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Research Article

Congenital anomalies of the kidney and urinary tract in Nigeria: A thirteen-year single Centre review

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ABSTRACT

Background: Congenital Anomalies of the Kidney and Urinary Tract (CAKUT) encompass a wide spectrum of structural anomalies of the kidney and urinary tract. Evidence from registries in high-income countries suggests that it is a major cause of morbidity in children, with up to 50-60% of Chronic Kidney Disease (CKD) due to CAKUT. The picture is not so clear in medium and low-income countries, mainly due to constraints of skilled personnel, diagnostic abilities, accurate data gathering and poor health-seeking behaviour.

Methods: Information on children managed from birth to 18 years with CAKUT, from January 2010 to end of December 2022 in the renal unit of the Paediatric Department, Jos University Teaching Hospital was extracted for review from the general paediatric renal database.

Results: One hundred and two children had CAKUT, (14.8%) of all children seen in the renal unit during the period. The prevalence of CAKUT among the general paediatric population during the same period was 0.54%. The mean age at diagnosis was 6.31 ± 5.52 years (76 months). There was male preponderance, n=72 (70.59%). The most common kidney anomaly was cystic kidney, n=17 (16.67%), while the most common urologic anomaly was pelviureteric junction obstruction (PUJo) n=26 (25.49%). Recurrent fever and abdominal pain were the commonest symptoms in older children. The antenatal detection rate was very low, only nine (8.82%).

Conclusion: Pelviureteric junction obstruction was the commonest anomaly. Antenatal ultrasound detection rate was very low and recurrent fever and abdominal pain were the most frequent symptoms in older children.

Keywords: Congenital anomalies, Kidney, Urinary tract, Children, Antenatal diagnosis

INTRODUCTION

The spectrum of Congenital Anomalies of the Kidney and Urinary Tract (CAKUT) is wide and includes varying degrees of faulty structural and functional developmental severity (Song et al., 2011). This condition may exist either as an isolated anomaly confined only to the kidney and urinary tract or as part of phenotypic features of a syndrome with other congenital malformations outside the kidney and urinary tract (Godron-Dubrasquet et al., 2017). The term CAKUT, loosely applied comprises anomalies affecting the kidney, collecting system, bladder and the urethra or of location. These include abnormal structure of the kidney and tract such as renal dysplasia and hypoplasia, cystic kidney disease, pelviureteric junction obstruction and megaureter, ureterocoele, vesico-

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ureteric reflux, posterior urethral valve, and of location such as ectopia (Godron-Dubrasquet et al., 2017; Egbe et al., 2015).
In the general paediatric population, prevalence of CAKUT ranges between 3-26.6% while in the prenatal period, it is the commonest malformation constituting 20-30% of all congenital anomalies (Egbe et al., 2015; Barakat et al., 1991). It is also estimated that CAKUT accounts for 25% of all paediatric surgical conditions (Elder et al., 1997).

Clinically, the diagnosis of CAKUT may be suspected when there is oligohydramnios in the mother, which may be due to impaired urine production by the foetus as a result renal of dysplasia or obstruction to urine flow. Another important prenatal indicator of CAKUT will be specific ultrasound features such as hydronephrosis seen in the foetus. Later in life after birth, other symptoms include urinary tract infections, abdominal swelling or recurrent abdominal pain which may be due to distended urinary bladder or enlarged kidneys (Queisser-Luft et al., 2002; Barakat et al., 1991; Yosypiv et al., 2012).

In developed countries, the routine prenatal ultrasound screening means that foetus with CAKUT will likely be identified as early as 18 weeks of gestation, with the most visible and easily noticeable feature being hydronephrosis. After delivery such children are then evaluated and appropriate management algorithms according to institutional or national guidelines are followed. In medium and low-income countries however, diagnosis of CAKUT is made predominantly postnatally and sometimes very late into adolescence. This is majorly, because of very low prenatal screening rate, fewer skilled personnel, limited ability for detection and diagnosis as well as poor health seeking behaviour.

