CHARACTERISTICS, OUTCOME AND FOLLOW UP OF CHILDREN WITH RENAL CYSTS: RESULTS FROM A SINGLE TERTIARY REFERRAL CENTRE IN CROATIA

Sara Grlić[1], Viktorija Gregurović[1], Mislav Martinić[2], Ivan Jakopčić[2], Hana Matković[2], Maja Ban[2],Maša Davidović[2], Ivanka Kos[2], Kristina Vrljičak[2], Lovro Lamot[1,2]

[1]University of Zagreb School of Medicine, Department of Pediatrics;

[2]Division of Nephrology, Dialysis and Transplantation, Department of Pediatrics, University Hospital Center Zagreb, Croatia;

AIMS

To provide a concise overview and analyze the real-life data from a **single center cohort followed over a 12-year period**, offering insights into the presenting symptoms, optimal treatment approaches, and **potential long-term prognosis**.

METHODS

A retrospective study of **112** patients examined from **July 2011 to March 2023** for cystic kidney disease (CyKD) in the Referral Center for Pediatric Nephrology of the Republic of Croatia. Data was collected from computerized medical records. Codes from the 10th revision of the International Classification of Diseases (ICD-10) designating CyKD (Q61.0-Q61.9) were used to search the database. Researchers collected comprehensive patient data, including **medical history**, **physical exams**, **ultrasound imaging data**, **creatinine** and **BUN levels** and **comorbidities**.

RESULTS

Out of 112 patients (54 female), **54 were diagnosed antenatally**, **14 in the first 28 days and 44 later in life** (median age 6.0 (3.8-12.3) years). During the follow-up period 59 patients remained asymptomatic. The most common presenting symptoms were **abdominal distension (21%) and abdominal pain (15%).** Total of **17 patients** progressed to chronic kidney disease **(CKD)**, with **13** having end stage renal disease **(ESRD)**, while coordinated **transition** to adult care occurred **in 8**. The median time from the initial detection of cysts on US to the onset of chronic kidney disease (CKD) was 59.0 (7.0-124.0) months, whereas onset of end-stage renal disease (ESRD) was in 127.0 (33.0-141.0) months. The **genetic testing** was performed on 20 patients in total (17.9%) and **was positive for a disease-causing mutation in 15 (75.0% of tested patients)**. The most frequently encountered **extrarenal manifestations** of CyKD included **hypertension (21%), ophthalmological findings (13%) and inguinal hernia (11%).**

DIAGNOSIS	ADPKD	ARPKD	BBS	IRC	JS	MCDK	NPHC	T13	TSC	Total
Patients (N, %)	**19** 17.0%	**16** 14.3%	**2** 1.8%	**15** 13.4%	**2** 1.8%	**52** 46.4%	**2** 1.8%	**1** 0.9%	**3** 2.7%	**112** 100.0%
Positive family history (N, %)	**17** 89.5%	**3** 18.8%	**1** 50.0%	**0**	**0** 0.0%	**5** 9.6%	**1** 50.0%	**0**	**2** 66.7%	**29** 25.9%
Positive genetics (N, % of tested)	**4** 100.0%	**2** 50.0%	**2** 100.0%	**0**	**2** 100.0%	**1** 33.3%	**2** 100.0%	**1** 100.0%	**1** 100.0%	**15** 75.0%
VUR detected (N,%)	**1** 25.0%	**1** 33.3%	**0**	**2** 50.0%	**0**	**13** 33.3%	**0**	**0**	**0**	**17** 31.0%
≥2 UTIs (N,%)	**4** 21.1%	**3** 18.8%	**1** 50.0%	**2** 13.3%	**1** 50.0%	**15** 28.8%	**0**	**0**	**2** 66.7%	**28** 25.0%
Uroprophylaxis (N, %)	**2** 10.5%	**3** 18.8%	**1** 50.0%	**2** 13.3%	**1** 50.0%	**20** 38.5%	**0**	**0**	**2** 66.7%	**31** 27.7%
ACEi (N, %)	**2** 10.5%	**7** 43.8%	**2** 100.0%	**0**	**1** 50.0%	**5** 9.6%	**0**	**0**	**3** 100.0 %	**20** 17.9%
CKD (N,%)	0	**7** 43.8%	**1** 50.0%	0	**2** 100.0%	**4** 7.7%	**2** 100.0%	0	**1** 33.3%	**17** 15.2%
ESRD (N,%)	0	**4** 25.0%	**1** 50.0%	0	**2** 100.0%	**4** 7.7%	**1** 50.0%	0	**1** 33.3%	**13** 11.6%

CONCLUSION

The greatest unmet need in CyKD is to **accurately predict, possibly prevent and safely delay progression into ESRD**. Detailed characterization of the disease and comparison with well described cohorts might provide an important clue for the creation of comprehensive follow-up and management plan. Often inconclusive, the clinical characteristics, laboratory investigations and outcome of patients in our cohort represents a real-life data which could serve as a signpost when seeing a new renal cyst in clinical settings.

CONTACT
lovro.lamot@kbc-zagreb.hr