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CLINICAL SPECTRUM OF CONGENITAL ANOMALIES OF KIDNEY AND URINARY TRACT IN CHILDREN

Paediatrics

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ABSTRACT

Introduction : Although some forms of CAKUT are a part of a syndrome associated with extra-renal manifestations or have positive family history, most cases of renal system anomalies are sporadic and isolated to the urinary tract. The majority of renal malformations are diagnosed antenatally, largely because of the widespread use and sensitivity of fetal ultrasonography **Aim and objectives:** to evaluate the clinical spectrum and patterns of clinical presentation in congenital anomalies of kidney and urinary tract. **Material and methods:** This is descriptive study conducted at tertiary medical centre in Maharashtra during March 2022 to March 2023 on 50 study subjects fulfilling the following inclusion and exclusion criteria. Purposive sampling methods were used for data collection. This descriptive study were conducted at our tertiary care hospital after obtaining approval from the Institute Ethics Committee. Informed consent was taken from the mothers/caregivers. All infants and children up to 12 years of age who were diagnosed to have CAKUT on imaging studies done either during antenatal period or after birth were enrolled in the study. **Result:** In our study we found that on antenatal USG scan VUR 5 (10%) was most common followed by bilateral hydronephrosis 4 (8%), left sided pelvic ectasia 3(6%) and right sided pelvic dilatation3 (6%). Out of 50 patients we found 23 (46%) patients were normal USG findings. clinical features such as Fever which is found in 11(22%) children and 39(78%) children having no fever. Out of 50 patients 37 (74%) had shown no grown on urine culture where as 10(10%) shown growth of E.Coli and 3(6%) patients show the growth of Protease. **Conclusions:** In conclusion, the commonest renal anomaly was PUJ, followed by PUV, then VUR. Most common presentation was urinary symptoms. The most prominent complication was UTI.

KEYWORDS

PUJ, Congenital kidney disease, UTI, VUR

Introduction

Congenital anomalies of the kidney and urinary tract [CAKUT] comprise a wide range of structural malformations that result from defects in the morphogenesis of the kidney and/or the urinary tract.[1]This wide range of renal system structural and functional malformations that occur at the level of the kidney, collecting system, bladder, or urethra account for about 40-50% of children with chronic kidney disease (CKD), and are the most common cause of end-stage renal disease (ESRD) in children.[2,3] That being said, it is important to diagnose these anomalies early enough and initiate therapy to minimize renal damage, prevent or delay the onset of ESRD, and provide supportive care to avoid complications of ESRD.[4,5]

Although some forms of CAKUT are a part of a syndrome associated with extra-renal manifestations or have positive family history, most cases of renal system anomalies are sporadic and isolated to the urinary tract.[6] The majority of renal malformations are diagnosed antenatally, largely because of the widespread use and sensitivity of fetal ultrasonography.[7]To date, 23 monogenic CAKUT causing genes have been identified in patients with isolated, familial, or syndromic CAKUT, each gene representing a monogenic recessive or dominant cause of CAKUT. The malformation phenotypes vary from normally appearing kidneys with intact kidney function (i.e. incomplete penetrance) to severe hypodysplasia and ESRD.[8]

In literature there is little description about warning signs and symptoms of urinary system impairment. The clinical presentation of CAKUT is variable. Some patients are diagnosed ante-natally during anomaly scan while others remainasymptomatic till adolescence. Similarly there is acute versus chronic presentation. Common upper urinary tract symptoms include polyuria, nocturia, short stature/failure to thrive, systemic hypertension and urinary tract infection. Lower urinary tract symptoms such as urgency, frequency, dysuria, straining, poor urinary stream, urinary retention and incontinence indicate a need for the evaluation of lower urinary tract. [9]

We conducted this study to evaluate the clinical spectrum and patterns of clinical presentation in congenital anomalies of kidney and urinary tract.

