VII CONGRESSO HISPANO-PORTUGUÊS DE NEFROLOGIA PEDIÁTRICA E XLVI CONGRESO ESPAÑOL DE NEFROLOGIA PEDIÁTRICA

LIVRO DE RESUMOS
ABSTRACT BOOK

DATA | DATE

18 E 19 DE MAIO DE 2023
18TH - 19TH MAY, 2023

LOCAL | PLACE

NOVA MEDICAL SCHOOL | FACULDADE DE CIÊNCIAS MÉDICAS
CAMPO DOS MÁRTIRES DA PÁTRIA 130, 1169-056 LISBOA

COM O APOIO DO TURISMO DE LISBOA SPONSORED BY LISBON TURISM

THURSDAY, MAY 18TH, 2023

08:30-09:00	**Registration**	
09:00-09:30	**Welcome session**	
	ROOM 1	**ROOM 2**
09:30-11:00	**ROUND TABLE: KIDNEY TRANSPLANTATION** ***Moderators:*** *Leire Madariaga Dominguez, Conceição Mota* • **Immunosuppression in kidney transplantation - new drugs** *Evgenia Preka, Evelina London Children's Hospital* • **Risk of neoplasm after transplantation** *Alejandro Zarauza, Hospital Universitário La Paz* • **Strategies to prevent chronic kidney graft dysfunction** *Filipa Durão, Centro Hospitalar Universitário de Lisboa Norte*	**ROUND TABLE: CAKUT** ***Moderators:*** *Ines Vergara Perez, Teresa Costa* • **Posterior urethral valves - markers of progression to chronic kidney disease** *Rute Baeta Batista, Centro Hospitalar Universitário de Lisboa Central* • **Vesicourethral reflux and urinary tract infection – antibiotic prophylaxis** *Juan David Gonzalez, Hospital General Universitario Santa Lucía de Cartagena* • **Preparation for transplantation** *Agustin Serrano, Hospital Universitari i Politècnic la Fe*
11:00-11:30	**COFFEE BREAK**	
11:30-12:00	**Industry-sponsored symposium**	
12:00-12:45	**Long communications** (4) ***Moderators:*** *Gema Ariceta Iraola, Raquel Santos*	**Long communications** (4) ***Moderators:*** *Ainhoa Iceta Lizarraga, Liliana Rocha*
12:45-13:10	**Poster presentation** (5) ***Moderators:*** *Rebeca Garrote Molpeceres, Filipa Durão*	**Poster presentation** (5) ***Moderators:*** *Carmen de Lucas Collantes, Liane Costa*
13:10-14:40	**LUNCH**	
14:40-15:10	**Industry-sponsored symposium**	
15:10-16:00	**Plenary Conference – IgA glomerulonephritis** - *Marina Vivarelli* ***Moderators:*** *Montserrat Anton Gamero, Margarida Abranche*	
16:00-16:30	**Young pediatric nephrologists** - *Cristina Blázquez, Patrícia Costa Reis,* ***Moderadores:*** *Pedro Arango Sancho, Telma Francisco*	
16:30-17:25	**Short communications** (6) ***Moderators:*** *Joaquin Escribano Subias, Liane Costa*	**Short communications** (6) ***Moderators:*** *Susana Ferrando, Graça Ferreira*
17:25 - 17:40	**COFFEE BREAK**	
17:40-18:30	**Registries**	**Registries**
20:30	**CONGRESS DINNER**	

FRIDAY, MAY 19TH 2023

	ROOM 1	ROOM 2
08:30-09:05	**Short communications** (4) ***Moderators:*** *Elvira Izquierdo Garcia, Liliana Rocha*	**Short communications** (4) ***Moderators:*** *Maria Angeles Fernandez Maseda, Célia Madalena*
09:05-09:30	**Poster presentation** (4) ***Moderators:*** *Marta Carrasco Hidalgo-Barquero, Paula Nunes*	**Poster presentation** (4) ***Moderators:*** *Esther Trillo Bris, Ana Zagalo*
09:30-11:00	**ROUND TABLE:** **INHERITED DISORDERS** ***Moderators:*** *Benito Amil, Caldas Afonso* • **Autosomal dominant polycystic kidney disease** *Ana Teixeira, Centro Materno Infantil do Norte* • **Alport's disease** *Joana Jardim, Centro Hospitalar Universitário do São João* • **Fabry disease** *Guillem Pintos-Morell Hospital Vall d'Hebron*	**ROUND TABLE: DIALYSIS** ***Moderators:*** *Javier Martín Benlloch, Maria do Sameiro Faria* • **Nutrition in dialysis** *Telma Francisco, Centro Hospitalar Universitário de Lisboa Central* • **Peritonitis in peritoneal dialysis** *Carolina Cordinhã, Centro Hospitalar Universitário de Coimbra* • **Current concepts in hemodialysis and venous access** *Alejandro Cruz, Hospital Vall d'Hebron*
11:00-11:30	**COFFEE BREAK**	
11:30-12:00	**Industry-sponsored symposium**	
12:00-12:45	**Long communications** (4) ***Moderators:*** *Teresa Alarcon Alacio, Ana Rita Sandes*	**Long communications** (4) ***Moderators:*** *José Eugénio Cabrera, Helena Pinto*
12:45-13:20	**Poster presentation** (5) ***Moderators:*** *Maria Isabel Luis Yanes, Madalena Almeida Borges*	**Poster presentation** (5) ***Moderators:*** *Fatima Fraga Bilbao, Teresa Costa*
13:20-15:00	**LUNCH – Industry-sponsored symposium**	
15:00-15:45	**Plenary Conference – Lupus nephritis** - *Patrícia Costa Reis* ***Moderators:*** *Álvaro Madrid Aris, Gisela Neto*	
15:45-16:40	**Long communications** (5) ***Moderators:*** *Pedro Ortega López, Carmen do Carmo*	
16:40-17:00	**Closure and best communications and posters awards**	
17:00 - 17:30	**COFFEE BREAK**	
17:30-19:00	**AENP PLENARY ASSEMBLY**	

LONG COMMUNICATION - COL 01

PEDIATRIC KIDNEY TRANSPLANT FROM DONATION AFTER CIRCULATORY DEATH: CLINICAL ASPECTS AND SHORT-TERM OUTCOMES

Alejandro Zarauza Santoveña1; Carlota Fernández Camblor1; Belén Estébanez Montiel2; Susana Rivas Vila3; Marta Melgosa Hijosa1; Ángel Alonso Melgar1; María José Martínez Urrutia3; Laura Espinosa Román1

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Introduction:
Donation after circulatory death (DCD) has become a valuable source of kidney grafts in the last decades. There are ethical, logistical and clinical aspects that differ from donation after brain death (DBD). Warm ischemia time determines a higher risk of delayed graft function, although medium and long-term outcomes in adult series are comparable to DBD.
In recent years, pediatric DCD is developing significantly. In our center, CDC accounts for up to 22% of all cadaveric donors in the last 3 years.

Objectives:
Describe characteristics and evolution of recipients receiving kidneys from DCD, and compare outcomes with DBD grafts.

Methods:
Retrospective analysis of medical records

Results:
Since January 2020 to December 2022 we transplanted 10 pediatric recipients with kidneys from DCD. All were type III donors and first transplants. 1 liver-kidney combined transplant. 8 male / 2 female. Age of recipients 11.2 ±4 y. Age of donors 19.7 ±9.6 (7/10 donors <18y). Time on waiting list 6.1±4.8 months.
Warm ischemia time 15.7±7.7 minutes. Total cold ischemia 12.6 ± 4.4 hours. No significant surgical complications. Standard immunosuppression with basiliximab, steroids, tacrolimus and micophenolate in all patients. 1 patient received also rituximab and plasma exchanges due to primary renal disease (FSGS). We compared the results with 35 recipients of DBD in the same period. Ages of donor and recipient, as well as total cold ischaemia time were comparable between the two groups. No graft loss and no delayed graft function in CDC group.
We found no differences in serum creatinine (0.71 ±0.27 mg/dL vs 0.69±0.34 mg/dL) and eGFR (91.83±25.2 mL/min/1.73m2 vs 91.78 ±29.8 mL/min/1.73m2) 1 month after transplantation.

Conclusion:
DCD is now an important source of kidney grafts. Immediate and short-term postoperative outcomes are good, and comparable to those of DBD. We need longer follow-up to verify if this good evolution is sustained in the long term.

Key words : Kidney transplantation, Donation after circulatory death

LONG COMMUNICATION - COL 02

ASSOCIATION BETWEEN TACROLIMUS VARIABILITY AND DEVELOPMENT OF DONOR-SPECIFIC ANTIBODIES IN PEDIATRIC KIDNEY TRANSPLANTATION

Jose Angel Caballero Gil2; Silvia Ojea Varona3; Julia Fijo López-Viota1; Rafael Bedoya Pérez1; Ana Sánchez Moreno1; Francisco De La Cerda Ojeda1

1 - Servicio de Nefrología Pediátrica Hospital Universitario Virgen del Rocío; 2 - Gerencia Atención Primaria A Coruña; 3 - Complejo Hospitalario Universitario de Ferrol

Introduction:
Donor-specific antibodies (DSA) is a well described predictor of antibody-mediated rejection and an important cause of graft loss in kidney transplantation. In adult population it has been demonstrated that high intrapatient tacrolimus variability is associated with the development of de novo DSA. There are very scarce publications regarding this condition in pediatric kidney transplant recipients.

Objectives:
To analyze intrapatient tacrolimus variability and its possible relationship with the development of de novo DSA in a cohort of pediatric kidney transplant recipients.

Methods:
A single-center retrospective study included pediatric kidney transplant recipients (<18 years), transplanted between 2015 and 2020. Inclusion criteria: patients on tacrolimus treatment, with more than 2 years follow-up and ≥ 3 measured tacrolimus trough levels. Patients with pre-transplant DSA were excluded. Intrapatient tacrolimus variability was defined using the coefficient of variation for all trough levels obtained after 3 months post-transplant.

Results:
From the 99 patients transplanted in our institution during the studied period, 61 patients were included in our final analyses. The median age at transplant was 11 (6,46-15,54) years. 14.75% of the patients developed de novo DSA. There was no significant difference in age and gender between those who developed DSA and those who did not. There was a significant association between de novo DSA development and thymoglobulin induction. A statistically significant association ($p<0.05$) was demonstrated between the increase in the variability of tacrolimus levels and the development of de novo DSA in the logistic regression model.

Conclusion:
In the pediatric renal transplanted population studied, an association between variability in tacrolimus trough levels and the development of DSA was demonstrated. These results can help to early identify the population at risk of developing de novo DSA, in order to modify the dose of immunosuppression preemptively.

Key words: Coefficient of variation (CV), Donor-specific antibody (DSA), Kidney, Pediatrics, Antibody, Immunosuppression

LONG COMMUNICATION - COL 03

ADULT LIFE PERCEPTION AFTER PEDIATRIC KIDNEY TRANSPLANTATION

Julia Fijo López-Viota1; Ana Sánchez Moreno1; Rafael Bedoya Pérez1; David Canalejo González1; Ángela García Rojas1; Francisco De La Cerda Ojeda1

1 - Pediatric Nephrology Unit. Virgen del Rocio Hospital. Sevilla. Spain

Introduction:
It is widely accepted that kidney transplantation (KT) is the best renal replacement therapy for children with end of stage renal disease (ESKD). However scarce data is published regarding feelings, social development and perceptions of adult patients who have undergone a pediatric KT.

Objectives:
Analysing social, educational and subjective outcomes of adult patients who previously underwent a pediatric KT

Methods:
A self report study in adult population (> 18 years old) who received a pediatric KT in our institution between 1979 and 2019, with > 1 year follow-up. Patients were contacted by e-mail or by phone and were invited to fulfill a self report survey about education/training, social and familial status, physical and emotional development, healthy lifestyle and free topic comments.

Results:
Of the 287 pediatric patients who received 345 KT in our institution, 17 were lost to follow-up, 45 died and 75 patients were < 18 years old. From the 150 remaining patients asked to fulfill the survey, 82 subjects (54,7%) accepted.
Only 39% cases became independent, most of them (37,8%) living together. Ten patients (12.2%) had children.
Regarding education, 28% completed upper secondary/post-secondary non tertiary education and 16 % tertiary education level. An early education dropout was referred by 42% cases. Fifty three subjects (64,6 %) were unemployed.
Only 26% cases reported a good quality of life (QOL), similar to rest of healthy population. Nearly one third (30%) referred serious comorbidities (defined as conditions affecting an extrarenal organ). Only 9% patients practiced regular sports. Five patients (6%) admitted drug addiction. Concerning to self corporal image, 13 (15.9%) and 4 patients (4.8%) were deeply troubled for their short stature and obesity, respectively.

Conclusion
Despite pediatric KT is the best treatment for children with ESKD, there are still many limitations and concerns that affect their development and QOL in the adult life.

Key words: Pediatric kidney transplantation, Quality of life, Adult Life

LONG COMMUNICATION - COL 04

EFFECT ON BLOOD PRESSURE AND CARDIOVASCULAR RISK (CVR) FACTORS IN PEDIATRIC PATIENTS DURING COVID-19 PANDEMIC CONFINEMENT (COBECOR STUDY)

Elena Codina Sampera1; Pedro Arango Sancho1; Ana Cristina Aguilar Rodríguez1; Bernat Gómez Herrera1; Marta Jiménez Moreno1; Yolanda Calzada Baños1; Raquel Jiménez García1; Verónica Coll Brito2; Osmar David Aguilar Rodríguez3; Álvaro Madrid Aris1

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Objectives:
Patients monitored in the CVR consultation usually present overweight/obesity,metabolic syndrome,hypertension(HT),unhealthy lifestyle with great resistance to change,influence of the environment and,frequently,psychosocial limitations.We asked whether home confinement for COVID-19(March-May 2020)could have negative effects in relation to CVR in these patients.

Methods:
Assess this hypothesis with respect to HT and the use of antihypertensive drugs in these patients.Secondary objective: assessed changes in other parameters(body mass index (BMI),level of physical activity and diet)

Results:
Retrospective cohort study with review of 738 ambulatory blood pressure monitoring(ABPM)between 2019-2022 obtaining,after applying the exclusion criteria (no overweight/obesity,poor therapeutic compliance,underlying renal pathology or failure to perform two ABPM in the study period),a final cohort of 46 patients divided into two groups (23 each):one group exposed to home confinement(G1)with one ABPM before and after home confinement and another group not exposed to confinement (G2)with two ABPM in different periods between 2021-2022.Blood pressure (BP)percentile values,dipper pattern,variability and blood pressure load,as well as the rest of the CVR parameters were compared in both periods.The mean age was 13 years(13.8 G1/13.2 G2)with a mean time between the 1st and 2nd ABPM of 11 months(11.08 G1/10.95 G2)and a greater reduction in BMI in G2 than in G1(1.05 G1/1.21 G2).Despite this,results were not statistically significant nor were the differences in HBP or worsening of the dipper pattern(30.4% in G1 and 21.7% in G2).We did observe differences($p<0.022$) in the use of antihypertensive drugs,although contrary to our initial hypothesis,with greater use of drugs in G2

Conclusion:
Although the low sample size,the biases inherent in the design and the lack of previous studies make the interpretability and statistical significance of some results difficult,they reinforce that the measures during confinement did not contemplate all spheres of health and the need to implement specific CVR consultations.Obesity and its associated pathologies are an important public health problem that pediatricians have the responsibility to address

Key words: Obesity, COVID-19, Ambulatory Blood Pressure Monitoring (ABPM), Environmental Pediatrics

LONG COMMUNICATION - COL 05

KIDNEY OUTCOMES IN CHILDREN WITH POSTERIOR URETHRAL VALVES

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Introduction:
Posterior urethral valves (PUV) affect approximately 1:5000 live male births, with about 50% progressing to end-stage renal disease within ten years.

Objectives:
We aimed to identify predictors of kidney outcomes for children diagnosed with PUV.

Methods:
This retrospective single-center cohort study included children who underwent a PUV ablation between 1st January 2015 and 30th June 2022. Patients with less than six months of follow-up were excluded. The primary outcome was a composite of glomerular filtration rate (GFR) below -2 SD of expected for age or renal replacement therapy therapy (RRT).

Results:
From a total of 41 patients in the cohort, 22 (54%) were diagnosed prenatally and 12 (29%) were referred to us from Portuguese-speaking African countries. Patients with a prenatal diagnosis were more frequently born preterm (54.5% versus 11.8%, p-value=0.006), were younger at first urethral catheter placement (7.8±15.1 days versus 1.1±2.5 years, p-value=0.07), and had higher baseline serum creatinine (1.5±1.5 versus 0.6±0.2 mg/dL, p-value=0.036) than patients diagnosed postnatally. There were no other significant differences in between-groups comparisons at baseline. Mean follow-up time was 3.1±1.4 years. At last follow-up, median age was 2.6 years [interquartile range (IQR) 0.9-3.9] and median GFR was 95.4 mL/min/1.73m2 [IQR 79.2-115.1]. Thirteen patients (31.7%) met the primary outcome. Three patients (7.3%) needed peritoneal dialysis and another 13 (31.7%) had a low GFR for age. In the logistic regression analyses adjusted for age at last follow-up, prenatal diagnosis (OR 8.1, 95% CI 1.3-50.7, p=0.025), prematurity (OR 2.8, 95% CI 1.3-6.3, p=0.012), and higher serum creatinine at baseline (OR 5.7, 95% CI 1.2-27.5, p=0.030) were significant predictors of the primary outcome.

Conclusion:
Prenatal diagnosis of PUV, prematurity and higher serum creatinine before VUP ablation may predict adverse kidney outcomes (low GFR and RRT). Our sample size and time of follow-up may have limited our conclusions.

Key words: Posterior urethral valves, chronic kidney disease, children

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LONG COMMUNICATION - COL 06

CARDIOVASCULAR AND RENAL MANIFESTATIONS IN A CASE SERIES OF WILLIAMS-BEUREN SYNDROME AT A TERTIARY-LEVEL HOSPITAL

Carmen Larramendi Hernández1,2; Jesús Ferrer Ferrer1,3,4; Héctor Ríos Duro1; Irene Valenzuela Palafoll5; Víctor Pérez Beltrán1; Alejandro Cruz Gual1; Saskia Natali Agámez Luengas1; Mercedes López González1; Gloria María Fraga Rodríguez1,3,6; Gema Ariceta Iraola1,6

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Introduction:
Williams-Beuren syndrome (SWB; OMIM 194050) is an autosomal dominant multisystemic disorder resulting from a 7q11.23 deletion. The SWB phenotype is characterized by distinctive dysmorphic facies, intellectual disability, characteristic personality and a cognitive profile, cardiovascular disease, and endocrine abnormalities, among others.

Objectives:
To define the features of our patients, as well as its prevalence, particularly in regards to nephrological and cardiovascular factors.

Methods:
We conducted a descriptive, retrospective study of all pediatric patients (0-18 age years old) genetically diagnosed with SWB at a tertiary-level hospital.

Results:
We present a case series of 23 patients ranging from 2 to 18 years old (with an average age of 11 years). Regarding cardiovascular manifestations, 20 patients had arterial stenosis (87%), with the most frequent being the typical supravalvar aortic stenosis either associated with other stenoses or not. Three patients presented with elevated blood pressure (13%), and eight arterial hypertension (34,7%), of which 75% were grade I and 25% were grade II. Six patients had renovascular hypertension (75%), most of whom required interventional radiology, and one case required additional vascular surgery. In terms of renal involvement, six patients were diagnosed with structural anomalies (CAKUT, 26%) but most patients (82%) had normal renal function (eGFR >90 ml/min/1.73m2, Schwartz 2009). Although enuresis and symptoms of bladder dysfunction have been reported, they were not commonly asked about in our patients. Four patients had hypercalciuria (17%), and two of them had nephrocalcinosis (8%) but hypercalcemia was detected only in one patient (4%).

Conclusion:
In our case series, fewer patients had arterial hypertension compared to the literature (50%), but most of them had renovascular hypertension. Additionally, a larger proportion had CAKUT than previously reported (10%). We should inquire more about bladder dysfunction, a frequent manifestation reported in the literature (60-70%), which can have significant implications for both quality of life and long-term kidney function.

Key words: Williams syndrome, renovascular hypertension, CAKUT, nephrocalcinosis, hypercalciuria, arterial stenosis

LONG COMMUNICATION - COL 07

EXTRAPOLATING URINARY SOLUTE EXCRETION AND URINE VOLUME FROM 12-HOUR OVERNIGHT SAMPLES TO A 24-HOUR PERIOD CAN PRODUCE INACCURATE RESULTS IN RENAL LITHIASIS: SPECIFIC REFERENCE VALUES ARE PROPOSED FOR 12-HOUR OVERNIGHT SAMPLES

María Concepción Mir Perelló1; María Concepción Rodrigo Jiménez1; Javier Lumbreras Fernández1; Iratxe Tapia Torrijos2; Adrián Rodríguez Rodríguez3; Cristina Gómez Cobo1; María Concepción Sáez-Torres Barroso4; Guillem Frontera1; Félix Grases5

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Objectives:
Collecting 24-hour urine samples can be complex for many patients. Shorter collections can be an alternative method. Extrapolation to 24-hour period for comparison with reference values is a common practice but its interpretation can be affected by circadian variability in urine composition.

Methods:
We aim to compare extrapolated urine composition from 12-hour overnight samples to real one obtained in 24-hour samples from healthy subjects and stone-formers.

Results:
Twenty-six pediatric stone-formers aged 5-17 years old were recruited from the outpatient clinic in a tertiary care center. Eighty-seven healthy individuals of the same age were recruited from local schools. Spontaneous ambulatory urine samples were obtained, fractioned in two consecutive 12-hour periods (daytime and overnight). Calcium, phosphate, magnesium, uric acid, oxalate, citrate, creatinine, pH and urine volume were measured for each period. Median excretions in 24 hours extrapolated from 12-hour overnight sample were compared with real 24-hour excretion for both groups of subjects. Urinary solute excretion and urine volume in the overnight period are described in healthy subjects.
In healthy subjects, phosphate and magnesium excretions were overestimated by an 11 and 15%, respectively, and calcium, uric acid, oxalate, citrate excretions and urine volume were underestimated by a 3, 23, 9.5, 25 and 18%. In stone-formers, calcium, phosphate and magnesium excretions were overestimated by a 4, 20 and 13%, and uric acid, oxalate, citrate excretions and urine volume were underestimated by a 14, 13, 22 and 1%. Upper normal limit (p95) in 12-hour overnight period for promotors were: calcium 2.2 mg/kg, phosphate 13.2 mg/kg, uric acid 424 mg/1.73 m2, oxalate 20.5 mg/1.73 m2; lower normal limit (p5) for inhibitors were: citrate 1.6 mg/kg, magnesium 0.6 mg/kg.

Conclusion:
Up to a 25% error was observed when extrapolating urinary solute excretion and urine volume from 12-hour samples to 24 hours. Specific reference values for 12-hour overnight samples are proposed.

Key words: Urine composition, Renal lithiasis, 12-hour sample, 24-hour sample

LONG COMMUNICATION - COL 08

A CONCEPTUAL REVIEW ABOUT THE DEFINITION OF RENAL PHOSPHATE LOSS

Juan David González Rodríguez1; María Isabel Luis Yanes2; José Eugenio Cabrera Sevilla1; María Cecilia Martín Fernández De Basoa3; Víctor Manuel García Nieto2

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Objectives:
Background Fractional tubular reabsorption of phosphate (TRP) has been used for over 60 years to establish the existence of renal phosphate loss. It is a parameter of corrected volume per decilitre of glomerular filtration rate (GFR). Later, a mass parameter per dl GFR called TP/GFR (tubular PO4 reabsorption per dl GFR) was devised which some authors have sought to substitute for TRP.

Methods:
The aim of the present work is to attempt to demonstrate that TRP and TP/GFR are similar parameters and, in certain aspects, TRP is more effective for diagnosis.

Results:
Methods Data were gathered on the metabolism of phosphate corresponding to a group of healthy children without hypophosphatemia (n= 47); a group of patients with idiopathic hypercalciuria (n= 27); and ten patients diagnosed with X-linked hypophosphatemia (XLH). The TRP, the TP/GFR and the percent tubular reabsorption of phosphate were calculated, with the latter being calculated from the blood PO4 levels and the TP/GFR.

Conclusion:
Results All the patients with XLH presented TRP values lower than 95 ml/dl GFR and of TP/GFR equal to or lower than 2.8 mg/dl GFR. In the total sample, a direct correlation was observed between TRP and TP/GFR (r= 0.65; p= 0.01). The TRP and the percent tubular reabsorption of phosphate values were the same in the three groups (r= 1; p= 0.01).
Conclusions TRP and TP/GFR are similar parameters. TRP is more effective than TP/GFR given that in renal hypophosphatemia it is always below 95% and above 95% in reduced phosphatemia and normal kidney proximal tubular function. There is no solid reason for using TP/GFR rather than TRP.

Key words: Phosphate metabolism, TRP, TP/GFR, Hypophosphatemia

LONG COMMUNICATION - COL 09

GENOTYPE-PHENOTYPE CORRELATION IN ALPORT SYNDROME

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1 - Hospital Regional Universitario de Málaga

Objectives:
Alport syndrome (AS) is a hereditary disease caused by mutations in collagen genes, specifically COL4A3 and COL4A4 (chromosome 2), and COL4A5 (chromosome X). It has a wide phenotypic spectrum, ranging from monosymptomatic microhaematuria to end stage renal disease (ESRD), even at early ages. Factors that may contribute to a more severe phenotype are still unclear.

Methods:
To describe epidemiological, genotypical and phenotypical characteristics from a series of patients with AS, and to analyse genotype-phenotype correlation.

Results:
Single-centre observational retrospective study, after reviewing electronical clinical records from patients under 18 years old diagnosed with AS between 2013 and 2022.

Conclusion:
We identified 21 cases; 81% of them were females, with a median age of 8.5 years (SD 4.4) at diagnosis. Family history was present in 12/21. Hearing and eye abnormalities were found in 33% of patients. From a renal perspective, haematuria was present in 90% of patients, and proteinuria in 62%. One patient presented renal cysts. 3/21 patients (14%) had hypertension, with ESRD at presentation. The remaining patients maintained normal renal function (mean age at last clinical review 12.2 years, SD 4.8). Most common inheritance was X-linked (47%), followed by autosomal-dominant (35%) and autosomal-recessive (18%). COL4A5 was the most frequent mutated gene (47%), followed by COL4A3 (35%) and COL4A4 (12%). Out of all patients with ESRD at diagnosis, one presented a mutation in COL4A3, a second one in COL4A5 and a third one, a digenic mutation in COL4A3+COL4A5. We could not find any statistically significant correlation between genotype and phenotype.
In our series, patients with ESRD at presentation were found to have a recessive inheritance (COL4A3) or X-linked (COL4A5). One of them had a digenic inheritance, which could have contributed to a more aggressive course. Unfortunately, a statistically significant correlation was not found, probably due to a small sample size.

Key words: Alport, genotype, phenotype, mutation, end stage renal disease

LONG COMMUNICATION - COL 10

A MULTICENTER STUDY OF PATIENTS WITH NEPHROCALCINOSIS SECONDARY TO MUTATIONS IN THE GENES ENCODING TWO RENAL PROXIMAL TUBULAR PHOSPHATE TRANSPORTERS

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Introduction:
It is known that mutations in the genes encoding two proximal phosphate transporters cause hypophosphatemia, hypercalciuria and nephrocalcinosis. The SLC34A3 gene encodes type IIc Na+/Pi cotransporter; its mutations cause Hypophosphatemic rickets with hypercalciuria. The SLC34A1 gene encodes type IIa Na+/Pi cotransporter; its mutations cause nephrolithiasis/osteoporosis, hypophosphatemic rickets

Objectives:
To determine the clinical, biochemical and genetic characteristics of patients with mutations in the SLC34A3, SLC34A1 and heterozygous genes

Methods:
Patients and methods. Multicenter study in which 17 patients (7V, 10M) with ultrasound nephrocalcinosis studied in nine Spanish hospitals were included. Several biochemical parameters were collected at diagnosis and at the end of the follow-up period

Results:
Nine patients had mutations in SLC34A3, six in SLC34A1 and, two, a heterozygous mutation in SLC34A1. Age at diagnosis was 5.3±3.9 years (range: 0.1-15.5). At diagnosis, all but one patient showed a TRP less than 95 ml/100 ml GFR, hypercalciuria was present in 10/17 (58.8%), hypophosphatemia in 5/17 (29.4%), lithogenic risk (elevated calcium/citrate ratio) in 15/17 (88.2%), concentration capacity defect in 12/15 (70.6%) and GFR less than 90 ml/min/1.73 m2 in 5/16 (29.4%). The treatment received was variable (in general, potassium citrate, thiazides and potassium citrate or phosphate salts). There were no statistically significant differences between phosphatemia levels at baseline and at the end of follow-up. In contrast, in the second period GFR (Schwartz) was lower (105.6± 8.5 vs. 90.9±30.3 ml/min/1.73 m2, p=0.03) and creatinine levels higher (0.47±0.19 vs. 0.66±0.19 mg/dl, p=0.01). At the end of follow-up 9/15 (60%) showed reduced GFR.

Conclusion:
In pediatric patients with nephrocalcinosis, the joint presence of hypercalciuria and hypophosphatemia should be investigated. If positive, mutations in the SLC34A3 and SLC34A1 genes are candidates. It is unknown whether the reduction of GRF is secondary to nephrocalcinosis or to other unknown mechanisms.

Key words: Monogenic Nephrocalcinosis, SLC34A1 gene, SLC34A3 gene, Hypophosphatemic rickets with hypercalciuria

LONG COMMUNICATION - COL 11

MULTICENTER STUDY OF PATIENTS WITH MUTATIONS IN GENES POTENTIALLY CAUSING HYPERCALCEMIA

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Introdução:
In recent years it has been known that mutations in the CYP24A1 gene (1,25-DiHydroxyvitamin D3 24-hydroxylase) cause hypercalcemia. Mutations in two other genes such as CaSR (calcium sensitive receptor) and SLC26A1 (solute carrier family 26 member 1) can also cause hypercalcemia

Objectives:
To determine the clinical, biochemical and genetic characteristics of patients with mutations in CYP24A1, CaSR, SLC26A1 and heterozygous genes

Methods:
Patients and methods. Multicenter study in which 9 patients (5V, 4M) with mutations in genes potentially causing hypercalcemia studied in nine Spanish hospitals were included. Various biochemical parameters were collected at diagnosis and at the end of the follow-up period

Results:
Four patients had mutations in CYP24A1, one in CASR, one in SLC26A1 and three had a heterozygous mutation in CYP24A1. Age at diagnosis was 6.1±5.6 years (range: 0.7-14.2). Two had normal ultrasound (CASR and SLC26A1 mutations) and the rest had ultrasound nephrocalcinosis. At diagnosis, eight patients showed calcemia levels above 10.5 mg/dl and a TRP between 82 and 95 ml/100 ml GFR. There was hypercalciuria in 5/9 (55.6%), hypophosphatemia in 3/9, lithogenic risk (elevated calcium/citrate ratio) in 2/9 (28.6%), concentration capacity defect in 7/9 (77.8%) and GFR less than 90 ml/min/1.73 m2 in 3/9 (33.3%). The treatment received was variable (in general, potassium citrate or thiazides together with potassium citrate). At the end of follow-up, the defect in concentrating ability persisted in seven patients and a reduction in GRF in two of them. In the two of the three cases in which calcitriol levels could be measured were higher than 120 pg/ml

Conclusion:
It is known that mutations in some genes can be associated with hypercalcemia with the consequence of nephrocalcinosis and/or renal lithiasis. The most frequent functional anomaly in these cases is the defect in renal concentrating capacity

Key words: Hypercalcemia, Monogenic Nephrocalcinosis, CYP24A1, SLC26A1, CaSR

LONG COMMUNICATION - COL 12

MOLECULAR FINDINGS IN A GROUP OF PATIENTS WITH PRIMARY HYPOMAGNESEMIA AND SALT LOSING TUBULOPATHY

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Introduction:
Disturbed Na+ reabsorption in the distal convoluted tubule (DCT) is associated with hypomagnesemia and hypokalemic alkalosis. Most of the Mg2+ delivered from the loop of Henle is reabsorbed in the DCT, so this segment plays an important role in maintaining Mg2+ homeostasis.

Objectives:
The aim of our study was to analyse the molecular findings in a group of patients with renal salt-losing tubulopathies associated with hypomagnesemia.

Methods:
37 patients from 34 families with hypokalemia and hypomagnesemia of renal origin. Initial study with NGS panel with 44 genes (including SLC12A3, CLCNKB, SLC12A1, KCNJ1, BSND, KCNJ10, FXYD2, CLCN5, CLDN16, CLDN19, ATP1A1, KCNJ10, CNNM2, HNF1B). Exome sequencing in those cases where no molecular alteration was found. MLPA for the search of gross deletions in the CLCNKB and the SLC12A3 genes.

Results:
The most common diagnosis were Bartter syndrome type 3 (BS3, 16/37 patients) and Gitelman syndrome (GS, 14/37 patients). In addition, 3/37 patient carried a heterozygous pathogenic variant in RRAGD, 1/37 carried a heterozygous pathogenic variant in CLCN5, and 1/37 patient carried a heterozygous pathogenic variant in HNF1B. Finally, in 2/37 patients no molecular alterations were found. Mean serum Mg2+ levels were similar in BS3 and GS patients (1.47 ± 0.21 & 1.46 ± 0.16 mg/dL; P=0.9) but significantly lower in RRAGD patients (1.06 ± 0.25 mg/dL, P<0.05). Mean serum K+ levels were significantly lower in BS3 patients than GS and RRAGD (2.1 ± 0.67 vs. 2.92 ± 0.62 vs. 3 ± 0.34 mEq/L, P<0.05).

Conclusion:
Mutations in different genes are associated with salt losing tubulopathies with hypomagnesemia, mostly SLC12A3 (GS) and CLCNKB (BS3). Potassium levels are lower in BS3 patients, indicating a stronger activation of the renin-angiotensin system. Although not universal, Mg2+ levels can be low in BS3 patients due to the ubiquitous expression of the ClC-Kb channel in the loop of Henle and in the DCT.

Key words: Hypomagnesemia, Salt-losing tubulopathy

LONG COMMUNICATION - COL 13

USE OF CUFFED-TUNNELED CENTRAL VASCULAR CATHETERS AS VASCULAR ACCESS IN PEDIATRIC HAEMODIALYSIS

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Objectives:
An adequate vascular access is essential for the performance of an effective haemodialysis. The use of cuffed-tunnelled central venous catheters (CCVC) as vascular access in paediatrics may be an option when a short duration of haemodialysis is expected.

Methods:
To describe the characteristics of our haemodialysis patients and their vascular accesses in the last 10 years.

Results:
We retrospectively reviewed the medical records of patients undergoing haemodialysis in our unit from 2012-2022.
Results: 105 patients (62% male) with mean age of 8.15 years (23% younger than 3 years) received haemodialysis during this period. The most frequent underlying primary renal disease was congenital kidney and urinary tract anomalies (48%).All of them used CCVC as vascular access.
The median time on haemodialysis was 8.5 months. 73 patients are currently transplanted. Haemodialysis is still being performed in 9% of our patients. Four patients died, none of them due to haemodialysis complications.
181 CCVC were used, 60% of the patients required a single catheter. The preferred vascular access was the right internal jugular vein (90% of cases). The catheters used in 146 cases were made of silicone (Perm-cath), the rest were made of polyurethane (Split-cath).
145 complications were described, affecting 106 of the CCVC. In seventy-two percent of the cases of complications, CCVCs required replacement for this reason. The most frequent complications were mechanical (45%), almost equal to infections (42%), whereas thrombotical events represented only 11% of the total. The complication rate was 1 complication per 10 catheter/month, with a probability of remaining complication-free at 6 and 12 months of 76 and 52% respectively.

Conclusion:
The vascular access preferentially used in our unit are CCVCs. In our series, their use allows adequate dialysis with a low complication rate. The median duration of haemodialysis in our sample is lower than 1 year.

Key words: Haemodialysis, Tunnelled central venous catheters, chronic renal disease, catheter related complications

LONG COMMUNICATION - COL 14

FACTORS INFLUENCING THE OCCURRENCE OF PERITONITIS IN A LEVEL III PORTUGUESE HOSPITAL

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Introduction:
Peritonitis is a frequent complication among children on peritoneal dialysis (PD). It remains as one of the main causes for technique failure, morbidity, and mortality among children on PD.

Objectives:
To estimate the rate of peritonitis episodes in patients on PD in a tertiary hospital in Portugal from 2013 to 2023 and to investigate clinical and analytical variables associated with higher development of peritonitis.

Methods:
Retrospective cohort study. Peritonitis diagnostic criteria followed the International Society for Peritoneal Dialysis 2022 guidelines. Clinical and demographic variables were analysed with SPSS® v28.0.1.0.

Results:
Total of 18 children reviewed (55.6% boys, median age at the beginning of PD 8.4 years, range 0.6-17.7 years, 94.4% white). Main cause of ESKD was CAKUT (n=5, 27.8%). PD was automated in 70.5% of the episodes. There were 35 peritonitis episodes occurring in 10 patients with a rate of peritonitis of 0.67 episodes per patient-year and median time to first peritonitis episode was 4.6 months (range 0.9-18.2). The most frequent agent was Staphylococcus aureus (n=31, 37.1%) and in 9 episodes (25.7%) the culture was negative. The most used antibiotic association was ceftazidime and vancomycin (n= 20, 57.1%) and median therapy duration 21 days (range 6-42 days). Peritonitis-associated catheter removal occurred in 4 patients and hemodialysis transfer in 1. There were 3 relapsing peritonitis and 5 repeat peritonitis by Staphylococcus aureus and Pseudomonas luteola and 1 refractory peritonitis by Pseudomonas aeruginosa. Automated peritoneal dialysis (APD) was associated with higher peritonitis rate ($p<0,05$).

Conclusion:
In this study peritonitis and negative culture rate were higher than the ISPD recommendations. The most common agent was Staphylococcus aureus and APD was associated with a higher peritonitis rate as previously reported in other studies. The main limitation of this study was the sample size. Review of optimal peritonitis prevention strategies should be made to reduce peritonitis occurrence.

Key words: Peritoneal Dialysis, Peritonitis, Pediatrics, Nephrology

LONG COMMUNICATION - COL 15

THIRTY YEARS EXPERIENCE ON THERAPEUTIC PLASMA EXCHANGES FOR PAEDIATRIC PATIENTS: INDICATIONS AND OUTCOMES. A SINGLE CENTRE EXPERIENCE

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Objectives:
Nowadays, the use of therapeutic plasma exchange (TPE) is accepted for several indications in paediatric diseases, both renal and non-renal.

Methods:
Review the indications and outcomes for TPE in our unit during the last 30 years.

Results:
We retrospectively reviewed all TPE procedures performed by the paediatric nephrology unit in the period 1990-2020.
Results: 151 patients (48% male) with a median age of 8.98 years (1 month - 20.4 years) underwent a total of 1059 TPE sessions. All procedures were performed using a hemofiltration technique and heparin as anticoagulation when it was needed. The central vascular access of choice was internal jugular vein in 59% of the patients. The exchange fluid used was plasma in 52% of the occasions. TPE was performed in combination with primary treatment for the disease concerned in 92% cases.
61 patients were treated for renal diseases, while 90 patients were treated for other indications such as neurological diseases (22), gastrointestinal and liver diseases (21), sepsis/multiorgan failure (18) or haemato-oncological diseases (22).
In the renal disease group 71% of the patients were classified as category I of the American Society of Apheresis (ASFA), while in non-renal indications only 23% belonged to this group.
Forty-two patients (70 %) improved after TPE in the renal disease group compared to 55% in the rest of the pathologies. 36 patients died, all in the non-renal disease group, one third of them in the context of multiorgan failure.

Conclusion:
The benefit of TPE in nephrological diseases associated with primary treatment of the underlying pathology is demonstrated and well categorized in selected conditions. However, in non-renal diseases, especially in critically ill patients, the current evidence is limited. In our series a higher percentage of failure and mortality was observed in this group.

Key words: Therapeutic plasma Exchange, apheresis, Antibody mediated rejection, rapidly progressive glomerulonephritis

LONG COMMUNICATION - COL 16

RENAL FUNCTION IN CHILDHOOD TUBULOPATHIES

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Objectives:
Childhood tubulopathies are a heterogeneous group of entities defined by “primary” or “secondary” abnormalities of renal tubular function. Each entity has its own age of onset, clinical and laboratory manifestations, severity and prognosis.

Methods:
To know the clinical-epidemiological, diagnostic-therapeutic and evolutionary characteristics of primary infantile tubulopathies.

Resultads:
Retrospective and observational cross-sectional descriptive study. Analysis of tubular and renal function. Study population: pediatric population from Valladolid (Spain) diagnosed with primary tubulopathy (≤ 15 years old) in last 10 years.
20 tubulopathies were recorded [10(50%) male, 10(50%) female]. Median age at diagnosis: 5.7 years (2.2-8.6), without differences by sex. Median evolution time: 6.7 years (1.7-10.3). Reasons for consultation in Nephro-Pediatric: 8(40%) poor general condition with hydroelectrolytic alterations, muscle cramps, palpitations and digestive pathology (vomiting, diarrhea and fever), 5(25%) renal colic, 4(20%) hypernatremic dehydration with polyuria/polydipsia and altered acid-base balance, 2(10%) glycosuria and 2(10%) bone deformities and short stature. Tubulopathies diagnosed 10 (50%) of the distal/collecting tubule, 6(30%) proximal and 4(20%) of the Loop of Henle. The most frequent was Gitelman's syndrome [6 patients (30%)]. Treatment was established based on the type of tubulopathy. Evolutively, 11 patients (55%) were diagnosed with impaired urinary concentration, 7(35%) hypercalciuria, 4(20%) developed chronic kidney disease, 10(50%) hypogrowth and 3(15%) required admission secondary to transgression therapy in adolescence due to rejection of medication.

Conclusion:
It is important to diagnose and treat childhood tubulopathies early to improve their renal and vital prognosis, as well as to evaluate the child and his family psychosocially to optimize psychosocial support.

Key words: childhood tubulopathies, renal function

LONG COMMUNICATION - COL 17

GUT PERMEABILITY IN LUPUS NEPHRITIS: RESULTS FROM THE GUT-LUPUS STUDY

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Introduction:
In a systemic lupus erythematosus (SLE) murine model, antibiotics and a vaccine prevented a gut pathobiont translocation, autoimmunity, and death. Furthermore, higher Ruminococcus gnavus abundance was identified in lupus nephritis (LN) patients, reflecting disease activity. We hypothesize, therefore, that dysbiosis, impaired intestinal barrier, and endotoxemia contribute to the chronic activation of the immune system seen in LN

Objectives:
To study diet, physical activity, body composition, gut microbiota, gut permeability, and endotoxemia in SLE patients, with and without LN, and healthy controls (HC).

