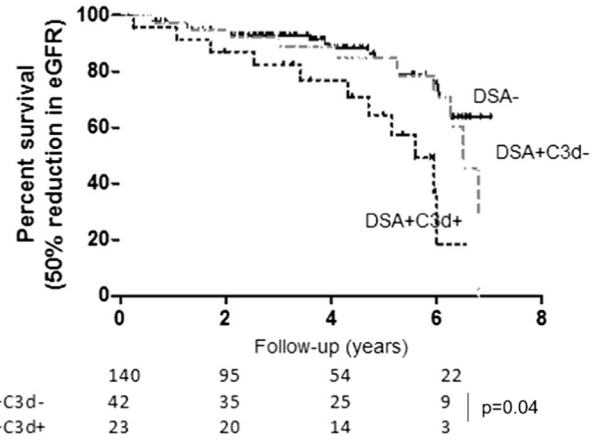
Introduction: We previously showed that children who developed *de novo* donor specific HLA antibodies (DSA) had greater decline in allograft function. We hypothesised that patients with complement activating DSA would have poorer renal allograft outcomes.
Material and methods: 75 children developed DSA in the original study. The first positive DSA sample was subsequently tested for C1q and C3d fixing. Primary event was defined as 50% reduction from baseline estimated glomerular filtration rate (eGFR) and was analysed using Kaplan-Meier estimator. Serial DSA positive samples were subsequently tested for C3d fixing and MFI results were correlated with eGFR using multi-level models.
Results: Of 65 patients tested, 32 (49%) and 23 (35%) were positive for C1q and C3d fixing respectively. 13/32 (41%) C1q positive patients did not show concomitant C3d fixing. MFI values of original IgG DSA correlated poorly with complement fixing positivity [C1q: adjusted $R^2 = 0.072$; C3d: adjusted $R^2 = 0.11$, $p < 0.05$]. C1q + antibodies were associated with acute tubulitis [C1q + 0.75(± 0.18) v C1q- 0.25(± 0.08) episodes per patient, $p < 0.05$] but not worse long-term renal allograft dysfunction [median time to primary event C1q + 5.9 v C1q- 6.4 years, $p = 0.58$]. C3d + antibodies were associated with positive C4d histological staining [C3d + 47% v C3d- 20%, $p = 0.04$] and significantly worse long-term allograft dysfunction [median time to primary event C3d + 5.6 v C3d- 6.5 years, $p = 0.04$]. Changes in C3d MFI did not correlate with eGFR over time.



Conclusions: Assessment of C3d fixing as part of prospective HLA monitoring can potentially aid stratification of patients at highest risk of long-term renal allograft dysfunction.

P-377 BILATERAL NEPHRECTOMIES FOLLOWED BY LIVING RELATED DONOR RENAL TRANSPLANTATION IN A 2 YEAR OLD CHILD WITH INTRA-RENAL ANEURYSMAL FIBROMUSCULAR DYSPLASIA

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Introduction: To report the successful outcome of a staged surgical approach with bilateral native nephrectomies, haemodialysis and living related donor (LRD) renal transplantation in an infant with bilateral intra-renal aneurysmal disease secondary to fibromuscular dysplasia (FMD) to avert the life-threatening risk of aneurysmal rupture.
Material and methods: Retrospective 12 month post-transplant follow up data of a 2 year old boy with FMD who presented at 7 months of age with macroscopic haematuria (after aneurysmal bleeding) and renovascular hypertension (presenting systolic blood pressure (SBP) of 250 mmHg at 7 months) requiring five anti-hypertensive agents with normal renal function. Multi-disciplinary team discussions led to a planned surgical approach in view of intra-renal aneurysm size (1.5 cm) and the life-threatening risk of rupture. He underwent bilateral native nephrectomies with histology confirming FMD, followed by haemodialysis with reduction to one anti-hypertensive agent. Three months later weighing 12 kg, he proceeded to LRD renal transplant from his mother (mismatch 1,1,0)

with anastomosis of the renal artery and vein onto his aorta and inferior vena cava respectively. He underwent excisions of inferior mesenteric artery and right internal iliac artery aneurysms without complications.
Results: 12 months post-transplant; he is on triple immunosuppression, normotensive with SBP of 104 mmHg and off all anti-hypertensive agents. He has had no biopsy-proven acute rejection nor significant infectious complications and has had no proteinuria nor donor specific antibodies. His renal allograft function remained stable (eGFR of 74mls/min/1.73m^2). A CT angiography at 12 months post-transplantation has shown no further aneurysmal changes but noted changes after ligation of the right internal iliac artery.
Conclusions: This paediatric renal transplant highlights the complexities that clinicians must consider in rare variants with anerysmal disease as part of the spectrum of FMD and is the first paediatric report of a bilateral nephrectomy for this indication.

P-378 POLYOMAVIRUS BK INFECTION IN PEDIATRIC RENAL TRANSPLANT PATIENTS

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Introduction: Polyomavirus BK (BKV) is a common infections and an important cause of graft loss. The purpose of this study was to determine the incidence, clinical features and risk factors of BKV infection in pediatric renal transplant patients in our center.
Material and methods: We retrospectively scanned the data files of 122 patients who had renal transplant. BKV values were recorded measured by quantitative polymerase chain reaction test in urine and serum. Patients with and without Polyoma BK infection were compared to assess risk factors in terms of chronic renal failure etiology, donor types, immunosuppressive treatments, ureteral stent, acute rejection episodes and accompanying viral infection.
Results: BKV infection were detected in 10 patients (8.19%). Ten patients (100%) developed BKV viruri; 7 (70%) BKV viremia; 4 (40%) BKV nephropathy. Acute humoral rejection was present in 3 of BKV nephropathy. Graft loss developed in 2 (1.6%) patients with BKV nephropathy. Patients with BKV infection gender, age of transplantation, donor type, etiology of renal failure, immunosuppressive treatment, ureteral stent, acute rejection were similar to patients without BKV infection. There was no significant difference between first year GFR of patients with BKV infection and those without BKV infection. Accompanying CMV infection were significantly higher in patients with BKV infection ($p = 0.013$). BK virus nephropathy had more acute rejection, ureteral stent, CMV association and higher rate of graft loss than BK viruri or viremia ($p < 0.01$).
Conclusions: The risk of developing graft loss is high in patients with BKV nephropathy. Close monitoring, early detection have a significant effect on the prognosis of polyomavirus BK infection.

P-379 CONVERTING IMMUNOSUPPRESSION FROM PROGRAF® TO MODIGRAF® IN PAEDIATRIC RENAL TRANSPLANT PATIENTS

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Introduction: Adherence to immunosuppression is of paramount importance for renal allograft recipients. In paediatric patients a granular formulation that allows flexibility for body weight-based dose adjustments is necessary. Our study aims at monitoring the impact of conversion from Prograf capsules to Modigraf granules in stable paediatric renal transplants.
Material and methods: Renal allograft recipients <18 years old followed up in our centre were eligible. Exclusion criteria were parental/patient preference to remain on Prograf, lactose intolerance and very low dose incompatible with measurement of the new granule formulation. Plasma creatinine and trough tacrolimus levels were obtained within 1 (Wk1) and 4–8 weeks post conversion (Wk4–8). Graft survival, side effects and creatinine levels were evaluated after 1 year.
Results: Twenty-six patients were finally converted. Mean patient age was 8+/− 4.1 years. Pre-conversion 69.2% of patients had no rejection episodes, 26.9% had 1 and 1 patient had 2. Only 1 patient had 1 rejection episode 1 year post conversion. Rejection rates for patients 1 year or more post-transplant were 0.2+/−0.4 and 0.1+/−0.2 episodes per year pre and post change respectively ($p = 0.25$). Graft survival at 12 months was 100% without any side effects. Trough tacrolimus and creatinine levels at D0, Wk1, Wk4–8 and 1 year did not differ ($p > 0.05$). No change in Modigraf dose was necessary in 42.3% of individuals whereas a 5–67% increase and a 7–25% decrease were implemented in 9 and 6 patients respectively. No statistical difference between number of dose changes at Wk1 and Wk4–8 was detected.
Conclusions: Conversion from Prograf to Modigraf in stable renal allograft patients is safe and efficacious. Close monitoring is though necessary with additional patient visits in order to optimise medication dose.

P-380 IMPROVING POST-OPERATIVE OUTCOMES IN PAEDIATRIC RENAL TRANSPLANT RECIPIENTS USING A CONTINUOUS TRANSVERSUS ABDOMINIS PLANE BLOCK INFUSION

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Introduction: Managing post-operative pain is fundamental for paediatric renal transplant recipients (pRTR). Opioid elimination is impaired in renal dysfunction, resulting in reduced clearance and opioid-related side effects. A transversus abdominis plane block (TAPB) uses local anaesthetic to interrupt sensory innervation from the surgical site. A catheter-mediated continuous TAPB infusion has seen improvements in postoperative morphine consumption and pain scores in adult renal transplant recipients, but has not yet been reported in pRTR.
Material and methods: Single center case-series of 27 pRTR of median age 12.0 years (Group 1) who received perioperative bolus TAPB and 3 pRTR of median age 16.6 years (Group 2) who received TAPB with continuous infusion of 0.125% levobupivacaine post-renal transplantation. Inclusion criteria: uncomplicated pRTR of dry weight > 20 kg receiving a living donor-related renal graft onto iliac vessels. Outcomes were assessed for each Group 2 individual and an auditing cycle applied to implement changes for subsequent patients.
Results: Median post-operative morphine consumption reduced from 22.0 (0.0–63.5) to 5.8 (0.3–50.8) mcg/kg/h in Groups 1 and 2 respectively. Proportion of pain scores ≥4 within two days post-transplant (%) decreased from a median of 4.6 (0.0–60.0) to 0.5 (0–29.2). Increase in median time to discharge (days) from 10 (6–25) to 13 (9–13) and time to first fluids (hours) from 19.0 (9.0–39.5) to 50.0 (24.0–53.0). Group 2 showed decreased prevalence of nausea/vomiting but increased prevalence of pruritus. Neither group experienced respiratory depression. There were no complications from the placement of the TAP block catheters intra-operatively.
Conclusions: This is the first study that demonstrates reduced postoperative morphine consumption in pRTR who have received TAPB with continuous infusion. Outcomes for these patients varied within the group, representing a learning curve for catheter placement. Further randomised controlled studies are required to study the benefits in a larger cohort of pRTR.

P-381 IMPROVEMENT OF CARDIAC FUNCTIONS IN PATIENTS WITH SEVERE CARDIAC RISK AFTER RENAL TRANSPLANTATION

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Introduction: Patients with chronic kidney disease (CKD) are at increased risk for cardiovascular morbidity and mortality. CKD evokes structural and functional cardiac changes such as left ventricular hypertrophy (LVH), LV dilatation, LV systolic and diastolic dysfunction. Restoration of renal function after renal transplantation (RTX) disrupts the negative cardiorenal interplay and may reverse some of the cardiac changes seen with CKD. We presented the patients with high cardiovascular risk and the success of renal transplantation on the cardiac functions.
Material and methods: Eleven RTX patients who had severe cardiac risk were evaluated by echocardiography before and after renal RTX. Left ventricular diastolic diameter, sistolic diameter, ejection fraction (%) were assesed by echocardiographic standart parameters.
Results: Mean transplantation age was 149.81 ± 43.2 months and mean follow-up period 26.0 ± 16.09 months after transplantation of eleven patients (F/M:6/5). After RTX, serum creatinine level was 0.67 ± 0.16 (0.5–1.06) mg/dl and glomerular filtration rate was 102.38 ± 23.29 (51–126) ml/dk/1,73m^2 in the sixth month. There was a statistically significant improvement ($p < 0.01$) in all cardiac parameters. Preoperative mean ejection fraction (EF) significantly increased after RTX within six months (37.45 ± 9.77%, 66.45 ± 8.39% respectively, $p < 0.01$). Preoperative mean left ventricular diastolic diameter (LVDD) and mean sistolic diameter (SD) were significantly decreased, after RTX with in six months (52.58 ± 8.58 vs 42.86 ± 9.25 and 42.57 ± 8.16, vs 27.05 ± 8.24 respectively, $p < 0.01$). There were 9 patients (81.8%) received multipl antihypertensive treatment before transplantation. Only 2 patients needed antihypertensive treatment after transplantation.
Conclusions: After RTX cardiac functions improve markedly and rapidly in ESRD patients with severe cardiac risk. RTX should be considered the treatment of choice for ESRD patients with systolic heart failure, because a longer duration of dialysis in these patients may result in progressive and ultimately irreversible myocardial dysfunction.