Material and methods

This is descriptive study conducted at tertiary medical centre in Maharashtra during March 2022 to March 2023 on 50 study subjects fulfilling the following inclusion and exclusion criteria. Purposive sampling methods were used for data collection.

Inclusion criteria:

- All infants who are diagnosed to have CAKUT on antenatal scan.
- All children up to 12 years age detected to have CAKUT on imaging studies done either incidentally or while being evaluated for illness suggestive of urinary tract involvement.

Exclusion criteria:

- Infants who present with illness suggestive of involvement of kidney and urinary tract but without evidence of CAKUT on imaging studies.
- Those found to have structural anomalies of kidney and urinary tract deemed to be of acquired origin (example P.U.J. obstruction due to calculi)

Detailed procedure of study conduct:

This descriptive study were conducted at our tertiary care hospital after obtaining approval from the Institute Ethics Committee. Informed consent was taken from the mothers/caregivers. All infants and children up to 12 years of age who were diagnosed to have CAKUT on imaging studies done either during antenatal period or after birth were enrolled in the study. A relevant data was collected and entered in pre structured proforma. Purpose of the study was explained and informed consent was taken. A pre structured proforma was used to record the relevant information from individual subject selected for the study. Thorough history, clinical examination, relevant biochemical tests and imaging studies were done based on individual cases.

STATISTICALANALYSIS:

Descriptive statistical analysis was carried out in the present study.Data was entered in Microsoft Excel and analysed in EpiData analysis software (version 2.2.2.178). Categorical data were summarized as percentages.

Result

In our study out 50 patients we found 32 (64%) were males and 18 (36%) were females. We also found that male to female ratio is higher. In our study we found that 20 (40%) patients between 1-24 months was most common age group. Followed by 16(32%) range 25 – 48 months. In our study we found that 30 (60%) age between 21-25 years was most common age group. Followed by 15 (30%) age between 26-30 years. During our study we found that 8 (16%) patients were having family history of CAKUT and 42(84%) were without any family history.

TABLE – 1: PERCENTAGE OF CASES DETECTED ANTENATALLY

ANTENATAL USG	No. Of cases	Percentage
Normal	23	46
BILATERAL HYDRONEPHROSIS, POSTERIOR URETHRAL VALVE	5	10
BILATERAL HYDROPNEPHROSIS	1	2
BLADDER EXOSTROPHY	1	2
HORSESHOE SHAPED KIDNEY	1	2
LEFT ECTOPIC KIDNEY	1	2
LEFT HYDRONEPHROSIS	2	4
LEFT KIDNEY NOT VISUALISED	1	2
LEFT POLYCYSTIC KIDNEY	1	2
LEFT SIDED PELVIC ECTASIA	3	6
POSTERIOR URETHRAL VALVE	1	2
RIGHT HYDRONEPHROSIS	1	2
RIGHT SIDED PELVIC DILATATION	3	6
RIGHT SIDED PUJ OBSTRUCTION	1	2
VUR	5	10

In our study we found that on antenatal USG scan VUR 5 (10%) was most common followed by bilateral hydronephrosis 4 (8%), left sided pelvic ectasia 3(6%) and right sided pelvic dilatation3 (6%). Out of 50 patients we found 23 (46%) patients were normal USG findings.

TABLE – 2 PERCENTAGE OF CASES DETECTED WITH ASSOCIATED ANOMALY

ASSOCIATED ANOMALIES	No. Of cases	Percentage
None	48	96
OPHALOCELE	1	2
VACTERAL ANOMALIES	1	2

Associated anomalies were found only in 4% cases out of that 2% were ophalocele and 2% were vacteral anomalies and 96% patients had no any other associated congenital anomalies in our study.

