Research Article

Clinical Characteristics of Congenital Anomalies of the Kidney and Urinary Tract, Minia District

Asmaa T. Abdelgawad*, Eman R. Ghazawy*, Eman S. Mohammed* and Eman Mahfouz*.
* Department of Public Health and Preventive Medicine, Faculty of Medicine, Minia University

Abstract
Background: Congenital anomalies of kidney and urinary tract (CAKUT) are characterized by structural and functional abnormalities of kidney, collecting system, bladder and urethra. CAKUT identified in 20% to 50% of all fetal congenital anomalies and represent the cause of 40–50% of pediatric end-stage renal disease worldwide. **Aim of the study:** This study was performed to report the patterns and clinical presentation of infants and children with CAKUT. **Methods:** Patients with diagnosed with CAKUT, presenting to Minia University Hospitals over one year duration were investigated and categorized based on underlying renal structural/functional malformation and associated extra-renal anomalies. **Results:** this study revealed that the most common type of CAKUT detected was uretro-pelvic junction obstruction (UPJO). The commonest pattern of presentation of CAKUT was fever, and urological manifestation was only in quarter of cases, genital anomalies were the most common associated malformation and the most common complication was recurrent urinary tract infection. **Conclusions:** CAKUT is a clinically heterogeneous group of diseases with diverse clinical phenotypes. More efforts should be aimed at improving antenatal detection as well as classification with comprehensive reference to the clinical features of the diseases.
Key Words: Congenital anomalies of the kidney and urinary tract (CAKUT), Uretro-pelvic junction obstruction (UPJO)

Introduction

Congenital anomalies of kidney and urinary tract (CAKUT) are characterized by structural and functional abnormalities of kidney, collecting system, blader and urethra[1]. CAKUT categorized into two main groups which differ in presentation and management: Obstructive phenotype including posterior urethral valves (PUV), complete renal duplex, ureterocele, megaureter and pelviureteric junction obstruction (PUJO) and non-obstructive phenotype as primary vesicoureteric reflux (VUR), multicystic dysplastic kidney (MCDK), renal agenesis (RA) and renal hypo-dysplasia[2] CAKUT are the most frequent form of malformation at birth, identified in 20% to 50% of all fetal congenital anomalies and affecting 3–7 out of 1000 live births in some populations[3]. CAKUT represent the cause of 40–50% of pediatric and 7% of adult end-stage renal disease (ESRD) worldwide[4].

In the Arab world, the incidence and prevalence of preventable renal diseases are more common than other regions which are often related to the high degree of consanguinity. Reflux nephropathy accounts for 53% of end stage renal diseases (ESRD) in Libyan children, hydronephrosis, hypo-plastic kidneys, posterior urethral valve have all been found to be major causes of kidney disease and renal failure in the Arab world. Congenital kidney diseases occur in 3.3 per 1000 births, 80% of which are due to urinary tract anomalies leading to congenital hydronephrosis[5]. In Egypt, CAKUT comprised 46% of the underlying etiology in Egyptian chronic kidney disease (CKD)in pediatric patients[6].

Methodology:

The study was conducted on 150 children aged ≤ 5 years old with established diagnosis of congenital anomalies of kidney and urinary tract (CAKUT). All children ≤ 5 years of age who were visited pediatric and urology department in Minia University Hospitals during the study period were recruited. Children' parents asked to participate in the study during hospital visits. The questionnaire includes: Child medical history: age, gender,

order, birth weight, mode of delivery, history of incubation, antenatal diagnosis, mode of diagnosis, type of anomalies, pattern of presentation of CAKUT, associated anomalies, disease duration, received therapy and complication.

Statistical analysis:

Statistical analysis Data management and analysis were performed using Statistical Package for Social Sciences (SPSS); Numerical data were summarized using means and standard deviations or medians and ranges, Categorical data were summarized as percentages. Graphics were done by Excel Microsoft office 2010.

Results

The characteristics of the participants were shown in (Table 1**).** 150 children diagnosed with CAKUT were included in the present study, 102 (68%) were males and 48 (32%) were females, with a median age of 10.5 months. UPJO ranked the first among different CAKUT types up to 30% of cases followed by non-obstructive cystic renal disease in 19.3% (figure 1). About three-quarters (73%) of CAKUT cases were obstructive in nature and 27% of them were non-obstructive type (figure 2). About 21.3% of cases were diagnosed during antenatal follow up and majority of cases diagnosed postnatal 78.7% (figure 3). 16% of cases diagnosed postnatally discovered accidentally without specific symptoms related to CAKUT, while the most common symptom was fever 32.7% followed by abdominal pain 17.3%. Urological related symptoms (retention and micturition difficulties, urosepsis and hematuria) represented nearly a quarter of presentations (25.9%). Figure 6 showed that 13.4% of CAKUT cases had associated malformations. Genital anomalies were the most common form of associated malformation (35%) followed by neurological anomalies (20%), then congenital heart diseases (15%).