Capacity for appropriate investigations are also limited and this significantly contributes to the poor detection rate of CAKUT in many of these low-income countries like Nigeria. Basic imaging such as ultrasonography and radiological studies such as Micturating Cysto-Urethrogram (MCUG) and Intravenous Urography (IVU) are all that many centers in this setting have access to. Some of these diagnostic tools are limited with regards to usability and sensitivity and may be associated with risks to patients and health workers, particularly high radiation dose. The more sensitive but very expensive tools like nuclear imaging, Magnetic Resonance Imaging (MRI) and Computerized Tomography (CT) scan are found in few centers with costs that are essentially prohibitive.

In young children, Urinary Tract Infection (UTI) should arouse a high index of suspicion to the likelihood of CAKUT. This becomes more significant in the face of non-*Escherichia coli* UTI, recurrent UTI episodes and those with poor or unsatisfactory response to appropriate pharmacological treatment. In many low-income and tropical countries like Nigeria however, there are further compounding factors. These include for example malaria, a very prominent and prevalent cause of febrile illness in children, which means that focus in the first instance is often diverted to this more common condition. This may result in some of these children having late diagnosis and treatment of UTI with resulting risk for renal scars, which may later in life lead to complications such as hypertension, CKD and End Stage Kidney Disease (ESKD).

Though there have been some studies on CAKUT in the paediatric population from Nigeria and other countries in sub-Sahara Africa. Many of these studies are constrained by their small study populations and limited knowledge of paediatric nephrology in many of these reports. Furthermore, many of these studies are retrospective folder reviews. These challenges have created gaps in the understanding of the true burden of this condition emanating from the region. This current study seeks to address some of these limitations.

MATERIALS AND METHODS

This study was commenced in January 2010. The successful completion of a fellowship program of International Paediatric Nephrology Association (IPNA) in 2009 provided the necessary advantage for improved knowledge and skills in the identification and diagnosis. The focus was to identify gaps if any, in the scope of earlier preliminary report on kidney disease in children published previously from this centre.

The study was carried out in the Paediatric Renal Unit of the Jos University Teaching Hospital, Jos, a federal teaching hospital that has a 500-bed capacity. It provides services for most of the states in the North central-region of Nigeria, including the Federal Capital Territory (FCT). The renal unit provides comprehensive paediatric renal care in the form of diagnosis of common childhood kidney diseases, treatment and follow up and renal replacement therapy, mainly dialysis.

The bulk of the study population came from Plateau state and as well as many who were referred from the surrounding states and FCT. This was a cross sectional and descriptive study and covered a period of thirteen years, from January 2010 and the end of December 2022. The study participants were aged from birth to 18 years.

Diagnosis of CAKUT was based on clinical and imaging findings, which included ultrasound, radiology, magnetic resonance imaging, computerized tomography particularly CT urography studies. Very few of the study participants had presumptuous antenatal ultrasound diagnosis and were referred subsequently after birth, while some were referred because of obvious clinical features observed at birth. Only one child was able to do mercapto acetyl triglycine (Tc99-MAG3) nuclear study from another center.

Details of each child seen and managed in the unit was entered into a case record form that was designed to capture relevant information. The information included demography, presenting complaints, relevant physical features, results of investigations done, final diagnosis, and the nature of care given. These data were subsequently entered into an excel spread sheet periodically until the end of the study period. Data on children

diagnosed with CAKUT was extracted from the general renal data for the present study.

Ethical approval was given by the Institutional Research Ethic Committee (IREC) of the Jos University Teaching Hospital.

Analysis was done using EPI info version 7.2.6.0. The results are presented as absolute frequency and percentage for qualitative variables while quantitative variables are presented as median with minimum and maximum values or mean with standard deviation.