Fig 1: TIME OF DETECTION OF ANOMALY

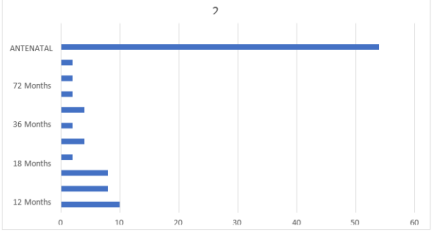

TABLE -3: PERCENTAGE OF CASES DETECTED WITH FEVER

FEVER	No. Of cases	Percentage
NO	39	78
FEVER	11	22
URINARY SYMPTOMS	15	30
OTHERS	12	24

During our study we found following clinical features such as Fever which is found in 11(22%) children and 39(78%) children having no fever. Out of 50 patients 37 (74%) had shown no grown on urine culture where as 10(10%) shown growth of E.Coli and 3(6%) patients show the growth of Protease.

TABLE -4 : PERCENTAGE OF CASES DETECTED ACCORDING TO CREATININE LEVEL

CREATININE	No. Of cases	Percentage
Deranged	12	24
Normal	38	76

Out of 50 patients blood urea creatinine level was found to be deranged in 11(22%) and 39(78%) patients was found within normal limits. Out of 50 patients creatinine level was found to be deranged in 12(24%) and 38(76%) patients was found within normal limits. We found that most common CAKUT was PUJO 16(32%). Out of that Left PUJO were 9(18%) and Right PUJO were 7(14%). Followed by PUV 11(22%), VUR 10(20%), ectopic kidney 4(8%), Renal Agenesis 3(6%), MCDK 3(6%), followed by other diagnosis such as bladder extropy, cross fused kidney, horse shoe shaped kidney.

Discussion

In our study we included total 50 consecutive children, upto 12 years of age (as 12 years is the upper age limit for registration in pediatric OPD and for admission to pediatric ward), with any one or more CAKUT with or without associated anomalies in other systems were enrolled in this descriptive study. The study was conducted for 18 months after written obtaining clearance from the institutional ethical committee and informed consent was taken from the parents/caregivers of all children included in the study.

GENDER

In our study out 50 patients we found 32 (64%) were males and 18 (36%) were females. We also found that male to female ratio is higher. The result is accordance to study conducted by Radhakrishna V, et. al. were they found male ratio is higher then a female ratio.[9] Ali SH et al also found the that 97 males (60.6%) and 63 females (39.4%) with male:female ratio 1.54:1.[10]

AGE GROUP

In our study the age distribution of the study population we found patients with 1-24 months 20 (40%) mostly diagnosed age group at presentation as the cases were detected during antenatal scan and presented to outpatient department for follow up. Radhakrishna et al. observed that Thirty-eight (47%) children presented in infancy, while 23 (28%) patients presented after 1 year of age.[9] Ali et al observed that the most frequent age group at time of presentation was 1-5 years age group including 84 patients (52.5%).[10]

ANTENATAL USG

In our study we found that on antenatal USG scan bilateral hydronephrosis 10 (20%), was most common followed by left sided pelvic ectasia 3(6%) and right sided pelvic dilatation3 (6%). Out of 50 patients we found 23 (46%) patients were normal USG findings. Study conducted by Nabeel S. Bondagji found that Fetal hydronephrosis is a common finding on antenatal ultrasound examination.[11]Though a significant number of children were detected antenatally, approximately 46% of the CAKUTs had not been antenatally identified, and a majority of this subgroup were identified after UTI.[12]

CLINICAL PRESENTATION

During our study we found following clinical features such as Fever which is found in 11(22%) children and 39(78%) children having no fever, however 15(30%) had Urinary symptoms like dribbling, urosepsis, hematuria, UTI etc. and other symptoms like abdominal pain, failure to thrive, a palpable abdominal mass were found in 12(24%) children. Similar result was found in the study done by Radhakrishna V, et. al. where they found following result. Twenty-six (32%) were asymptomatic, while 18 (22%) patients presented with UTI, 18 (22%) had dribbling, 15 (19%) had abdominal pain, 7 (9%) a palpable abdominal mass, 6 (7%) urosepsis, 3 (4%) hematuria, and 1 (1%) urinary ascites. Twenty-two (27%) patients were found to have hypertension. Twenty-six (32%) were stunted and underweight, while 22 (27%) patients were underweight, and 4 (5%) patients were stunted.[9]