Methods:
Evaluation of HC and SLE patients (children and adults), who fulfill the 2019 EULAR/ACR SLE classification criteria. Diet and physical activity assessed by three 24h-dietary recalls, PREDIMED, KIDMED, and IPAQ questionnaires. Body composition analyzed by whole-body air-displacement plethysmography. Gut microbiota studied by Next Generation Sequencing (16S rRNA analysis). Lactulose/mannitol test used for direct gut permeability assessment, measured by mass spectrometry. Zonulin and sCD14 evaluated by ELISA. Endotoxemia assessed using TLR4 reporter cell line.

Results:
We studied 16HC (88% females; [14-50Y]) and 45SLE patients (87% females; [11-57Y]; 64% had LN). SLE patients had lower physical activity, lower Mediterranean diet adherence, higher fat mass and decreased gut microbiota a-diversity compared to HC ($p<0.05$, in all). The Rikenellaceae family was increased in SLE, particularly in LN ($p<0.05$). SLE patients had increased gut permeability (Lactulose/Mannitol test 0.0180 vs 0.0140; $p<0.05$), which was higher in LN patients than SLE patients without LN ($p<0.05$). We detected in SLE overexpression of zonulin ($p<0.01$), a gut permeability modulator that disassembles tight junctions between cells of the gut wall. sCD14 ($p<0.05$) and endotoxemia were also increased in SLE.

Conclusion:
SLE patients have dysbiosis and increased gut permeability comparing to HC, particularly if they have renal involvement. The microbiota, the gut barrier and endotoxemia may have a role in LN pathogenesis, being new potential therapeutic targets

Key words: systemic lupus erythematosus, lupus nephritis, microbiota, gut permeability, endotoxemia

LONG COMMUNICATION - COL 18

RENAL OUTCOMES OF CHILDREN WITH IGA NEPHROPATHY

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Introduction:
Oxford classification of IgA nephropathy (IgAN) defines histologic criteria (MEST-C) that provide prognostic information based on kidney biopsy. The International IgAN Prediction Tool (IIgAN-PT) predicts, at the time of kidney biopsy, the risk of a 50% decline in estimated glomerular filtration (eGFR) or progression to end-stage kidney disease (ESKD).

Objectives:
We aimed to correlate MEST-C and IIgAN-PT scores at the time of kidney biopsy with renal outcomes in a pediatric cohort of IgAN.

Methods:
Retrospective cohort study of paediatric patients with biopsy-proven IgAN diagnosed between 2010 and 2022. Demographic, clinical, laboratorial, and histologic variables were analysed. The primary outcome was a composite of eGFR loss, ESKD, or incident proteinuria.

Results:
MEST-C was available for 23 patients (52% were male). Median age at biopsy was 13.8 years (interquartile range (IQR) 9.6;16.3). The MEST-C scores were M1-87%, E1-22%, S1-39%, T1/2-13% and C1-26% and the five-year risk of a 30% decline in eGFR or progression to ESKD according to the IIgAN-PT was 10.5% (IQR 5.5;15.5). At baseline, eGFR was 129 ml/min/1.73m2 (IQR 109.8-150.3) and 20 (87%) patients had significant proteinuria. Over 3.1 (IQR 0.7;7.5) years of follow-up, the median annual eGFR decline was -1.9 (IQR -13.6;1.1) ml/min/1.73 m2 corresponding to -11.8% (IQR -19.1;0.61), and eGFR decreased ≥30% in 3 (13%) patients. Proteinuria remission occurred in 6 (27%) cases. The composite outcome was met by 15 (65%) patients (eGFR decreased in 14 (61%) and 1 (5%) patient received a kidney transplant). No significant correlations were found between MEST-C and IIgAN-PT scores at the time of biopsy and the occurrence of individual and composite renal outcomes over follow-up.

Conclusion:
The sample size may have limited our ability to find significant correlations. Adapted tools may be needed to accurately predict renal outcomes in the pediatric population with IgAN after a period of observation post-biopsy.

Key words: IgA Nephropathy, Paediatrics, MEST-C,

LONG COMMUNICATION - COL 19

NEW TREATMENTS, NEW CHALLENGES: NEPHROTOXICITY-ASSOCIATED-NAXITAMAB IN PEDIATRIC PATIENTS WITH HIGH-RISK NEUROBLASTOMA

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Introduction:
Neuroblastoma is the most common extracranial solid tumor in pediatrics,having a poor survival in high-risk(HR)tumors.Naxitamab(hu3F8)is a humanized monoclonal antibody anti-dysialoganglioside (GD2) approved for treatment of >1-year-old and adults with refractory/relapsed HR-neuroblastoma limited to bone or bone marrow.Our hospital has been the first center(2017)worldwide use it(clinical trials/compassionate use),obtaining excellent results

Objectives:
Describe renal AE associated-Naxitamab in treatment of HR-pediatric-neuroblastoma in our hospital,from its establishment until today.

Methods:
Retrospective descriptive study including 244 patients(41% female-101-and 59% male-143-)using Naxitamab(monotherapy and/or associated with chemotherapy)from June 2017 to current day(6 years)in whom renal involvement and/or hypertension(HT)was evaluated

Results:
Mean age was 8 years,presenting nephrotoxicity of some type up to 26.6%(65):HT(11.9%/29),acute renal damage(ARD10.2%/25)and proteinuria(5.3%/13),developing all of them during the infusion or the first 3 cycles.In case of HT,only in 6 patients an ABPM was performed,observing:2 nocturnal-HT,2 diurnal-HT without specific-pattern and 2 disautonomic-pattern,not previously observed.Among the ARD,all cases were tubular except for one patient who presented clinical-analytical pattern of acute tubule-interstitial nephritis(AIN)and another who presented thrombotic microangiopathy(TMA)with subsequent confirmation of heterozygous CFHR1-CFHR4 deletion.Eight of them(32%)presented possible confounding factors in the development of(previous chemotherapy,ibuprofen or radiotherapy).Among patients with proteinuria(none nephrotic range):38% tubular,38% glomerular and 23% mixed.2 patients presented ARD+AHT and 3 a combination of AHT+ARD+proteinuria.Of these last,all of them received prior chemotherapy,leaving 2 of them with chronic renal damage(CKD stage 2 and 3).67 patients(27.4%)died due to progression of their underlying disease.

Conclusion:
Management of HR-neuroblastoma remains a daily challenge.Naxitamab is an emerging therapy in this type of tumors,although there are few studies describing its AE.Previous studies of our group in mice explain the involvement of the myelin sheaths of the autonomic nervous system with this drug,which could explain,among others,the dysautonomic pattern of blood pressure presented.Short- and long-term follow-up,the systematic performance of ABPM and the use of early markers of renal damage,could lead to a more efficient management of complications derived from this new treatment

Key words: Naxitamab, Nephrotoxicity, Onco-Nephrology, Disautonomic-Pattern

LONG COMMUNICATION - COL 20

PAEDIATRIC REFERENCE VALUES FOR URINE PH AND URINARY AMMONIUM IN SINGLE SPOT URINE

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Introduction:
The kidney plays a crucial part in the regulation of acid-base balance by replacing the bicarbonate consumed in the buffering of non-volatile acids through urinary acidification and the excretion of ammonium (NH4+). These processes suggest that urine pH (UpH) and urinary NH4+ (UNH4+) are valuable tools to assess urinary acidification. However, the technical
difficulties for direct UNH4+ quantification and the lack of data on these parameters in children, have limited their application in clinical practice.

Objectives:
To establish reference values of UpH and UNH4+ (as the NH4+/creatinine ratio) in single spot urine samples in paediatric population, in order to promote the use of spot urine samples for the evaluation of urinary acidification.

Methods:
The first morning urine sample, after an overnight fasting period, was collected in healthy continent children, aged from 4 to 14 years. UpH was immediately determined by direct potentiometry and the samples were frozen at -80°C after centrifugation, until UNH4+ quantification (Cardo et al. Clin Chem Lab Med, 2019). Results were expressed as median value and range. Reference values were determined by the non-parametric method (as 2.5th and 97.5th percentiles), and 90 % confidence intervals were indicated for each limit.

Results:
157 subjects were recruited (44.6% female). Median value obtained for UpH, was 5.69 (range 4.83 - 7.60), and for UNH4+/creatinine ratio, was 5008 mmol/mol (range 1395 - 14826). Reference values for UpH and NH4+/creatinine ratio ranged from 4.99 (4.83 - 5.10) to 6.85 (6.71 - 7.60), and from 1737 mmol/mol (1395 - 2227) to 12784 mmol/mol (10695 - 14826), respectively.

Conclusion:
This study provides, to our knowledge, the first reported paediatric reference values for the simultaneous evaluation of UpH and UNH4+. These reference values will facilitate the reliable use of an isolated urine sample to assess urinary acidification in children in clinical practice.

Key words: urine pH, urinary ammonium, single spot urine, urinary acidification

LONG COMMUNICATION - COL 21

CORRELATION AND CONCORDANCE BETWEEN URINARY INDEXES FOR THE STUDY OF RENAL LITHIASIS BETWEEN 12-HOUR OVERNIGHT AND 24-HOUR SAMPLES: STRENGTHS AND WEAKNESSES

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Objectives:
Collecting 24-hour urine samples can be complex for many patients. Incorrect collection can derive in biased results. Shorter periods can palliate this problem and detect peaks of risk for crystallization. Interpretation of results from this samples compared to standard 24-hour ones is debatable.

Methods:
We compare urinary indexes in overnight 12-hour samples and over 24 hours regarding correlation and concordance.

Results:
Twenty-six pediatric stone-formers aged between 5 and 17 years old were recruited from the outpatient clinic in a tertiary care center. Eighty-seven healthy individuals of the same group of age were recruited from local schools. Spontaneous ambulatory 24-hour urine samples were obtained, fractioned in two consecutive 12-hour periods (daytime and overnight). Calcium, phosphate, magnesium, uric acid, oxalate, citrate, and creatinine were measured for each period. Urinary indexes for overnight 12-hour sample and 24 hours were compared regarding correlation (Spearman test) and concordance in detecting abnormal results according with reference values for 24-hour samples.
In healthy subjects, correlation coefficients for calcium, phosphate, uric acid, oxalate, magnesium and citrate to creatinine ratios, and calcium to citrate ratio, were, respectively: 0.92, 0.86, 0.89, 0.90, 0.91, 0.92 and 0.92. In stone-formers, correlation coefficients were, also respectively: 0.87, 0.90, 0.76, 0.96, 0.92, 0.92 and 0.90. Most of abnormal results over a 24-hour period were detected in overnight samples. Remarkable exceptions are 11% individuals with normal calcium-to-creatinine ratios in overnight samples when abnormal in 24 hours and 18% ones with abnormal calcium-to-citrate ratio in overnight samples when normal in 24 hours, both in stone-formers.

Conclusion:
Correlations between indexes from overnight 12-hour samples and over a 24-hour period were high for all parameters. Nevertheless, circadian pattern should be evaluated before using 12-hour samples for follow-up, especially for calcium. 12-hour samples can better detect increased calcium-to-citrate ratio. Specific reference values for 12-hour samples could give a more accurate interpretation.

Key words: Urinary indexes, Renal lithiasis, 12-hour sample, 24-hour sample

SHORT COMMUNICATION - COC 01

HOW ARE WE MANAGING PATIENTS WITH UNILATERAL RENAL AGENESIS? A SURVEY OF THE YPNN-AENP

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Introduction:
Unilateral renal agenesis is usually diagnosed prenatally. Due to the occurrence of complications in adults, debate arises about the need of monitoring during childhood and adolescence. It is not controverted that follow-up should comprise blood pressure measurement and proteinuria and renal function calculation. However its frequency has not been properly established.

Objectives_
To determine the current practice of Spanish paediatricians in the diagnostic and therapeutic management of patients with primary unilateral renal agenesis.

Methods:
Cross-sectional study on the management of prenatally-diagnosed unilateral renal agenesis in children and adolescents by paediatricians. An anonymous online survey was conducted, excluding patients with a single functioning kidney of other etiology.

Results:
103 paediatricians participated in the study. 64(62.1%) work in hospital care, while 39(37.9%) work in primary care. 3.1%(2) of those in specialty care do not monitor renal agenesis. 47(73.4%) paediatric nephrologists initially perform renal scintigraphy, 35(54.7%) estimate glomerular filtration rate and 46(71.9%) perform urine tests. Only 21(32.8%) look for genital congenital malformations by abdominal ultrasound during puberty. No recommendation is made to reduce protein intake. 57(89.1%) transition these patients to primary care while 5(7.8%) to adult nephrology departments. 94.9%(37) primary care paediatricians refer these patients to paediatric nephrology clinic. All primary care paediatricians can request a renal ultrasound but only 61.5%(24) have access to paediatric blood pressure cuffs.

Conclusion:
-There is great variability in the follow-up and treatment of these patients by paediatric nephrologists.
-Follow-up of these patients in primary care could be limited by the unavailability of paediatric blood pressure cuffs.
-The creation of a protocol for the diagnosis and therapeutic management of congenital solitary kidney in the paediatric age should be considered.

Key words: primary unilateral renal agenesis, survey

SHORT COMMUNICATION - COC 02

CONTRAST-ASSOCIATED ACUTE KIDNEY INJURY IN CHILDREN WITH CONGENITAL HEART DISEASE UNDERGOING CARDIAC CATHETERIZATION. INCIDENCE AND FEATURES

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Introduction:
Contrast-Associated Acute Kidney Injury (CA-AKI) entails sudden deterioration of kidney function within 7 days after intravenous contrast media administration. Variable incidence, 1-30% in adults, considered lower in children, especially since hipo/iso-osmolar contrast agents generalization.

Objectives:
To analyse the incidence and related factors of CA-AKI in children with heart disease.

Methods:
Prospective analytical study, enrolling patients 0-18 years old with heart disease undergoing cardiac catherization using Optiray-300 iodized injectable solution (July-2021 to April-2022). CA-AKI prophylaxis protocol was applied.
Demographic, clinical, and specific intervention variables were collected. Kidney function parameters were collected from blood and urine samples prior to catheterization and after 6-24-48 hours, and 7 days. In patients considered to be at higher baseline risk, NGAL and KIM in urine were also obtained.

Results:
126 patients were included. Median age: 7.6 (60% females). 75% came from Paediatric Cardiology ward, 21% Neonatal Intensive Care Unit (ICU) and 3% Paediatric ICU. Median amount of intravenous contrast within every catheterization 3.2 ml/kg, total iv fluids: 11 ml/kg.
14 (11%) patients developed CA-AKI. 11 (79%) came from Neonatal ICU. 2 had prior established chronic kidney disease. Significant association ($p<0.05$) with CA-AKI development was observed with: $^{3}4$ ml/kg of intravenous contrast, adverse events during catheterization, transfusion and nephrotoxic drugs prior to catheterization.
CA-AKI management was habitual according to hospital's protocols. Extrarenal clearance techniques were not necessary. All patients recovered kidney function.
Samples for NGAL and KIM1 urinalysis were collected from 13 patients: 2 (15%) developed CA-AKI; NGAL and KIM1 were elevated in the first 6 hours.

Conclusion:
CA-AKI is rare in children with heart disease undergoing cardiac catheterization using intravenous contrast after a prophylaxis protocol. It is associated with previous risk factors, higher contrast amount or complications during procedure. NGAL and KIM1 could have an added value in early diagnosis of CA-AKI.

Key words: contrast-associated acute kidney injury, cardiac catheterization.

SHORT COMMUNICATION - COC 03

POST-TRANSPLANT OUTCOMES AND COMPLICATIONS IN PEDIATRIC KIDNEY TRANSPLANT RECIPIENTS: A SINGLE CENTRE TWELVE-YEAR COHORT

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Introduction:
Kidney transplant (KT) reference centres regularly assess information regarding post-transplant complications, patient, and graft survival aiming to identify opportunities for clinical care improvement and research questions.

Objectives:
To update data on paediatric post-KT events at a reference centre.

Methods:
Retrospective cohort study of all paediatric patients who underwent KT at a tertiary centre between 2011-2022. Baseline characteristics, post-KT events, and outcomes were evaluated. Chi-squared or Mann-Whitney-U tests were used for comparisons and age-adjusted regression models for event prediction.

Results:
77 KT were performed (76 recipients; 57% male) - 21 (27%) preemptive and 13 (17%) from a living donor. Age at KT was 12.3 [7.2;15.8] years. Induction with thymoglobulin was used in 35 (46%) and triple-drug maintenance regimen in all cases. Over 3.7 [2.3;5.59] years of follow-up, two (2.6%) patients died and two (2.6%) lost the graft. Acute rejection was diagnosed in 19 (25%) and chronic nephropathy in 12 (16%). Early surgical and vascular complications were seen in 9 (12%) and 11 (14%), respectively. Neoplasia occurred in 4 (5%). Regarding cardiometabolic complications, hypertension occurred in 64 (84%), dyslipidaemia in 17 (22%), and post-transplantation diabetes mellitus in 8 (11%). Crude incidence of bacterial infections was 54 (73%), including urinary infections (n=39; 51%). Viral infections occurred in 27 (41%), including CMV (n=16; 21%) and EBV (n=14; 18.2%) infections, which were more frequent among “donor+/recipient-” pairs than other pairs combined (CMV: 47% vs 13%, p=0.001; EBV: 63% vs 20%, p=0.024). No significant predictors were identified in the regression models.

Conclusion:
Compared with previously published data from our centre (1995-2010), the proportion of living donors and preemptive KT has increased, while graft loss has decreased. Infectious and cardiometabolic complications remain similar. Improved patient and graft outcomes over time may lead to less intensive immunosuppression protocols tailored according to risk profile, possibly decreasing the burden of infectious and cardiometabolic complications.

Key words: Pediatric Renal Transplant, Complications

SHORT COMMUNICATION - COC 04

AMBULATORY BLOOD PRESSURE MONITORING IN 40 KIDNEY TRANSPLANT PATIENTS

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Introduction:
Hypertension is a common complication in post-renal transplantation. Ambulatory blood pressure monitoring (ABPM) is the best method of assessment as it can help to detect alterations that might not be observed in the consultation room.

Objectives:
Describe ABPM findings in children being followed for kidney transplant in our centre.

Methods:
Retrospective and preliminary observational descriptive study in patients with their first renal transplant who underwent an ABPM study.

Results:
ABPM was performed in 40 patients, 72.5% male and 55% from cadaveric donor. Median age at transplantation was 7 years (IQR: 6.75). 19 patients (47.5%) were under antihypertensive treatment prior to recording.
There were 18 patients (55%) with hypertension on ABPM (group 1): 9 had masked hypertension revealed by ABPM and in the other 9 the consultation finding was confirmed. 10 of the 18 patients (55.5%) were receiving antihypertensive treatment at the time of recording and, of these patients, 3 were diagnosed with masked hypertension. A total of 72% had lost the dipping pattern and almost half had left ventricular hypertrophy (47%).
22 patients had no hypertension on ABPM (group 2). 9 patients were under antihypertensive treatment. 73% had no nocturnal dipping.
Statistical analysis revealed no significant differences between groups (group 1; group 2) in the following variables: recipient age (6.83±4.35; 8.86±4.24 years; p-value: 0.145), donor age (22.67±13.14; 24.55±14.58 years; p-value: 0.674), glomerular filtration rate estimated according to Schwartz (71.43±23.94; 66.31±22.78 ml/min/1.73 m2; p-value: 0.49) and time since transplantation (55.86±42; 76.28±54.19 months; p-value: 0.187).
Only 2 cases had renal artery stenosis (1 in each group).

Conclusion:
50% of our children with confirmed hypertension have poor blood pressure control. Given the necessity of controlling hypertension and the high incidence of masked hypertension, ABPM is an essential complementary test for the management of kidney transplant recipients.

Key words: Ambulatory Blood Pressure Monitoring, Kidney transplantation, Children, Blood pressure

SHORT COMMUNICATION - COC 05

INFECTIOUS COMPLICATIONS ASSOCIATED WITH THE USE OF CUFFED-TUNNELED CENTRAL VASCULAR CATHETERS FOR HAEMODIALYSIS, 10-YEAR EXPERIENCE IN A SINGLE PAEDIATRIC CENTRE

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Objectives:
The use of cuffed-tunnelled central venous catheters (CCVC) as vascular access for haemodialysis in paediatrics is common clinical practice in our setting. Its management is not free of complications. Infections are of special relevance given their high morbidity and risk of vascular access failure.

Methods:
Describe infectious complications, as well as their influence on survival of vascular access in our patients.

Results:
We retrospectively reviewed the medical records of patients carriers of CCVC undergoing haemodialysis in our unit int the period from 2012-2022.
181 CCVC were used during this period in 105 patients (60% patients required a single catheter). 63 infectious complications were recorded in 60 catheters over 10 years: 25 catheter-related bacteraemia, 28 exit site infections and 10 tunellitis. The infection rate was 1.42/1000 catheter-days with a probability of remaining infection-free at 6 and 12 months of 82% and 64% respectively. 32% of infectious events resulted in vascular access replacement. Catheter-related bacteraemia was the most frequent infectious cause of failure (20% failures in the sample). There was no case of mortality related.
Staphylococcus aureus was the most frequent isolated micro-organism (36 occasions), followed by Staphylococcus epidermidis (12) and Pseudomonas aeruginosa (6). Seventy percent of the S.Aureus related cases were nasal carriers.
Median catheter survival in the infection group in our sample was lower (10.7 months vs. 15.66 months in the absence of infection), although this difference was not statistically significant (p=0.14).

Conclusion:
Infectious complications related to CCVC are an important cause of CCVC failure, leading in our sample to decreased catheter survival. Our infection rate was similar to that reported in the literature. We observed a higher rate of S. Aureus nasal carriers in those patients with infectious complications.

Key words: Haemodialysis, Cuffed-tunnelled central venous catheters, central venous catheter-related infections, chronic kidney disease

SHORT COMMUNICATION - COC 06

THE RELATION BETWEEN COVID-19 PANDEMIC AND HEMOLITIC UREMIC SYNDROME.

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Introduction:
Hemolytic uremic syndrome (HUS) is a life-threatening disease classified as a thrombotic microangiopathy (TMA), where complement dysregulation plays an important role. Infections are common triggers of HUS, being Shiga toxin-producing Escherichia coli (STEC) the most frequent associated pathogen.
Since 2020 some authors have hypothesized that a relation between COVID-19 and TMAs could exist, but mechanisms are unclear.

Objectives:
To study how COVID-19 pandemic has influenced the epidemiology of HUS, we conducted a single-center retrospective medical chart review.

Methods:
We analyzed clinical and epidemiological characteristics of HUS patients before (2008-2019) and during COVID-19 pandemic (2020-2022).

Results:
We had 18 patients with HUS, 17 typical and 1 atypical. Since COVID-19 pandemic, we observed a higher incidence (3 versus 0,75 cases/year), and affected patients were older (5,2 versus 2,5 years old, p=0,034). Their laboratory workup showed lower creatinine (1,43 versus 4,23 mg/dl, p=0,02, CI 95% 0,45-5,13) and LDH (2094 versus 3099 UI/l, p=0,043, CI95% 37,1-1972,35) at admission. Only 5/9 were admitted in the pediatric intensive care unit (PICU), and the need for renal replacement was lower (22% versus 100%, RR 4,5, p=0,02, CI95% 1,32-15,27). We could not document active SARS-CoV2 infection or recent vaccination in post-pandemic patients.
Among pre-pandemic patients, 5/9 were lost to follow up, while 50% of the rest have persistent renal dysfunction. All post-pandemic patients exhibited complete renal function recovery.

Conclusion:
In conclusion, we observed a surprising incidence peak of milder cases of HUS during the pandemic. Despite not being able to document recent active SARS-CoV2 infection or vaccination, we cannot rule out previously undiagnosed oligosymptomatic infections in our patients,as has been described in the literature. Possible explanations include microbiome shifts, lack of microbial exposure derived by extensive hygiene, or COVID-19-related hyperinflammatory immune response deriving in a lower threshold for Shiga-toxin to trigger HUS.

Key words: Covid-19, hemolitic uremic syndrome

SHORT COMMUNICATION - COC 07

OUR INITIAL EXPERIENCE WITH BUROSUMAB IN CHILDREN WITH X-LINKED HYPOPHOSPHATEMIC RICKETS (XHR)

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Introduction:
X-linked hypophosphatemic rickets (XHR) is caused by PHEX gene mutations, which lead to serum FGF23 elevation causing renal phosphate wasting with hypophosphatemia, low calcitriol levels, rickets, osteomalacia and linear growth impairment. Treatment with Burosumab, a humanized monoclonal antibody against FGF23, has modified XHR management and disease outcome. We describe our case series of patients treated with Burosumab for a year.

Methods:
7 children with confirmed PHEX mutations are described. We analyzed their disease characteristics at the initiation of burosumab (thus after at least one year with classic treatment with phosphate and calcitriol supplements in all, rhGH in 5, and rickets severity score (RSS) ≥2), and at 3,6,9 and 12 months later.

Results:
After 1 year with Burosumab mean grouped serum Phosphate (P) increased from 2.29 to 3.49mg/dl, and 6 out of 7 patients normalized their phosphatemia due increased tubular reabsorption (TmP from 2.16 to 3.01), whereas RSS diminished from 3.5 to 0.79. However, growth catch up was not observed and 4 patients resumed treatment with rhGH. No side effects occurred but local pain at puncture area.

Conclusion:
Burosumab treatment in XLH patients achieved rickets healing due correction of renal P wasting, with normalization of phosphatemia but limited impact on growth.

Key words: X-linked hypophosphatemic rickets, PHEX, burosumab, growth

SHORT COMMUNICATION - COC 08

OUR EXPERIENCE WITH LUMASIRAN TREATMENT IN TWO PATIENTS WITH PRIMARY HYPEROXALURIA TYPE 1 (PH1)

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Introduction:
PH1 is a rare and severe disease, caused by AGXT gene mutations. It is characterized by hepatic overproduction of oxalate, leading to elevated urine oxalate/glycolate. PH1 manifestations are kidney stones, nephrocalcinosis, CKD, kidney failure and oxalosis. Standard treatment based on hyperhydration and alkaline citrate is poorly effective. However, vitamin B6 (pyridoxine), is helpful in some PH1 patients with specific missense or mistargeting mutations, achiving reduction/ normalization of urinary oxalate and glycolate. Lumasiran a novel approved RNA interference(RNAi) drug reduces liver oxalate production by targeting glycolate oxidase, and represents a very promising therapy for PH1.

Objectives:
To evaluate the efficacy and safety of lumasiran plus standard treatment in two PH1 children

Results:
Two PH1 moroccan children from unrelated consanguineous families treated with lumasiran:
Patient 1: 11y boy with homozygous AGXT c.731T>C (p.I244T) mutation at the minor haplotype. He presented repeated lithiasis-obstruction episodes with pyelonephritis and acute renal injury (nadir of eGFR 29 mL/min/1,73 m2). Before lumasiran he was CKD G2A2 with Uoxalate/Cr 130,06 mmol/mol (normal <70). After 19 months, he remains CKD G2A1, free of stone events with Uoxalate/Cr 113,5 mmol/mol.
Patient 2: 12y girl, with homozygous AGXT c.731T>C (p.I244T) mutation at the minor haplotype. She consulted at 1 year with abdominal pain, feeding difficulties, bilateral nephrocalcinosis and normal eGFR. Before lumasiran she was CKD G1A1 with Uoxalate/Cr 432,78 mmol/mol (normal <70). After 19 months, she is CKD G2A1 with Uoxalate/Cr 145,5 mmol/mol.
We describe those patients follow up with lumasiran. No side effects were observed.

Conclusion:
Lumarisan is a new and highly effective RNAi treatment for PH1. Lumasiran is indicated in unresponsive patients to supportive treatment and pyridoxine. Long-term experience is needed to see the impact of lumasiran on PH1 natural history, and design patient individual management based on genotype and disease severity.

Key words: hyperoxaluria, lithiasis, CKD, pyridoxine, RNAi, lumasiran

SHORT COMMUNICATION - COC 09

ANOMALIES OF PHOSPHO-CALCIUM METABOLISM IN A CASE SERIES OF CHILDREN WITH OSTEOGENESIS IMPERFECTA IN RELATION TO TREATMENT WITH DENOSUMAB AND BIPHOSPHONATES

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Introduction:
Osteogenesis imperfecta (OI) is a heterogeneous group of inherited diseases which are affecting the connective tissue with an estimated incidence of 1/15,000-20,000 births. It's characterized by bone fragility but there is a broad clinical spectrum. Around 85-90% of cases are caused by mutation in COL1A1/COL1A2, with up to 18 non-collagenous genes involved in the remaining 10%.
In severe forms, biphosphonates are indicated in the initial treatment, employing denosumab in those with poor response to previous treatment or in some special forms. The use of these drugs could produce alterations in phosphor-calcium metabolism.

Objectives:
Genetic, blood analysis and urinalysis and ultrasound description of a case series diagnosed with severe OI in combined treatment with denosumab and biphosphonates.

Methods:
Retrospective and observational study, by reviewing the medical history of 5 patients diagnosed with severe OI under treatment with denosumab to which zolendronic acid was subsequently associated.
The following variables are included: genetics, age at diagnosis, ultrasound alterations and blood and urine parameters at different points (after one year of denosumab [C1], before start the combined treatment [C2], one year after start the combination [C3] and the most recent [C4]).
Qualitative variables are expressed as percentages and quantitative variables as median and interquartile range.

Results:
Five patients (60% female) were included. 3/5 had mutation in COL1A1, one in COL1A2 and one in SERPINF.
Calcaemia had medians between 9.19-9.33mg/dl (normal 8.2-10.2mg/dl), phosphataemia 4.9-5.35mg/dl (normal 4.5-6.5mg/dl), PTH 15.5-26.5pg/ml (normal 18.5-88pg/ml), vitamin D 19-27ng/ml (20-55ng/ml) and beta-cross 0.39-0.93ng/ml.
No glomerular filtration rates <90ml/min/1.73m2 were observed in the analytical controls.
Approximately 4/5 (80%) presented hypercalciuria at any time during evolution. The median calciuria during monotherapy treatment was 0.4mg/mg and with combined treatment 0.2mg/mg.
1/5 (20%) developed nephrocalcinosis during follow-up with combination therapy.

Conclusion:
A decrease in calciuria was observed with the association of bisphosphonates but was not statistically significant.

Key words: Osteogenesis imperfecta, Hypercalciuria, Denosumab, Biphosphonates

SHORT COMMUNICATION - COC 10

PSEUDOHYPOALDOSTERONISM TYPE 1 IN THE LAST DECADES: A CASE SERIES FROM OUR PERSPECTIVE

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Introduction:
Pseudohypoaldosteronism type 1 (PHA1) is a rare hereditary disorder characterized by resistance to the actions of mineralocorticoids. It presents with hyperkalemia, hyponatremia, metabolic acidosis and elevated plasma aldosterone and renin levels. The autosomal recessive form is a systemic disease with worse prognosis than the autosomal dominant one, which is more frequent and leads exclusively to renal involvement.

Objectives:
We aim to summarize the differences and similarities between two patients with PHA1 who were diagnosed in our nephrology unit.

Methods:
We have compared the clinical, diagnostic and therapeutic aspects of two cases of PHA1.

Results:
We present two cases, both of them were term newborns with appropriate birth weight. Disease onset occurred on the sixth and fifth day of life respectively. The first one, a male patient born in 1995, has a familiar history of excessive salt craving. The second case is also a male patient born in 2022, with consanguineous parents and a deceased sister due to severe dehydration as a newborn.
In both cases, the main symptoms were severe dehydration associated with hyponatremia, hyperkalemia and weight loss in the first week of life (16.6% and 17% respectively). PHA1 remained as the most probable diagnosis due to elevated plasma aldosterone and renin levels without arterial hypertension or anatomical signs of uropathy. Treatment was promptly initiated in both cases with fludrocortisone, sodium ion exchange resin, bicarbonate and sodium supplements. The second patient required peritoneal dialysis.
In the first case, a genetic study was performed in 2016, finding homozygous deletion c.1305delC (p.Y436IfsX46) in exon 8 in the SCNN1A gene. For the second case, homozygous duplication c.1339dup (p.Tyr447LeufsTer13) in SCNN1A recently confirmed the diagnosis.

Conclusion:
The evolution of genetic techniques has allowed a swift diagnosis of PHA1. Early treatment is essential for these patients, even before confirmatory diagnosis has been reached.

Key words: pseudohypoaldosteronism, genetics, case series

SHORT COMMUNICATION - COC 11

GENETICS AS A TOOL TOWARDS DIAGNOSIS

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Objectives:
Since the sequencing of the genome, genetics has experienced an exponential development. Today, given its accessibility, it is considered yet another tool for the diagnosis of our patients.

Methods:
To analyse the cost-effectiveness of genetic studies performed in our hospital in patients with non-filiated nephrourological pathology or with an atypical course and to determine their influence on clinical management.

Results:
Retrospective observational study of genetic studies requested by our Unit in the last 5 years in patients with uncertain clinical diagnosis. Patients were grouped according to the following clinical syndromes: haematuria, nephrotic syndrome/proteinuria, congenital anomalies of the urinary tract (CAKUT), cystic nephropathy, lithiasis and arterial hypertension.
The genetic studies were carried out from peripheral blood samples performing a massive sequencing of the complete exome, bioinformatic analysis with own pipeline (Karma©) and subsequent filtering and prioritization of genetic variants or virtual gene panels according to the reason of the request with sequential analysis. Genetic variants were then classified as: pathogenic, probably pathogenic, uncertain significance, probably benign and benign.

Conclusion:
A total of 69 genetic studies were carried out with a positive diagnosis in 37 of them.
When dividing the patients into subgroups, the most frequently requested panels were those associated with renal cysts (isolating a pathogenic variant in 59%), followed by CAKUT (positivity <1%), nephrotic syndrome/proteinuria (positivity 25%), haematuria (positivity 70%), lithiasis (positivity 25%), tubulopathy (positivity 100%) and hypertension (3 patients only, positivity 100%).
Performing genetic studies in our hospital showed a high cost-effectiveness (>50%) for detecting pathogenic variants in patients with uncertain clinical diagnosis.
The clinical group with the highest profitability was the one of cystic nephropathies being the less one the subgroup of CAKUT. Genetic diagnosis had implications in a significant number of patients, both in terms of treatment and follow-up, confirming the importance of genetic studies in clinical practice.

SHORT COMMUNICATION - COC 12

CIRCADIAN VARIABILITY OF URINE COMPOSITION AND LITHOGENIC RISK SHOWS A SIMILAR PATTERN BETWEEN PEDIATRIC HEALTHY SUBJECTS AND STONE-FORMERS

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Objectives:
Urine composition can be vary modulated by oral intake, activity and individual intrinsic factors, often producing a circadian pattern. This variability can affect the individual risk for urinary crystallization along the day. Whether this variability is similar or different between healthy and stone-forming pediatric subjects is uncertain.

Methods:
We aim to describe the circadian variability in urinary parameters related to crystallization in children and adolescents comparing healthy individuals with stone-formers.

Results:
Twenty-six pediatric stone-formers aged 5-17 years old were recruited from the outpatient clinic in a tertiary care center. Twelve of them had been diagnosed with renal lithiasis and 14 ones presented with pre-lithiasic symptoms (hematuria, dysuria, overactive bladder or chronic abdominal pain related to urinary crystallization). Eighty-seven healthy individuals of the same group of age were recruited from local schools as a control group. Spontaneous ambulatory 24-hour urine samples were obtained, fractioned in two consecutive 12-hour periods (daytime and overnight). Calcium, phosphate, magnesium, uric acid, oxalate, citrate, creatinine, pH and urine volume were measured for each period at a research specialized laboratory.
In both groups, overnight excretion of phosphate and magnesium was higher; on the contrary, uric acid and citrate excretion, as well as pH and urine volume, were lower. Calcium and oxalate average excretion did not differ between both periods but we observed day to night differences according to each subject. Lithogenic risk was higher in the overnight period in both healthy and stone-forming subjects according to higher calcium to citrate ratio, as well as the previously mentioned findings.

Conclusion:
An altered pattern of the circadian variability in urinary solute excretion does not seem to play a role in kidney stone formation, as it is similar in healthy subjects and patients. A higher overnight risk for stone formation is confirmed in this study.

Key words: Urine composition, Circadian, Renal lithiasis

SHORT COMMUNICATION - COC 13

LONG TERM OUTCOMES IN IGA VASCULITIS: SINGLE CENTRE EXPERIENCE

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Objectives:
IgA vasculitis (IgAV) is a small vessel vasculitis, characterized by IgA deposits presenting between 2-10 years (incidence 3-26 cases 100 000), associated with renal involvement in 20-30% of cases. The risk of progression to chronic kidney disease (CKD) is small although it may increase in presentations with nephrotic syndrome and/or renal failure. In recent years it has been proposed to be more aggressive in treatment as historically it had been considered a benign entity.

Methods:
A descriptive, retrospective study was designed to describe the long-term evolution of patients with a diagnosis of IgAV referred to our clinic from 1983 to 2022.

Results:
We reviewed 91 patients (43 males) with mean age 8.1 (SD +/- 2.79 years) and median follow-up 11.5 (1-5 years). At the beginning of the cutaneous symptoms 49% had urinary alterations and 50% received steroids due to extrarenal indication.
All were referred for persistence/appearance of urinary signs (15% hematuria, 81% hematuria and proteinuria, 3% isolated proteinuria; 14% proteinuria in nephrotic range), 5% decreased glomerular filtration rate (GFR) and/or 9.8% arterial hypertension. In 13 cases the diagnosis was confirmed by biopsy.
After our evaluation 10 patients received immunosuppression (10/10 methylprednisolone, azathioprine 2/10, cyclophosphamide 2/10).
75/91 continued follow-up, and 22% received ACE inhibitors. Throughout evolution 14 presented outbreaks of macroscopic hematuria.
At the end of follow-up 49% were in remission with normal GFR, 20% maintained microhematuria, 22% proteinuria, 76% of them were controlled with ACE inhibitors. 4 developed CKD: 1 dialysis 3 years after presentation and 3 with borderline GFR (81-85ml/min/1.73m2).

Conclusion:
In our historical series a low percentage of patients were biopsied or received immunosuppression. Although 22% maintain low grade proteinuria, GFR evolution has been favorable, apart from 1 patient who had a rapidly progressive renal failure. The clinical impact of persistent urinary alterations must be evaluated in longer term studies.

Key words: IgA vasculitis

SHORT COMMUNICATION - COC 14

A COMPARATIVE STUDY OF ACUTE POST-STREPTOCOCCAL GLOMERULONEPHRITIS IN THE CURRENT STREPTOCOCCAL EPIDEMIC VERSUS PREVIOUS SEASONS. HAS THE SARS-COV2 PANDEMIC INFLUENCED RESULTS?

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Introduction:
Streptococcus pyogenes (SP) causes multiple variety of infection (local, invasive, toxin-induced, immunological). Acute poststreptococcal glomerulonephritis (PEGN) is a form of immunological disease that usually triggers a nephritic syndrome whose prognosis depends on the persistence of massive proteinuria and a rapidly progressive presentation.
On December 2022, United Kingdom informed about a SP epidemic with an increase in infant mortality due to invasive disease. It is postulated that an "immunological debt" acquired during SARS-CoV2 pandemic could be responsible.
In our center, we have shown an increase in PEGN in accordance with the aforementioned epidemic peak.

Objectives:
To analyze mean characteristics of PENG in our hospital in the last 10 seasons and compare them with the current one.
To assess whether the renal immunological disease produced by SP has changed in terms of aggressiveness in relation with SARS-CoV2 pandemic.

Methods:
This is a descriptive and retrospective observational study. We have selected all PEGN diagnosed in our hospital from 2010-2011 season to the current one (n=23).

Results:
Our study shows seven times increase in the number PEGN this season; a higher prevalence of acute kidney injury, nephrotic range proteinuria and edema in the current season, but without statistically significant differences.
Comparing children under 5 with older ones (for having less streptococcal contact during SARS-CoV2 pandemic) within the current season, we have a higher prevalence of hypertension and a higher average proteinuria in younger children; but without statistically significant differences.

Conclusion:
Our data show an increase in PEGN in the current season, according to the epidemiological situation at the time. Although, it has not been proved more aggressiveness.

The "immune debt" produced during SARS-CoV2 pandemic could be one of the triggers for the higher prevalence.
Our lack of statistical significance can be due to the small sample size. We may obtain a greater statistical power with a multicenter study.

Key words: Acute poststreptococcal glomerulonephritis, SARS-CoV2

SHORT COMMUNICATION - COC 15

ALPORT SYNDROME: 15 YEARS IN A TERTIARY HOSPITAL

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Introduction:
Alport syndrome is a rare genetic condition caused by a defect in type IV collagen that affects the kidney, ear and eye, and is most commonly inherited by X-linked pattern. Most patients progress to ESRD in adulthood.

Objectives:
To characterize an Alport syndrome cohort followed in a Pediatric Nephrology unit of a tertiary hospital.

Methods:
Retrospective observational study of Alport syndrome patients followed between 2007 and 2022.

Results:
Twenty-two patients were followed for suspected Alport Syndrome. Four were excluded (lost follow-up or had negative genetic tests) and three had the genetic diagnostic while adults. Median age at first evaluation was 4,7 years-old, 55% females. Fifteen had X-linked and three had recessive inheritance. Reasons of referral were hematuria (13), hematoproteinuria (2), family history of Alport syndrome (2), and established genetic diagnosis (1). Thirteen patients had a family history of CKD, nine with renal transplants in 1st or 2nd degree relatives. At admission, 10 had hematoproteinuria, five isolated hematuria and two isolated proteinuria, all non-nephrotic. All maintained follow-up with regular evaluations of blood pressure, renal function and proteinuria/hematuria to monitorize disease progression. During follow-up, six patients started ACE inhibitors and three added an ARB to control worsening proteinuria. On the last evaluation, all of them maintained hematuria, three no longer had proteinuria, three had progressed to nephrotic proteinuria and one had new-onset proteinuria. Two of the patients with recessive inheritance had the worst outcomes: one with stage 2 CKD and the other with stage 5 and hypertension, both on higher dosages of medication to control proteinuria.

Conclusion:
Alport syndrome is a rare condition, but should be suspected in children with family history of CKD, proteinuria and/or hematuria.Family history of CKD should be taken into consideration and prompt an early referral or closer surveillance in a pre-symptomatic phase.

Key words: Alport, hematuria, proteinuria, genetic

SHORT COMMUNICATION - COC 16

LUPUS NEPHRITIS - THE TRANSITION FROM YOUTH TO ADULTHOOD

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Introduction:
Lupus nephritis (LN) in childhood usually presents after the age of 10 years. Female children and adolescents develop LN more commonly. The incidence of clinically evident renal disease is greater at childhood-onset than in adults (82%), usually with proteinuria (nephrotic syndrome in 50%).

