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Congenital Abnormalities of Kidneys and Urinary Tract in Children at the Dakar University Hospital

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Abstract

Congenital abnormalities of the kidneys and urinary tract (CAKUT) represent all the morphological abnormalities associated with a disorder of the embryonic development of the kidneys and their excretory tract. Its prevalence is underestimated in developing countries compared to developed countries. This was a cross-sectional and descriptive study between January 1st, 2015 and February 28th, 2020, carried out in the pediatric department of Aristide Le Dantec Hospital. The prevalence of CAKUT was 8.77% for all pediatric nephrologic pathologies (n = 58), the median age at diagnosis was 77.28 months and the sex ratio was 1.6. The circumstances of the discovery of CAKUT were dominated by urinary tract infection (n = 21), the antenatal diagnosis was poor (n = 13) and confusion between cysts and calyx dilation in antenatal was noted. Ultrasound was the main postnatal imaging test requested. The most common renal abnormality was kidney cysts (n = 18) (cystic kidneys and multicystic dysplastic kidney) and the most common urological abnormality was the posterior urethral valves (n = 8). Kidney failure was a pejorative factor in the evolution of these CAKUT.

Keywords

Urinary Tract, Antenatal Diagnosis, Congenital Abnormalities, Children

1. Introduction

Congenital anomalies of the kidneys and urinary tract (CAKUT) represent all morphological anomalies related to a disorder of the embryonic development of

the kidneys and their excretory tract [1]. CAKUT can affect the entire urinary tract, from the kidneys to the urethra. They may be cystic or dilated, or abnormal in number or position [2] [3] [4] [5]. CAKUTs are discovered in a variety of circumstances, during a urinary tract infection [6], an imaging examination or in patients presenting urinary signs with sometimes an alteration of the renal function [7]. They remain among the leading causes of chronic renal failure in children in developed and developing countries [6]. They may be associated with other malformations or form part of a syndromic pattern [8]. Indeed, ultrasound is the first-line diagnostic imaging of CAKUT [7] for which the medical and surgical management has recently been modified by advances in antenatal diagnosis [6]. CAKUT are the main reason for prenatal consultation in paediatric surgery and represent a vast pathological field [1]. They are estimated to occur in 1% of all pregnancies [9] and account for 25% of childhood surgical conditions [9]. This makes them a major public health problem, ranking fifth behind neurological, cardiac, otorhinolaryngological and digestive malformations [10]. This prevalence is underestimated in developing countries compared to developed countries [9]. The aim of this study was to describe the epidemiological and diagnostic aspects of CAKUT in the paediatric department of Aristide Le Dantec Hospital (HALD) to assess the quality of antenatal diagnosis of CAKUT and to identify children with associated renal failure at the time of diagnosis.

2. Patients & Methods

2.1. Type and Time Period of Study

This was a cross-sectional, descriptive study led between January 1st 2015 and 28 February 28th 2020.

2.2. Study Setting

The study was led in paediatric nephrology unit housed in the paediatric department of the HALD in Dakar. It is the only paediatric nephrology unit in the whole country (Senegal) with two paediatric nephrologists. HALD is a level 3 teaching hospital where all children with kidney disease requiring advanced management are referred to.

2.3. Study Population

This is the total number of children hospitalised or seen in consultation in the paediatric nephrology unit for CAKUT.

2.4. Inclusion and Non-Inclusion Criteria

All children aged 0 - 15 years with CAKUT were included. Unusable or incomplete files were not included, as well as lost files and those managed by surgeons without going through the paediatric nephrology unit. In Senegal, the patients files are not computerized. Incomplete or lost files could not be filled out because we were unable to contact the parents.

2.5. Data Collection

Data were collected from hospitalization and consultation files and a survey form was used to conduct our study.

2.6. Definitions of Operational Variables

2.6.1. Diagnosis of CAKUT

We have identified and defined CAKUTs on the basis of imaging findings (antenatal or postnatal kidney and urinary tract ultrasound and/or neonatal computerized tomography (CT) urogram) [11] [12] [13].

2.6.2. Biological Parameters

- Glomerular filtration rate (GFR) was calculated from blood creatinine and height according to the 2009 Schwartz formula [14].
- Acute renal failure was confirmed by the acute (less than 3 months) decline in GFR according to the child's age-related standards [14].

2.6.3. Management

The treatment consisted of medical and surgical management in collaboration with nephrologists and paediatric surgeons.

2.6.4. Statistical Analysis

After entering the data into the Excel database, the analysis was done using Excel 2016 and RStudio1.3.1093. Qualitative variables were presented as absolute frequency and percentage and quantitative variables as median with minimum and maximum or mean ± standard deviation, depending on their validity condition.