Ali SH et al also found Urinary symptoms were most common presented in 93 patients (58.1%), followed by fever detected in 77 patients (48.1%). UTI was the most frequent complication accounted for (62.5%), followed by renal impairment (34.4%).[10]

CULTURE POSITIVE UTI

Out of 50 patients 37 (74%) had shown no grown on urine culture where as 10(20%) shown growth of E.Coli and 3(6%) patients show the growth of Protease. Children with known urinary tract problems such as CAKUT are very prone to developing recurrent UTI. Previous studies on the prevalence of UTI in CAKUT patients ranged from 25-59%. Ramayani OR et al found that the prevalence of UTI thestudy reached 64%. It may be due to the average age of the CAKUT patients found to be detected at a relatively older age (except for PUV),and the CAKUT abnormality may persist until adolescence.[13] Shahnam Askarpour et al observed that The causative agent was Escherichia coli in 111 (88%), Klebsiella in 8 (6%), Proteus in three, Staphylococcus suprophyticus in two and others in two patients.[14]

BIOCHEMICAL FINDINGS

Out of 50 patients blood urea level was found to be deranged in 11(22%) and 39(78%) patients was found within normal limits. And creatinine level was found to be deranged in 12(24%) and 38(76%) patients was found within normal limits. Shahnam Askarpour et al observed that In 121 (96%) patients, serum creatinine level was normal for the age of patient but in five three girls and two boys was elevated.[14]

OBSTRUCTION AND NON OBSTRUCTION CAKUT

The types of CAKUT in this study were divided into obstruction and non-obstruction types, where the obstruction type was divided into vesicoureteral junction obstruction (VUJO), pelvic-ureteric junction obstruction (PUJO), posterior urethral valve (PUV) and non-obstruction type divided into reflux, neurogenic and hypoplasia. In our study obstructive CAKUT was found in 37 (74%) patients which was more common, among them PUJO 16(32%) was more common obstructive CAKUT. And Non obstructive CAKUT was found in 13 (26%). Soemyarso NA . et. al observed that the most obstruction group was VUJO type with 26 (27.7%), followed by PUJO type with 21 (22.3%) and PUV 1 type (1.1%). CAKUT was the most neurogenic type of non-obstruction group with 28 (29.8%), followed by hypoplasia 13 (13.8%) and reflux 5 (5.3%) the result was similar to our study.[15]While Ali SH et al observed that obstructive CAKUT was found in 33 (22.60%) patients. And Non obstructive CAKUT was found in 113 (77.39%). Which is in contrast to present study.[10]

DIAGNOSIS

We found that most common CAKUT was PUJO 16(32%). Out of that Left PUJO were 9(18%) and Right PUJO were 7(14%). Followed by PUV 11(22%), VUR 10(20%), ectopic kidney 4(8%), Renal Agenesis 3(6%), MCDK 3(6%), followed by other diagnosis such as bladder extropy, cross fused kidney, horse shoe shaped kidney and 2 (4%) had normal findings.

Radhakrishna V, et. al. found the Thirty-two (40%) patients had pelviureteric junction obstruction (PUJO), 26 (32%) patients had posterior urethral valves (PUV), 15 (19%) had vesicoureteric reflux (VUR), 5 (6%) had a multicystic dysplastic kidney (MCDK), 2 (3%) had ureterocele, and 1 (1%) had vesicoureteric junction obstruction (VUJO). Of the 11 female patients of the study group, 6 (55%) had PUJO and 5 (46%) had VUR.[9]