Table (1): Frequency distribution of the studied children according to their characteristics, Minia governorate, May 2019 to December 2020

Variables	CAKUT cases N=150	Controls N=150	P-value	X^2(DF)
Age of child (months)				
Range	1-60	1-60	0.1	1.8 (298)
Median (IQR) #	10.5 (27)	12 (36)		
Sex				
Male	102 (68%)	95 (63.3%)	0.3	0.7 (1)
Female	48 (32%)	55 (36.7%)		
Child order				
1st child	47 (31.3%)	33 (22%)	0.07	3.3 (1)
2nd child or more	103 (68.7%)	117 (78%)		
Mode of delivery				
Normal vaginal	85 (56.7%	105 (70%)	0.01*	5.7 (1)
Ca**e**sarean section (CS)	65 (43.3%)	45 (30%)		
Maturity				1.8 (1)
Preterm	18(12%)	11 (7.3%)	0.1	
Full-term	132 (88%)	139 (92.7%)		

***** Statistically significant # Mann–Whitney U test.

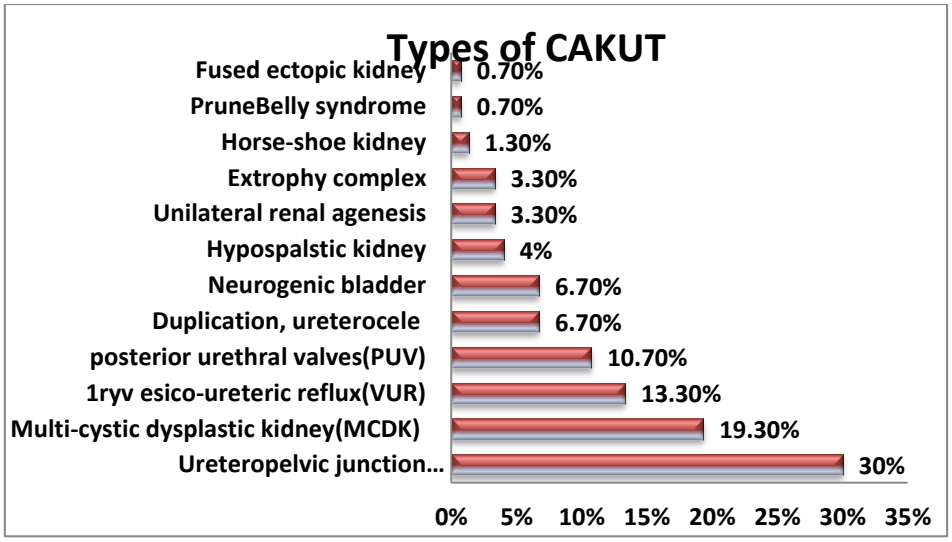

Figure (1): Frequency distribution of congenital anomalies of kidney and urinary tract (CAKUT)

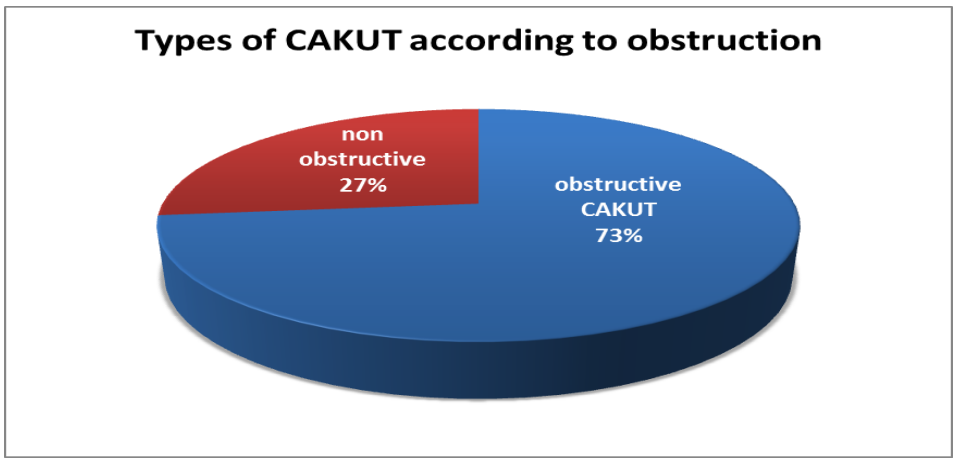

Figure (2): Types of congenital anomalies of kidney and urinary tract (CAKUT) according to obstruction, Minia governorate, May 2019 to December 2020

