MCDK not excluded by virtue of function on renal scan

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Introduction: *The conservative management of multicystic dysplastic kidneys (MCDK) has been very successful, largely due to advances in diagnostic imaging. Classically, MCDK is described as a non-functioning cystic renal mass. However, we noticed that the increasing sensitivity of renal scans is able to demonstrate function in MCDK that previously would not have been detected.*

Methods: *We describe eight cases of MCDK with elements of function on renal scan, and their follow up. Results and conclusions: One half (4/8) of these children underwent nephrectomy, and histology was consistent with MCDK. The remaining 4/8 were successfully followed to radiographic involution, without any complications. Therefore, we believe that minimal function on nuclear scintigraphy does not preclude the diagnosis of MCDK, and is yet another adjustment in our understanding of this entity.*

Key Words: multicystic dysplastic kidney, nuclear medicine, diagnosis, function

Introduction

Multicystic dysplastic kidney (MCDK) is a congenital anomaly that results in an atretic ureter and non-communicating cysts.[1] Prior to the advent of prenatal ultrasonography this entity presented as an abdominal mass, required surgical exploration, and this usually resulted in nephrectomy. However, with the increased use of prenatal ultrasonography, the natural history has become more apparent.[1, 6] Conservative management has become widely accepted as the initial fears of infection, pain, and malignancy are proving to be rare.[2] The condition is usually detected with a prenatal ultrasound, and confirmed postnatally with a radioisotope scan that demonstrates non-function.

Classical teaching dictated that a MCDK was a non-functioning entity; any function implied another lesion, such as severe UPJ obstruction, or cystic nephroma.[3] However, a review of MCDK at our institution shows that some function demonstrable with scintigraphy may exist without compromising the diagnosis. A 10-year review demonstrated that 16% of patients eventually shown to have a MCDK demonstrated some function of the kidney on radioisotope imaging.[4] We feel this is a result of the

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increased sensitivity of the scintigraphy, and should not compromise the ultrasonographic diagnosis.

Methods

We undertook a 10-year retrospective review of all cases of MCDK at our tertiary care referral centre. Follow up was a minimum of 1 year. Cases were examined for demographic information, incidence of prenatal detection, postnatal investigations, and clinical outcomes.

Results

Fifty-four cases were discovered, with slightly more being male (59%) and on the left (56%); 96% were detected ante-natally, and 93% referred to a pediatric urologist. Twenty-eight (54%) were correctly identified as MCDK by prenatal ultrasonography. Other diagnoses suggested on the basis of the initial ultrasound included: 15 (29%) hydronephrosis, 7 (13%) as cysts, and 4 (8%) with an unknown mass.

Postnatal investigations were obtained for all children in the study. All 54 children had ultrasonography; the average number of studies was 3, with an average follow up period of 28 months. Fifty cases (93%) had radioisotope scans. Dimercaptosuccinic acid (DMSA) was the most common imaging agent, used for 20/50 scans (40%). Eighteen (36%) of patients had glucoheptanate, 10 (20%) DPTA, and 2(4%) had MAG-3 scintigraphy.

Eight (16%) of the scans demonstrated activity in the renal unit. Three (37.5%) were DMSA, 3 (37.5%) glucoheptanate, 1 (12.5%) was MAG-3, and 1 (12.5%) used DPTA. Of these, four were observed with serial ultrasonography and four underwent nephrectomy.

Table 1a gives basic data on the four patients who were followed ultrasonographically. Three (75%) were identified as MCDK prenatally, and all four were postnatally confirmed as MCDK by virtue of ultrasound findings. The average number of ultrasound exams was 2.5, with a range of one to four studies. Two of the kidneys demonstrated complete involution ultrasonographically, while two showed no function on repeat scintigraphy. No complications had occurred during a minimum of a 3-year follow-up.