Where another study conducted by Kumar BH et. al. found that VUR was the commonest CAKUT followed by PUJO, MCDK, non-obstructive hydronephrosis and PUV.[12] Also Study conducted by Shakoor, J et. al. found that he most common anomaly was VUR 83 (59.25%) while PUVs being 2nd most common anomaly 48 (34.3%).[16] While Ali SH et al observed that VUR was the commonest abnormality (41.9%), followed by renal agenesis (15.0%), then PUJ (12.5%).[10]

Conclusions

In conclusion, the commonest renal anomaly was PUJ, followed by PUV, then VUR. Most common presentation was urinary symptoms. The most prominent complication was UTI. Improving antenatal diagnosis, collaborative management between the obstetrician, the pediatric nephrologist and the urologist in order to provide optimal care for children with renal anomalies.

REFERENCES

1. Schedl A. Renal abnormalities and their developmental origin. Nature Review Genetics 2007; 8:791–802.
2. Hildebrandt F. Genetic kidney diseases. Lancet 2010; 375:1287-95.
3. Harambat J, van Stralen KJ, Kim JJ and Tizard EJ: Epidemiology of chronic kidney disease in children. Pediatric Nephrology 2012; 27: 363-73.
4. Song R and Yosypiv IV. Genetics of congenital anomalies of the kidney and urinary tract. Pediatric Nephrolgy 2011; 26 (3): 353-64.
5. Bingham C, Bulman MP, Ellard S et al. Mutations in the hepatocyte nuclear factor-1 beta gene are associated with familial hypoplastic glomerulocystic kidney disease. American Journal of Human Genetics 2001; 68:219-24.
6. Seikaly MG, Ho PL, Emmett L, Fine RN and Tejani A.Chronic renal insufficiency in children: the 2001 Annual Report of the NAPRTCS. Pediatric Nephrology 2003; 18(8):796-804.
7. Vanderheyden T, Kumar S and Fisk NM. Fetal renal impairment. Seminars in Neonatology 2003; 8:279-289.
8. Vivante A, Kohl S, Hwang D, Dworschak GC and Hildebrandt F. Single-gene causes of congenital anomalies of the kidney and urinary tract (CAKUT) in humans. Pediatric Nephrology 2014; 29:695-704.
9. Radhakrishna V, Kumaravel S, Priyamvada PS, Hanumanthappa N, Jindal B, Govindarajan K, Naredi BK, Rijhwani A. Clinico-biochemical profile of children with congenital anomalies of the kidney and urinary tract: a cross-sectional study. Kidney Diseases. 2019;5(1):51-7.
10. Ali SH, Kadhim SA, Al-Obaidy QM. Comparative Study between Obstructive and Non-Obstructive Renal Anomalies Among a Group of Iraqi Children. IRAQI JOURNAL OF MEDICAL SCIENCES. 2021;19(2).
11. Bondagji NS. Antenatal diagnosis, prevalence and outcome of congenital anomalies of the kidney and urinary tract in Saudi Arabia. Urology annals. 2014 Jan;6(1):36.
12. Kumar BH, Krishnamurthy S, Chandrasekaran V, Jindal B, Ananthakrishnan R. Clinical spectrum of congenital anomalies of kidney and urinary tract in children. Indian pediatrics. 2019 Jul;56(7):566-70.
13. Ramayani OR, Eyanoer PC, Ritarwan K, Siregar B, Siregar RS. Prevalence of recurrent urinary tract infection in children with congenital anomalies of the kidney and urinary tract (CAKUT). InIOP Conference Series: Earth and Environmental Science 2018 Mar 1 (Vol. 125, No. 1, p. 012112). IOP Publishing.
14. Ahmadzadeh A, Askarpour S. Association of urinary tract abnormalities in children with first urinary tract infection. Pakistan journal of medical sciences. 2007 Jan 1;23(1):88.
15. Soemyarso NA, Prasetyo RV. The Correlation of Congenital Anomalies of The Kidney and Urinary Tract with Renal Function in Children. Indian Journal of Forensic Medicine & Toxicology. 2021 Apr 1;15(2).