RESULTS

One hundred and two (102) children were diagnosed with CAKUT during the study period. This represents a prevalence of 14.8% of the total number of children with kidney and urinary tract diseases and 0.54% of the general paediatric population, which included all new admissions to the ward and those seen in the paediatric outpatient clinic. Of the 102 patients, 72 (70.60%) were males, with a male to female ratio of 2.4:1. Their mean age was 6.31 ± 5.52 years (75.6 ± 66.37 months). The median age at presentation was 60 months, interquartile range (IQR) 12-144 (Table 1).

Table 1. Shows the distribution of the types of CAKUT diagnoses.

Site of CAKUT	Diagnosis	Frequency (n)	Percentage (%)
Kidney	Cystic kidneys	10	9.8
	Hypoplastic kidneys	2	1.96
	Multi cystic dysplastic kidneys	9	8.82
	Ectopic kidneys	9	8.82
	Single kidneys	10	9.8
	Cross-fused kidneys	5	4.9
Urinary tract	Pelvi-ureteric junction obstruction	26	25.49
	Posterior urethral valves	21	20.59
	Congenital megaureter	2	1.96
	Uretrocoele	2	1.96
	Others	6	5.9
Total		102	100
Note: (Others: Duplex system, congenital meatal stenosis, mesoblastic nephroma, VUR, Congenital neuropathic bladder, Non-PUV bladder outlet obstruction)			

Only 9 (8.82%) of the children had prior antenatal detection of anomalies. Of these number, five were males and four females. Postnatal diagnostic confirmation showed that four of these children had posterior urethral valves, two had PUJo while three had Multicystic Dysplastic Kidney Disease (MCKD).

Table 2 shows the age at diagnosis of the different kidney anomalies and their gender distributions. Children whose anomalies were detected antenatally are included in the group under twelve months.

Table 2. Age at diagnosis and gender distributions of children with CAKUT.

	Gender		
Age distribution (months)	**Male n (%)**	**Female n (%)**	**Total n (%)**
<12	16 (22.22)	8 (26.67)	24 (23.53)
12-60	22 (30.56)	8 (26.67)	30 (29.41)
>60-144	23 (31.94)	6 (20.00)	29 (28.43)
>144	11 (15.28)	8 (26.67)	19 (18.63)
Total	72 (100)	30 (100)	102 (100)

The distribution of the different types of diagnoses according to age is shown in Table 3. With regards to specific anomaly such as PUJo, of the 26 children, 17 (65.38%) were males, 4 (15.38%) had bilateral obstructions, while left and right sided presentations occurred in 12 (46.15%) and 10 (38.46%) respectively. The other major anomaly was single kidney in 10 children, six and four had single kidney on the right and left respectively (Figures 1-4).

Table 3. Distribution of the different types of diagnoses according to age.

Age categories (months)					
CAKUT types	**<12 n (%)**	**12-60 n (%)**	**>60-144 n (%)**	**>144 n (%)**	**Total n (%)**
Collecting system	10 (35.71)	7 (26.92)	9 (33.33)	3 (14.29)	29 (28.43)
Kidney	6 (21.43)	10 (38.46)	13 (48.15)	17 (80.95)	46 (45.10)
Bladder/Urethra	12 (42.86)	9 (34.62)	5 (18.52)	1 (4.76)	27 (26.47)
Total	28 (100)	26 (100)	27 (100)	21 (100)	102 (100)

