

Multicystic dysplastic kidney (MCDK) in the neonate: The role of the urologist

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Background¹

The urological management of multicystic dysplastic kidneys (MCDK) in the pediatric population is controversial. Historically, MCDK was rare, presenting with a palpable mass or symptoms, and was managed with open nephrectomy. Wilms tumor (WT) was listed in the differential diagnosis. Introduction of antenatal ultrasound (US) transformed unilateral MCDK into a common condition (1/4300 live births),² which is generally asymptomatic at presentation. Observation through a MCDK registry determined and reported in 1993 that the low risk of WT developing in these kidneys did not justify prophylactic nephrectomy and that observation with periodic US was safe, but as of 2007, a cost-effective protocol had not been established. With widespread adoption of pediatric laparoscopy, there was potential for the pendulum to swing back to prophylactic nephrectomy as management for MCDK. However, the indications cited for intervention were more to relieve the iatrogenic symptoms caused by long-term observation, those being parental anxiety and costs to the healthcare system, rather than any confirmed medical indications.

This CUA Guideline was initially developed in 2008. The recommendations were based on a literature review carried out in 2007 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.¹

In 2015, this CUA Guideline was updated following a 2014 literature review to determine whether management controversies continued to exist, to obtain up-to-date information regarding the medical implications of having a MCDK, and to determine the appropriate investigation

and followup for same. Following this literature review, “Renal cell carcinoma and other adult renal malignancies” and “Anomalies of the internal genitalia” were added to the list of “Medical conditions perceived or shown to be associated with MCDK.” Levels of evidence and grades of recommendations are based on the modified Oxford Centre for Evidence-Based Medicine grading system for guideline recommendations.

Literature review

This revision includes the findings of the previous literature review, in addition to the following literature review:

- Databases: Embase, Pubmed and Papers Conference Index, with key word searches:
 - “multicystic kidney” and “multicystic dysplastic kidney” paired with “Wilms tumor,” “hypertension,” “vesicoureteral reflux,” “natural history,” “nephrectomy” (2007–2014)
 - “multicystic kidney” and “multicystic dysplastic kidney” paired with “renal cell carcinoma” and “cancer” (no year restriction)
- Database: Pubmed, with key word search:
 - “multicystic kidney” and “multicystic dysplastic kidney” paired with “genitalia” and “genital anomalies” (no year restriction)

The titles of all identified articles were reviewed. Abstracts were read if titles were pertinent. Abstracts of articles related to pertinent articles were reviewed. English articles were read if pertinent to the questions to be addressed in the update. Relevant articles referenced in the read articles were reviewed for pertinence.

2014 update on management controversies

Since 2008, a nonsurgical approach remains the initial management for MCDK. The main controversy revolves around the appropriate intensity and length of US followup. On

one extreme is the recommendation that only two renal USs be performed, with the second and final one undertaken at one year of age, ordered by the primary care physician, with no need for repeat imaging unless the MCDK has grown.³ On the other end of the spectrum is the recommendation for ongoing follow up with US imaging, starting with a renal US every six months for the first two years and yearly thereafter, until the MCDK completely involutes.⁴ Nephrectomy continues to be considered by some as a suitable treatment option for MCDKs that do not involute or are unlikely to involute. The argument provided is that there is not enough data to draw conclusions regarding the appropriate frequency and duration of US followup.⁵ Based on the described literature review, the only medical condition that could potentially develop in a non-involuted MCDK, which was not previously addressed in the 2008 guideline, is a malignancy in adulthood. Therefore, this topic is newly addressed in this version of the guideline.

New since 2008 is that there has been a significant number of nephrology articles reporting on the risk of hyperfiltration injury in children born with a congenital solitary functioning kidney. The compensatory hypertrophy of the contralateral kidney, previously seen as reassuring for preserved renal function, may reflect glomerular hypertrophy, which can lead to hyperfiltration injury.⁶ These authors advocate for ongoing followup allowing for early identification of those at risk and timely intervention.⁶⁻⁹

Regarding the use of voiding cystourethrograms (VCUGs), the literature suggests some urologists continued to routinely perform them in all children with MCDK until as recently as 2012,¹⁰ with more recent publications advocating for judicious use of VCUG, only being ordered when US shows contralateral anomalies,³ or only ordering a VCUG if the child has a urinary tract infection, regardless of the presence of mild contralateral hydronephrosis.¹¹

Though not included in the initial guideline, it has been shown that 15% of children with MCDK have malformations of the ipsilateral internal genitalia, warranting followup.¹² Therefore, this “medical condition” has been added to this version of the guideline.

