Research article

INVOLUTION OF UNILATERAL MULTICYSTIC DYSPLASTIC KIDNEY: LONG TERM DATA FROM A SINGLE CENTER

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ABSTRACT

Unilateral multicystic dysplastic kidney (MCDK) is congenital anomaly which can be detected by ultrasound during antenatal period. The reported rate of MCDK ranges between 1 in 2,200 to 1 in 4,300 live births. This study was aimed to assess the determinants associated with the MCDK with follow-up until teens to evaluate the evolution of the disorder. This was an observational longitudinal study, involution degrees were observed following the baseline measurement of the size of the MCDK as documented on postnatal ultrasound, and long-term complications and renal function were also recorded. A total of 350 patients were identified, while 90 patients fulfilled the criteria of the study and completed the study duration of 10 years. Out of which 19 (21.1%) patients reported involution of MCDK size during follow-up as complete and partial, 7 (7.7%) within first year of age, 5 (5.5%) during first 2 years and 7 (7.7%) within 4 years of age. Large-sized MCDK at the time of diagnosis is less likely to involute through the first decade of life. However, conventional treatment is rational with the absence of complications.

Key Words: Multicystic Dysplastic Kidney, Solitary functioning Kidney, Involution of MCDK

INTRODUCTION

Multicystic dysplastic kidney (MCDK) is a congenital renal anomaly causing chronic renal failure in children (1). It is histologically characterized by abnormal and incomplete differentiation of metanephric mesenchymal tissue and ureteric buds, having fibromuscular tissue around the cysts(2). Unilateral Multicystic dysplastic kidney is commonly diagnosed during antenatal ultrasound. It's rate of occurrence ranging from 1 in 2,200 to 1 in 4,300 live births (3). The prevalence is higher in male fetuses, especially bilateral MCDK associated with oligohydramnios (4). Involution of MCDK with time has been reported in many studies, longer follow-up studies identified an increased risk of hypertension and renal malignancies in MCDK patients with time (5). The literature suggesting etiology of MCDK remains inconsistent, where some studies suggested positive relation with familial risk where there was a history of solitary kidney in first family members and MCDK incidents (6). However, associated disorders such as hypospadias, bladder diverticulum, Ehler-Danlos syndrome, and DiGeorge syndrome are known to be linked with MCDK (7).

The histopathological differences between polycystic kidney and MCDK are the presence of scanty and dysplastic abnormal cells between cysts and the absence of parenchyma in MCDK (8). Although conservative management of MCDK has been acknowledged as an improved management route instead of nephrectomy but the literature is still not sufficient as the follow-up of an MCDK child till adulthood is limited (9). First identification of MCDK is usually through antenatal ultrasonography, or fetal abnormality scan performed in many established health care setups to identify any anatomical abnormality in the fetus before delivery. The number of antenatal MCDK documentation is not done in routine clinical practice as antenatal ultrasonography is not an established regulation in all obstetric setups. If not identified in antenatal scans, the child

may present with palpable abdominal mass. Vesicoureteral reflux (VUR) is reported to be seen in 20% of MCDK cases, while pelvic ureter junction obstruction (PUJO) can also be seen on the contralateral side of MCDK as additional findings (10). The kidney with moderate to severe hydronephrosis may be identified as PUJO (11). The management choices include a conventional method of investigation of the kidney dimension based on ultrasonography as MCDK usually endure involution by the age of 10 years. This observation also aids in identifying malignant variations or obstructive features in the contralateral kidney (12, 13). Though the condition is not very rare in Pakistan but there is limited literature available. Therefore, this was conducted aimed is to assess the frequency of MCDK in children presented in our institute, and the determinants associated with the disease with follow-up to ten years to evaluate the progression of the disorder

METHODOLOGY

Study Design:

This was a longitudinal observational study conducted in the Pediatric urology department of Liaquat University of Medical and Health Sciences, Jamshoro, Pakistan. Institutional approval from the ethical committee was obtained to conduct the study. The patients were enrolled via eligibility criteria of presented in the hospital within first month of life with unilateral MCDK. All the bilateral cases of MCDK were excluded from the study. Upon successful enrollment the demographic details were obtained from medical records starting 2012 till 2022, a total of 10 years of data was used to assess follow-up and involution of MCDK.

A total of 350 patients was brought in and 90 patients were deemed eligible to be recruited in this study and they were evaluated for involution frequency and determinants of MCDK.

