1198 PERIPHERALLY INSERTED CENTRAL VENOUS CATHETERS IN NEONATE: A QUICK AND EFFICIENT STANDARDIZED TECHNIQUE

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Background and Aims Peripherally inserted central venous catheters (PICCs) are commonly used for neonatal vascular access. Early catheter-related sepsis and catheter non infectious complications are linked with initial dressing method. We evaluate efficacy of our specific PICCs insertion protocol with trained nurses and doctors.

Methods We observed prospectively 1686 PICC procedures in neonate (0–28d) from 2002 to 2011. Silicon PICCs were inserted from 0 to 28 days corrected age. Procedure followed our specific protocol.

Results 1686 PICCs were attempted with a success rate of 96.2% in neonates with a mean gestational age of 29.7 weeks at a median age of 3 (0–94d) days of life.

In the successful PICCs, median number of venous puncture was 1 (1–13), median time spent was 20 (5–120) minutes, device change in 11.7% and site change in 5.9%. Median temperature difference between the beginning and the end of the procedure was very low: -0.2°C (-1.5 to 1.8). 187 complications (11.5%) occurred: 105 diffusions of which 6 pericardic effusions, 25 occlusions, 37 porous catheters, 7 surrounding catheter tissue inflammations. 13 PICCs were removed for infection (sepsis or local mycosis).

Conclusions Standardized protocol with specific nurse and doctor leads to a success rate of 96.2% with a small time spent for insertion and a median of one attempt mostly in the initial chosen site. This quick method leads to low neonate cooling and expose patients to minimal infection risk and complications.

1199 METABOLIC RISK FACTORS AND PROGNOSIS IN CHILDREN WITH UROLITHIASIS

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Objective To investigate the demographic characteristics, clinical features, and metabolic risk factors of children with urolithiasis.

Methods This retrospective study enrolled 52 boys, 51 girls with urolithiasis diagnosed by ultrasonography. Mean age at presentation was 60(1–192) months, and mean follow-up period was 5.5 (1–27) months.

Results The most common symptom was restlessness in infants (< 1 year), while it was abdominal or flank pain in older children ($p<0.001$). Microcalculi (stone diameter < 3mm) and calculi (>3mm) were found in 26% and in 74% of patients, respectively. Hypercalciuria was the most common abnormality, followed by hypomagnesiuria (Table 1). Recurrent urinary tract infection (UTI) was detected in half of the patients. Four patients underwent ESWL, five underwent open surgery, and the other 94 were treated with conservative therapies. Spontaneous passage occurred in 19 patients. Stone analysis revealed calcium-oxalate in 85%. At the time of their last visit, in 70% of the patients with conservative therapies, the

stones were disappeared or diminished in size by appropriate therapy such as water intake, diet, hydrochlorothiazide and potassium-citrate.

Conclusion Identifying the underlying metabolic risk factor is important in order to choose the appropriate treatment modality, prevent stone recurrence and renal damage. Patients presenting with restlessness, especially infants must be evaluated in terms of renal stone disease by ultrasonography.

1200 THE CORRELATION BETWEEN RENAL DAMAGE AND CLINICAL AND LABORATORY FINDINGS IN CHILDREN WITH ACUTE PYELONEPHRITIS

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Objective The aim of this study was to determine the risk factors such as age, sex, duration of fever, voiding dysfunction, constipation, history of recurrent UTI, the levels of C-reactive protein (CRP), erythrocyte sedimentation rate (ESR), white blood cell (WBC), polymorphonuclear leukocytes (PNL), platelet count, mean platelet volume (MPV), serum urea, creatinine, proteinuria, type of microorganisms, presence of vesicoureteral reflux(VUR) for the development of renal damage in patients with acute pyelonephritis.