P-382 CLINICAL CHARACTERISTICS OF PATIENTS WITH MTORI CONVERSION AND EFFECTS ON RENAL OUTCOME

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Introduction: Sirolimus is an immunosuppressive agent that offers potentially significant benefits for pediatric transplant patients. In this study, we investigated the effects and efficacy of sirolimus in pediatric renal transplant recipients.
Material and methods: We performed a retrospective analysis of 19 renal transplant recipients who underwent sirolimus/everolimus conversion.
Results: Between years 2002–2012, 226 patients were transplanted and sirolimus/everolimus was not used as a baseline immunosuppressive therapy. During follow-up, 17 patients (7 girls, 10 males) were converted to sirolimus and 2 patients were converted to everolimus (2 males). Five patients were transplanted from deceased donors and the rest from living related donors. The most common etiology for chronic renal failure was congenital anomalies of kidney and urinary tract ($n = 8$). The median age of transplantation was 10.7 years (IQR; 8.0–14.6). These 19 patients were converted to sirolimus/everolimus at 24.5 ± 19.1 months after transplantation for biopsy-proven interstitial fibrosis/tubular atrophy (IF/TA) ($n = 7$), BK-virus associated nephropathy (BKVAN) ($n = 4$), progressive decline of renal function ($n = 3$), gingival hypertrophy/tremor ($n = 2$), posttransplant lymphoproliferative disease (PTLD) ($n = 1$), cyclosporine nephrotoxicity ($n = 2$). Median follow-up after switch was 33 months (IQR; 14–59 months). Three patients with declining renal function and 6 out of 7 patients with IF/TA had stabilized graft function after sirolimus/everolimus. Patients with BKVAN ($n = 4$) had functioning grafts after sirolimus along with anti-BKV treatment. Patient with PTLD had diminished cervical lymph node sizes and complete remission occured after sirolimus. There was no graft loss during observation period. Most common side effects of sirolimus were hyperlipidemia ($n = 7$), development of proteinuria ($n = 3$), increase in proteinuria ($n = 2$) and they were controlled with angiotensin converting enzyme inhibitors.
Conclusions: In conclusion, conversion to sirolimus/everolimus is an effective option for selected patients with tolerable side effects.

P-383 10 YEAR EXPERIENCE OF PAEDIATRIC RENAL TRANSPLANTATION - A REVIEW OF 420 CASES

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Introduction: Renal transplantation is the gold standard treatment for end-stage kidney disease. There are increased challenges in performing transplantation for prospective paediatric renal transplant recipients (pRTR) under 20 kg, especially when placing an adult kidney into a small abdomen.
Material and methods: Data was retrieved from a prospectively collected database, electronic records and hospital notes from two paediatric transplant units in UK. eGFR was calculated using the Schwartz formula. Death-censored renal allograft survival and patient survival were assessed using Kaplan-Meier analysis.
Results: 420 children, (between 2005 and 2015), were divided according to weight: Group A (<20 kg): 116 (28%) and Group B (>20 kg): 303 (72%) pRTR. Median age for Group A was 3 (IQR 2–4) and Group B, 13 (IQR 10–15) years ($p < 0.001$). Patient survival was 98% and 99.7% and renal allograft survival was 93% and 90% in Group A and B respectively, at median follow up of 2 (IQR 1–5) years. Three (38%) and four (48%) renal allograft losses in Group A and B, respectively were due to rejection. 1/116 in Group A and 29/303 in Group B were re-transplants. The last median eGFR was 61 (IQR 48–74) and 51 (IQR 40–63) mls/min/1.73 m2 in Group A and B respectively ($p < 0.001$). There was no significance difference between the groups with respect to patient and renal allograft survival ($p = 0.13$ and 0.28 respectively).
Conclusions: Despite the obvious difference in age between the two groups, the overall patient and graft survival was similar between children <20kgs and >20kgs in this cohort. Transplantation in small children is feasible with good outcomes.

P-384 BK VIRAEMIA AND NEPHROPATHY IN PAEDIATRIC RENAL TRANSPLANT RECIPIENTS

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Introduction: Patient and allograft survival rates for paediatric renal transplant recipients (pRTR) have improved due to more potent immunosuppressive agents. However, they are associated with more infectious complications, such as BK viraemia (BKV) which can lead to BK viral associated nephropathy (BKVAN) and progressive renal allograft dysfunction. We conducted this prospective and longitudinal study to compare incidence of BKV and BKVAN in pRTR.
Material and methods: One hundred and thirty-four pRTR transplanted were included and divided into two groups: Group 1 (15%) were newly transplanted patients and Group 2 (85%) were previous transplanted patients. Group 1 were prospectively monitored with plasma BKV PCR DNA from the time of transplantation, weekly for the first month, fortnightly for the second and third months, and monthly thereafter. Group 2 were patients already transplanted and had plasma BKV PCR DNA checked monthly and during episodes of renal allograft dysfunction.
Results: BKV was detected in four (20%) and seven (6%) patients in Group 1 and 2 (with median ages at renal transplantation of 14.2 and 14.1 years in Group 1 and 2), respectively. Patients who received induction therapy were 75% in Group1 BKV pRTR (vs 43% in Group 2). The median time to detection of BKV post- transplantation was 44 and 142 days in Group1 and 2, respectively. BKVAN was diagnosed histologically in three patients (2 and 1 pRTR in Group 1 and 2, respectively) all of whom were receiving tacrolimus, mycophenolate mofetil and corticosteroids as maintenance immunosuppression. Reduction in BKV PCR DNA was attained in all patients with reducing immunosuppression.
Conclusions: Monitoring of BKV and early intervention to reduce BKVAN are important for pRTR. The first line of treatment remains careful reduction of immunosuppression and close monitoring of renal allograft function.

P-385 EPIDEMIOLOGY AND MORBIDITY OF BK POLYOMAVIRUS (BKPYV) REPLICATION AND BKPYV-ASSOCIATED NEPHROPATHY (BKPYVAN) IN EUROPEAN PEDIATRIC KIDNEY ALLOGRAFT RECIPIENTS: ANALYSIS OF THE CERTAIN REGISTRY

Britta HÖcker [1], Lukas Schneble [1], Lars Pape [2], Paul Schnitzler [2], Luisa Murer [2], Birgitta Kranz [2], Luca Dello Strologo [2], Nikoleta Printza [2], Kristina Heindl-rusai [2], Alexander Fichtner [1], Kai Krupka [1], Lennart KÖster [1], Members Of The Certain Research Network [2], Hans H. Hirsch [2], Burkhard TÖnshoff [1]
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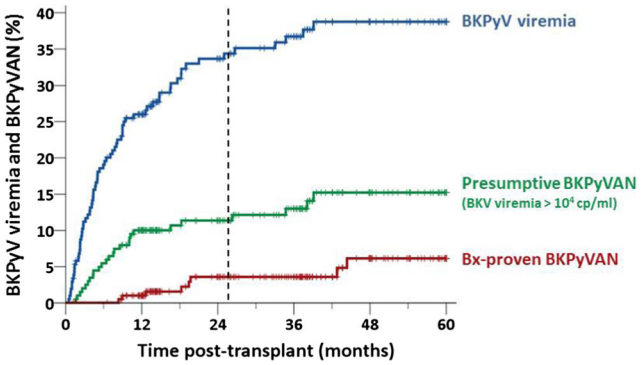
Introduction: BK polyomavirus-associated nephropathy (BKPyVAN) constitutes a serious cause of allograft failure. In adults, BKPyV viremia occurs within weeks to months after kidney transplantation (KTx). According to international guidelines, frequent BKPyV DNA surveillance and judicious reduction of immunosuppression is therefore recommended at least during the first two years post-transplant. While numerous studies in adults are available, larger multicenter studies of BKPyV in pediatric KTx patients are scarce.
Material and methods: As part of the *Cooperative European Paediatric Renal Transplant Initiative* (CERTAIN), we investigated the prevalence and impact of BKPyV replication and BKPyVAN in a cohort of 209 pediatric RTx recipients (mean age 10.9 ± 5.5 years) in whom regular BKPyV DNA surveillance in urine and/or plasma was performed.
Results: In the 1st year post-transplant, 35.7% of patients developed BKPyV viruria; 24.3% exhibited a high viral load in urine (> 10^7copies/mL). BKPyV viremia was observed in 28.6% of recipients. 12.9% of patients developed a high plasma BKPyV viral load (> 10^4 copies/mL) in the 1st year post-transplant fulfilling the criteria of presumptive BKPyVAN. Biopsy (Bx)-proven BKPyVAN was diagnosed in 2.8% of patients during the 1st year post-transplant. Fig. 1 depicts the time-to-BKPyV viremia and Bx-proven BKPyVAN over five years post-transplant. 4/26 (15.4%) of presumptive BKPyVANs and 2/8 (25%) of Bx-proven BKPyVANs were found after two years post-transplant. One patient with Bx-proven BKPyVAN lost his graft 4 months after BKPyVAN diagnosis; in 4 recipients (50%), transplant function stabilized at a lower level (60% of eGFR prior to BKPyVAN). Graft function recovered completely in 3 patients (37.5%) after minimization or conversion of the immunosuppressive regimen. Risk factors associated with BKPyV viremia were younger recipient age ($p = 0.002$), acute rejection before onset of BKPyV viremia ($p < 0.001$) and tacrolimus-based immunosuppression ($p = 0.001$).
Conclusions: Up to 25% of BKPyVAN develop later than two years after KTx, possibly as a consequence of a reduced BKPyV-specific cellular immune response of pediatric patients. Hence, in pediatric kidney allograft recipients, surveillance for BKPyV viremia should be extended beyond the 2nd year post-transplant.



P-386 INCOMPLETE VACCINATION RATES IN EUROPEAN CHILDREN WITH END-STAGE KIDNEY DISEASE PRIOR TO RENAL TRANSPLANTATION – AN ANALYSIS OF THE CERTAIN REGISTRY

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Introduction: Infections constitute a major cause of morbidity and mortality in paediatric renal allograft recipients on immunosuppressive therapy. Hence, avoidance of preventable systemic infections by vaccination prior to transplantation is of utmost importance. However, data on the completeness of vaccinations and risk factors associated with an incomplete vaccination status in paediatric renal transplant candidates are scarce.
Material and methods: In the framework of the Cooperative European Paediatric Renal Transplant Initiative (CERTAIN), we therefore launched a multi-centre, multi-national, retrospective study investigating the vaccination status prior to transplantation of 254 European children with end-stage renal disease (mean age 10.0 ± 5.6 years).
Results: Only 22 of 254 patients (8.7%) presented a complete vaccination status conforming to country- and era-specific immunisation schemes. In particular, the respective vaccination rates against human papillomavirus

(27.3%), pneumococci (42.0%) and meningococci (47.9%) were low. Patients with a complete pneumococcal vaccination status had numerically less lower respiratory tract infections during the first three years post-transplant than children without vaccination or with an incomplete status (16.4% vs. 27.7%, $p = 0.081$). Altogether, vaccine-preventable diseases post-transplant occurred 4.7-fold more frequently in unvaccinated (14/17, 82.4%) than in vaccinated patients (3/17, 17.6%). By multivariate analysis, risk factors associated with an incomplete vaccination status were non-Caucasian ethnicity (OR 9.21, $p = 0.004$), chronic dialysis treatment before transplantation (OR 6.18, $p = 0.001$), and older age at transplantation (OR 1.33, $p < 0.001$) (Table 1).

Table 1

Variable	Unadjusted OR (95% CI)	*p*-value	Adjusted OR (95% CI)	*p*-value
Older age at RTx	1.25 (1.12–1.39)	<0.001	1.33 (1.17–1.52)	<0.001
Gender (ref. male)	0.46 (0.16–1.28)	0.135		
Ethnicity (ref. Caucasian)	3.74 (1.11–12.7)	0.034	9.21 (1.99–42.6)	0.004
History of immunosuppressive therapy	1.57 (0.51–4.82)	0.431		
Time on waiting list	1.46 (0.88–2.42)	0.140		
Dialysis prior to RTx (ref. pre-emptive RTx)	2.80 (1.16–6.80)	0.022	6.18 (2.10–18.2)	0.001
Donor type (ref. living donor)	1.67 (0.69–4.01)	0.254		

OR, odds ratio; CI, confidence interval; RTx, renal transplantation

Conclusions: The vaccination rates in paediatric kidney transplant candidates are incomplete. Paediatric nephrologists, together with primary-care staff and patients' families, should therefore take every effort to improve vaccination rates prior to kidney transplantation.