Involution:

Involution suggests the reduction in the number or dimension of the multiple cysts present in kidneys. Whereas complete involution is the significant loss of cyst, and partial involution was taken as a reduction in the number or size of cysts. The baseline at the first encounter was considered as the initiation point while disease initiation was taken as the date of child birth, while the endpoint was the date when imaging showed no residual cysts or the last follow-up.

Complications:

An increase in Serum creatinine and blood pressure was monitored as complications along with radiological investigations, documented urinary tract infections, or any sign of lower urinary tract symptoms (LUTS) were also analyzed. Vesicoureteral reflux was identified as an associated complication on the contralateral side.

Statistical Methods

The data was entered and analyzed in Statistical package of social sciences (SPSS version 22.0), mean and standard deviation (±SD) were calculated for age, and years of MCDK diagnosis. The size on follow-up ultrasound were defined as Stasis of MCDK (Similar to the first measurement), Decrease in MCDK measurement (size reduction), and, growth in MCDK (increase in size). A p-value of <0.05 was considered significant.

RESULTS

A total of 90 children were enrolled in the study for 10 years, the mean age at presentation was 93.2 ± 137.3 days, with 70 (77.8%) males and 20 (22.2%) females. 06 (6.7%) of the children were diagnosed during antenatal radiological investigations while 84 (93.3%) were diagnosed post-delivery and referred to the urology department. Laterality of MCDK was 13 (14.5%) on the right side while 39 (43.3%) on the left side. The associated factors were reported as twin pregnancy in 7 (7.8%) and intrauterine growth restriction in 3(3.3%) of patients. While in antenatal scans amniotic liquor quantity was adequate in 81 (90%) of the patients, only 9 (10%) reported inadequate liquor during pregnancy scan. A summary is presented in Table 01. The mean serum creatinine at the time of presentation was 0.4 ± 0.1 mg/dL. Upon follow-up 07 (7.8%) children

indicated an increase in MCDK size within one year (in 1 patient), at 4 years of age (in 1 patient), and at 8 years (1 patient). A summary of the duration is given in Figure 01.

The size increase in these subjects was measured between 3.1 – 4.0 cms to 6.1 – 7.0 cms, and 02 patients with maximum size improvements proceeded with nephrectomy to avoid the relevant risk of malignancy. The MCDK was reportedly decreased in the size in 19 (21.1%) patients after conservative management and in-depth evaluation during follow-up. Involution was identified in 7 (7.7%) patients within the first year of age, while 5 (5.5%) within 2 years and another 7 (7.7%) in 4 years of age. A summary is given in Figure 2. Involution was documented as complete, partial, and unchanged. With respective of laterality of MCDK, results indicated that right (n=13) sided MCDK showed partial involution in 5 (5.5%) patients while 2 (2.2%) patients were unchanged. The left (n=77) sided MCDK indicated complete involution in 2 (2.2%) patients, 12 (13.3%) showed particle involution and 33(36.6%) were unchanged. The contralateral side showed pelvic ureter junction obstruction in 3 (3.3%), while vesicoureteral reflux was diagnosed in 5 (5.5%) of patients.

Table I: Demographic details of Multicystic dysplastic kidney (MCDK) children presented in Out Patient Department

Details of study subjects		
Variables (n=90)		Frequency N(%)
Gender	Male	70 (77.8%)
	Female	20 (22.2%)
Presentation	Antenatal	6 (6.7%)
	Postnatal	84 (93.3%)
Referral	Primary	51 (56.7%)
	Secondary	39 (43.3%)
MCDK side	Right	13 (14.5)
	Left	77 (85.6%)
Associated factors	Twin pregnancy	7 (7.8%)
	IUGR	3 (3.3%)
	None	80 (88.9%)
Liquor quantity	Adequate	81 (90%)
	Not adequate	9 (10%)
Term on delivery	Full term	81 (90%)
	Preterm	9 (10%)

