tolerate therapy and the difficulties regarding care of vascular-access ports in a younger cohort.¹ Given the lack of data surrounding PLEX in Paediatrics, protocols are developed based on extrapolations from best-practice in adult populations, an area where work must be carried out to improve patient safety and outcome. We present a large cohort of paediatric patients undergoing therapeutic plasma exchange therapy for solely renal indications, across a period of 17 years.

A retrospective chart review was conducted for all patients (under 16 years) undergoing PLEX therapy for a renal indication, as specified by the ASFA Guidelines, between January 2002 & June 2019.² The following data were extracted, for each individual case: Age; gender; indication; complications; pre-medications; therapeutic outcome. Patients were stratified into groups as follows: STEC HUS; aHUS; Nephritis (C3GN, PIGN, anti-GBM, ANCA Vasculitis); IgA Nephropathy/HSP; Post-Transplant (recurrent FSGS, acute humeral rejection). This review was performed with permission from the CHI at Temple Street Research & Ethics Committee.

A total of n=58 patients were identified, 39.7% were male (n=23) and 60.3% were female (n=35). 1137 exchanges were performed. The median age of patients undergoing PLEX was 35.5 months. The most common indication was STEC HUS (n=29). Fluid substitution was performed using 5% Albumin-Saline or Plasma. Complications occurred in n=38 patients, with most experiencing minor complications. Asymptomatic hypocalcaemia was the most common complication experienced (n=25). There were no deaths as a result of PLEX therapy.

We present our experience of plasma exchange (PLEX) therapy, spanning 1,137 exchanges across 17 years, proved a well-tolerated, highly efficacious therapy for a variety of renal pathologies, as listed above. Most complications experienced were minor in nature, and with therapy conducted in specialised centres, with appropriately paediatric-trained staff, there are very low levels of adverse events – most of which can be anticipated.

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NEUROGENIC BLADDER DYSFUNCTION – EXPERIENCES OF THE DEPARTMENT OF PEDIATRICS IN UNIVERSITY HOSPITAL CENTRE SPLIT

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Neurogenic bladder is a bladder dysfunction caused by impaired innervation of the lower urinary tract. The most common causes of neurogenic bladder in children are congenital anomalies of the spinal cord. Acquired causes of neurogenic bladder in children can be: cerebral palsy, transverse myelitis, spinal cord tumors, trauma, progressive neurodegenerative diseases, multiple sclerosis, Guillain-Barre syndrome and iatrogenic damage to the pelvic plexus.

Urodynamic testing is the gold standard in the diagnosis and monitoring of children with neurogenic bladder.

The most common form of treatment is clean intermittent catheterization. In terms of drugs, anticholinergics are most commonly used. The main goals of treatment are to preserve renal function and achieve continence.

Children with neurogenic bladder are monitored in the Laboratory for Urodynamics and in the Voiding Disorders Clinic, Department of Pediatrics in University Hospital Centre Split. We retrospectively analyzed data from the medical records of children with neurogenic bladder in the period from Year 2015 to 2019.

Over a period of five years, neurogenic bladder was diagnosed in total of 20 girls and 16 boys. The mean age of diagnosis of neurogenic bladder was 8.3 years. The mean age of children with congenital anomalies of the spine and spinal cord was 4.9 years, while the mean age of children with acquired patterns of neurogenic bladder dysfunction was 11.0 years. Urodynamic examination was done in all children. The diagnosis of neurogenic overactive detrusor and detrusor sphincter dyssynergia was made in a total of 13 children with congenital anomalies and in 9 children with acquired anomalies. Thus, 61% of children had the most severe form of bladder dysfunction. A total of fifteen children were treated with clean intermittent catheterization.

The anticholinergic was administered to ten children with neurogenic overactive detrusor and congenital anomalies and to eight children with acquired neurogenic bladder causes.

In most children with neurogenic bladder, early urodynamic diagnosis and early initiation of treatment may preserve renal function.

Further monitoring of our patients is needed.

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THE VALUE OF CONTRAST-ENHANCED VOIDING UROSONOGRAPHY (CEVUS) IN THE DIAGNOSIS OF INTRARENAL REFLUX (IRR) IN CHILDREN WITH LOW-GRADE VESICOURETERAL REFLUX (VUR) – A CASE REPORT

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Objective Many studies have shown that intrarenal reflux (IRR) is an important risk factor for renal scarring and reflux nephropathy in children with vesicoureteral reflux (VUR). The incidence of IRR diagnosed by fluoroscopic voiding cystourethrography (VCUG) ranges below 1% to a maximum of 10% and is detected only in children with higher grades of VUR.

In our institution we have been using ultrasound methods for the diagnosis of VUR since 2006. Contrast-enhanced voiding urosonography (ceVUS) combined with harmonic imaging and second-generation ultrasound contrast media, which we introduced in 2013, has high diagnostic accuracy compared to VCUG in the detection of VUR. This method enabled IRR detection in almost 12% of our patients with VUR.

The diagnostic criterion for IRR using ceVUS is the appearance of contrast microbubbles outside the contours of the duct system or renal calyx and the entry of contrast into the renal parenchyma. By March 2021 we have had ten children with VUR gr II and IRR which demonstrates the possibility of ceVUS to detect IRR in children with low-grade VUR as well. We report one of the children with low-grade reflux and IRR.

