

# Wait and Watch: Management Strategies of Multi Cystic Dysplastic Kidney in Pediatric Population

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## ABSTRACT

**Background:** One in 2,200 to 1 in 4,300 babies have unilateral multicystic dysplastic kidney (MCDK), one of the most common abnormalities found by prenatal ultrasound.

**Objective:** The study aims to quantify the prevalence of MCDK in children who are brought to our facility and the characteristics associated with the disease.

**Methods:** Participants in this retrospective cohort study were added after the fact if they met the eligibility requirements of being diagnosed with unilateral MCDK and presenting to the hospital within the first month of life. Version 22 of the Statistical Package for Social Sciences was used to enter and analyze the data. For age and years of MCDK diagnosis, the mean and standard deviation were calculated. Frequencies, percentages, mean values, and standard deviations will be analyzed for independent data.

**Results:** The 15-year research included 90 children, with a mean age of  $93.2 \pm 137.3$  days, 70 (77.8%) males and 20 (22.2%) females. The average size of MCDK was  $6.2 \pm 1.2$  cms on the right side and  $5.4 \pm 1.63$  cms on the left. During follow-up, 7 (7.8%) children showed an increase in MCDK size within <1 year, 2 years, 4 years, and 8 years.

**Conclusion:** Studying 15 years of our MCDK patient records included evidence supporting the current literature, primarily positively advocating non-operative delivery of MCDK with periodic follow-up and radiological tests.

**Keywords:** Solitary functioning Kidney, Multicystic Dysplastic Kidney, pediatric patients, Renal health,

## INTRODUCTION

One of the most prevalent renal abnormalities that causes chronic renal failure in children is multicystic dysplastic kidney disease (MCDK).<sup>1</sup> The characteristic histology of the ureteric buds and metanephric mesenchymal tissue's aberrant and incomplete development, with fibromuscular tissue encircling the cysts, serves as the basis for the diagnosis.<sup>2</sup> With an incidence of 1 in 2,200 to 1 in 4,300 births, unilateral multicystic dysplastic kidney (MCDK) is one of the most prevalent abnormalities found by prenatal ultrasound.<sup>3</sup> Male fetuses had a higher prevalence, particularly bilateral MCDK linked to oligohydramnios.<sup>4</sup> Numerous investigations have documented the involution of MCDK over time, and longer follow-up studies have found that patients with MCDK have a higher chance of developing hypertension and renal malignancies over time.<sup>5</sup> MCDK has an irregular etiology, and some research has found a positive correlation between an MCDK occurrence and a history of a single kidney in first family members.<sup>6</sup> The presence of sparse, dysplastic

aberrant cells between cysts and the lack of parenchyma in MCDK are the histological differences between polycystic kidney disease and MCDK.<sup>7</sup> The literature is still insufficient since it is uncommon to follow up with MCDK children into adulthood, despite the fact that conservative care of the disease has been recognized as a better management option than nephrectomy.<sup>8</sup> In many well-established healthcare facilities, antenatal ultrasounds, also known as fetal abnormality scans, are used to detect any anatomical abnormalities in the fetus prior to delivery. This is typically how MCDK is initially identified.<sup>9</sup> Since prenatal ultrasonography is not a standard procedure in all obstetric settings, the number of antenatal MCDK findings or documentation is the lowest. Additionally, MCDK is known to be connected with related conditions such as hypospadias, bladder diverticulum, Ehler-Danlos syndrome, and DiGeorge syndrome.<sup>10</sup>

The infant may have a palpable abdominal mass if it is not detected by prenatal imaging. A patient with polyc-