3. Results

In this study, 58 children with CAKUT were identified, representing a prevalence of 8.77% of renal pathologies followed up in the paediatric nephrology unit and 1.78% of all pediatrics hospitalizations. Of these children with CAKUT, 43 (74.14%) were diagnosed during hospital admission. The median age at diagnosis was 77.28 months [0.43 - 192 months]. Most CAKUT were diagnosed after one year (n = 40, 68.96%) with almost half of them (n = 19, 47.50%) diagnosed after 10 years (**Figure 1**). The sex ratio was 1.6. The circumstances of discovery of CAKUT were dominated by urinary tract infection (n = 21, 36.20%), abdominal pain (n = 15, 25.86%) and antenatal diagnosis (n = 13, 22.41%). Antenatal diagnosis, through antenatal ultrasound, was made in less than a quarter of the children (n = 13, 22.41%). In most cases (n = 12, 92.31%), cysts were found on antenatal ultrasound; in one case (7.69%) it was a hydronephrosis. Postnatal ultrasound was performed in all children (n = 58, 100%). Cysts (n = 19, 32.76%) and ureteral and/or caliceal dilatation (n = 19, 32.76%) were mainly found. Among children who had a post-natal ultrasound, 13 (22.41%) had an antenatal ultrasound. Sometimes the postnatal ultrasound confirmed the ante-natal ultrasound (n = 10, 76.92%), in other times their results were divergent (n = 3, 23.08%). In the discrepant cases (n = 3), antenatal ultrasound showed cysts,

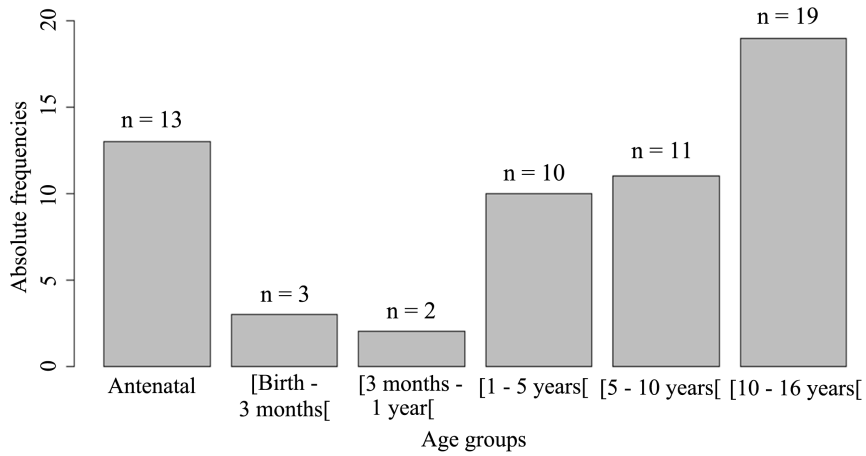

Figure 1. Age at time of diagnosis of CAKUT.

while postnatal ultrasound showed caliceal dilatation. The CT urogram (**Figure 2**) was performed in 20 children (34.48%) and had confirmed the postnatal ultrasound findings in all cases. Static renal scintigraphy (**Figure 3**) was performed in 2 children (3.45%) and dynamic renal scintigraphy in 10 children (17.24%) (**Figure 4**). Retrograde urethrocystography (RUC) was performed in 9 out of 15 (60%) children who had the indication. It was normal (n = 3) or showed vesico-ureteral reflux (VUR) (n = 6). The mean creatinine level was 10.16 mg/L [2.11 - 69.4 mg/L] and the mean GFR was 73.32 ml/min/1.73m^2 [10 - 165 ml/min/1.73m^2]. Twenty-five children (43.10%) had renal failure at time of diagnosis. Cystic kidneys were the main diagnosis of CAKUT (n = 18, 31.04%). The distribution of children with cystic kidneys was as follows: multi-cystic kidney dysplasia (n = 6), autosomal dominant polycystic kidney disease (n = 5) and cystic kidneys of unspecified diagnosis (n = 7). The remaining diagnoses were represented in **Table 1**. The other diagnoses selected were shown in **Table 1**.

4. Discussion

CAKUT account for approximately 20% - 30% of all prenatal anomalies and are considered the leading cause of chronic kidney disease (CKD) in children [15].

