Involution Rate of Multicystic Renal Dysplasia

Oded J. Kessler, MD*; Niza Ziv, MD§; Pinchas M. Livne, MD*; and P. Merlob, MD§

ABSTRACT. Objective. To document the involution rate and long-term results of management of multicystic dysplastic kidney.

Materials and Methods. Data were collected retrospectively for all 23 infants (16 boys) with multicystic dysplastic kidney who were treated at our center over the last 19 years (1977–1995). The diagnosis was based on prenatal ultrasound in 18 patients and on palpable abdominal mass in 5, and confirmed in all patients by postnatal ultrasound and radioisotope scan. Voiding cystography was performed in 18 patients to exclude vesicoureteral reflux. Mean follow-up was 46 months (range, 3 months to 5 years) and included serum creatinine measurements and renal ultrasonography.

Results. Two groups of patients were identified. Ten (43.6%) with other urologic abnormalities (group A) and 13 patients without other urologic abnormalities (group B). Vesicoureteral reflux was observed in 4 patients. Nephrectomy was performed in 4 patients, all from group B. The other 19 patients were treated conservatively. Complete involution was observed in 8 patients in group A and 6 in group B after a mean follow-up period of 9.2 and 10 months, respectively. Two patients, 1 from each group, later underwent nephrectomy not because of no involution but because of an increase in the size of the kidney involved.

Conclusion. Patients with multicystic renal dysplasia have significant associated urologic malformations, and the natural history of the disease is unpredictable. All patients require appropriate investigation of the urinary tract and long-term follow-up. The most outstanding finding of the study is the much higher involution rate of multicystic renal dysplasia and the rate of associated urologic abnormalities than that reported in the literature. Surgery remains an option for those patients in the absence of no involution. Surgery was performed in 18 patients (73.6%) and by palpation of an abdominal mass in 5. All patients underwent postnatal ultrasonography, and in all cases, the diagnosis had been noted on prenatal ultrasound in 18 patients (73.6%) and by palpation of an abdominal mass in 5. All patients underwent postnatal ultrasonography, and in all cases, the diagnosis was confirmed by renal radionuclide scan with technetium-99 m-dimercaptosuccinic acid. Two patients with suspected ureteropelvic junction (UPJ) stenosis also underwent renal scanning with diethylenetriamine-pentaacetic acid and Lasis. Voiding cystography was performed within 1 month of diagnosis in all but 3 patients.

Once vesicoureteral reflux (VUR) was documented and graded according to internationally accepted reflux classification, prophylactic antibacterial therapy was instituted. Serum creatinine level, blood urea nitrogen, electrolytes, and blood pressure were measured, and urine culture was performed. Long-term follow-up was primarily by ultrasonography. Cystography was repeated only in those patients with VUR at 2 and 3 years of age.

RESULTS

Of the 23 children with newly diagnosed MCRD, 16 were boys. Fourteen multicystic kidneys were on the left, eight were on the right, and one was crossed ectopic to the right. Patient age at presentation was birth to 3 months. Five patients presented with flank mass.

Another urologic anomaly was found in 43.6% of the patients, including four (22% of the cohort) with contralateral VUR (grade II/V, 3 patients; grade III/V, 1 patient) (Table 1). The VUR resolved spontaneously in all 4 patients by 24 to 36 months, and all were free of urinary tract infections during follow-up. Among the patients without contralateral VUR, other problems of the contralateral kidney included ureterovesical junction (UVJ) obstruction (1 patient) and UPJ obstruction (2 patients), suspected on initial ultrasonography and confirmed by subsequent radiologic examination (Table 1). The UVJ obstruction has revealed that MCRD is apparently even more prevalent than had been assumed. The association of hypertension, malignancy, and incomplete involution in MCRD mandate long-term follow-up, and much controversy has surrounded the question of whether nephrectomy is indicated routinely on diagnosis. To assess the natural history of this condition and, for purposes of proper management, the associated urinary malformations, we conducted a 19-year retrospective study of all infants with MCRD who had been diagnosed and treated in our tertiary care medical center.

MATERIALS AND METHODS

We reviewed the charts and radiologic records of the 25 patients with MCRD who were diagnosed and treated at our center (Beilinson Campus and later at Schneider Children’s Medical Center of Israel) between 1977 and 1995. Two patients died of neurologic causes in the first week of life. In the present study, we refer to the 23 remaining patients. Nineteen patients were born at our center, and 4 were referred from other hospitals. The malformation had been noted on prenatal ultrasound in 18 patients (73.6%) and by palpation of an abdominal mass in 5. All patients underwent postnatal ultrasonography, and in all cases, the diagnosis was confirmed by renal radionuclide scan with technetium-99 m-dimercaptosuccinic acid. Two patients with suspected ureteropelvic junction (UPJ) stenosis also underwent renal scanning with diethylenetriamine-pentaacetic acid and Lasis. Voiding cystography was performed within 1 month of diagnosis in all but 3 patients.

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ABBREVIATIONS. MCRD, multicystic renal dysplasia; UPJ, ureteropelvic junction; VUR, vesicoureteral reflux; UVJ, ureterovesical junction.

Multicystic renal dysplasia (MCRD) is the most common form of cystic disease of the kidney in childhood. The increasingly widespread use of prenatal diagnostic techniques has revealed that MCRD is apparently even more prevalent than had been assumed. The association of hypertension, malignancy, and incomplete involution in MCRD mandate long-term follow-up, and much controversy has surrounded the question of whether nephrectomy is indicated routinely on diagnosis. To assess the natural history of this condition and, for purposes of proper management, the associated urinary malformations, we conducted a 19-year retrospective study of all infants with MCRD who had been diagnosed and treated in our tertiary care medical center.
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 1.** Prevalence of Contralateral Renal Abnormalities in Patients With MCRD

<table>
<thead>
<tr>
<th>Condition</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>VUR Obstruction</td>
<td>5 (22%)</td>
</tr>
<tr>
<td>UPJ Obstruction</td>
<td>10 (47%)</td>
</tr>
<tr>
<td>UVJ Obstruction</td>
<td>5 (22%)</td>
</tr>
<tr>
<td>Total</td>
<td>20 (91%)</td>
</tr>
</tbody>
</table>

**TABLE 2.** Other Anomalies in Patients With MCRD

<table>
<thead>
<tr>
<th>Condition</th>
<th>No. of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>VATER association</td>
<td>1</td>
</tr>
<tr>
<td>Undescending testis</td>
<td>3</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>1</td>
</tr>
<tr>
<td>Transposition of great vessels</td>
<td>1</td>
</tr>
<tr>
<td>Total</td>
<td>6 (27%)</td>
</tr>
</tbody>
</table>
(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).

REFERENCES

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