Objectives:
We aim to evaluate LN with childhood-onset and the transition to adulthood.

Methods:
Retrospective study of patients being followed by pediatric nephrology with subsequent transition to adult nephrology since 1995 to 2020. Demographic and clinical data were collected and analyzed using IBM SPSS Statistics®.

Results:
Fourteen patients (female:78,6%) with a median age of 26,3 ± 5,2 years (range 21-44) were included. The median age of LN diagnosis was 13,6 years (range 9-17). The presentation and clinical development of LN was mild proteinuria (n=9, 64,3%), acute kidney injury (n=5, 35,7%), nephrotic syndrome (n=5, 35,7%), asymptomatic hematuria (n=4, 28,6%) and nephritic syndrome (n=3, 21,4%). All patients underwent kidney biopsy: diffuse LN (class IV) - 78,6% (n=11), membranous LN (class V) - 14,3% (n=2), and focal LN (class III) - 7,1% (n=1). Induction therapy included methylprednisolone pulses and intravenous cyclophosphamide (n=5, 35,7%) or mycophenolate mofetil (MMF) (n=9, 64,7%). Maintenance treatment consisted of oral glucocorticoids (GC), with azathioprine (n=3, 21,4%), MMF (n=11, 78,6%) or intravenous cyclophosphamide (n=6, 42,9%). A complete remission occurred in all patients. In the transition to adulthood, MMF was discontinued in 3 patients (21,4%) after 17±9 months; currently, 11 patients (78,6%) are on MMF and oral GC. Six patients (42,9%) had renal flares equally distributed in youth or adulthood. None of these patients developed ESRD or died.

Conclusion:
The long-term outcome in this group of children with LN was excellent, with 100% patient and renal survival at a mean follow-up of 11 years. Nevertheless, it is important to notice the high dependence of immunosuppressive agents in adulthood probably related with severe disease at childhood-onset.

Key words: Lupus nephritis, outcomes, treatment, kidney biopsy

SHORT COMMUNICATION - COC 17

BLOOD PRESSURE IN OUR ENVIRONMENT. PEDIATRIC RISK FACTORS

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Introduction:
Arterial Hypertension (AHT) in pediatrics is an independent risk factor for developing as an adult. The American Academy of Pediatrics recommends
screening from the age of 3 in pediatrics check-ups, especially in children predisposed.

Objectives:
To find the prevalence of arterial hypertension in the age groups studied and the relationship with risk factors(RF).

Methods:
We made a prospective observacional study in Lorca´s medical centers. The population included are children who come for check-up at ages 4 and 13 between April and December
2022. We carried out an analysis of BP figures and different risk factors (sex, age, obesity, medical history, medication intake, type of nutrition, screen hours, exercise and stimulating drinks)

Results:
We made 113 determinations(53 of 4 years and 70 of 13 years). In two medical centers this review is carried out by pediatricians and nursery, and in the rest it is carried out only by nursery.
In 23% of the cases, AHT or prehypertension figures are obtained, 14% correspond to men, the majority being 4 years old (53.8%). In 61.5%, a second confirmatory determination is not performed, 21.2% confirm measurement with a manual cuff. Regarding RF, men stand out among the altered figures, despite being the ones who exercise the most(70%), drink up 3 times more stimulating drinks and triple of screening hours than women.

Conclusion:
Children with AHT are usually asymptomatic, the person responsible for screening must know the normal BP values for its correct interpretation. In 61.5%, a second confirmatory measure isn´t taken, so we intend to raise awareness of the importance of correct determination and management of BP from medical centers. The prevention of cardiovascular diseases should start from the pediatric age, favoring healthy lifestyle and limiting the stimulating drinks/screening hours from schools and health centers, since we have observed a clear difference between sexes, ages and BP figures.

Key words: Arterial Hypertension

SHORT COMMUNICATION - COC 18

"INFLUENCE OF BIRTH WEIGHT DISCORDANCE IN TWINS, ON GLOMERULAR FILTRATION RATE AND KIDNEY VOLUME, DURING CHILDHOOD"

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Introduction:
Low birthweight, prematurity and intrauterine growth restriction (IUGR) increased risk of having decreased nephron endowment. Total kidney volumen correlates closely with nephron mass and kidney function (GFR).

Objectives:
To know the influence of IUGR in twins, related to birth weight discordance (BWD), on GFR, TKV and renal resistive index (RRI), during childhood.

Methods:
We studied twins without severe perinatal pathologies with BWD≥14.6% and >20% (BWD-calculation: sum of the joint-weight and its difference between them): serum creatinine (standardized-Jaff) and CysC (standardized-immunonephelometry), GFR (mL/min/1.73m2) assessed by 24-h creatinine clearance and estimated using National Kidney Foundation equations. B-mode kidney ultrasound and KV were performed using ellipsoid´s equation. RRI for every kidney (right-RK, left-LK), as the average of three values pulsed-wave-Doppler. Statistical analysis: comparing means between pairs of twins (Student's t-test, Wilcoxon´s signed-rank-test).

Results:
Age-study (years): 9.3±4.2 (range: 2.6-16.0). Gestational-age (days): 241±14 (range: 198-268).

BWD≥14.6% *(n=46)*	***Large-twin****		***Small-twin****		***p-value***
Birth-weight (g)	2.236±461		1.472±370		p<0.001
Cr (mg/dL)	0.50±0.15		0.55±0.18		p<0.001
CysC (mg/L)	0.79±0.10		0.82±0.10		p=0.054
C_{Cr} *(n=36)*	152±27	(0%)*	141±31	(1.5%)*	p=0.027
Schwartz-IDMS-2009	115±18	(4%)*	104±16	(10%)*	p<0.001

SHORT COMMUNICATION - COC 18

CysC-based-CKiD-2012	103±12	(8.5%)*	99±12	(15.5%)*	p=0.065
Cr-CysC-based-CKiD-2012	105±9	(2.5%)*	97±12	(19.5%)*	p<0.001
TKV (mL/m²)	138±17	(8.5%)ᵇ	132±20	(22%)ᵇ	p=0.023
RRI	*RK*[1]	*LK*[2]	*RK*[1]	*LK*[2]	[1]p=0.095 [2]p=0.559
	0.64±0.03	0.66±0.04	0.66±0.04	0.66±0.04	
BWD>20% *(n=23)*	***Large-twin****		***Small-twin****		***p-value***
Birth-weight (g)	2.187±457		1.301±321		p<0.001
Cr (mg/dL)	0.53±0.19		0.59±0.20		p=0.002
CysC (mg/L)	0.79±0.10		0.85±0.09		p=0.023
C_{Cr} *(n=18)*	143±29	(0%)*	124±28	(2.5%)*	p=0.009
Schwartz-IDMS-2009	112±21	(6.5%)*	100±20	(13%)*	p<0.001
CysC-based-CKiD-2012	102±11	(8.5%)*	96±9	(17.5%)*	p=0.035
Cr-CysC-based-CKiD-2012	102±9	(2%)*	92±8	(28%)*	p<0.001
TKV (mL/m²)	136±16	(13%)ᵇ	126±17	(30%)ᵇ	p=0.029
RRI	*RK*[1]	*LK*[2]	*RK*[1]	*LK*[2]	[1]p=0.951 [2]p=0.578
	0.65±0.04	0.66±0.03	0.66±0.04	0.66±0.05	

Conclusion:
Smaller twins and greater BWD have higher Cr and CysC concentrations, lower GFR, lower TKV, but no differences in RRI. CCr reported the lowest percentage of twins with decreased GFR and Cr-CysC-based-equations the highest.

Key words: Nephron endowment. Total kidney volume. Cystatin C. Twin. Renal clearance. Intrauterine growth restriction. Renal resistive index.

VII CONGRESSO HISPANO-PORTUGUÊS DE NEFROLOGIA PEDIÁTRICA E XLVI CONGRESO ESPAÑOL DE NEFROLOGIA PEDIÁTRICA
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SHORT COMMUNICATION - COC 19

VARIABILITY IN CREATININE SERUM DETERMINATION EVEN AFTER IDMS STANDARDIZATION

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Introduction:
With the aim of reducing variability in creatinine (Cr) measurements, a standardization through calibration procedures by Isotope dilution mass spectrometry (IDMS) was carried out by the manufacturers. As a consequence, pediatric reference intervals for standardized Cr in serum have been published as well as modifications in the equation for eGFR in children (Schwartz- IDMS), data necessary for the staging of CKD.

Objectives:
To compare the results obtained in the measurement of serum creatinine by Beckman- Jaffe and Abbott-Jaffe methods both standardized to IDMS, versus Abbott-enzymatic assay in pediatric samples. To compile the methods used for serum Cr determination in the Spanish laboratories of the pediatric nephrology units.

Methods:
Serum specimens (n=175) from children 15 days-12 years of age with creatinine ≤0.8mg/dL were included. Creatinine measurements by Abbott-Jaffe, Beckman-Jaffe and Abbott-enzymatic methods were compared. Furthermore, we evaluated 819 specimens comparing Abbott-Jaffe and Abbott-enzymatic procedures.
We carried out a survey to find out the methods used in the Spanish laboratories of the different pediatric nephrology units, through PEDIANEF network

Results:

Linear regresion	N	a	b	R^2	p
Jaffee-Beckman-Enzymatic-Abbott	175	-0.02	1.01	0.94	<0.001
Jaffee-Abbott-Enzymatic-Abbott	175	0.25	0.71	0.83	<0.001
Jaffee-Abbott-Jaffee-Beckman	175	0.28	0.66	0.80	<0.001
Jaffee-Abbott-Enzymatic-Abbott	819	0.21	0.67	0.59	<0.001

Equations: $y = a + b*x$

The largest increases in Cr-Jaffe vs Cr- enzymatic are found in the range of Cr<0.30 mg/dL and at the age of 1-24 months with values (mean and median) of 60-64%.
Out of 42 laboratories that responded to the survey, 83% use Jaffe-IDMS: 40%-Roche, 26%-Abbott, 20%-Siemens, 14%-Beckman. Only 17% enzymatic assay

Conclusion:
The standardization of measurement to IDMS does not ensure the homogeneity of creatinine values mainly those measured with the Abbott-Jaffe method that overestimate it, especially with the lowest creatinine levels. In these cases, enzymatic method seems reasonable.
There´s significant variability in the measurement methods used in Spanish hospital laboratories

Key words: Pediatric, Jaffe creatinine, Variability, Enzymatic creatinine, Isotope dilution mass spectrometry

SHORT COMMUNICATION - COC 20

INDICATIONS, FINDINGS AND COMPLICATIONS OF PERCUTANEOUS KIDNEY BIOPSIES IN PEDIATRIC PATIENTS. EXPERIENCE OF A REFERRAL HOSPITAL

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Introduction:
Kidney biopsy is an essential tool for the diagnosis of many renal conditions. It allows not only optimizing and monitoring the treatment but also establishing a prognosis. The ultrasound-guided performance have reduced the complications rate in both native and transplanted kidney biopsies.

Methods:
We performed a retrospective descriptive study of patients <18 years, who underwent a percutaneous kidney biopsy in the Pediatric Nephrology Unit of our institution, between 2007 and 2021. We analyzed demographic and clinical data, description and complications of the procedure and the histological findings. The variables were expressed in total/percentages (qualitative) and median/range (quantitative).

Results:
186 biopsies were performed, 70 in women. The median age was 12.2 years (range 0.8-18.4). One hundred biopsies were performed in native kidneys, 73 in cadaveric grafts and 13 in living-donor grafts. The main indications were steroid-resistant nephrotic syndrome and impaired graft function.
In most cases the 18G needle was used (90.9%) and two shots were given (64.5%). The median number of obtained glomeruli was 17 (range 2 – 73). Conscious sedoanalgesia (midazolam + ketamine) in Pediatric Nephrology ward was performed in 145 cases (78%), with no serious related side effects. Valid material was obtained in 182 cases (97.8%). The main findings were minimal change disease and acute cellular rejection.
After biopsy, 34 cases developed macrohematuria and 3 required blood transfusions. There were 4 urinary retentions, 1 suture dehiscence and 1 transplantectomy. Postbiopsy ultrasound was performed only in 23 cases, which previously have developed clinical complications. There were no fistulas, infections or post-hospital discharge complications.

Conclusion:
Percutaneous kidney biopsy is a safe procedure in pediatric population. Conscious sedoanalgesia in ward setting is safe and valid, avoiding the side effects of general anesthesia. Performing the postbiopsy ultrasound only in selected patients is a valid approach.

Key words: Percutaneous Kidney Biopsy, Children, Procedure, Effectiveness, Complications

POSTER DEFENSE- POD 01

ESTIMATED FILTRATION GLOMERULAR RATE IN ADOLESCENCES:

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Introduction:
The measurement of serum creatinine (Crs) is the most common method to assess glomerular filtration rate (GFR). In the clinical setting, estimation of GFR (eGFR) is recommended. Different formulas are used for adults (Cockcroft–Gault [CG], Modification of Diet in Renal Disease [MDRD], Chronic Kidney Disease Epidemiology Collaboration [CKD-EPI) and for children (Bedside Schwartz 2009) which can be a problem, especially in adolescents.

Objectives:
To assess eGFR in patients 10-18 years of age using paediatric and adult formulas

Methods:
Crs values (Jaffe traced to IDMS) from 10-18 years old patients performed during 2022 were obtained from the database of the central lab. We search weight/height (+/- 6 months of the date of the blood test) in the electronic medical record of a subset of 15 patients for each age/sex (5 lowest, 5 highest and 5 patients with a Crs in the mean for the age group). eGFR was calculated using Schwartz and CG formulas for those patients with height/weight available and CKD-EPI/MDRD for all patients.

Results:
From 5782 patients we had 8024 Crs results. eGFR was <90ml/min/1.73m2 in 377 (4.7%) samples according to MDRD and in 90 (1.1%) when using CKD-epi.
We could calculate Schwartz and CG in 186 patients (80% of patients ≤14 years of age, 55% of the patients >14 y). eGFR was <90ml/min/1.73m2 in 72 by Schwartz and in 62 by CKD-epi. The average difference between eGFR by Schwartz and CKD was 23 for the whole population and 13 for the group 17-18 y. In this former group, 8/30 patients would be in a different stage.

Conclusion:
The lack of anthropometric data to assess eGFR is frequent.
Significant differences between Schwartz and CKD-epi results ,even at 17-18y
Transfer from paediatric to adult physician could cause a change in stage of kidney disease just because of the methods to assess eFG

Key words: Glomerular filtration rate, Adolescents

POSTER DEFENSE- POD 02

VARIABILITY IN THE ESTIMATION OF GLOMERULAR FILTRATION RATE BASED ON CREATININE AND CYSTATIN C IN PEDIATRIC KIDNEY TRANSPLANT PATIENTS - IMPACT OF BODY MASS INDEX AND PROTEINURIA

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Introduction:
The estimation of the Glomerular Filtration Rate (eGFR) is essential in the evaluation of kidney function in Pediatric Kidney Transplant (P-KT) patients.

Objectives:
With this study, we intended to compare eGFR by different equations using serum creatinine(Cr) and/or Cystatin C(CysC) in P-KT patients and to evaluate the relationship with Body Mass Index(BMI), and proteinuria.

Methods:
We conducted a retrospective study of P-KT patients in a Pediatric Nephrology unit, with stable graft function for more than 6 months post-P-KT; eGFR was calculated by CKiDCr(Schwartz bedside), CKiD-CysC, Schwartz combined(Cr/CysC), Zapitelli-CysC, and Zapitelli combined(Cr/CysC) formulas.

Results:
Forty-five patients were included, with median age 14.6(11.2-16.7) years, 69%(n=31) male and median BMI 20.7(17.3-23.6)kg/m2. Seven patients(15.6%) were obese(BMI zscore>+2). Proteinuria occurred in 40% (n=18) of patients. The median eGFR(ml/min/1.73m2) was: CKiD-Cr 61.5(51.2-72.3); CKiD-CysC 49.8(41.0-57.3), CKiD-Cr/CysC 50.0(42.3-60.2); Zapitelli-CysC 51.7(41.8-60.4) and Zapitelli-Cr/CysC 53.6(43.7-66.3). Correlations between eGFR by all formulas proved to be strong and statistically significant, especially between combined formulas(r=0.98). When compared, CysC-based and combined formulas showed statistically lower median eGFR than CKiD-Cr ($p<0.01$), and this difference remained statistically significant in patients with proteinuria. There were no significant differences between eGFR by the different formulas in obese patients. When comparing the CKiD-Cr vs CKiD-CysC and CKiDCr/CysC equations, 53.3%(n=24) and 46.7%(n=21) of patients, respectively, had discordant estimates of CKD stage, with a higher stage estimated by CKiD-CysC and CKiD-Cr/CysC equations.

Conclusion:
CysC-based and combined formulas showed statistically lower medians of eGFR in P-KT patients compared with CKiD-Cr, suggesting that equations based solely on Cr may overestimate GFR. In obese patients, this difference was not statistically significant. In about half of the patients, there was discordance between CKD stages determined by eGFR by the different formulas, with a higher stage estimated by CysC-based and combined formulas.

Key words: Pediatric Kidney Transplant, Estimation of Glomerular Filtration Rate, Body Mass Index, Proteinuria

POSTER DEFENSE- POD 03

NEAR INFRARED SPECTROSCOPY AS CONTINUOUS REAL-TIME MONITORING FOR KIDNEY GRAFT PERFUSION IN THE FIRST POSTRANSPLANT WEEK

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Introduction:
Near Infrared Spectroscopy (NIRS) is a non-invasive technique employed to detect regional hemoglobin oxygen perfusion (rS02). It has the capacity to cover an area of 1-2 cm3 and penetrate up to 3-4 cm beneath the skin's sensor placement. NIRS technology does not rely on pulse or temperature variations or operator intervention. This methodology may prove valuable for monitoring pediatric renal transplant because of the superficial position of the allograft and the thinness of the superficial tissues. By continuously monitoring real-time assessment of graft perfusion status, healthcare providers can take prompt action when blood flow complications or ischemia arise - both known factors capable of contributing over time towards eventual renal dysfunction or loss if left unaddressed.

Results:
An 11-year-old patient suffering from chronic kidney disease caused by nephronophthisis (homozygous NPHP1 deletion), exhibiting symptoms of polyuria, normal blood pressure, BMI 18 kg/m2 underwent a preemptive deceased donor kidney transplantation. The surgical procedure was followed by continuous measurement of regional oxygen saturation (rS02) of the kidney graft using an INVOS 5100 device (Medtronic) over seven days, which did not lead to any adverse effects. The patient showed good progress with rapid reduction in plasma creatinine levels along with appropriate vascular resistance indices observed through Doppler ultrasound. Analysis based on the NIRS trend indicated lower rS02 values at around 75% during the first day following transplantation and subsequent readings stable within the range of 85-90% throughout the rest of study duration.

Conclusion:
RS02 provides continuous measurement of renal graft perfusion in selected patients, which could be an additional tool to evaluate vascular alterations in a critical phase where early detection is crucial. Following this case, we have initiated a prospective study that has already included 15 kidney transplant recipients and is currently in the data analysis phase, with results to be presented soon.

Key words: Near Infrared Spectroscopy, perfusion, kidney transplantation.

POSTER DEFENSE- POD 04

URINARY CITRATE IN SINGLE SPOT SAMPLES: PAEDIATRIC REFERENCE VALUES

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Introduction:
Hypocitraturia is a condition associated to recurrent kidney stones, nephrocalcinosis and distal renal tubular acidosis, among others. To make the diagnosis, citraturia is usually measured in 24-hour urine, which is difficult to collect. The citrate-to-creatinine ratio (Cit/Cr) in spot urine can be a useful, simpler parameter to diagnose hypocitraturia. However, its application in clinical practice is hindered by the lack of robust reference values in paediatrics. A Cit/Cr > 400 mg/g is commonly considered to rule out hypocitraturia, but various cut-off points between 250 and 420 mg/g have also been proposed, most of these values being derived from 24-hour samples.

Objectives:
To establish paediatric reference values of urinary citrate (expressed as Cit/Cr) in single spot urine samples.

Methods:
The first morning urine sample from healthy continent children aged from 4 to 14 years was collected in tubes without additives. After centrifugation, samples were frozen at -80°C until analysis. Results were expressed as median value and range. Reference values were determined by the non-parametric method (as 2.5th and 97.5th percentiles), and 90 % confidence intervals were indicated for each limit.

Results:
A total of 154 subjects were recruited (44.8% female, median age 8.5 years). Median Cit/Cr was 474 mg/g (range 50 – 1626). No differences were found between males and females. Reference values ranged from 113 mg/g (50 – 177) to 1341 mg/g (1086 – 1626). 38.9% of the subjects had a Cit/Cr < 400 mg/g.

Conclusion:
Our findings suggest that the threshold values used for the diagnosis of hypocitraturia may be excessively high, which could give rise to a high rate of false positives, and point out the need for more studies to establish a lower cut-off point to determine the condition of hypocitraturia.

Key words: urinary citrate, single spot sample, citrate-to-creatinine ratio, hypocitraturia

POSTER DEFENSE- POD 05

TRAINING IN PAEDIATRIC NEPHROLOGY ACROSS SPAIN, WORKING CONDITIONS AND OPPORTUNITIES TO RESEARCH: A SURVEY OF THE YPNN-AENP NATIONAL SOCIETY NETWORK

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Objectives:
The aim of our study was to describe the training and the working situation of paediatric nephrologists in Spain.

Methods:
Multicentre cross-sectional study through a survey of paediatricians working in Paediatric Nephrology in hospitals in Spain in January to February 2023.

Results:
60 paediatricians answered the survey. 72.9% are women. 63.3% are working in third-level hospital hospitals, 26.7% in second-level hospital hospitals 6,7%(4) in first-level hospitals.
Median length of mandatory training in Paediatric Nephrology for a general paediatrician during their residency was 2(0-6) months. For those subspecialised in Paediatric Nephrolog median training lasted 12(1-48) months.
45.8% held a permanent position. 72.9% of participants declared that job openings in Paediatric Nephrology in Spain should be broadly advertised.
61.7% are or have been enrolled in a PhD programme, while 26.7% completed it. 86.4% conduct research routinely but more than 58.3% do it in their free time. 91.5% reckoned that Paediatric Nephrology should be considered a paediatric subspeciality. Most of them belong to AENP 96.9% as full members 62.7% and 33.9% associate members, whereas only one third belong to ESPN or IPNA.

Conclusion:
There are important discrepancies in training programs in Paediatric Nephrology. Strategies to support the recognition and to standardise training should be implemented.
Better working conditions for paediatric nephrologists are necessary to promote research and networking.

Key words: Paediatric Nephrology, Medical Education, Spain, Working Conditions

POSTER DEFENSE- POD 06

RIM ÚNICO CONGÉNITO FUNCIONANTE NA CRIANÇA – SEGUIMENTO E EVOLUÇÃO

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Introduction:
O rim único congénito funcionante (RUFc) é um defeito congénito comum, com uma prevalência de 1/1500 nascimentos, sendo mais frequente no género masculino. Na maioria dos casos representa uma agenesia renal unilateral ou um rim displásico multiquístico.

Objectives:
Analisar as caraterísticas e evolução da população pediátrica com RUFc.

Methods:
Análise retrospetiva dos processos clínicos das crianças com RUFc, seguidas na consulta de Pediatria – Patologia Renal de um hospital de nível II entre 2003 e 2022. Foram colhidos dados demográficos, clínicos e analíticos, com posterior análise estatística.

Results:
A amostra inicial incluiu 31 crianças, das quais 6 foram excluídas por perda de seguimento. Das 25 incluídas, 60% (n=15) eram do género masculino.
Identificaram-se 19 casos de agenesia renal (dos quais 16 com diagnóstico pré-natal e 12 agenesias renais esquerdas) e 7 casos de rim displásico multiquístico.
Em 16% (n=4) dos doentes identificou-se alguma malformação associada: 2 casos com útero didelfo e 1 caso de Síndrome de Zinner (com agenesia renal), 1 caso com refluxo vesicoureteral (com rim displásico multiquístico).
Adicionalmente, a ecografia renovesical revelou: rim contralateral vicariante (n=20), discreta ectasia do bacinete (n=7), discreta ectasia do ureter (n=1) e atenuação da diferenciação parenquimo-sinusal (n=1).
A cintigrafia renal com ácido dimercaptosuccínico revelou cicatrizes renais em 6 crianças.
Foi instituída profilaxia antibiótica em 23 casos (19 ao nascimento), com duração média de 5,7 meses.
Na evolução verificaram-se: infeção do trato urinário de repetição (2 crianças) e tensão arterial elevada (numa criança com 10 anos). Todos apresentam função renal e microalbuminuria normais.
O tempo médio de seguimento foi de 7,6 anos.

Conclusion:
O RUFc é habitualmente uma condição isolada com evolução benigna. No entanto, pode estar associado a outras malformações e cursar com complicações que podem conduzir a doença renal crónica, pelo que deve ser mantida vigilância e proteção da função renal durante toda a vida.

Key words: Rim único, Agenesia renal, Rim displásico multiquístico

POSTER DEFENSE- POD 07

POSTERIOR URETHRAL VALVES: CASUISTRY OF THE LAST 20 YEARS IN A LEVEL II HOSPITAL

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Introduction:
Posterior urethral valves (PUV) are one of the main causes of urinary tract obstruction in the pediatric age. They have an estimated incidence between 1 in 5000 to 1 in 8000 births, and approximately 30% of patients with this anomaly progress to chronic kidney disease (CKD). With recent technological advances, the diagnosis of PUV is made earlier, in the prenatal period, which has had an important influence in terms of prognosis.

Objectives:
The aim of this study was the clinical and evolutionary characterization of pediatric patients diagnosed with PUV.

Methods:
Retrospective and descriptive study of 6 patients, diagnosed with PUV in the last 20 years, followed in pediatric nephrology consultations in a level II hospital. By consulting the clinical process of each patient, the following variables were analyzed: clinical presentation at diagnosis, blood work analysis, imaging data, with focus on timing of therapy, clinical evolution and current follow-up.

Results:
Of the 6 patients studied, the diagnosis of 4 of them was suggested by prenatal ultrasound. In the first postnatal renal and bladder ultrasound, 5 patients had bilateral hydroureteronephrosis, and 2 of them had bladder thickening. In 5 patients, the diagnosis was confirmed by retrograde cystography, and 3 of them had concomitant vesicoureteral reflux. In all patients, the initial treatment was resection of the valves by urethrocystoscopy, with a median age of the first intervention of 1 month. One of the patients evolved to terminal CKD at 15 years of age.

Conclusion:
Despite the decrease in mortality associated with earlier diagnosis of PUV, it continues to be an important cause of CKD in children. The presence of secondary complications related to this pathology imply a continued monitoring of these patients, in order to reduce the morbidity of the disease.

Key words: posterior urethral valves, urethrocystoscopy, hydroureteronephrosis, chronic kidney disease

POSTER DEFENSE- POD 08

MICROBIOLOGICAL COMPARISON OF URINE COLLECTION METHODS IN A PEDIATRIC EMERGENCY DEPARTMENT: 2-YEAR ANALYSIS

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Introduction:
Urinary tract infection (UTI) is a common cause of admissions in the pediatric emergency department (PED). Urine sample collection isn't always easy nor quick, and when necessary, invasive methods should be sensitive and specific.

Objectives:
To evaluate contamination rates of urine cultures from different collection methods.

Methods:
Retrospective observational study with analysis of urine cultures in a PED from July 2020-June 2022. Patients' age, gender, previous UTIs and CAKUT were collected. Urine collection methods, culture results, isolated microorganism and respective drug resistance were evaluated. When non specified, sphincter continence was assumed from 48 months of age. Statistical analysis was performed using SPSS V28.

Results:
3206 urine cultures were collected. Of these, 26.6% had no information about the collection method, and were excluded (N=2354). Mean age was 5.5 years, with 65% urine cultures from female individuals. 26.6% were from urine collection bag (UCB), 19.3% urethral catheterization (UC), 53.9% midstream and 6 samples from suprapubic aspiration (SA). Overall positivity rate of urine cultures was 27.4%, and 33.6% were contaminated. E.coli was the predominant bacteria (N=450). 1.5% of urine cultures was positive for Multidrug-resistant agents. Contamination rate in each method was 50.7% in UCB, 8.8% in UC, 34.3% in midstream and 0% in SA. Fisher's test showed statistical significance of these differences ($p<0.001$). When age-adjusted, compared to mid-stream (the most used method), UCB has bigger odds of contamination (OR 3,39, CI 2,55-4,51, $p<0,001$) and UC has less (OR 0,31, CI 0,21-0,46, $p<0,001$).

Conclusion:
Contamination rates difference between collection methods is statistically significant ($p<0.001$), haaving UCB bigger odds of contamination. However, contamination rate in the midstream is not negligible (34.3%), being this the most overall used method (N=1269). Through this analysis, institution's UTI protocol and methods for valorization of urine cultures were reviewed. Currently a study is underway to analyze these changes' impact on contamination rates.

Key words: urine cultures, urinary tract infections, contamination, collecting methods

POSTER DEFENSE- POD 09

PADRÃO DA RESISTÊNCIA ANTIMICROBIANA NAS INFEÇÕES DO TRATO URINÁRIO NOS PRIMEIROS 2 ANOS DE VIDA: A EXPERIÊNCIA DE UM HOSPITAL DE NÍVEL III

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Introduction:
A infeção do trato urinário (ITU) é uma das doenças infeciosas mais frequentes em pediatria. O uso inadequado de antibióticos para tratar estas infeções tem levado a um aumento da resistência dos microrganismos.

Objectives:
Determinar a prevalência dos principais microrganismos causadores de ITU e o padrão de resistência aos principais antibióticos utilizados empiricamente no tratamento ambulatório.

Methods:
Estudo retrospetivo descritivo realizado num hospital de nivel III, de doentes com idade inferior a 2 anos, com o primeiro episódio de ITU entre janeiro de 2018 e janeiro de 2022. As amostras foram obtidas através de algaliação e foi utilizado o ponto de corte >104-105 unidades formadoras de colónias/mL para definição de caso. Foi realizada análise estatística dos dados demográficos e das uroculturas.

Results:
Foram analisados 886 doentes (67% do sexo feminino), com idade média de 7 meses. O microrganismo mais frequentemente isolado foi a Escherichia coli (62%), seguida do Proteus mirabilis (9.7%), Klebsiella pneumoniae (4%) e Enterococcus faecalis (2%). Analisando o padrão de resistências aos antibióticos: na Escherichia coli a resistência foi maior para amoxicilina/ácido clavulânico (31.5%), seguida da cefuroxima (3%); no Proteus mirabilis também se observou maior resistência à amoxicilina/ácido clavulânico (7%) e não se verificou nenhum caso de resistência à cefuroxima; na Klebsiella pneumoniae também se observou maior resistência à amoxicilina/ácido clavulânico (25%) do que à cefuroxima (22%). O microrganismo com maior prevalência de multirresistência foi a Klebsiella pneumoniae (17%).

Conclusion:
Os autores concluem que o agente etiológico mais prevalente na ITU demonstra uma elevada resistência ao antibiótico empírico mais utilizado no seu tratamento e um baixo nível de resistência à cefuroxima. Nos restantes microrganismos também a resistência à amoxicilina/ácido clavulânico foi superior à da cefuroxima. Destaca-se a importância de mais estudos, idealmente a nível multicêntrico, com o objetivo de atualizar as orientações terapêuticas na abordagem da ITU em idade pediátrica.

Key words: infeção do trato urinário, Escherichia coli

POSTER DEFENSE- POD 10

TRATAMIENTO CON CORTICOIDES EN LA PIELONEFRITIS AGUDA PARA EVITAR CICATRICES: REVISIÓN SITEMÁTICA Y METANANALISIS

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Introduction:
La pielonefritis aguda en niños puede ocasionar una cicatriz renal permanente. El tratamiento con antibióticos no se ha mostrado suficientemente eficaz para erradicar la aparición de cicatrices, por lo que se ha postulado la terapia coadyuvante con corticoides para disminuir el número de cicatrices
Objectivos
Evaluar la eficacia de la terapia con corticoides en niños con infección urinaria febril para la prevenir la aparición de cicatrices renales

Methods:
Revisión sistemática y metaanálisis. Población de estudio: niños con primer episodio de ITU febril. Intervención: administración de corticoides. Comparación: placebo o no intervención. Resultado principal: aparición de cicatrices. Estrategia de búsqueda de ensayos clínicos randomizados en PubMed, Embase y Central de la Cochrane

Results:
Se incluyeron cinco ensayos clínicos aleatorios (464 niños), con un diseño heterogéneo. En cuatro con datos confirmados de pielonefritis por DMSA en fase aguda, y en un quinto con niños con ITU febril. Uno de ellos analiza niños con pielonefritis grave, con gran daño en el DMSA inicial. Los pacientes recibieron dexametasona o prednisolona durante un periodo corto (2-4 días). Analizados conjuntamente los corticosteroides no son eficaces para reducir el riesgo de cicatrización renal en comparación con placebo. Se realizó metaanálisis con un modelo de efectos aleatorios, según el método de Mantel Haenszel resultando una OR: 0,59 (IC 95%: 0,32-1,09). El grado de heterogeneidad fue moderado: Q 2,95, I2 12%. El análisis por subgrupos no mostró tampoco efectos protectores significativos. En los niños con pielonefritis confirmada y bajo sesgo de selección: OR 0.95 (IC95% 0,2 a 2,9) y en el grupo de ITU febril con importante sesgo de selección: OR 0.46 (IC 95% 0,03 a 7, 64).

Conclusion:
Existe evidencia de calidad moderada que sugiere que la terapia adyuvante con corticosteroides de corta duración, junto con terapia antibiótica, en niños con ITU febril aguda no previene de la formación de cicatrización renal

Key words: pielonefritis, corticoides, cicatriz renal

POSTER DEFENSE- POD 11

PREDITORES DE DOENÇA RENAL DIABÉTICA EM IDADE PEDIÁTRICA NUM HOSPITAL DE GRUPO I

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Introduction:
A doença renal diabética (DRD) é uma das principais complicações da diabetes mellitus (DM), cujo rastreio é realizado através da deteção de albuminúria moderadamente aumentada (microalbuminúria), um marcador precoce de doença. O rastreio inicia-se aos 11 anos ou início da puberdade após 2-5 anos de DM1.

Objectives:
Determinar a prevalência de microalbuminúria e outros fatores de risco associados a DRD em adolescentes e adultos jovens com diagnóstico de DM1 em idade pediátrica num hospital grupo I.

Methods:
Estudo retrospetivo descritivo dos dados clínicos e analíticos dos doentes <24 anos acompanhados em consulta entre 2017-2023. Considerou-se microalbuminúria excreção de albumina >30mg/L em amostra isolada de urina ou rácio albumina/creatinina urinária (RAC) >30mg/g, em ≥ 2 avaliações.

Results:
Selecionados 54 doentes (31 do sexo feminino) com critério para rastreio. Idade média 15,5 (±4,7) anos; duração média de doença 7,6 (±4,9) anos; 36 sob sistema de perfusão subcutânea de insulina e 18 sob múltiplas administrações de insulina. HbA1c média 7±3,7%; número médio de internamentos por DM desde o diagnóstico 1,2. Dezanove doentes apresentavam fatores de risco para DRD, nomeadamente excesso de peso (8), obesidade (7), dislipidemia (4), hipertensão arterial (3) e tabagismo (3). Foi identificada microalbuminúria em 2 doentes sem outros fatores de risco, com 5 e 9 anos de doença, valor médio de HbA1c 7,5% e 9,4%. Dois doentes apresentavam RAC>30mg/g, após 9 e 12 anos de doença, com HbA1c média 11,6% e 8,6%, um deles com obesidade. Nenhum doente apresentava diminuição da taxa de filtração glomerular. Um doente medicado com dapagliflozina.

Conclusion:
A prevalência de microalbuminúria encontrada foi 7%, sendo concordante com a literatura. Apenas um doente tinha outro fator de risco para DRD além de HbA1c ≥ 7,5%. Este trabalho pretende alertar para a importância do rastreio de DRD, vigilância dos fatores de risco e instituição precoce de medidas renoprotetoras com impacto no prognóstico.

Key words: Albuminúria, Diabetes Mellitus, Nefropatias Diabéticas

POSTER DEFENSE- POD 12

POST-STREPTOCOCCAL GLOMERULONEPHRITIS: IS IT INCREASING AFTER COVID-19?

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Introduction:
Acute post-streptococcal glomerulonephritis (APSGN) is a prototype of post-infectious glomerulonephritis associated with a previous skin or throat infection by group A streptococcus (Streptococcus pyogenes). It usually presents as nephritic syndrome: hematuria, proteinuria, edema, hypertension (AHT), and oliguria.
Recently, due to the removal of masks, a significant increase in infections caused by this bacterium has been detected, with a cluster of APSGN cases reported during December - February (2022-2023).

Objectives:
To analyze the cases of APSGN detected and to raise awareness of this recent increase.

Methods:
Retrospective descriptive study conducted between 2020-2023 on APSGN cases. Analyzed epidemiology, clinical-analytical variables, treatment, and evolution.

Results:
11 cases were detected, predominantly in males (63.6%), median age of 5 years. Only one case was detected each year in 2020 and 2021. However, between December and February (2022-2023), a total of 9 cases were detected.
6 were associated with a previous S.pyogenes infection, while the other 5 had registered infection in their family. 6 maintained GFR > 90 mL/min/1.73 m2 at their debut, while the minimum recorded was 62 mL/min/1.73 m2. All patients presented elevated ASLO and decreased C3 at admission. In one case, a concomitant decrease of C4 was detected, without any repercussions on renal function. Only 2 developed sustained AHT (above P99 for their age), requiring treatment with furosemide. 7 developed proteinuria in the nephrotic range without hypoalbuminemia and 9 patients presented macrohematuria at the time of consultation.
No patient required extrarenal clearance techniques. All patients were followed up in the Nephrology department, with subsequent normalization of renal function and complement.

Conclusion:
In all patients presenting with symptoms of hematuria, edema, or oliguria, we should inquire about previous history of S. pyogenes infection.
When nephritic syndrome is suspected, we must have a proactive attitude in the early detection of APSGN to avoid complications, monitoring diuresis and blood pressure.

Key words: Acute post-streptococcal glomerulonephritis, nephritic syndrome, glomerular hematuria

POSTER DEFENSE- POD 13

SUCCESSFUL REPLACEMENT OF ECULIZUMAB BY RAVULIZUMAB AND ITS THE NOVO USE FOR AHUS TREATMENT

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Introduction:
Ravulizumab is a new long lasting humanized monoclonal antibody C5 blocker engineered from eculizumab and approved for treatment of aHUS. Ravulizumab allows extending intervals between IV infusions every 4–8 weeks based on patient weight. We describe our case series of aHUS patients treated with Ravulizumab.

Methods:
6 children (4 boys) with aHUS onset at age 3.6 years (0.75-11.2) were treated with ravulizumab. Complement genes pathogenic variants were observed in 5, and one was a kidney transplant recipient.

Results:
2 girls with new onset aHUS received ravulizumab as first treatment achieving complete response with hematologic remission and renal function recovery. After 5.2 years they are free of disease activity preserving normal eGFR (111 & 118). Further, in 4 boys treated with eculizumab for 5.5 years (0.7-8.9), eculizumab was successfully and uneventfully replaced by ravulizumab. All patients remain in remission with normal eGFR (129,107,125,88). Further, patients and families refer huge improvement in quality of life after ravulizumab initiation.

Conclusion:
Ravulizumab is a new long lasting C5 blocker for treatment of aHUS as first option or by replacing eculizumab successfully and uneventfully.

Key words: ravulizumab, eculizumab, atypical hemolytic uremic syndrome

POSTER DEFENSE- POD 14

RAVULIZUMAB "DE NOVO" IN PEDIATRIC PATIENTS WITH ATYPICAL HEMOLITIC UREMIC SYNDROME (AHUS): FIRST WORLDWIDE CASES

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Objectives:
Ravulizumab is a long-acting C5 inhibitor that has recently demonstrated its effectiveness for the control of hemolytic uremic syndrome compared to eculizumab,allowing average annual infusion times (up to 70% less).There´s still no evidence in the literature of naïve treatment with this drug in pediatrics

Methods:
Present the first two pediatric cases worldwide using "de novo" Ravulizumab (in the onset of the disease and post-kidney transplant)

Results:
13-year-old girl with history of bloody stools,vomiting and compromise of consciousness with TMA,AKI III evolves to anuria and convulsive episode requiring corticosteroid boluses,6 plasmapheresis sessions and 4 intermittent hemodialysis.Normal ADAMTS-13,negative direct Coombs and decreased complement.Due to persistent TMA and requirement of renal replacement therapy (RRT),ravulizumab was started with a loading dose (2400mg)and a second one after 2 weeks.The need for RRT ceased with improvement of hemolysis and renal function.Genetic:CFHR3-CFHR1 deletion.Case 2:7-year-old girl in chronic hemodialysis secondary to aHUS (CD46 mutation)was admitted for kidney transplant from a living donor.Low-intermediate immunological risk and high CMV infectious risk.Induction treatment:Basiliximab,tacrolimus,mycophenolate and steroids.First dose of Ravulizumab was infused the day before transplantation (900mg),well tolerated.The patient has had a favorable evolution of renal function with normal creatinine value at discharge.Protein/Creatinine urine ratio increased to a maximum of 8mg/mg (negative DSA levels).The option of renal biopsy was discarted due to a decrease in proteinuria.She received second dose after 2 weeks,remain stable with no data on recurrence of her underlying disease today

Conclusion:
Ravulizumab was satisfactory both in the acute phase of the disease and in the immediate post-transplantation. In the first case we observed a functional recovery from the first dose with no notable adverse effects up today, as in post-transplant patient, maintaining a good control of TMA despite more spaced dosing (8 weeks). The inclusion of this drug in the therapeutic arsenal opens a new safe treatment route in pediatric patients with aHUS

Key words: aHUS, Ravulizumab, TMA, Kidney Transplant

POSTER DEFENSE- POD 15

ALPORT SYNDROME: CHANGES AT DIAGNOSIS

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Introduction
Alport syndrome is an inherited collagenopathy with renal, hearing and visual involvement, and it usually presents with haematuria and proteinuria. Currently, diagnosis by biopsy has been replaced by genetic diagnosis.

Objectives:
Our aim is to study the demographic, clinical and genetic features of patients with Alport syndrome under follow-up in paediatric nephrology in two hospitals.

Methods:
Twenty-two patients were included, diagnosed between 2010 and 2023. Demographic, clinical and genetic variables were collected from medical records.