Questions remain as to the natural history of MCDK remnants;¹³ however, it could be argued that the majority of those born with a MCDK who are over 35 years of age (born prior to the age of routine antenatal US imaging) still live with the remnants. In spite of this, MCDK is not felt to be a problem identified in adults by urologists or nephrologists, at least not in its recognizable form. When identified in an adult, it is likely to be given the diagnosis of congenital solitary kidney.¹³

Looking at the big picture, an overview of the recent literature gives the impression that as more is learned about the sequelae of MCDK, the condition becomes less urological and more nephrological as the child ages. While recommendations regarding the need for long-term followup

continue to be made, the impression is that in the event that intervention is ultimately required, it is more likely to be under the direction of a nephrologist rather than a urologist. While the role of the urologist may be further limited to the early years, or even months, this early interaction with the parents, which allows opportunity to provide information and direction, has the potential to be a valuable contribution to the child’s future well-being.

Medical conditions perceived or shown to be associated with MCDK

1. Wilms tumor
2. Hypertension
3. Chronic renal insufficiency/end-stage renal disease
4. Urinary tract infection and vesicoureteric reflux
5. Renal cell carcinoma and other adult renal malignancies (*new*)
6. Anomalies of the internal genitalia (*new*)

Evidence

1. Wilms tumor (WT)

- a. Between 1983 and 1998, there were five cases of WT associated with a MCDK in the U.S.,¹⁴⁻¹⁶ resulting in an estimated risk of 0.03–0.1%^{14,15}
- b. There were no published reported cases from 1997–2007¹
- c. A review of all published cohort studies of MCDK from 1986–2004 suggested the risk of WT developing in MCDK is nil¹⁷
- d. A U.K. consensus panel suggested that renal US surveillance should be offered to children at >5% risk of WT¹⁸
- e. **2014 update**
 - I. There are no new reported cases of WT developing in MCDK since 2007
 - II. Although no longer considered a WT, there is a case report of a malignant rhabdoid tumour diagnosed in a five-year-old girl with a postnatal diagnosis of an ipsilateral MCDK. No imaging was performed after the initial diagnosis at birth and prior to the presentation of a “huge” palpable abdominal mass at age five years. No long-term followup is provided¹⁹

Conclusion

The increased risk of developing WT appears negligible, if not nonexistent, and does not warrant surveillance (*no change*)

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 by nephrectomy of the MCDK, even if the kidney has shown involution on US, suggesting MCDK can be the primary etiology²⁰
 - II. HTN can develop following remote nephrectomy for MCDK, suggesting an abnormal contralateral kidney may be the etiology²¹
- b. A 2005 review of published cohort studies of MCDK suggested the risk of developing HTN was no higher than that of the general pediatric population²²
- c. **2014 update**
 - I. Cases of HTN cured by nephrectomy of a MCDK continue to be published^{23,24}
 - II. There are no recent publications that specifically address the question of whether children with MCDK are at higher risk for developing HTN
 - III. Retrospective⁸ and prospective⁹ studies show that HTN, which can result from hyperfiltration injury, is significantly higher in children with a radiographically normal congenital solitary kidney (which includes those with MCDK) in comparison to normal two-kidney controls

Conclusion:

Routine blood pressure monitoring assessing for HTN should be performed on children with MCKD, and if identified, appropriately managed. If HTN is identified, the possibility exists that nephrectomy may cure the HTN if no other etiologies are identified (*no change*)

Level of evidence: 3

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

- a. MCDK can be subclassified into "Simple" and "Complex" MCDK²⁵
 - 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 five 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 seven years is 29% and 21%, respectively

- b. Children with a solitary functioning kidney of any etiology have a small increased risk of proteinuria and renal insufficiency in adulthood^{26,27}

c. 2014 update

- I. A 2008 study in the pediatric nephrology literature confirmed the findings that "simple" MCDK was not associated with renal insufficiency and suggested that once compensatory hypertrophy was confirmed, screening for proteinuria and HTN could be continued by the general pediatrician²⁸
- II. More recently, pediatric nephrology articles are reporting concerns regarding the increased risk of hyperfiltration injury, represented by HTN and proteinuria, in spite of compensatory hypertrophy. The resultant recommendation is that there be systematic monitoring of blood pressure, urine for proteinuria, and renal function in all children with a MCDK or other etiology for a congenital solitary kidney, in order to identify those with markers of renal injury. No specific followup regimens are described with respect to time intervals, specific tests performed, or whether such monitoring is best performed by a nephrologist^{6-9,29}
- III. Sporting activities can result in significant renal trauma and loss of function.³⁰ Parents should be counselled on these risks in order to make informed decisions regarding their children's sporting activities.³¹ It is recommended that the Canadian Urological Association's Guideline on "Sports and the solitary kidney: What parents of a young child with a solitary kidney should know"³¹ be followed

