

# **Multicystic Dysplastic Kidney (MCDK) in the Neonate: The role of the Urologist**

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**Conflict of Interest of Author: None**

**I. Background<sup>1</sup>:** The urological management of multicystic dysplastic kidneys (MCDK) in the paediatric patient is controversial. Historically, MCDK was rare, presenting with a palpable mass or symptoms, and was managed with open nephrectomy. Wilms' Tumour was listed on the differential diagnosis. Introduction of antenatal ultrasound (US) transformed MCDK into a common condition which is generally asymptomatic at presentation. Observation through a MCDK registry established that the low risk of WT developing in these kidneys did not justify nephrectomy, and that observation with periodic ultrasound was safe. However, a cost-effective protocol for follow-up was never established. With widespread adoption of paediatric laparoscopy, there is potential for the pendulum to swing back to surgical management of this condition. However, the current indications cited for surgical intervention are more to relieve the iatrogenic symptoms caused by long-term observation, those being parental anxiety and cost to the health care system, than for any confirmed medical indication.

With evidence-based medicine being the new standard, a literature review was carried out to determine whether perceived medical concerns regarding MCDK could be substantiated, and to determine what degree of urological investigation and observation was actually necessary for the neonate with a MCDK. The findings of this literature search are detailed in the article: Psooy K. Long-term urological follow-up of MCDKs: Is it still indicated in 2007? *Can Urol Assoc J* 2007;1(2):113 and are summarized below. A separate literature search was carried out regarding vesico-ureteral reflux.

## **II. Literature Review:**

Databases: Embase, Pubmed and Papers Conference Index.

Key word search: "multicystic kidney" and "multicystic dysplastic kidney" paired with "Wilms' tumor", "hypertension" "vesicoureteral reflux" and "natural history"

The titles of all related articles were reviewed on all key articles. Abstracts were read if the titles were pertinent. English articles were read if pertinent to the questions to be addressed in the review.

## **III. Medical conditions perceived to be associated with MCDK:**

1. Wilms' Tumour
2. Hypertension
3. Chronic renal insufficiency/End-stage renal disease
4. Urinary tract infection & Vesicoureteric Reflux

## 1. Wilms' Tumour (WT)

- a. Between 1983-1998 there were 5 cases of WT associated with a MCDK in the United States,<sup>2-4</sup> resulting in an estimated risk of 0.03%-0.1%<sup>2,3</sup>
- b. There have been no published case reports since 1997<sup>1</sup>
- c. A review of all published cohort studies of MCDK from 1986-2004 suggests the risk of WT developing in MCDK is nil<sup>5</sup>
- d. A United Kingdom consensus panel has suggested that renal US surveillance should be offered to children at >5% risk of Wilms' tumour<sup>6</sup>

**Conclusion:** The increased risk of developing WT appears negligible, if not nonexistent, and does not warrant surveillance.

**Level of Evidence: 3**

## 2. Hypertension (HTN)

- a. Case reports suggest MCDK can be associated with HTN
  - i. In some cases, HTN has been cured with nephrectomy of the MCDK, even if the kidney has shown involution on US, suggesting MCDK can be the primary etiology<sup>7</sup>
  - ii. HTN can develop following remote nephrectomy of the MCDK, suggesting an abnormal contralateral kidney may be the etiology<sup>8</sup>
- b. A review of published cohort studies of MCDK suggested the risk developing HTN was no higher than that of the general paediatric population<sup>9</sup>

**Conclusion:** Routine blood pressure monitoring should be performed. If HTN is identified, the possibility exists that nephrectomy may cure the HTN if no other aetiologies are identified

**Level of Evidence: 3**

## 3. Chronic renal insufficiency/End-stage renal disease (CRF/ESRD)

- a. MCDK can be sub-classified into "Simple" and "Complex" MCDK<sup>10</sup>
  - i. "Simple" is defined as: unilateral dysplasia with a normal contralateral kidney with compensatory hypertrophy and no associated genitourinary anomalies detected by US or physical examination
    1. In "simple" MCDK, the risk of CRF or ESRD at 5 years is nil
  - ii. "Complex" is defined as: bilateral dysplasia or abnormalities of the contralateral kidney or genitourinary tract detected by US or physical examination
    1. In "complex" MCDK, the risk of CRF and ESRD at 7 years is 29% and 21% respectively
- b. Children with a solitary functioning kidney of any aetiology have a small increased risk of proteinuria and renal insufficiency in adulthood<sup>11,12</sup>

**Conclusions:** The contralateral kidney in those with “simple” MCDK does not warrant urological follow-up. The contralateral kidney in those with “complex” MCDK warrants urological and/or nephrological follow-up depending on the associated abnormalities identified.