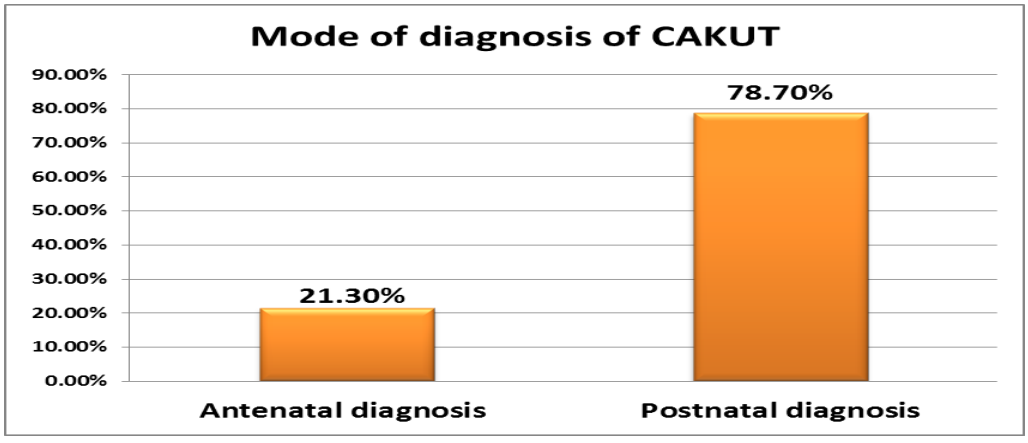

Figure (3): Mode of diagnosis of congenital anomalies of kidney and urinary tract (CAKUT) Minia governorate, May 2019 to December 2020

Table (2): Frequency distribution of most common pattern of post natal presentation of congenital anomalies of the kidneys and urinary tract (CAKUT), Minia governorate, May 2019 to December 2020

Pattern of presentation of CAKUT (n=118)	Frequency	Percent
Asymptomatic accidentally discovered by Ultrasound (US)	24	16%
Symptomatic[a]		
Fever	49	32.7%
Abdominal pain	26	17.3%
Retention and micturition difficulties	20	13.3%
Failure to thrive	19	12.7%
Urosepsis	14	9.3%
Hematuria	5	3.3%
Defect in abdominal wall	5	3.3%
Abdominal swelling	4	2.7%
[a] Numbers do not add to 100% as respondents might have more than 1 presentation.		

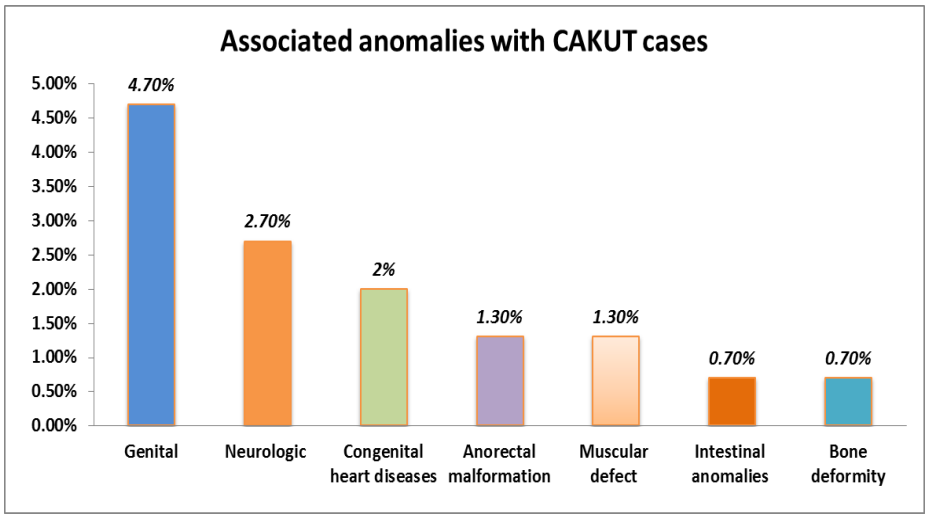

Figure (6): Frequency distribution of associated anomalies with congenital anomalies of kidney and urinary tract (CAKUT) in studied cases, Minia governorate, May 2019 to December 2020

Discussion

Congenital anomalies of the kidney and the urinary tract (CAKUT) one of the most common forms of birth defects and the leading cause of end stage renal disease in children less than 5 years of age[7].