The frequency of CAKUT was 8.62%. This frequency is variously assessed around the world and seems to be correlated with the number of years of the census. This is the case in African, American and European studies [6] [7] [16] [17]. The small sample in our study could be explained by the fact that some children were managed directly in the paediatric surgical wards without being seen by the paediatric nephrologist. In our study, the mean age at diagnosis was higher than in other African and European studies [7] [8] [17]. At the international level, antenatal screening for CAKUT is in the order of 60% to 70% [6] [15]. In Africa, antenatal screening is low as reported in our study (22.41%) and the age at diagnosis is later [6], explained on one hand by the lack of systematisation of prenatal ultrasound scans and the lack of expertise of sonographers; on the other hand, symptomatic treatment of recurrent urinary tract infections

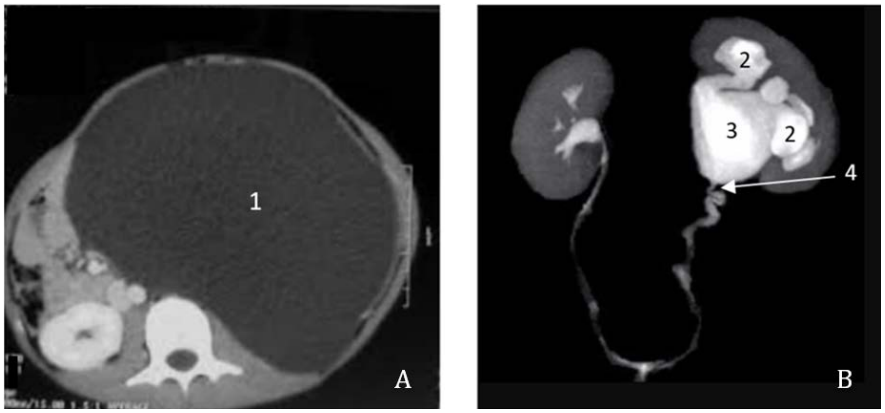

Figure 2. Abdominal CT scan in coronal (right) and axial (left) sections. (A) Left syndrome of the pyelo-ureteral junction (PUJS) (left) in an 11-month-old infant; 1: Significant dilatation of the left renal pelvis, resulting in an abdominal cystic mass. (B) Left PUJS (right) in a 9-month-old infant; 2: Dilatation of calyxes, 3: Renal pelvis dilatation, 4: level of stenosis (pyelo-ureteral junction).

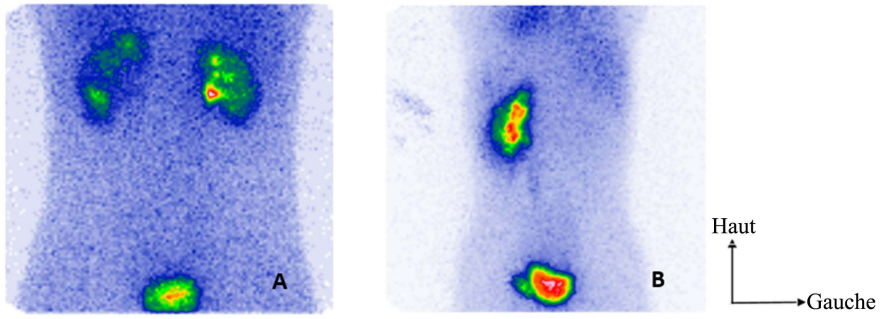

Figure 3. Images from a static renal scan. (A) Left PUJS (right): right kidney of normal position and size with relatively homogenous radiotracer fixation (right kidney); left pyelocalic dilatation (left kidney). (B) Right single kidney (right): right kidney of normal position and size with relatively homogeneous radiotracer fixation (right kidney); vacuity of the left renal cavity and absence of radiotracer fixation (left kidney).

Table 1. Final distribution of the different CAKUT diagnoses.

Diagnosis		Frequency (n)	Percentage (%)
Kidney Anomalies	Cystic kidneys	18	31.04
	Hypodysplasia of the kidney	8	13.79
	Single kidney	5	8.62
	Ectopic kidneys	4	6.90
Urinary Tract Anomalies	PUJS	7	12.07
	Congenital mega-ureter	3	5.17
	Urethral duplication	5	8.62
	PUV	8	13.79
	Total	**58**	**100**

PUV = posterior urethral valves; **PUJS =** pyelo-ureteral junction syndrome.

