mostly grade II (41; 60.3%) and grade III (20; 29.4%). Of the total number of children with a prenatal diagnosis of hydronephrosis, 28,6% had VUR.

In the group of children with first febrile UTI, VUR was discovered in 30 (36,6%). With regard to VUR, no significant difference was found between normal pathologic ultrasound findings (38.1% vs. 41.7%; $p = 0.739$). No difference was found regarding severity of UTI: urine Leukocyte < 20 vs. urine Leukocyte > 20 (40.3% : 43.7%; $p = 0.787$), CRP < 50 vs. CRP > 50 (37.5% : 35.5%; $p = 0.46$) as well as between age of children < 1 year vs. > 1 year (34,5% : 41,3%; $p = 0.141$). Significant differences were found between girls and boys (M vs.F = 19.0% vs.34.6%; $p = 0.0466$).

Analyzed data point to a higher likelihood of background VUR in recurrent UTI; whereas gender, age, urinary tract ultrasound examination, severity, and other causative agents of UTI have no predictive value for VUR detection. ceVUS is one of the best choices in modern nephrology for the detection of VUR with high sensitivity and negligible side effects.

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A RARE CASE OF POLYCYSTIC KIDNEY DISEASE AND MULTICYSTIC DYSPLASTIC KIDNEY IN A PEDIATRIC PATIENT

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10.1136/archdischild-2021-europaediatrics.357

Introduction Cystic kidney disease (CyKD) is one of the most important causes of chronic kidney disease (CKD) in children. Multiple kidney cysts can occur unilaterally (e.g. multicystic dysplastic kidney – MCDK) or bilaterally (e.g. autosomal dominant or autosomal recessive polycystic kidney disease) due to genetic or non-genetic (developmental or acquired) disorders. Here we present a rare case of combined polycystic kidney disease (PCKD) and MCDK in a pediatric patient.

Case Report The patient was admitted to our hospital for the first time at the age of two. She was normally developed (weight: 10.8 kg – 19th percentile, height: 90 cm – 80th percentile) with blood pressure (BP) within normal limits.

Estimated glomerular filtration rate (eGFR) at that time was 70.2 mL/min with elevated albumin/creatinine ratio 5,2 mg/mmol and she was diagnosed with G2A2 stage of CKD. During her prenatal period oligohydramnios and polycystic kidneys were detected. At birth a palpable mass of the left abdomen was found. First sonography after birth showed hydronephrosis of the right kidney and multiple cysts on the left kidney. Micturating cystourethrography was negative for vesicoureteral reflux. Initial magnetic resonance imaging (MRI) showed enlargement of both kidneys with multiple cysts of various sizes on the left kidney and lesser number of diffuse renal cysts on the right kidney. Cysts weren't found neither on the brain nor on the other abdominal organs. Echocardiography was normal. Family history was negative for kidney diseases. The abdominal and kidney ultrasound of both parents and older brother was normal. At the age of two dynamic scintigraphy with Tc-99m revealed nonfunctional left kidney leading to diagnosis of MCDK.

Follow-up ultrasound showed progressive involution of the MCDK left and multiple very small cysts that generate abnormal parenchymal echogenicity (eg, salt-and-pepper sign) on the right kidney. Functional MRI urography showed non-functional MCDK left and functional right kidney with lesser number of small cysts. At the age of four arterial hypertension (130/70 mmHg) was diagnosed and ACE inhibitor was introduced. There were no signs of liver disease. Abdominal ultrasound revealed no cysts of liver, spleen or pancreas. Gynecological ultrasound and ophthalmological examination were also normal. Due to parental disapproval, genetic testing wasn't performed.

Conclusion We conclude that, although rare, different types of CyKD can be associated and we should consider it when setting the diagnosis. Due to vast differential diagnosis and overlapping clinical presentations of CyKD genetic testing should be performed whenever possible.

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RECURRENT MULTI DRUG RESISTANT URINARY TRACT INFECTIONS IN A THREE-YEAR-OLD HOSPITALIZED CHILD WITH HYPOTONIC CEREBRAL PALSY

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10.1136/archdischild-2021-europaediatrics.358

Background Catheter associated urinary tract infection (CAUTI) is a common device-acquired infection and represent a potentially harmful reservoir of resistant uropathogens. Guidelines recommend limitation of catheter use, aseptic catheter insertion, sterile equipment, strict hand hygiene, use of smallest catheter possible and maintenance of a closed drainage system.

Klebsiella Pneumoniae is a non-mobile aerobic rod causing a large spectrum of hospital-acquired infections, especially pneumonia or urinary tract infections (UTI), developing intrinsic resistance genes. Treating multi drug resistant(MDR) gram negative pathogens becomes a challenge for the caregiver.

Vesicoureteral reflux(VUR) consists of backflow of urine from the bladder into the ureters. It can be primary or secondary due to abnormal lower urinary tract function and elevated intravesical pressure. Post void residual(PVR) is a hallmark of detrusor underactivity(DUA) in children.

Case Presentation Summary We present the case a three years old boy, hospitalized for viral encephalitis, undergoing artificial respiratory support and urine catheterization for 6 weeks. Neurological status was hypotonic cerebral palsy and secondary urine incontinence in a previously toilet trained child.

First febrile UTI developed two days after removing urine catheter. High resistant Klebsiella pn. (ESBL+, AAC(3)-II) was treated with a ten-day course of Cephtriaxone and Amikacin. Clinical response to treatment was good with sterile urine culture after 96 hours. After treatment, the child had asymptomatic bacteriuria with MDR Klebsiella pn. in spite of rigorous local hygiene, proper hydration and oral Fosfomycin. Second febrile UTI was accompanied by febrile seizures, and urine culture was positive with MDR Klebsiella (+ESBL or +HL AmpC, Carbapenem impermeability). Treatment with high dose Meropenem (40mg/kg/dose) for twelve days was successful. Third febrile UTI occurred five days after finishing treatment with same MDR Klebsiella strain. Once again, fourteen-day high dose Meropenem course was successful. Fourth

febrile UTI with *Escherichia Coli* occurred two weeks later and was treated successfully with ten-day course of Ciprofloxacin.