P-387 HIGH RATE OF SECONDARY VACCINATION TITRE LOSSES IN PAEDIATRIC RENAL TRANSPLANT RECIPIENTS: AN ANALYSIS OF THE CERTAIN REGISTRY

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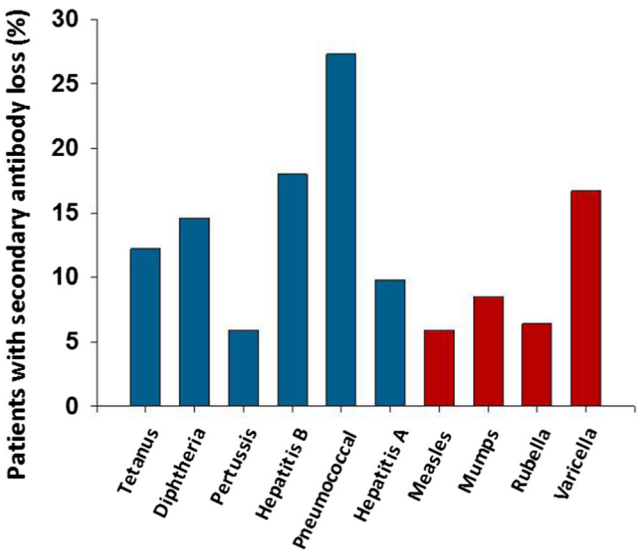
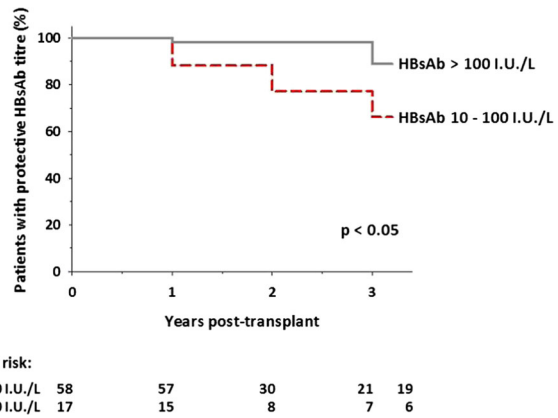
Introduction: Infections constitute a major cause of morbidity and mortality in paediatric renal allograft recipients on immunosuppressive therapy. Hence, avoidance of vaccine-preventable systemic infections is of utmost importance. However, the development and maintenance of protective vaccination titres may be impaired in this patient population due to their need of immunosuppressive medication.

Material and methods: In the framework of the *Cooperative European Paediatric Renal Transplant Initiative* (CERTAIN), we therefore performed a multicentre, cross-sectional study in 254 patients and analysed the post-transplant course of vaccination titres in a subset of 150 patients for whom serial titre measurements were available.

Results: The rate of protective vaccination titres in uraemic children prior to transplantation was low, especially for diphtheria (38.5%) and pertussis (21.3%). As few as 58.1% of patients developed a hepatitis B titre (HBsAb titre) > 100 I.U./L prior to RTx. 39.6% of patients showed a secondary vaccination titre loss post-transplant, especially against the life-virus vaccines varicella and mumps as well as the inactivated vaccines tetanus, diphtheria, pneumococcus and hepatitis B (Fig. 1). Patients with a HBsAb titre between 10 and 100 I.U./L prior to RTx (baseline) experienced significantly ($p < 0.05$) more often a hepatitis B vaccination titre loss post-transplant than patients with a baseline HBsAb titre >100 I.U./L (Fig. 2). The revaccination rate post-transplant was low and failed to induce protective titres in a considerable amount of patients: only 37.5% developed a sufficient HBsAb titre, and as few as 14.3% of patients showed a protective pertussis titre after revaccination. Treatment with rituximab was associated with a significantly increased risk of a vaccination titre loss post-transplant (odds ratio 3.9; $p = 0.044$).





Patients at risk:

HBsAb > 100 I.U./L	58	57	30	21	19
HBsAb > 10 I.U./L	17	15	8	7	6

Conclusions: We observed a high rate of secondary vaccination titre losses post-transplant also for the inactivated vaccines tetanus, diphtheria, pneumococcus and hepatitis B. We therefore recommend in accordance with the *AST* guidelines to measure vaccination titres including tetanus, hepatitis B and pneumococcus at regular intervals post-transplant in order to induce timely revaccination and thus to avoid the development of vaccine-preventable diseases.

P-388 URINARY EXOSOMES ISOLATION: DIFFERENT METHODS TO DISCOVER NOVEL BIOMARKERS OF KIDNEY REJECTION

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Introduction: Exosomes are lipid membrane-bound nanoparticles (40–150 nm of size) released from different cells type. They could carry different types of cargo (e.g. miRNA, proteins) that reflect the physiopathological status of the cells and/or organ they originated from. For instance, the exosomes present in the urine called Urinary Exosomes (UExs) arise from all the different nephron cells. Thus, an accurate characterization of their content could be helpful to identify novel reliable non-invasive biomarkers for kidney allograft injury. However, due to their low amount, UExs concentration and characterization remain a challenge. This study aims to identify the most efficient UExs isolation method both for RNA profiling and proteomic analysis to discover novel renal transplant rejection biomarkers.
Material and methods: UExs were isolated from urine samples using four different methods: three commercial kits (Norgen, ExoQuick and Qiagen) and the ultrafiltration. UExs were quantified by qNano. Total RNA, included small RNA species, was extracted using column kit. Biochemical assay was applied to extract proteins. The miRNA and proteins integrity and concentration were evaluated by Agilent Bioanalyzer 2100.
Results: The UExs isolated with the four methods differ in raw concentration (3.5×10^8–3.5×10^{12}) and size (90–130 nm). qNano analysis showed that Qiagen kit was the most efficient isolation method for UExs. However, Agilent Bioanalyzer RNA analysis emphasized that the highest amount of miRNAs was obtained using Norgen Kit. Same kind of experiments are currently ongoing for the analysis of the UExs proteins content.
Conclusions: Based on our results, all four methods yielded UExs with a high variability in number and diameter. Furthermore, Norgen kit is the most productive method for miRNA UEXs isolation. The next step of this study will be the evaluation of UExs characterization in correlation with the different outcome observed in the pediatric population recruited in our renal transplanted center.

P-389 PENTRAXIN-3 AND INFLAMMATORY STATUS IN PEDIATRIC RENAL TRANSPLANT RECIPIENTS

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Introduction: Chronic inflammation is mostly linked to chronic kidney disease (CKD) and is associated with progression of renal disease, cardiovascular disease and poor outcome. Renal transplantation (RTx) may improve this condition whereas it may also contribute to inflammation via oxidative stress due to ischemia reperfusion injury, acute rejection, graft dysfunction, hypertension and increased body mass index. In the present study, we evaluated inflammatory status in pediatric RTx recipients.
Material and methods: A total of 24 RTx patients (17 males, aged between 5.0–18.6 years) with a functioning kidney [median (IQR) of eGFR; 66.5 (27) ml/min/1.73m^2] were enrolled in the single-center study. Control group consisted of 12 age, gender and eGFR similar patients with CKD. For the assessment of inflammatory status, Pentraxin-3, IL-6 and IL-10 were measured by ELISA method. Routine biochemical parameters and high sensitive (hs)-CRP were recorded from the patients' file. Twenty-four hour MAP was also recorded from the last ABPM within 6 months. Patients who had a 24-h MAP ≥ 2SDS were defined as hypertensive.
Results: The median age at the transplantation was 11.8 (2.2–17.0) years and the median follow-up was 35.1 (3–87) months. All except one patient received their first graft and 19 (79%) received a kidney from a living donor. Four patients (17%) had a history of acute rejection episodes. There was no difference considering inflammatory markers except Pentraxin-3 between the two groups. The median (IQR) of serum Pentraxin-3 levels was significantly lower in renal transplant recipients as compared to CKD controls [1.00 (0.99) vs. 1.51 (1.02) ng/mL, $p = 0.026$) and it was positively correlated with 24 h–MAP SDS ($r = 0.453$, $p = 0.034$) but not with any other clinical or laboratory parameters. Hypertensive patients ($n = 6$) showed higher medians (IQR) of Pentraxin-3 [1.90 (1.12) vs. 0.86 (0.57) ng/mL, $p = 0.027$] and lower IL-10 levels [6.42 (4.66) vs. 10.7 (10.8) pg/mL, $p = 0.040$].
Conclusions: Pentraxin-3, a marker of vascular inflammation is lower in RTx recipients compared to CKD patients, whereas a high level of pentraxine-3 is closely associated with hypertension.

P-390 TACROLIMUS VARIABILITY: A CAUSE OF DONOR SPECIFIC ANTIBODY FORMATION IN RENAL TRANSPLANTATION?

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Introduction: Immunsupresive drug compliance is required for long-term graft survey in renal transplant recipients. Tacrolimus is one of the major immunsupresive agents and fluctuations of blood levels may lead to antibody development. The aim of the study was to investigate whether tacrolimus variability is an effective factor for antibody development and graft survival in pediatric renal transplant recipients.
Material and methods: Pediatric living-donor renal transplant recipients followed-up at our center at the last two years and who were using tacrolimus were retrospectively evaluated. Patients who had pretransplant donor spesific antibodies (DSA) were excluded. Tacrolimus blood levels, serum creatinine and DSA were recorded. Estimated glomerular filtration rate (eGFR) were calculated using Schwarz formula. Tacrolimus variability (Tac CV = standard deviation of mean/mean) was calculated separately at the first post-transplant 6 months, between 6 and 12 months and from the end of the first year to the last follow-up period. Renal biopsy was performed in all patients with positive DSA.
Results: A total of 65 patients, 48 males (73.8%), with a mean age of 15.16 ± 4.43 years, were included in the study. The mean age at the time of transplantation was 11.20 ± 3.88 years and mean follow-up period was 50.82 ± 26.84 years. DSA positivity was detected in 12 patients (18.4%). eGFRs were similar between DSA negative and positive groups (78.72 ± 2.86 vs 77.45 ± 8.08, respectively; $p > 0.05$). HLA mismatches >3, presence of acute celluler rejection, Tac CV at post-tx 6–12 months and >12 months were found to be significant factors associated with the development of DSA. According to the logistic regression analysis, Tac CV > 0.3 at post-tx 6–12 months was associated with the development of DSA (B:1.440, $p = 0.046$). Also, Tac CV > 0.3 at post-tx 6–12 months negatively correlated with eGFR. Within recipients who underwent renal biopsy, mean Tac CV over 12 months were significantly higher in IF/TA positive patients than IF/TA negative patients (6.58 ± 0.44 vs 6.29 ± 0.20, respectively; $p = 0.031$).
Conclusions: Tacrolimus variability was associated with DSA formation and negatively correlated with estimated glomerular filtration rate in pediatric living-donor renal transplant recipients. We suggest to calculate

tacrolimus variability in all pediatric living-donor renal transplant recipients.

P-391 COMPLEMENT ACTIVATION IN RELATION TO DONOR SPECIFIC ANTIBODIES AFTER KIDNEY TRANSPLANTATION

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Introduction: Antibody mediated rejection (AMR) is the major immunological cause of kidney transplant failure also in the paediatric population. According to current knowledge, late AMR is classically caused by the development of donor specific antibodies (DSA) and the complement system plays a critical role in its pathomechanism. The aim of this work was to determine the levels of various complement activation products in kidney transplant recipients.
Material and methods: 106 adult kidney transplanted patients who had detectable DSA after transplantation (DSA+, 189 days to 29 years post-transplantation) were involved in the study. One hundred six DSA-negative patients were matched as controls using 1:1 propensity score matching. Two patients were excluded due to poor sample quality. The levels of complement activation products (C3a, SC5b-9, C4d and Bb) were measured by enzyme-linked immunosorbent assay (ELISA) using EDTA plasma samples.
Results: Activation product levels were compared between the DSA-positive and negative groups. Mean C3a concentration in the plasma of DSA+ patients was 97.3 ng/ml (SD 38.9), compared with 93.86 ng/ml (SD 41.38) in the DSA- group ($P > 0.05$). Mean SC5b-9 concentration in the DSA+ patients was 223.3 ng/ml (SD 86.8), compared with 222.8 ng/ml (SD 83.5) in the DSA- group ($P > 0.05$). Mean level of C4d in the DSA+/− groups was 2.7 μg/ml (SD 1.19) compared with 2.93 μg/ml (SD 1.65) ($P > 0.05$). Mean Bb levels of the DSA+/− patients found to be 1.3/1.15 μg/ml (SD 0.66/0.38) ($P > 0.05$).
Conclusions: Levels of complement activation products in kidney transplant recipients were determined. No significant difference in plasma levels of activation markers for C3 (C3a), terminal pathway (SC5b-9), classical pathway (C4d) and alternative pathway (Bb), was observed in DSA positive and negative patients. Further analysis is necessary to identify association of complement activation with the histological signs of AMR.