**Level of Evidence: 3**

Children with a normal solitary functioning kidney have a small risk of future renal insufficiency.

**Level of Evidence: 3**

#### **4. Vesicoureteral reflux (VUR) & Urinary tract infection (UTI)**

- a. Using only published reports of referred and live birth populations with MCDK where >90% of the patients with unilateral MCDK had a voiding cystogram, the percent with contralateral VUR ranges from 4.5-28% (weighted mean = 16%, total N = 889)<sup>8,13-28</sup>
  - i. In select studies<sup>25-29</sup>, the presence of contralateral renal abnormalities (including dilatation of the collecting system, ectopia and agenesis, but excluding the absence of compensatory hypertrophy) documented on renal US conferred a risk of contralateral VUR of 21.829 times more than those without (Chi squared = 56.705 with 1 df, p<0.0001)
  - ii. It is worth noting that the often quoted rate of VUR up to 43% from the Multicystic Kidney Registry is prone to selection bias as only 15% of patients in the registry had a voiding cystogram<sup>30</sup>
- b. The risk of UTI in “simple” MCDK over 5 years is 7%<sup>10</sup>
- c. The risk of UTI in “complex” MCDK is 29%<sup>10</sup>

**Conclusions:** The overall incidence of contralateral VUR is higher in those with MCDK than the general population; however, the likeliness of having MCDK associated VUR is significantly higher in those whose ultrasound shows contralateral renal abnormalities, in comparison to those who do not.

**Level of Evidence: 3**

On a continuum, those with “simple” MCDK have the lowest risk of UTI; those with “complex” MCDK are at the highest risk of UTI.

**Level of Evidence: 3**

### **IV. Recommendations:**

#### **The role of the Urologist in MCDK:**

##### **1. Confirm the diagnosis of MCDK is correct (Grade A)**

- a. US criteria are clear and make misdiagnosis of a cystic malignancy unlikely when these criteria are identified by a an ultrasonographer experienced in paediatrics
- b. In indeterminate cases, a renal scan showing lack of function is supportive of a diagnosis of MCDK, but evidence of some function does not necessarily rule it out.

2. **Use clinical judgement to determine if voiding cystogram is indicated (*Grade D*)**
  - a. The decision to perform a voiding cystogram should take into consideration the risks of the child having VUR (lowest in those without contralateral hydronephrosis) or developing UTI and CRF/ESRD (lowest in those with “simple” MCDK and no history of UTI, and highest in those with “complex” MCDK).
    - i. The management of VUR, if diagnosed, is beyond the scope of this guideline
3. **Determine if the MCKD is “Simple” or “Complex” and manage accordingly (*Grade B*)**
  - a. “Complex” MCDK warrants urological and/or nephrological follow-up depending on the associated abnormalities identified.
    - i. Annual blood pressure monitoring should be included in follow-up
  - b. Confirmation of “Simple” MCKD warrants a repeat renal US at 12-24 months to confirm compensatory hypertrophy
    - i. Those with “Simple” MCKD do not warrant further urological follow-up.
    - ii. Discharge should include:
      1. Counselling the parents on sports and the solitary kidney (See CUA Guideline for specific recommendations<sup>31</sup>)
      2. Providing recommendations to the primary care physician regarding:
        - a. Annual blood pressure monitoring
        - b. Monitoring of the solitary functioning kidney
          - i. Amongst Canadian paediatric nephrologists there is local variation in terms of whether children with a solitary functioning kidney should be seen by a nephrologist, and what investigations the child should have as they age in order to monitor for renal insufficiency. A dialogue with the local nephrologist(s) on this topic would allow the urologist to pass on these recommendations, if any, to the primary care physician

## V. References

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