

Multicystic dysplastic kidney in the neonate: the role of the urologist

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Background

The urological management of multicystic dysplastic kidneys (MCDK) in the pediatric 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 transformed MCDK into a common condition which is generally asymptomatic at presentation. Observation through a MCDK registry established that the low risk of Wilms' tumour 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 the widespread adoption of pediatric 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, such as parental anxiety and cost to the health-care system, caused by long-term observation 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 were actually necessary for the neonate with a MCDK. The findings of this literature search are detailed in a 2007 article on the long-term urological follow-up of MCDKs¹ and summarized below. A separate literature search was carried out regarding vesicoureteral reflux.

Literature review

The following databases were reviewed: Embase, PubMed and Papers Conference Index. Key word searches were: "multicystic kidney" and "multicystic dysplastic kidney" paired with "Wilms' tumour", "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.

Medical conditions perceived to be associated with MCDK

Wilms' tumour

1. Between 1983 and 1998 there were 5 cases of Wilms' tumour associated with a MCDK in the United States,²⁻⁴ resulting in an estimated risk of 0.03% to 0.1%.^{2,3}
2. There have been no published case reports since 1997.¹
3. A review of all published cohort studies of MCDK from 1986 to 2004 suggests the risk of Wilms' tumour developing in MCDK is nil.⁵
4. A United Kingdom consensus panel has suggested that renal ultrasound surveillance should be offered to children at greater than 5% risk of Wilms' tumour.⁶

Conclusion: The increased risk of developing Wilms' tumour appears negligible, if not nonexistent, and does not warrant surveillance (**Level 3 evidence**).

Hypertension

1. Case reports suggest MCDK can be associated with hypertension.
 - a. In some cases, hypertension has been cured with nephrectomy of the MCDK, even if the kidney has shown involution on ultrasound, suggesting MCDK can be the primary etiology.⁷
 - b. Hypertension can develop following remote nephrectomy of the MCDK, suggesting an abnormal contralateral kidney may be the etiology.⁸
2. A review of published cohort studies of MCDK suggested the risk developing hypertension was no higher than that of the general pediatric population.⁹

Conclusion: Routine blood pressure monitoring should be performed. If hypertension is identified, the possibility exists that nephrectomy may cure the hypertension if no other etiologies are identified (**Level 3 evidence**).

Chronic renal insufficiency/end-stage renal disease

1. Multicystic dysplastic kidneys can be subclassified into “simple” and “complex” MCDK.¹⁰
 - a. “Simple” is defined as unilateral dysplasia with a normal contralateral kidney with compensatory hypertrophy and no associated genitourinary anomalies detected by ultrasound or physical examination. In “simple” MCDK, the risk of chronic renal insufficiency or end-stage renal disease at 5 years is nil.
 - b. “Complex” is defined as bilateral dysplasia or abnormalities of the contralateral kidney or genitourinary tract detected by ultrasound or physical examination. In “complex” MCDK, the risk of chronic renal insufficiency or end-stage renal disease at 7 years is 29% and 21%, respectively.
2. Children with a solitary functioning kidney of any etiology have a small increased risk of proteinuria and renal insufficiency in adulthood.^{11,12}

Conclusion: 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 3 evidence**). Children with a normal solitary functioning kidney have a small risk of future renal insufficiency (**Level 3 evidence**).

Vesicoureteral reflux and urinary tract infection

1. Using only published reports of referred and live birth populations with MCDK where more than 90% of the patients with unilateral MCDK had a voiding cystogram, the percent with contralateral vesicoureteral reflux ranges from 4.5% to 28% (weighted mean 16%, total n = 889).^{8,13-28}
 - a. In select studies,²⁵⁻²⁹ 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 ultrasound conferred a risk of contralateral vesicoureteral reflux of 21.829 times more than those without (Chi-squared 56.705 with 1 degree of freedom, $p < 0.0001$).
 - b. It is worth noting that the often-quoted rate of vesicoureteral reflux 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.³⁰
2. The risk of urinary tract infection in “simple” MCDK over 5 years is 7%.¹⁰
3. The risk of urinary tract infection in “complex” MCDK is 29%.¹⁰