P-392 EN-BLOC KIDNEY TRANSPLANTATION FROM INFANT DONORS INTO PEDIATRIC RECIPIENTS: A SINGLE CENTER EXPERIENCE

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Introduction: En-bloc kidney transplantation (EBT) from infant donors may be a treatment option for children with end stage renal disaese. The aim of this study was to evaluate the clinical outcome of children who received en-bloc kidney from infant donors at our center.
Material and methods: Medical records of children with performed EBT were reviewed retrospectively from hospital computerized database system.
Results: 8 children with a median age at transplantation of 14.7 years (3.5–17 years) and a median follow-up period of 57.0 months (2–95 months) were included in the study. Donor age and weight ranged from 7 to 38 months and 4.4 to 15 kg (median 9.5 kg), respectively. All recipients showed immediate graft function. No post-operative thrombotic complications were seen. Urine leakage developed in one patient at the 7nd day of transplantation, which spontaneously resolved after continuously adequate drainage. The size of left and right kidneys increased from 73.37 ± 5.75 mm to 95.66 ± 16.35 mm and 70 ± 8.2 mm to 91.57 ± 19.53 mm, respectively. Only one patient developed acute humoral rejection due to non-adherence at the 15th month of transplantation and had an estimated glomerular filtration rate of 16.5 ml/min/1.73 m2 at the last follow-up. The others had stable graft functions.
Conclusions: According to our experience, en-bloc kidney transplantation from infant donors into pediatric recipients seems to be safe and effective.

P-393 EVALUATION OF SAFETY AND EFFICACY OF FOLLOW-UP BIOPSIES IN MONITORING RENAL PAEDIATRIC TRANSPLANT

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Introduction: Surveillance allograft biopsies could provide the opportunity to tailor the patient's management based on evidence of subclinical histological changes with potentially important implication for graft outcome.
Material and methods: We evaluated all protocol biopsies performed in our Center from January 2007 to December 2015 at 6, 12 and 24 months post-transplant. The primary aim of our study was to evaluate the efficacy of the histological surveillance in preventing functional decay of renal allograft. The secondary aim was to evaluate the prevalence/severity of post-bioptic complications.
Results: 234 biopsies have been performed in 115 patients (M/F 74/41, m. age 10.9-6.9 years). Subclinical histological changes were observed in 73/234 that lead to clinical therapies change in 67/73 cases. Forty-one subclinical acute rejection (34% Banff type 3, 53% Banff type 4 IA/IB, 12% Banff type 2) have been trated with methylprednisolone pulses and immunosoppression adjustement, plus Ig ev and Rituximab in the 5 cases of acute humoral rejection. Eighteen cases with CNI toxicity and 6 cases of IF/TA have been treated with low calcineurin doses +/− mTOR inhibitor. One case of intrarenal PTLD was treated with Rituximab. We observed 24/234 complications: in 4/234 (1.7%) procedures there were major complications including three intraparenchimal AVF resolved with endovascular procedure. 20/234 (8.5%) minor complications all solved spontaneously.
Conclusions: Our study confirmed the efficacy and safety of protocol biopsies for guiding post-transplant management and improving the renal allograft outcome in paediatric patients.

P-394 SUCCESSFUL KIDNEY ALLOGRAFT IN PATIENT WITH DENSE DEPOSIT DISEASE

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Introduction: To describe a case of successful kidney transplant in a patient with Dense-deposit- disease (DDD), without recurrence of the initial pathology after prophylactic treatment with eculuzimab.

Material and methods: Dense-deposit disease (DDD) is a rare glomerulopathy characterized by electron-dense deposits in the glomerular basement membrane. About 50% of patients with DDD progress to end-stage kidney disease and require dialysis within 10 years of diagnosis. The disease often recurs post-transplant. Some clinical reports show an efficacity of eculizumab traitement, a humanized monoclonal antibody that binds to C5 complement protein.
Results: We describe an 18-year-old girl with a successful kidney allograft for end-stage renal disease due to DDD. At the age of 9 years she was diagnosed with steroid resistant nephrotic syndrome associated to microscopic hematuria, severe hypocomplementemia and renal failure. Kidney biopsy revealed DDD. Despite treatment with steroids and mycophenolate mofetil (MMF), the kidney function further declined over 2 years, and hemodialysis was started at the age of 14 years. She received a cadaveric transplant at 17 years and we chose an immunosuppressive treatment including one IV steroid pulse of 500 mg/m2, oral steroids over 3 months, tacrolimus, MMF and eculizumab (8 infusions over the first 2 months post-transplant). Biological follow up was done with weekly complement C3 level analysis and proteinuria. There was complete terminal complement pathway inhibitionno proteinuria recurred and eculizumab treatment was discontinued after 2 months. Protocol kidney biopsies were performed at M1, 4, and 11 without signs of disease recurrence. Graft function remained normal and proteinuria negative after eculizumab discontinuation.
Conclusions: Eculizumab might be an interesting tool to prevent disease recurrence post-transplant in patients with DDD. Careful monitoring of histological and biological parameters during and in particular after eculizumab treatment are necessary to reduce the potential interval between disease recurrence and eculizumab (re)-treatment.

P-395 IS CONTRACEPTIVE ADVICE BEING GIVEN TO ADOLESCENT RENAL TRANSPLANT PATIENTS?

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Introduction: Adolescents are at risk group so contraceptive advice is paramount. Risk taking behaviours and disengagement from the medical team is common, therefore delivering advice can be challenging. Many of these patients will never have received contraception advice before and many find the prospect of talking about it embarrassing and daunting. Providing appropriate advice can often be challenging due to co-existing learning difficulties. Our objective was to assess whether adolescents within the young adult transplant service had received accurate advice on contraceptive and pregnancy transplant safety.
Material and methods: We designed and distributed patient experience questionnaire to all patients within the renal young adult service who had a functioning transplant. This focused upon two main issues: advice they had been given regarding contraception and the risks of pregnancy and current use of contraceptives. We also asked for their opinions of how to improve the information provided.
Results: Nine responses were received from this small patient group. Only 55% had received contraceptive advice since transplant, with 67% wanting more information relating to contraceptive choices. Only 22% had received contraceptive advice at their paediatric centre. Only 55% of patients had received advice regarding pregnancy and transplant. When asked how they would like to receive further information, a leaflet format and further conversations with the young adult team were preferred.
Conclusions: Although a small sample, this demonstrates that contraceptive advice is not regularly being given out to adolescent renal transplant patients. The patient cohort is extremely diverse with many patients suffering from learning difficulties; however this should not stop contraception being discussed. This has resulted in an adolescent specific leaflet being designed to provide detailed information regarding sex, contraception and STIs. A framework to aid clinicians in providing advice is under development.

P-396 RENAL FUNCTION AFTER PEDIATRIC COMBINED LIVER KIDNEY TRANSPLANTATION

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Introduction: Combined liver and kidney transplantation (CLKT) is a viable option for end–stage organ failure altering both liver and kidney. Our aim was to evaluate renal function after pediatric CLKT with measured and estimated glomerular filtration rate (GFR).
Material and methods: All pediatric patients (age < 16 years) who underwent CLKT by 2011 were included (n = 11). GFR was measured (mGFR) with ^{51}Cr-EDTA clearance (n = 10). In addition, GFR was estimated (eGFR) with bedside Schwartz eq. [0.413 x (height (cm) / creatinine (mg/dL))] (n = 10). GFRs are shown with unit of ml/min/1.73m^2. Triple immunosuppression with cyclosporine or tacrolimus, azathioprine or mycophenolatemofetil and methylprednisolone was used. Results are shown with mean (SD). Paired t-test with 5000 bootstrap samples was used. In addition, comparison between eGFR and mGFR with limits of agreement (LOA) was made.
Results: Primary diagnoses for CLKT were autosomal recessive polycystic kidney disease (n = 7), primary hyperoxaluria type I (n = 2), methylmalonicacidemia (n = 1) and atypical hemolytic uremic syndrome (n = 1). Four (36.4%) patients were male. Mean age at CLKT was 4.9 (4.3) years. Mean mGFR at 1 year was 68.3 (24.0) [13.9 (2.6) months from CLKT] and 60.0 (11.6) at last follow-up [121.9 (47.1) months from CLKT]. Difference in mGFR between 1 year and last follow-up was 8.3 (95% CI - 6.1 to 26.5). Mean eGFR at 1 year was 82.1 (29.7) and 73.5 (18.3) at last follow-up. Difference in eGFR between 1 year and last-follow-up was 8.6 (95% CI - 6.6 to 21.6). Mean difference between eGFR and mGFR at 1 year was 13.8 (95% LOA - 46.8 to 74.4) and at last follow-up 13.5 (95% LOA - 10.4 to 37.5).
Conclusions: Renal function remained acceptable level in CLKT patients during the follow-up. Bedside Schwartz equation overestimated measured GFR, however, wide limits of agreement between these two methods justifies careful interpretation.

P-397 THE CHANGING LANDSCAPE OF PAEDIATRIC KIDNEY TRANSPLANTATION OVER TWO DECADES

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Introduction: Kidney transplantation is a life transforming procedure for children and their families. However, graft and recipient outcomes can be compromised by early technical difficulties, non-concordance with medication and the side effects of immunosuppression. Knowledge of the clinical course of children who have received kidney transplants is essential to guide practice.
Material and methods: Our objectives were to review the demographics of paediatric kidney transplants performed in Northern Ireland over the past 20 years. In addition, we wanted to evaluate the long term graft and recipient outcomes after paediatric kidney transplantation. All recipients of paediatric kidney transplants performed in Northern Ireland from 1995 to 2016 were

included. Donor and recipient demographics and transplant details are recorded at the time of transplantation. Recipient and donor outcomes were collected prospectively. Recipients were followed up until death or 1st December 2016.
Results: There were 78 transplants performed during the study period; 61% of recipients were male. The median age was 12 years. The median duration of pre-transplant renal replacement therapy was 14 months. Eighteen patients were transplanted pre-emptively; 50% of transplants performed since 2010 have been pre-emptive.The median donor age was 29 years and kidneys were donated after brain death in 65% of cases. Since 2010, donor demographics have shifted in favour of living donation and the majority of transplants performed in the past five years have been from living donors. The median number of HLA mismatches was two and the ischaemic time ranged from 143 to 2785 min (median 1143 min). The median follow up time was 9.6 years. Seventy-one patients were discharged with a functioning graft. Discharge creatinine was available for 66 patients with a range of 22–197 μmol/L and a median of 60 μmol/L. One and two years after transplant, the median creatinine was 76 μmol/L and 87 μmol/L respectively.The median graft survival was 84.5 months. There were 22 cases of death-censored graft loss in the study period. In five, graft loss was secondary to early technical problems; these all occurred between 1995 and 2005. In the other 17, grafts functioned for 5–18 years prior to failure. In multivariate analysis, recipient age, donor age and era of transplantation were associated with graft survival. Three deaths occurred in the follow up period. Two patients died within the first month and a third died from post-transplant lymphoproliferative disorder 17 years after transplantation.
Conclusions: There have been many improved developments over the past twenty years after paediatric kidney transplantation in Northern Ireland. There have been no early graft losses or deaths in the past decade. The expansion of the living donor programme means that 50% of children are now transplanted pre-emptively, removing the detrimental effect of dialysis therapy on their health and development. The overall outcomes are now excellent and boast an impressive foundation for future transplant outcomes.

P-398 PSYCHIATRIC ASPECTS OF CHILDREN WITH RENAL TRANSPLANTATION

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Introduction: Renal transplantation is the rescue treatment for End Stage Renal Diseases in all ages. It 's well known that; various factors such as difficulties in daily life arising from chronic renal disease, complications, waiting for a donor and the need for social support, marriage status and need for having children cause multiple psychiatric problems with the majority of anxiety and depression, in these patients. The recent studies reported the persistence of anxiety and depression in 25–40% of cases after renal transplantation. We aimed to evaluate the psychiatric profiles of children and teenagers after renal transplantation and detect the variables in this preliminary study.
Material and methods: The study was consisted of 31 children and teenagers. All patients and parents were acknowledged about the study and only the volunteers were included. All patients were asked to fill out the socio demographic form, family assessment device (FAD), state-trait anxiety inventory II (STAI-II) and the symptom check list-90-R (Scl90-R). The demographic and clinical data including the donor type, the waiting time for donor, data about immunosuppressive treatment were recorded from the charts of patients. All data were analyzed on SPSS 20.0 statistical program. Correlation coefficients were used to analyze the relationship between standard definitive tests and multiple variables.
Results: The statistically significant increase in the Scl 90 obsession score was found in children of families, which are in very low socio economic status. Although the finding of the more immunosuppressive drugs used, the increased scores of FAD, STAI-II and Scl 90-R were found; the only statistically significant relation was seen between the role score of FAD and obsession score of Scl 90. There was statistically difference between depression and anxiety scores of Scl-90 belonged to cases having living related donor. However, in all cases role, attention and behavioral control score of FAD were worse. STAI-II scale was found moderately high in all cases. There weren't any significant correlation between drug usage time, donor type and the other multiple variables. The moderate and high correlation was found between STAI-II and all scores of Scl90 except for anger.
Conclusions: In this study we want to emphasize the worse results found in all cases scored for role, attention and behavioral control scale of FAD. Therefore we vigorously suggest considering the cases in relation with their families during both pre and posting transplant period and following up for psychiatric problems with the care of their developmental course, even in the post transplant long-term.