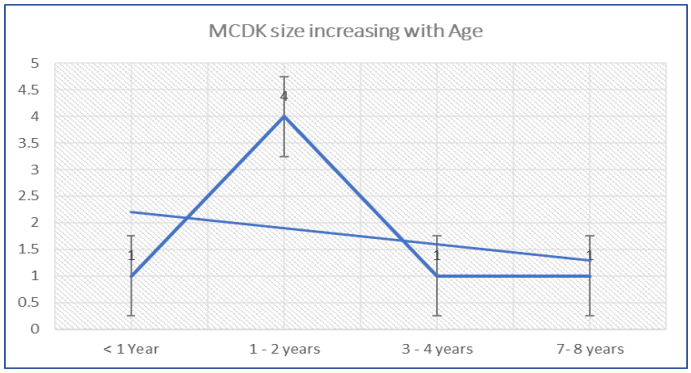

Figure 1. Frequency of MCDK size improvement with advancement in age of study subjects

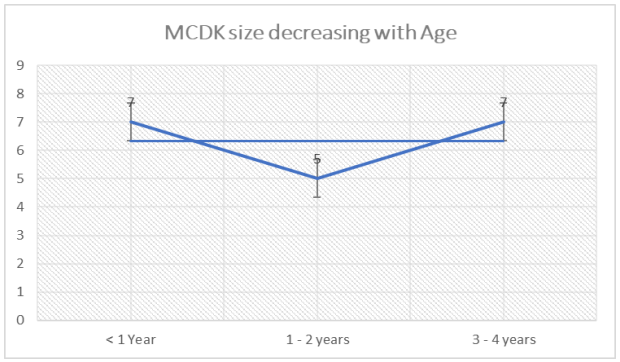

Figure 2. Frequency of MCDK size declines with advancement in age of study subjects

DISCUSSION

This study evaluated the involution of unilateral MCDK presented in single-center, our results indicated the maximum involution within first 10 years of age. Multicystic dysplastic kidney frequently seen congenital anomaly in children in urology practice, with a risk renal failure later in life. The renal abnormality developed during fetal development at gestational age with a higher prevalence in male infants as compared to females (14). The diagnosis during the antenatal scan is less frequent in developing countries, the reason for minimal antenatal diagnosis of renal anomalies including MCDK might be limited access to antenatal care and denial of radiological investigations during pregnancy by parents. This limitation of antenatal diagnosis has been reported in prior studies as well (15-16). Twin pregnancy was associated with congenital anatomical abnormalities in the fetus, in this study 7 (7.8%) of infants were twins, and intrauterine growth restriction was presented in 3 (3.3%) of infants indicating minimal risk of MCDK in IUGR children, this was also reported in previous studies (17). The literature supported our results as studies evaluated maximum positive involution, complete or partial before adulthood. MCDK size was reported as increased during the first 7 years of age with a maximum of 4.00 cms from the first reported size of MCDK, another study indicated a 15% MCDK increase requiring nephrectomy, while another study of 46 patients reported a 39.1% increase in MCDK size during the first year of age (13,18). While partial involution of MCDK was previously reported in 40.9% on the right side while 50% on the left side, however, in our study partial involution was reported as 5.5% and 13.3% on right and left sides respectively. While complete involution was reported in 27.2% of the same study, our study had 2.2% of complete involution of patients 19. The reason for the drastic difference in the involution rates between these studies is probably follow-up duration. We have evaluated patients for a longer period as compared to previously reported studies. Associated problems such as vesico ureter reflux, pelvic ureter junction obstruction, and hydronephrosis were identified in 5 (5.5%) and 3 (3.3%) respectively on the contralateral side of the affected kidney in our study. A previously reported study including 80 children diagnosed with MCDK reported VUR in 13 (16.3%) children while PUJO was in 2 (2.5%) only (20). Other congenital anatomical abnormalities of the renal system were not reported except for recurrent urinary tract symptoms in our study. Nephrectomy was performed in 2 (2.2%) of patients indicating a ≤ 4.0 cms increase in MCDK size, another study reported 7 (5.5%) nephrectomies due to improved MCDK size (21).

This study was reported from a single center with long term follow-up. However, the sample size was smaller, which is considered as the limitation of the study. The follow-up was also shorter, further long term follow-

up and regular measurement of eGFR is also suggested to be evaluated to understand natural progression of the disorder.

CONCLUSION

The study concluded that a small number of patient cyst involute with passage of time, though large-sized MCDK at the time of diagnosis is less expected to involute through the first decade of life. However, conventional treatment is rational with absence of complications. Further long term follow-up studies are recommended to understand the disorder so that proper management guidelines can be designed.

Ethical Consideration: The study was approved by local Research Ethics Committee
Conflict of Interest: There is no conflict of interest.
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