

TABLE 1. Prevalence of Contralateral Renal Abnormalities in Patients With MCRD

	No. of Patients		
	VUR	UPJ Obstruction	UVJ Obstruction
Right MCRD (<i>n</i> = 8)	2	2	—
Left MCRD (<i>n</i> = 14)	2	—	—
Left crossed ectopic (<i>n</i> = 1)	—	—	1
Total cases	4	2	1
(%)	(22)	(8.7)	(4.3)

and 1 case of UPJ obstruction were corrected surgically at ages 2 weeks and 5 days, respectively; in the second case of UPJ obstruction, renal scan with Lasix showed good renal function and T^{1/2} (half-life of the wash out of the isotope) of 12 minutes that remained stable on follow-up with no deterioration in renal function. Other associated anomalies are presented in Table 2.

Nephrectomy was performed in 4 patients with no other urologic abnormalities, all diagnosed before 1985. Diagnosis was confirmed by postoperative pathologic study that showed the multiple cysts lined by cuboidal cells surrounded by immature stroma. Primitive tubules and glomerular structures are present as islands of cartilage and fibromuscular tissue. The 19 patients diagnosed after 1985 were treated conservatively. Follow-up ranged from 3 months to 5 years. Complete involution was noted in 14 patients (73.6%) after a mean of 9.6 months (3 to 28 months) (Table 3). Three patients showed no change in kidney size (mean follow-up period of 18 months; range, 8 to 24 months) and still are under observation, and 2 (1 from each group) underwent nephrectomy after 36 and 48 months because of a 1-cm increase along the longitudinal axis of the kidney involved. No hypertension or neoplastic changes were found, and renal function was normal in all.

DISCUSSION

This retrospective study of infants with MCRD yielded several interesting findings. The prevalence rate of MCRD in our hospital during the period of study was 1 in 3310 live births (3/10 000) for the 23 patients who were included in the study. Most of the cases of MCRD (73.6%) were diagnosed by prenatal ultrasound that became available routinely in 1982. A second urologic abnormality was noted in 43.6% of patients; the most common was VUR in the contralateral kidney (22.1% of patients). None of the patients had urinary tract infection. Complete involution occurred after a mean of 9.6 months in 63.1%. Follow-up revealed normal renal function and no hypertension or malignancy in any of the patients.

TABLE 2. Other Anomalies in Patients With MCRD

	No. of Patients
VATER association	1
Undescending testis	3
Tetralogy of Fallot	1
Transposition of great vessels	1
Total	6 (27%)

Today fetal genitourinary anomalies are being detected with increasing frequency with the aid of prenatal ultrasound.⁵⁻⁷ This was true for 115 (47%) of the 245 cases registered at the National Multicystic Kidney Registry.⁸ Indeed, the most common reason for referral of the patients in the present study was a prenatal diagnosis of a renal abnormality.

Several previous studies have documented a variety of contralateral renal anomalies in patients with MCRD. The most significant is bilateral disease, which may be fatal.⁹ Other anomalies, found in 20% to 80% of patients, are UPJ obstruction, horseshoe kidney, segmental ureteral disparity, pyelonephritis, hydroureteronephrosis, renal agenesis, and glomerulonephritis.¹⁰⁻¹⁹

Table 3 outlines some of the findings from previous studies on MCRD. One outstanding feature is the significant incidence (22.1%) of VUR. A similar incidence was found in the present study on voiding cystography during the first month of life. The question of routine use of voiding cystography in MCRD has not yet been resolved. Some authors support this practice,⁹ whereas others such as Gough and associates²⁷ do not, based on the assumption that VUR generally is benign. We found that all of our affected patients had complete resolution of the VUR by age 36 months, and there was no breakthrough infection in those receiving prophylactic antimicrobial treatment. However, because of the high likelihood in MCRD of contralateral VUR, which is life-threatening in the presence of pyelonephritis, routine voiding cystography should be considered. Moreover, the high incidence of VUR in the asymptomatic siblings of children with reflux has made routine screening of this group an accepted practice for many clinicians.²⁸ When performed properly, voiding cystography is a safe procedure. We prefer the contrast media VCUG as the first test to get the best anatomic details. However, follow-up VCUG can be performed with either contrast or isotope, with the latter causing less irradiation. Prophylactic antimicrobial therapy is warranted in children with proven VUR. The presence of a single kidney mandates a more active approach in the form of prophylactic antibiotic treatment to prevent possible renal damage of the single kidney.

Despite reports of associated malignancy and hypertension in MCRD, the trend toward conservative management is gaining momentum because of the possibility of spontaneous involution. Our review of the literature, however, indicated only a 33% rate of complete involution in one study.²⁹ Cumulative data on 380 patients with MCRD revealed that only 189 (49.7%) had ultrasonographic involution (Table 3). In the Multicystic Kidney Registry study of 260 patients with MCRD, who were managed nonoperatively and followed for varying periods up to 5 years, 31% of the kidneys were undetectable by age 3 years and 54% by age 5 years.³ A higher involution rate was noted in our study, 14 of 19 patients (73.6%) who were treated conservatively. This high involution rate is similar to the 75% rate reported by Orejas and colleagues²⁰ in a small group of patients not diagnosed prenatally, but much higher than the rate

TABLE 3. Review of Literature on MCRD

Reference	No. of Patients	Diagnosed Prenatally	Observation Only	Involution	Contralateral VUR*
Deklerk et al (1977) ¹⁹	7				2/7
Dungan et al (1990) ¹⁵	14	14	7	5	
Orejas et al (1992) ²⁰	12		12	9	
Richwood et al (1992) ²¹	44	44	39	20	
Atiyeh et al (1992) ⁹	49				9/49
Kamal (1992) ²²	24	17			0/10
Strife et al (1993) ²³	81	28	48	7	5/44
Flack and Bellinger (1993) ²⁴	29	21			8/29
al Khaldi et al (1994) ²⁵	44	44			20/30
Kaneko et al (1995) ²⁶	7	5	1		3/7
Gough et al (1995) ²⁷	62	62	25	8	12/62
Wacksman et al (1993) ³	441	288	260	140	28/65
Total	814	518	380	189	87/303
Percentage		63	46.6	49.7	29.4
Present study	23	73.6		73.6	22.1

* Reflux was studied only in the patients who underwent voiding cystography.

(49.7%) obtained from all previous studies taken together (Table 3).

In conclusion, all patients with MCRD should be screened for urologic abnormalities of the contralateral kidney. Regarding management, 17% of the our patients underwent nephrectomy compared with 41% in the multicystic kidney registry. Therefore, nephrectomy is an eventual option that should be explained to the parents. Most nephrectomies in the registry were performed at 7 to 12 months of age because there was insufficient parenchyma to allow reconstruction. This age is similar to the mean time to complete involution in our patients (9.6 months).

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