Conclusions:

The contralateral kidney in those with "simple" MCDK does not warrant urological followup. The contralateral kidney in those with "complex" MCDK warrants urological and/or nephrological followup, depending on the associated abnormalities identified

Level of evidence: 3

Children with a normal solitary functioning kidney, with evidence of compensatory hypertrophy, have a small risk of future renal insufficiency (*no change*). However, the increased risk of hyperfiltration injury, which may be marked by HTN and proteinuria, has led to the recommendation that even these children warrant long-term systemic followup of this nephrological issue (*new*)

Level of evidence: 3

Parents of children with a solitary functioning kidney should be counselled on the issue of sports and the solitary kidney (*new*)

Level of evidence: 4

4. Vesicoureteral reflux (VUR) and 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)¹
 - I. In select studies, 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 relative risk of contralateral VUR to be 21.829 compared to those without (Chi squared=56.705 with 1 df, p<0.0001)¹
- b. The risk of UTI in “simple” MCDK over five years is 7%²⁵
- c. The risk of UTI in “complex” MCDK over five years is 29%²⁵
- d. **2014 update**
 - I. The previously reported rate of contralateral VUR in MCDK remains valid^{32,33}
 - II. Contralateral hydronephrosis alone, when all other significant congenital anomalies and other genito-urinary anomalies are excluded, is not predictive of VUR³³

Conclusions:

The overall incidence of contralateral VUR is higher in those with MCDK than the general population (*no change*). The likelihood of having MCDK associated contralateral VUR is significantly higher in those whose US shows contralateral renal abnormalities when compared to those whose do not (*no change*); however, when the only contralateral anomaly is hydronephrosis, the likelihood of having MCDK associated VUR is not significantly different from those with a completely normal contralateral kidney (*new*).

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 (*no change*)

Level of evidence: 3

5. Renal cell carcinoma (RCC) and other adult renal malignancies (*new*)

- a. As of a 2003 review of renal neoplasms associated with cystic renal diseases, there were five reported RCCs and one “mesothelioma” that may have been a sarcomatoid RCC, which were felt to develop from

a MCDK. Age of presentation was 15–68 years and all were metastatic and lethal. The conclusion of the review was that “the current data suggest that kidneys with MCDK are *not* predisposed to the development of RCC.”³⁴ Beckwith since commented that the burden of proof to confirm a primary MCDK was not met for the majority of these six cases.¹⁵ Also mentioned in the review is a case of transitional cell carcinoma in a 63-year-old with flank pain, weight loss, and microhematuria. Pathology suggested the surrounding tissue to be consistent with a MCDK. No information regarding previous urological history or followup is provided.³⁵ The review authors felt the association probably occurred by chance³⁶

b. Since 2003:

- I. There is a 2006 case report of a collecting duct RCC developing in a 19-year-old with a diagnosis of congenital absence of the ipsilateral kidney made prior to six months of age. She presented as an adult with flank pain and fever. Tissue surrounding the tumour was a rim of compressed benign cystic dysplastic renal parenchyma. At 11 months postoperatively, the woman had no evidence of disease³⁷
- II. There is a 2008 case report of a metastatic clear cell RCC developing in a 34-year-old male with a history of ipsilateral absent kidney diagnosed by US several years prior. He presented generally unwell, with a cough and thoracic pain. Lung metastases were identified. Search for a primary focus identified a pelvic mass. Tissue surrounding the tumour had dysplastic features, including primitive ducts. Final diagnosis was RCC developing in a congenital ectopic MCDK³⁷
- III. There is a 2008 case report of a primary angiosarcoma arising from a multicystic kidney in a 68-year-old who presented with flank pain. No previous abdominal imaging is reported and the information provided does not make a convincing argument that the kidney was a MCDK³⁸
- IV. Cambio, in 2008, calculated that if the risk of RCC developing in a MCDK was equal to that of normal kidneys, there would be 10 cases of RCC in a MCDK per year in the USA. Given that at the time, there were only six cases reported in the English literature, the conclusion was that the risk of RCC developing in MCDK was equal to or less than that developing in normal kidneys³⁹

Conclusion:

Based on the literature, there is potential for an adult malignancy to develop in a MCDK. This risk appears to be very low considering the majority of persons currently over the age of 35 years born with a MCDK did not have it removed. There is no evidence to suggest that this risk is higher in MCDKs that have failed to undergo radiographic involution (*new*)