P-399 TRANSIENT HYPERPHOSPHATASEMIA AFTER RENAL TRANSPLANTATION IN 2 CHILDREN

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Introduction: Transient, isolated hyperphosphatasemia has been rarely reported in pediatric and adult transplant patients.
Material and methods: We describe two cases in boys after living donor kidney transplantation (Tx).
Results: They were 7 and 9 years old respectively in good general condition, without clinical problems, on immunosuppression with tacrolimus and mofetil mycophenolate with stable blood levels and doses. In the first case, 10 months after Tx increased level of serum alkaline phosphatase (AP) was noticed on regular blood check. All other blood tests, enzymes, PTH and bone markers were within normal range as well as no data for infection. Bone izoenzyme of AP was elevated. It peaked at 4868 U/l 1 month later and went down to normal values after 3 months. In the second case, 7 months after Tx increased level of AP was registered on regular blood check. All other blood tests, enzymes, PTH and bone markers were within normal range as well as no data for infection. Liver izoenzyme of AP was elevated. It peaked at 6284 U/l 1,5 months later and went down to normal values after 3 months. However, 10 days latter CMV infection, presenting as pneumonia was detected by PCR and treated accordingly.
Conclusions: Transient, isolated hyperphosphatasemia could be seen in children after renal Tx and is benign condition, which resolves without treatment. However, it might precede some infections.

P-400 RENAL CELL CARCINOMA IN A KID WITH TRANSPLANTED KIDNEY

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Introduction:
Background: Renal cell carcinoma (RCC) is an uncommon renal cancer in paediatric population with the occurrence of 0.1–0.3%. Cumulative frequency 10 years subsequent to kidney transplantation is 186/100.000, in kids under the age of 18 years at the moment of transplantation.
Material and methods: We would prefer to represent the case of a girl age 13 years and 5 months identified at the habitual echo-sonographic assessment seven years following kidney transplantation, with a native renal cyst. The identification of an illness that initiated the end phase renal disease was infantile steroid resistant nephritic syndrome with path histology confirmation of disseminated mesangial sclerosis established at the

age of 10 months. Hemodialysis was begun at the age of three years, followed by existing allied kidney transplantation (grandfather) at the age of six. Through post-transplantation follow up there were two severe rejections, cured with metylprednisolone.
Results: It was detected an inadequate reaction to cure chronic allograft nephropathy built up. Six years subsequent to transplantation growth hormone (GH) therapy was begun. At the time of incidental result of kidney cyst, girl had no record of fever, tiredness, urinary symptoms or hematuria. Magnetic resonance imaging (MRI) was carried out and left side nephrectomy was completed. Path histology demonstrated renal cell carcinoma, grade I (Fuhrman), whereas additional investigation illustrated no metastasis.
Conclusions: In kids with renal transplant, native renal cysts and GH therapy standard echosonographic screening of native kidneys is essential. Nephrectomy of cystic native kidneys smust be thought prior to transplantation and/or GH healing.

P-401 METABOLIC SYNDROME AND LEFT VENTRICULAR HYPERTROPHY IN CHILDREN AFTER RENAL TRANSPLANTATION

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Introduction: Whereas renal transplantation(RTx) reverses uremic abnormalities; additional risk factors for cardiovascular disease like hypertension, diabetes and metabolic syndrome (MS) may arise. The present study aims to evaluate the prevalence of MS in pediatric RTx recipients and the association of MS with left ventricular hypertrophy(LVH) as a risk factor for cardiovascular disease.
Material and methods: A total of 36 RTx recipients (15 female, aged between 10 to 21 years) who were transplanted before 18 years of age were enrolled in the study. Anthropometric measurements were performed and their SD scores were calculated according to the national percentiles. Casual blood pressure (BP)s were measured and their z scores were calculated. Data on primary renal disease, modality and duration of dialysis, donor and transplant properties and certain laboratory findings were retrospectively documented from patient's records. Left ventricular mass index (LVMI) was calculated from echocardiographic measurements, and LVH assessed for height-age. MS was defined by International Diabetes Federation (IDF) criteria.
Results: Ten out of 36 patients (28%) showed MS. Patients with MS had shorter transplant vintage compared with those without MS (25.9 ± 14.9 vs 49.1 ± 29.9 months, $p = 0.04$); however there were no differences considering age, gender, weight gain after transplantation and other clinical data between the groups. Patients with MS had higher LVMI than the patients without MS (42.8 ± 8.7 g/m$^{2.7}$ vs 35.7 ± 9.4 g/m$^{2.7}$, $p = 0.044$). The prevalence of LVH was 60% in the patients with MS, but 38.5% without MS ($p = 0.285$). There was no correlation between LVMI and metabolic syndrome criteria including SD scores of waist circumference, z scores of systolic and diastolic BP, triglycerides and HDL levels the patients with MS and in the whole group.
Conclusions: Metabolic syndrome is a common condition in pediatric RTx recipients and associated with increased risk of LVH.

P-402 JUVENILE NEPHRONOPHTHISIS: TIMING OF DIAGNOSIS AND RESULTS OF KIDNEY TRANSPLANTATION

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Introduction: Nephronophthisis (NPH) is the most common genetic cause of end-stage renal disease (ESRD) in childhood and adolescence. Diagnosis is often late as it is difficult to suspect until the complications of slowly decreasing renal function manifest. Treatment of NPH is symptomatic until ESRD, then renal replacement therapy (RRT) is started. We report our centre's experience in timing of NPH diagnosis and results of kidney transplantation.
Material and methods: Medical records of 22 patients with juvenile NPH (by clinical findings or genetic tests) in our centre during 1990–2016 were analysed. Presentation of extrarenal symptoms, timing of diagnosis and ESRD, RRT and graft survival was evaluated.
Results: 14 boys and 8 girls were diagnosed with NPH at age 9.1 ± 3,6 years, three confirmed by genetic testing. Extrarenal symptoms were identified for 5 patients. Average age of ESRD were 11,9 (± 3,4) years. RRT was started for 20 patients, 2 pre-emptive transplantations performed. For 5 patients RRT was started during the first 4 months after diagnosis. During observation time 7 transplants were lost for 6 patients: 5 due to chronic graft nephropathy, 2 due to infection induced graft failure. Overall graft survival after 5, 10, 15 and 20 years was 90%, 70%, 70%, 60% respectively. Disease recurrence after transplantation was not observed.
Conclusions: Diagnosis of NPH was usually delayed for our patients with one fourth of them requiring RRT soon after the diagnosis. Renal graft function was preserved for long periods among transplant recipients, disease recurrence was not observed.

P-403 THE EFFECT OF PRETRANSPLANT LONG TERM ANURIA ON GRAFT OUTCOME IN CHILDREN

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Introduction: There is little data on the course of allografts in patients with long term anuria. We aimed to determine the effect of pre transplant long term anuria on graft outcomes in children.
Material and methods: We retrospectively scanned the data files of 133 renal transplant recipients. Patients divided two groups as long term and short term pre transplant anuria (less and more than six months). These two groups compared with non-oliguric patients in terms of graft outcomes.
Results: 21 of 133 patients were short term anuric (10 male, 11 female), 25 of them were long term anuric (11 male, 14 female) and 87 were non-anuric (53 male, 34 female). Bladder capacity of non-anuric patients was 295 ± 135 ml, and 248.3 ± 140.83 ml in short term anuric patients, 126.8 ± 72.38 ml in long term anuric patients($p < 0.05$). There was no difference in glomerular filtration rates of the groups in the 1st and 3rd years. Graft loss rate were; % 5.7 in non-anuric patients, %14 in six months short anuric patients, %24 in six months long anuric patients. There was a positive correlation between the rate of graft loss and anuria in patients with anuria (r: 0.25 $p = 0.03$).
Conclusions: Long-term outcomes in anuric patients prior to transplant worse than non-anuric patients. This result emphasizes the importance of early term or preemptive kidney transplant.

P-404 VIRAL INFECTIONS AFTER RENAL TRANSPLANTATION IN BULGARIAN CHILDREN

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Introduction: Viral infections are common problem in pediatric patients after kidney transplantation (Tx).
Material and methods: The aim of this retrospective study was to analyze the frequency of the most important viral infections after Tx. In period of 10 years (2007–2016) we followed 22 children, 14 boys and 8 girls after renal Tx. They were from 2 to 17 years old and follow up time was from 1 to 10 years.
Results: Of them 10 (45.5%) have never had viral infections. In 11(50%) patients CMV infection was registered by PCR, in 4 (18.2%) EBV, in 1 (5%) BKV, 2 (9%) have had Varicella disease, 1 (5%) acute hepatitis B, 1 (5%) chronic anemia, due to Parvo virus B19 infection and in 1 (5%) JCV was detected in renal biopsy. More than one viral infection was seen in 7 (32%) patients.
Conclusions: Various viral infections were observed in Bulgarian children after renal Tx. Although almost half of them have never had, one third had more than one. The most frequent was CMV, affecting half of the patients.

P-405 LONG-TERM OUTCOME OF KIDNEY TRANSPLAN TATION IN CHILDREN WITH FSGS

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Introduction: Recurrence of focal segmental glomerulosclerosis (FSGS) occures in aproxymatelly 30% of patients after renal transplantation (Tx) and negatively impacts graft survival. Risk faktorsfor recurrence are age 6 years-15 years, presence of mesangial hyper cellularity. at onset of nephrotic syndrome and rapid progression of the disease to ESRD in less than 3 years in the native kidneys. Retransplantation of a recipient with recurrence of FSGS in a perevious graft brings 80% risk a recurrence rate. Therapeutic options to produce a remission following recurrence of NS include Plasmaexchange (PE), high dose of cyclosporine, PE and cyclophosphamide, Plasma protein absorption, and recently rituximab, but there were not control studies.
Material and methods: Medical records from 121 kidney transplanted (kTx) children from 1982 to 2016 were collected and analysed. Most of them were transplanted in our country, some in abrod (France, Austria, Italy). In 17/121 (20.5%) primary renal disease were FSGS. In all patients FSGS as original diesease and as a recurrence was biopsy proven. Three pts. received second graft. One of them had FSGS recurrence in the second graft as well as the first. The patient with mesangioprolipherative GN in his native kidneys developed massive proteinuria on the first day after Tx. Graft biopsy showed FSGS. Imunosupression was CyA, Aza, Steroids earlier and CyA or tacrolimus, MMF, Steroids recently. Plasmaexchange (PE) was preformed in 3 pts. with FSGS recurrence and rituximab with PE in 2. In Patient with HBV infection only steroid boluses were administered.
Results:
Age ys 11.46 ± 7.2.
Follow up ys 6.8 ± 21.2.
Source of graft DD/LD 14/3.
Retransplanted- second graft 1.
Age at onset NS 6.1 ± 7.4.
Reccurence of FSGS 5/17.
Onset of proteinuria after Tx 1 day-7 mos.
Treatment modality: PE, ACE, Rituximab, high doses of methylprednisolone.
Outcome: Graft loss after 1 months - 3,5 years Complete remissIon in one after preemptive Rituximab and PE.
Conclusions: Recurrence of FSGS after kTx is associated with impared graft function and graft loss. Current therapy with PE and rituximab is not always effective. We neeed more controlled studies to answer the issue when to start therapy, which therapy and how long.