Table 1b gives information about the four patients who underwent nephrectomy. The indications for surgery were hypertension (n=2), and radiologic question of diagnosis (n=2). Post-operatively, the hypertension resolved, and no complications were observed in any of the four patients. Gross and microscopic examination

TABLE 1. **Eight patients showed a degree of ipsilateral function at scintigraphy**

TABLE 1a. **Involution. Four of the patients' kidneys were safely observed to involution**

Case #	Pre natal u/s Dx	Post natal u/s Dx	Clinical note
1	MCDK	MCDK	horseshoe kidney
2	MCDK	MCDK	no function on repeat at 4 months of age
3	MCDK	MCDK	
4	hydronephrosis	MCDK	VATER assoc.

TABLE 1b. **Nephrectomy. Four patients underwent nephrectomy. All four specimens were histologically confirmed to be MCDK**

Case #	Pre natal u/s Dx	Age at Nephrectomy	Pathologic diagnosis	Clinical note
5	MCDK	7 months	MCDK	hyper-tension resolved post-op
6	renal cysts	3 months	MCDK	diagnosis uncertain
7	MCDK	9 months	MCDK	diagnosis uncertain
8	MCDK	7 months	MCDK	hyper-tension resolved post-op

confirmed MCDK in all cases; Figure 1 describes one of the operative cases.

Discussion

The success of the conservative management of MCDK is directly related to advances in diagnostic imaging; with improved accuracy and confidence in both antenatal and postnatal imaging.[5] Since the initial exams are usually able to distinguish MCDK from other masses, such as neuroblastoma and Wilms' tumor, diagnostic surgery is now rarely required.[4] Also, the improved accuracy has allowed us to confidently follow these patients and further elucidate the benign natural history of this entity.[3] However, along with the improvements in ultrasonography, nuclear medicine has also become increasingly sensitive. Therefore, a multicystic kidney that may not have shown any function on radioisotope scanning now can have enough uptake to be detected.

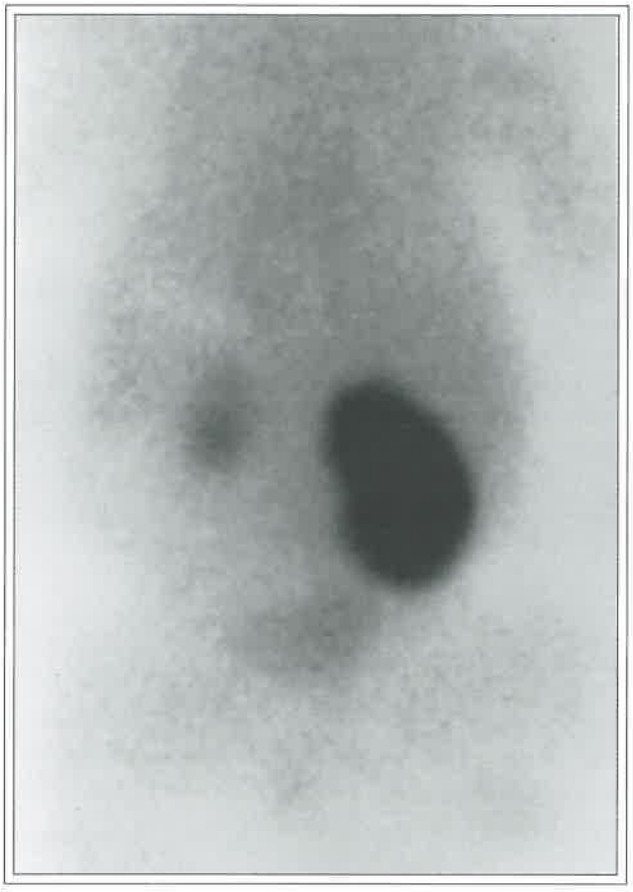

Figure 1a. DMSA scan: normal hypertrophied right kidney and minimal but detectable left renal function.