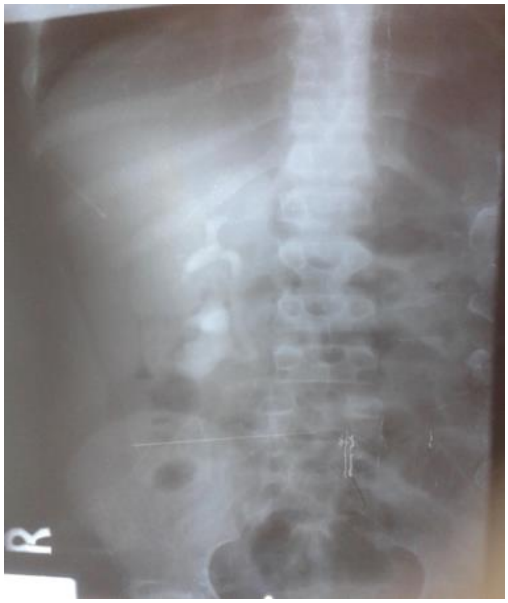

Figure 1. Intravenous urography of one of the children with right sided cross-fused ectopia.

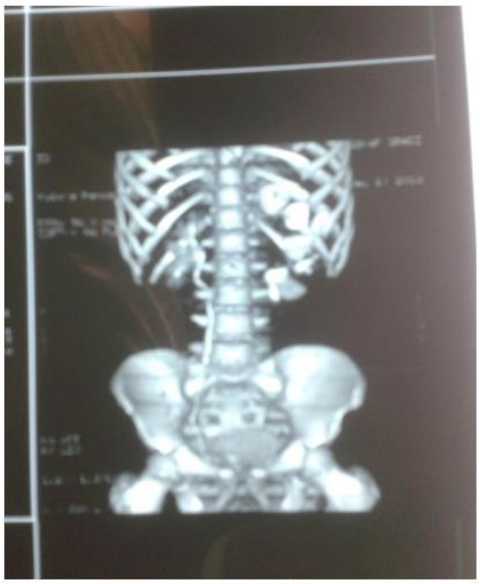

Figure 2. 3D Volume rendered image from computerized urography in a child with left sided pelvis-ureteric junction obstruction.

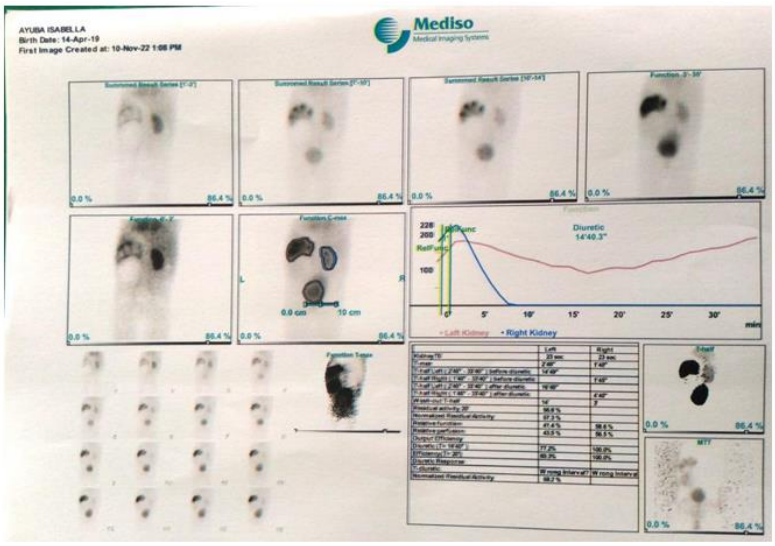

Figure 3. MAG3 renogram of a 3-year old girl with left PUJo.

The coexistence of renal and non-renal disease conditions with CAKUT was observed as well. These comorbidities are shown in Table 4.

Table 4. Coexistence of CAKUT with other renal and non-renal morbidities.

Types of CAKUT	Comorbidity	Frequency
Posterior urethral valves	Down syndrome	1
Unilateral dysplastic kidney	Hypertrophic pylorus	1
Unilateral dysplastic kidney	Nephrotic syndrome	1
Single kidney	Sickle cell anaemia	2
Posterior urethral valves	Sickle cell anaemia	1
Pelvi-ureteric junction obstruction	Renal abscess	1
Single kidney	Nephrotic syndrome	1
Single kidney	Acute kidney injury	1
Unilateral dysplastic kidney	Congenital hydrocephalus	1
Total		10

With regards to management, only six children with PUJo had surgical treatment. Four had dismembered pyeloplasty while two had outright nephrectomies. The remaining were managed conservatively in the form of regular follow up with ultrasound review.