Material and Methods A total of 197 children (44 boys, 153 girls) were enrolled in the study, mean age was 4.7 ± 4.0 years (1 month–16 years). Ultrasonography, renal cortical scintigraphy (RCS) with ^{99m}Tc-Dimercaptosuccinic acid and voiding cystourethrography were performed in all patients. The patients were grouped by age according to presumed risk of renal damage: high risk(≤ 1 year), moderate risk(1–5 years), and low risk (>5 years).

Results Renal lesions on RCS were detected in 91 patients. Abnormal RCS were found in 35% of infants younger than 1 year, in 57% of children between 1–5 years and in 42% of children older than 5 years. Abnormal RCS was found in 66% of the children with VUR and in 42.7% of those without VUR($p=0.005$). There was a significant positive correlation between abnormal RCS and VUR, duration of fever (>2 days), history of recurrent UTI, high levels of ESR, CRP, WBC, and PNL at the presentation.

Conclusion The patients with VUR were 2.6 times more likely to have renal damage. The risk of renal damage should be considered in all age groups.

1201 WHOLE EXON DELETION IN THE CLAUDIN 16 GENE, A NOVEL MUTATION IN FAMILIAL HYPOMAGNESEMIA/HYPERCALCIURIA/NEPHROCALCINOSIS (FHHNC) AND SENSORINEURAL DEAFNESS (SND)

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Abstract 1199 Table 1 Urine metabolic analysis

Risk factor	Patients(%)	Risk factor	Patients(%)	Risk factor	Patients(%)	Risk factor	Patients(%)
Hypercalciuria	32(31.1)	Cystinuria	4(3.9)	Hyperoxaluria	2(1.9)	Hypercalciuria+hypernatruria	2(1.9)
Hypomagnesiuria	13(12.6)	Hyperuricosuria	3(2.9)	Hypercalciuria+hypocitraturia	4(3.9)	Hypocitraturia+hypomagnesiuria	2(1.9)
Hypernatruria	11(10.7)	Hypocitraturia	3(2.9)	Hypercalciuria+hyperoxaluria	3(2.9)	Normal	24(23.3)

Background FHHNC is a rare autosomal recessive tubulopathy of the thick ascending limb due to inactivating mutations in the *Claudin-16* and *Claudin-19* genes which are responsible for the paracellular reabsorption of calcium and magnesium. Clinically, FHHNC is characterized by urinary tract infections, nephrolithiasis, nephrocalcinosis and progressive renal failure.

Objective To present clinical and molecular data on 2 siblings with FHHNC and early onset SND, found to have a new mutation in the *Claudin-16* gene.

Methods A 16-year-old male and his 14-year-old sister were diagnosed with chronic kidney disease since early infancy, manifested by recurrent UTIs, polyuria and polydipsia, poor growth, mild mental retardation and delayed speech. SND was diagnosed at the age of 6 and 7 years. Both have hypomagnesemia, hypermagnesuria, hypercalciuria and nephrocalcinosis. Genotyping was performed by PCR using tetranucleotide repeat polymorphisms. Specific primer pairs of genomic DNA were used as template for sequencing the *Cldn16* gene.

Results Exons 1, 2, 4 and 5 were amplified and revealed wild type sequencing but exon 3 failed in amplification by PCR. Long range PCR spanning exon 2 to 4 of the gene yielded a 2.5 kb fragment shared by the patients. Sequencing of this fragment reveal a 2630 bp deletion including the entire exon 3 resulting in deletion and frame shift of *Cldn16* protein.

Conclusion This family demonstrates the first identified homozygous mutation in the *Cldn16* gene causing the deletion of an entire exon. This, however, is unlikely to explain the SND, since this gene is not expressed in the cochlea.

1202 PROTEOLYTIC ENZYMES IN GLOMERULONEPHRITIS

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Aim of the investigation: Study of activity of proteolytic enzymes in children with acute (AGN) and chronic (CGN) glomerulonephritis.