P-406 KIDNEY TRANSPLANTATION IN SMALL CHILDREN: A MATCH-CONTROLLED RISK ASSESSMENT

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Introduction: Paediatric renal transplantation (RTx) is the treatment of choice for children with end-stage renal failure. Infants <10 kg of body weight, however, represent a small patient group which is often not considered suitable for RTx. The objective of this retrospective, matched cohort study is the comparison of short-term outcomes (for 2 years post-transplant) in low-weight children (<10 kg) and in a control group of children with 10–15 kg body weight at time of engrafting in order to refute or confirm the common current weight threshold (10 kg) for paediatric RTx.
Material and methods: We conducted a multicentre, retrospective cohort study on paediatric renal transplant recipients with a body weight below 10 kg at RTx (low-weight group, $n = 38$) compared to a matched control group (cases matched with two controls each) with a body weight of 10–15 kg at RTx (control group, $n = 76$) from data entered into the Cooperative European Paediatric Renal Transplant Initiative (CERTAIN) Registry. RTx from non-heart beating donors and AB0-incompatible transplantations were excluded. The matching criteria were as follows: (1) primary renal disease group (defined by risk of potential recurrence of primary renal disease and/or systemic organ impairment), (2) graft source (living or deceased donor), (3) immunosuppressive therapy (including calcineurin-inhibitor (cyclosporine microemulsion or tacrolimus), antiproliferative agent (mycophenolate mofetil (MMF) or azathioprine) with or without corticosteroids, and antibody induction therapy), and (4) number of HLA-mismatches (with a maximum difference of 1). Outcome variables were categorized as follows: (i) variables related to transplant surgery and the immediate postoperative period, (ii) transplant function (eGFR), (iii) number of treated acute rejection episodes (ARE) and (iv) transplant-related viral infections.
Results: Cases and controls were well comparable regarding demographics and clinical characteristics beside the expected differences in anthropometric data. Surgical-related complications were

rare and not significantly different (8% in the low-weight group, 17% in the control group; $P = 0.379$) (Table 1).

Table 1. Outcome measures of the surgical transplant procedure

Complications	Low-weight group ($n = 38$)	Control group ($n = 76$)	*P*-value
Vascular			
Renal artery thrombosis	0 (0)	1 (1)	1.000
Renal vein thrombosis	0 (0)	0 (0)	---
Laceration of the renal artery	0 (0)	0 (0)	---
Renal artery stenosis	0 (0)	0 (0)	---
Ureteral			
Leakage	0 (0)	0 (0)	---
Obstruction	2 (5)	5 (7)	1.000
Necrosis	0 (0)	0 (0)	---
Fluid collection			
Lymphocele	1 (3)	4 (5)	0.663
Perirenal hematoma	0 (0)	0 (0)	---
Seroma/urinoma	0 (0)	0 (0)	---
Miscellaneous*			
Graft compression	1 (3)	2 (3)	1.000
Wound infection	1 (3)	1 (1)	1.000
Total *	3 (8)	12 (17)	0.379

* leading to surgical re-intervention

For the first 2 years post-transplant, patient survival in both groups was 100% and graft survival 97% ($P = 1.00$). Estimated GFR at 2 years post-transplant was excellent and not significantly different between groups (low-weight, 95 ± 14 ml/min/1.73 m^2; control, 94 ± 12 ml/min/1.73 m^2). The respective frequencies of ARE, arterial hypertension, CMV-, EBV- or BKV-replication, CMV disease and BK-virus associated nephropathy, and malignancies were comparable. EBV disease occurred more frequently in the low-weight group ($p = 0.003$).

Conclusions: Our data demonstrate that RTx in low-weight children (<10 kg of body weight at time of engrafting) is associated with excellent graft function and overall outcome at 2 years post-transplant. It therefore represents a feasible option in these patients to potentially improve long-term outcomes, at least in selected centers with appropriate surgical and medical expertise.

P-407 PHARMACODYNAMIC EVALUATION OF ADV7103, AN INNOVATIVE PROLONGED-RELEASE ORAL ALKALISING FORMULATION

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Introduction: A new prolonged-release multi-particulate oral formulation, ADV7103, consisting of a fixed-dose combination of potassium citrate and potassium bicarbonate, was developed in order to improve the dosing scheme and reduce gastro-intestinal discomfort, main drawbacks of current alkalising treatments. The aim of the study was to evaluate the effect of two daily doses of ADV7103 on urine pH values.

Material and methods: A randomised, placebo-controlled, double-blind, two-period incomplete cross-over study was performed in healthy adults ($n = 16$). During period 1, subjects received two intakes (morning and evening) of one of three different doses of ADV7103 or placebo during 5 days. Urine pH was measured every 2 h during 24 h: at baseline (before treatment), during the two last treatment days, and during follow-up (after treatment was stopped).

Results: The treatment afforded significantly increased urine pH values compared to placebo ($p < 0.05$), with positive dose-response and non-saturating effect observed within the dose-range tested (1 to 3 mEq/kg/day). It showed alkali power of long duration, maintaining a stable urine pH between 7.0 and 7.5 throughout 24 h, with only two daily administrations of the highest dose. A strong circadian rhythm of urine pH was observed (Fig.1), composed of 2 cycles of 12 h in the absence of treatment. After administration of ADV7103 twice a day the circadian rhythm was still observed, with acrophase and bathyphase, respectively, +2–6 h and +8–12 h after administration, but the low urine pH phases were shortened with increasing ADV7103 doses. The observed circadian rhythm highlights the importance of the choice of the timing for the evaluation of urine pH values in clinical practice. No serious adverse events occurred and only one episode of nausea of mild intensity was observed with the highest dose.

Conclusions: These results of pH maintenance support further clinical evaluation of ADV7103 in patients requiring alkali.

P-408 AP(CAOX) INDEX AND CALCIUM/CITRATE RATIO MAY REPRESENT USEFUL TOOLS TO ASSESS THE RISK OF CRYSTALLIZATION IN PEDIATRIC RENAL LITHIASIS

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Introduction: Renal lithiasis is a complex process in which not only metabolic abnormalities but interaction among solutes concentrations play a major role. Several tools have been developed in order to quantify the risk of crystallization but there is little information about clinical utility in children. We have explored calcium/citrate ratio and AP(CaOx) index in stone-forming children (SF) and in healthy children (HC).

Material and methods: 24-h urine was collected from 87 HC and 14 SF. Pharmacological treatments were discontinued 3 days before analysis. Urine volume, calcium, magnesium, phosphate, urate, citrate and oxalate were determined. Calcium/creatinine, citrate/creatinine, calcium/citrate, and AP(CaOx) index were calculated.

Results: We found significant differences between HC and SF regarding calcium concentration (P_{50} 7.2 mg/dl; P_{25}-P_{75} 4.5–12 vs 21; 9.9–26.5, $p < 0.001$), calcium/creatinine (0.08 mg/mg; 0.05–0.11 vs 0.16; 0.15–0.23, $p < 0.001$), citrate/creatinine (542 mg/g; 367–724 vs 385; 188–613, $p = 0.05$), calcium/citrate (0.15 mg/mg; 0.09–0.24 vs 0.45; 0.32–1.13, $p < 0.001$) and AP(CaOx) (0.55; 0.33–0.83 vs 1.33; 0.81–1.81, $p < 0.001$). Nevertheless, most of SF had calcium/creatinine and citrate/creatinine values in normal range (calcium/creatinine >0.21 mg/mg, 3/14 patients, citrate/creatinine <250 mg/g, 4/14), but calcium/citrate >0.33 mg/mg was observed in 11/14 patients.

Conclusions: Most of SF had calciuria and citraturia in normal range but calcium/citrate and AP(CaOx) index showed significant differences between SF and HC. Calculation of risk formulas may be supplementary useful tools for decision making in the evaluation of the stone-forming patients. Higher values of the indexes seem to be associated with increased lithogenic risk.

P-409 ALKALISING TREATMENTS USED TO TREAT DISTAL RENAL TUBULAR ACIDOSIS (DRTA) IN CLINICAL PRACTICE – OBSERVATIONS DURING A CLINICAL STUDY

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Introduction: Treatments used in dRTA consist of oral alkalising agents, usually as bicarbonate and/or citrate salts. The aim of this work was to present information gathered on current treatments in order to assess the standard of care (SoC) approach in clinical practice and compare it to treatment with ADV7103, a new prolonged-release combination of potassium citrate and potassium bicarbonate.

Material and methods: Adults, adolescents, children and infants with dRTA, considered to be adequately controlled with their SoC therapy, were enrolled in a clinical study (B21CS): 35 patients in France, one in Slovakia and one in Serbia. They received their usual SoC and then ADV7103 at appropriate doses, during consecutive 5-day periods. Descriptive analysis of the SoC treatments was performed and their limitations discussed in comparison with ADV7103.

Results: Pharmacy preparations were used by all patients in the form of immediate release formulations (capsules, powders, syrups, solutions). A great diversity of products was observed, mainly consisting of bicarbonate or citrate, either as potassium or sodium salts, or as mixtures (Table 1).

Table 1. Number (%) of patients taking each SoC medication

Alkalinising used (whatever the formulation type)	**Adults (≥18 yrs) *N* = 7**	**Adolescents (12–17 yrs) *N* = 10**	**Children (4–11 yrs) *N* = 15**	**Infants (0.5–3 yrs) *N* = 5**	**TOTAL *N* = 37**
Potassium citrate	4 (57.1)	3 (30.0)	1 (6.67)	0 (0.0)	8 (21.6)
Sodium bicarbonate	1 (14.3)	1 (10.0)	2 (13.3)	3 (60.0)	7 (18.9)
Potassium bicarbonate	0 (0.0)	2 (20.0)	1 (6.67)	0 (0.0)	3 (8.1)
Modified Shohl's solution(sodium citrate, citric acid, potassium citrate)	0 (0.0)	0 (0.0)	1 (6.67)	0 (0.0)	1 (2.7)
Potassium bicarbonate +Potassium citrate	0 (0.0)	1 (10.0)	2 (13.3)	0 (0.0)	3 (8.1)
Potassium bicarbonate + Sodium bicarbonate	1 (14.3)	0 (0.0)	3 (20.0)	1 (20.0)	5 (13.5)
Sodium bicarbonate + Potassium citrate	1 (14.3)	2 (20.0)	5 (33.3)	1 (20.0)	9 (24.3)
Mod. Shohl's solution + sodium bicarbonate	0 (0.0)	1 (10.0)	0 (0.0)	0 (0.0)	1 (2.7)
Taking > 1 medication	**2 (28.6)**	**4 (40.0)**	**10 (66.7)**	**2 (40.0)**	**18 (48.6)**

Half of the patients took two alkalising products. Some of them (11%) additionally took potassium supplements, such as potassium chloride, whilst ADV7103 (alone) afforded to treat both metabolic acidosis and, if required, hypokalaemia. For 84% of the patients, the number of daily intakes of SoC was ≥3 and a notable proportion of patients (30%) had to take their medication during the night. In contrast, only 2 doses of ADV7103 (morning and evening) were provided.

Conclusions: Great variability of products used as SoC was observed. Medication data highlighted the limitations of current SoC treatments in terms of number of daily intakes, need to take the medication during the night, and need of concomitant treatments. These issues, impacting the compliance, could be overcome with ADV7103 formulation.

P-410 NEPHROPATHIC CYSTINOSIS: 25 YEAR MULTICENTRE FOLLOW UP

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Introduction: We describe our experience of managing patients with nephropathic cystinosis across three centres and report their clinical course during follow-up of up to 25 years.

Material and methods: A retrospective observational study was undertaken of patients attending from 1990 to present day. Nineteen patients were identified with complete data available for eighteen. Data collected included age, height, weight and e-GFR at presentation and last follow-up. Clinical and biochemical data was recorded, including need for renal replacement therapy (RRT) or transplant (Tx). Data is presented as median and range.

Results: Weight, height and e-GFR at presentation are shown in Table 1.

Table 1: Features at Presentation

	Age (days)	Weight (kg)	Weight (z-score)	Height (cm)	Height (z-score)	e-GFR (ml/min)
Median	635	8.1	−2.33	74.5	−2.63	58
Minimum	91	5.65	−3.44	61.4	−5.39	24.5
Maximum	3585	53.4	2.55	140.7	0.53	141.1

Patients were followed-up for median 12.0 years (1.8;23.2). Four patients required RRT with 3 transplanted. Median time from presentation to RRT/Tx 11.4 years (7.5;13.5). At study end 3/18 were receiving thyroxine, all had a normal HBA1C and, excluding 3 transplanted patients, all had significant proteinuria. The median white cell cystine level at last follow-up was 0.745 (0.22;1.91) at a Mecaptamine dose of 42.9 mg/kg/day (26.1;59.9). 14/18 required gastrostomy placement. Median z-score at study end for height was −1.19 (−7.7;0.3) and weight − 1.07

(−7.1;2.41). Change in z score from presentation for height was 1.045 (−2.31;3.49) and weight 1.07 (−3.66;2.46).
Conclusions: The clinical course of patients with nephropathic cystinosis has improved since the introduction of cysteamine, presenting adult nephrologists with a cohort of patients previously unseen. At our institution we undertake a combined clinic with adult nephrologists. Four of five patients transitioned were transplanted or receiving RRT. We demonstrate that whilst growth improves from presentation patients with cystinosis remain significantly shorter than their peers despite growth hormone and supplemental nutrition.