Results:
Eleven (50%) were male. Median age of symptoms onset was 3.8 years (IQR 2-7.5 years) and median age of diagnosis delay was 4.5 years (IQR 2-9.1 years). The initial symptom of consultation was: macrohaematuria (10/22), persistent microhaematuria (4/22), proteinuria (2/22) and positive family background (5/22; all of them were affected by haematuria with/without proteinuria afterwards).
One patient progressed to chronic kidney disease at the age of 15 years, and required dialysis and kidney transplant. Four (18%) had sensorineural hearing loss and one (4.5) had anterior lenticonus.
Diagnosis was confirmed by genetic study in 20 patients: 12 with COL4A5 mutation (X-linked dominant inheritance), 4 with COL4A4 (2 with autosomal recessive inheritance, AR, and 2 with autosomal dominant inheritance, AD), 2 with COL4A3 (both AD), 1 with COL4A3-COL4A4 (AD with possible digenic effect) and 1 with COL4A3-DYNC2H1 (AD). Two patients are expecting genetic confirmation (both families with COL4A5 mutation).
Six patients underwent renal biopsy before genetic studies. Two of them needed a second biopsy because the diagnosis of IgA nephropathy differed from the clinical course.
Eight patients were patient zero, from which the mutation was detected in four families.

Conclusion:
Renal biopsy is an invasive test which might not be conclusive. On the other hand, genetic studies have become key for diagnosis because of clinical and genetic variability. Renal biopsy might be dispensable/non-essential.

Key words: Alport syndrome, renal biopsy, genetics

POSTER DEFENSE- POD 16

NOVEL HYPOKALAEMIC TUBULOPATHY DUE TO MUTATIONS IN THE POTASSIUM CHANNEL KIR5.1 (KCNJ16 GENE)

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Introduction:
KCNJ16 encodes basolateral potassium channel Kir5.1 forming heteromeric channels with Kir4.2 (KCNJ15) at the proximal tubule and with Kir4.1 (KCNJ10) distally. Biallelic loss-of-function variants in KCNJ16 cause a novel tubulopathy with hipokalemia, salt wasting, disturbance acid-base homeostasis and sensorineural deafness. We present the case of a patient with severe hypokalemia due to a mutation in this gene.

Results:
A 21-month-old Senegalese girl with consanguineous parents presented with failure to thrive since 9 months of age (from p3-10 in weight and p25-50 in height to <p1). Clinically she began with muscle weakness, and hypokalemia(1.3mEq/L) associated with electrocardiographic alterations was detected. Initially, intravenous potassium chloride therapy was started, followed by oral supplementation, normalizing electrocardiographic alterations. Polyuria was evidenced and arterial blood pressures were normal. The laboratory findings showed elevated EFK and GTTK, increased renin and aldosterone, and hypocalciuria. All other ions and gasometry parameters were normal. Neither ultrasound nor abdominal MRI scan alterations were identified. The genetic analysis demonstrated two homozygous nonsense variants (p.Lys48 and p.Tyr57) in the KCNJ16 gene.
She was discharged from the hospital with oral potassium chloride (6meq/Kg/day), and salt and dextrinomaltase supplementation in the diet. Plasma potassium levels were maintained at 2.5-3 meq/L. Indomethacin was added subsequently as an attempt to improve polyuria and potassium citrate after the onset of metabolic acidosis. Weight and height improved significantly (currently p3 and p25, respectively) and psychomotor development was normal at all times. The auditory truncal evoked potentials were normal.

Conclusion:
Tubulopathy due to mutation in KCNJ16 has been recently discovered and there are few cases described in the literature. Clinically it presents with failure to thrive, metabolic acidosis/alkalosis, polyuria/polydipsia, salt wasting, hyperaldosteronism, hypokalemia and hypocalciuria. During their evolution they develop sensorineural hearing loss and glomerular function remains intact. This mutation should be included in the differential diagnosis of patients with hypokalemic tubulopathy.

Key words: hipokalemia, tubulopathy

POSTER DEFENSE- POD 17

HNF4A MUTATION: HYPERINSULINISM AND/OR LATE-ONSET DIABETES ASSOCIATED WITH RARE AUTOSOMAL DOMI-NANT FANCONI SYNDROME

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Introduction:
Mutations in the HNF4A gene are commonly associated with neonatal hyperinsulinism (HI) and maturity-onset diabetes of the young (MODY). Recently. p.R63W mutation specifically linked to a rare autosomal dominant Fanconi syndrome (FS) and liver dysfunction has been described.

Objectives:
We present two patients with FS and HI caused by HNF4A mutations.

Results:
Case 1: A full-term large-for-gestational age boy developed hypoglycaemia due congenital HI. He was treated with diazoxide until the age of three months. At six months of age, failure to thrive, hepatomegaly and elevated transaminases were detected. At 18 months of age, physical exam and X-ray findings were typical of rickets Complementary tests revealed: normal eGFR, nongap metabolic acidosis with acidified urine, glycosuria, generalised aminoaciduria and tubular proteinuria in addition to hypophosphatemia of renal origin (TRP 72%), hypercalciuria and carnitine deficiency. 25OHD was normal with supplementation, PTH and alkaline phosphatase were elevated.
Case 2: A 3-year-old girl with a history of benign myoclonus of infancy and genu varum presented with afebrile tonic-clonic seizures, prompting a metabolic study. Hypoglycemia secondary to HI was found , requiring diazoxide to the present day, as well as generalized hyperaminoaciduria. Further tests revealed chronic kidney disease (CKD) (eGFR 70 ml/min/1.73m2), nephrocalcinosis and FS features. X-rays also demonstrated rickets. No liver abnormalities were observed.
In both cases, genetic testing detected the pathogenic variant p.R63W (c187C>T) in HFN4A.
After proper metabolic and Fanconi treatment, both children showed catch-up growth and rickets healing. Case 1 progressed to CKD (8 years-old, eGFR 35 ml/min/1.73m2), case 2 (5 years-old) remained estable.

Conclusion:
Early identification of HNF4A mutations will improve earlier and proper management of FS and HI. Close long-term follow-up is needed to monitor CKD progression and potential MODY onset. Family counselling is required.

Key words: HNF4A, Fanconi syndrome, hyperinsulinism, diabetes

POSTER DEFENSE- POD 18

TREATMENT OF PRIMARY HYPEROXALURIA TYPE 1 WITH LUMASIRAN: A CASE SERIE

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Introduction:
Primary hyperoxaluria (PH) is an ultra-rare genetic disorder caused by an inherited metabolic disorder of glyoxylate. This leads to excessive production of oxalate, which is eliminated through the kidneys. Accumulation of oxalate can result in kidney stones, nephrocalcinosis, and even end-stage renal disease (ESRD). PH can present in various forms, from urinary tract infections and hematuria to ESRD or recurrence in a kidney transplant.

Objectives:
The objective of this study is to describe our experience in treating primary hyperoxaluria type 1 (homocigosis p. I244T) with Lumasiran, a synthetic RNA interference therapy.

Methods:
We describe three cases, and their follow up.

Results:
The first a 7-year-old boy with PH1 who had chronic kidney disease and nephrocalcinosis at diagnosis. After starting Lumasiran treatment, his oxalate levels decreased by 70%, and after two years of treatment, his condition remained stable with a 30% reduction in oxalate levels.
The second case was a 3-year-old girl with extensive nephrocalcinosis but normal kidney function. She had a moderate response to treatment and achieved a 42% reduction in oxalate levels during the induction phase of Lumasiran treatment. After six months of maintenance therapy, her oxalate levels decreased by 55%.
The third case was an 11-month-old infant with extensive nephrocalcinosis and normal kidney function. She was asymptomatic at diagnosis and had a moderate response to treatment with Lumasiran. During the induction phase, her oxalate levels decreased by 91%. After six months of maintenance therapy, her oxalate levels decreased by 94%.

Conclusion:
We suggest that new therapeutic strategies like Lumasiran are a promising option for PH patients who previously lacked effective treatments. Our experience showed that response to treatment can vary even within the same genotype and family. Individualized treatment plans could potentially provide better disease control in the future.

Key words: Lumasiran, Hyperoxaluria, Nephrolithiasis, New therapies, End-stage renal disease, Genetic

POSTER DEFENSE- POD 19

ACUTE KIDNEY INJURY IN ALLOGENIC HEMATOPOIETIC STEM CELL TRANSPLANTATION IN THE PICU.

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Objectives:
Renal dysfunction is a major complication of allogenic hematopoietic stem cell transplantation (alloHSCT). The risk of kidney damage is directly related to the conditioning method, previous comorbidities and basal creatinine levels.

Methods:
The study objective was to describe the casuistry of acute kidney injury(AKI) in our serie of paediatric intensive care patients undergoing alloHSCT and to investigate the renal factors associated with a poorer prognosis.

Results:
Observational, retrospective, descriptive, analytical study of renal complications presented by paediatric patients undergoing alloHSCT, admitted to PICU (paediatric intensive care) for 10 years. 132 allogeneic HSCT interventions were performed at Hospital de Santa Creu i Sant Pau (Barcelona) in 112 patients, 54(48.2%) of whom (accounting for 68 transplants) required PICU admission.

The overall actuarial survival rate was 41.7% at 12 years. Of the 54 patients who required PICU admission, the main reasons were respiratory failure (41.1%), followed by neurological (22.7%) or haemodynamic (21.2%) disorders, and kidney (6.0%) or liver (6.0%) failure. pSOFA:5.67, OPRISM:12.97.
On PICU admission, the univariate analysis showed significantly increased weight (p=0.007), creatinine (p=0.007), and a significantly decreased glomerular filtration rate (p<0.001). AKI affected 22 children: eleven KDIGO1, four KDIGO2 and seven KDIGO3. Lower survival was associated in the univariate (p=0.036) and multivariate (OR:6.08, p=0.014) study with KDIGO3.
During PICU stay, 42 patients presented AKI: six KDIGO1, ten KDIGO2 and twenty-six KDIGO3. 27 patients require dialysis techniques(40.9%): Continuous renal replacement (23), conventional dialysis(1), combined techniques(3). Mean duration days of therapy: 19.56±18.14days. Significant differences in mortality were found in patients with AKI(p=0.002), KDIGO3(p=0.011) and those in need of dialysis(p<0.001).

Conclusion:
Children requiring PICU admission after alloHSCT have a significant increase in creatinine levels and a significant decrease in glomerular filtration on admission. The highest degree of acute kidney disease (KDIGO3) and the need of dialysis are risk factors significantly associated with mortality.

Key words: Acute kidney injury, Allogenic stem cell transplantation, Paediatric intensive care

POSTER DEFENSE- POD 20

ESTAREMOS A DIAGNOSTICAR A LESÃO RENAL AGUDA? - DESCRIÇÃO DA LRA NUMA UNIDADE DE CUIDADOS INTENSIVOS PEDIÁTRICOS

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Objectives:
A lesão renal aguda (LRA) parece ser ainda um diagnóstico pouco identificado na nossa prática clínica. Contudo está associada a um aumento de risco significativo de morbimortalidade em idade pediátrica.

Methods:
O objetivo deste estudo é determinar a frequência de LRA identificada pelo médico e a sua incidência numa unidade de cuidados intensivos pediátricos (UCIP).

Results:
Estudo retrospetivo de colheita de dados entre Janeiro de 2021 a Dezembro de 2022 numa UCIP. Crianças com mais de um mês de vida até 18 anos foram incluídas. Foram analisados os dados clínicos e bioquímicos de todos os doentes admitidos na UCIP e aplicada a classificação Kidney Disease: Improving Global Outcomes (KDIGO). Doentes com duração de internamento inferior a 24 horas, com doença renal crónica e ausência de valores de creatinina durante a permanência na UCIP foram excluídos do estudo.

Conclusion:
Durante o período do estudo, 156 crianças ficaram internadas na UCIP; 121 cumpriram os critérios de inclusão; 21 apresentaram LRA (18%). Os casos identificados distribuíram-se homogeneamente pelos graus de KDIGO 1, 2 e 3 (1/3 em cada). A média das idades foi de 104 meses e a média dos níveis de creatinina máxima foi de 1.1 mg/dL. A LRA foi identificada pelos médicos em 3 de 21 casos de LRA.
Os casos identificados apresentaram todos LRA grau 2 ou 3, a média de idades foi de 120 meses e a média dos níveis de creatinina máxima foi de 1.9mg/dL. A LRA é uma complicação frequente na UCIP, mas encontra-se subdiagnosticada. No nosso estudo só 14% tinham o diagnóstico de LRA na nota de alta ou em diário clínico. Os casos identificados eram de faixa etária superior, com LRA mais grave e níveis de creatinina superiores aos de referência. São por isso necessários novos biomarcardores de lesão renal para um melhor reconhecimento e tratamento da LRA.

Key words: lesão renal aguda, diagnóstico, UCIP

POSTER DEFENSE- POD 21

TUBULOINTERSTITIAL NEPHRITIS (TIN): DIFFERENT SIDES OF THE SAME COIN

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Objectives:
TIN in pediatrics represents 7% of the causes of acute kidney injury(AKI).It have a variety of manifestations,even oligosymptomatic,with the classic triad (fever,eosinophilia and exanthema)being observed in only 10%.The causes are multiple(pharmacological 70%)and can be associated with clinical syndromes.The diagnosis is defined by histology,but renal biopsy is not always performed due to the rapid and good evolution of some patients.The pathogenic mechanism is immune-mediated,self-limited and reversible,could evolve to tubulointerstitial fibrosis and CKD

Methods:
The aim of our study is to describe the clinical characteristics of patients with TIN

Results:
9 years(2013-2022)retrospective descriptive study of a series of cases with TIN(with/without biopsy)evaluating demographic,etiological,clinical-analytical,evolution and treatment parameters.Exclusion criteria:previous renal disease/renal transplantation.KDIGO 2012 criteria were used to evaluate AKI.eGFR was estimated using Schwartz 2009 equation in >1 year,by CKD-EPI in >12 years an by Filler equation in Cystatin C.Proteinuria was expressed as iPr/Cr(mg/mg) and Alb/Cr(mg/mmol).Fractional excretion of solutes were analyzed using urinary Beta-2-microglobulin(ug/ml)as a marker of tubular injury.Eighteen cases were identified,median age of 14 years(2-7 years).11 renal biopsies were performed (61.1%).More frequently indication:Persistent AKI of non-filial etiology despite supportive treatment.11 pharmacological cases were identified(61.1%),3 infectious(2 M.Pneumoniae),2 TINU and 2 idiopathic.Most common symptoms and sings were abdominal pain(94%)and fever(94%).The classic triad was detected in two cases(11.1%).On admission,all patients had normal BP,with only one patient oliguric on debut.The delay from clinical onset to diagnosis was a median of 8.5 days(IQR 20.5).Renal ultrasound showed renal hyperechogenicity(25%)and nephromegaly(12.5 %).Median eGFR(1 month)was 79.72mL/min/1.73 m2,with only 1 case of recurrence and 2 of chronicity.7 cases received treatment with corticosteroids and 2 with immunosuppressants

Conclusion:
In AKI,TIN is one of the causes that we must always keep in mind in our differential diagnosis.In the series we confirm the wide forms of presentation of the disease and its various etiologies.Likewise,the evolution and prognosis will depend on the cause and the early diagnosis,conditioning the treatment

Key words: Acute Kidney Injury (AKI), Tubulointerstitial Nephritis with Uveitis (TINU), Tubulointerstitial Nephritis

POSTER DEFENSE- POD 22

PATIENT JOURNEY IN CYSTINOSIS: FOCUS IN NON-ADHERENCE AND DISEASE MANAGEMENT

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Introduction:
Cystinosis is a rare lysosomal storage disorder with renal and extra-renal manifestations. It is a chronic and progressive disease with potentially severe complications, that can be reduced and /or delayed with early and long-life cysteamine treatment. However, the challenging side-effects and strict dose schedule of cysteamine lead o risk of non-adherence, which is associated with worse prognosis and fast disease progression.

Objectives:
This project aims to evaluate, together with patients, the impact of cystinosis and treatment-associated problems on therapeutic compliance, in order to detect potential leverages of change.

Methods:
In March 2022, standardized online interviews were performed to six patients with cystinosis treated with cysteamine, aged between 12 to 40 years, and two caregivers. Further, in April 2022, two online workshops were organized, each one with the participation of an advisory board conformed of up to 4 patients and 6 caregivers. As result, the first patient journey mapping was developed considering pre-diagnosis, diagnosis, and post-diagnosis steps, prescription of treatment, laboratory tests and daily life for patients, categorized by age (children, teenagers, adults). A patient support program PSP was also considered.

Results:
Patients described fatigue, sleep interruption and extra-renal affections as main impact features related to cystinosis although the first two points are mainly linked to cysteamine. Adherence worsened during the paediatric-adult transition process. Patients were not completely aware of disease complications risks associated to non-adherence. Main factors explaining poor therapeutic compliance were GI symptoms and cysteamine night dosing schedule, which impaired sleep pattern and had a negative impact on daily life.

Conclusion:
Although more research is needed, patients with cystinosis and caregivers provide key information to identify topics for optimizing adherence and disease management. Thus, adherence could be improved by implementing patient suggestions and following lines of action arisen from this project.

Key words: Cystinosis, cysteamine, Adherence, patients, caregivers, patient journey

POSTER DEFENSE- POD 23

IDIOPATHIC NEPHROTIC SYNDROME: AN OVERVIEW OF OUR PRACTICE

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Introduction:
Idiopathic nephrotic syndrome(INS) is the most common glomerulopathy in paediatrics. Prognosis is determined by corticosensitivity(CS), with around 20% developing corticoresistance(CR). In addition, 50-70% of CS will have frequent relapses(RF) or corticodependence(CD).

Objectives:
- Performing a descriptive study of INS patients diagnosed in the last 12 years.
- Trying to identify parameters to predict evolution.

Methods:
Retrospective observational analysis of INS patients≤16years between 2010-2022. It included 79 patients(69.6%male). Median diagnostic age:3.83years(2.75-5.37). Sample distribution: CR=15.1%;CS=84.9% with 73.1%CD/RF.

Results:
- CD/RF: 49 patients(73.5%male). Median diagnostic age was 3.58years(2.7-5.12). Remission in the first outbreak was achieved at a median of 11.5 days(7-26.75), with 7 relapses(5-12.5) on average. 85.7%(42/49) received corticosteroid-sparing drugs: 28/42 levamisol (8 as single drug), 30.6%(15/49) mycophenolate, 24.4%(12/49) cyclophosphamide, 8.1%(4/49) cyclosporine, 6.1%(3/49) Tacrolimus and 6.1%(3/49) Rituximab.11 patients required >3drugs. After 9.9 years(3-9.95) of follow-up, none had developed chronic kidney disease and 42.8% were in remission without treatment. Only 4 children had infectious complications; thrombotic was not presented.
Age at diagnosis was higher in patients requiring >2 additional drugs(6.29vs.4.23 years;p=0.01). Time required for response in the first flare-up was longer in those who subsequently presented >4 Relapses(R)(R<4:7,6días vs.R>4:17,3días;p=0.02).
- CR: 12 patients(41.7%male): Median diagnostic age was 5.6years(2.9-8.9). Cyclosporine was the most frequent additional drug(83.3%). All underwent biopsy, with focal segmental glomerulosclerosis(FSGS 58.3%) followed by minimal change disease (MCD 25%). Genetic study was performed in 91.7% patients, identifying specific mutations in 50%. 41.6%(5/12) required renal replacement therapy(RRT) 22 months(19-27) after diagnosis.
RRT risk was lower in MCD patients compared to other histological patterns(p=0.01).

Conclusion:
- Disease onset at a later age and longer initial response time in the first outbreak, would be associated with a worse evolution in CD/RF. Levamisol as single drug was useful in 28.5% patients
- 41% INS-CR patients required RRT after a median of 22 months post-diagnosis. MCD patients have a better prognosis.

Key words: Idiopathic nephrotic syndrome, paediatrics, prognosis

POSTER DEFENSE- POD 24

CYSTIC KIDNEY DISEASE: A RARE DIAGNOSIS PRESENTATION

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Introduction:
Pediatric cystic kidney diseases (CKD) include a diversity of hereditary or non-hereditary conditions, whose phenotypic presentation can vary from asymptomatic to end-stage renal disease.

Results:
We report a case of a male with a pre-natal diagnosis of pyelectasis at 22 weeks' gestation, delivered at 39weeks, with no peri-natal complications. Throughout the first 2 years of life, he had serial renal ultrasounds, with regression of pyelectasis, but increased renal echogenicity, firstly identified at 3 months-old. Family history of renal diseases was negative. Voiding cystourethrography showed no signs of vesicoureteral reflux and MAG3 renogram revealed equally functioning kidneys. He was on trimethoprim prophylaxis until he turned 1 year-old, with no reported urinary tract infections. At 29 months-old, elevated transaminases and two cortical cysts on the right kidney were detected, raising the suspicion of a CKD. Although hepatic ultrasound was normal, he had a hepatic biopsy done, which had no signs of fibrosis. At this time, he was tested for ARKPD, which was negative. Renal ultrasounds showed progression of the number of cysts bilaterally, with diffuse hyperechogenicity and reduction of parenchymal-sinus differentiation. Throughout the years, he maintained normal blood pressure and elevated transaminases with raising levels of serum creatinine from the age of 13. A NGS panel for polycystic diseases detected 17q12 deletion syndrome, that causes renal cysts and may include other urinary tract malfunctions, as well as MODY diabetes, hyperparathyroidism, hypoacusis, altered hepatic function and behavioral and psychiatric conditions. On last appointment (14 years-old) ultrasound showed bilateral cysts (10mm) and worsened renal function (GFR of 71ml/min/1.73m2), elevated transaminases and normal glycemia.

Conclusion:
In this case, an atypical presentation of CKD led to an extended investigation resourcing to other genetic panels, which allowed not only the etiological diagnosis, but also enabled the early identification and monitoring of possible extra-renal comorbidities, commonly associated with this syndrome.

Key words: cystic kidney disease, genetic, 17q12 deletion, renal cysts

POSTER DEFENSE- POD 25

AUTOSOMIC RECESSIVE POLYCYSTIC KIDNEY DISEASE: CLINICAL VARIABILITY OF THE SAME DISEASE

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Introduction:
Autosomal recessive polycystic kidney disease (ARPKD) is a rare hereditary disease secondary to mutations in the PKHD1 gene. Multiple pathogenic variants in this gene have been described, with a broader spectrum of clinical expression than the classically recognized. We present the clinical manifestations and the mutational study of the cases of ARPKD identified in the fetal and pediatric stage, in our Hospital.

Results:
Seven cases were included. Five of them were identified after prenatal suspicion due to oligo/anihydramnios and bilateral nephromegaly. One of them ended up in a voluntary termination of pregnancy. Two cases were diagnosed accidentally at 5 and 8 years of age, in the context of evaluation of liver fibrosis and abdominal pain, respectively. Two of the prenatally diagnosed cases presented oligoanuria and acute renal failure in the immediate neonatal stage requiring, in the first 24 hours of life, renal replacement therapy. In one of them, due to comorbidities, limitation of therapeutic effort was carried out. The other two cases of prenatal diagnosis presented chronic kidney disease (CKD) from birth. Currently, one patient is on chronic hemodialysis, another is on the waiting list for renal transplantation, two are in advanced CKD and one has normal renal function.In the absence of one of the results of a genetic study, the rest of the cases, presented mutations in PKHD1, with a predominance of pathogenic variants in compound heterozygosity. The family segregation study demonstrated the presence of the mutations in the parents

Conclusion:
The manifestations described in our patients confirm the heterogeneous clinical expression of this disease, ranging from cases with early and severe manifestations in the fetal and neonatal stage, to others with late presentations and predominance of extrarenal involvement. The genetic study confirmed the disease in all cases, this will allow further knowledge of its phenotypic correlation with larger clinical series.

Key words: AUTOSOMI RECESSIVE, POLYCYSTIC, KIDNEY

POSTER DEFENSE- POD 26

A IMPORTÂNCIA DA CINTIGRAFIA-DMSA NO SEGUIMENTO DE INFEÇÕES DO TRATO URINÁRIO EM PEDIATRIA

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Introduction:
As infeções do trato urinário (ITU) são infeções bacterianas frequentes em pediatria, sendo fundamental identificar os doentes com risco de lesão renal a longo prazo. A ecografia reno-vesical (ERV) é um exame de primeira linha e a cintigrafia-DMSA o exame gold-standard para identificação de cicatrizes renais. Segundo a Academia Americana de Pediatria (AAP), esta última deve ser considerada se alterações ecográficas e ponderada em alguns casos de ITU recorrente. Contudo, na nossa Instituição, o protocolo de atuação inclui indicações mais alargadas: ITU < 6 meses, ITU atípicas até aos 3 anos de idade, ITU recorrente, alterações ecográficas.

Objectives:
Comparar o seguimento das ITU num Hospital de Grupo I com o instituído na norma da AAP.

Methods:
Estudo observacional retrospetivo dos exames complementares de diagnóstico realizados em crianças diagnosticadas com ITU em 2021 e 2022. Análise estatística com nível de significância a 5%.

Results:
Foram diagnosticadas 472 ITU em 398 crianças, entre estas, 63 foram submetidas a ERV. Foram realizadas 28 cintigrafias-DMSA (4 ITU atípicas; 4 alterações ecográficas; 10 ITU recorrentes e 10 ITU <6 meses), 71,4% sexo feminino, mediana de idades 5,5 meses. Destas, 8 apresentaram alterações (28,6%), apenas 1 doente (12,5%) com ecografia alterada, não tendo sido encontrada uma relação estatisticamente significativa entre ERV normal e ausência de lesões na cintigrafia (p=0,466). Todos estes doentes com cicatrizes renais mantiveram seguimento, nenhum apresentando hipertensão arterial, diminuição da taxa de filtração glomerular ou proteinúria.

Conclusion:
Apesar da ERV ser um exame inócuo e de fácil realização, não parece detetar a maioria das cicatrizes renais. A cintigrafia-DMSA utiliza radiação e deve ser realizada de forma criteriosa, no entanto, o alargamento das indicações para a sua realização parece ser importante para identificação de cicatrizes renais com potencial risco de progressão a longo prazo, que requerem acompanhamento regular com implementação de medidas reno-protetoras.

Key words: Cintigrafia, Pielonefrite, Ecografia, Pediatria

POSTER DEFENSE- POD 27

EFFECT OF HYPERPRESSURE AND LOSS OF RENAL PARENCHYMA ON THE RESULTS OF BASIC RENAL FUNCTION TESTS IN CHILDREN WITH VESICOURETERAL REFLUX

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Introduction:
Severe vesicoureteral reflux (VUR) is accompanied by impaired concentrating ability in 100 % of cases. Something similar occurs in the loss of parenchyma; with GFR <70ml/min/1.73m2 the maximum urinary osmolality (UOsm) is always reduced.

Objectives:
We wanted to check whether there is a summative effect of both circumstances on renal function.

Methods:
Retrospective cross-sectional study in which 175 children (117V, 58M) were included. They were classified into three groups: absence of VUR (No VUR; n=118), mild-moderate VUR -VUR grades I-III- (VUR I-III; n=30) and severe -VUR grades IV and V- (VUR IV-V; n=27). Twenty-eight cases of parenchyma loss [atrophic kidney (n=15), unilateral scar (n=10), hypoplasia (n=2), hypoplasia and scar (n=1)] were confirmed. In addition to UOsm, the values of the albumin/creatinine and NAG/creatinine ratios were collected.

Results:
Some of the three parameters studied were abnormal in 51% of the cases of the No VUR group, in 60% of the VUR I-III group, and in 100% of the VUR IV-V group. There were differences between groups in UOsm ($p<0.001$) but not in the values of the ratios. The frequency of UOsm reduction in the group No VUR was 40% and 25% in the subgroups without (n=110) and with loss of parenchyma (n=8) respectively, and in VUR I-III it was 68% and 13% in the subgroups without (n=22) and with loss of parenchyma (n=8) ($p=0.02$); there was a difference in UOsm and albumin/creatinine values between these two subgroups.

Conclusion:
VUR influences renal water management probably due to hyperpressure. The loss of parenchyma did not affect in the cases on the absence of VUR and in severe VUR. The differences in UOsm and albumin/creatinine in the mild-moderate VUR subgroups may be due to the shortness of the sample, although the effect of compensatory hypertrophy is possible, a mechanism that our Group has verified in the single kidney.

Key words: vesicoureteral reflux, concentrating capacity

POSTER DEFENSE- POD 28

ENURESE NOTURNA – EXPERIÊNCIA DE 6 ANOS DA CONSULTA EXTERNA DE NEFROLOGIA DE UM HOSPITAL DE GRUPO I

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Introduction:
A enurese noturna (EN) é uma patologia comum, caracterizando-se por incontinência urinária durante a noite em crianças ≥ 5 anos. A sua prevalência varia com a idade, é mais frequente nos rapazes, tendo causas multifatoriais.

Objectives:
Caracterizar a população referenciada por EN à consulta externa de nefrologia pediátrica dum Hospital de grupo I entre 2017-2022.

Methods:
Estudo retrospetivo descritivo, através da consulta de processos clínicos. Análise estatística com recurso a SPSS®, versão 27.0.

Results:
Incluídas 116 crianças, 58,6% do sexo masculino, a maioria referenciada a partir de outras consultas externas (83,6%). A idade média na primeira consulta foi de 9,5 anos [desvio-padrão=2,7] e 50,9% apresentaram antecedentes familiares de EN.
As comorbilidades mais frequentes foram: patologia do neurodesenvolvimento (28,4%; PHDA 19,8%), obstipação (20,7%), obesidade/excesso peso (13,8%), roncopatia/SAOS (12,9%), asma/rinite (12,1%), patologia do foro psiquiátrico (12,1%).
A EN foi classificada como primária em 66,4% e monossintomática em 73,3%. A análise sumária de urina foi normal em todas as crianças e verificou-se disfunção vesical em 76,7% das ecografias reno-vesicais realizadas. 33,6% das crianças já tinham efetuado/efetuavam algum tratamento. A maioria iniciou farmacoterapia (85,3%): 51,5% com desmopressina e 42,4% com desmopressina e oxibutinina; e 8 crianças usaram alarme/despertador. 40,5% das crianças foram seguidas em consulta externa de Psicologia.
44,8% tiveram alta, com uma idade média de 11,3 anos [desvio-padrão=3,1] e 18,5 meses até alta. Taxa de abandono de 24,1%.

Conclusion:
A maior prevalência de EN primária monossintomática, sexo masculino e história familiar positiva, está de acordo com a literatura. Verificámos uma elevada presença de comorbilidades que devem ser ativamente procuradas e tratadas. O grande número de crianças seguidas noutras consultas externas demonstra a multidisciplinaridade da intervenção.
Pretendemos chamar a atenção para esta patologia multifatorial, com grande impacto na qualidade de vida. É importante identificar precocemente e implementar medidas comportamentais, sendo fundamental a motivação da criança e sua família.

Key words: Enurese, Incontinência, Qualidade de vida, Pediatria

POSTER - PO 01

ACUTE KIDNEY INJURY IN HYPOXIC-ISCHEMIC ENCEPHALOPATHY NEONATES TREATED WITH THERAPEUTIC HYPOTHERMIA

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Objectives
Neonatal asphyxia causes hypoxic-ischemic encephalopathy (HIE) and multisystem affectation, includying acute kidney injury (AKI). This acts as an independent morbidity and mortality risk factor, and can cause chronic kidney disease (CKD).

Methods
To evaluate neonatal AKI in HIE subjected to therapeutic hypothermia (TH) and its renal prognosis. Assess the Renal Angina Index (RAI) score predictive of neonatal AKI. Relate AKI with neurological affectation.

Results
Ambispective observational study. Population: Neonates >35 weeks GA with HIE treated with TH in the Neonatal ICU of the Hospital Clínico Universitario de Valladolid from 2011-2019. Sources: Clinical histories. Variables: clinical-epidemiological, diagnostic-therapeutic, evolutionary, renal biomarkers, DRA-Neonatal-KDIGO-2012 criteria. Assessment of the RAI Score. Comparison between neonates with/without AKI.

Conclusion
Results: 34 neonates (58.8% male) were registered, 21 (61.8%) had moderate HIE and 13 (38.2%) severe HIE. 18 (52.94%) presented AKI [7(38.9%) grade I, 11(61.1%) grade II], with a median plasma Cr-24h life of 1.09 (0.95-1, 22) mg/dl. There is a relationship between a higher degree of AKI and HIE, cesarean delivery, longer stay in the NICU, coagulopathy, resuscitation with cardiac massage, elevated troponin T, days of intubation, need for inotropes and blood products ($p<0.05$). 21 neonates (61.8%) presented RAI Score alteration, 18 (85.7%) with AKI [sensitivity (100%), specificity (81.25%), PPV (85.7%) and NPV (100%), Spearman's correlation coefficient: 0.791]. 4 patients died, 3 (75%) with AKI. 12(48%) survivors with AKI developed renal abnormalities, 2(8%) CKD G2A1. The differences in neurological prognosis were not statistically significant.
Conclusions: Neonatal AKI implied higher morbidity and mortality. The RAI Score is valid for its prediction (high sensitivity, specificity, PPV and NPV), and can be used in its screening to optimize its treatment and prognosis.

Key words: acute kidney injury, hypoxic-ischemic encephalopathy, therapeutic hypothermia, chronic kidney disease, Renal Angine Index

POSTER - PO 02

LESÃO RENAL AGUDA E RABDOMIÓLISE GRAVE – UM DIAGNÓSTICO GENÉTICO RARO A CONSIDERAR

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Introduction:
A rabdomiólise caracteriza-se por necrose do músculo esquelético, manifestando-se pela tríade de mialgias, fraqueza muscular e mioglobinúria, podendo complicar-se de lesão renal aguda (LRA). Tem causas variadas como trauma, imobilização prolongada, esforço físico extenuante, e em casos mais raros miopatias metabólicas/mitocondriais.

Results
Sexo masculino, 15 anos. Irmã com episódios de rabdomiólise recorrentes, em investigação. Acompanhado em consulta de Doenças Neuromusculares, desde os 11 anos, por mialgias/cãibras para médios esforços, seguidas de período de mioglobinúria. Abandonou seguimento aos 13 anos, sem ter completado investigação.
Recorreu ao serviço de urgência por, no decorrer de prova de corrida, ter iniciado mialgias/cãibras intensas e limitadoras da marcha, posteriormente associadas a micções de urina com coloração de vinho do porto. Sem alterações no exame objetivo, incluindo pressão arterial (<P90), frequência cardíaca (80bpm), peso habitual (53Kg), edema periférico ausente.
Analiticamente apresentava LRA (creatinina habitual 0,6mg/dL, na admissão 1mg/dL), aumento de CK (488100UI/L), mioglobina (13561ng/mL), AST (1909UI/L), ALT (699UI/L), sem alterações no ionograma/gasimetria.
Decidido internamento para fluidoterapia intensiva por via endovenosa, terapêutica com bicarbonato para alcalinização da urina, monitorização clínica/analítica e investigação complementar.
Fez estudo genético com identificação de duas variantes patogénicas raras no gene PYGM descritas em doentes com Glicogenose tipo V.
Após normalização da função renal, enzimas musculares e perfil hepático, teve alta referenciado às consultas de Nefrologia/Neuropediatria.

Conclusion
A Glicogenose tipo V é um subtipo de miopatia metabólica, sendo a doença de armazenamento do glicogénio que mais frequentemente afeta o músculo. Como no presente caso, manifesta-se mais frequentemente na adolescência com rabdomiólise, por vezes complicada de LRA, sendo o diagnóstico confirmado por estudo genético.
O caso descrito alerta para um diagnóstico raro, que deve ser equacionado em crianças com história familiar de rabdomiólise, e pessoal de episódios recorrentes de mialgias/cãibras, intolerância durante e hematúria após o esforço físico, de forma a adequar a investigação etiológica.

Key words: Rabdomiólise, Lesão Renal Aguda, Doença Genética

POSTER - PO 03

LSEVERE KIDNEY IMPAIRMENT AT THE EMERGENCY CARE: ROLE OF A PROMPT ACTUATION.

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Introduction
·The etiological diagnosis of stage 3 acute kidney injury in emergency settings is a challenge that we can face in our daily practice. There are occasions when a high index of suspicion is necessary to confirm rare causes that may require specific treatment.

Results
·A 12-year-old girl presented to the emergency department with a 24-hour history of anuria. Urethral catheterization was performed, but no urine sample was obtained. The patient reported progressive oliguria over the last week, without urinary symptoms or macroscopic changes in urine. There were no significant personal or family medical history, and the patient had no history of medication use or previous trauma.
·Creatinine 14.1 mg/dL, urea 147 mg/dL, and uric acid 11.3 mg/dL. Hemoglobin 12.8 g/dL, and platelets 387,000/µL. An ultrasound showed two kidneys with good corticomedullary differentiation, normal size for height, and mild grade I right-sided dilation. Abdominal X-ray detected a stone of less than 5 mm in the left kidney. An abdominal CT scan confirmed the suspected diagnosis of bilateral urolithiasis.
·An urgent urological intervention was performed, which involved bilateral double-J stent placement. The patient developed post-obstructive diuresis and a rapid decline in plasma creatinine levels, which at 48 hours post-intervention, was 0.92 mg/dL.
·The urine study revealed an increased excretion of cystine, with hexagonal crystals observed in the fresh sample. The Brand test was positive, confirming the diagnosis of Cystinuria.

Conclusion
In cases where there is suspicion of obstructive acute renal failure based on the patient's history and clinical presentation, but ultrasound and abdominal radiography are inconclusive, an abdominal CT scan should be performed to detect small or ureteral stones. Urgent surgical intervention of the urinary tract is crucial to prevent permanent kidney damage.

Key words: Kidney impairment, cystinuria.

POSTER - PO 04

IMPORTED ACUTE KIDNEY INJURY: ARE WE PREPARED?

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Introduction:
Malaria is an endemic disease in African countries, imported by globalization and migratory movements.
Kidney damage is related to the type of plasmodium that causes it (especially P.falciparum) and the degree of parasitemia.
The treatment of choice includes intravenous (IV) artesunate, which is not exempt from renal complications.

Objectives:
To report causes of Acute Kidney Injury (AKI), infrequent in our environment, and the evolution in 2 pediatric patients born in Spain who traveled to endemic countries.

Methods:
Two cases of severe malaria due to P.falciparum with AKI. They were admitted to PICU for IV treatment (cefotaxime, artesunate, fluid-therapy, serum albumin and red-blood-cell transfusion).

Results:
Case 1: A 15-month-old female who traveled to Nigeria and returned two weeks before the onset of symptoms: fever, jaundice, choluria, oligoanuria and impaired consciousness. In initial examinations: parasitemia 12%, AKI (eGFR 40.78mL/min/1.73m2), proteinuria and arterial hypertension. She receives IV artesunate, furosemide and conservative measures. She is discharged after 8 days with eGFR improvement.
Case 2: A 13-year-old woman who traveled to Senegal and returned 11 days before the episode: fever, abdominal pain, jaundice, choluria, hepatomegaly, arterial hypotension and oligoanuria. In examinations: parasitemia 8.9%, AKI (eGFR 55mL/min/1.73m2) with normalization after 72h of treatment with artesunate and conservative measures of the AKI. Seven days after, there is clinical and analytical worsening without parasitemia. Diagnosed of hemolytic crisis due to artesunate with AKI, high-dose corticosteroid-therapy is prescribed with progressive improvement. She is discharged after 17 days with eGFR 82mL/min/1.73m2.

Conclusion:
Due to globalization, it is increasingly common to find imported pathologies with potential renal involvement.
AKI due to malaria, rare in children, has different pathophysiological mechanisms and damage degree is proportional to parasitemia.
Artesunate hemolytic crisis, which usually appears 1-4 weeks after initial treatment, aggravates AKI and should be treated with high-doses of corticosteroids to prevent disease progression.

Key words: malaria, kidney, failure, artesunate, hemolytic.

POSTER - PO 05

IACUTE RENAL FAILURE IN AUTOIMMUNE HEMOLYTIC ANEMIA

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Introduction:
Autoimmune hemolytic anemia (AIHA) is a rare inmune disorder due to autoantibodies directed against erythrocytes, causing shortened erythrocyte survival, with or without complement activation. The disorders can appear as a primary disorder(idiopathic) or secondary to other autoimmune-disorders, malignancies or infections. Treatment involves immune modulation.

Results:
We report an 5-year-old girl who presented fever, hyporexia and a cough in the last <12 hours. In the emergency room, she had unstable pediatric assessment triangle(PAT) due to shock. She was transferred to PICU and started IV empiric antibiotics. In blood test, she presented leukocytosis with neutrophilia, CRP 182mg/L, hemolytic anemia with normal platelets and acute kidney injury. Direct Coombs was positive (C3D+). Chest X-ray revealed atelectasis of the right upper lobe. She had negative pneumococcal antigenuria, negative urine and blood cultures. After 48 hours, she had higher rate of creatinine and urea, the hemolytic anemia worsened and had significant thrombocytopenia. That required red blood cell transfusion and initiation of continuous hemodiafiltration, which was maintained for 3 days.
Having differential diagnosis between AIHA and atypical hemolytic uraemic syndrome (aHUS) due to pneumococcus without isolating the microorganism, we started IV-corticosteroids. Hemolysis decreased at 72 hours, presenting worse renal function up to urea of 235mg/dL with ions and controlled blood pressure, improving after 6 days, without requiring renal replacement therapy. At discharge, 21 days after admission, she presented normal glomerular filtration.
Agglutinin test was negative on several occasions, until 14 days after admission, direct Coombs positive for IgG and C3D was obtained, being compatible with AIHA -IgG+ complement by warm antibodies.

Conclusion:
In hemolytic anemias with associated renal damage, we must establish differential diagnosis of thrombotic microangiopathy versus AIHA. Given the suspicion of autoimmunity, early initiation of steroids is important to improve the kidney function prognosis. Immunosuppressive drugs may be indicated as second level treatment,once the diagnosis is established.

Key words: Acute renal failure, renal replacement therapy, hemolytic anemia

POSTER - PO 06

PAEDIATRIC PATIENT WITH ANURIC ACUTE KIDNEY INJURE WITH A PREVIOUS HISTORY OF DIARRHOEA, NOT EVERYTHING IN PAEDIATRICS IS HAEMOLYTIC UREMIC SYNDROME.

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Objectives:
We define osmotic nephrosis as the presence of vacuolisation and oedema in the cells belonging to the proximal renal tubule. This non-specific finding is associated with various aetiologies of acute kidney injure (AKI) and is rarely described in children.

Methods:
To report a clinical case with this histological finding, as well as review its causes and outcomes.