Materials and Methods Were investigated 50 children with acute and chronic glomerulonephritis, age from 7 to 15 years, including 30 children with AGN plus nephrotic syndrome, and 20 children with CGN, nephrotic form, period of activation and remission. Control group including 20 practically healthy children. In serum determined activity of trypsin, a-antitrypsine, and a₂-macroglobuline.

Results In AGN with nephrotic syndrome, at the initial stage of the process, as well as in activation of CGN, were determined significant increasing of trypsin, a-antitrypsine, and a₂-macroglobuline in comparison to control group (P<0.01). The activity of a₂-macroglobuline in children with AGN plus nephrotic syndrome at the initial stage of disease was 11.9±1.42 g/l, P<0.001, what 1.9 fold higher than in control group (6.2±0.030 g/l). In activation of CGN, nephrotic form, the activity of a₂-macroglobuline was in 1.5 fold higher the indexes of control group (9.5±0.90 g/l, P<0.01).

In remission, the values of trypsin, a-antitrypsine, and a₂-macroglobuline decreasing, but did not attain the basic level of control group, which suggests the persistence of activity of pathological process in kidneys.

Conclusion Determining of activity of proteolytic enzymes in serum in diagnostic criterion for determining of severity, activity of pathological process in kidneys, and determining the outcome the complications of glomerulonephritis.

1203 THE DIABETIC PREGNANCY AND OFFSPRING BLOOD PRESSURE IN CHILDHOOD: A SYSTEMATIC REVIEW AND META-ANALYSIS

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Background and Aims Offspring of diabetic mothers (ODM) are at increased risk of the metabolic syndrome in later life. We aimed to perform a systematic review and meta-analysis of studies examining offspring systolic and diastolic blood pressure (SBP, DBP) in childhood in relation to maternal diabetes.

Methods Citations were identified in PubMed. Authors were contacted for additional data where necessary. SBP and DBP in ODM and controls were compared. Subgroup analysis was performed according to type of maternal diabetes and offspring gender. A fixed effect meta-analysis was performed, and a random effects analysis where significant heterogeneity was present. Meta-regression was used to test the relationship between offspring SBP and maternal pre-pregnancy BMI.

Results Fifteen studies were included in the systematic review and thirteen in the meta-analysis. SBP was 1.88mmHg higher in ODM (95% CI 0.47, 3.28; p=0.009). The increase in SBP was similar in both offspring of mothers with gestational diabetes (1.39mmHg [0.00, 2.77]; p=0.05) and type 1 diabetes (1.64mmHg [0.09, 3.18]; p=0.04). Male ODM had higher SBP (2.01mmHg [0.93, 3.10]; p=0.0003) and DBP (1.12mmHg [0.36, 1.88]; p=0.004) than controls, but the differences in SBP and DBP between female ODM and controls were not statistically significant. Offspring SBP was positively correlated with maternal pre-pregnancy BMI; however, the association was not significant (p=0.37).

Abstract 1203 Figure 1

Conclusions ODM have higher SBP than controls. This increase is independent of type of maternal diabetes and may be related to maternal pre-pregnancy BMI. Gender-specific differences require further investigation.

1204 UTILITY OF SERUM PROCALCITONIN TO PREDICT RENAL SCARS IN INFANT WITH FEBRILE URINARY TRACT INFECTION

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Background and Aims The aim of this study was to evaluate the usefulness of Procalcitonin(PCT) as a marker for renal scars in infants with a first febrile urinary tract infection(UTI).

DMSA scintigraphy, the gold standard for detection of cortical scarring, has clearly shown that not all febrile UTI are associated with renal lesions and that common clinical and laboratory evaluations are not reliable to distinguish between acute pyelonephritis(APN) and simple UTI. Scarring secondary to APN is a common event occurring in approximately 30% of all cases.

PCT, prohormone of calcitonin has been measured in various systemic inflammatory response syndromes, because it appears to be correlated with the severity of microbial invasion and it can be used to check for the presence of parenchymal scars.