DISCUSSION

The prevalence of 14.8% of CAKUT in the current report falls within the prevalence rates among paediatric population with kidney diseases, which ranges between 3.5% and 43% from studies in both low and high-income countries. For instance, it is comparable with the 20% reported from South Africa but higher than the 8.62% from Senegal. These differences probably arose from study methodology such as the duration of study, the age of study participants, the study population. While the current study was prospective, spanning a period of 13 years and among children aged from birth to 18 years, the studies from South Africa and Senegal were retrospective by design. The age brackets of the study populations in both groups are similar to the present study, 2 weeks to 18 years and from birth to 15 years respectively, but vary however in population characteristics. Our study population comprised of children in both general paediatric population and those managed in the renal unit, but theirs were only from patients seen primarily in the renal unit with established diagnosis or who were being assessed for CAKUT because of risk such as urinary tract infections. The study from Senegal covered a period of five years in contrast to the longer duration in this study.

In some other reports, the study designs included both children and adults or those primarily diagnosed antenatally with CAKUT. These factors could have contributed to the varying prevalence rates of CAKUT as reported in those studies

compared with the current study.

Pelvi-ureteric junction obstruction was the most common CAKUT identified in this study. This finding is at variant with other reports that showed posterior urethral valves to be more common. In the report from South Africa, the prevalence of PUV was 45% of all cases of CAKUT reviewed over a 10-year period from a single paediatric renal unit. Similarly, a PUV rate of 36.4% was reported among 107 different types of CAKUT in an Egyptian study. It is difficult to state what plausible explanation to advance for these contrasting figures, though it is known that PUJo is the commonest cause of antenatal hydronephrosis. Furthermore, since PUJo can be seen in both genders, it is possible more individuals will present with this condition commonly unlike PUV that only affects the male gender. Secondly, in our institution, some of these patients with obvious CAKUT such as PUV are also seen and managed in the paediatric surgical unit without recourse to the paediatric renal unit except and until they observed evidence of impaired renal function either on admission or during subsequent follow up after valve ablation. In the present study, PUJo is predominant in males (65.38%) and a predilection for left kidney (12/22), while in four bilateral PUJo was observed.

Posterior urethral valve was the second most common CAKUT (20.59%). The mean age at diagnosis was 3.51 years (range from day one to 14 years). Only four (19.05%) of the 21 children with PUV were diagnosed in the first year of life as compared with the 58% and 77% in two previous South African studies. This difference is due to their higher rates of antenatal detection compared to the abysmally low antenatal detection rate of 8.82% in the current study. The antenatal detection rate observed in the current study is much lower than the international detection level of 60% to 70%, and even the 22.41% reported from Senegal.

CONCLUSION

In conclusion, late diagnosis of children with CAKUT in Jos remains a challenge. The commonest type of CAKUT is PUJo. Male children are more frequently affected than females. Antenatal detection rate is unacceptably very low. The surgical management of children with CAKUT particularly those with PUV and PUJo is still pose a huge challenge as trained and skillful personnel, as well as non-availability of enabling surgical equipment in the public tertiary health facilities.

RECOMMENDATIONS

It is recommended that regional and national societies of paediatric and other relevant sub-specialists develop management guidelines that take into consideration our peculiarities. This will enable care givers rise to the occasion of proactive and effective care for children with CAKUT with regards to early diagnosis and treatment. Such early diagnosis should include advocating for and improving skills of health care providers for prenatal detection. In addition, effort at building capacities and improvement of infrastructures through both collaborations with established centers or government commitment should be made to ensure effective treatment is provided for affected children.

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