P-411 AN INNOVATIVE TREATMENT AS PROLONGED-RELEASE GRANULES FOR DISTAL RENAL TUBULOPATHY ACIDOSIS (DRTA) SPECIFICALLY DESIGNED FOR TWICE A DAY TREATMENT

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Introduction: There is no registered treatment for dRTA. Current treatments are based on alkalizing agent and potassium supplements to correct bicarbonataemia value and restore normal kalaemia. But those standards of care have important limitations impacting efficacy and compliance, such as:
• Inappropriate pharmaceutical form especially for children
• Repeated daily intakes due to short duration of action
• Poor gastro-intestinal tolerability
The objective was to develop pharmaceutical form suitable for children and adults in order to (i) prolong efficacy of alkalizing treatment, (ii) decrease the number of intakes to only 2 per day to reduce the burden of the regimen and to improve compliance.
Material and methods: Advicenne has developed a fixed-dose combination with two specific types of coated prolonged-release, neutral taste granules containing a high load of potassium citrate and potassium bicarbonate respectively. The release features of the new formulation were evaluated in an *in-vitro* study and the efficacy with a human *in vivo* study.
Results: Advicenne developed a prolonged release, tasteless, sodium free, 1/3–2/3 potassium citrate/potassium bicarbonate, 2 mm coated granules (mini-tablets) that combines the advantages of both agents in one appropriate pharmaceutical oral form for children and adults with only a twice-a-day administration.
Their dissolution profiles were respectively defined reflecting physiological reasons:

- Release of potassium citrate over three hours with aim to target liberation of the drug in the upper intestinal tract where citrate is mostly absorbed
- Prolonged release of potassium bicarbonate over twelve hours, to achieve the long duration of action needed to cover a twice-a-day administration and to decrease the release in the stomach responsible of GI side events.

The prolonged efficacy and GI improvement was demonstrated in the human study.
Conclusions: The combined patented granules form, provides a prolonged effect on the normalization of alkalizing effect over 24 h with only a twice-a-day intake.

P-412 PHENOTYPICAL DIFFERENCES IN PATIENTS WITH DISTAL RENAL TUBULAR ACIDOSIS CLASSIFIED ACCORDING TO THE UNDERLYING GENE DEFECT

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Introduction: *Collaborators: Ainhoa Iceta Lizarraga, Juan Alberto Piñeiro Fernandez, Gema Ariceta y Sara Chocron, Virginia Cantos Pastor, Natalia Mejía, Neus Roca Saladrigues, Ana Peña Busto, Cristina Aparicio López, Rosa Delia López Ramirez, Inmaculada Nadal Lizabe, Susana Ferrando Monleón, Raquel Lopez Hidalgo, Fiona Blanco Kelly, Luz Esthella Gonzalez Chaparro, Inés Vergara Pérez, Ainhoa Iceta Lizarraga, Alejandro Cruz Gual, Gloria María Fraga Rodriguez. This study was designed to find out if patients with primary distal renal tubular acidosis (dRTA), caused by mutations in ATP6V1B1, ATP6V0A4 or SLC4A1 genes, have different clinical and biochemical manifestations.
Material and methods: Twenty nine (19 males) pediatric patients from the RenalTube database, aged from 1 month to 15 years, were grouped according to whether had mutations in ATP6V1B1 ($n = 10$), ATP6V0A4 ($n = 14$) or SLC4A1 ($n = 5$) genes. The following diagnostic characteristics were compared among the 3 groups: demographic data, family history, age, growth, biochemical values, hearing assessment and image studies.
Results: The three groups were not different in sex, ethnicity, severity of acidosis, growth retardation and the presence of nephrocalcinosis. Patients having mutations in SLC4A1 presented later onset (median and interquartile range) than the other two groups: 120 (45) months *versus* 7.50 (30) months and 3 (12) months in ATP6V1B1 and ATP6V0A4, respectively. Moreover, they had a higher serum potassium concentration (X ± SD) (3.66 ± 0.44 versus 3.31 ± 0.57 and 2.93 ± 0.57 mEq/l, respectively) ($p = 0.05$) and presented higher serum bicarbonate values: 18.34 ± 1.79 versus 15.19 ± 4.48 and 13.35 ± 4.66 mEq/l ($p = 0.104$). Patients with ATP6V1B1 mutations had the highest prevalence of hypoacusia (8/10 patients versus 2/14 ATP6V0A4 patients).
Conclusions: This study confirms that deafness is more frequent in patients with ATP6V1B1 defects, in spite of recent clinical reports suggesting that hypoacusia cannot be used to distinguish patients with ATP6V1B1 and ATP6V0A4 mutations. In addition, dRTA caused by SLC4A1 gene defect, very rarely involved in western population, presents later and tends to be milder.

P-413 TOLVAPTAN USED SUCCESFULLY TO TREAT SYNDROME OF INAPPROPRIATE ANTI DIURETIC HORMONE SECRETION IN A CHILD

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Introduction: Hyponatremia is one of the most common electrolyte abnormality encountered in the clinical setting in hospitalized patients. Syndrome of inappropriate anti-diuretic hormone secretion (SIADH) is the leading cause of hyponatremia in most of the cases. Tolvaptan is a vasopressin type-2 receptor antagonist used for treatment of hyponatremia due to cirrhosis and congestive heart failure. There is limited data about tolvaptan usage in children for treatment of SIADH in the literature. Here we present an adolescent case of idiopathic SIADH that is treated successfully with tolvaptan. Sixteen year-old, female patient admitted with the complaint of headache and nausea. Physical examination and medical history was unremarkable. She was found to have hyponatremia (Na:110 mmol/l), hypochloremia (89 mmol/l), hypouricemia (1,4 mg/dl) with normal renal function tests, electrolytes and normal blood gas analyses. Serum osmolality was 276 ng/ml and urine osmolality was 565 ng/ml. Fractional excretion of Na was 1.9% with urine sodium level of 173 mmol/l, tubular phosphorus re-absorption was 78% and uric acid excretion rate was 834 mg/1.73 m2/d. She did not have any signs of dehydration and urine output was normal. Serum ADH levels during hyponatremia was also found to be decreased; 8.3 ng/mL. Her clinical and

laboratory findings, led us to the diagnosis of SIADH. She didn't respond well to fluid restriction. Thus, Tolvaptan was started on the second week at a dose of 0.28 mg/kg (15 mg/d) once a day and titrated down. Her response to treatment was perfectly well without any side effects. Treatment was continued for 2 weeks. In conclusion, tolvaptan is a safe treatment and it should be emphasized that tolvaptan could be an effective treatment option for refractory cases with careful monitoring of fluid and electrolyte status of the patient.
Material and methods:
Results:
Conclusions:

P-414 A NOVEL HOMOZYGOUS W99G MUTATION IN CLDN-16 GENE CAUSING FAMILIAL HYPOMAGNESEMIC HYPERCALCIURIC NEPHROCALCINOSIS IN TURKISH SIBLINGS

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Introduction: Familial hypomagnesemic hypercalciuric nephrocalcinosis (FHHNC) (OMIM: 248,250) is chracterized by hypomagnesemia, hypercalciuria and nephrocalcinosis. FHHNC inevitably progresses to end-stage renal disease in decades. Mutations in CLDN-16 and CLDN-19 genes are associated with distrupted magnesium handling in the thick ascending limp of Henle's loop. Patients with mutations in these genes share similar clinical features, and those with CLDN-19 gene mutations have ocular findings in addition.
Material and methods: Case I & II.
Results: Case I A 2-month-old boy, was admitted to our clinic with the complaints of upper respiratory tract infection. He was the first born child of consanginous parents. Laboratory findings revealed hypocalcemia and hypomagnesemia. Bilateral medullary nephrocalcinosis was detected on abdominal ultrasound. His ophthalmologic examination was unremarkable. With hypomagnesemia, hypercalciuria and nephrocalcinosis, patient was considered to have FHHNC. Oral magnessium supplementation was initiated. Four years of follow-up has been completed uneventfully. Case II A 6-day-old term male infant was admitted with seizure. The patient was resistant to calcium and anticonvulsant drugs and the seizure activity could only be controlled after magnesium infusion. His parents were consanguineus and his 4-year old brother was being followed due to hypercalciuria by the pediatric nephrology department. Biochemistry profile revealed hypocalcemia and hypomagnesemia. Calcium extraction was 11 mg/kg/day. Medullary nephrocalcinosis was reported on renal ultrasound. His eye examination, echocardiography, transfontanel ultrasound and electroencephalography were normal. Due to the triad of hypomagnesemia, hypercalciuria and nephrocalcinosis, and the medical history of his elder brother, he was diagnosed with FHHNC. After correction of the electrolyte abnormalities, he was discharged from hospital and is currently being followed-up without any problem.
Conclusions: In this manuscript, we shared our experience about a novel homozygous mutation (W99C) in CLDN-16 gene causing FHHNC in a couple of Turkish siblings.

P-415 DISTAL RENAL TUBULAR ACIDOSIS WITH TRANSIENT PROFOUND HYPERCALCEMIA IN TWO SIBLINGS

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Introduction: Metabolic acidosis, alkaline urine, hypokalemia, nephrocalcinosis secondary to hypercalciuria and hypocitraturia are frequent findings in distal renal tubular acidosis (dRTA). But profound hypercalcemia is rare in dRTA. We present here two siblings with dRTA and ATP6V1B1 mutations and hypercalcemia.
Material and methods: Case reports.
Results: Case 1: Two and half year's old girl admitted with hearing loss and growth retardation. The height/weight SDS was found −1.70 and 3.77 consecutively. In laboratory investigation renal function tests was normal and pH 7.26, HCO_3 15.6 mmol/l, potassium 3.0 mmol/l, calcium 13 mg/dl, phosphorus 5.1 mg/dl, magnesium 1.71 mg/dl, PTH <3 pg/l. Urine pH was changed between 7.0–8.5 with positive urinary anion GAP and hypocitraturia. She was diagnosed with dRTA and oral bicarbonate was started. She had height/weight SDS −0.44 and −0.91 consequently in last control at 5 years old. **Case 2**: Patient 2 (sibling of first patient) was investigated for family history at 40 days old. The height/weight SDS was found +1.01 and +0.62 consecutively. In laboratory investigation was found pH 7.32, HCO_3 14.1 mmol/l, potassium 4.0 mmol/l, calcium 13.6 mg/dl, phosphorus 5.5 mg/dl, magnesium 2.5 mg/dl, PTH <3 pg/l. Urine pH was changed between 7.0–8.5 with positive urinary anion GAP and hypocitraturia. She was diagnosed with dRTA and oral bicarbonate was started. She had height/weight SDS −2.77 and −1.55 consequently in last control at 2 years and one month old. We found bilaterally neurosensorial hearing defect and p.Pro346Arg homozygote mutation in ATP6V1B1 gene in both case. Serum calcium concentration was found normal in both sisters after starts of treatment.
Conclusions: Hypercalcemia is not frequently seen in patients with dRTA. Hypercalcemia should also be in mind in the definitive diagnosis of dRTA. In the cases with hearing loss with RTA, ATP6V1B1 mutation should be analyzed.

P-416 FUNCTIONAL STATE OF TUBULO-INTERSTITIAL TISSUE OF KIDNEYS IN CHILDREN WITH POISONINGS OF TOPICAL DECONGESTANTS

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Introduction: The aim of the study was to analyze activity of enzyme neutral α-glycosidase (NG) in children with poisoning of topical decongestants (TD).
Material and methods: We examined 66 children aged from 1 month up to 12 years with poisoning of TD. Control group had 50 healthy children similar age and sex. Activity of NG was determined in urine on a spectrophotometer "SF-103" (Russia).
Results: We established increase of excretion of NG in urine (212.22 ± 41.69 μmol/l) in children with poisoning of TD. The level of NG in urine in children of control group was 111.34 ± 18.75 μmol /l.
Conclusions: So, children with poisoning of TD had acute toxic damage of proximal tubules in tubulo-interstitial tissue of kidneys which was reversible.