Case Report A four-month-old girl with *Klebsiella pneumoniae* febrile urinary tract infection (UTI) was diagnosed by ceVUS with VUR grade II on the right kidney. The appearance of contrast microbubbles outside the contours of the channel system and the entry of contrast into the renal parenchyma showed that the girl had IRR in the lower pole of the right kidney. A 99m Tc 2,3-dimercaptosuccinic acid (DMSA) renal scan, which was performed 8 months after the UTI, revealed a small scar on the lower pole of the right kidney, where IRR was found by ceVUS.

Conclusion Our case report showed that not only the grade of reflux but also the presence of IRR is important to predict the severity of the clinical picture and the development of possible scarring. It is important to find IRR because it may cause renal scars. VUR with IRR should be managed actively to decrease the chances of renal scarring or the development of new scars.

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A CASE REPORT OF AN INFANT WITH IDIOPATHIC HYPERCALCIURIA AND NEPHROLITHIASIS ASSOCIATED WITH CYP24A1 ENZYME POLYMORPHISM

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CYP24A1 is an enzyme that inactivates vitamin D and encodes vitamin D 24-hydroxylase. Mutations in this enzyme have been linked with idiopathic infantile hypercalcemia, nephrolithiasis, and nephrocalcinosis. The genetic testing for this mutation should be considered in the presence of calciuria, elevated serum calcium, elevated 1,25-dihydroxyvitamin D, and suppressed parathyroid hormone.

We present a previously healthy eight-month-old male infant with macroscopic hematuria. He was born full-term with no perinatal risks. Infant was breastfed for up to four months, and afterwards fed with milk formula, some mixed fruits and vegetables, supplemented with vitamin D according to the recommendations. The family history was negative for nephrolithiasis and urinary tract abnormalities. On the admission, he was in good general condition, afebrile, with normal vital parameters. His body weight was 8.05 kg (15th centile), length 76 cm (94th centile), and head circumference 46.1 cm (77th centile), without any deviations in his clinical examination. In laboratory findings, there were 90% dysmorphic erythrocytes in urine and elevated calcium/creatinine ratio (1.5 mmol/mmol, 2.1 mmol/mmol). 24-hour urine sample showed hypercalciuria (6 mg/kg/24h) and albuminuria (54 mg/24h). The values of alpha-1-microglobulin, parathyroid hormone, vitamin D, serum electrolytes, antinuclear antibody, anti-neutrophil cytoplasmic antibodies, amino acids, glycols, oxalates and citrates in urine and coagulation tests were normal. Immunoglobulin blood test, C3, and C4 levels were normal. The urinary tract ultrasound revealed kidney stone of 6 mm in diameter in the middle cup of the left kidney. Genetic testing excluded suspected Dent's disease but confirmed heterozygous missense variant CYP24A1 c.469C>T, p.(Arg157Trp) classified as a polymorphism. He was treated with hydrochlorothiazide (1 mg/kg) with the recommendation of increased fluid intake with higher citrate content and a low-salt diet. Given the findings of genetic testing, we omitted vitamin D

supplementation. Initially we had a good therapy response, but considering relapse of hypercalciuria after lowering the dose of hydrochlorothiazide (0.5 mg/kg) and known risk of non-melanoma skin cancer in patients on hydrochlorothiazide, the therapy was changed to potassium citrate (2 mmol potassium ion/kg/day). During the follow-up, there was no relapse of macrohematuria, the infant was in good general condition with all tests within reference values. The ultrasound of the urinary tract remained unchanged.

Children presenting with hypercalcemia, hypercalciuria and nephrolithiasis should be tested because of the importance of recognition, genetic diagnosis and proper treatment of CYP24A1 mutations that can present with a wide range of phenotypic presentations, from asymptomatic to chronic renal disease.

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HYPERTENSIVE CRISIS IN A 16-YEAR OLD GIRL WITH ACCESSORY RENAL ARTERY: A CASE REPORT

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Introduction The most common pattern of kidney vascularisation is a single renal artery originating from the abdominal aorta. However, in 20-30% of general population an accessory renal artery can be found being more frequently present (up to 80%) in patients with essential hypertension.

A possible pathomechanism of hypertension is the impaired renal perfusion since the diameter of a single renal artery is usually larger than when multiple arteries are present. Whether an accessory renal artery could be a cause of hypertension is still controversial.

Case Presentation We describe a case of a 16-year old girl who presented with hypertensive crisis. She was previously healthy with no record of abnormal blood pressure (BP).

At admission she reported nausea and severe headache while her BP was 220/120 mmHg. Her body mass index was normal and physical examination unremarkable.

Initial workup showed normal renal function with normal serum electrolytes and plasma glucose. Urin dipstick and urin toxicology screen were also normal. She had hypercholesterolemia and mild proteinuria but no signs of other target organ damage (electrocardiogram, echocardiography, fundoscopy and computed tomography of the brain were normal). Ambulatory blood pressure monitoring confirmed severe ambulatory hypertension.

Further evaluation was aimed at determining the possible cause of secondary hypertension. Urine metanephrines, urinary free cortisol, plasma cortisol, ACTH and thyroid function tests were within reference ranges. High normal plasma renin with elevated plasma aldosterone led to a suspicion of renovascular hypertension.

Magnetic resonance imaging revealed no pathology of the adrenal glands. Although renal ultrasonography with Doppler was normal, magnetic resonance angiography and later CT angiography showed two nonstenotic right renal arteries.

Conclusion Although nonstenotic, an accessory renal artery should be considered as a possible cause of renovascular hypertension in children and adolescents.