Results:
Description of a case-report and review of the literature.
Case Report: 7-year-old girl was admitted at our center due to anuric AKI. She presented previous history of diarrhoea, hyporexia and gross haematuria for one week.
Laboratory findings showed decreased glomerular filtration rate(GFRe) (maximum serum creatinine 6.4 mg/dl) associated with mild hypertransaminasemia and elevated LDH, with no abnormalities in the blood count. Renal doppler was normal. Mycrobiological and immunological studies were negative.
During the first 24 hours, fluid therapy and diuretic treatment were administered without response. On the third day of anuria, a central catheter was placed and a kidney biopsy was performed. The biopsy showed a pattern compatible with osmotic nephrosis with no glomerular alterations.
With these findings and without specific treatment other than ensuring adequate hydration, and no need for dialysis, the patient began spontaneous diuresis on the fifth day of admission. She recovered renal function in less than 48 hours. After 9 months of follow-up, she maintains a normal GFRe with no evidence of tubular involvement.

Conclusion:
AKI related to osmotic nephrosis has been described in association with the administration of various intravenous osmotic agents, such as immunoglobulins, dextrans or mannitol, as well as oral antidiabetics of the SGLT2 family. In our case, however, we could not establish a causative agent. AKI is usually reversible, with fully recover of renal function, although it can sometimes leave long-term sequelae and require transient dialysis. Kidney biopsy is essential for diagnosis.

Key words: acute kidney injury, osmotic nephrosis, Kidney biopsy, osmotic agents

POSTER - PO 07

PINCIDÊNCIA DE INJÚRIA RENAL AGUDA EM PACIENTES ADMITIDOS EM UMA UTI PEDIÁTRICA PARTICULAR DO ESTADO DE MATO-GROSSO - BRASIL, ENTRE OS ANOS DE 2020-2021

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Introduction
A INJÚRIA RENAL AGUDA É DENOMINADA COMO ALTERAÇÃO E QUEDA ABRUPTA DA FILTRAÇÃO GLOMERULAR RENAL, PREJUDICANDO DESSA FORMA A MANUTENÇÃO DA HOMEOSTASE CORPÓREA DEVIDO AO ACÚMULO DE EXCRETAS E COMPOSTOS NITROGENADOS, TÓXICOS AO ORGANISMO.

Objectives:
DEMONSTRAR ESTATISTICAMENTE A FREQUÊNCIA DE INJÚRIA RENAL AGUDA EM PACIENTES PEDIÁTRICOS ADMITIDOS E INTERNADOS EM UMA UTI PEDIÁTRICA DE UM HOSPITAL PARTICULAR DE CUIABÁ-MT, BRASIL.

Methods:
OS DADOS DE 100 PACIENTES ADMITIDOS NA UTI PEDIÁTRICA SUPRACITADA ENTRE 2020 E 2021, FORAM ANALISADOS COM O AUXÍLIO DO MICROSOFT EXCEL E EPI INFO.

Results:
DE 100 PACIENTES ANALISADOS APENAS 01 FOI ADMITIDO COM A HIPÓTESE DIAGNÓSTICA DE INJÚRIA RENAL AGUDA. PORTANTO, 1% DAS INTERNAÇÕES ANALISADAS ESPECIFICAMENTE NESTE PERÍODO E DENTRO DESTA CASUÍSTICA. ADEMAIS A CAUSA DA IRA FOI REVELADA COMO INTOXICAÇÃO POR DIGITÁLICOS.

Conclusion:
CONCLUI-SE QUE COMPARANDO ESTE ESTUDOS COM OUTRAS PUBLICAÇÕES CIENTÍFICAS QUE REVELAM UMA PORCENTAGEM DE 5% DE IRA EM ADMISSÕES HOSPITALARES E 30% EM ADMISSÕES EM UNIDADE DE TERAPIA INTENSIVA. HOUVE 4% A MENOS DE INTERNAÇÕES POR IRA COMO DIAGNÓSTICO INICIAL. E 29% A MENOS DE DIAGNÓSTICOS DE IRA EM ADMISSÕES NA UTI PED ESTUDADA.

Key words: doença renal, nefrotoxicidade, digitalicos, UTI PED

POSTER - PO 08

IMPORTANCIA DEL SEGUIMIENTO NEFROLÓGICO EN PACIENTES CON 'SOSPECHA' DE VEJIGA NEURÓGENA'

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Introduction:
Destacar la importancia, a propósito de 3 casos, de un seguimiento nefrológico en pacientes afectos con vejiga neurógena.
La vejiga neurógena es una patología compleja, seguida por múltiples especialistas, pero en ocasiones el nefrólogo no es uno de ellos. Alertamos de la aparición de casos de pacientes con daño renal agudo y/o enfermedad renal crónica con patología vesical sin seguimiento nefrológico.

Methods:
CASO 1. Varón de 12 años con Neuropatía axonal gigante. Micciones 'voluntarias'; impresiona por rebosamiento. Acude a su pediatra por cefalea. TA 180/110, derivado a UCI. Creatinina 3,36 mg/dl junto con dilatación uretero-pielocalicial bilateral severa con vejiga distendida (CVE 1000 ml). Alta con Creatinina 0,5 mg/dl y Cistatina. Inicia sondajes intermitentes.
CASO 2. Varón de 13 años diagnosticado de mielomeningolipoma al nacimiento. Portador de pañal, goteo miccional contínuo. Micciones voluntarias 'sin esfuerzo'. Acude a su pediatra por astenia. Analítica sanguínea Creatinina 5,79 mg/dl; en ecografía dilatación uretero-pielocalicial severa con hiperecogenicidad renal bilateral. Vejiga multidiverticular. Inicia sondajes intermitentes. Alta Cr 2,3 mg/ml. Actualmente ERC G3A2.
CASO 3. Varón de 13 años diagnosticado de Enuresis primaria 'monosintomática'; con incontinencia diurna a diario. Orina sin referir esfuerzos al inicio ni al finalizar. Tratamiento con desmopresina sin respuesta. Acude a su pediatra por astenia. Analítica con Creatinina de 6,69 mg/dl; Hb 6,3 g/dl y ecografía con dilatación ureteropielocalicial bilateral severa, con hiperecogenicidad renal. A la EF destaca expresión facial, con risa invertida; se confirma en estudio genético, Síndrome de Ochoa. Inicia sondajes intermitentes.

Conclusion:
Destacamos la importancia de establecer un consenso unificado de seguimiento nefrológico ante pacientes con patología de base vesical. En 2 de los 3 pacientes debido al diagnóstico tardío presentan actualmente ERC G3, lo cual podría haber sido evitado ante un seguimiento precoz por parte de nefrología.

Key words: SEGUIMIENTO NEFROLÓGICO. VEJIGA NEURÓGENA.

POSTER - PO 09

SEVERE HYPERCALCEMIA IN WILLIAMS-BEUREN SYNDROME

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Objectives:
Williams-Beuren Syndrome (WBS) is a neurodevelopmental disorder with multisystemic manifestations caused by a 1.55-1.83 Mb deletion at 7q11.23. It is characterized by heart disease, intellectual disability, characteristic facial features and systemic abnormalities. Hypercalcemia appears in 15% of patients with WBS, generally transient and of unknown cause.

Methods:
To present the treatment of severe hypercalcemia in an infant with SWB.

Results:
Methods: Clinical history review.
Clinical Case: A 7-month-old male infant with a WBS begins with difficulties in feeding, constipation and irritability. A blood test was performed that showed severe hypercalcemia, requiring admission to the Pediatric Intensive Care Unit (PICU) for close monitoring, being treated with hyperhydration, furosemide and intravenous calcitonin. After normalization of blood calcium, he was discharged to the ward, presenting again on the 3rd day with severe hypercalcemia, and was readmitted to PICU with the same treatment as in the previous admission. Oral treatment with methylprednisolone (2 mg/kg/day) was started with initial normalization of calcium levels, following a descending regimen with suspension of medication in 8 weeks. Despite this, he developed several peaks of hypercalcemia, most of them asymptomatic, requiring the association of intravenous pamidronate. It required administration of up to 4 doses of pamidronate (at 9, 10, 12 and 15 months of life), associated with oral phosphorus supplements. Subsequently, calcemia has remained stable. In the first admissions, the patient was diagnosed with nephrocalcinosis and arterial hypertension, ruling out stenosis of the renal arteries by means of angiography. Hypertension was controlled in treatment with oral propranolol and nifedipine.

Conclusion:
Some patients suffer from recurrent or persistent hypercalcemia after the resolution of the hypercalcemic crisis, as ours. Although hypercalcemia is generally transient, it may last for several months, result in significant morbidity. There are no guidelines for the management of persistent or recurrent hypercalcemia in patients with WBS.

Key words: severe hypercalcemia, Williams-Beuren Syndrome, nephrocalcinosis, hypertension

POSTER - PO 10

REVISIÓN DE PACIENTES CON DILATACIÓN PIELOCALICIAL EN SEGUIMIENTO EN LA CONSULTA DE NEFROLOGÍA DE RECIENTE CREACIÓN

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Introduction:
La dilatación pielocalicial (DPC) es un hallazgo frecuente en las ecografías renales, se incluye dentro de las anomalías congénitas del riñón y del tracto urinario (CAKUT), y su gravedad es variable.

Objectives:
Dado el aumento de casos tras la mejora del screening prenatal, se ha llevado a cabo una revisión sistemática de los pacientes valorados en nuestra consulta con este diagnóstico, para tipificar las características de los pacientes y conseguir un mejor manejo de esta patología.

Méthods:
Se han incluido un total de 112 pacientes (2017-2022). Siendo estudiadas las siguientes variables: diagnóstico prenatal, sexo, lateralidad, grado, necesidad de profilaxis, número de infecciones urinarias, pruebas complementarias realizadas durante su seguimiento y cirugía.

Results:
De los 112 pacientes incluidos, un 64% corresponde a varones y un 35% a mujeres. 1 de cada 3 pacientes ya tenían un diagnóstico prenatal cuando se inició su seguimiento en consulta, debido al gran avance en este campo en los últimos años.
En cuánto a la lateralidad, la mitad de ellos poseen una DPC bilateral y la otra mitad unilateral, con predominancia del lado izquierdo 3:1.
En torno a un 70% presentan un grado leve, un 20% moderado y un 8% severo. Porcentajes similares a los reflejados en el número de ITUS: 68 ninguna, 35 han presentado 1, y 9 ≥2 ITUS.
Un 6% presentaban FG<90 ml/min/1,73m2 y en 1-2% <60ml/min/1,73m2). En un 6-14% se objetivan alteraciones en las pruebas realizadas (Renograma, Gammagrafía y CUMS) . Tratamiento quirúrgico en un 9% de los casos.

Conclusion:
Con este estudio queremos destacar que a pesar de que predominantemente son casos leves, es prioritario una detección precoz de aquellos que puedan presentar una peor evolución para reducir el deterioro de la función renal.

Key words: DPC, PRENATAL, FUNCION RENAL

POSTER - PO 11

INFEÇÃO DO TRATO URINÁRIO FEBRIL EM CRIANÇAS COM FATORES DE RISCO DE NEFROUROPATIA

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Introduction:
A infeção do trato urinário (ITU) é uma das infeções bacterianas mais comuns na idade pediátrica. A importância do seu diagnóstico, tratamento e seguimento deve-se ao facto de se poder associar a malformações do aparelho urinário e resultar em lesão renal permanente, potencialmente responsáveis por morbilidade significativa a longo prazo.

Objectives:
Avaliar a incidência de nefrouropatia em lactentes com ITU febril associada a fatores de risco de anomalia do aparelho urinário.

Methods:
Foi selecionada uma amostra dos recém-nascidos e lactentes (0-12 meses) seguidos na Consulta de Pediatria/Patologia Renal do Centro Hospitalar Póvoa de Varzim-Vila do Conde, de 2016 a 2021, com diagnóstico de ITU febril, na presença de fatores de risco de nefrouropatia (ITUs recorrentes, infeção por agentes não Escherichia coli (E.coli), elevação da creatinina sérica, bacteriémia, má resposta à antibioterapia, diagnóstico pré-natal de anomalia do trato urinário, história familiar de refluxo vesicoureteral (RVU)ou doença renal crónica). Avaliado o estudo realizado, diagnósticos e evolução.

Results:
Dos 70 lactentes incluídos no estudo, 42 (60%) eram do sexo feminino. A mediana das idades foi de 4 meses (± 3,53).
Os fatores de risco identificados foram: ITUs recorrentes (19; 27,1%), infeção por agente não E.coli (13; 18,6%), diagnóstico pré-natal de patologia do trato urinário (11; 15,7%), bacteriémia (5; 7,1%), elevação da creatinina (3; 4,3%); má resposta à antibioterapia (3; 4,3%).
Identificaram-se alterações no estudo em 78,6 % (55; 32 sexo feminino): dilatação pielocalicial em 35 (50%), refluxo vesico-ureteral em 13 (18,6%), lesão renal cicatricial em 27 (38,6%) e hipofunção renal em 2 (2,9%).
Verificou-se nas crianças com mais do que um fator de risco, uma maior incidência de nefrouropatia ($p<0.05$).

Conclusion:
Salienta-se a importância de identificar os factores de risco nas crianças com ITU febril para diagnóstico de nefrouropatia (nomeadamente RVU) e doença renal cicatricial.

Key words: Infeção do Trato Urinário, Fatores de Risco, Nefrouropatia

POSTER - PO 12

O QUE PODE ESCONDER UMA AGENESIAL RENAL – A PROPÓSITO DE UM CASO CLÍNICO

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Introduction:
A Síndrome de Zinner é uma malformação genitourinária congénita rara, caraterizada pela tríade: agenesia renal unilateral, cisto da vesícula seminal (CVS) ipsilateral e obstrução do ducto deferente. A maioria dos doentes são assintomáticos até à 3ª-4ª década, altura em que pode surgir disúria, dor na ejaculação, infeções urinárias/epididimites recorrentes, disfunção vesical e infertilidade.

Results:
Criança, sexo masculino, seguido em consulta de Patologia Renal por diagnóstico pré-natal de agenesia renal esquerda. História de tio paterno com insuficiência renal crónica de causa desconhecida. Gravidez vigiada, 38 semanas. Parto eutócico hospitalar. Somatometria adequada à idade gestacional. Período neonatal sem intercorrências. Sob profilaxia com trimetoprim logo após o nascimento, que manteve até aos 3 meses.
Ecografia renovesical, aos 8 dias de vida, confirmou a ausência de rim à esquerda e rim direito de caraterísticas normais. A cintigrafia renal com ácido dimercaptosuccínico, em D43 de vida, revelou: rim esquerdo funcionante (97,13%) e exclusão funcional do rim direito (2,8%). A cistoureterografia miccional seriada, realizada aos 2 meses, não apresentou alterações.
Uma ecografia renovesical de controlo, aos 3 anos, identificou ureter distal remanescente à direita, cuja porção distal comunicava com estrutura multiquística paramediana na pelve renal, compatível com CVS. Esta alteração foi confirmada por tomografia computorizada abdomino-pélvica que revelou lesão hipodensa, multisseptada, cística, de 18 mm, na região da vesícula seminal direita. Esta associação de agenesia renal com CVS é típica do Síndrome de Zinner.
Atualmente, com 4 anos e 5 meses, a criança permanece assintomática.

Conclusion:
Os CVS, em idade pediátrica, são, na maioria das vezes, diagnosticados incidentalmente durante a investigação de anomalias do trato urinário. O tratamento conservador é preferido, reservando-se a abordagem cirúrgica para os casos muito sintomáticos. É recomendado um seguimento destes doentes até à idade pós-púbere, uma vez que os sintomas podem aparecer apenas após o início da atividade sexual.

Key words: Agenesia renal, Cisto da vesícula seminal, Malformação congénita, Síndrome de Zinner

POSTER - PO 13

POSSIBLE CHANGE IN THE PARADIGM OF PATIENTS WITH SINGLE-KIDNEY AND COMMON ORIGIN WITH GENITAL ABNORMALITIES: ZINNER SYNDROME AND BICORNUATE UTERUS.

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Introduction:
Genital anomalies may be associated with renal abnormalities due to common embryological development (mesonephric or Wolffian ducts).
The Single Functioning Kidney (SFK) can associate ipsilateral cystic dilatation of the seminal vesicle in men (Zinner syndrome) and different uterovaginal malformations in women. These malformations are diagnosed incidentally in teenagers but may have implications for health and fertility.

Objectives:
The objective is to propose pelvic ultrasound screening in single-kidney patients for diagnosis of an entity that requires high clinical suspicion.

Methods:
We present 2 patients with SFK in whose follow-up (between 2022 and 2023) urological/gynecological abnormalities were detected.

Results:
Case 1. A 16-year-old male who underwent an uroCT scan for renal lithiasis, showing evidence of a retrovesical cystic lesion. Magnetic resonance imaging (MRI) of the pelvis is performed with confirmation of cystic hypoplasia of both seminal vesicles and dilation of the distal third of the right ureter, compatible with Zinner syndrome. He is referred to Urology, where an expectant attitude is decided since he is asymptomatic.
Case 2. A 18-year-old woman with a left SFK with scarring nephropathy due to reflux. Ultrasound control reveals a bicornuate uterus. She is referred to Gynecology, who recommends a study when there is reproductive desire.

Conclusion:
The association of SFK and reproductive organ involvement is known, but its incidence is still uncertain, since current guidelines do not establish an active search for this association.
Although most are asymptomatic, there are symptoms that can impair the quality of life of patients (urinary infections, recurrent testicular pain) and their early recognition could prevent situations of infertility.
The initial diagnostic test is an ultrasound, and since it is an affordable and inexpensive test, pelvic-ultrasound could be protocolized in adolescents with SFK. If there is possible suspicion, a certain diagnosis will be made using MRI.

Key words: single, kidney, zinner, mesonephric.

POSTER - PO 14

SÍNDROME DE ZINNER – UM DIAGNÓSTICO RARO COM IMPORTANTES CONSEQUÊNCIAS

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Introduction:
O Síndrome de Zinner (SZ) é uma doença congénita rara que resulta de uma perturbação no desenvolvimento embrionário do ducto mesonéfrico e se manifesta pela tríade: quistos da vesícula seminal, obstrução do ducto ejaculatório e anomalias do trato urinário superior ipsilateral, como a doença renal poliquística displásica ou a agenesia renal.

Results:
Adolescente 17 anos, acompanhado em consulta de patologia nefro-urológica pediátrica por rim único, diagnostico confirmado no período neonatal. Não apresenta sintomas urinários, hipertensão, proteinúria ou diminuição da taxa de filtração glomerular. Em ecografia de vigilância foi detetada agenesia da vesícula seminal esquerda. Por suspeita de SZ realizou RMN que confirmou o diagnóstico, apresentando: agenesia renal esquerda completa, acompanhada de agenesia da vesícula seminal esquerda e pequeno quisto periuretral (9x8mm). Mantém-se assintomático e com seguimento em consulta.

Conclusion:
O SZ é tipicamente assintomático em idade pediátrica, e o seu diagnóstico é frequentemente incidental ou realizado durante a investigação ou seguimento de outras patologias nefro-urológicas. Durante a adolescência, podem surgir sintomas relacionados o efeito de massa do quisto da vesícula seminal ou com a obstrução do ducto ejaculatório, sendo mais comuns a dor abdominal, pélvica ou perineal e sintomas urinários baixos.
Para além da vigilância da função renal, pressão arterial, proteinúria e hipertrofia renal compensatória, alertamos para a importância de equacionar o diagnóstico de SZ em crianças e adolescentes com rim único, com estudo ecográfico das vesículas seminais. Uma vez que pode estar associado a infertilidade, em até 45% dos adultos com este diagnóstico, deverá realizar-se um estudo da fertilidade quando é atingida a idade adulta. O tratamento cirúrgico está reservado aos casos sintomáticos, mas são necessários estudos para avaliar a pertinência da excisão precoce do ducto ejaculatório com o objetivo de preservar a fertilidade, sendo o diagnóstico atempado essencial neste sentido.

Key words: agenesia renal, doença renal poliquística, infertilidade

POSTER - PO 15

OHVIRA SYNDROME

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Introduction:
Abdominal pain is one of the most frequent reasons for consultation in paediatric emergency departments. Its possible aetiology is very variable depending on the age of our patients, with infant colic, gastroenteritis and viral infections standing out among the youngest and pneumonia, asthma and appendicitis becoming more important among adolescents, without ever leaving aside dysmenorrhoea and gynaecological pathologies as causes of abdominal pain at the end of the paediatric age group.

Results:
12-year-old patient who consulted the emergency department on several occasions for non-specific complaints. Regular, non-painful menarche started the same year. An abdominal ultrasound scan showed absence of the left kidney and a retrovesical collection of 11-12cm, a diagnosis of haematocolpos was made and the patient was admitted for assessment and drainage by gynaecology. Blind haemivagina, haematocolpos and 2 hemiuteri were confirmed by transvaginal ultrasound. Ultrasound examination of the urinary tract showed dilatation of the pelvis, calyces and complete ureter in the right kidney. The left kidney was not visible. Compatible with left renal agenesis and complete dilatation of the right kidney (reflux and stenosis of the bladder junction). Finally, MRI was performed: findings compatible with complete bicornuate uterus. She was diagnosed with Ohvira Syndrome and underwent surgery to maintain vaginal patency, asymptomatic since then.

Conclusion:
With this case we want to emphasise that we cannot leave aside gynaecological causes as a factor of abdominal pain in girls close to the onset of menarche, as we can overlook common pathologies such as dysmenorrhoea, endometriosis, surgical abdomens such as ovarian torsion or rarer cases such as this one, which also required surgery. This case has led us to rule out concomitant gynaecological disorders in all our single-parent patients.

POSTER - PO 16

REFLUJO VESICOURETRAL, IMPORTANCIA DE LA DISFUNCIÓN VESICAL.

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Introduction:
La disfunción vesical puede tener un papel clave en la patogenia del RVU y modificar de forma sensible su evolución. En niños con RVU y disfunción vesical las ITUs recurrentes son más frecuentes, la resolución del RVU se retrasa y es más común la aparición de daño renal.

Results:
Niña de 3 años con RVU izquierdo grado I e ITUs de repetición, deterioro de la función renal izquierda (FRD 20.5%) e importante aplazamiento de la micción. Se indica profilaxis antibiótica además de normas de uroterapia estándar para corregir la disfunción miccional. Al cabo de un año, sin presentar nuevas ITUs, se repite CUMS que evidencia aumento significativo del RVU (bilateral grado III permiccional y grado II izquierdo en fase retrógrada), así como la gammagrafía con discreto descenso de la función renal izquierda (FRD 19.54%). Se realiza estudio urodinámico que muestra actividad esfinteriana durante la micción, por lo que se le hace hincapié en el control de la disfunción vesical con ejercicios de entrenamiento del suelo pélvico/técnicas de biofeedback. No se consigue resolución del RVU y se realiza corrección endoscópica del mismo con inyección de Deflux bilateral. En la cistografía de control 5 meses después persiste RVU izquierdo grado III en fase permiccional, y por ello se somete a una segunda inyección de Deflux izquierda. Sin dejar de lado las normas de uroterapia y las técnicas de biofeedback, se repite la CUMS pasado un año que no evidencia RVU. La niña no consigue normalizar la función vesical, continua con importante aplazamiento de la micción y pasados 7 meses vuelve a presentar una ITU.

Conclusion:
Es fundamental valorar la función vesical en todo niño con RVU.
La disfunción vesical puede empeorar un RVU congénito.
La asociación de RVU, ITU y disfunción vesical es bien conocida.

Key words: Disfunción vesical, Reflujo vesicoureteral, infección urinaria.

POSTER - PO 17

HEART FAILURE IN END-STAGE RENAL DISEASE. SHOULD WE DO JUST A KIDNEY TRANSPLANT?

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Objectives:
Patients with chronic kidney disease are at increased risk of cardiovascular disease and this often manifests clinically like heart failure.

Methods:
We present a child with end-stage renal disease (ESRD) who developed a severe heart failure and her manage.

Results:
12 year-old girl referred (March/21) for evaluation and renal biopsy due to nephrotic syndrome, hypertension and microhematuria. No relevant personal history. Family history of focal segmental glomerulosclerosis (FSGS). Autoimmunity and infectious study were negative. Renal ultrasound showed increased cortical echogenicity. Renal biopsy demonstrated NOS-type FSGS. No response to oral prednisone. Genetic study revealed TRCP6 mutation. Cardiological evaluation in June 2021 was normal.
She presented a progressive deterioration of renal function and started peritoneal dialysis (October/2021) whereas waited for a deceased donor renal transplant.
In February/2022, she was admitted due to impaired functional class, hypertension and edema with anuria. Echocardiogram showed ventricular dilatation, severe ventricular dysfunction and mitral regurgitation. Ejection fraction (EF) 39%. Infectious screening and genetic for cardiomyopathies were negative. Cardiac magnetic resonance without gadolinium showed severe ventricular dysfunction with mitral regurgitation. Nt-proBNP >35000ng/L. Sacubitril/valsartan was started. Cardiac catheterization without contrast to measure pressure, resistance and cardiac output: no pulmonary hypertension; tolerated anesthesia well. She remained hypertensive despite treatment with sacubitril/valsartán, carvedilol, amlodipine and clonidine; started levosimendan.
In June/2022, kidney transplant was performed (3 missmatches, standart induction). Good renal allograf function. EF 24-hours after trasplantation 44%. The main complication was hypertension requiring up to seven drugs in the peritransplant period.
Current situation (February/2023): excellent graft function (creatinine 0.74mg/dL, no proteinuria). Blood pressure controlled with sacubitril/valsartán, carvedilol, amlodipine. Echocardiogram: improvement of left ventricule cardiac function. EF 60%. Nt-proBNP 66mg/L.

Conclusion:
Renal transplantation in the setting of cardiac dysfunction and the effect of renal transplantation on this progression remain poorly studied in children. In our case, recovery of cardiac function were observed after transplantation.

Key words: Heart failure, end-stage renal disease, kidney trasplant.

POSTER - PO 18

END-STAGE KIDNEY DISEASE WITH UNKNOWN ETIOLOGY - THE IMPORTANCE OF GENETIC TESTING – CASE REPORT

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Introduction:
The etiology of chronic and end-stage kidney disease in pediatrics is not always clear, and genetic testing can be a very important tool in some cases.

Objectives:
To discuss the complex etiology of chronic and end-stage kidney disease, as well as the importance of genetic testing.

Methods:
Case report.

Results:
An eight year-old girl was transferred to our pediatric nephrology tertiary center due to acute kidney injury (creatinine 2.8 mg/dL; uremia 154 mg/dL), hypertension stage 2 and nephrotic range proteinuria (urine protein/creatinine ratio 2.86). During the 6 months prior to this hospitalization, the patient presented recurrent vomiting, with no other accompanying symptoms. There was no relevant medical background neither familiar history of kidney disease. Kidney ultrasound was normal, and kidney pathology revealed “interstitial nephritis, sclerosed glomeruli and 1 crescent”. Genetic panel for glomerulosclerosis was requested, and the patient was discharged at the 10th day of hospitalization.
Due to progressive loss of kidney function, hemodialysis was started 5 months after first admission. Genetic testing revealed 2 variants in the TTC21B gene, 1 pathogenic and 1 with unknown significance, consistent with the diagnosis of nephronophthisis. Kidney transplant was performed 3 months after dialysis initiation, with normalization of kidney function (0.39 mg/dL) and proteinuria (urine protein/creatinine ratio 0.223), while blood pressure was controlled with 2 antihypertensives.

Conclusion:
In this case report, most probably both glomerular and tubular disorders, together with dehydration were combined for the development of rapid progressive kidney disease. Kidney transplant was successfully performed, with favorable outcome. Genetic testing was very important for the etiological diagnosis, requiring future counselling and family planning.

Key words: End-stage kidney disease, Genetics

POSTER - PO 19

DIFFICULT TO CONTROL ARTERIAL HYPERTENSION. TUMOR AS MAIN ACTOR

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Objectives:
Arterial hypertension (HT) is one of the main causes of morbidity and mortality worldwide.Its prevalence has increased mainly due to the epidemic of childhood obesity.Nevertheless,it continues to pose a diagnostic challenge due to the different clinical forms of presentation and the higher incidence of secondary hypertension in this population compared to adults.

Methods:
Present a case of HT secondary to a tumor process

Results:
14-year-old patient under follow-up by Neurology due to a flare-up of left optic neuritis(suspected relapsing-remitting multiple sclerosis).History of deceased father due to laryngeal cancer.She presented with acute headache and left fasciobrachiocrural hemiparesis with HT (194/127 mmHg).Cranial scan showed posterior bulbar hematoma.She was admitted to the ICU where perfusion urapidil was started,followed by nifedipine and oral labetalol.As etiological diagnosis:urine with mild proteinuria(iPr/Cr 0.56mg/mg);renal function,renal doppler ultrasound,echocardiography,thyroid,basal cortisol and ACTH were normal.Renin 63.6 IUU/ml, aldosterone 0.27 nmol/L.Catecholamines and metanephrines elevated in blood and urine.In view of these findings,the study was extended with MIBG scintigraphy,PET-CT dota,CT and abdominal MRI,showing a right retroperitoneal mass and another left one,as well as bone uptake at the atlas level.In view of the suspicion of metastatic paraganglioma,nifedipine was suspended and fenoxybenzamine was started.After achieving good blood pressure control,surgical excision was performed,confirming the diagnosis.Genetic study:Mutation in SDHD gene.Currently presents a good blood pressure control without drugs and a stable bone lesion.

Conclusion:
Paragangliomas are endocrine tumors derived from chromaffin cells of the autonomic nervous system.Given their low incidence in childhood,a high degree of suspicion is required and they should be considered in the differential diagnosis of difficult-to-control AHT.Metastases are present in >10%and cloud the prognosis.Prior to surgery,sequential blockade of alpha/beta adrenergic receptors is required,as well as good volume expansion to avoid intraoperative instability and fluctuation of blood pressure with tumor manipulation.30-50% are part of hereditary syndromes(especially multiple/bilateral forms),genetic testing should be considered in patients with a confirmed tumor.

Key words: Hypertension, Tumors, Paraganglioma

POSTER - PO 20

WHAT A “NORMAL” BLOOD PRESSURE HIDES.

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Introduction:
Pheochromocytoma is a rare disease (0.3 cases/1000000 inhabitants/year). Presentation is often sporadic although it can be associated with Von Hipple-Lindau, MEN2, neurofibromatosis type 1. Management involves blood pressure control and later surgical resection which can cause blood pressure (BP) lability and arrhythmias.

Results:
We report a 14-year-old patient transferred to our center for study of hypertension (170/110mmHg). He presented headache of 6 months of evolution, predominantly nocturnal, and on questioning associated palpitations, chest pain and sweating. At the beginning of the presentation he had seen his pediatrician where he presented normal BP (110/60mmHg) and was referred to neurology. Initial analytical study showed hypokalemia, creatinine 0.62mg/dl and other ions were normal, cranial CT showed signs of intracranial hypertension (HTIC) and ultrasound of the abdomen showed a right adrenal mass 4x5.3mm. Treatment with amlodipine was started and a study with catecholamines in blood (noradrenaline 8486ug/24h) and urine (vanillylmandelic acid 72mg/24h, homovaline acid 4.3mg/24h) was completed. An extension study was performed with SPET-CT and abdominal-pelvic MRI with a single uptake in the right adrenal gland. On admission he presented echocardiogram with diastolic dysfunction and moderate LVH, an ocular fundus with severe hypertensive retinopathy and brain MRI with signs of HTIC. BP control was controlled with phenoxybenzamine (maximum 0.88mg/kg/day), and before surgery propranolol 0.85mg/kg/day was prescribed, presenting significant orthostatism managed with fluidotherapy. During surgery, BP was regulated using esmolol and after removal of the mass, noradrenaline was started and maintained until 48 hours post-surgery due to hemorrhage. Normalization of BP at discharge (105/53mmgH) without treatment.

Conclusion:
Although pheochromocytoma is rare in children, we must always rule it out as the management is very specific. It is important to assess blood pressure repeatedly since catecholaminergic symptoms may be paroxysmal. Adequate control prior to surgery is essential to avoid complications during the intervention.

Key words: pheocromocitoma

POSTER - PO 21

AN UNUSUAL DEBUT OF POLYCYSTIC KIDNEY DISEASE

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Introduction:
Autosomal dominant polycystic kidney disease (ADPKD), with a prevalence of 1/400-1000 newborns, is usually asymptomatic until the fourth decade of life. The most common genetic mutation is in PKD1 and PKD2 gene. The incidence of arachnoid cysts is higher than in general population, but neurological tests are not routinely performed.

Objectives:
We present a patient with an atypical genetic variant and severe neurological involvement before diagnosis.

Results:
In 2016, a 12-year-old male presented hemiparesis and involvement of the left third nerve secondary to a chronic subdural hematoma associated with an arachnoid cyst of the left middle fossa. He required drainage and a subduro-peritoneal shunt valve. In 2022, in the context of headache and asthenia of one month's evolution, an abdominal evaluation of the shunt valve was performed, incidentally detecting multiple bilateral cortical cysts and a hepatic cyst compatible with polycystic kidney disease on ultrasound. A genetic study was carried out where we found involvement of the PKD1 gene with a mutation of uncertain clinical significance and of LRP5 associated with polycystic liver disease. In the cosegregation study, the 44-year-old mother carries the LRP5 gene mutation with renal cysts and without liver involvement. None of the brothers presented ultrasonographic renal cysts.
At present, our patient presents normal glomerular function; while his mother has a chronic kidney disease G2A1. No results of father's cosegregation study.

Conclusion:
- The presence of arachnoid cysts and subdural hemorrhage could be a guiding sign to detect polycystic kidney disease, which would go unnoticed for valuable time to preserve renal function.
- The mutation of the LRP5 gene associated with polycystic liver disease and with a better renal prognosis, could in some cases where mutations in other genes also coexist, not have such a favorable prognosis and with greater renal than hepatic involvement.

Key words: POLYCYSTIC KIDNEY DISEASE, PKD1/2, LRP5

POSTER - PO 22

HEREDITARY RENAL HYPOURICEMIA IN TWO SIBLINGS

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Objectives:
Hereditary renal hypouricemia (HRH) is a rare genetic disorder due to dysfunction of the tubular uric acid transporters, URAT1 and GLUT9, encoded by the genes SLC22A12 (Type 1) and SLC2A9 (Type 2) respectively. Most of the patients are asymptomatic and sometimes they can debut with complications such as nephrolithiasis, hematuria or acute kidney damage after intense physical exercise.

Methods:
We report two siblings afected by hereditary renal hypouricemia confirmed by genetic study.
Resultados
An 11-year-old boy, with history of prematurity (32 weeks), twin pregnancy (IVF) and ADHD under treatment. Weight and height in p10. Non-consanguineous parents with no known history. In a routine blood test, a serum uric acid level of 0.8 mg/dl is detected and confirmed. Renal function was studied. In an isolated urine sample without glycosuria, proteinuria, or hypercalciuria, the fractional excretion of uric acid being of 54%. Estimated GFR, acid-base balance, and renal ultrasound were normal. Family data are reviewed, the mother had normal uricemia figures. His sister shows sustained hypouricemia between 0.6-0.8 mg/dL with a fractional excretion of uric acid being of 40%. Weight and height in p25. Glycosuria, proteinuria, or hypercalciuria were not detected in an isolated urine sample. Estimated GFR, acid-base balance, and ultrasound were normal. Given the suspicion of renal hypouricemia, a genetic study of both siblings was carried out, and a homozygous variant in the SLC22A12 gene was detected. The mother presents the same variant in heterozygosity.

Conclusion:
All hypouricemia below 2 mg/dl should be studied. Molecular analysis of the SLC22A12 and SLC2A9 genes is important to confirm the diagnosis of HRH. Prevention of its complications is the main treatment.

Key words: HEREDITARY RENAL HYPOURICEMIA

POSTER - PO 23

AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE WITH SEVERE PHENOTYPE IN A NEWBORN

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Introduction:
·Polycystic kidney disease, autosomal dominant (ADPKD), is a genetic kidney disease that typically causes mild symptoms during early childhood and gradually worsens over time. However, there are cases where severe symptoms can appear early in life. Genetic testing in these cases can provide an explanation and help with family genetic counseling.

Results:
·Preterm female (week 34+3, weight 2850 g) with intrauterine finding of bilateral nephromegaly with hyperechogenic kidneys. History of chronic kidney disease in father's family. She presented early hypertension requiring high dose of ACE inhibitors and calcium channel blockers. Plasmatic creatinine went up to 1,5 mg/dl with decreasing tendency reaching 0,6 mg/dl at discharge with 25 days of life. In post-natal ultrasound polycystic kidney disease was confirmed: bilateral nephromegaly, up to 9 cm, absence of cortico-medullar differentiation and numerous small cysts widely distributed with predominance of subcortical location. The patient was found to have two heterozygous variants in the PKD1 gene: c.10405+5G>A mutation inherited from the father and c.6395T>G p.Phe2132Cys from the mother. Based on the clinical features and co-segregation of the variants found in PKD1, it was determined that the c.10405+5G>A mutation was the origin of the disease, while the p.Phe2132Cys mutation, being present in the other allele, was aggravating the patient's phenotype. The patient's condition progressed to chronic kidney disease, and she was placed on the preemptive deceased donor transplant waiting list at the age of 36 months.

Conclusion:
This case highlights the importance of early detection of polycystic kidney disease, particularly in high-risk families. It also emphasizes the value of genetic testing in providing a definitive diagnosis and guiding treatment decisions. This information could be useful in guiding genetic counseling for the patient's family members and in developing targeted therapies for polycystic kidney disease.

Key words: Polycystic Kidney disease, genetics, newborn

POSTER - PO 24

EVOLUTION OF ISOLATED MICROORGANISMS AND ANTIMICROBIAL SUSCEPTIBILITY PATTERNS IN URINE CULTURES OF PEDIATRIC POPULATION AT A SECONDARY UNIVERSITY HOSPITAL OVER 13 YEARS

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Introduction:
Urinary tract infections are one of the most prevalent infections in pediatric patients. Adequate empiric treatments require knowing the most prevalent microorganisms in our environment, and their susceptibility patterns.
Objectivos
Describe the susceptibility patterns of the most prevalent isolated microorganisms in pediatric population at our institution. Analyze antimicrobial susceptibility trends throughout the study period, including the impact of the COVID-19 pandemic in antibiotic resistance.

Methods:
Retrospective observational study on significant urine cultures at Hospital Universitario Príncipe de Asturias from January 2010 to September 2022 in pediatric patients under 16. Demographic variables and susceptibility data for each clinical isolate was retrieved from clinical records. We evaluated the susceptibility trends by differentiating three periods: pre-pandemic (years 2010-2019), pandemic (2020) and post-pandemic (2021-2022

Results:
4215 positive urine cultures included. The most frequent microorganism were Escherichia coli (68.9%), Proteus mirabilis (9.6%) and Enterococcus faecalis (6.6%).
Regarding the most frequently isolated microorganism, E. coli, the three oral antibiotics that showed higher susceptibility rates were fosfomycin(98.5%), cefuroxime(95.8%) and ciprofloxacin(92.4%). We found a significant increase in the resistance of amoxicillin/clavulanate(from a rate of 11.9% in the pre-pandemic period, through a 15.8% in the pandemic and up to 43.2% in the post-pandemic period, $p<0.001$).
Finally, regarding the overall susceptibility patterns including all strains and focusing in the post-pandemic period, fosfomycin was the most active antibiotic (94.9%), followed by cefuroxime (88.4%) and cefotaxime (88.4%). We found a significant increase of resistance through the three study periods for amoxicillin/clavulanate. Inversely, we found a decrease in the resistance rates for amoxicillin, first, second and third generation cephalosporins and trimethoprim/sulfamethoxazole.

Conclusion:
The most active antibiotics to be used as empiric therapy in our patients are fosfomycin, and second and third-generation cephalosporins.The COVID-19 pandemic impact in the antibiotic resistance has been marginal in our study population, except for a significant increase of the antibiotic resistance for amoxicillin/clavulanate.

Key words: Urinary tract infections, antibiotics, antibiotic, resistance, susceptibility

POSTER - PO 25

THROMBOTIC MICROANGIOPATHY (TMA) IN A PATIENT WITH ECMO SUPPORT

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Introduction:
Thrombotic microangiopathy (TMA) is a group of disorders characterized by damage to the vascular endothelium mediated by activation of the alternative pathway of complement primarily or secondary to environmental factors and/or triggers. The complement system is also triggered upon initiation of extracorporeal support (ECMO).
Objectivos
We present a case of TMA in a patient with ECMO support.

Results:
A 12-year-old girl, previously healthy. She was admitted to the PICU due to hypoxemic respiratory failure.During initial stabilization, severe cardiac dysfunction with cardiorespiratory arrest and subsequent initiation of ECMO.
On admission, progressive pancytopenia with nadir values of Hb 9.7 gr/dl, platelets of 55,000 x10^9/L and leukopenia of 200 x10^9/L, prior to ECMO. It was associated with oliguria, mild renal dysfunction (eGFR by Schwartz 73 ml/min/1.73m2), without hematuria but severe proteinuria. After PCR, anuria requiring renal replacement therapy.
Ultrasound with severe bilateral decrease in the doppler signal and exclusively in renal flow, being almost imperceptible at the arterial level. Presence of hemolysis data on extracorporeal support (decreased haptoglobin, increased LDH), decrease in C3 (33.9 mg/dl) and elevation of the soluble membrane attack complex (C5sb9) 694.77 mg/dl (standard 127-303). The clinical diagnosis of suspected TMA is established, and urgen empirical treatment with Eculizumab is started.
Subsequently, bilateral renal blood flow improved at 8 days, diuresis started at 2 weeks and withdrawal of support with CRRT at 6 weeks. ECMO support for 49 days. After removing extracorporeal support, a renal biopsy was performed, which confirmed the diagnosis of TMA. Molecular and genetic study of complement in progress.

Conclusion:
It is important to maintain a high diagnostic suspicion in incomplete cases of TMA and start treatment early to reduce morbidity and mortality and preserve renal function.
In the case that we present, renal failure with absence of renal blood flow was key for diagnosis and early initiation of treatment.

POSTER - PO 26

BONE PAIN, MUSCLE WEAKNESS AND HYPOPHOSPHATEMIA - A CASE OF TUMOR-INDUCED OSTEOMALACIA IN CHILDREN

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Introduction:
Tumor-induced osteomalacia (TIO) is a rare and frequently underdiagnosed paraneoplastic condition characterized by hypophosphatemia and inappropriately low or normal 1,25-dihydroxy vitamin D. It's usually caused by elevated levels of fibroblast growth factor 23 (FGF23), secreted by mesenchymal tumors typically very small and difficult to locate. A high index of suspicion is required to prevent diagnostic and treatment delays.

