RESEARCH BRIEF

# Multicystic Dysplastic Kidney: A Retrospective Study

***Sathish Sharada, Mahalingam Vijayakumar, Prahlad Nageswaran, Sudha Ekambaram and Amish Udani**
*From the Departments of *Pediatrics and Pediatric Nephrology, Mehta Children's Hospital, Chetpet, Chennai, India.*

*Correspondence to:*
*Dr M Vijayakumar,*
*Consultant Pediatric Nephrologist,*
*Mehta Children's Hospital,*
*No.2(e) Mc Nichols Road, 3rd Lane,*
*Chetpet, Chennai 600 031, Tamilnadu,*
*India. doctormvk@gmail.com.*
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**Objective:** To report the renal structural and functional anomalies in children with multicystic dysplastic kidneys. **Methods**: Retrospective descriptive analysis of 47 children with multicystic dysplastic kidney seen in a pediatric nephrology unit over a period of 6 years. **Results**: Antenatal diagnosis of multicystic dysplastic kidney was made in 34 (72.3%) patients. On follow up of 31 children for more than 12 months, 21 (68%) had involution, 4 [13%] had non-regression, and 4 (13%) were nephrectomized. Vesico-ureteric reflux (*n=13; 28%)* was the commonest renal abnormality. The serum creatinine values were higher (*P*=0.006) in children with contralateral reflux. Sub-nephrotic proteinuria was noted in 9 (29%) and was significantly associated with complete involution (*P*=<0.023). None of the patients developed hypertension and 2 (6.4%) had renal failure. **Conclusion**: Close nephrological follow-up is needed in children with multicystic dysplasia of kidneys.

**Keywords**: *Hyperfiltration injury, Multicystic dysplastic kidney, Proteinuria*.

Better and advanced antenatal ultrasound screening has led to an increased diagnosis of fetal renal abnormalities. Multicystic dysplastic kidney (MCDK) is the most common form of cystic renal dysplasia with an incidence of 1:4000 live births [1]. The natural course of unilateral MCDK is either complete or partial involution which begins early in fetal life and progresses throughout the postnatal life. The renal function depends on the functioning contralateral kidney [2]. Thus close nephrological follow-up becomes essential. This retrospective analysis of the records of children with MCDK was done to describe the follow-up of the clinical parameters and associated renal and urological abnormalities.

## Methods

Records in Pediatric nephrology services at Mehta Children's Hospital, Chennai between the years 2004-2010 were reviewed. Children with antenatally and/or postnatally detected MCDK were included. MCDK was defined as presence of multiple non communicating cysts of various sizes detected sonographically with no evidence of functioning renal parenchyma by dimercaptosuccinic acid renal scan (DMSA) [3]. Children with other cystic renal diseases and those with less than 12 months of follow-up were excluded.

Initial assessment was done by demographic data collection, blood pressure measurement along with investigations, including blood urea, serum creatinine and electrolytes, urinalysis, spot urine protein-creatinine ratio (UPCR), ultrasound abdomen, DMSA renal scan and micturating cysto-urethrogram (MCU). 24-hours urine protein analysis was done, if considered necessary. During follow-up (every 6 months), blood pressure measurements, renal function tests and ultrasonography were done. MCU and DMSA scans were repeated, if necessary.

Hypertension was defined as blood pressure more than 95th centile for age, gender and height [4]. Proteinuria was considered when spot UPCR was >0.2 mg/mg or 24-hours urinary protein was >150 mg/1.73 $m^2$/day. Urinary Tract Infection (UTI) was defined on the basis of urine culture done during febrile episodes associated with urinary symptoms. Compensatory hypertrophy of the contralateral kidney was defined as renal length of >2 SD of normal kidneys for age [5]. Modified Schwartz' formula was used to calculate estimated glomerular filtration rate (eGFR). Complete involution was defined as disappearance of the MCDK by USG abdomen [5]. Statistical analysis was done using SPSS software. Involution rate was calculated using Kaplan Meier survival analysis. Chi-squared test, Fisher exact test and Student t-test were applied to find out the association between variables.

## Results

Of the 4200 children who attended the out-patient department from 2004 to 2010, 47 children with unilateral MCDK were enrolled; their characteristics are described in ***Table* I**.

**TABLE I** CHARACTERISTICS OF CHILDREN WITH UNILATERAL MULTICYSTIC DYSPLASTIC KIDNEY

| *Patient characteristics* | *Number (%)* |
|---|---|
| *At baseline (n=47)* | |
| Males | 27 (57.4) |
| Right sided | 25 (53.2) |
| Antenatal diagnosis | 34 (72.3) |
| Urological abnormalities | 15 (31.9) |
| VUR | 13 (27.7) |
| Ipsilateral / Contralateral | 5/2 |
| Unilateral / Bilateral | 9/4 |
| Ureterocele | 1 (2.1) |
| Ectopic ureter | 1 (2.1) |
| *At follow up (n=31)* | |
| Ipsilateral | |
| Complete regression | 7 (22.6) |
| Partial regression | 14 (45.2) |
| Increase in size | 4 (12.9) |
| No change | 2 (6.4) |
| Nephrectomy | 4 (12.9) |
| Contralateral compensatory hypertrophy | 31 (100) |
| Renal failure | 2 (6.4) |
| Sub-nephrotic proteinuria | 9 (29) |

