Case description: We present a case of a ten-year-old boy with multiple comorbidities who developed MIS-C after asymptomatic SARS-CoV-2 infection. The patient initially presented hematuria, persistent fever and elevated markers of inflammation, with no other sign of renal affection. Besides, he had a discrete erythematous maculopapular rash on the right lower leg. Within the next two days, his condition worsen despite the broad-spectrum antibiotic therapy. He started to vomit and developed abdominal pain, conjunctivitis, arrhythmia and mild left ventricular systolic dysfunction with hypotension and pleural effusion. High level of clinical suspicion for MIS-C was supported by laboratory findings (elevated ESR, CRP, proBNP, D-dimers and fibrinogen) along with positive IgG SARS-CoV-2 antibodies and negative microbiological cultures. The patient was given intravenous immunoglobulin (IVIG) at a dose of 2 g/kg and began to show instantaneous clinical improvement, including downtrend of fever and inflammatory markers.

Conclusion:

Despite the growing reports of the MIS-C in the literature, there is still paucity of studies describing the various clinical manifestation and laboratory finding in this serious condition which can be easily mistaken for many others inflammatory diseases. Therefore, pediatric professionals must be aware of (many) unusual presentations of COVID-19 associated disease in order to early recognize and treat such challenging patients.

EP-120 HYPERTENSIVE CRISIS AS A PRESENTATION OF COVID-19 IN CHILDREN

Nakysa Hooman , Golnaz Gharehbaghi , Masoud Hashemzadeh - esfahani , Ali Manafi-anari , Behzad Haghighi-aski , Maryam Sakhaei
Aliasghar Clinical Research Development Center, Aliasghar Children Hospital, Iran University Of Medical Sciences, Tehran

Background & Aim: Hypertension consider as a risk factor for the severity of Covid-19 but there is no report of hypertension crisis as an earlier presentation of Covid-19 in pediatrics that is the aim of this study.

Methods: This is a case series collected from March to February 2021. Confirmed SARS-Covid 2 defined by positive RT-PCR of nasopharynx swab, or antibody against it (IgG or IgM), or imaging pattern in favor of it on chest CT-scan . suspicious cases described by clinical sign (fever, diarrhea, cough) and laboratory tests (leucopenia, lymphopeia, rise of CRP, d-dimer, LDH, CPK-MBA, Ferritin, or ALT, with negative cultures for bacteria) and close contact with an infected person. Hypertensive crisis elucidated by acute rise of blood pressure more than stage 2 cut off with potential risk of end organ damage. Results: Four confirmed and two suspicious SARS-COvid-19 cases (4 males, 2 females) were reported. The mean age of patients was 4.2 (range, 1-12) years. Four cases with newly diagnosis of nephrotic syndrome who were on 2mg/kg /day prednisolon developed rapid rises of blood pressure. The other two cases were suffered from Chronic kidney disease stage 5 being on regular dialysis with no edema or sign of fluid overload (HD and CAPD). The lowest systolic blood pressure at the time of admission was 160 mmHg and the highest was 200 mmHg. The diastolic blood pressures were between 100 -155 mmHg. All the patients had normal blood pressure prior to the admission. Three patients (two CKD) received anticovid 19 medications. All patients treated with labetalol infusion titrated to maximum dosage and continued for at least one week and because of poor control of oral antihypertensive medications were added. On follow up, one patient on CAPD died , patient on HD complicated with pericardial effusion, pancreatitis and chronic diarrhea. All nephrotic syndrome patients were resistant to steroid, went on renal biopsy, and calcineurin inhibitor started for massive proteinuria despite being on ARBS /ACEI. All children had resistant hypertention and their blood pressure controlled with four or five antihypertensive medications.

Conclusion: Whether the virus itself can cause exacerbation of hypertension is not yet known but it seems that there is a correlation between COVID-19 infection and hypertension crisis in these cases. Further clinical research with a larger population would be required to determine the clinical significance of these findings.