ystic kidney disease has a smooth palpatory surface, whereas a patient with MCDK has an uneven region with decreased appetite, trouble eating, and distension of the abdomen.<sup>11</sup> Twenty percent of MCDK cases have vesicoureteral reflux (VUR), and the contralateral side of MCDK is associated with pelvic ureter junction obstruction (PUJO).<sup>12</sup> The kidney with Moderate to severe hydronephrosis may be identified as PUJO, Hydronephrosis needs to be under strict surveillance for functioning check to assess the requirement of intervention and preserve the functionality of solitary kidney and avoid renal failure as MCDK kidney is completely non-functioning and shows 0% uptake in DMSA scan. Though the results are nearly identical to those of an MCDK kidney that is not working, some medical professionals prefer MAG-3 scans, which are only useful for evaluating the kidney's condition prior to nephrectomy.<sup>13</sup> Since MCDK often undergoes involution by the age of ten, one of the available therapy options is a traditional approach to kidney dimension evaluation based on ultrasonography. Additionally, this observation helps detect obstructive characteristics or malignant changes in the contralateral kidney. Antibiotics used to treat VUR-associated UTIs may or may not be to blame for this. Regardless, a youngster with a single kidney should be kept under close supervision for stones or hyperfiltration injury.<sup>12</sup> Monitoring for hypertension should be of utmost importance till adulthood. Although the open method of removing the affected kidney (nephrectomy) is less invasive than the surgical approach to managing MCDK, the decision to remove the kidney surgically is based on the following factors: a high risk of malignancy changes, failure of involution, imperative mass outcome, or hypertension on follow-up radiological investigations.<sup>13</sup> This study's objectives are to determine the prevalence of MCDK in children who are admitted to our institution and the factors linked to the illness, as well as to track the disorder's progression into adulthood.

## METHODS

This retrospective cohort study was carried out at Liaquat University of Medical and Health Sciences, Jamshoro, at the pediatric urology department. The study was approved by the ethics committee (LUMHS/REC/129), and patients who met the qualifying requirements of being admitted to the hospital within the first month of life and having a diagnosis of unilateral MCDK were enrolled retroactively. The study did not include any bilateral MCDK cases. After enrollment was successful, demographic information was taken from medical records from 2007 to 2022. This 15-year data set was utilized to evaluate MCDK follow-up and evolution.

A decrease in the number or size of the numerous cysts in the cystic area was referred to as involution. Partial involution was described as a decrease in the size or quantity of cysts in a chronic lesion, while complete involution was characterized as adequate removal of the cyst. The age of the patient at which imaging first revealed a decrease in cysts was the initiation point of involution, and the age of the patient at which imaging revealed no remaining cysts was the endpoint.

Blood pressure and serum creatinine levels were tracked as complications occurred, and radiographic tests, confirmed UTIs, and any indication of lower urinary tract symptoms (LUTS) were also examined. On the opposite side, vesicoureteral reflux was found to be a related consequence.

For independent data, frequencies, percentages, mean value, and standard deviation will be examined. The data was input and examined using Statistical Package of Social Sciences version 22, where mean and standard deviation were computed for age and years of MCDK diagnosis. urinary tract infection, period until full involution, further urological variations in the ipsilateral and contralateral regions, and the evolution of the contralateral kidney. On follow-up ultrasound, multicystic dysplastic kidneys were divided into three groups based on size: MCDK stability (same to the initial measurement), MCDK growth (increasing size), and MCDK decrease (reduction in size). The Shapiro-Wilk test was used to determine whether the data was normally distributed; if not, the median (IQR) will be presented. For quantitative data, frequency and percentage will be computed. Stratification and post-stratification were used to control effect modifiers such as duration of disease, antenatal diagnosis, and involution. Fisher's exact and Mann-Whitney U tests were also conducted, and a chi-square test was used to maintain a P-value < 0.05 as significant.

## RESULTS

A total of 90 children were enrolled in the study 70 (77.8%) male and 20 (22.2%) female, were enrolled in the study for 15 years. The mean age at presentation was  $93.2 \pm 137.3$  days. Of the children, 84 (93.3%) were diagnosed after delivery and sent to the urology department, while 06 (6.7%) were diagnosed during prenatal radiological exams. The related factors were twin pregnancy in 7 (7.8%) and intrauterine growth restriction in 3 (3.3%), with a p-value of 0.08. The laterality of MCDK was 13 (14.5%) on the right side and 39 (43.3%) on the left side, with a p-value of 0.21. Only 9 (10%) of the patients reported insufficient amniotic liquid during the prenatal