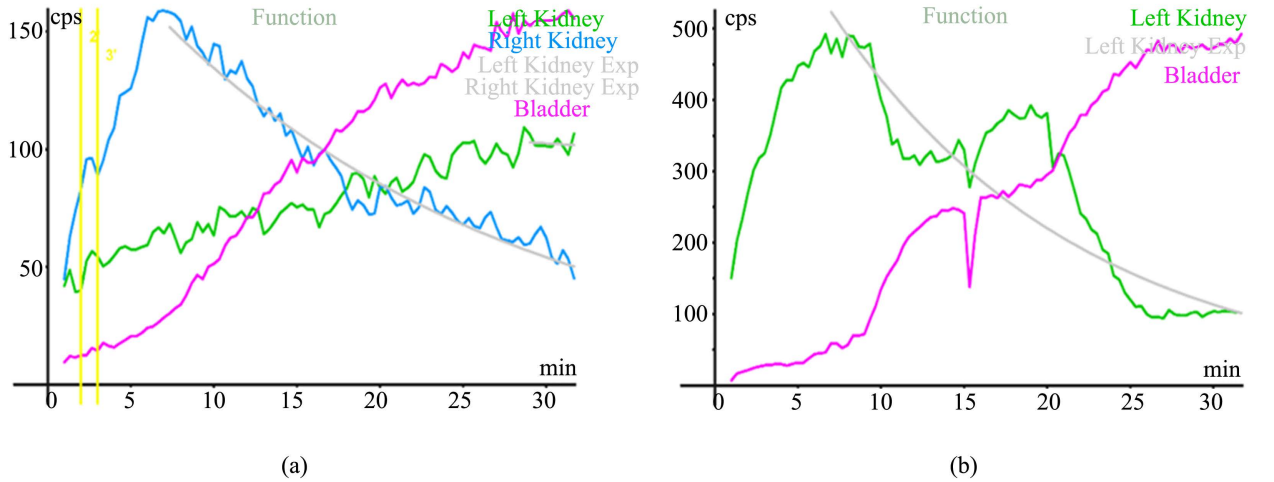

Figure 4. Dynamic renal scan images; right kidney uptake and excretion (blue), left kidney uptake and excretion (green) and bladder filling (purple). (A) Left PUJS (left): weak and very delayed capture of the tracker with a Tmax of 28'40. Absence of spontaneous excretion with a continuously rising curve until the 25th minute, then a plateau until the end of the examination, indicating significant dilatation of the pyelocalic cavities which is not very sensitive to Lasilix (left kidney); higher uptake of the radiotracer with a Tmax within the normal range (7'). Spontaneous excretion more or less rapid, not very sensitive to Lasilix because almost complete at the 20th minute (right kidney). (B) Single left kidney (right): capture of the radiotracer in time with a Tmax of 6'40", followed by a more or less rapid and almost complete spontaneous excretion at the end of the dynamic acquisition.

without extensive imaging may delay or mislead the diagnosis of CAKUT [18] [19]. In our study, urinary tract infection was the main presenting symptom of postnatal CAKUT. This finding was found in most studies; anatomical abnormalities of the urinary tract hinder proper drainage of urine, resulting in stasis that can lead to urinary tract infection [6] [17]. Despite the adjustment of some antenatal diagnoses, postnatal ultrasound confirms in most cases the results of antenatal ultrasound. Adjustments were in all cases due to confusion between cysts and antenatal calyceal dilatation. The main differential diagnosis of renal cysts in antenatal care is calcific dilatation [13] [19]. In cases where the CT urogram was done, it confirmed the results of the postnatal ultrasound. CT comes after ultrasound in the exploration of the urinary tract [13]. Despite its importance in assessing the renal parenchyma and the distribution of renal function between the two kidneys, it was only performed in 12 children. These data are similar to those from Tunisia [6]. In cases where RUC was performed, VUR was found in the majority of cases. VUR is the main indication for performing a RUC [6] [13]. Almost half of our patients had a renal failure. The progression of urinary malformations to renal failure is the most serious and potentially life-threatening complication [20]. In the European and American series, renal failure due to CAKUT was variable. Diagnostic delays and lack of early management might be detrimental factors in the occurrence of impaired renal function, hence the importance of antenatal diagnosis by ultrasound, possibly leading to termination of pregnancy in non-viable forms of CAKUT, and of early multidisciplinary management from birth [6]. At the end of our study, we make the following recommendations:

- To improve the quality of prenatal consultations and insist on antenatal obstetrical ultrasounds;
- To improve training in fetal echomorphology for obstetricians and radiologists;
- To Involve the pediatric nephrologist as soon as CAKUT is suspected in the antenatal or postnatal period;
- To consolidate the collaboration between obstetricians, pediatric surgeons and pediatric nephrologists in the ante- and post-natal care of these children, as well as in pre- and post-operative care;
- To avoid treating recurrent urinary tract infections without doing the necessary investigations;
- To improve the record-keeping system and archives organization;
- To open a register of CAKUT for a better appreciation of their frequency and a better organization of their management.

5. Conclusion

CAKUT are relatively common in paediatric nephrology. Antenatal diagnosis is poor and the age at the time of diagnosis is late, due to the lack of collaboration between obstetricians, nephrologists and paediatric surgeons. Urinary tract infection is the main circumstance of discovery and renal failure is a major factor in their evolution.

Conflicts of Interest

The authors declare that they have nothing to disclose.

Ethical Approval

This study was conducted in accordance with the Declaration of Helsinki. To respect confidentiality, an identification code was assigned to each patient. This study was hospital-based research conducted in routine conditions. "Oral informed consent" was taken from patients because the majority of them were not literate.

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