EP-121 PHENOCOPIES OF CLINICALLY DIAGNOSED ARPKD REVEALED IN CHILDREN

Abdul Halawi [1], Kathrin Burgmaier [1], Anja Buescher [2], Ismail Dursun [3], Matthias Galiano [4], Michaela Gessner [5], Ibrahim Gökce [6], Claudia Kowalewska [7], Djalila Mekahli [8], Sevgi Mir [9], Rukshana Shroff [10], Stella Stabouli [11], Maria Szczepanska [12], Ana Teixeira [13], Lutz Weber [1], Elke WÜhl [14], Simone Wygoda [15], Katarzyna Zachwieja [16], Jörg Dötsch [1], Franz Schaefer [14], Max Liebau [1]
[1] Department Of Pediatrics, Faculty Of Medicine, University Hospital Cologne And University Of Cologne, Cologne, Germany
[2] Department Of Pediatrics Ii, University Hospital Essen, Essen, Germany
[3] Department Of Pediatric Nephrology, Erciyes University, Faculty Of Medicine, Kayseri, Turkey
[4] Department Of Pediatrics And Adolescent Medicine, University Of Erlangen-nÜrnberg, Erlangen, Germany
[5] Department Of General Pediatrics And Hematology/ Oncology, Children's University Hospital Tuebingen, Germany
[6] Research And Training Hospital, Division Of Pediatric Nephrology, Marmara University, Istanbul, Turkey
[7] The Children's Memorial Health Institute, Warsaw, Poland
[8] Department Of Pediatric Nephrology, University Hospitals Leuven, Leuven, Belgium
[9] Department Of Pediatric Nephrology, Ege University Medical Faculty, Izmir, Turkey
[10] Great Ormond Street Hospital For Children Nhs Foundation Trust, London, United Kingdom
[11] First Department Of Pediatrics, Hippokration Hospital, Aristotle University Of Thessaloniki, Thessaloniki, Greece
[12] Department Of Pediatrics, Faculty Of Medical Sciences In Zabrze, Sum In Katowice, Katowice, Poland
[13] Pediatric Nephrology, Centro Hospitalar São João, Porto, Portugal
[14] Division Of Pediatric Nephrology, Center For Pediatrics And Adolescent Medicine, University Of Heidelberg, Heidelberg, Germany
[15] Clinic For Children And Adolescents, Hospital St. Georg, Leipzig, Germany
[16] Department Of Pediatric Nephrology And Hypertension, Faculty Of Medicine, Jagiellonian University Medical College, Krakow, Poland

Objectives: Autosomal recessive polycystic kidney disease (ARPKD) is a rare genetic disorder ciliopathy with an incidence estimated to 1:20,000. ARPKD is mainly caused by variants in the *PKHD1* gene and rarely in *DZIP1L*. The disease presents with a phenotypical spectrum with high variation in severity that could lead to an early impact on child health. While there are defined criteria for the clinical diagnosis, the clinical variability can make it challenging to differentiate ARPKD from other genetic causes of cystic kidney diseases. In some of these cases of phenocopies genetic testing can lead to a change in the initial clinical diagnosis.

Methods: Patients secondarily excluded from ARegPKD due to genetic diagnosis of another cystic kidney disease were analyzed. Variants found in phenocopy genes were evaluated using *in silico* pathogenicity prediction tools and classified using ACMG criteria. Initial and follow-up clinical presentation data in this cohort was then compared to ARPKD cases with detection of relevant *PKHD1* variants (at least one variant ACMG class ≥3).