The characteristics of children with VUR is depicted in ***Table II***. Resolution of reflux was documented only in 5 children who had unilateral low grade contralateral reflux with no associated complications. Surgical intervention for VUR (ureteric reimplantation) was performed in one child with high grade contralateral reflux which was associated with recurrent UTI, proteinuria and multiple scars on DMSA. The mean (SD) serum creatinine values were higher in children with contralateral VUR than in those without VUR [0.93 (0.31) *vs.* 0.67 (0.14); $P$=0.006]. The median eGFR was 91 mL/min/1.73 $m^2$ (range: 46-163 mL/min/1.73 $m^2$]. The median eGFR was 94 mL/min/1.73$m^2$ [range: 73-137 mL/min/1.73$m^2$] in children with proteinuria and 87 mL/min/1.73$m^2$ (range: 67-121 mL/min/1.73$m^2$] in those who underwent nephrectomy. Other associated ureteric anomalies were ipsilateral ectopic ureter and ipsilateral ureterocele. Six of the 34 children diagnosed antenatally showed compensatory hypertrophy of the contralateral kidney at the initial postnatal scan. Two children diagnosed postnatally showed scars on DMSA during their initial visit at 7 and 8 years of age.

A total of 31 children were followed-up for more than 12 months with a mean follow up period of 34.55 months (range 41-142 months). Their characteristics are described in ***Table* I**. Twenty-one (67.8%) had involution of MCDK. The mean period of involution was 36 months with a minimum period of 7 months. The estimated median (95% CI) time during follow-up for complete involution of MCDK was 136 (107-169) months by Kaplan Meier survival analysis. All 31 children had compensatory hypertrophy of the contralateral kidney. The probability of complete involution at 1, 2 and 5 years were 10%, 15% and 20%, respectively. The probability of children without complete involution at 10 year follow-up was 50%. The mean follow-up of those who did not have involution of MCDK was 26 months.

**TABLE II** CHARACTERISTICS OF CHILDREN WITH MULTICYSTIC KIDNEY DISEASE AND ASSOCIATED VESICOURETERIC REFLUX ($N$=13)

| *Characteristics* | *Number* |
|---|---|
| *Unilateral VUR* | 9 |
| Ipsilateral (Grade IV) | 1 |
| Contralateral | 8 |
| Grade III | 4 |
| Grade II/I | 2 each |
| *Bilateral VUR* | |
| Ipsilateral | 4 |
| Grade III | 2 |
| Grade I or II | 1 each |
| Contralateral | 4 |
| Grade III or II | 2 each |

Four children underwent nephrectomy. Two among them had increasing size of MCDK of whom one had hypertension which resolved after nephrectomy. One child with ipsilateral ectopic ureter and another with ipsilateral high grade reflux underwent nephro-ureterectomy. Both had scars on contralateral kidney by DMSA of which the child with ectopic ureter presented at 7 years of age and the other child diagnosed antenatally, developed scars on follow-up. Eight children developed at-least one episode of culture positive UTI with 3 children having contralateral grade IV VUR and proteinuria. None of them had recurrent UTI, renal failure or scars by DMSA. None of the children showed acute kidney injury during their infection episodes. Among 9 children with proteinuria, 3 had undergone nephrectomy and 3 had VUR. There was significant association between proteinuria and complete involution of MCDK ($P$<0.023). All of them had compensatory hypertrophy of the contralateral kidney. There was no significant association between grades of reflux and proteinuria. There was no significant difference in the

**WHAT THIS STUDY ADDS?**

- Associated anomalies in the contralateral kidney are common in multicystic dysplastic kidneys.

eGFR between proteinuric and non-proteinuric children. Two children with postnatal diagnosis had renal failure. Both had high grade reflux into contralateral kidney with multiple scars on DMSA. The child who underwent ureteric reimplantation showed decreased kidney size on follow-up.

## DISCUSSION

MCDK is increasingly being recognized due to routine antenatal ultrasonographic screening [6]. Our retrospective analysis showed that antenatal diagnosis was made in majority of children. Associated renal anomalies were seen in one-third of patients, and contralateral VUR was seen in one-fourth. The proteinuria in completely involuted MCDK was probably due to hyperfiltration. The usual course in any single kidney status is progressive hyperfiltration, glomerulosclerosis and decrease in renal function [7]. Hence, these children need to be under close monitoring.

Singh, *et al*. [8] described predominant postnatal diagnosis of MCDK. Rabelo, *et al*. [9], suggested a median time of 122 months for the MCDK to become undetectable by USG, which is similar to our study. The overall involution rate in our study was lesser than that reported by Kessler, *et al*. [10]. We found significant difference in the involution rates at the 6-year follow-up between MCDK with an initial size of ≥5 cm *versus* MCDK with an initial size of ≤5 cm, similar to the finding of study by Hayes, *et al*. [11]. The rate of associated abnormalities of the contralateral kidney has been reported to be 7-43% [12]. According to Mansoor, *et al*. [2], children with contralateral anomalies are at risk for developing decreased renal function. Hence, early diagnosis and treatment of contralateral anomalies to prevent renal injury is necessary. Seeman, *et al*. [13] suggested that the main risk factor for developing hypertension is contralateral kidney damage. In our study, even though the contralateral renal anomalies were high, no child developed persistent hypertension.

In conclusion, long term nephrological follow up is needed in children with MCDK. As renal functions depend on the contralateral functioning kidney, its anomalies need to be detected early and managed appropriately.

*Contributors*: SS, SE, MVK: formulated the manuscript; MVK, NP: were in-charge of the patients; MVK: will act as a guarantor.
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