Results:
A previously healthy nine-year-old girl from São Tomé, presented with a one-year history of bilateral and symmetrical pain of the lower limbs with progressive gait impairment. She was transferred to Portugal for further investigation. She had normal stature (15th percentile) and there were no bone deformities. Lower limbs X-ray showed diffuse and symmetrical metaphyseal lucent areas and osteopenia. MRI revealed low-signal bands in all physeal plates. Laboratory workup revealed hypophosphatemia(1,7mg/dL), increased alkaline phosphate (1091U/L), with normal calcium (9,5mg/dL), PTH (72,1pg/mL) and 1,25-Vitamin D (30,50pg/mL), low tubular reabsorption of phosphate (TRP 46.9%), decreased TmP/GFR (0,94mmol/L) and increased FGF23 (284UA/mL, RR <230). Physical rehabilitation and supplementation with phosphorus and calcitriol were implemented, with progressive pain and gait improvement, but without an increase in phosphatemia. The diagnosis of TIO was considered and a PET(68Ga-DOTANOC) was performed, which didn't identify the tumor. As TIO remained the most probable diagnosis, Burosumab was started empirically, with a progressive increase of serum phosphate (3,0mg/dL), normalization of TmP/GFR, and great improvement of physical capacity.

Conclusion:
TIO is a debilitating disease characterized by a long diagnostic delay leading to metabolic disturbances and skeletal impairment. FGF23 production is usually associated with benign mesenchymal tumors however recent metanalysis revealed up to 10% of malignant histology. Although rare, TIO can occur at any age, so it should be suspected and looked at in cases where childhood inherited conditions cannot be demonstrated. Increasing awareness of TIO is crucial to decrease its diagnostic delay and clinical consequences.

Key words: hypophosphatemia, tumor-induced osteomalacia, fibroblast growth factor 23, burosumab

POSTER - PO 27

FIFTIETH ANNIVERSARY OF THE SPANISH ASSOCIATION OF PEDIATRIC NEPHROLOGY. BIBLIOMETRIC STUDY OF THE COMMUNICATIONS PRESENTED IN THE FIRST TEN MEETINGS

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Introduction:
Asociación Española de Nefrología Pediátrica (AENP) was founded on December 8, 1973.
Objectivos
On the fiftieth anniversary of its creation, we believe that it is a good time to investigate its origins and quantify the characteristics of the 275 communications presented in the ten first meetings.

Methods:
The contents of the first seven meetings were recorded from the abstracts published in the journal Anales Española de Pediatría and those of the last three were extracted from the communication books. The number of speakers and the names of the hospitals of origin (the last nomination written in the book of the X Meeting was collected) were noted and distributed in sixteen topics.

Results:
In the II Meeting (Sevilla 1975) nine papers were presented; the maximum number of presentations corresponded to the IX Meeting (n=65;Seville 1982). The number of signatories per paper ranged, in general, between four and six (most frequent: five authors; 28.7%). The hospitals that presented the most communications were Vall d'Hebrón, Barcelona (n=34;12.4%), San Juan de Dios, Barcelona (n=28;10.2%), Hospital Provincial, Madrid (n=26), La Paz, Madrid (n=22) and Hospital Infantil de la Seguridad Social, Bilbao (n=20). The eleven communications of the I Meeting (Madrid 1974) were presented by members of six hospitals. At the end of the X Meeting (Tenerife 1983), papers from 26 different Spanish hospitals had already been heard. The most frequent topics were tubulopathies (n=55;20%; especially, Bartter and tubular acidosis), glomerulopathies (n=36;13.1%; more common, Schönlein-Henoch), nephrotic syndrome (n=26), neonatal nephrology (n=22), CAKUT malformations (n=22; priority was vesicoureteral reflux) and chronic renal failure (n=22; hemodialysis was the most common).

Conclusion:
The review of the contents of these meetings is enriching. In them, major papers on many common diseases and topics in pediatric nephrology were presented. In fairness, the effort of the pioneers of the AENP must be recognized.

Key words: History, pediatric nephrology

POSTER - PO 28

RENAL EXPRESSION OF BERARDINELLI-SEIP CONGENITAL LIPODYSTROPHY

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Introduction:
Berardinelli-Seip congenital lipodystrophy is a rare metabolic disorder characterized by leptin deficiency, as well as other lipodystrophies, which is associated with a severe form of metabolic syndrome. It is an autosomal recessive disorder usually diagnosed at birth or in the first year of life.

Results:
We report a case of this form of lipodystrophy in a 17-year-old Chinese male who was diagnosed at the age of four months because of arterial hypertension and severe obstructive hypertrophic cardiomyopathy, confirmed genetically (BSCL2 gen). He was treated with propranolol until twelve months of age with clinical improvement, so it was stopped afterwards. Because of his return to China, the follow up was lost until he was seven years old. He presents the classic phenotype such as severe generalized muscular hypertrophy and absence of adipose tissue, umbilical hernia, signs of insulin resistance like acanthosis nigricans, hepatomegaly, concentric hypertrophic cardiomyopathy and xanthomas. Actually, he is in a regular medical multidisciplinary pediatric care (endocrinology, cardiology, gastroenterology, neurology, nephrology, otolaryngology, ophthalmology). About nephrological symptoms, he started the follow up because of arterial hypertension but in the evolution the red flag was bilateral nephromegaly (at the age of nine, size at length > 90 percentile) and glomerular protein loss (protein/creatinine 1500 mg/g; albumin/creatinine 1100 mg/g; ß-2-microglobuline negative), normal renal function (eGFR 117 ml/min/1,73m2) and urinary sediment, negative autoimmunity study. Due to beta-adrenergic antagonist and angiotensin-receptor blocker, both the high blood pressure as well as the proteinuria were reduced to normal values. Now he maintains blood pressure close to 50-75 percentile and very low albuminuria (protein/creatinine 134 mg/g and albumin/creatinine 65 mg/g).

Conclusion:
Finally, highlight that it has been described in literature patients with chronic kidney disease related to diabetic nephropathy as well as its association with focal segmental glomerulosclerosis and membranoproliferative glomerulonephritis.

Key words: Berardinelli-Seip Congenital Lipodystrophy, Renal expression

POSTER - PO 29

HYPERCARBIA IN HOSPITALISED CHILDREN AND ADOLESCENTS WITH ANOREXIA NERVOSA AS A PREDICTIVE MARKER FOR READMISSION: A PROSPECTIVE STUDY

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Objectives:
Anorexia nervosa (AN) is a severe eating disorder (ED) with multifactorial causes and important psychiatric and organic comorbidities. AN-associated mortality ranges from 2 to 8%, which is the highest mortality rate for a psychiatric disorder, mainly because of suicide and organic complications.
Readmissions in AN are a common though understudied outcome. No organic alterations have been related to the likeliness of readmission to date. This study evaluated clinical and laboratory alterations associated to the risk of AN-related hospital readmissions in children and adolescents.

Methods:
The aim of this study was to assess clinical and laboratory alterations associated to the risk of AN-related hospital readmissions in children and adolescents.

Results:
A prospective study was performed with every patient ≤18 years old admitted due to AN destabilization to the Eating Disorder Ward of a freestanding children's hospital in Madrid (Spain) from November 2018 to October 2019. Both subtypes of AN were included. The participants were evaluated upon admission, at discharge and six months after discharge.
154 patients were admitted during the study period. 131 met the inclusion criteria. Median age was 15.1 years (interquartile range 13.5-16.4). 71% of participants were malnourished at admission. 33 participants (25 %) had been previously admitted due to an eating disorder.

Conclusion:
Hypercarbia and respiratory acidosis are frequent findings in children and adolescents admitted due to AN destabilization. This anomaly generally persists for at least 6 months after discharge, despite clinical amelioration. Venous blood gases alterations should be thoroughly considered when reassessing physiological impact of AN, as pCO2 elevation at discharge is associated with higher odds of readmission. To our knowledge, no laboratory alterations had been previously reported as potential indicators of readmission.

Key words: Anorexia nervosa, Feeding and Eating Disorders, Medical Complications, Respiratory Acidosis, Hospital Readmission, Acid-Base Imbalance

POSTER - PO 30

NUTCRACKER SYNDROME

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Introduction:
Nutcracker syndrome is caused by compression of the left renal vein between the aorta and superior mesenteric artery.
If the compression of the left renal vein originates between the aorta and the vertebral body, it is called posterior nutcracker syndrome.
The clinical manifestations are unspecific, with the possibility of finding unspecific alterations in the sediment
(hematuria and/or proteinuria), abdominal pain or left varicocele.
Its prevalence is unknown and there may be asymptomatic cases.

Objectives:
We present a series of patients under follow-up in our hospital

Methods:
We review the pacient histories with radiological diagnosis

Results

Age/Gender	9 years/ F	14 years/M	11 years / M	14 years / F
Weight/Height/BMI	33 Kg/139 cm/17	57 kg/174 cm/18.8	38 kg/151 cm/16.7	37 kg/151 cm/16.5
BP	110/55	106/69	102/69	96/51
Symptoms	Gross Hematuria	Persistent proteinuria LeftS varicocele	Gross hematuria with flank pain	Microhematuria with left lumbar pain

POSTER - PO 30

9y, F	Angio SCAN:decrease in caliber of the left renal vein passage between the abdominal aorta (AA) and the outflow of the superior artery mesenteric (SAM) ;compression ratio 3.1 (NV: < 2.25), aorto angle-SAM 25-30ª (NV > 38ª)
14y,M	Angio SCAN: Left renal vein that crosses the aortic angle mesenteric (AAM):AAM 10º . Distance between AA and SAM 3mm.
11y,M	US: AAM 25ª. Distance between AA and SAM 3-4 mm.
14y,F	US: AAM 21ª; Distance AA-SAM 3 mm

Conclusion:
Microhematuria is a frequent clinical sign of follow-up in our patients in pediatric nephrology clinics, without an etiological
diagnosis in a high percentage.
Nutcracker syndrome is a rare cause of hematuria.
Ultrasound and SCAN can be diagnostic techniques.
It usually does not require specific treatment since physical development increases the deposit of fatty and fibrous tissue at the origin
of the superior mesenteric artery, and decreases compression of the left renal vein.

Key words: NUTCRACKER SYNDROME, HEMATURIA, PROTEINURIA, VARICOCELE

POSTER - PO 31

CASES OF NUTCRACKER SYNDROME IN A TERTIARY HOSPITAL

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Introduction:
Hematuria is a common finding in pediatrics, often without a specific etiology. Orthostatic proteinuria accounts for more than 60% of asymptomatic proteinuria in children and is more prevalent in tall teenage boys with lumbar hyperlordosis. Nutcracker syndrome is a rare cause of hematuria and an important cause of orthostatic proteinuria, characterized by compression of the left renal vein between the aorta and superior mesenteric artery. It can present with abdominal pain, varicocele, dyspareunia, dysmenorrhea, and asthenia. Nutcracker syndrome is associated with a decrease in body mass index due to reduced retroperitoneal fat and aortomesenteric angle.

Objectives:
To describe the epidemiology, symptoms, diagnosis, and management of nutcracker syndrome cases diagnosed in our tertiary hospital over the last 5 years.
Métodos
Medical records and tests of the three patients diagnosed with nutcracker syndrome in the last 5 years in our hospital were reviewed.

Results:
The average age at diagnosis was 10 years, with a male-to-female ratio of 2:1. Two patients presented with proteinuria, and one with persistent microhematuria. Two of the patients had a BMI <15 with weight percentiles p7 and p18, respectively, and one of them presented with constitutional symptoms prior to proteinuria diagnosis. Magnetic resonance angiography confirmed the diagnosis in all three cases. Only one patient required treatment with ACE inhibitors.

Conclusion:
Orthostatic proteinuria is a benign condition, and its proper diagnosis can avoid additional tests and invasive interventions. Nutcracker syndrome is an infrequent but important cause of hematuria and orthostatic proteinuria that should be suspected in appropriate clinical settings. A renal doppler ultrasound or magnetic resonance angiography can confirm the diagnosis. Management strategies vary from spontaneous resolution to treatment with ACE inhibitors, surgical intervention, or stent placement, depending on the severity of the syndrome

Key words: Hematuria, Nutcracker syndrome, orthostatic proteinuria

POSTER - PO 32

DOUBLE NUTCRACKER SYNDROME WITH "BENIGN" LONG-LASTING PROTEINURIA. HOW MUCH SHOULD WE WAIT?

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Introduction:
The nutcracker syndrome (NCS) is the set of symptoms produced as a result of the compression of the left renal vein (LVR), which usually occurs between the aorta and the superior mesenteric artery (anterior NCS). Posterior NCS is defined as the compression against spine.
It may presents with a wide variety of symptoms. Although macroscopic hematuria is the most frequent symptom, orthostatic proteinuria can be the only manifestation.
The most of cases resolves completely with conservative management. Surgery should only be applied for persisting or severe clinical manifestations.

Results:
Women 14 years old followed up because of persistent isolated proteinuria. It was a casual diagnostic in urine analysis in a fever episode when she was 4 years old.
There were not personal or familial relevant background.
Along the time, proteinuria persists being selective glomerular, under nephrotic range.
Not other alterations in renal function were found.
Orstostatism tests were performed in several times, being negative. Doppler ultrasound showed NCS, meeting pathological criteria of angle and venous outflow ratios and diameter .
The patient remained asymptomathic, normotensive and with normal glomerular filtration.
A conservative approach with weight gain was recommended, but proteinuria not modified. Then, treatment with angiotensin-converting enzyme inhibitor and subsequently, angiotensin receptor blocker was initiated, disrupted because of side effects (dizziness).
As the proteinuria persists for 10 years, rule out differential diagnoses were performed. Abdominal scan showed double NCS with circumaortic LVR. Collateral circulation neither other complications were found.
As spontaneous improvement or even complete resolution is widely described in long-term studies, and the stable condition of our patient, surgery it has not been considered by the moment as a therapeutic option.

Conclusion;
Double anterior and posterior NCS because of circumaortic LRV is extremely rare. We raise hypotheses that this variant can predispose to a greater amount and long-lasting proteinuiria.

Key words: Nutcracker syndrome, persistent proteinuria

POSTER - PO 33

SCHISTOSOMIASIS: A RELEVANT CAUSE OF HEMATURIA DUE TO MIGRATION AND GLOBALIZATION.

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Introduction:
Human schistosomiasis is the parasitic disease with the highest morbidity and mortality worldwide after malaria. At least 90% of people affected by bilharziasis live in Africa. However, the number of imported cases in Western countries has increased due to the arrival of a significant number of migrants from endemic regions. Schistosoma haematobium must be screened in patients with non-glomerular hematuria with positive epidemiological backgrounds.

Results:
A nine-year-old child, born in Spain, whose parents are from Mauritania, was attended because of gross hematuria (intermittent fresh blood drops at the end of diuresis). Moreover, he presented mild left testicular pain for two weeks and sequentially a self-limited periumbilical pain. His blood pressure was within the normal range and had no edema.
Six months ago, he had traveled to Mauritania. On arrival in Spain, he presented with generalized skin lesions with ulceration and indurated margins. Microbiological studies were negative. He was treated with cefadroxil with good evolution.
The blood test showed a GF of 130 ml/min/1.73m2, hemoglobin 10.6 g/dL, without eosinophilia or thrombopenia. Urinalysis revealed erythrocytes, and urine protein-creatinine ratio was 0.22. Urine culture was negative. Doppler ultrasonogram of the abdomen and kidneys was solicited. Finally, based on the epidemiological background, schistosomiasis was screened. The microbiology department recommended the collection of three urine samples on different days between noon and 3 pm after physical activity, finding Schistosoma haematobium eggs in all samples. He started with praziquantel.

Conclusion:
The classical sign of urogenital schistosomiasis is hematuria. Bladder and ureter fibrosis and kidney damage are usually diagnosed in chronic parasitism. Long-term irreversible consequences such as bladder cancer and infertility may occur in the later stages.
It is important to highlight personal backgrounds, including epidemiological history. For the diagnosis, the urine samples must be taken properly. Praziquantel is safe and effective.

Key words: Hematuria, Schistosoma, Gross hematuria, Schistosoma haematobium

POSTER - PO 34

DEL OJO AL RIÑÓN, EXPERIENCIA EN RECAIDA DE UN TINU.

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Introduction:
El síndrome de nefritis túbulo-intersticial aguda y uveítis (TINU) es una entidad rara (predominante en mujeres). Tiene una etiología multifactorial y suele evolucionar favorablemente, aunque las recaídas son probables.

Results:
Mujer de 10 años derivada por hiperemia, fotofobia y dolor en ojo derecho desde hace 3 días. Refiere episodios similares en ojo izquierdo en los últimos 2 meses. Es valorada por oftalmología con sospecha de uveítis. Completan estudio objetivando alteración de la función de renal (Creatinina 1,26 mg/dL). Inicia seguimiento en reumatología y nefrología con aumento de B2-microglobulina (máxima 24000 microgramos/24 horas) y creatinina estable. Se inicia corticoterapia oral (1 mg/kg) al mes del inicio de la sintomatología. Se realiza biopsia renal donde se objetiva nefritis túbulo-intersticial aguda con focos de necrosis tubular. Rápida estabilización de función renal (Creatinina 0,75 mg/dL) sin datos de afectación ocular. Al mes de iniciar corticoterapia se instaura pauta de descenso. A los 8 meses de iniciado el decalaje (0,5 mg/kg/48 horas) sufre empeoramiento clínico con focos de vasculitis ocular. Se realiza control analítico con aumento de B2-microglobulina (843 microg/24h) por lo que se instaura dosis terapéutica a 1 mg/kg y se asocia micofenolato mofetilo como ahorrador de corticoides en relación con fenotipo marcadamente cushingoide y aumento de 10 kg. En esta segunda pauta de tratamiento presenta rápida mejoría ocular con lento descenso de niveles de B2-microglobulina (creatinina estable). Se consigue práctica normalización de B2- microglobulina a los 5 meses. Precisa derivación a psicología infantil a causa de su cambio físico.

Conclusion:
EL síndrome TINU cursa con afectación ocular y renal. El tratamiento se basa en corticoterapia que debe mantenerse hasta estabilización clínica con una pauta descendente lenta. No existe pauta estandarizada en caso de recaídas.
En caso de tratamiento prolongado es importante descartar efectos secundarios de la corticoterapia; valorando asociar ahorradores de corticoides si fuera necesario.

Key words: nefritis túbulo-intersticial, uveítis, recaída, micofenolato mofetilo

POSTER - PO 35

SARS-COV-2 INFECTION AND TUBULOINTERSTITIAL NEPHRITIS AND UVEITIS (TINU) SYNDROME

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Introduction:
It is firmly established that severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) has a characteristic kidney tropism, but tubulointerstitial nephritis (TIN) is not a common association with this virus. We present a patient with TIN and uveitis (TINU syndrome) where SARS-CoV2 spike protein was identified in the kidney tissue.

Results:
A 12-year-old girl was assessed for an elevation of serum creatinine, anorexia, vomiting, weight loss, and asthenia following a febrile respiratory infection with no known infectious cause several weeks before. Data of incomplete proximal tubular dysfunction (hypophosphatemia and hypouricemia with inappropriate urinary losses, low molecular weight proteinuria, and glucosuria) were also associated. Eight weeks after the onset of symptoms, the patient tested positive in PCR for SARS-CoV-2 (Omicron variant). Subsequently, a kidney biopsy revealed TIN and immunofluorescence staining with confocal microscopy detected the presence of SARS-CoV-2 protein S within the kidney interstitium. Steroid therapy was started with gradual tapering. After ten months of follow-up, a routine ophthalmological examination revealed asymptomatic bilateral anterior uveitis, and a second kidney biopsy was performed, without demonstrating acute inflammation or chronic changes, but SARS-CoV-2 protein S within the kidney interstitium was again detected.

Conclusion:
We report a patient where SARS-CoV2 spike protein was observed in kidney tissue several weeks and ten monthsfollowing the onset of a clinical manifestations corresponding with TINU syndrome. Although simultaneous PCR for SARS-CoV-2 infection could not be demonstrated, we hypothesize that SARS-CoV-2 could have been involved in tiggering and clinical course of the patient's disease

Key words: SARS-COV-2, TUBULOINTERSTITIAL NEPHRITIS, TINU

POSTER - PO 36

THE IMPORTANCE OF MAGNESIUM DETERMINATION IN THE ED.

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Introduction:
The clinical picture of hypomagnesaemia varies from the asymptomatic patient, anorexia, nausea, vomiting, lethargy, weakness, personality changes, tetany, tremors and muscle twitching and even generalised tonic-clonic seizures in cases of severe hypomagnesaemia. In familial hypercalciuric hypomagnesaemia with nephrocalcinosis (HHFNC) we find hypomagnesaemia of renal origin with nephrocalcinosis with almost invariable evolution towards renal failure.

Results:

	1	2
Age	17 months	4 years
AP	Normal metabolic screening. Psychomotor retardation. Congenital bilateral cataract	Normal metabolic screening. Normal psychomotor development. Left unilateral horizontal nystagmus from with normal RMN.
Clinic (debut)	22 days of life had seizures	Asymptomatic
Analytics (debut)	Ca 5.9 Mg 0.9. pH 7.37 pCO2 40 HCO3 23.1	Ca 9.5 Mg 1.1 7.360 pCO2 49 HCO3 27.7 mmol/L
Renal function and blood gases (current)	FGe Schwartz 83.41 ml/ min/ m2. ERC G2A3 Ca 11 Mg 1.6 PTH 97.1 pg/ml pH 7.520 pCO2 23 mmHg HCO3 22.4	FGe. Schwartz 85.84 ml/ min/ m2. ERC G2A2. Ca 10.1 Mg 1.4 PTH 61 pg/ml pH 7.32 pCO2 45 mmHg HCO3 22.8
Renal ultrasound (current)	Both kidneys with diffuse nephrocalcinosis.	Both kidneys with medullary nephrocalcinosis.
Genetics	Claudin 19. Mutation in exon 4 Heterozygous carrier parents	Claudin 19. Mutation in exon 4. Heterozygous carrier parents
Others pathologies	HTA in treatment with amlodipine. failure to grow Non-IgE mediated cow's milk protein allergy. Dehydration cataract	Bilateral nigtasmus. With congenital high myopia, bilateral macular scars and right coloboma.
Treatment	Oral supplements	

POSTER - PO 36

Conclusion:
The determination of serum magnesium is not performed urgently in all laboratories, and its suspicion is particularly important in cases of hypocalcemia refractory to treatment, as was the case in our first patient. Thanks to this genetics diagnosis, the second case was identified. Symptomatic treatment, with abundant fluid intake, sodium restriction, control of hyperparathyroidism, does not, as a rule, slow the progression of CKD towards end-stage renal failure.

Key words: familial hypercalciuric hypomagnesaemia with nephrocalcinosis (HHFNC)

POSTER - PO 37

UVEITIS AND RENAL INVOLVEMENT: AN UNUSUAL CASE.

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Introduction:
Since its description in 1975 by Dobrin et al., approximately 500 hundred cases of tubulointersticial nephritis and uveitis syndrome (TINU) have been published. It mainly affects adolescent women and its etiology is unknown. Diagnosys of this syndrome is based on clinical, analytical and histologycal criteria of bilateral uveitis and tubulointersticial nephritis (TIN).

Objectives:
To carry out an analysis and review of an infrequent entity through a clinical case.

Methods:
A summary of the clinical history and a review of literature related to TINU has been performed.

Results:
We present the case of a 10-year-old male patient, admitted to the Emergency department with a 2-week history of eye redness and photophobia, without any other relevant symptom. The ophtalmological examination revealed signs of bilateral acute anterior uveitis (Tyndall effect, posterior synequiae and mutton-fat keratic precipitates). After initiating topical treatment with corticosteroids and cycloplegics, an analytical study was carried out, in which an alteration in renal function including elevation of alfa-1-microglobulin was detected.
Pediatric nephrology monitoring was started and broader analytical tests were undergone. After ruling out other infectious, inflammatory and autoimmune diseases, treatment with oral prednisone was started, with TINU as the main clinical suspicion. Given the persistence of renal involvement and incomplete Mandeville criteria, it was decided to perform a renal biopsy, which confirmed TIN. Due to exacerbations coinciding with the decrease in prednisone dose and side effects related to corticosteroid therapy, mycophenolate-mofetil was started. Currently, renal function has stabilized and uveitis symptoms have resolved.

Conclusion:
TINU is a rare syndrome, although it is estimated that one in 50 cases of bilateral uveitis in children is due to this entity. Most data related to TINU come from small case series, so more studies are necessary to establish management guidelines for patients.

Key words: bilateral uveitis, tubulointersticial nephritis, tubulointersticial nephritis and uveitis syndrome, TINU, case report

POSTER - PO 38

IMPORTANCE OF SENSORY INTEGRATION IN PEDIATRIC NEPHROLOGY.

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Introduction:
Voiding disorders are commonly encountered in Pediatric Nephrology, and functional disorders constitute an essential part of them. Proper identification, clinical history, physical examination, and tests are necessary to rule out organic pathology in some cases. The consequences of these disorders can be multiple, such as urine infection, vesicoureteral reflux, secondary uropathies, and social impact.

Objectives:
To emphasize the importance of appropriate detection and treatment of voiding disorders associated with sensory disorders.

Methods:
We conducted a bibliographic research and reviewed the medical records of two patients diagnosed with voiding disorder and sensory disorder. We also studied the interventions received by occupational therapy.

Results:
We describe two cases of girls aged 5 and 6, who consulted for pollakiuria and vesical tenesmus, with enuresis appearing in one of them. Both patients presented methodical behavior, with some manias and tactile hypersensitivity. Tests performed were normal, but sensory disorder was suspected, and adequate urinary hygiene guidelines were given. Both were referred to occupational therapy, and a progressive clinical improvement was observed, with complete disappearance of symptoms in one patient.

Conclusion:
Sensory integration is an important issue to explore in pediatrics as it can interfere with the basic activities of daily life and the correct psychomotor development of our patients. Thus, adequate referral to occupational therapy when we suspect alterations is crucial. The occupational therapist plays an essential role, analysing the patient's participation in toileting routines, hypothesizing about the factors involved in the challenge of participating in this activity, and creating individualized interventions that maximize participation. The most used therapies in these cases are those based on reactivity and sensory perception/discrimination, ranging from symbolic play to stimulation with different textures in case of tactile hypersensitivity.

Key words: functional disorders, voiding disorders, sensory disorders, occupational therapy, sensory integration

POSTER - PO 39

GLOMERULAR FILTRATION RATE ESTIMATION IN HOSPITALISED CHILDREN AND ADOLESCENTS WITH ANOREXIA NERVOSA: A PROSPECTIVE STUDY

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Objectives:
Although eating disorders (ED) are mental health disorders, they cause serious physical comorbidities. The consequences of anorexia nervosa (AN) and other ED on kidney function in children and adolescents have been scarcely reported. Chronic damage on glomerular filtration in malnourished patients is firmly established. In AN patients, 5 % of them will develop end-stage kidney disease after 20 years of evolution of AN. This complication is even more prevalent in patients with purging behaviours. However, estimation of glomerular filtration rate (GFR) in children and adolescents diagnosed of ED are challenging, as standard methods are ofter inadequate for these patients.

Methods:
The objective of this study was to assess the GFR of children and adolescentes admitted due to ED complications and to determine if there are differences between the different approaches for GFR estimation.

Results:
A prospective study was performed in the Eating Disorder Ward of a public-funded freestanding children's hospital in Madrid (Spain). Participants were recruited from November 2018 to October 2019. 172 admissions were included in this study.
Serum creatinine, serum cystatin C and 24-hour urine creatinine were measured upon admission, at discharge and 6 months after discharge. GFR was calculated and compared by using 10 paediatric-validated equations that included serum creatinine, serum cystatin C or both.

Conclusion:
Creatinine-based equations showed lower and more heterogeneous GFR in children and adolescents admitted due to AN when compared to cystatin C and combined equations. Up to 50 % of patients showed a GFR < 90 mL/minute/1,73 m2 upon admission. Further research is required to determine the best method to estimate GFR in this population.

Key words: Glomerular Filtration Rate, Anorexia nervosa, Feeding and Eating Disorders, Chronic Kidney Disease, Cystatin C, Creatinine

POSTER - PO 40

HALLAZGO CASUAL DE HTA E INSUFICIENCIA RENAL EN PACIENTE QUE CONSULTA POR ENURESIS NOCTURNA

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Introduction:
La HTA en niños tiene una incidencia creciente y a menudo está infradiagnosticada. Se define HTA como valores de PA >p95 según edad, sexo y talla, en 3 o más ocasiones. Cuanto menor es la edad del niño y más altos los valores de TA, es más probable que la causa sea secundaria.

Methods:
Paciente de 7 años que consulta en Nefrología por enuresis nocturna primaria, con fracaso tras 2 meses con desmopresina.
En la exploración, obesidad con IMC de 25.4 > p99 y TA 144/90 mm Hg (>p99) y en la ecografía en consulta, riñones hiperecogénicos e indiferenciados con aspecto de nefropatía médica.
Se solicita MAPA que confirma HTA. Se realiza estudio etiológico y de repercusión orgánica destacando en analítica proteinuria en rango nefrótico: 1.129 g/ 24 horas y función renal disminuida con filtrado glomerular de 27 ml/min. En la ecocardiografía destaca hipertrofia ventricular secundaria a la HTA. Se decide ingreso para completar estudio y realizar biopsia renal.
El informe anatomo-patológico de biopsia renal describe lesión de esclerosis segmentaria sin depósitos inmunes.
En el perfil de autoinmunidad destaca: Ac(IgG) anti SSA/Ro-52 (DOT) ** 22. Estudio serológico negativo excepto Ig G VHH tipo 6, VEB y VHA.
Durante el ingreso se inicia corticoterapia, inicialmente con bolos iv durante 3 días a 1 g/día, 21 días a 2 mg/Kg/día estando actualmente con 2 mg/Kg cada 48 horas y disminución de la proteína a 0.8 g/24 horas.

Conclusion:
Aunque la hipertensión arterial secundaria a enfermedad renal crónica era la más frecuente en niños, hay un aumento de hipertensión primaria asociada a obesidad y sedentarismo.
El diagnóstico de HTA en la infancia es incidental, por lo que es importante el cribado de HTA para detectar precozmente HTA silente.
La MAPA es útil para descartar HTA de bata blanca, evaluar la gravedad y eficacia del tratamiento.

Key words: HTA, INSUFICIENCIA RENAL

POSTER - PO 41

SPONTANEOUS PERIRENAL URINOMA IN A CHILD

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Introduction:
Urinoma is an encapsulated collection of extravasated urine into the perirenal space. The main causes are renal trauma and accidental perforation of the collecting system during surgical procedures; they rarely occur due to congenital obstruction of the urinary tract or spontaneously.

Methods:
Clinical case description

Results:
A previously healthy 11-year-old boy, a tourist, came to emergency department due to intense pain in the left flank (LF) of 2 hours' evolution, without fever or previous trauma. Physical examination with regular general condition, abdomen with intense pain in LF. Blood tests highlight leukocytes count 18,200 /µL, neutrophils 93%, haemoglobin 13 g/dL, C reactive protein 22.31 mg/dL, serum creatinine 1.17 mg mg/dL (glomerular filtration rate-GFR- 54 mL/min/1.73m^2). Urine analysis was normal. Abdominal ultrasonogram showed left kidney (LK) with grade IV pyelocaliceal dilatation without ureteral dilatation, hypoechoic fluid accumulation in perirenal space measuring 11.4 x 6.4 cm, suggestive of ureteropelvic junction obstruction (UPJ-O) complicated with perirenal urinoma.
Abdominal computed tomography (CT) scan revealed LK with severe pyelocaliceal dilatation due to UPJ-O; fluid accumulation in the posterior-inferior portion of the perirenal space (310 ml), inferior fornix rupture with urinoma open to the peritoneal space, moderate ascites. Left pleural effusion of moderate quantity.
Percutaneous nephrostomy was performed, draining copious hematuric urine; received intravenous antibiotic treatment. Control creatinine 0.68 mg/dL (93 ml/min/1.73m2); negative urine and blood cultures. Transferred to his country in stable condition.

Conclusion:
Obstructive congenital anomalies are the most common cause in neonates and infants, but in older children the most common obstructive cause is UPJ-O secondary to ureteral compression, inflammation, retroperitoneal fibrosis, lithiasis, scar tissue.
CT scan is the test of choice for diagnosis and serial ultrasonogram for follow-up. Management is usually conservative in smaller urinomas, reserving drainage or percutaneous nephrostomy for complicated cases. Its adequate management is essential for the functional prognosis of the affected kidney.

Key words: urinoma, urinary tract obstruction

POSTER - PO 42

ACUTE PYELONEPHRITIS OR RENAL ABSCESS? – A CASE REPORT

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Introduction:
Urinary Tract Infections are an important pathology in paediatrics. Pyelonephritis can cause renal scarring, hypertension, and chronic kidney disease. Renal abscesses can arise as a possible complication of urological infections, or due to hematogenous dissemination.

Results:
We present the case of a 6-year-old child, without significant medical history, who went to the Emergency Department with a fever with 24h of evolution, temperature of 39.5°C, apyrexia every 4h, good response to antipyretics, and uncontrollable vomiting, without genitourinary symptoms. There was no ingestion of suspicious food, epidemiological context, respiratory/gastrointestinal symptoms. On examination, had a preserved general condition, sunken eyes, dry lips, but hydrated mucous membranes. After ondansetron, he started intravenous fluid therapy. Laboratory tests revealed leukocytosis (30900uL) with neutrophil predominance (86.2%), urea/creatinine 42/0.6mg/dL, C-Reactive Protein of 168.20mg/L. Due to an episode of urinary frequency, urine was collected, revealing leukocyturia and active urinary sediment and a first ultrasound without any changes, so the patient started amoxicillin+clavulanic acid 50mg/kg/dose 8/8h IV. Urine culture later isolated Enterococcus faecalis multisensitive. Due to persistent fever under antibiotic therapy, he repeated the ultrasound that identified a "streaking in the right renal parenchyma, with hypo/hyperechogenic areas, prominent in the lower 2/3, anechoic area of 5mm, with the hypothesis of liquefaction". After discussing the case, considering the possibility of renal abscess, the therapy was changed to ceftriaxone 75mg/kg/day IV for 2 weeks. A follow-up ultrasound showed resolution of the abscess, and the patient completed the rest of the treatment orally at home.

Conclusion:
Renal abscesses are severe diseases depending on the extent and risk factors of each host. If there are anatomical anomalies predisposing to infection, pyelonephritis is associated with an increased incidence of renal abscesses. The authors would like to emphasize the importance of considering a renal abscess whenever there is no response to treatment or clinical worsening.

Key words: Acute Pyelonephritis, Renal Abscess, Urinary Tract Infections

POSTER - PO 43

WHAT IS HIDDEN BEHIND ENURESIS?

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Introduction:
Ciliopathies are a group of rare genetic disorders that occur due to dysfunction of primary cilia and can affect multiple organ systems, to which Bardet–Biedl syndrome (BBS) belongs to. It is an autosomal recessive disorder characterized by retinal dystrophy, obesity, polydactyly, learning disability, hypogonadism, renal malformations and hypertension.

Results:
A 12 years-old female patient present with primary non-monosymptomatic enuresis.
She had been diagnosed with right hand and foot polidactyly at 2 years-old; intellectual disability (IQ 74) and developmental delay with serious behavioural abnormalities at 5 years-old. Beyond that she is obese (BMI >P97), has metabolic syndrome with insulin resistance. Besides little refractive error, she has no other visual abnormalities. Following evaluation showed renal ultrasonography with no other abnormalities besides horseshoe kidneys; renal scintigraphy with differential renal function 46-54% corresponding to right and left kidney.
Meanwhile, her younger brother was diagnosed BBS, as he presented with polydactily, prenatal pyelocalyceal dilation and renal microcysts. Because of that, she underwent genetic evaluation, which confirmed the diagnosis of BBS.
Enuresis has resolved with time, although she still has daytime urinary incontinence sporadically. Renal function is still preserved, with no urine concentration defect or other signs of renal dysfunction. Blood pressure is within normal reference range for age, height and sex (<P90).

Conclusion:
A multidisciplinary diagnostic approach may be necessary in unclear cases, as well as, a high index of suspicion based on the clinical picture for further evaluation.
Although there is no therapy to prevent progressive organ involvement, patients require multidisciplinary health team to manage comorbidities. Visual prognosis is often poor while kidney dysfunction is the major cause of morbidity and mortality.
As seen in the case above, BBS exhibits a variable expressivity and intrafamilial variation.

Key words: Bardet Biedl, ciliopathies, polydactyly

POSTER - PO 45

TRPM7 MUTATION CAUSING HYPOMAGNESAEMIA WITH SECONDARY HYPOCALCAEMIA

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Objectives:
Hypomagnesaemia with secondary hypocalcaemia (HSH) is a rare autosomal recessive disorder caused by pathogenic variants in TRPM6, encoding the channel-kinase transient receptor potential melastatin type 6. TRPM6 forms tetramers with its close homologue TRPM7 and this interaction is essential for TRPM6 activity. Patients have very low serum magnesium (Mg2+) levels and suffer from muscle cramps and seizures. Despite genetic testing, a subgroup of HSH patients remains without a diagnosis.

Methods:
Our objective was to describe the clinical characteristics of a Spanish patient with HSH and to identify the mutation causing the disease.

Results:
A Spanish family with an HSH phenotype was subjected to whole exome sequencing. Bioinformatics tools were used to predict variant pathogenicity.
The patient suffered from seizures and muscle cramps due to Mg2+ deficiency and episodes of hypocalcaemia. Nystagmus has been observed since birth. We identified a heterozygous point mutation resulting in a p.Gly1046Asp change in the pore domain. This variant was predicted pathogenic with very high scores by pathogenicity prediction tools. The amino acid change was predicted to disturb the pore structure. Parental testing by Sanger sequencing did not show the variant, indicating de novo origin. No other rare variants were detected in known hypomagnesaemia-causing genes.

Conclusion:
We describe for the first time the presence of a rare variant of the TRPM7 gene in a patient with HSH. The identified variant impairs TRPM7 channel activity. Screening of unresolved patients with hypocalcaemia and secondary hypocalcaemia may further establish TRPM7 mutations as the cause of a novel type of HSH.

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Key words: Rare genetic disease, HSH, magnesium deficiency, TRPM6, TRPM7

POSTER - PO 46

MAY THE TUBULE HAVE PROPELLERS?

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Objectives:
HELIX syndrome is a new tubulopathy of genetic cause(claudin-10 deletion) characterized by salt-losing nephropathy and dyselectrolythemia (hypokalemia, hypermagnesemia, hypercalcemia and hypocalciuria) together with alterations in the homeostasis of ectodermal glands and skin integrity (lacrimal dysfunction, xerostomia, ichthyosis and heat intolerance)

Methods:
Illustrate with a clinical case, a novel salt-losing tubulopathy

Results:
3-year-old boy with history of polyhydramnios and need for sodium intake (up to 1.5 mEq/kg/day) from birth due to persistent hyponatremia together with low-limit potassium with a high transtubular potassium gradient (GTTK) and hyperreninemia. Subsequent controls with hypochloremic metabolic alkalosis with persistence of borderline potassium. Preserved renal function and tubular study with increased fractional excretions of sodium, potassium, chloride and hypocalciuria. Renal ultrasound without nephrocalcinosis. Given these findings, a genetic study was performed, which was negative for classic and antenatal Bartter syndromes, including MAGED-2, Gitelman syndrome, CaSR gain mutations, and pseudohypoaldosteronisms. As an important finding, clinically he presented xerostomia, so it was decided to expand the genetic study due to suspicion of an alteration in Claudin-10, which detected a deletion in the CLDN-10B gene.He maintained normal calcium levels, as well as magnesium levels at the upper limit of normal along with hypomagnesiuria.During his follow-up, he has required potassium and sodium contributions, markedly increasing his requirements in the event of decompensation due to intercurrent processes

Conclusion:
Salt-wasting tubulopathies are underdiagnosed at any age, especially when dealing with infrequent or studied entities and in the face of high phenotype-genotype variabilities and phenotypes not yet clearly described.That is why the progressive knowledge of the pathophysiological processes of the renal tubule and the integral proteins of the tight junction, expressed not only in the kidney, but also in the skin and salivary glands (claudin-10b) will allow an early diagnosis and better management of these patients, allowing adequate genetic counseling and possible prevention of kidney disease progression

Key words: HELIX syndrome, Salt-wasting tubulopathies, Claudin-10

POSTER - PO 47

TWO NEW FAMILIES WITH RRAGD GENE TUBULOPATHY, CHARACTERIZED BY SEVERE HYPOMAGNESEMIA AND DILATED CARDIOMYOPATHY

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Introduction:
RRAGD gene encodes a small Rag guanosine triphosphatasa D expressed in hearth and kidney, specifically in the thick ascending limb and distal convoluted tubule. Variants in this gene lead to a recently described tubulopathy called Autosomal Dominant Kidney Hypomagnesemia. Patients have a severe hypomagnesemia and/or dilated cardiomyopathy with early heart failure resulting in heart transplantation in a substantial subset of patients.

Objectives:
to describe the clinical phenotype of four patients (two families) affected with this new tubulopathy.
Métodos
medical records review, molecular diagnosis by Whole Exome Sequencing and later sequencing by Sanger.

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Results
Clinical data is detailed in Table 1

Table 1. Clinical data of patients

	Family A			Family B
	A-II-1	**A-II-2**	**A-III-1**	**B-II-1**
Polyhydramnios	No	No	Yes	-
Gestational age (weeks)	ND	ND	33	35
Age at renal manifestation/last-follow-up	3,5/47	3,5/47	6/8	11/15
Polyuria and hypomagnesemia or hypocalcemia symptoms at diagnosis	Yes	Yes	Yes	Yes
Length last follow up, SD	Normal	Normal	-1,4	-0,8
Dilated cardiomyopathy (age diagnosis, years)	Yes (32)	Yes (32)	No **	Yes (5)
Heart transplantation (age, years)	No*	Yes (42)	No	No
Initial laboratory findings (plasma)				
Na^+, 135-145 meq/l	136	135	137	139
K^+, 3,5-4,5 meq/l	2,8	2,8	3,3	3,5
Cl^-, 95-105 meq/l	92	92	97	104
Mg^{2+}, 1,7-2,5 mg/dl	1,1	1,3	0,9	1,2
Ca^{2+}, 9-10,8mg/dl	6,7	Low	-	9,7

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Metabolic alkalosis	Yes	Yes	Yes	Yes
Renal function last follow-up	Normal	Normal	Normal	Normal
Nephrocalcinosis (age first finding, years)	Yes	Yes	Yes (6)	Yes (11)
Therapy				
Mg/Ca/K supplementation	Yes/yes/yes	Yes/yes/yes	Yes/No/yes	Yes/No/yes
Heart failure medication	Yes	Yes	No	No
Other	No	Myfortic	Citrate	No
Variant *RRAGD* gene	p.(Ser76Leu)			

* On the waiting list

** Cardiac apical trabeculations

Conclusion:
In the Autosomal Dominant Kidney Hypomagnesemia the cardiomyopathy has poor prognosis, whereas the renal prognosis appears to be good. However, further long-term studies of this new entity are needed.