Conclusion: The overall incidence of contralateral vesicoureteral reflux is higher in those with MCDK than the general population; however, the likeliness of having MCDK associated vesicoureteral reflux is significantly higher in those whose ultrasound shows contralateral renal abnormalities, in comparison to those who do not (**Level 3 evidence**). On a continuum, those with “simple” MCDK have the lowest risk of urinary tract infection; those with “complex” MCDK are at the highest risk of urinary tract infection (**Level 3 evidence**).

Recommendations

The role of the urologist in MCDK is to:

Confirm the diagnosis of MCDK is correct (*Grade A recommendation*)

1. Ultrasound criteria are clear and make misdiagnosis of a cystic malignancy unlikely when these criteria are identified by an ultrasonographer experienced in pediatrics.
2. 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.

Use clinical judgement to determine if voiding cystogram is indicated (*Grade D recommendation*)

1. The decision to perform a voiding cystogram should take into consideration the risks of the child having vesicoureteral reflux (lowest in those without contralateral hydronephrosis) or developing urinary tract infection and chronic renal insufficiency/end-stage renal disease (lowest in those with “simple” MCDK and no history of urinary tract infection, and highest in those with “complex” MCDK).
 - a. The management of vesicoureteral reflux, if diagnosed, is beyond the scope of this guideline.

Determine if the MCKD is “simple” or “complex” and manage accordingly (*Grade B recommendation*)

1. “Complex” MCDK warrants urological and/or nephrological follow-up depending on the associated abnormalities identified.
 - a. Annual blood pressure monitoring should be included in the follow-up.
2. Confirmation of “simple” MCKD warrants a repeat renal ultrasound at 12 to 24 months to confirm compensatory hypertrophy.
 - a. Those with “simple” MCDK do not warrant further urological follow-up.
 - b. Discharge should include counselling the parents on sports and the solitary kidney (see CUA Guideline

for specific recommendations³¹); and providing recommendations to the primary care physician regarding annual blood pressure monitoring and monitoring of the solitary functioning kidney.

Amongst Canadian pediatric 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 on this topic would allow the urologist to pass on these recommendations, if any, to the primary care physician.

