Unilateral multicystic dysplastic kidney: the case for nephrectomy

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Abstract

Management of unilateral multicystic dysplastic kidneys (MCDK) presents physicians and surgeons with a significant dilemma. Recent studies have indicated that the incidence of short term complications of MCDK is low and many authors have recommended conservative non-operative treatment. Surgery has been proposed by some because of the potential complications of hypertension, infection, and malignant change. Three children with hypertension secondary to MCDK seen at this institution in the past four years, one of whom had been discharged from follow up as a result of 'disappearance' of the cystic kidney on ultrasound examination, are reported. We believe that the risks of hypertension secondary to MCDK have been understated, and that based on the conclusions of these studies, many children may be receiving suboptimal follow up. We currently favour elective nephrectomy as the treatment of choice for this lesion.

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Before the widespread use of high resolution antenatal ultrasonography, multicystic dysplastic kidneys (MCDK) were either undetected or presented as abdominal masses in the neonate. Diagnosis was established at laparotomy, with nephrectomy often being performed. Recently, there has been an increase in the number of reported cases, the current incidence being estimated at one in 4300 live births.[1] Most MCDK are now diagnosed by antenatal ultrasound examination with subsequent confirmation by ultrasound assessment and radionuclide imaging in the neonatal period. The increased confidence in ultrasonography as a diagnostic tool has also resulted in a change in the management of MCDK. Recent short-term retrospective and prospective studies have indicated that the incidence of complications of MCDK is low, and with supportive evidence from serial ultrasound examination which suggests that the lesion involutes with time, many authors have recommended conservative non-operative management.[2–6]

Surgery has been proposed by some[8] as the treatment of choice for MCDK because of the potential complications of hypertension,[7] infection,[5] and malignant change to Wilms' tumour,[8] adenocarcinoma,[9] and embryonal tumour.[10]

As histological examination is not possible in patients receiving conservative management, those who advocate such an approach are making the diagnosis from the findings of a non-functioning kidney on dimercaptosuccinic acid (DMSA) scan with a marked cystic appearance on ultrasound examination. This group will include a few kidneys that do not meet the strict pathological criteria for diagnosis of MCDK, but which behave in a similar manner clinically. In this article, we shall consider them together under the diagnostic heading of MCDK.

We report on three children with hypertension secondary to MCDK seen at this institution over the past four years, one of whom had been discharged from follow up as a result of 'disappearance' of their cystic kidney on ultrasound examination.

Case reports

CASE 1

A boy was investigated in the neonatal period after the detection of cystic changes in the left kidney on antenatal scanning at 16 weeks' gestation. A DMSA scan at the referring hospital showed absent function in this kidney and abdominal ultrasonography showed a cystic left kidney with complete absence of any normal renal tissue but an entirely normal right kidney. No vesicoureteric reflux was found in the contralateral system on micturating cystourethrogram (MCUG). At outpatient assessment at 3 months of age, he was noted to be hypertensive with a blood pressure consistently above the 97th centile for age (130/75 mm Hg) and was referred to this institution for further investigation. Echocardiography showed mild global left ventricular hypertrophy (left ventricular posterior wall diameter 9 (normal range 3.8+/-0.8) mm and interventricular septum diameter 9 (3.8+/-0.8) mm) with an otherwise structurally normal heart, indicative of hypertension of some duration. His resting plasma renin activity was raised at 4.0 (normal range 0.5–2.7) ng/ml/hour.

Nephrectomy was performed at 6 months of age. Surgery was uneventful and he was discharged the following day. Histological examination of the kidney confirmed multicystic dysplasia with an atretic ureter.

At follow up he is well and normotensive. Recent echocardiography shows complete resolution of the left ventricular hypertrophy.

CASE 2

A boy was investigated in the neonatal period after the detection of cystic changes in the left kidney on antenatal scanning. Ultrasonography and a DMSA scan performed postnatally at the referring hospital showed a multicystic dysplastic non-functioning left kidney with a normal right kidney. He was treated conservatively. He was found to be hypertensive (130/90 mm Hg) in the outpatient clinic at 23 months of age, and was admitted for further assessment.