Key words: RRAGD gene, tubulopathy, hypomagnesmia, cardiomyopathy

POSTER - PO 48

WHEN TO STOP BREASTFEEDING? CYP24A1 MUTATION

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Introduction:
Autosomal recessive hypercalcaemia of infancy also known as vitamin D hypersensitivity is a rare disorder of phosphocalcium metabolism. It is due to mutations in the CYP24A1 gene that reduces or eliminates the enzyme 24 hydroxylase responsible for transforming vitamin D to its inactive form. It is characterised by early-onset hypercalcaemia, hypophosphataemia, hypercalciuria, decreased parathyroid hormone and medullary nephrocalcinosis. It typically manifests with medro failure, hypotonia, vomiting, constipation and/or polyuria.

Results:
6-month-old infant admitted for severe asymptomatic hypercalcaemia. Personal history: admission at 12 hours of life due to infectious risk and hyperbilirubinaemia. Follow-up in the gastrointestinal tract due to medro failure and suspected IPLV. Laboratory tests showed hypercalcaemia (serum calcium 16.6 mg/dL), normocalciurea, hypophosphataemia, low parathyroid hormone and high vitamin D levels. They denied vitamin D dosage error. During admission, hyperhydration was performed, reducing the figures, but hypercalcaemia persisted in subsequent controls. Normal CGH-array. Renal ultrasound showed medullary nephrocalcinosis. The patient was referred to the nephrology department, where two pathogenic variants in heterozygosity in the CYP24A1 gene were detected. Calcium intake was limited, avoiding sun exposure, abundant fluid intake, without achieving normal calcaemia figures until total suppression of breastfeeding.

Conclusion:
Although the most frequent cause of hypercalcaemia in children is iatrogenic, we must not forget, in the differential diagnosis, the rare or genetic aetiologies that may be the cause of hypotonia or failure to thrive, as in our case. In our case, it should be pointed out how difficult it was to withdraw breastfeeding, given that its multiple benefits are so well known. This was the only way to achieve clinical and analytical normalisation in our patient.

Key words: CYP24A1 mutation

POSTER - PO 49

A RARE CAUSE OF COMPLEX PROXIMAL TUBULOPATHY IN MODERN TIMES

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Introduction:
Tyrosinemia is a rare genetic disorder that disrupts the breakdown of tyrosine, an aminoacid. It affects approximately 1 in every 100,000 to 120,000 individuals globally and can impair kidney function by causing tubulopathy – dysfunction of renal tubules. The toxic metabolic agent involved is succinylacetone which accumulates over time leading to damage. At present, the prevalent treatment for all instances of tyrosinemia is nitisinone therapy. Consequently, tubular toxicity has become exceedingly uncommon since the introduction of this medication.

Results:
We present the case of a 7-year-old girl who arrived in Spain with a prior diagnosis of tyrosinemia made in infancy, but who had not received treatment since the age of two. The patient's diagnosis was made early because she had a brother who had passed away from the same disorder. Upon physical examination, the patient had short stature, malnutrition, splenomegaly, and marked bilateral genu valgus with thickened wrists, characteristic signs of rickets. Laboratory investigations revealed generalized proximal tubular dysfunction with hyperchloremic metabolic acidosis, hypophosphatemia, hypouricemia, hypokalemia, glycosuria, and tubular proteinuria. Mild liver dysfunction was also noted, with nodular lesions found on liver ultrasound. Imaging studies confirmed radiological rickets and normal renal ultrasound. Genetic testing confirmed a homozygous mutation in the FAH gene, providing a definitive diagnosis for the patient's condition.
Treatment with Nitisinone and a low-protein diet was initiated, along with supplements of bicarbonate, phosphorus, potassium, and vitamin D. Following this treatment regimen, the patient showed positive progress, with a gradual correction of the previously observed analytical abnormalities, and improvement in overall health condition, as evidenced by an increase in growth speed and weight.

Conclusion:
The implementation of nitisinone and appropriate water and electrolyte correction has resulted in the gradual improvement of the condition. Overall, early diagnosis and prompt initiation of nitisinone therapy can prevent long-term complications associated with tyrosinemia, including tubulopathy.

Key words: tyrosinemia, proximal tubulopathy, nitisinone

POSTER - PO 50

PRESCHOOL CHILD WITH PRIMARY HYPEROXALURIA TYPE 3

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Objectives:
Preschool male with bilateral renal lithiasis and recurrent renal colic secondary to primary hyperoxaluria.

Methods:
think about infrequent causes of recurrent lithiasis rule out the most common ones

Results:
Healthy 18-month-old infant, asymptomatic, with bilateral urolithiasis on abdominal ultrasound. Lithogenic risk study: frank hyperoxaluria (500mg/1.73m2/day), without other alterations.
At the age of 21 months, the first renal colic, partially obstructive, resolved with expulsion of lithiasis, the analysis of which reveals a composition of 40% calcium oxalate dihydrate and 60% calcium oxalate monohydrate.
In successive months, he suffered two new renal colics, not complicated. Hyperoxaluria persists, in addition to mild hyperuricosuria and hypocitraturia. Start treatment with oral potassium citrate and a low-oxalate diet.
With the suspicion of primary hyperoxaluria, a molecular genetic study is requested, which shows two heterozygous variants of the HOGA1 gene, c.700+5G>T(IVS+5G>T) and c.818T>C(p.Ile273Thr). associated with Type 3 Primary Hyperoxaluria, with each parent carrying one of them.

Conclusion:
Primary hyperoxalurias are rare diseases, genetically based, with an increase in endogenous oxalate due to enzymatic alterations involved in its metabolism. They present with early recurrent nephrolithiasis, generally associated with nephrocalcinosis and occasionally end-stage renal disease. Currently, 3 types with different clinical presentations are distinguished: Type 1 (more common and worse prognosis), type 2 (less frequent) and type 3 (rare and better prognosis). It is convenient to make a differential diagnosis with secondary causes of hyperoxaluria that will have a specific management. The treatment of PH is based on hyperhydration and alkalizing of the urine. In some forms pyridoxine may be effective. Severe forms require renal or hepatorenal transplantation. A high index of suspicion and early diagnosis allow adequate management that can improve the prognosis by slowing down the progression to end-stage renal disease.

Key words: PRIMARY HYPEROXALURIA

POSTER - PO 51

QTC PROLONGATION AS A SIGN OF TUBULOPATHY: A CASE REPORT.

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Introduction:
Dyselectrolytemias, such as hypokalemia and hypomagnesemia, can be the cause of heart rhythm disturbances and can be indirectly observed on electrocardiogram studies.
Gitelman syndrome is an autosomal recessive inherited tubulopathy due to inactivating mutations of the SLC12A3 gene, that codifies for the sodium-chloride cotransporter (NCC) in the distal convoluted tubule with saline urinary loss, thus producing metabolic alkalosis with hypokalemia, hypomagnesemia and hypocalciuria. Although it is a benign disease, severe ventricular arrhythmias and sudden cardiac death have been described due to prolongation of the QTc interval.

Objectives:
We present a case of a patient in whom electrocardiographic changes were key in the diagnosis of this tubulopathy.

Results:
The patient was a 13-year-old girl, daughter of consanguineous parents of Romani ethnicity. During the study of transient arterial hypertension secondary to obesity resolved with hygienic-dietary recommendations, a persistent QTc prolongation was observed on the electrocardiogram. Laboratory tests were performed, hypokalemia (2.6 mEq/L) and hypomagnesemia (1.14 mg/dL) were detected with elevated urinary excretions, metabolic alkalosis (pH 7.46, bicarbonate 32), hypocalciuria (Ca/Cr 0.007), and high renin levels. The remaining renal studies (GFR 130ml/min/1.73m2) and blood pressure showed normal results, and physical examination was unremarkable.
The patient reported water and salt craving, paresthesias in the hands and asthenia. A brother of her was taking oral potassium supplements for persistent hypokalemia.
Using analytical data of metabolic alkalosis, hypokalemia, hypomagnesemia, hypocalciuria, and normal blood pressure, Gitelman syndrome was diagnosed, for which oral potassium and magnesium supplementation was started and a family genetic study was carried out (pending). The patient clinically improved after the start of treatment and QTc interval normalized.

Conclusion:
Gitelman syndrome is a tubulopathy that should be considered in cases of hypokalemia, hypomagnesemia, and metabolic alkalosis. Its diagnosis is based on clinical symptoms and biochemical abnormalities, with possible genetic confirmation.
Proper treatment prevents dangerous complications, such as arrhythmias.

Key word: Gitelman Syndrome, Electrolyte imbalance, Prolonged QTc

POSTER - PO 52

SÍNDROME DE GITELMAN: A PROPÓSITO DE UN CASO.

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Introduction:
El síndrome de Gitelman (SG) es una tubulopatía de herencia autosómica recesiva debida a mutaciones inactivantes en el gen SLC12A3 que codifica el cotransportador sodio-cloro sensible a tiacidas (NCC) del túbulo contorneado distal. Es una nefropatía pierde sal con alcalosis metabólica hipopotasémica, hipomagnesemia e hipocalciuria.

Results:
Niño de 7 años que presenta hipopotasemia (2.9mEq/L) en una analítica realizada para control de dislipemia. Padres no consanguíneos. No presenta antecedentes personales ni familiares de interés. Únicamente destacan avidez por alimentos salados y ocasionales dolores musculares con el ejercicio intenso. No presenta retraso del crecimiento y la tensión arterial es normal. Se descartó ingesta de diuréticos. El estudio de hipopotasemia mostró los siguientes resultados: alcalosis metabólica (pH 7.41, pCO2 54.3mmHg, bicarbonato 34.9mmol/L), hipomagnesemia (1.32mg/dl), activación del eje renina-angiotensina-aldosterona (aldosterona 308pg/ml, renina activa 68.5pg/ml), hipocalciuria (cociente calcio/creatinina 0.016mg/mg), pérdida renal de potasio (EFK 35.6%, GTTK 7.5%) y de magnesio (EFMg 4.09%). El electrocardiograma y la ecografía abdominal fueron normales. Con la sospecha de SG se inició tratamiento con suplementos de cloruro potásico y de magnesio. El estudio genético identificó 2 variantes en heterocigosis en el gen SLC12A3, p.A589D y p.Q817X. Actualmente, 11 años, con suplementos orales de K (1.7mEq/Kg/día) y Mg (8.5mg/Kg/día) mantiene niveles séricos normales y permanece asintomático.

Conclusion:
Muchos pacientes con SG se encuentran asintomáticos o presentan síntomas leves neuromusculares.
Con frecuencia el diagnóstico es fortuito, a partir del hallazgo casual de hipopotasemia.
El diagnóstico diferencial se realiza con el síndrome de Bartter tipo III (hipocalciuria < 0.05mg/mg es típico del SG) y con la hipomagnesemia renal con hipocalciuria.
El tratamiento se basa en dieta rica en sal y en suplementos de cloruro potásico y de magnesio; importante evitar fármacos que alarguen el QT por el riesgo de arritmias.

Key words : Tubulopatia, hipocalciuria, hipomagnesemia

POSTER - PO 53

SHIPERCALCEMIA EN SÍNDROME DE GITELMAN: CASUALIDAD O CAUSALIDAD

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Introduction:
El síndrome de Gitelman (SG) es una tubulopatía de herencia autosómica recesiva, en la cual se produce una alteración del cotransportador NaCl en el túbulo contorneado distal (TCD). Esta alteración conlleva disminución de la reabsorción de cloro y sodio e hipovolemia, activando el eje renina-angiotensina aldosterona y el intercambiador Na/Ca basolateral. Los pacientes presentan alcalosis metabólica con hipopotasemia, hipocalciuria e hiperaldosteronismo hiperreninémico. Frecuentemente, asocian hipomagnesimia por una reducción de los canales de Magnesio de la membrana apical de las células del TCD. La calcemia suele ser normal. Presentamos un caso de SG que durante ingreso en su hospital de referencia presenta cifras elevadas de calcemia. Inicialmente, relacionan este hallazgo con su enfermedad de base, por lo que nos lo derivan para estudio y tratamiento.

Results:
Niño de 15 años, de etnia gitana, en seguimiento en consultas de Nefrología desde los 2 años por SG. En tratamiento con suplementos de potasio y magnesio. Clínicamente asintomático con controles analíticos normales. Presenta deshidratación con descompensación hidroelectrolítica en el contexto de gastroenteritis aguda, por lo que ingresa en su hospital de referencia para tratamiento intravenoso. En analítica al ingreso calcemia de 11,9 mg/dl en ascenso progresivo a pesar de adecuada hidratación, por lo que nos lo derivan.
Ampliamos estudio de función renal, perfil tiroideo y metabolismo fosfocálcico donde destaca paratohormona 76,3 pg/ml, 25OH vitamina D 7 ng/ml, calcio 13,1 mg/dl, fósforo 3,8 mg/dl y Ca/Cr 0,2. Resto normal. Ante dichos hallazgos, solicitamos ecografía de cuello con nódulo hipoecogénico con captación patológica en gammagrafía. Es diagnosticado de hiperparatiroidismo primario por adenoma paratiroideo, intervenido con evolución favorable.

Conclusion:
Como conclusión, señalar que la hipercalcemia tiene múltiples causas, algunas graves como patología maligna. Su aparición de nuevo en un paciente con enfermedad renal de larga evolución, como el SG, debe ser investigada para realizar un diagnóstico y tratamiento correcto.

Key words: Hipercalcemia; Gitelman; Hiperparatiroidismo; Adenoma

POSTER - PO 54

PLEURAL EFFUSION SECONDARY TO EVEROLIMUS IN A PEDIATRIC RENAL TRANSPLANT PATIENT

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Objectives:
The mTOR inhibitors are macrolides obtained from the fungus Streptomyces hygroscopicus,and their use as immunosuppressants in renal transplantation has increased due to their lower nephrotoxicity and their antilymphoproliferative action,which has been related to a lower risk of tumors and infection

Methods:
However,they are not free of adverse effects.We present a case of pleural effusion associated with the use of everolimus

Results:
8-year-old girl with CKD secondary to renal dysplasia underwent early cadaveric donor kidney transplantation.Induction:Basiliximab,tacrolimus,mycophenolate and methylprenisolone.Maintenance;Tacrolimus,mycophenolate and prednisone.Then she presented polyomavirus BK viremia,minimizing mycophenolate and starting monthly immunoglobulins with initial improvement but deterioration 4 months later so mycophenolate was changed to everolimus.Went to emergency room due to abdominal distension and pain in the graft.Exploration with hypoventilation of the right hemithorax(chest x-ray and ultrasound:right pleural effusion 13.6x10cm and collapsed lung).Normal renal function and sediment,CRP 18 mg/L,abdominal ultrasound with free fluid.Thoracocentesis was performed showing a transudate.Etiological study:normal cardiological study. echocardiography and abdominal ultrasound without portal hypertension, non-nephrotic proteinuria, normal thyroid,serum albumin,auto-antibodies,quantiferon,respiratory array,PCR for S.Pneumoniae and viral test(CMV,EBV,VH6/VH7/VH8 and adenovirus) pleural fluid ADA,PCR,staining and cultures.Thoracic CT and MRI ruled out thromboembolism and malignancy.Pharmacological cause was suggested as etiology,everolimus was suspended and progressive improvement was observed with complete resolution

Conclusion:
Lymphedema and visceral effusions have been reported as rare adverse effects in transplant patients receiving imTOR.In pediatrics there have been associated with the development of lymphedema,but no cases of visceral effusions have been reported.Pleural effusion secondary to everolimus in a pediatric renal transplant patientIn our patient,the condition was related to everolimus when other etiologies were ruled out and resolved after discontinuation of the drug.The imTORs prevent lymphangiogenesis by inhibiting the proliferation of lymphatic endothelial cells driven by vascular endothelial growth factor.Therefore,after excluding other etiologies,the pharmacologic cause should be considered in transplant recipients who develop visceral effusions.The condition usually resolves after discontinuation of the medication

Key words: mTOR inhibitors, Everolimus, Kidney Transplantation, Pleural Effusion

POSTER - PO 55

BK VIRUS INFECTION IN PEDIATRIC KIDNEY TRANSPLANT: THE POWER OF AN OLD DRUG

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Introduction:
Post-transplant infections are an important cause of morbidity and mortality in pediatric kidney transplant recipients. BK virus is a DNA virus that is widespread in the general population with an estimated seroprevalence of >90% by early school-age years. BK virus infection is common after pediatric kidney transplantation and while this infection is clinically silent in immunocompetent hosts, in kidney transplant recipients it may progress to BK nephropathy, resulting in graft dysfunction and graft loss.

Results:
Caucasian 9 years-old boy submitted to kidney transplant at 5 years for end-stage renal disease secondary to posterior ureteral valves. Transplant immunosuppression induction therapy with basiliximab, tacrolimus, mycophenolate mofetil (MMF) and methylprednisolone and maintenance with tacrolimus, MMF and prednisolone. He developed CMV infection 18 days after transplant. Valganciclovir was increased to therapeutic dosage, MMF was reduced with no improvement and then switched to everolimus. BK virus plasmatic copies were detected 56 days after the transplant and begun to rise since then. Tacrolimus dose reduction was attempted but ineffective on BK viral load and serum creatinine started to rise. Adjuvant treatment with multiple immunoglobulin administrations and ciprofloxacin was associated with transient improvements on BK virus load but sustained graft disfunction. Afterwards tacrolimus was switched to cyclosporin. The treatment with cyclosporin achieved control of BK virus plasmatic load despite the discontinuation of immunoglobulin. Nowadays, 4 years after the transplant, with cyclosporin and everolimus on a low dosage, BK viral load is 228UI/ml and serum creatinine 87umol/L.

Conclusion:
Regular and early monitoring for BK virus is an effective strategy for detecting the virus and allows for anticipated treatment to insure a longer graft function. Although there is no standardized therapy algorithm for post-transplant infection, this clinical case emphasizes the effective role of cyclosporine in the management of BK infection.

Key words: BK Virus, Kidney Transplantation, Nephrology, Pediatrics, Cyclosporine

POSTER - PO 56

CHRONIC ANTIBODY-MEDIATED REJECTION IN AN ADOLESCENT

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Introduction:
In the follow-up of kidney transplant patients, the support of pathologists interpreting renal biopsy is crucial to this day. One particular scenario is the adolescent period, during which the highest number of kidney graft losses occur due to poor compliance. In the case described below, the pathological diagnosis was crucial for the patient and of great educational value due to the images obtained.

Results:
A 15-year-old patient with displastic solitary kidney underwent deceased donor kidney transplantation. In the second year post-transplantation, developed worsening renal function. He was on prednisone, tacrolimus, and mycophenolate-mofetil. Tacrolimus levels were irregular. Percutaneous renal biopsy showed mixed active antibody-mediated rejection (AMR) and acute T-cell-mediated rejection. Microvascular inflammation and the other features of active AMR were also present, ie, C4d deposition and presence of circulating donor specific antibodies (DSA). Treatment was methylprednisolone, immunoadsorption, immunoglobulins, and rituximab. No subsequent infectious complications. Renal function improved and the patient continued to follow up in outpatient clinics.
Two years later a creeping creatinine was observed. The patient stated taking medication properly. There were no fluctuations in tacrolimus levels. DSA monitoring negative. A new renal biopsy was performed to assess for active humoral rejection. The renal biopsy showed typical chronicity findings, such as transplant glomerulopathy and severe peritubular capillary basement membrane multilayering on electron microscopy.There were no features of active AMR like in the first biopsy, and C4d deposition was absent. Thus, the diagnosis of chronic AMR was made.With this diagnosis, an active AMR was ruled out and therefore intensive immunosuppressive treatment was not employed. We opted for a positive reinforcement of compliance along with closer monitoring in the patient's appointments.

Conclusion:
Thanks to the pathological diagnosis, the appropriate therapeutic response could be provided. Additionally the case described has important educational value because high-quality samples were obtained in combination with the interpretation of pathologists.

Key words: Kidney transplantation, rejection, antibody-mediated.

POSTER - PO 57

NEW VARIANTS IN THE GENE LAMA5 CAUSING NEPHROTIC SYNDROME IN CHILDREN

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Objectives :
Infantile Nephrotic Syndrome (INS) is a disorder characterized by massive proteinuria, hypoalbuminemia and edema that appears in the first year of life. It may be due to congenital infections, systemic or inmunological diseases. However, the main causes is genetic, between 60-80% is due to those affected in the NPHS1, NPHS2, WT1 and LAMB2 genes. New associated genes have recently been identified, including LAMA5.

Methods:
5-month-old infant who attended with bilateral leukocoria, he was diagnosed with cataracts. Second child of non-consanguineous parents with no relevant family history. Delayed intrauterine growth, full-term birth with low birth weight, adequate posterior weight-height development. Working diagnosis was galactosemia, complementary investigations showed hypoalbuminemia, hyperchloremic metabolic acidosis, glycosuria, microhematuria, generalized hyperaminoaciduria and glomerular proteinuria in the nephrotic range stand out. Polyuric patient without acute renal damage or arterial hypertension. Nephrotic syndrome associated with incomplete Fanconi Syndrome was diagnosed. No known past history of toxic exposure. Once congenital infections and inmunological diseased had been ruled out, nephrotic syndrome of genetic origin was suspected.

Results:
Clinical exome secuencing identifies two compound heterozygous variants in the LAMA5 gene, classified as probable pathogenic (c.7036_7037delCA p.Gln2346fs*8888) and of uncertain significance (c.4300G>A p.Gly1434Arg), each inherited from a parent. They are considered to cause the disease.

Conclusion:
The sequencing of the complete exome in the study of the INS has allowed the discovery of new genes involved in its etiology, expanding the genetic heterogeneity of the disease. The alpha5 subunit, encoded by LAMA 5, is the major component (excluding collagen) of the glomerular basement membrane. Abnormalities in this protein can cause different degrees of proteinuria as well as microhematuria. It is also expressed in the basal membrane of neuromuscular, ocular and vascular tissue and extrarenal anomalies may be associated with nephrotic syndrome. Fanconi syndrome secondary to massive proteinuria INS has been described in the literature.

Key words: nephrotic syndrome, fanconi syndrome, LAMA5

POSTER - PO 58

THESE DEPOSITS SHOULD NOT BE TAKEN LIGHTLY

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Objectives:
Angiogenesis is critically important in tumor growth,invasion and metastasis development.VEGF play this role,overexpressed in a wide variety of tumors,associated with progression.Bevacizumab(BVZ) is a humanized anti-VEGF monoclonal antibody widely used and studied in oncologic patients.Its short-term role in the development of renal toxicity(hypertension,proteinuria and TMA)is well known,although the pattern of renal toxicity associated with prolonged treatment is still unknown

Methods:
Present a case of nephrotoxicity due to prolonged use of BVZ

Results:
18-year-old woman with neurofibromatosis type II started treatment with BVZ.18 months later developed proteinuria(iPr/Cr 0.68 mg/mg)with preserved renal function.Started antiproteinuric therapy(candesartan)with good control but need to increase to maximum dose.Nephrotic proteinuria persisted and forced to change to olmesartan and later to lisinopril(both maximum doses).Despite this,developed HT needing amlodipine(maximum dose).Due to these findings BVZ was suspended on multiple occasions,worsening her underlying disease.Such decompensation together with the finding of persistent microhematuria (suspected TMA),promotes renal biopsy:glomeruli with mesangiocapillary pattern and PAS-positive pseudothrombus.Conventional DIF:IgM (+++),Kappa (+++)and Lambda(+++),deposits for C1q(+)and very focal and segmental positivity for IgG together with images of pseudothrombus impregnation for IgA.Congo red staining negative,receiving initially the diagnosis of cryoglobulinemic glomerulonephritis.While awaiting the definitive results,prednisone was started(60mg/day)with subsequent decrease until discontinuation in the absence of response,after performing a proteinogram(normal)and cryoglobulins(negative).Electron microscopy showed electrodense deposits at subendothelial,mesangial and paramesangial levels compatible with glomerular microangiopathy associated with chronic use of bevacizumab,and in order to improve the quality of life of the patient as an alternative,it was decided to start Brigatinib,reducing the dose of BVZ and progressive renal involvement

Conclusion:
The deposition of antibodies,either by immunocomplex formation or anti-BVZ antibody production,could explain the cases of difficult-to-control nephrotic proteinuria in patients with prolonged treatment with BVZ.Knowledge of the mechanisms of nephrotoxicity,as well as its long-term effects,is essential for the development of new guidelines and preventive strategies to minimize the risk and impact on the survival of these patients

Key words: Bevacizumab, Neurofibromatosis, Amyloidosis, Nephrotic Proteinuria

POSTER - PO 59

IMMUNOADSORPTION-DEPENDENCE IN A CHILD WITH MULTI-DRUG RESISTANTNEPHROTIC SYNDROME. IS IT A NEW STEP IN THE CONFIRMATION OF CIRCULATING FACTOR DISEASE.

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Introduction:
We present the case of a 13-year-old boy with Multi-drug-Corticoresistant Nephrotic Syndrome(SN) in which the introduction of Immunoadsortion(IA) has induced partial remission.

Results:
A 13-year-old boy, with no personal history of interest, was referred for presenting edema and dyspnea. He didn´t refer any infectious triggering episode. Physical exploration showed significant weight gain(30kg), tension edema, without clinical data of systemic disease.
The analysis showed proteinuria in the nephrotic range(108mg/m2/h), hypoalbuminemia(1.2 g/dl) and acute kidney disease (GF80 ml/min/1.73m2). Ultrasound showed bilateral pleural and peritoneal effusion, with normal kidneys. Differential diagnosis of secondarisms showed negative serology for virus, normocomplementemia and negative autoimmunity study(ANA, ENA and ANCA).
Our patient received initial treatment with oral prednisone 60 mg/m2/day for eight weeks and intravenous methylprednisolone bolus(1gram/1.73m2) without achieving remission. In the state of steroid-resistance, a renal biopsy was performed, showing FSGS and associated genetic pathogenic variants were ruled out. Sequential immunosuppressive treatment with cyclophosphamide (2.5mg/kg-12 weeks), cyclosporinA and Rituximab (375mg/m2-2 doses) didn´t achieve any remission response. Given the Multi-drug-resistance, LDL-apheresis was started with no response. Immunoadsortion was then started, administering three weekly sessions. During the first year of treatment, the patient showed a slow improvement achieving partial remission (proteinuria from 8gr/day to 1gr/day, albuminemia 4gr/dl). He currently maintains stability with prednisone low doses, cyclosporine(levels 40-50mg/dL) and two weekly IA sessions. Attempts have been made to reduce the number of IA sessions with increased proteinuria, raising the possibility of a dependency on immunoadsorption.

Conclusion:
IA can be an effective technique for inducing complete or partial remission in children resistant to sequential immunosuppressive treatment in native kidney being a good option to delay the evolution to end-stage kidney disease and need for a first kidney transplant, especially in the child population that will require retransplants throughout their lives.
IA-dependence can develop in this patients.

POSTER - PO 60

SEVERE COLLAPSING GLOMERULOPATHY LEADING TO KIDNEY TRANSPLANTATION

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Introduction:
WT1 associated collapsing glomerulopathy is a rare and aggressive form of kidney disease. This type of glomerular harm causes tufted glomeruli to collapse, alongside the covering podocytes. It is triggered by mutations found within the Wilms tumor 1 (WT1) gene which performs an important function in both development and functionality of kidneys. Despite medical intervention, it usually advances rapidly to chronic kidney disease (CKD) G5.

Results:
A nine-year-old male with no significant medical history presented with palpebral swelling and asthenia. He had tested positive for asymptomatic SARS-CoV-2 infection two months prior and influenza A shortly before presenting to the clinic. The patient was diagnosed with nephrotic syndrome, kidney failure (estimated GFR 30 ml/min/1.73m^2), and hypertension without oliguria or hematuria. Follow-up investigations revealed normal hematological parameters, negative autoimmunity markers, and normal complement levels. Ultrasound confirmed bilateral cortical hyperechogenicity.
Percutaneous biopsy showed global sclerosis in 26% of glomeruli and focal segmental sclerosis. Segmentary visceral podocytes hyperplasia was outlined, demonstrated by immunohistochemical antiWT1 and KI67, leading to glomerular capillaries to complete collapse. No relevant deposits in the immunofluorescence. Electronic microscopy reveals global wriklenning of glomerular basement membrane with subendothelial edema, hyperplasia of mesangial cells, hypertrophic podocytes with microvellositary transformation and pedicel fusion and capillary collapse.
The diagnosis of collapsing glomerulopathy was made. Steroid treatment was initiated, followed by two doses of Rituximab, without response. Subsequent genetic testing revealed a heterozygous pathogenic mutation in the WT1 gene, the underlying cause of steroid-resistant nephrotic syndrome. The patient's condition rapidly progressed to chronic kidney disease, and they underwent a preemptive deceased donor kidney transplantation 8 months after initial presentation.

Conclusion:
According to our analysis, it is plausible that a double-hit mechanism has occurred in this case involving both the presence of WT1 mutation since birth and subsequent infection with SARS-CoV-2, leading to an intensified form of glomerulopathy.

Key words: WT1, collapsing glomerulopathy, kidney transplantation

POSTER - PO 61

KIDNEY DISEASE SECONDARY TO PRIMARY COENZYME Q DEFICIENCY: A CASE SERIES REPORT

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Introduction:
Coenzyme Q10 (CoQ10) is involved in the mitochondrial energy production process. Primary coenzyme Q10 deficiency (PCQD) is caused by autosomal recessive mutations in genes involved in its synthesis. Symptoms can affect several organs including kidney.

Objectives:
Our aim is to focus on PCQD as a rare cause of nephrotic syndrome and end-stage renal disease (ESRD).

Methods:
Review of electronic medical records of 3 patients with PCQD and kidney involvement.

Results:
Case 1: 7-year-old boy from China. Incidental finding of nephrotic range proteinuria and hypertriglyceridemia with normal glomerular filtration rate (GFR). Ultrasound showed nephrocalcinosis. Biopsy, performed because of treatment resistance, showed focal and segmental glomerulosclerosis (FSGS) and genetic testing confirmed 2 heterozygous mutations in COQ8B. Proteinuria improved with ACE inhibitors but only dropped below nephrotic range after replacement therapy. 5 years after the diagnosis he stays asymptomatic, proteinuria is controlled and has normal GFR.
Case 2: 15-year-old boy from Morocco with intellectual disability of unknown etiology. Incidental finding of ESRD with severe proteinuria. Biopsy showed terminal nephrosclerosis. After 2 years of hemodialysis, he received deceased kidney transplant and has normal graft function 1 year after. Genetic testing recently confirmed a homozygous mutation in COQ8B.
Case 3: 12-year-old girl from Morocco with dilated cardiomyopathy of unknown etiology. Incidental finding of ESRD with nephrotic range proteinuria. No biopsy was performed, and genetic testing confirmed a homozygous mutation in COQ8B. After 3 months of hemodialysis, she received deceased kidney transplant. She is on CoQ10 replacement therapy with no signs of recurrence 1-year post-transplant.

Conclusion:
- PCQD should be considered as a possible cause of resistant nephrotic syndrome.
- COQ8B is the most frequently affected gene.
- FSGS is the most prevalent pathological finding.
- CoQ10 replacement therapy should be started before irreversible organ damage occurs and may change prognosis and prevent or delay progression to ESRD.

POSTER - PO 62

ALTERATIONS IN GLOMERULAR FILTRATION MARKERS WITHOUT ALTERATIONS IN KIDNEY FUNCTION

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Introduction:
The assessment of estimated glomerular filtration rate (eGFR) in patients with childhood neoplasms is important. This value is used to adjust different prescriptions in light of their frequent nephrotoxic effects. We present two cases with an alteration in the filtration rate according to cystatin C calculation but within normal range according to the creatinine calculation and nuclear medicine techniques.

Results:
Clinical cases:
The first patient is an 11-year-old girl with acute myeloid leukaemia, whom, prior to the start of treatment, showed an alteration in cystatin C of 2.74 mg/l (eGFR by Filler 30 ml/min/1.73m2), despite serial creatinine levels within the normal range (0.55-0.65 mg/dl, eGFR by Schwartz 95-105 ml/min/1.73m2). Analytical study ruled out the presence of thyroid disorders and the use of corticoids. Renal ultrasound showed a slight non-specific parenchymal echogenicity. Finally, the GFR was measured by Cr-EDTA: 102 ml/min/1.73m2, confirming a normal GFR.
The second patient was a 12-year-old boy also with acute myeloid leukaemia, who had received an allo-transplant of haematopoietic progenitors 1 year earlier, referred to the clinic for progressive worsening of renal function due to high cystatin C levels (1.44 mg/dl, GFR by Filler of 61 ml/min/1.73 m2) and creatinine within normal levels (0.43 mg/dl, GFR by Schwartz 137 ml/min/1.73m2). Initially, the discrepancy was ascribed to baricitinib and acyclovir nephrotoxicity. Laboratory analysis ruled out thyroid disorders and the patient did not receive corticotherapy. His glomerular filtration rate was 99mTc-DTPA of 135 cc of plasma/minute/1.73 m2, within the normal range.

Conclusion:
Cystatin C can be an unreliable marker filtration marker in the context of leukemia. Nuclear medicine techniques are an alternative for the calculation of glomerular filtration rate in patients with discrepancies between different markers.

Key words: Cystatin C, Estimated glomerular filtration rate, Leukemia, Creatinine

POSTER - PO 63

ACUTE RENAL FAILURE IN A PATIENT WITH HENOCH SHONLEIN NEPHRITIS

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Objectives:
Renal involvement determines the long-term prognosis in Henoch-Schonleïn Purpura (HSP). Nephrotic syndrome (NS) is a rare presentation form that increases the risk of end-stage renal failure. We present the case of a patient who developed NS after being diagnosed with HSP and afterwards presented a rapidly progressive renal failure of extrarenal ethiology

Methods:
A 7-year-old boy recently diagnosed of HSP was admitted to our hospital because of edema, proteinuria (Qprot/creat 15.7) and hypoalbuminemia (1.8g/dl). A percutaneous renal biopsy was performed without incident and Prednisone 60mg/m2/d was started. Renal pathology revealed secondary HSP ISKDC III nephritis. Afterwards, the patient initiated with persistent macrohematuria and 7 days later initiated abdominal pain and fever and was diagnosed of an ileo-ileal intussusception. He was admitted with intravenous cefoxitin with spontaneous resolution of the intussusception, but he started with progressive worsening of renal function and edemas. Suspecting rapidly progressive glomerulonephritis, 3 high-dose of corticosteroids were administered and a second biopsy was performed, detecting secondary HSP ISKDC III nephritis with acute tubular necrosis due to hematic casts. Despite the initial treatment, he developed a severe acute renal failure (urea 222mg/dl, creatinine 7.5mg/dl), so renal replacement therapy with hemodialysis was started for 5 days and also oral treatment with Mycophenolate. He also developed anasarca and arterial hypertension, requiring high-flow oxygen therapy, daily infusions of serum albumin and Amlodipine. Subsequently, progressive clinical improvement with increased urine output and decreased creatinine and edema, being able to be discharged after 2 weeks with GFR 64ml/min/1.73m2 and decreasing proteinuria. Current renal function recovered without proteinuria or hematuria.

Conclusion:
Acute tubular necrosis due to hematic casts is a rare complication described in patients with persistent macrohematuria that can lead to acute renal failure. It is a pathology to take into account in patients with HSP nephritis with gross hematuria and acute renal failure.

Key words: : Henoch Shonlein nephritis, acute renal failure, actue tubular necrosis

POSTER - PO 64

RAPIDLY PROGRESSIVE GLOMERULONEPHRITIS IN A PATIENT WITH RENO-PULMONARY INVOLVEMENT: ADULT PATHOLOGY ONLY?

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Objectives:
Anti-glomerular basement membrane antibody (anti-GBM) disease is a very rare small vessel vasculitis in pediatrics,caused by the presence of antibodies directed against GBM and alveolar basement membrane(ABM),which presents as rapidly progressive glomerulonephritis(RPGN)and/or alveolar hemorrhage

Methods:
We present the case of a patient diagnosed with anti-glomerular basement membrane antibody disease

Results:
8-year-old girl,no history of interest,consulted the emergency department for prostration and pallor,presenting the previous days with epigastralgia without other associated symptoms.No edematization and she reported preserved diuresis.Constant findings:tachypnea,hypoxemia(SatO2 89%)and hypertension(138/89 mmHg).Physical examination:poor general condition and cutaneomucous pallor,crackles in the right lung base and pansystolic heart murmur.Blood analysis:severe non-hemolytic anemia (Hb4.1mg/dl),acute renal damage(urea 249mg/dl,creatinine 10.78mg/dl)with hyperkalemia(K 6.7mmol/l)and metabolic acidosis.Proteinuria in nephrotic range (IPr/Cr 5mg/mg)and microhematuria.The study was completed with a chest X-ray showing signs of acute pulmonary edema,cardiomegaly and peripheral cottony infiltrate in right fields suggestive of alveolar hemorrhage.An abdominal ultrasound was performed showing globular,hyperechogenic kidneys with loss of corticomedullary differentiation.A red blood cell transfusion was administered.A bladder catheter was placed and oligoanuria was observed despite diuretic treatment.He was transferred to the PICU for placement of a hemodialysis catheter and empirical treatment with methylprednisolone (300mg/m2/day,3 doses)was administered.Renal biopsy was performed after 48 hours:necrotizing glomerulonephritis with active and chronic lesions,with positive immunofluorescence for linear IgG in GBM.With the diagnostic orientation of anti-GBM antibody disease treatment with cyclophosphamide and plasmapheresis is associated.The detection of anti-GBM antibodies in the analytical controls is repeatedly negative,and the presence of pANCA antibodies is confirmed.A bronchoalveolar lavage confirming the presence of hemosiderophages with a satisfactory evolution.She persists in ESRD requiring RRT until she receives a renal transplant at 10 years of age

Conclusion:
Anti-GBM disease is a rare cause of ESRD in pediatrics.The serologic spectrum is variable,antibodies may not be found in plasma(12%)or may be found together with ANCA(10-50%).Renal biopsy is essential to establish the diagnosis and prognosis of the disease

Key words: Vasculitis, Goodpasture Syndrome, ANCA, Anti-GBM

POSTER - PO 65

HYPOCOMPLEMENTEMIC NEPHROTIC SYNDROME: WHO IS WHO?

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Objectives:
Alport Syndrome (AS) is the second most common hereditary kidney disease caused by alterations in type IV collagen genes,with varied symptoms, from asymptomatic or hematuria ,proteinuria, sensorineural hearing loss and ocular involvement,to end-stage renal disease(ESRD)

Methods:
Present a case of AS with atypical presentation

Results:
14-year-old boy who,at age of 6,presented episode of generalized edema, nephrotic proteinuria(iPr/Cr 11.24mg/mg), hematuria, hyperkalemia, hypoalbuminemia, elevated creatinine (0.77mg/dl), metabolic acidosis, and hypocomplementemia (C3 175mg/l,C4 230mg/l) with normal autoimmune tests. Hypertension (HT) was documented, requiring treatment with nifedipine. The diagnosis was oriented to acute glomerulonephritis (GN) in the context of viral sickness. Renal biopsy was performed, obtaining data of post-infectious-GN with positive immunofluorescence (C3,IgG and IgM). He progressively recovered renal function,decreasing proteinuria and hematuria. Given persistence of HT, hyperkalemic metabolic acidosis with low renin activity and decreased transtubular potassium gradient (TTKG), diagnosis of type IV ATR/Gordon syndrome was proposed,initiating treatment with hydrochlorothiazide, achieving adequate evolution, without genetic confirmation. 3years later, second episode of similar characteristics. Enalapril and amlodipine were added. However,due to persistence of C3 consumption, renal biopsy was made, diagnosing C3-glomerulopathy. During follow-up, the presence of hypocitraturia together with high lithogenic index was highlighted (no nephrocalcinosis /lithiasis) which has made it necessary to maintain treatment with potassium citrate. Progressive withdrawal of antihypertensives has been achieved until omitting them, as well as hydrochlorothiazide due to normalization of calciuria. Currently, he presents tubular damage with partial resolution and worsening of renal function (eGFR 50-70ml/min/1.73m2),drawing attention to persistent hypomagnesemia, hypovitaminosis D, without proteinuria or hematuria, maintaining low GTTK. For all these reasons,it was decided to study the clinical exome, observing a mutation in COL4A3 in homozygosis, producing SA

Conclusion:
When faced with clinically variable diseases, the importance of genetic tests in the face of uncertain diagnoses is highlighted,and in this case, without usual clinical presentation(up to 40% ocular involvement and sensorineural hearing loss in childhood), masked by symptoms and analytical findings that led to a completely different pathology. Once the affected gene has been identified, a family segregation study is necessary to determine the inheritance pattern and to be able to provide genetic counseling,as well as prognosis and prevention of progression towards ESRD

Key words: Alport Syndrome, Hypocomplementemia, C3 glomerulopathy

POSTER - PO 66

HYPOCOMPLEMENTEMIC GLOMERULONEPHRITIS OR HYPOCOMPLEMENTEMIA WITH GLOMERULONEPHRITIS?

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Objectives:
Postinfectious gomerulonephritis(APIGN)is the most common cause of acute glomerulonephritis (AGN) and glomerular hematuria worldwide in pediatrics,with most of the manifestations usually resolving in 2 weeks,with persistent proteinuria 6-8 weeks,decrease of complement (C3) 10-12 weeks and microhematuria 12-24 months

Methods:
Cases that present deviations on this evolution force to raise the differential diagnosis with other ANG with C3 decrease (glomerulonephritis by C3 or systemic pathologies like SLE).We present a case with a torpid evolution

Results:
4-year-old patient consulted for macrohematuria of 10 days of evolution and oliguria in the last 48h,who had presented a febrile episode 2 weeks earlier.Mild bilateral palpebral edema and stage II hypertension together with nephrotic proteinuria(iPr/Cr4.4mg/mg).Blood tests showed normal renal function parameters and negative antibodies,hypocomplementemia with decreased C3,normal C4 and elevated ASLO.He was admitted for blood pressure control,receiving furosemide and nifedipine.In subsequent controls,normalization of proteinuria and decrease of hematuria was observed,with persistence of low C3 levels after 12 weeks of follow-up,so a renal biopsy was indicated.The results showed a mesangial proliferative glomerulonephritis with only C3 deposits and no scarring or necrotizing lesions.Ultrastructurally we identified small nodular deposits predominantly in mesangium and very isolated paramesangial/parietal deposits.Ultrastructural findings ruled out a dense deposit disease and raised the possibility of a C3 glomerulopathy/evolved postinfectious glomerulonephritis/glomerulonephritis in remission.Nephritic factor was negative.A genetic study of the complement found a heterozygous variant of the C3 gene(aberrant protein)that would justify the partial deficit of C3.Same mutation was found in both the father and his sister.She has had no new flares during follow-up,with negative proteinuria and hematuria.