The present study showed that among infants with CAKUT, 68% were males and 32% were females. with male to female ratio 2.1:1 these results were similar to what reported in Egyptian study described the pattern and clinical presentation of CAKUT among 107 CAKUT children with an approximate male to female ratio 2.3:1 ,also the same finding reported in Turkish children by Bulum et al., (2013) with male to female ratio 1.9:1. This male predominance may be due to occurrence of some types of CAKUT exclusively in males e.g. PUV[2][8]

Obstructive form of CAKUT detected in 73% of cases in this study where UPJO ranked the first among different CAKUT types up to 30% of cases the same finding reported by Aksu et al., (2005) and Radhakrishna et al., (2019) where PUJO was the most frequent type of CAKUT in both studies [9] [10]

In contrast, Soliman et al., (2015) reported that

PUV was the commonest abnormality (36.4%) followed by VUR (19.6%) then PUJO (18.7%), but this difference may be attributed to different age group included in his study[2].
Unfortunately in this study, less than quarter of cases diagnosed early (21.3% antenatal and 34% early postnatal) which lower than Soliman et al., (2015) study (36.6% antenatal diagnosis) and much lower than what reported by Radhakrishna et al., (2019) in India (65% antenatal an 47% early postnatal)[2] [10].

In the current study it was found that 16% of children were asymptomatic and nearly 25% of them presented by urinary tract symptoms. In line with these findings, previous study conducted in India found that 32% of cases were asymptomatic and 22% presented with UTI[10]. Soliman et al., (2015) reported urinary manifestation only in 37% and about half of cases presented with nonspecific abdominal symptoms (e.g. abdominal mass, distention, pain, and diarrhea)[2].

In this study, there were 13.4% of CAKUT patients had associated non-renal anomalies, the most common being genital anomalies (35%). Radhakrishna et al., (2019) found associated extra renal anomalies in 13.6% of cases, Bondagji et al., (2014) in 26.2% of cases Ahmadzadeh et al., (2009) in 22.4% of cases and Soliman et al., (2015) in 31.8% of cases[10,11,12,2]

Reference

1. Song, R. and I.V. Yosypiv, Genetics of congenital anomalies of the kidney and urinary tract. Pediatric Nephrology, 2011. 26(3): p. 353-364.
2. Soliman, N.A., et al., Pattern of clinical presentation of congenital anomalies of the kidney and urinary tract among infants and children.Nephrology,2015.20(6):p.413-418
3. Tain, Y.-L., et al., Incidence and risks of congenital anomalies of kidney and urinary tract in newborns: A population-based case–control study in Taiwan. Medicine, 2016. 95(5).
4. Capone, V.P., et al., Genetics of congenital anomalies of the kidney and urinary tract: the current state of play. International journal of molecular sciences, 2017. 18(4): p. 796.
5. Shaheen, F.A. and A.A. Al-Khader, Preventive strategies of renal failure in the Arab world. Kidney International, 2005. 68: p. S37-S40.
6. Safouh, H., et al., Causes of chronic kidney disease in Egyptian children. Saudi Journal of Kidney Diseases and Transplantation, 2015. 26(4): p. 806.
7. Rodriguez, M.M., Congenital anomalies of the kidney and the urinary tract (CAKUT). Fetal and Pediatric Pathology, 2014. 33(5-6): p. 293-320.
8. Bulum, B., et al., High frequency of kidney and urinary tract anomalies in asymptomatic first-degree relatives of patients with CAKUT. Pediatric Nephrology, 2013. 28(11): p. 2143-2147.
9. Aksu, N., et al., Postnatal management of infants with antenatally detected hydronephrosis. Pediatric Nephrology, 2005. 20(9): p. 1253-1259.
10. Radhakrishna, V., et al., Clinico-biochemical profile of children with congenital anomalies of the kidney and urinary tract: a cross-sectional study. Kidney Diseases, 2019. 5(1): p. 51-57.
11. Bondagji, N.S., Antenatal diagnosis, prevalence and outcome of congenital anomalies of the kidney and urinary tract in Saudi Arabia .Urology annals, 2014. 6(1): p.36.
12. Ahmadzadeh, A., M. Tahmasebi, and M.M. Gharibvand, Causes and outcome of prenatally diagnosed hydronephrosis. Saudi Journal of Kidney Diseases and Transplantation, 2009. 20(2): p. 246.