P-417 LONG TERM FOLLOW-UP OF PATIENTS WITH PRIMARY TUBULOPATHIES: GITELMAN SYNDROME (GS) AND DISTAL RENAL TUBULAR ACIDOSIS (DRTA)

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Introduction: To analyze the long term outcome of a group of patients with DRTA and GS diagnosed in pediatric age. The underlying gene defect had been identified in all cases.
Material and methods: Six Gypsy patients with GS, aged between 18 and 37 years, and 5 patients with DRTA, from 19 to 40 years of age, were studied after 17.6 ± 3.3 and 21.0 ± 10.8 years (X ± SD) of follow-up, respectively Final height (Z-Score), renal function (FGE: CKD-EPI formula), comorbidities, therapeutic compliance and quality of life by the SF-36 questionnaire were analyzed.
Results: Only 2 DTRA and 1 GS patient reached the final height according to their genetic potential. FGE was normal except in 1 DRTA case (57 ml/min). All DRTA patients revealed a good adherence to potassium citrate treatment and to annual follow-up by a nephrology unit. All DRTA patients had nephrocalcinosis and hearing impairment. Only 2 out of the 6 GS cases declared an adequate compliance with magnesium and potassium supplements. Two patients received spironolactone. Two patients had several hospital admissions because of syncopal crisis. The rest of GS patients had refused to follow medical follow-up visits and did not take the treatment on their own initiative. The analysis of the quality of life showed that while GS patients presented the lower values in physical limitations and corporal pain, DRTA cases had the lower scores in social and emotional roles.
Conclusions: Most adults having GS and DRTA diagnosed in pediatric age do not achieve their genetic height potential. They usually maintain a normal glomerular function rate. The adherence to treatment is good in DRTA patients and poor in GS individuals. The impact on the quality of life was greater on physical parameters in GS whereas DRTA patients reported a low score in regard with psychological items.

P-418 TINU SYNDROME - CASE REPORT

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Introduction: Tubulointerstitial nephritis and uveitis syndrome (TINU) is a rare disorder occuring in less than 2% of cases of uveitis. Diagnosis requires the presence of both TIN and uveitis. The most common signs and symptoms of uveitis include photophobia, eye pain and redness, eyelid edema and progressive loss of vision. Renal impairment is characterized by abnormal renal function and abnormal urinalysis, symptoms of systemic illness, including fever, fatigue and weight loss.
Material and methods: Case report 14 -year old boy treated for acute anterior uveitis was admitted to our department because of increased serum creatinine level. On admission, physical examination was normal and uveitis was in remission with only persistence of mild visual impairment of the right eye. He had no other clinical symptoms except of polyuria (4000 ml /day) and associated polydipsia.
Results: Laboratory tests confirmed increased serum creatinine (189 μmol/L), other biochemical parameters were within normal reference range. Urinalysis showed normoglycemic glycosuria, non-nephrotic glomerulo-tubular proteinuria and high levels of β-2 microglobulin. Renal biopsy was consistent with tubulointerstitial nephritis, with chronic inflammatory changes and tubular atrophy. To complete the diagnostic work-up of TINU, we excluded infection, systemic and autoimmune causes. Because of persistent impairment of renal function we decided to use oral corticosteroids, which resulted in renal function improvement. However, after tapering of corticosteroids the boy had recurrence of uveitis and nephritis with decreased renal function. Treatment with topic and oral corticosteroids led to uveitis control, but renal functions remain impaired.
Conclusions: TINU syndrome is probably an underdiagnosed disorder and must be actively searched in either patients with uveitis or tubulointerstitial nephritis. Symptoms of uveitis and nephritis are not always present at the same time. Tubulointerstitial nephritis in our patient very probably preceded the manifestation of uveitis by several months.

P-419 ATTENTION TO CHROMOSOME 16: A MALE CHILD WITH ASSOCIATION OF SCL12A3 AND M694 V HOMOZYGOUS MUTATION

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Introduction: A 14 year old boy, who had recurrent abdominal pain and fever episodes since 3 years old, diagnosed familial mediterrian fever (FMF) at the age of 9 and started colchicine treatment.
Material and methods: Genetic analysis showed that homozygote M694 V mutation on chromosome 16p13.3.
Results: Hypokalemia, hyponatremia, metabolic alkalosis, hypomagnesemia and hypocalciuria was detected and Gitelman syndrome was diagnosed at the age of 14. Diagnosis confirmed by genetic analysis and it showed that homozygote mutation in the SCL12A3 gene on chromosome 16.
Conclusions: The patient was presented as an interesting case due to the fact that both genetic mutations are on chromosome 16. Although the association of FMF and Gitelman syndrome is rare, it should not be forgotten.

P-420 A CLINICAL CASE OF PATIENT WITH CYSTINURIA AND NEPHROLITHIASIS

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Introduction: Cystinuria (OMIM 220100) is an autosomal disorder characterized by impaired epithelial cell transport of cystine and dibasic amino acids (lysine, ornithine, and arginine) in the proximal renal tubule and gastrointestinal tract. It is a rare, but important cause of urinary stone disease, that has high recurrence rate.
Material and methods: We present the clinical case of 6.5 year old boy with cystinuria and complicated treatment of nephrolithiasis.
Results: The boy was observed by the pediatrician from 4 months with a bilateral nephrolithiasis without urinary obstruction. The family history for renal lithiasis was negative. In 1 year old (IX/2011) renal colic was noted, after which - only in the left kidney stones were visualized. Urinalysis revealed numerous red blood cells per high-power field on microscopic examination, in the absence of proteinuria. Urine analytes such as calcium, oxalate, and uric acid were within normal ranges, cystinuria was not detected. At the age of 4.5 year old (II/2015) he had the signs of a repeated renal colic, and imaging studies, including ultrasonography, computer tomography demonstrated bilateral large radiodense calculi in left major calyx and left pelvis. Urologist evaluated the patient and extracorporeal shockwave lithotripsy (ESWL) on the left side was performed. During few weeks steinstrasse and ureter dilatation was developed, and repeat ESWL with cystoscopy was held (IV/2015). Chemical composition of a stone showed 100% cystine. Urine aminoacid analysis revealed increased levels of ornithine and arginine. Medical treatment was initiated with sodium bicarbonate to achieve urine alkalinization to a pH of 7.0 and captopril in a dose of 0.5 mg/kg/day, and high fluid intake and a low-sodium diet was suggested. Within 2 months the signs of obstruction were increased, imaging studies

demonstrated recurrence calculi; 99 m Tc-Dimercaptosuccinic acid renal scan (DMSA) revealed abnormal left kidney and normal right kidney, with differential renal function of 13% and 87% respectively. Due to the obstruction and nephrolithiasis, an urologist performed percutaneous nephrolithotomy (PCNL) with installation of an internal ureteral stent (VI/2015), which was removed in a three months (IX/2015). According to the results of conducted studies the stones was not recorded, but there was a loss of function of the left kidney (on DMSA 0% of renal function) and the laparoscopic nephrectomy was performed (IV/2016). Genetic analysis identified previously reported heterozygous mutations in the SLC3A1 gene (in 4 exon (c.808C > T) and 8 exon (c.1400 T > C) – compound heterozygous). The 1-year follow-up showed stable GFR ~ 112 mL/min per 1.73 m2, microalbumin/creatinine level < 10 mg/g, normal rate of cystine excretion is around 0.10 mmol/day, normal arterial blood pressure (98/60 mmHg). The treatment (high fluid intake, low-sodium diet, urine alkalinization, captopril) is continued.
Conclusions: Cystinuria is a rare disease requiring the cooperation between a nephrologist and urologist. Morbidity from stone formation and repeated urological interventions can be reduced by early diagnosis and adequate medical treatment. The high recurrence rate of stone formation along with the early age of first stone formation necessitates long and close follow-up.

P-421 PSEUDOHYPOALDOSTERONISM – REPORT OF THREE CASES

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Introduction: Pseudohypoldosteronism type I (PHA-I) is a heterogeneous syndrome characterized by the salt waste as a result of target organ unresponsiveness to mineralocorticoids. It is inherited in an autosomal recessive or autosomal dominant pattern, not rarely as a result of mutation *de novo*. It can be subclassified into two distinguishable clinical entities: renal PHA (type 1 PHA-I) and systemic/multiple target organ defects (type 2 PHA-I). The third recognizable entity is the secondary (transient) PHA (type 3 PHA).
Material and methods: Three patients were studied in order to characterize PHA in infants. Patients were selected by chart review.
Results: Three cases of PHA, 2 with renal type PHA-I and 1 with type 3, have been described. First two described cases were diagnosed with renal PHA-I. In one case salt supplementation was performed for a short period (for 24 h) but in the second patient it was performed for 1.5 year. Because of oliguira and severe electrolyte dysbalans, peritoneal dialysis was performed for five days in our third patient with transient PHA. After acute renal impairment, complete renal function recovery wasn't achieved and 1st grade chronic renal insufficiency developed.
Conclusions: In all patients with salt-wasting and dehydration differentiation between congenital adrenal hyperplasia and PHA should be performed. Also, in the cases with hyperkalaemia, hyponatremia and metabolic acidosis, urinary tract infection and obstructive uropathy should be excluded.

P-422 GITELMAN SYNDROME WITH EARLY ONSET

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Introduction: Gitelman syndrome (GS) is caused by genetical mutation resulting in dysfunction of the thiazide-sensitive sodium-chloride cotransporter located in the distal convoluted tubule. The disease usually produces first visible symptoms during adolescence or adulthood.
Material and methods: This paper describes case of a boy who developed first symptoms of GS at the age of 6 and was diagnosed when 16.
Results: Case report.
At the time of admission of the 16 year old boy the following information was provided: the child suffered from upper limb muscle cramps recurring every few weeks since the age of six. Lower limb and orbicularis oris muscle cramps were also observed. In addition, during fever, the patient presented additional symptoms: numbness of the face and limbs. The patient was treated in an ED with anxiolytics and magnesium. Two weeks before admission he developed diarrhea and fever, followed next day by severe muscle cramps with forced limb position and finally short term loss of consciousness. The boy was admitted to local hospital, where laboratory tests showed hypokalemia, hypomagnesemia and metabolic alkalosis, and then transferred to Clinic of Paediatric Nephrology. Blood tests confirmed earlier results and in addition revealed hypochloremia and increased plasma renin activity. The urine tests had following results: basic pH, decreased specific gravity, increased excretion of potassium and magnesium and reduced excretion of calcium. Blood pressure and USG abdomen were normal. Genetic testing was performed and two heterozygous mutations: 2221 G → A (Gly741Arg) oraz 1315 G → A (Gly439Ser) were found in *SLC12A3* gene.
Conclusions: GS may occur early in life and produce only short passing episodes when symptoms are observable. The presence of said symptoms requires considering tubulopathy during diagnosis. If not diagnosed and not treated GS may lead in the long term to somatic complications and decreased quality of life.

P-423 A CASE REPORT OF EFFECTIVE HYPOTHIAZIDE TREATMENT IN A 15-YEARSE-OLD GIRL WITH PSEUDOHYPOALDOSTERONISM TYPE II

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Introduction: Pseudohypoaldosteronism type II (Gordon syndrome, familial hyperkalemic hypertension) is a rare, autosomal-dominant disorder characterized by sever hypertension, hyperkalemia, metabolic acidosis, and low serum renin level due to mutations in the genes *WNK1, WNK2, CUL3, KLHL3*.
Material and methods: We report a patient with pseudohypoaldosteronism type II.
Results: A girl, the second child of unrelated parents with positive family history of cardiovascular disease includes hypertension, heart attack, cerebrovascular insult. At the age of 10 years first measurement of BP was revealed hypertension, stage 2 (180/120 mmHg). Treatment with ACE-I was started at the dosage of 0.2 mg / kg/d, but without effect of hypertension. The girl was refined to our clinic further evaluation. On admission at the age of 15 years the girl had short stature (height 154 cm (3 pc), weight 52 kg (25–50 pc)), hypertension, stage 2 without any phenotypical features. On examination she has high serum potassium level 6.0 mmol/l (N: 3.5–5.2), metabolic acidosis (pH 7.31; base excess −6.2 mmol/l). Serum hormones showed low renin (0.2 ng/ml (N 1.9–6.0)), high aldosterone (540.0 pg/ml (N: 12.0–340.0)); thyroid hormones and cortisol were normal. Excretion of catecholamine metabolites with urine was normal. She had normal renal function, eGFR 100 ml/min/1,73m^2. She had hypercalciuria with high level of Ca/creatinine (1.2 mmol/mmol ($N < 0.6$)) ratio. Renal US scan, echocardiography, X-ray of the hands, MRI of the brain and adrenal glands were normal. In adolescent with severe hypertension, hyperkalemia, metabolic acidosis with normal GFR, low serum renin was diagnosed pseudohypoaldosteronism type II. The girl has been treated with low doses of hypothiazide (0.46 mg/kg/d). This led to normalized of BP (mean BP<90‰) and all laboratory findings, including potassium levels (3.7–4.0 mmol/l), acid-base composition, Ca/creatinine (0.4 ($N < 0.6$ mmol/mmol) ratio after 7 days therapy. Follow-up after 6 months of hypothiazide treatment showed normal BP, blood electrolytes and acid-base composition.
Conclusions: We have not performed the molecular genetics test in our patient, due to clear phenotype of disease with effective hypothiazide treatment. However, according to recent date about phenotype-genotype associations we can suggest that our girl with early appearance of hypertension, severe hyperkalemia has a mutation, based on *CUL3* gene.

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