Conclusion:
APIGN has an excellent prognosis with complete remission in 95% and infrequent recurrences;other pathologies should be suspected in case of recurrences or torpid evolution.Our case was oriented as an episode of APIGN in a patient with genetic complement deficiency,justify the persistently low C3 values without associated abnormal complement activation

Key words: C3 glomerulonephritis, Post-infectious glomerulonephritis, Nephrotic Proteinuria

POSTER - PO 67

ALOPECIA AREATA IN TWO PATIENTS WITH STEROID-RESISTANT NEPHROTIC SYNDROME

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Objectives:
Alopecia areata is an autoimmune-based non scarring alopecia with low prevalence in children. It has been associated with various autoimmune diseases (such as Hashimotos's thyroiditis), some HLA (DQ3, DQ7, DR4 y DR11) and triggers like stress. However, there is little bibliography about its association with nephrotic syndrome.
There are local treatments that can help regrow hair and systemic treatments that influence the course of the disease, including Janus kinase (JAK) inhibitors.

Methods:
We present two patients with steroid-resistant nephrotic syndrome who developed alopecia areata.

Results:
CASE 1
15-year-old male diagnosed with steroid-resistant nephrotic syndrome, treated with tacrolimus, mycophenolate mofetil and low-dose prednisone. In 2020, he presented an alopecia areata patch on the scalp. Initially, he received topical treatment with corticosteroids, tacrolimus and minoxidil without response. Progressivelly, patchy alopecia progressed to alopecia universalis. He remained in complete remission of the nephrotic syndrome during this time. For these reason, change to systemic cyclosporine was ruled out. Oral treatment with JAK inhibitor was offered but it was rejected by the family. Currently, he presents universal alopecia with stable nephrotic syndrome.
CASE 2
12-year-old woman with steroid-resistant nephrotic syndrome since the age of 3. She received tratment with tacrolimus and mycophenolate mofetil. At eleven years old she presented alopecia areata patch on the scalp that was treated with minoxidil and topic corticosteroids without improvement. On the contrary, it progressed towards alopecia universalis. Treatment with mycophenolate mofetil was stopped. Meanwhile systemic tofacitinib (JAK inhibitor) was started. She remains currently in complete remission of the nephrotic syndrome. The follow-up time is still insufficient to evaluate the new treatment efficacy.

Conclusion:
There is not enough information about the association between nephrotic syndrome and alopecia areata. The use of JAK inhibitors may be an alternative for the concomitant treatment of both pathologies

Key words:: alopecia areata, nephrotic syndrome, Janus kinase inhibitors

POSTER - PO 68

A RELEVÂNCIA DO ESTUDO GENÉTICO NA GLOMERULOESCLEROSE SEGMENTAR FOCAL: A PROPÓSITO DE UM CASO CLÍNICO

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Introduction:
A glomeruloesclerose segmentar focal (GESF) é uma glomerulopatia com prevalência crescente em idade pediátrica. A determinação da sua etiologia assume-se de extrema importância pelas implicações terapêuticas e prognósticas, subdividindo-se em causas primárias, secundárias e genéticas, a última constituindo até 30% de todas as formas de GESF.

Results:
Adolescente, 13 anos, sexo masculino, com história paterna de doença renal crónica (DRC) estadio 5 diagnosticada aos 18 anos de etiologia indeterminada, referenciado à consulta por proteinúria significativa persistente (rácio proteínas:creatinina 0.8 mg/mg; albumina:creatinina 603 mg/g) documentada em estudo de enurese secundária não monossintomática. Normotenso e sem alteração da função renal (TFG 118 mL/min/1.73m2 pela fórmula de Schwartz). Excluídas causas tubular e de sobrecarga de proteínas, com prosseguimento de estudo para causa glomerular. Neste âmbito, realizou ecografia renal com evidência de discreta hiperecogenicidade das pirâmides renais; estudo imunológico normal e excluídas causas tóxicas e infeciosas. Dada idade do doente e história familiar, realizou biópsia renal com evidência histológica de 1 glomérulo com esclerose segmentar mas restantes achados de diagnóstico impreciso, e estudo genético por painel NGS com deteção de mutação em heterozigotia c.605A>G p. (Asn202Ser) no gene INF2, associada a GESF tipo 5, de transmissão autossómica dominante. Neste contexto, iniciou terapêutica com inibidor da enzima de conversão da angiotensina. Até ao momento, mantém-se com proteinúria significativa, sem manifestações de síndrome nefrótico.

Conclusion:
Com o presente caso, pretende-se relembrar a necessidade de considerar uma doença monogénica na investigação diagnóstica de GESF, particularmente em doentes com história familiar de DRC. De facto, a identificação de causa genética tem impacto na gestão terapêutica do doente, obviando o recurso à imunossupressão e aos efeitos laterais desta pela reconhecida associação da etiologia genética a formas corticoresistente de proteinúria / síndrome nefrótico. O estudo genético possibilita ainda o aconselhamento genético familiar e seleção de dadores renais apropriados.

Key words: GESF, estudo genético, proteinúria

POSTER - PO 69

AGLOMERULOPATHY IN HORSESHOE KIDNEY

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Objectives:
Horseshoe kidney (HR) is the most common fusion anomaly. It is usually asymptomatic, although some cases develop complications. Its association with polymalformation syndromes and chromosomopathies has been described. There are few reports on its association with glomerulopathies, these being especially proteinuric (membranous GN, nephrotic syndrome or FSGS). Renal biopsy is essential for diagnosis but can be challenging for the nephrologist.

Methods:
We report a patient affected by a glomerulopathy associated with horseshoe Kidney.

Results:
We report an 15-year-old Ecuatorian boy who had presented an episode of gross glomerular hematuria with fever. Among the family history, terminal CKD of unknown cause in maternal uncle. Normal GFR and no HTA data. Ultrasound showed horseshoe kidney. Normal immune profile. AngioMRI shows 3 accessory polar arteries. During follow-up, he presented episodes of gross hematuria coinciding with infectious symptoms and persistent microscopic hematuria associated with significant proteinuria rising to the nephrotic range. Normal GFR and no HTA data. Ophthalmological study and normal audiometry. Treatment with antiproteinurics was started and a laparoscopic percutaneous renal biopsy was performed without obtaining an optimal sample. The study was extended with a genetic panel of glomerulopathies and familial hematuria (COL4A3, COL4A4 and COL4A5), which was negative. A new percutaneous renal biopsy with interventional radiology was performed, obtaining a viable sample where minimal glomerular alterations were evidenced without deposits in direct immunofluorescence. That suggested the possibility of disease with minimal changes. An electron microscopy study was requested and he started treatment with steroids at 60 mg/m2/day. The result of the study was compatible with thin basement membrane nephropathy, so the steroids were interrupted. He currently maintains microscopic hematuria and significant almost nephrotic proteinuria with normal GFR.

Conclusion:
The coexistence of glomerulopathy, especially hematuric, and HR is rare. Renal biopsy is an essential tool for diagnosis, which can be a real challenge.

Key words: HORSESHOE KIDNEY

POSTER - PO 70

FAMILIAL HEMATURIA: CASE RESPORT FROM A SECONDARY HOSPITAL

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Introduction:
Alport syndrome (AS) and thin basal membrane disease (TBMN) have glomerular hematuria as a clinical fact. However, genetics and clinical progression remain different.
AS is characterized by renal failure, hearing impairment, lenticonus and retinal flecks, glomerular basement membrane (GBM) lamination with abnormal collagen IV composition, and a COL4A3;4;5 gene mutation. 85% of families have a X-linked inheritance.
TBMN have isolated hematuria caused by heterozygous mutations in the COL4A3 or COL4A4 genes; It usually represents the carrier state of an autosomal recessive AS.

Objectives:
To describe a case report series of patients diagnosed of familial hematuria in our institution.

Methods:
Case report communication.

Results:
We have documented 4 cases of this disease, so far. All of them start follow up due to familial antecedents (FA).
Case 1. Woman. Starts study at 13. Asymptomatic. Blood and urine analysis normal. Genetics: heterozygous mutation in the COL4A3 gene, similiar to her father. FA: Father with sensorineural deafness.
Case 2. Male. Starts study at 6. Asymptomatic. Isolated microhematuria. Genetics with heterozygous mutation in the COL4A3 gene, similar to her mother. AF: mother with hematuria and proteinuria with normal renal function.
Case 3. Woman. Starts study at 7. Blood and urine analysis normal. Genetics with heterozygous mutation in the COL4A3 gene, similar to her mother. AF: mother asynmptomatic; maternal uncle with renal failure and kidney transplantation.
Case 4. Woman. Starts study at 2. Isolated myopia. Microhematuria. Genetics similar to the one of her father (mutation in COL4A5 gene). AF: father with sensorineural deafness and kidney transplantation.

Conclusion:
No renal biopsy was performed in any patient. Patients 1,2,3 fulfill TBMN standards; patient 4 meets SA criteria.
To carry out a correct management of familial hematuria both genetics and inheritage mechanisms are vital. They provide accurate information for a follow up, prognosis and the appropriate genetic counseling in each case.

Key words: Hematuria, Alport Syndrome, Thin basement membrane nephropathy

POSTER - PO 71

CHRONIC KIDNEY DISEASE SECONDARY TO ANCA-POSITIVE FOCAL SEGMENTAL GLOMERULOSCLEROSIS.

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Objectives:
Focal segmental glomerulosclerosis (FSG) is a clinical-pathological entity that causes pediatric chronic kidney disease (CKD). Cases associated with positive autoimmunity are infrequent in Pediatrics.

Methods:
Describe the case of a pediatric patient with CKD with a histological pattern of FSG and positive autoimmunity for antineutrophil cytoplasmic antibodies (ANCA).

Results:
Review of clinical history and renal biopsy. We present the case of a 9-year-old female patient, referred to the hospital due to iron deficiency anemia detected in the context of an upper respiratory tract infection. Initial glomerular renal function is preserved (eGFR Schwartz 2009 100ml/min/1.73m2) although there is an increase in ESR up to 95mm/hour with hypergammaglobulinemia (IgG 2326 mg/dL) and microhaematuria. She has no relevant personal history. As a family history, the mother underwent splenectomy due to autoimmune hemolytic anemia in adolescence. In the following 6 months, she develops decreased renal function (GFR Schwarzt 2009 48 ml/min/1.73m2) with non-nephrotic proteinuria and microalbuminuria, requiring oral enalapril. Renal ultrasound and scintigraphy are normal. In the evolution, Hashimoto's Thyroiditis is also diagnosed, well controlled with thyroid hormone. She reached a stage of G3bA3 chronic kidney disease, controlled with bicarbonate, oral iron, and subcutaneous erythropoietin. In extended studies (serology, immunology, and autoimmunity), positivity for p-ANCA (MPO-ANCA > 134 U/mL) as well as for circulating immunocomplexes in serial analyzes are detected. Renal biopsy is performed at a referral center with a finding of focal segmental sclerosis (NOS variant) with global glomerular sclerosis of 42%. Genetic study without finding a mutation. Given the autoimmune component, it was decided to add immunosuppressive therapy with rituximab.

Conclusion:
Despite not presenting clinically or histopathologically like classic vasculitis, we consider that the etiopathogenic role of ANCA is essential in this patient. Therefore, following the therapeutic recommendations of the current literature, she is currently on immunosuppressive therapy with rituximab.

Key words: Chronic kidney disease, Focal segmental glomerulosclerosis, Autoimmunity, Antibodies, Anti-neutrophil cytoplasmic antibody (ANCA)

POSTER - PO 72

NEPHROTIC SYNDROME IN PEDIATRIC SHÖNLEIN-HENOCH PURPURA

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Objectives:
Schönlein-Henoch Purpura (SHP) or IgA vasculitis is a systemic small-vessel vasculitis characterized by leukocytoclasty and deposition of IgA1 immune complexes in vascular wall. Renal manifestations in childhood SHP are hematuria and/or proteinuria with different prognosis.

Methods:
To know kidney function in children with Schönlein-Henoch Purpura (SHP) or IgA vasculitis.

Results:
METHODS: Clinical history review.
CLINICAL CASE: 5-year-old girl diagnosed with SHP reviewed in the Pediatric Nephrology clinic with initially normal kidney function. She had joint pain, and prescribed treatment with oral prednisone (0.5mg/kg/d). After 5 days, she consults again due to abdominal pain, pretibial edema and persistent joint pain. Blood biochemistry showed increased nitrogen products with acute renal failure. Urinalysis showed microscopic hematuria and nephrotic proteinuria. Abdominal ultrasound was normal. Treatment with ACE inhibitors was associated and it was decided to perform a renal biopsy, which was compatible with SHP nephritis. Oral corticosteroid therapy with gastric protection and anti-osteoporotic prophylaxis was administered according to infant nephrotic syndrome protocol. In absence of response, she required an 8-week cycle of cyclophosphamide, with nephrotic syndrome remission, disappearance of hematuria, and normal renal function. After 2 years, she presented a 2nd outbreak of SHP with reappearance of steroid-resistant nephrotic syndrome, requiring oral tacrolimus to induce remission.

Conclusion:
Renal manifestations in childhood SHP appear in 20-55% cases, being the most important prognostic factor. Glomerulonephritis occurs due to mesangial IgA deposition that leads to progressive sclerosis. Up to 20% will develop nephritic/nephrotic syndrome with poor prognosis, corticosteroid resistance and the need for immunotherapy. Its early diagnosis and treatment are essential to improve the vital prognosis of these children.

Key words: IgA vasculitis, Schönlein-Henoch Purpura, chronic kidney disease, steroid-resistant nephrotic syndrome

POSTER - PO 73

RAPIDLY PROGRESSIVE GLOMERULONEPHRITIS (RPGN) IN A PATIENT WITH MICROHEMATURIA AND EPISODES OF MACROMEHATURIA FROM THE INFANCY

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Objectives:
Rapidly progressive glomerulonephritis (RPGN) is an acute nephritic syndrome, with progressive loss of renal function in a short period of time. It constitutes <15% of glomerulopathies, and histology shows epithelial crescents that evolve to fibrous. Pauciimmune(type 3 RPGN) doesn't present immunoglobulin deposits. Negative-ANCA has a worse prognosis.

Results:
We report an 11-year-old boy, with a personal history of microhematuria under study between the age of 2 and 4, suspicion of IgA nephropathy, and mother with microhematuria and episodes of macrohematuria. He presented high creatinine (GFR-Schwartz 22 mL/min/1.73 m2), hypoproteinemia, normocytic anemia and hypercholesterolemia in blood test made because of knee pain.
In anamnesis, he has referred episodes of macrohematuria since the age of 4, every 2-3 months, in context of infections. In the last 6 months, he has also presented nausea, fatigue, halitosis, day-night incontinence and abdominal pain. He had received ibuprofen two months before for knee pain.On physical examination, he only had uremic fetor with normal blood pressure.
In diagnostic study, he had poliuria, high cratinine and urea (GFR-Schwartz 17 mL/min/1,73m2), hyperkalemia and hyperparathyroidism.The immunity study was normal. Urine test had proteinuria in nephrotic range, glycosuria and hematuria. Serologies were negative and urinary tract ultrasound was normal. He started treatment for chronic kidney disease.
Ultrasound-guided percutaneous renal biopsy was performed and it reported "RPGN extracapillary pauciimmune sclerotic class (>50% of sclerotic glomeruli globally) with presence of epithelial/fibroepithelial crescents in 36% and data of acute tubular necrosis. Negative IF.Negative ANCA".
The patient was treated with three bolus doses of IV-steroid on consecutive days, followed by two bolus doses of cyclophosphamide, with no improvement in renal function.

Conclusion:
RPGN is a rare cause of acute kidney damage in children, especially RPGN ANCA-negative pauciimune. Although prognosis is bad, early initiation of treatment with steroids and cyclophosphamide may minimize the degree of irreversible renal damage.

Key words: Rapidly progressive glomerulonephritis, Microhematuria, Macrohematuria, Pauciinmune vasculitis

POSTER - PO 74

MORE THAN ORTHOSTATIC PROTEINURIA?

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Introduction:
Orthostatic proteinuria (OP) is the most frequent cause of isolated proteinuria in children, especially in adolescent males (70%). Drug-induced acute interstitial nephritis (AIN) is suspected when there is a temporal link between the onset of kidney dysfunction and drug exposure, as well as its resolution after the drug discontinuation.

Results:
A 16-year-old male patient presented with proteinuria. He had been diagnosed with psoriatic arthritis two years before, for which he was being treated with methotrexate for one year. He was also on minocycline for acne vulgaris treatment for six months.

A routine urinalysis detected proteinuria (3+). He reported no history of lower urinary tract symptoms, macroscopic urine changes, recent infections, or trips. Physical examination was unremarkable, including blood pressure within the normal range for age, height, and sex (<P90).

Daytime 12h-proteinuria was 27 mg/m2/hour, while nighttime 12h-proteinuria was 4 mg/m2/hour. He had no haematuria, glycosuria, urine concentration defect or other signs of tubular dysfunction. Urine sediment was normal. Eosinophil blood count was normal. Serum creatinine was 0,8 mg/dL. Albumin, C3, C4, ANAs, and ANCAs were also within the normal reference values.

Isolated orthostatic proteinuria was confirmed in at least three occasions over a six-month period. Doppler ultrasound did not show signs of nutcracker phenomenon.

As articular complaints subsided, methotrexate was stopped as per Rheumatology's team recommendation.

Three months after stopping therapy with methotrexate, which coincided with the replacement of minocycline with isotretinoin, 12h-daytime proteinuria normalised (1.2 mg/m2/h). Therefore, kidney biopsy was postponed and the patient is under watchful waiting surveillance.

Conclusion:
The differential diagnosis between orthostatic and persistent proteinuria is crucial to define patient prognosis and guide further workup and management.

Key words: orthostatic proteinuria, proteinuria

POSTER - PO 75

EMERGING HEMATURIA, ALSO RELATED TO WORSE SOCIO-SANITARY CONDITIONS

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Objectives:
We present th case of a 4-year-old boy, under follow-up for hematuria whose etiological findig was due to other extra-urinary affectations.

Methods:
Third child of parentes from Pakistan. Second degree consanguineous. It begins whith macroscopic hamaturia, whithouth urinary symptoms, mild general involvement justified by previous viral infection. The child hasn't got edema, his tension and analysis are normal. Renal function of 98 for height, C3, C4, coagulation, electrolytes ant hematymetry normal. Macroscopic hematuria whith a protein content of 22 mg/m2/h. Normal kidney and bladder ultrasound. Symptomatic measures are taken, rest, continues for 18 days with hematuria, pregressively decreasing until it appears only microscopically at 26 days. Mild proteinuria, index 0,4 mg/g, No MAU.
In subsequent follow-up, he began wiht astenia, diarrheal stools and weight loss. Normal analysis, except for mild lymphocytosis and eosinophilia of 6%. Stool crop performed, schistosomiasis appears. Urine is analyzed, microscopic hematuria persists, but schistosome eggs are not visualized. Normal kidney ultrasound. Treatment with praziquantel was started, the intestinal symptoms progressively subsided and hematuria resolved in 20 days. Analysis of siblings and parents was negative for schistosome and hematuria. They had traveled to Pakistan 8 months ago. Subsequent follo-ups have not returned with hematuria. He manteins a weight in p3, bad eater but no new episodes. Rehistorying the onset of hematuria, they reported posible skin erythema and radiological analysis evaluated normal, a 12 mm post void residue was described, which could be considered subsequently as a schistosome nodule.

Conclusion:
The origin could have been a bladder schistosomiasis, it was no thougt, so the evaluation 2,5 months later, it may not be found at a sufficient level for its detection. In addition, urine collection was not done several days. The evolution after treatment suggest that it was indeed bladder schistosomiasis and should be included in the differential diagnosis.

Key words: Hematuria, schistosomiasis

POSTER - PO 76

EGENETIC STUDY IN FAMILIAL NEPHROTIC SYNDROME

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Introduction:
The clinical evaluation of children with idiopathic nephrotic syndrome (NS) is based on a complete medical history, examination, and laboratory tests. Renal biopsy, unlike adults, is generally not performed early in the disease. The genetic study is recommended in people over 12 years of age, extrarenal manifestations, family history of NS and in cases of corticosteroid resistance.

Methods:
Clinical case description

Results:
A 9-year-old male from Morocco diagnosed with NS at 16 months, treated with prednisone with good response but frequent relapses. At 4 years old, he started mycophenolate mofetil (MMF) with a good response, withdrawn before 2 years due to suspected hepatotoxicity, continuing with high-dose prednisone until he arrived at our center.
Family history: Parents from Morocco, not consanguinity, healthy. A 3-year-old brother with corticosensitive NS since he was 2 years old. A 10-year-old cousin (paternal family) with NS since he was 3 years old, corticosensitive, without current treatment.
Physical exploration: Weight 30.6 kg, height 130 cm; Blood pressure 107/66. Male phenotype. No dysmorphia or cushingoid fascies, mild eyelid edema; globular abdomen, without masses or megalia. No other alterations.
Complementary studies: Biochemistry with NS pattern. Hemogram, C3 and C4 normal. Negative study for autoimmune hepatitis.
Genetic study by massive sequencing: INF2 gene change in heterozygosis (c.2419-7A>G,) with potential effect to produce dysfunctional proteins. LAMB2 gene, change in heterozygosity (c.1423> T, p.R475W), with probably deleterious or neutral effect. Both, not reported in databases or scientific literature, classified as variants of uncertain significance (USV)

Conclusion:
The genetic study is essential in cases of familial and/or syndromic NS, being indicated even before performing a renal biopsy.
Genetic diagnosis is important to define the disease, help in making therapeutic decisions and provide genetic counseling in the face of the risk of recurrences.

Key words: nephrotic syndrome, genetic nephrotic synd

POSTER - PO 77

INFEÇÃO A YERSINIA ENTEROCOLÍTICA COMPLICADA COM GLOMERULONEFRITE: UMA ENTIDADE RARA, A PROPÓSITO DE UM CASO CLÍNICO

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Introduction:
A infeção por Yersinia enterolítica cursa, habitualmente, com clínica de gastroenterite aguda. Raramente, ocorre disseminação sistémica e associam-se complicações.

Results:
Apresentamos o caso de uma adolescente de 12 anos, previamente saudável, que se apresentou com febre, dor abdominal e diarreia. À admissão com tensões arteriais sistólicas no percentil 95-99. O estudo analítico revelou creatinina 1,4mg/dL, ureia 38mg/L e proteína C reativa 155mg/L. A taxa de filtração glomerular (TFG), segundo Schwartz/2009, era 47mL/min/1.73m2. Iniciou fluidoterapia endovenosa e 12h depois apresentava creatinina 1,3mg/dL, ureia 40mg/dL, cistatina C de 1,21mg/L – TFG 51mL/min/1.73m2. O estudo da urina revelou 10 eritrócitos/uL no sedimento, um rácio proteínasU/creatininaU de 0,5 e fração excretada de Na+ 2.9%. A ecografia abdomino-pélvica mostrou adenite mesentérica e rins de dimensões aumentadas (eixo polar esquerdo 13,3cm e direito 13,9cm), com acentuação da diferenciação cortico-medular por aumento da ecogenicidade cortical, a favor de nefropatia médica.
Admitida no internamento de Pediatria, apresentou melhoria clínica progressiva, diurese preservada e TA abaixo do percentil 95-99. Cerca de 48 horas depois, apresentava creatinina de 1,3mg/dL, ureia 34mg/dL, TFG 51mL/min/1,73m2. O exame microbiológico de fezes revelou Yersinia enterolítica e, dada a melhoria clínica, a antibioterapia não foi instituída.
Um mês depois, em consulta, verificou-se normalização da função renal com TGF 112mL/min/1,73m2, estudo imunológico normal, mantendo ainda proteinúria moderada. Aos 12 meses de evolução apresentava diminuição progressiva da proteinúria, conservação da função renal e desaparecimento das alterações ecográficas renais. Assim, pode presumivelmente assumir-se um diagnóstico de glomerulonefrite associada à infeção por Yersinia enteroclitica.

Conclusion:
Com este caso clínico os autores pretendem alertar para um diagnóstico raro, pouco descrito na literatura, e para as possíveis complicações renais associadas à infeção por Yersinia enterocolítica.

Key words: Glomerulonefrite, Yersinia enterocolítica, Nefropatia

POSTER - PO 78

PERIORBITAL EDEMA, WHAT NOT TO FORGET

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Introduction:
Nephrotic syndrome can affect children of any age, but is most common among school-aged children and adolescents. It is characterized by significant proteinuria and hypoalbuminemia (less than 3 /dL) initial presence of edema can improve clinical suspicion. The age of onset may be orientative to discern its etiology.

Results:
A previously healthy 6-year-old male, presents to the emergency department with bilateral morning-predominant periorbital edema, over the course of
three days, and a 2kg weight gain. Fever, abdominal pain and urine complains were denied. In the first days of the symptoms an allergic reaction was suspected and medicated with an anti-histaminic without clinical improvement. On physical examination, the patient was hemodynamically stable, with generalized edema but no associated redness or pain. Dipstick test performed showed proteins 3+, and urinalysis confirmed proteinuria (500mg/dL),with a Protein/Creatinine ratio of 11.1mg/mg in a single urine sample. Blood analysis revealed hypoalbuminemia (2.2g/dL), hypoproteinemia (4.0g/dL), and hyperlipidemia (total cholesterol 418 mg/dl, LDL 294 mg/dl). There were no changes in renal function, coagulation studies and the complement levels were normal. The 24 hour-urinary collected confirmed the proteinuria. Oral prednisone was initiated (60mg/m2 per day), which resulted in complete remission after 10 days. After both clinical and analytical improvement, the patient was discharged while maintaining corticosteroid treatment and regular follow-up, with no relapse of symptoms. Given the diagnostic criteria and clinical findings, idiopathic nephrotic syndrome was considered.

Conclusion:
The authors believe it is relevant to share this case to emphasize the necessity of considering this entity as a potential cause of edema at all levels of care. In fact, periorbital edema is a common manifestation that can easily be misinterpreted as an allergic response. Furthermore, we wish to highlight the importance of regular follow-up appointments and monitoring for proteinuria. This can help detect relapses early and minimize the risk of complications.

Key words: Idiopathic Nephrotic Syndrome, Differential diagnosis, Periorbital edema

POSTER - PO 79

NEPHROTIC SYNDROME ATYPICAL PRESENTATION: A CASE REPORT

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Introduction:
Nephrotic syndrome (NS) is the commonest glomerular disease in children but it's incidence in the first year of life is very low. When it's present at birth or develops in the first three months, it is considered congenital, while if it develops between three and twelve months of age, it is called infantile. Most of these situations have a genetic cause and a poor outcome.

Results:
We report a case of a previously healthy 9-month-old caucasian twin girl with no prenatal history. Non-consanguineous parents, history of hypertension in her father. The patient was admitted in the Emergency Department in september 2022 with two days of fever, three days before admission, and odynophagia. Parents also reported hypoactivity and lower oral intake, along with a single episode of vomiting at admission. No other symptoms were reported. On physical examination, she appeared sick, was hypoactive, with sunken fontanelle. Apyretic, blood pressure in the 5-50th percentile for age and gender. No evidence of edema or significant weight changes. Laboratory work-up revealed nephrotic range proteinuria (urinary protein:creatinine ratio 98.1mg/mg), hyperlipidemia (cholesterol 291mmol/L), hypoalbuminemia (2.2g/dL), hyponatremia (125mmol/L), no evidence of renal injury, normal haemoglobin and inflammatory markers. Negative microbiologic and serologic tests for infectious causes. Normal abdominal ultrasound. Treatment was initiated with prednisolone 60mg/m2/day. Albumin perfusions were performed on admission and on the second and third day of hospitalization. After the initial approach, the child was transferred to a reference center. Progressive improvement was achieved and proteinuria completely resolved after eight days of glucocorticoid therapy. A genetic study is ongoing.

Conclusion:
This case reports a rare type of infantile NS, diagnosed in the absence of characteristic symptoms. Most cases are unresponsive to corticosteroids and progress to chronic kidney disease. However, the steroid responsiveness of this patient predicts a favorable outcome. Even so, continuous monitoring is mandatory.

Key words: Nephrotic Syndrome, edema, proteinuria

POSTER - PO 80

IDIOPATHIC NEPHROTIC SYNDROME AND STEROID DEPENDENCE – MANAGEMENT OF A CLINICAL CASE

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Introduction:
Idiopathic nephrotic syndrome (INS) is the most common cause of glomerular disease in children. Despite their numerous known adverse effects, steroids remain the first line treatment.
About 50% of INS patients relapse frequently or are steroid-dependent. In these cases, steroid-sparing agents play an important role.

Results:
Case Presentation:
A 22-month-old male child, with a history of prematurity of 35 weeks, with no other relevant medical history, was diagnosed with INS, during a viral infection. His mother was battling multiple sclerosis. As for the other family relatives, medical history was unremarkable.
The patient responded to steroid treatment 7 days after admission and was discharged on day 8, medicated with prednisolone 60 mg/m2/day and a proton pump inhibitor. Steroid tapering began three weeks after discharge, under guidance from a Paediatric Nephrologist. During tapering, he had two relapses - the first at 24 months and the second at 2 years and 2 months, both with 20 mg prednisolone on alternating days. Given steroid-dependent INS, he was referred to a Nephrology Reference Centre. A new relapse occurred once tapering was attempted, and the patient was started on mycophenolate mofetil (333 mg/m2/dose). Currently, at 2 years and 6 months of age, the patient is on steroid tapering and mycophenolate increase, with no record of new relapses.

Conclusion:
The 2022 International Association of Pediatric Nephrology's Guidelines (IPNA) no longer recommend a tapering schedule of steroids during alternate day dosing (below 40 mg/m2). Furthermore, recent studies suggest that extending initial steroid treatment doesn't appear to improve clinical outcomes as time to first relapse or the number of relapses.
With presenting this case, based on the young age of presentation and the dependence on high steroids dosing, we intend to encourage discussion about the increasingly need for individualized therapeutic regimens.

Key words: nephrotic syndrome, steroid treatment, steroid-sparing agents

POSTER - PO 81

NEPHROTIC PROTEINURIA IN AN ADOLESCENT WITH SYMPTOMS OF MULTISYSTEMIC DISEASE

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Introduction:
Childhood-onset systemic lupus erythematosus (cSLE) is a rare and chronic autoimmune disease that typically results in multiorganic involvement, with lupus nephritis being one of the major indicators of poor prognosis.

Objectives:
Our objective is to expose a case that illustrates a diagnosis of cSLE with lupus nephritis at presentation, in an adolescent with multisystemic symptoms since several months ago.
Métodos
We report a case of an adolescent admitted in the emergency room (ER) of a level III portuguese hospital.

Results:
Previously healthy teenager with intermittent pain, swollen joints and accentuated loss of hair with 6 months of evolution, requiring multiple medical observations, arrives at the ER complaining of arthralgias and edema of both hands and legs, fever, anorexia, abdominal distension and oral ulcers. Physical examination revealed hypertension, cervical adenopathies, discrete cheilitis, bibasal diminished vesicular murmur, in association with vasculitic lesions and livedo reticularis of lower limbs. The laboratory workup showed nephrotic proteinuria, renal impairment, normochromic and normocytic anemia, leucopenia, complement consumption, elevated erythrocyte sedimentation rate and positive antinuclear and anti-double stranded DNA antibodies. Ultrasound revealed bilateral pleural effusion and low volume ascites. A diagnosis of cSLE with renal, joint, hematologic and serous involvement was established. She immediately initiated hydroxychloroquine 5mg/kg/day in association with methylprednisolone pulses 20 mg/Kg/day during 5 days, which were later replaced by oral prednisolone 40mg/Kg/day. Later on, a renal biopsy was performed in a specialized unit that revealed lupus nephritis class III.

Conclusion:
This case enhances the importance of an holistic approach when we evaluate a patient with multisystemic disease like cSLE. A prompt diagnosis and treatment initiation is crucial to improve patients prognosis in cSLE.

Key words: Childhood-onset systemic lupus erythematosus, Lupus nephritis, Nephrotic Proteinuria

POSTER - PO 82

EFFICACY OF LEVAMISOLE FOR MAINTENANCE OF REMISSION IN CHILDREN WITH STEROID-DEPENDENT NEPHROTIC SYNDROME OR WITH FREQUENT RELAPSES

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Introduction:
Nephrotic syndrome is the most common primary glomerulopathy in childhood. It is defined as the presence of proteinuria (>40 mg/m2/h) together with dyslipidemia, hypoalbuminemia (<2.5 g/dL), edema and endocrine disorders. Corticosteroids are the mainstay of treatment, but about 50% of patients evolve to corticodependence, which exposes them to high doses of corticosteroids throughout their lives. Levamisole is an anthelmintic with not well known immunomodulatory activity, without immunosuppressive component unlike the other drugs used. It is currently relegated in Spanish guidelines as second or third line despite its reduced number of side effects

Objectives
To determine the effectiveness of levamisole to keep children with corticodependent nephrotic syndrome in remission.

Methods:
This is an observational, descriptive, longitudinal, retrospective, case series study of the population with corticodependent nephrotic syndrome who received levamisole in the pediatric nephrology unit of the Hospital Universtario Santa Lucia from January 1, 2007 to February 28, 2023

Results:
Ten patients were included, 60% male. The mean age at diagnosis was 3.3 years. A dose of 2.5 mg/kg every 48 hours was prescribed. 50% of the sample took cyclophosphamide before taking levamisole. Two (20%) patients had mild side effects. The median cumulative corticosteroid dose before levamisole was 6040 mg/m2 (RI 2422) and decreased 1513 mg/m2 (3523) after the end of levamisole treatment ($p<0.05$). The median number of relapses was 1 relapse every 6 months before treatment, reduced to 0.50 relapses/6 months during treatment and finally decreased to 0.12 relapses/6 months ($p<0.05$). No significant differences were found between sexes or in those who had previously taken cyclophosphamide, although this group started with higher doses of corticoid and a greater number of relapses.

Conclusion:
Levamisole is a useful drug to keep children with corticodependent nephrotic syndrome in remission and reduces the doses of corticosteroids to which they are exposed.

Key words: Nephrotic syndrome, levamisole, pediatrics, corticodependent, relapses.

POSTER - PO 83

HOW TO TREAT ANTI-FACTOR H ASSOCIATED AHUS – CASE REPORT

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Introduction:
Anti-factor H antibodies (aFH) associated aHUS is a unique subgroup of aHUS that is more prevalent in the pediatric population, occurring in about 24% of all children with aHUS. Around 50% of aHUS cases arise from genetic mutations that encode regulatory proteins of the alternate complement pathway. Factor H (FH) is one of these important regulatory proteins. The autoantibodies impair the interaction of FH with C3b causing dysregulation of the alternate pathway of the complement.Eculizumab, an anti-C5 antibody that targets terminal complement activation, has transformed outcomes in aHUS. Eculizumab is preferred in patients who possess FH mutations, it further reduces the likelihood of recurrences and seems to be a safe option for the treatment of aFH aHUS in the paediatric population.
Several immunosuppressors, like prednisone, cyclophosphamide, azathioprine, mycophenolate mofetil (MMF) and rituximab, have been used as inhibitors of aFH production and as maintenance therapy, reducing relapse risk.

Results:
We present the case of a 6 year-old boy, previously healthy who was diagnosed with aHUS after exclusion of STEC-HUS, thrombotic thrombocytopenic purpura and infectious etiologies. Eculizumab was started about 24h after admission, with excellent clinical and laboratorial response. He was discharged after eight days, maintaining eculizumab treatment fortnightly. Further investigation showed elevated aFH (11100 UA/mL) and a homozygous CFHR3-CFHR1 deletion. Therapy with MMF was started, 1200 mg/m2/day. The titre of aHF have decreased but not normalized. The interval between eculizumab doses was progressively widened up to every 5 weeks. Currently, 15 months after diagnosis, he remains clinically stable with no relapses.

Conclusion:
The management of this patient presents a challenge, since there are very few studies regarding the optimal duration of eculizumab, under what circumstances its discontinuation may be safe, and for how long additional immunosuppression should be carried out.

Key words: ANti-factor H Antibodies ;aHUS; Eculizumab

POSTER - PO 84

AN ATYPICAL CASE OF ATYPICAL HEMOLYTIC-UREMIC SYNDROME

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Introduction:
Haemolytic-uremic syndrome (HUS) is caused by thrombotic microangiopathy, which leads to endothelial injury, platelet activation and aggregation, resulting in haemolytic anaemia and acute kidney injury. The classic triad of symptoms are non-immune haemolytic anaemia, thrombocytopenia, and acute kidney disease. While these are the main symptoms, it may also develop lesser-known and frequent neurological, pulmonary, hepatic, and musculoskeletal disorders.
Objectivos
To describe a clinical case of atypical-HUS with hypertension, acute kidney failure, rhabdomyolysis, and neurological involvement as leading symptoms.

Methods:
Clinical case report and literature review.

Results:
A 14-year-old male presented with complex feverish seizure and visual disorders. On admission, he presented with hypertension, creatinine of 2.95 mg/dl, hyperuricemia of 17.7 mg/dl, without other ionic disturbances. Haemoglobin was 17.7 g/dl, platelet count 104,000/mm3, and in 24 hours, it dropped to 12.2 g/dl and 35,000/mm3, respectively. The direct Coombs test was negative, haptoglobin 25.1 mg/ml. GOT 2033 U/L, GPT 1161 U/L, bilirubin 3.1 mg/dl, indirect bilirubin 1.7 mg/dl, LDH 2445 U/L, amylase 356 U/L, lipase 150 U/L, CPK-NAC 98186 U/L. C3 74 mg/dl, with normal C4 and immunoglobulin levels. Hemofiltration was initiated 36 hours after admission and maintained for the next 8 days. The first dose of eculizumab was administered on the fifth day, with early improvement in neurological and haematological findings, as well as renal function, with creatinine of 1.22 mg/dl and eGFR Schwartz 2009 of 61 ml/min/1.73m2. In the follow-up, the patient showed progressive improvement in blood pressure, treated with amlodipine, and in renal function with an eGFR Schwartz 2009 of 83 ml/min/1.73m2. Factor I was 8 ug/ml, C3 was 60 mg/dl. Genetic testing is not yet available.

Conclusion:
a-HUS is a multisystemic disease, and diagnosis remains clinical. We must suspect this entity if hypertension due to kidney failure is associated with neurological disorders. Rhabdomyolysis, although very rare, is a possible finding in a-HUS.

Key words: Atypical Haemolytic-uremic Syndrome, Renal failure, Eculizumab, Complement

POSTER - PO 85

SÍNDROME HEMOLÍTICO-URÉMICO TÍPICO DE CURSO ATÍPICO.

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Introduction:
El síndrome hemolítico-urémico (SHU) forma parte del espectro de las microangiopatías trombóticas. Supone hasta el 90 % de los casos. Cursa con anemia hemolítica, insuficiencia renal y trombopenia. Es importante un diagnóstico y tratamiento precoz.

Results:
Varón de 4 años con cuadro de diarrea sanguinolenta de 4 días de evolución. Entre las pruebas complementarias destaca crecimiento de E. Coli 0157:H7 productor de toxina Shiga (STEC) en coprocultivo. Sufre un empeoramiento en los primeros días de ingreso con alteración en cifra de plaquetas (mínimo de 17.000/mmc) y hemoglobina (mínima 6.7 g/dL, con datos de hemólisis) manteniendo función renal dentro de la normalidad (creatinina máxima 0.5 mg/dl, basal del paciente 0,42 mg/dL). Precisa transfusión de concentrado de hematíes y medidas de soporte, con adecuada respuesta. Mantiene tensión normal. Se realiza ecografía durante el ingreso con riñones aumentados de tamaño, sin otros hallazgos.
Al mes del alta reingresa por daño renal agudo en contexto de síndrome emético de escasos vómitos (creatinina 2.39 mg/dL) manteniendo hemoglobina y plaquetas dentro de valores normales, sin datos de hemólisis. Nuevamente responde adecuadamente a fluidoterapia, pudiendo darse de alta con una creatinina de 0.59 mg/dL.
Tras el alta mantiene función renal normal.

Conclusion:
Ante un paciente con anemia hemolítica y afectación de órganos diana es importante descartar una microangiopatía trombótica. El SHU-STEC es la causa más frecuente.
Se deben minimizar transfusiones (tanto de plaquetas como de hematíes) en la medida de lo posible.
El tratamiento principal se basa en medidas de soporte.
Es importante un seguimiento estrecho de estos pacientes tras el alta, sobre todo en relación con procesos concomitantes que puedan alterar la función renal en las primeras semanas.

Key words: síndrome hemolítico-urémico, anemia, hemolisis, plaquetopenia, tratamiento soporte

POSTER - PO 86

SA PROPÓSITO DE UN CASO: SÍNDROME HEMOLÍTICO-URÉMICO ATÍPICO, OTRAS FORMAS DE PRESENTACIÓN.

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Introduction:
El síndrome hemolítico-urémico (SHU) forma parte del espectro de las microangiopatías trombóticas. Cursa con anemia hemolítica, insuficiencia renal y trombopenia. Se clasifica en típico y atípico. La causa más frecuente de SHU-atípico son las alteraciones en la vía del complemento; aunque también puede deberse a infecciones por Streptococcus pneumoniae.

Results:
Mujer de 6 años que acude por vómitos y diarrea de 5 días de evolución. En estudio analítico realizado por afectación del estado general destaca elevación de marcadores de función renal (creatinina 2,46 mg/mL). Asocia anemia hemolítica (Hemoglobina 5,5 g/L con elevación de LDH, hiperbilirrubinemia y haptoglobina baja) y descenso de plaquetas. En frotis sanguíneo se observan esquistocitos. Se traslada a UCI pediátrica para monitorización y medidas de soporte; precisando transfusión de plaquetas y hematíes. En controles posteriores sufre deterioro renal (índice proteína/creatinina máximo de 24 mg/mg) y elevación de tensión arterial por lo que se inicia tratamiento con amlodipino. En estudio ampliado se objetiva alteración del complemento (Factor H en límite bajo (30 mg/dL), C3 46 mg/dL y C4 12 mg/dL) con ADAMTS-13, Shiga y resto del estudio normal. Ante probable SHU atípico se inicia tratamiento con eculizumab al décimo día de ingreso (recibe profilaxis vacunal y amoxicilina) con buena evolución y mejoría en las primeras 24 horas de los parámetros analíticos. Estudio genético pendiente de resultado. Continúa seguimiento con una función renal normalizada (Creatinina 0,34 mg/dL e índice proteína/creatinina de 0,5 mg/mg). Mantiene tratamiento con amlodipino con tensiones dentro de la normalidad.

Conclusion:
Ante un paciente con anemia hemolítica y afectación de órganos diana es importante descartar una microangiopatía trombótica.
El pronóstico es peor que en el SHU-típico por lo que ante sospecha se debe iniciar tratamiento con anticuerpos monoclonales (Eculizumab o más novedoso Ravulizumab si cumple criterios) lo antes posible. Como alternativa, valorar plasmaféresis.

Key words: SHU atípico, anemia hemolítica, trombopenia, transfusión, eculizumab