Ultrasound examination revealed normal kidneys and bladder, while voiding cystography was evocative for bilateral VUR and post void residual.

Neurologic status improved over a three-month period from almost complete hypotonic palsy to mild hypotonic paraplegia.

Discussions Long hospitalization and prolonged urine catheterization increase the risk of CAUTI. High dose Meropenem remains efficient in treating MDR *Klebsiella* strains. Recurrent UTI should be further investigated for presence of VUR.

Neurological damage, like hypotonic cerebral palsy can be associated with underactive bladder and detrusor underactivity.

359 A NOVEL COL4A4 MUTATION IN THE PROBAND INITIALLY DIAGNOSED AS IGAN WITH AUTOSOMAL RECESSIVE ALPORT SYNDROME

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10.1136/archdischild-2021-europaediatrics.359

Immunoglobulin A nephropathy (IgAN) is recognized as the most common form of glomerulonephritis all over the world. The diagnosis of IgAN is generally made according to clinical findings and histologic lesions on renal biopsy. Alport Syndrome is an inherited rare glomerular disorder characterized by hematuria, proteinuria, sensorineural hearing loss and progressive kidney disease. We here present a patient initially diagnosed as IgAN with biopsy findings and then diagnosed as Alport Syndrome by genetic screening.

An eight-year-old boy presented with a history of macroscopic hematuria attacks preceded by upper respiratory tract infections. Renal biopsy was compatible with IgAN showing positive staining of IgA in the mesangium and the basement membrane in immunofluorescence. Furthermore, the thinning and thickened of the glomerular basement membrane and splitting of lamina densa was found in electron microscopic examination. During his follow-up, persistent microscopic hematuria and proteinuria were noticed.

Sensorineural hearing loss was developed at ten years of age. Also the proband had a family history of hematuria with proteinuria. Despite the positive mesangial IgA staining in immunofluorescence, atypical renal phenotypes for IgAN including persistent hematuria and hearing loss, and positive family history and electron microscopic findings of renal biopsy aroused suspicion of Alport Syndrome. Genetic analysis (whole exon sequencing) demonstrated a homozygous mutation in COL4A4 (chr 2:g.227922261 c.2438delG (p. Gly813AspfsTer56)) in the proband. Heterozygous mutation was identified by Sanger sequencing of gene COL4A4 in the carrier of all member of his family.

Due to this very rare coincidence, we emphasize that atypical clinical findings should warn the clinicians for other possible diagnosis. The diagnosis of this case also highlights the importance of genetic test in diagnosis of inherited kidney disease. Genetic screening has been recommended as the gold standard for the proper clinical diagnosis and understanding the mode of inheritance when Alport syndrome has suspected in a patient.

360 UNILATERAL RENAL AGENESIA : A 28-YEAR SINGLE CENTER EXPERIENCE

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10.1136/archdischild-2021-europaediatrics.360

Unilateral renal agenesis (URA) is defined as the one-sided congenital absence of renal tissue resulting from failure of embryonic kidney formation [1] URA is often associated with congenital anomalies of the kidney and urinary tract and extra-renal anomalies, such as cardiac, genital or gastrointestinal malformations. The impaired clinical outcome of those children may be explained by the hyperfiltration hypothesis which implies that children with a solitary functioning kidney are at risk to develop hypertension, proteinuria, and chronic kidney disease. [2] Aim of this paper was to estimate the incidence of URA and associated anomalies in our population.

We reviewed 44 consecutive cases of unilateral renal agenesis diagnosed at our hospital in this 28 years (1991-2019) retrospective study During that period 28776 children were evaluated by 56921 renal ultrasonography (RUS). Patient age at diagnosis ranged from newborn to 18 years. There were 23 (52%) boys and 21(48%) girls. Our patients were evaluated for urinary tract infection or abdominal pain and during examination for congenital malformations or pathological fetal US. The left kidney was absent in 25(56%) patients and the right kidney was absent in the remaining 19 (44%). Associated genital anomalies were present in 5 (23%) of 21 girls with URA including agenesis et hypoplasia uteri, agenesis et polycystic ovarii, vaginal duplicity. One boy (4%) had contralateral hydronephrosis with bladder diverticulum. Ectopic kidney was diagnosed in 15 patients.

In our population the frequency of URA was 1 per 1330 births. The most frequent non urinary malformations were genital anomalies among girls.

Careful screening should be proposed throughout childhood to detect early signs of glomerular hyperfiltration and prevent its progression to more serious complications. Ultrasound has been effective for early detection of renal and urinary tract anomalies.

361 PRETREATMENT MORNING URINE OSMOLALITY PREDICTS ORAL DESMOPRESSIN LYOPHILISATE TREATMENT OUTCOME IN PATIENTS WITH PRIMARY MONOSYMPTOMATIC ENURESIS

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10.1136/archdischild-2021-europaediatrics.361

To determine the association between urine osmolality in patients with primary monosymptomatic enuresis (PMNE) and their response to desmopressin.

We hypothesized that pretreatment morning urine osmolality is lower in PMNE patients with complete response to desmopressin treatment compared to the cases with partial or no response.

This was a prospective cohort study that included 419 patients with enuresis seen in our outpatient clinic between October 2017 and October 2019. Patient workup included symptom checklist, bladder diary, kidney and bladder