# Multicystic Kidney in Siblings

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● **Two siblings (one girl and one boy), with a left multicystic kidney in whom a renal abnormality had been recognized prenatally, are reported. A large renal mass was present in both patients and the second sibling also had hypertension. Early surgical resection was carried out with satisfactory clinical progress and resolution of the hypertension. Multicystic kidney is considered a developmental abnormality with a sporadic incidence. These cases and other reports of familial incidence in the literature indicate that there may also be a genetic basis for the abnormality.**


**INDEX WORDS: Multicystic kidney, siblings.**

MULTICYSTIC KIDNEY is frequently diagnosed after prenatal ultrasonographic (US) recognition.[1] It is considered the most common form of renal cystic disease and represents the most common cause of neonatal abdominal mass.[1-3] It is part of the spectrum of renal dysplastic abnormalities that are considered developmental in origin with a sporadic incidence.[4] However, familial cases of multicystic kidney or renal dysplasia have also been reported, suggesting a genetic basis to the disease.[5,6] We report two siblings, one girl and one boy with a left multicystic kidney. The second sibling also had hypertension, and both were managed by early surgical resection.

## CASE REPORT

A gravida 6, para 5, diabetic woman on insulin, married to a nonconsanguineous man, was the mother of two siblings, born 21 months apart and each having a left multicystic kidney. A fetal renal abnormality had been recognized on prenatal US scan in both babies, but there was no past or family history of renal disease. The first sibling was a girl, the second a boy, and both had a large left renal mass. The second sibling was discovered to have hypertension on routine clinical examination at 20 days of age. The systolic blood pressure was consistently around 110 in the next 10 days and the diastolic pressure was between 65 and 75. Hydralazine, 2.5 mg administered orally twice a day, was effective in controlling the hypertension. The diagnosis of multicystic kidney was based on US scan findings (Fig 1), and renal nonfunction was discovered on isotope scan. Early surgical resection of the multicystic kidney was carried out in both children. Pathological examination of the specimen confirmed the diagnosis of multicystic renal dysplasia, although the gross appearance of the second kidney was atypical (Fig 2). Follow-up was satisfactory with a normal right kidney and normal renal function in both patients. Hydralazine was discontinued in the second patient who has remained normotensive with blood pressure of 91/45.

## DISCUSSION

Multicystic kidney is the most common form of cystic disease in childhood, with an incidence of about 1 in 4,000 live births. The disease has been recognized more frequently since the advent of prenatal US scan but it is possible that there is a real increase in its incidence.[1] Multicystic kidney is part of the spectrum of renal dysplastic abnormalities with a sporadic incidence.[4] However, our experience in two siblings and isolated reports of familial cases in the literature indicate that a genetic factor may also be implicated.[4-6] The overall risk of sibling recurrence is said to be less than 10%, but one known teratogenic factor for renal dysplasia is maternal diabetes.[4]

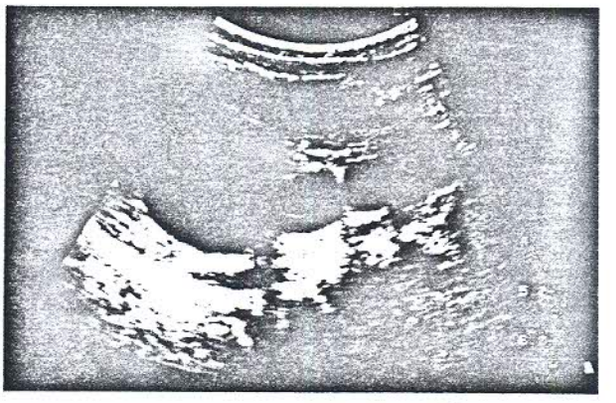

**Fig 1. Renal US scan in the first sibling showing multiple cysts in the left dysplastic kidney.**

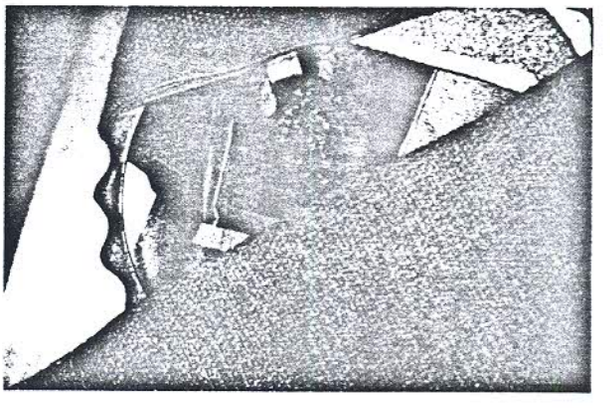

**Fig 2. Intraoperative photograph showing multicystic kidney with an atypical appearance in the second sibling.**

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With regard to surgical management, multicystic kidneys can be treated nonsurgically anticipating spontaneous resolution.[2] However, cyst resolution on surveillance implies absorption of fluid only and renal cellular elements persist. These have the potential for long-term complications including malignancy, hypertension, and infection.[1,3] In contrast, surgical resection of multicystic kidney is a simple procedure with little morbidity and brief hospitalization for 1 to 2 days. Further follow-up of the patient is not necessary provided there are no other nephrological or urologic abnormalities.[1]

We conclude that there is a case for assessment of new siblings of patients in whom multicystic kidney has been diagnosed. This assessment may already have been made by a prenatal US scan failing, in which case a postnatal US scan is justifiable. This is further justified on the basis that a significant number of patients with multicystic kidney also have other nephrourologic problems requiring investigation, long-term observation, or surgery.[1] Whereas some cases of multicystic kidney can be managed nonsurgically, surgical resection is the more definitive treatment.

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