Results: At the time of analysis 665 patients were included in the ARegPKD registry. 284 patients had relevant *PKHD1* variants. We identified 32 individuals with variants in other cystic kidney disease-causing genes (18 *PKD1,* 5 *TMEM67,* 5 *HNF1B,* 2 *NPHP3,* 1 *PKD2,* 1 *LRP5*) but without detection of *PKHD1* or

DZIP1L variants. All variants identified in these genes showed *in silico* predicted disease-causing pathogenicity and compatible results in variant frequencies in the genome aggregation database. The majority of these patients presented prenatally or postnatally in the first year of life with 34% and 41%, respectively. Extrarenal manifestations were seen in 9 patients. 8 patients started kidney replacement therapy with a median age at onset of 4.4 years.
Conclusions: Utilizing our international cohort, we extend the data on ARPKD phenocopies and present their genetic and clinical data.

EP-122 AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE IN TUNISIAN CHILDREN: A SINGLE CENTRE EXPERIENCE

Ben Guedria Mouna, *Selsabil Nouir*, *Mabrouk Sameh*, *Mansour Kais*, *Majdoub Fadwa*, *Ajmi Houda*, *Tfifha Meniar*, *Chemli Jaleleddine*, *Zouari Noura*, *Abroug Saoussen*
Pediatric Department, Sahloul Hospital, Tunisia

Introduction: Autosomal dominant polycystic kidney disease (ADPKD) is a systemic disease involving cysts in the kidneys and abdominal organs as well as abnormalities in the heart and vasculature. Although it typically presents in adults, ADPKD has been diagnosed in foetuses, infants, children, and adolescents. The aim of this study was to describe the clinical features and outcomes of ADPKD among Tunisian children.
Material and methods: Cross sectional observational study of the paediatric cases of ADPKD managed in the department of paediatrics in the University hospital (Sousse – Tunisia)
Results: 21 patients, predominantly girls (57%) were included. Family history of cystic disease was positive in 80% of cases. Mean age of diagnosis was 11 years, however one patient was diagnosed at neonatal period. Half of our patient were symptomatic when the disease was diagnosed. Main symptoms were abdominal pain, fever and gross hematuria. Hypertensionwas found in 16% of patients. Renal function was normal at the diagnosis in 20 cases. One patient had End stage renal disease at presentation. Renal ultrasound showed renal enlargement with multiple cysts. For the extra renal manifestations, liver cysts were found in 3 patients and pancreatic cysts in one patient. The main complication was Hypertension in 20% of patients, and Urinary tract infection occurred in 3 cases. Management was based on ACE inhibitors.14% 3 patients progressed to end-stage renal disease requiring haemodialysis in 2 cases followed by renal transplantation, and one was on peritoneal dialysis.
Conclusions: ADPKD in children is relatively rare in children, often asymptomatic, however symptoms may be observed during childhood, mainly hypertension and hematuria. Besides End stage renal disease can occur during childhood

EP-123 KIDNEY DIMENSION IS THE MOST IMPORTANT PARAMETER ASSOCIATED WITH DETERIORATION IN KIDNEY FUNCTION IN CHILDREN WITH AUTOSOMAL RECESSIVE POLYCYSTIC KIDNEY DISEASE

Neslihan Cicek [1], *Ibrahim Gokce* [1], *Ceren Alavanda* [2], *Sercin Guven* [1], *Mehtap Sak* [1], *Ozde Nisa Turkkan* [1], *Ece Demirci Bodur* [1], *Serim Polat* [1], *Pinar Ata* [2], *Nurdan Yildiz* [1], *Harika Alpay* [1]
[1] *Marmara University School Of Medicine, Department Of Pediatric Nephrology*
[2] *Marmara University School Of Medicine, Department Of Genetics*