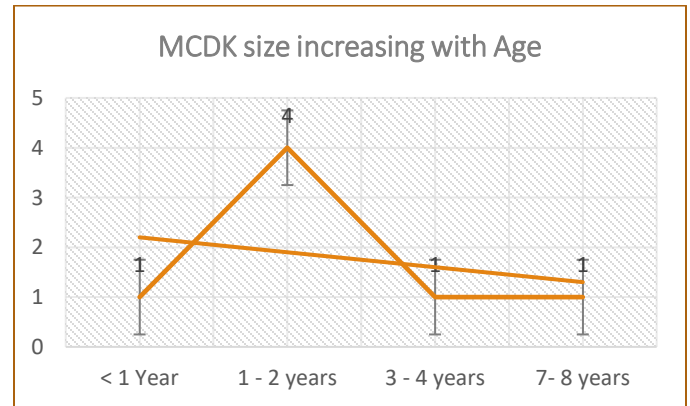
scan, whereas 81 (90%) of the patients reported adequate liquor during the antenatal scan (Table- 1).

**Table 1:** Subjective and Demographic Details of MCDK Children Presented in OPD

| Details of study subjects |                |            |
|---------------------------|----------------|------------|
| Variables (n=90)          |                | P-value    |
| 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%)    |

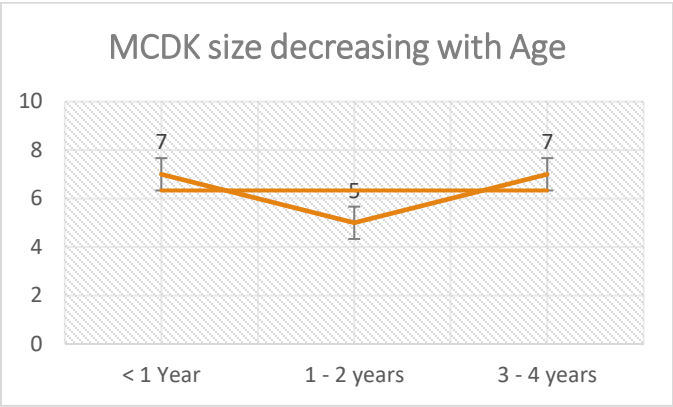
The antenatal ultrasound was performed at  $21.6 \pm 5.9$  weeks of gestation, and the mean size of the MCDK kidney from ultrasound at presentation was  $5.91 \pm 1.3$  cms. While laterality differentiated the mean size of  $6.2 \pm 1.2$  cms on the right side while  $5.4 \pm 1.63$  cms on the left side. At the time of presentation, the average serum creatinine level was  $0.4 \pm 0.1$  mg/dL. Following up, the seven (7.8%) children reported that their MCDK size had increased within the following time frames: less than a year (in one patient), two years (in zero patient), four years (in one patient), and eight years (1 patient). (Figure-1).

These subjects' size increases ranged from 3.1 to 4.0 cm to 6.1 to 7.0 cm, and two patients who experienced the greatest size improvements underwent nephrectomy in order to reduce the associated risk of cancer.



**Figure 1:** Frequency of MCDK Size Improvement with Advancement in Age of Study Subjects

After conservative therapy and thorough evaluation during follow-up, the MCDK was reported to have shrunk in size in 19 (21.1%) of the individuals. Involution was found in 7 (7.7%) of the patients within the first year of life, 5 (5.5%) over the next two years, and another 7 (7.7%) within the fourth year (Figure-2).



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

Lost to follow-up patients were contacted via available contact numbers and asked to resume their follow-ups, however, 22 (24.4%) patients were identified and documented as lost to follow-up, and a maximum number of patients were denied to resume their follow-up schedule during 5-6 years of age. The reason for a big number of losses to follow-up during these days could be fear of missing school as health care facility is located at distance and require a minimum of 3 days for patients coming from other cities or nearby villages. According to the laterality of MCDK, involution was recorded as complete, partial, and unchanged. The results showed that 5 (5.5%) of the 13 patients with right-sided MCDK had partial involution, whereas 2 (2.2%) had no change. In contrast, 2 (2.2%) of the 77 patients with left-sided MCDK had full involution, 12 (13.3%) showed partial involution, and 33 (36.6%) remained unaltered. The p-value was deemed statistically significant and was 0.04 on the right side and 0.007 on the left (Table- 2).