Echocardiography, performed in view of persisting hypertension, showed left ventricular hypertrophy with an otherwise structurally normal heart (left ventricular posterior wall diameter 10.3 (normal range 4.5+/-0.9) mm and interventricular septum diameter 5.5 (4.7+/-0.9) mm). His resting plasma renin activity was raised at 11.4 (normal range 0.5–2.7) ng/ml/hour.

An uneventful nephrectomy was performed at 26 months of age and he was allowed home the following day. Histological examination of the kidney confirmed multicystic dysplasia with an atretic ureter.

He is currently well and normotensive. Recent echocardiography shows complete resolution of the left ventricular hypertrophy.

CASE 3

A 14 year old girl was referred to her local paediatrician because of a two month history of headaches with no other associated symptoms. After delivery at 38 weeks' gestation, a mass had been found in the right loin, and an intravenous pyelogram and DMSA scan were performed. These revealed total absence of excretion of contrast and uptake of isotope respectively. A diagnosis of right MCDK was made. The mass was no longer palpable at the time of discharge at 2 weeks of age. Follow up appointments were made, and a repeat DMSA scan at the age of 12 months confirmed a complete lack of isotope uptake. Plans were made for her to undergo cystoscopy in the future, though the family moved from the area when she was 18 months of age.

Urological review at 4 years of age showed a normal left kidney on ultrasonography and no right renal tissue. The presumptive diagnosis of right MCDK with natural resolution was made and the family reassured. No further follow up was arranged.

On admission she was found to have severe hypertension (180/110 mm Hg) with papilloedema on fundoscopy. Examination was otherwise normal. Serum creatinine, serum renin activity, and urinary catecholamines were normal. Ultrasonography revealed absence of the right kidney with a normal left kidney. DMSA images confirmed total absence of function on the right side. There was no microscopic haematuria nor evidence of microangiopathy on her blood film. She was initially given a labetolol infusion, which successfully produced controlled reduction in her blood pressure, and then converted to atenolol, although this was later changed to captopril because of side effects. Cystoscopy and a retrograde pyelogram were subsequently performed and a ureteric orifice was found; injection of contrast showing remnant renal tissue in the right loin. This remnant was removed at exploration of the right loin. Histological examination was consistent with a diagnosis of a severely dysplastic kidney.

At follow up she is normotensive with normal renal function.

Discussion

Treatment of hypertension associated with MCDK by nephrectomy has previously been reported in three infants,[7 11 12] two children aged less than 12 years,[12 13] and eight adults and children older than 12.[8 11 14 15] Resolution of hypertension after nephrectomy seems to decrease with increasing age at presentation, occurring in the three infants, both children less than 12 years of age, but only in two (12 and 21 years) of the eight patients aged 12 years or more. The poor results of nephrectomy in older patients may be attributable to hypertensive arteriolar damage in the contralateral originally normal kidney.[12] In addition to these 13 well described cases, Emmert and King have received personal correspondence about a further three patients in whom hypertension was cured by nephrectomy.[16] Moreover, a survey of 48 paediatric urologists reported that 15% had seen hypertension attributable to MCDK.[17] There are also five reported cases of hypertension in children with MCDK in which the authors have not attributed hypertension to the renal lesion, although they provide no supportive evidence to substantiate any alternative cause.[3 5] Plasma renin activity was measured in only a small number of these patients, being raised in two[7 15] and normal in one.[12] Hyperplasia of the juxtaglomerular apparatus was reported in the nephrectomy specimen from one 6 year old girl.[13]

We report a further three patients with hypertension directly attributable to MCDK with resolution of hypertension following nephrectomy. Both patients who underwent echocardiography studies had evidence of left ventricular hypertrophy indicative of hypertension of some duration, which also resolved after nephrectomy. The youngest two children had raised plasma renin activity; although the 14 year old girl with the most severe hypertension had normal activity. All samples were collected under optimal conditions, with patients in the resting state.

Disappearance of MCDK both clinically and on ultrasound examination is seen in most patients by 5 years of age,[5] and this may lead to cessation of follow up, as occurred in case 3. Resolution of the cysts on ultrasonography or disappearance of the mass clearly does not equate with resolution of the risk of hypertension, as significant stromal tissue may still be present. We have previously reported four children initially managed conservatively in whom progressive involution of MCDK was observed by ultrasound examination and whose parents later elected for surgical intervention.[18] While preoperative ultrasonography in all children

and enhanced computed tomography scanning in one failed to detect either cystic or solid tissue, all had significant residual cellular masses excised at operation. We believe that the risks of hypertension secondary to MCDK have been understated, and that based on the conclusions of these studies, many children may be receiving suboptimal follow up. The risk of hypertension alone is such that conservative management of MCDK should entail lifelong follow up with regular assessment of blood pressure.