Level of evidence: 4

6. Anomalies of the internal genitalia (new)

- a. The coexistence of renal and reproductive duct anomalies is well established. Schlegel et al found that 26% of men with unilateral congenital absence of the vas deferens (CAVD) and 11% of men with bilateral CAVD had an absent ipsilateral kidney.⁴⁰ Similarly, uterus didelphys and obstructed hemivagina have an increased association with ipsilateral renal agenesis⁴¹
- b. More recently, the association with MCDK and anomalies of the internal genitalia has been identified.^{42,43} Merrot identified ipsilateral malformation of the internal genitalia in 15% of MCDK patients,¹² which has the potential to lead to genitourinary complaints⁴⁴
- c. The known tendency for MCDKs to involute, the low rate of prenatally diagnosed renal agenesis, and the prospective observation of children with MCDK having anomalies of the internal genitalia that persist in spite of MCDK involution, has led to the conclusion that the congenital anomalies previously identified in those with “congenital renal agenesis” extends to those with a history of MCKD^{42,43,45}
- d. Screening for female anomalies of the internal genitalia is felt to be beneficial, as the management of female anomalies of the internal genitalia at the onset of puberty may prevent the symptoms of acute abdominal pain and dysmenorrhoea.⁴³ Opportunities for diagnosis in the asymptomatic female include an early pelvic US in the neonate with the assistance of vaginal infusion of saline.⁴³ Alternatively, and less invasively, diagnosis may be obtained later, with a pelvic US done upon reaching the stage of advancing thelarche. [Dr. Jenna McNaught, Department of Obstetrics & Gynecology, University of Manitoba, personal communication, November 21, 2014 & Dr. Tarek Motan, Department of Obstetrics & Gynecology, University of Alberta, personal communication December 8, 2014]
- e. The role of screening for male anomalies of the genital ducts is unclear. These conditions alone, in the absence of symptoms, do not require intervention.⁴⁴ Additionally, the true risk of developing infertility or becoming symptomatic from such anomalies is

unknown, with the exception of bilateral CAVD, which may be diagnosed on physical examination as a teenager. Therefore, routine US screening for such anomalies in boys is unlikely to provide patient benefit, and if identified, almost certainly will result in parental anxiety [Dr. Ross MacMahon, Section of Urology, Department of Surgery, University of Manitoba, personal communication December 14, 2014]

Conclusions:

Any recommendations that refer to the assessment of the internal genitalia in patients with a congenital solitary kidney, should be applied to patients with a history of a MCDK (*new*)

Level of evidence: 3

Females with MCDK should have a screening pelvic US following advanced puberty (*new*)

Level of evidence: 4

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 an ultrasonographer experienced in pediatrics⁴⁶
 - I. Those inexperienced with recognizing the US features of MCDK may misdiagnose it for severe ureteropelvic junction obstruction. This error could result in loss of renal function that might have otherwise been salvageable
- b. In indeterminate cases, renal scan showing lack of function is supportive of a diagnosis of MCDK, but evidence of some function does not necessarily rule it out.⁴⁷ (*no change*). Magnetic resonance urography may also provide this information⁴⁸

2. Use of clinical judgement to determine if VCUG is indicated (Grade D)

- a. The decision to perform a VCUG should take into consideration the risks of the child having VUR and developing a UTI (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. However, if the urologist has no intent to intervene on any identified VUR in the absence of symptoms, it would seem appropriate to defer the investigation of a VCUG until such symptoms occur

3. Determine if MCDK is “simple” or “complex” and manage accordingly (Grade B)

- a. “Complex” MCDK warrants urological and/or nephrological followup, depending on the associated abnormalities identified
 - I. Annual blood pressure monitoring and screening for proteinuria should be included in followup
- b. Confirmation of “simple” MCDK warrants a repeat US at 12–24 months to confirm compensatory hypertrophy
 - I. Those with “simple” MCDK do not warrant further urological followup (Grade B)
 - II. Discharge from urological followup should include:
 1. Determining locally if the child should have ongoing followup by pediatric nephrology or their primary care physician (PCP) for blood pressure monitoring and signs of hyperfiltration (proteinuria and decreased renal function) (*new*) (Grade C)
 - a. Among Canadian pediatric nephrologists there is local variation in terms of whether these children should be followed by a pediatric nephrologist or a PCP and what that followup entails. A dialogue with the pediatric nephrologist(s) who serve(s) the urologist’s community would determine whether the urologist should redirect these children to their PCP with the appropriate recommendations, or refer them to the pediatric nephrologist
 2. Recommendation of a screening pelvic US for females following advanced puberty, with referral to a gynecologist if anomalies are identified (*new*) (Grade C)
 3. Counselling the parents on sports and the solitary kidney (*new*) (Grade C)
 - a. See CUA Guideline³¹ for specific recommendations

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