**Table 2:** Involution Details of Patients with MCDK

| Variables (n=90) |              | Complete involution | Partial involution | Unchanged involution | P-value |
|------------------|--------------|---------------------|--------------------|----------------------|---------|
| MCDK side        | Right (n=13) | 0                   | 5 (5.5%)           | 2 (2.2%)             | 0.04    |
|                  | Left (n=77)  | 2 (2.2%)            | 12 (13.3%)         | 33 (36.6%)           | 0.007   |

**DISCUSSION:**

Male newborns are more likely than females to have multicystic dysplastic kidney (MCDK), the most common cause of renal abnormalities during fetal development at gestational age.<sup>14-16</sup> Prenatal diagnosis of kidney abnormalities, including MCDK, is less common in developing countries; this could be because to parental refusal of radiological investigations during pregnancy and limited access to prenatal treatment.<sup>17</sup> Prior research has also documented this limitation of prenatal diagnosis;<sup>18</sup> twin pregnancies were linked to congenital anatomical abnormalities in the fetus; 7 (7.8%) of the infants in this study were twins, and 3 (3.3%) of the infants had intrauterine growth restriction, suggesting a low risk of MCDK in IUGR children.<sup>19-21</sup> The involution of unilateral MCDK shown in a single area was examined at this study; the results showed that the maximal involution took place during the first 10 years of life.<sup>22-24</sup> Our findings were supported by the literature, which evaluated the maximum positive involution—whether whole or partial—prior to maturity. A 15% increase in MCDK size necessitated a nephrectomy, according to one study; a 39.1% growth in MCDK size during the first year of life was recorded by another study of 46 individuals; and MCDK size was found to expand by a maximum of 4.00 cm throughout the first 7 years of life.<sup>13</sup> Partial involution of MCDK was detected at 5.5% on the right side and 13.3% on the left side in our analysis, although it was previously shown to be 40.9% on the right side and 50% on the left side with a p-value of 0.74. Our analysis revealed 2.2% of patients had complete involution, whereas the same study reported 27.2%. We evaluated patients over a longer period of time, which explains the considerable difference in involution between the two trials.<sup>26-27</sup> On the contralateral side of the afflicted kidney, related issues such hydronephrosis, pelvic ureter junction blockage, and vesicoureter reflux were found in 5 (5.5%) and 3 (3.3%) of the patients, respectively. Thirteen (16.3%) of the 80 children with MCDK in the research had VUR, whereas just two (2.5%) had PUJO.<sup>28</sup> Other than recurrent urinary tract complaints, no more congenital renal structural abnormalities were found during our research.<sup>29</sup> Nephrectomy was conducted in 2 (2.2%) of patients, showing a < 4.0 cms increase in MCDK size. Another research showed 7 (5.5%) nephrectomies due to enhanced MCDK size.<sup>30</sup> Despite the large patient population, this study has limitations because it was conducted retrospectively. However, the cohort study design contributed to the study's strength, yet secondary data limitations are evident. Another study with a multicenter, high sample size, and prospective design is suggested.

**CONCLUSION:**

Studying 15 years of our MCDK patient records carried data subsidizing existing literature mostly positively encouraging non-operative administration of MCDK with regress follow-up and radiological investigations. The sample size was not enough to conclude any ultimate answer, however, the duration and time of follow-up should be adjusted as per the involution course of the patient exclusively.

**ETHICAL PERMISSION:** The Research Ethical Committee of Liaquat University of Medical & Health Sciences approved this study vide letter No. LUMHS/REC/130.

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**AUTHORS' CONTRIBUTION:**

**JAJ:** Acquisition of data, manuscript writing, statistical analysis and data interpretation final approval of manuscript.

**WA:** Acquisition of data, data analysis and interpretation,

**MMA:** Data analysis and interpretation final approval of manuscript.

**MIN:** Data analysis and interpretation, manuscript writing

**ST:** Acquisition of data, analysis and interpretation, manuscript writing, statistical analysis and data interpretation final approval of manuscript.

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