The other major reported complication of MCDK is malignancy. A computer search reported by Gordon *et al* identified six cases, including two Wilms' tumours, three adenocarcinomas, and one embryonal tumour, over a 20 year period.[1] On the basis of these data, Wacksman and Phipps estimated that 8000 MCDK would need to be removed to prevent one Wilms' tumour.[5] Beckwith, basing his calculations on the increased incidence of nodular renal blastema in patients with both MCDK and Wilms' tumour, calculated this value to be 1600.[19] These calculations do not take into account the risk of malignancies other than Wilms' tumour. While this risk of malignant transformation may seem to be small, there are considerable difficulties associated with its early detection. Significant regression in the size of MCDK, leading in many to disappearance of the kidney on ultrasound examination, precludes the early detection of tumour growth in stromal tissue, and in those not followed by ultrasonography, the retroperitoneal location of the tumour may preclude early physical detection.

Additionally, the high incidence of ureteral atresia precludes early detection by either urine cytology or the development of macroscopic haematuria.[8] Tumours have developed in patients of various ages (Wilms' tumours in children and adenocarcinomas at ages of 15, 26, and 68 years), confirming the need for lifelong follow up of conservatively managed patients.

Early discharge from follow up is clearly not indicated given the available information about the incidence of complications of MCDK. We disagree with recent publications[2 3 6] which, on the basis of small series of patients, recommend an entirely conservative approach to the management of this lesion as we believe that the incidence of complications, particularly hypertension, has been generally underestimated.

After diagnostic confirmation of MCDK (ultrasound examination, DMSA scanning, and a MCUG to determine whether contralateral vesicoureteric reflux is present), we have shown that nephrectomy using a retroperitoneal approach results in minimal morbidity with most children being allowed home the day after surgery.

Between 1982 and 1992, 62 infants with MCDK diagnosed after antenatal detection of a renal anomaly were followed up prospectively at this institution. Some 49 (79%) underwent nephrectomy, 37 shortly after diagnosis and 12 after a period of follow up. There were no perioperative complications, and with the exception of two children with significant contralateral anatomical abnormalities all currently have normal renal function.[18] Surgery is therefore a safe and valid option.

In those patients who are managed conservatively, there are at present no clear guidelines as to how frequently ultrasound examination should be performed. Wacksman and Phipps have recommended that ultrasonography be performed every 3–6 months in infancy, every 6–12 months until 5 years of age, and annually thereafter.[5] Such demanding follow up of the seemingly asymptomatic child may clearly result in significant clinic non-attendance, especially in countries where there are financial costs associated with attendance at a doctor's surgery. This follow up may also provoke parental anxiety. Viewing the management dilemma in purely financial terms, the costs of nephrectomy with a 24 hour hospital stay and one postoperative follow up visit are less than those of 12 ultrasound examinations in the first five years of life with annual examinations thereafter, in addition to those costs associated with repeated attendance for monitoring blood pressure.

We currently favour elective nephrectomy from 3 months of age onwards as the treatment of choice for MCDK. Families are provided with balanced information, fully informing them of the risks of surgery and the potential morbidity associated with conservative management, thereby allowing them to take an active part in the decision making process. They must clearly understand that a conservative approach entails lifelong follow up with regular attendance for measuring blood pressure and ultrasound examination. Logically, the same approach should be taken with prenatally diagnosed and involuted MCDK, although the precise incidence of this is unclear and questions may be raised about the accuracy of the original ultrasound diagnosis of such lesions.

The arguments for and against surgical management of MCDK will continue, though based on current information regarding the safety of operation and the complications that we have encountered in conservatively managed patients, surgery will remain our treatment of choice. In centres in which conservative management is undertaken, physicians unaware of the potential complications of MCDK need to be dissuaded from discharging patients from follow up once 'disappearance' of the kidney on ultrasound examination has occurred. National registries with complete long term follow up data are necessary to provide complete information about the true incidence of complications of both the medical and surgical approaches to treatment to help resolve this dilemma.

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