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References

1. Psooy K. Long-term urological follow-up of multicystic dysplastic kidneys: Is it still indicated in 2007? *Can Urol Assoc J* 2007;1:113-6.
2. Homsy YL, Anderson JH, Oudjhane K, et al. Wilms tumour and multicystic dysplastic kidney disease. *J Urol* 1997;158:2256-9.
3. Beckwith JB. Editorial comment. *J Urol* 1997;158:2259-60.
4. Pérez LM, Naidu SI, Joseph DB. Outcome and cost analysis of operative versus nonoperative management of neonatal multicystic dysplastic kidneys. *J Urol* 1998;160:1207-11.
5. Narchi H. Risk of Wilms' tumour with multicystic kidney disease: a systematic review. *Arch Dis Child* 2005;90:147-9.
6. Scott RH, Walker L, Olsen ØE, et al. Surveillance for Wilms tumour in at-risk children: pragmatic recommendations for best practice. *Arch Dis Child* 2006;91:995-9.
7. Webb NJA, Lewis MA, Bruce J, et al. Unilateral multicystic dysplastic kidney: the case for nephrectomy. *Arch Dis Child* 1997;76:31-4.
8. Kuwertz-Broeking E, Brinkmann OA, Von Lengerke H-J, et al. Unilateral multicystic dysplastic kidney: experience in children. *BJU Int* 2004;93:388-92.
9. Narchi H. Risk of hypertension with multicystic kidney disease: a systematic review. *Arch Dis Child* 2005;90(9):921-924.
10. Feldenberg LR, Siegel NJ. Clinical course and outcome for children with multicystic dysplastic kidneys. *Pediatr Nephrol* 2000;14:1098-101.
11. Argueso LR, Ritchie ML, Boyle ET Jr, et al. Prognosis of patients with unilateral renal agenesis. *Pediatr Nephrol* 1992;6:412-6.
12. Argueso LR, Ritchie ML, Boyle ET Jr, et al. Prognosis of children with solitary kidney after unilateral nephrectomy. *J Urol* 1992;148:747-51.
13. Kaneyama K, Yamataka A, Satake S, et al. Associated urologic anomalies in children with solitary kidney. *J Ped Surg* 2004;39:85-7.
14. Eckoldt F, Woderich R, Wolke S, et al. Follow-up of unilateral multicystic kidney dysplasia after prenatal diagnosis. *J Matern Fetal Neonatal Med.* 2003;14:177-86.
15. Flack CE, Bellinger MF. The multicystic dysplastic kidney and contralateral vesicoureteral reflux: protection of the solitary kidney. *J Urol* 1993;150:1873-4.
16. Guarino N, Grazia M, Casamassima S, et al. Natural history of vesicoureteral reflux associated with kidney anomalies. *Urology* 2005;65:1208-11.
17. Onal B, Kogan BA. Natural history of patients with multicystic dysplastic kidney – what followup is needed? *J Urol* 2006;176(4 Pt 1):1607-11.
18. Al-Khaldi N, Watson AR, Zuccollo J, et al. Outcome of antenatally detected cystic dysplastic kidney disease. *Arch Dis Child* 1994;70:520-2.
19. Miller DC, Rumohr JA, Dunn RL, et al. What is the fate of the refluxing contralateral kidney in children with multicystic dysplastic kidney? *J Urol* 2004;172:1630-4.
20. Selzman AA, Elder JS. Contralateral vesicoureteral reflux in children with a multicystic kidney. *J Urol* 1995;153:1252-4.
21. Rottenberg GT, Gordon I, DeBruyn R. The natural history of the multicystic dysplastic kidney in children. *Br J Radiol* 1997;70:347-50.
22. Damen-Elias HAM, Stoutenbeek PH, Visser GHA, et al. Concomitant anomalies in 100 children with unilateral multicystic kidney. *Ultrasound Obstet Gynecol* 2005;25:384-8.
23. Rabelo EAS, Oliveira EA, Diniz JSS, et al. Natural history of multicystic kidney conservatively managed: a prospective study. *Pediatr Nephrol* 2004;19:1102-7.
24. Alconcher L, Tombesi M. Multicystic dysplastic kidney detected by prenatal ultrasonography: conservative management. *Pediatr Nephrol* 2005;20:1024-5.
25. Ismaili K, Avni FE, Alexander M, et al. Routine voiding cystourethrography is of no value in neonates with unilateral multicystic dysplastic kidney. *J Pediatr* 2005;146:759-63.
26. Fanos V, Sinaguglia G, Vino L, et al. Multicystic dysplastic kidney and contralateral vesicoureteral reflux: renal growth. *Minerva Pediatr* 2001;53:95-8.
27. Krzemie G, Roszkowska-Blaim M, Kastro I, et al. Urological anomalies in children with renal agenesis or multicystic dysplastic kidney. *J Appl Genet* 2006;47:171-6.
28. Van Eijk L, Cohen-Overbeek TE, Den Hollander NS, et al. Unilateral multicystic dysplastic kidney: a combined pre- and postnatal assessment. *Ultrasound Obstet Gynecol* 2002;19:180-3.
29. Zerin JM, Leiser J. The impact of vesicoureteral reflux on contralateral renal length in infants with multicystic dysplastic kidney. *Pediatr Radiol* 1998;28:683-6.
30. Wacksman J, Phipps L. Report of the multicystic kidney registry: preliminary findings. *J Urol* 1993;150:1870-2.
31. Psooy K. Sports and the solitary kidney: what parents of a young child with a solitary kidney should know. *Can Urol Assoc J* 2009;3:67-8.

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