This improved sensitivity forces us again to reconsider our thinking on this entity, since the original criteria for MCDK necessitated demonstration of an absence of function with IVP or radioisotope imaging.[7]

Our survey included 8/50 (16%) of patients who demonstrated some renal function (isotope uptake) in a dysplastic kidney. This agrees with other recent literature,[9] but much higher than previously believed.

Of the eight kidneys to demonstrate some function,

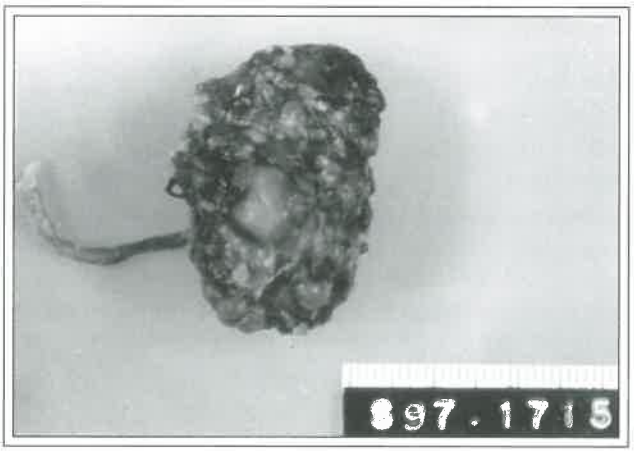

Figure 1b. Gross appearance of nephrectomy specimen showing multiple cysts measuring 0.1-0.7 cm diameter.

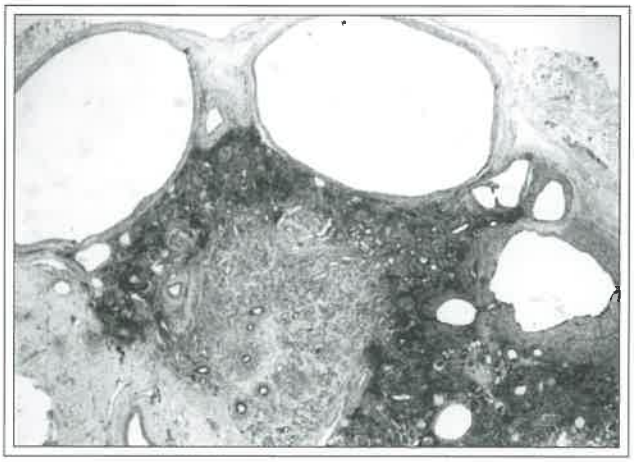

Figure 1c. Histologic section showing renal dysplasia characterized by large and medium-sized epithelial lined cysts surrounded by collars of concentric fibromuscular mesenchyme. Scattered immature glomeruli (best seen in right lower corner) and small primitive ducts surrounded by concentrically orientated nesenchyme (lower middle region). No foci of metaplastic cartilage are seen. Hematoxylin & Eosin, magnification 26x.

Figure 1. Scintigraphy and pathology from a single patient (case #7).

four ultimately involuted by ultrasonographic criteria. We are confident in the diagnosis of the non-surgically managed group, as two were radiologically observed to involute; and the remaining observed to be decreasing in size, and have continued a benign clinical course.

Four were removed surgically for a variety of reasons. However, the original diagnosis was subsequently proven histologically, confirming our hypothesis that MCDK may show miniscule function with current detection methods.

We believe that this realization adds credence to the hypothesis of MCDK evolves secondary to obstructive mechanisms.[1,8] These findings may suggest a continuum with obstructive hydronephrosis, with a non-functioning MCDK at the extreme end of the obstructive picture.

Summary

The conservative management of MCDK has successfully decreased the number of nephrectomies performed in these patients. Diagnostic technology has allowed us to accurately diagnose and monitor the dysplastic kidneys to ensure minimal morbidity. However, the classical description as a non-functioning entity may no longer be valid, as increasingly sensitive scintigraphy may demonstrate function, albeit minimal. We have shown that demonstration of function does not preclude conservative management, and these kidneys can be observed to involution. □

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