Introduction: In this study, we described clinical, biochemical, imaging and genetic findings and investigated parameters that may influence renal prognosis in pediatric patients with autosomal recessive polycystic kidney disease(ARPKD).
Material and methods: Patients who were diagnosed before birth or during the first month were classified as perinatal presenters(9 patients), diagnosed later classified as non-perinatal presenters (10 patients). We also divided patients according to estimated glomerular filtration rates(eGFR) at last visit as patients with eGFR≥30 and eGFR<30 ml/min/1.73m^2. We analyzed clinical, demoghraphic findings, kidney dimensions adjusted to height, urinary protein excretions, liver functions, office blood pressures at diagnosis and at last visit, genetic analysis and genetic types. The correlation between time of diagnosis (perinatally or non-perinatally, eGFR at last visit(eGFR≥30 or eGFR<30 ml/min/1.73m^2) and other parameters were evaluated.
Results: Seven patients(36.84%) were diagnosed antenatally, the mean follow up time was 7.7±5.21 years. Among six(31.6%) patients who reached stage-5-CKD, two had a renal transplantation. Eleven patients(57.9%) had hypertension and five(26.3%) had proteinuria. Liver disease was diagnosed in 13 patients(68.4%). Genetic tests of 15 patients were evaluated. Eleven patients had homozygous mutation in PKHD1 gene, 1 truncating and 10 non-truncating. Four patients had compound heterozygous mutation in PKHD1 gene. When two groups were compared, kidney dimension at diagnosis was higher in perinatal presenters(p=0.01) and although not statistically significant, eGFR at diagnosis was lower in perinatal presenters (p=0.065). Liver and spleen involvements were higher in non-perinatal presenters(p=0.04, p=0.033). When patients are compared according to eGFR at last visit, kidney dimension at diagnosis was higher in patients with eGFR<30 ml/min/1.73m^2(p=0.045).
Conclusions: In patient with perinatal presenters kidney dimension at diagnosis is higher. Kidney dimension is the parameter most associated with low eGFR at last visit. Non-perinatal presenters, on contrary, had more liver and spleen involvements.

EP-124 CLINICAL AND MUTATIONAL SPECTRUM OF CHILDREN WITH AUTOSOMAL RECESSIVE AND AUTOSOMAL DOMINANT POLYCYSTIC KIDNEY DISEASE

Ozum Tutal [1], *Bora Gulhan* [2], *Emine Atayar* [3], *Selçuk Yüksel* [4], *Birsin Özçakar* [5], *Oğuz Söylemezoğlu* [6], *Seha Saygili* [7], *Mihriban İnözü* [8], *Esra Baskin* [9], *Ali Düzova* [2], *Mutlu Hayran* [10], *Rezan Topaloglu* [2], *Fatih Ozaltin* [2]
[1] *Hacettepe University School Of Medicine, Department Of Pediatrics, Ankara, Turkey*
[2] *Hacettepe University School Of Medicine, Department Of Pediatrics, Division Of Pediatric Nephrology, Ankara, Turkey*
[3] *Hacettepe University School Of Medicine, Nephrogenetics Laboratory, Ankara, Turkey*
[4] *Pamukkale University School Of Medicine, Department Of Pediatrics, Division Of Pediatric Nephrology, Denizli, Turkey*
[5] *Ankara University School Of Medicine, Department Of Pediatrics, Division Of Pediatric Nephrology, Ankara, Turkey*
[6] *Gazi University School Of Medicine, Department Of Pediatrics, Division Of Pediatric Nephrology, Ankara, Turkey*
[7] *Istanbul University CerrahpaŞa School Of Medicine, Department Of Pediatrics, Division Of Pediatric Nephrology, Istanbul, Turkey*
[8] *Ankara City Hospital, Department Of Pediatric Nephrology, Ankara, Turkey*
[9] *Baskent University School Of Medicine, Department Of Pediatrics, Division Of Pediatric Nephrology, Ankara, Turkey*
[10] *Hacettepe University School Of Medicine, Department Of Preventive Oncology, Ankara, Turkey*

Introduction: In this study, we aimed to investigate clinical and genetic features of autosomal recessive polycystic kidney disease (ARPKD) and autosomal dominant polycystic kidney disease (ADPKD) in a group of Turkish patients.
Material and methods: A total of 69 children with genetically confirmed ARPKD (10 females, 11 males) or ADPKD (28 females, 20 males) from