Unilateral multicystic dysplastic kidney: long term outcomes

M Aslam, A R Watson, on behalf of the Trent & Anglia MCDK Study Group

See end of article for authors’ affiliations

Correspondence to: Prof. A R Watson, Consultant Paediatric Nephrologist, Nottingham University Hospitals NHS Trust, City Hospital Campus, Hucknall Road, Nottingham NG5 1PB, UK; judith.hayes@nuh.nhs.uk

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PATIENTS AND METHODS
Since 1985 we have maintained a prospective regional registry of patients with antenatally detected MCDK using a common protocol of investigation and follow up agreed among members of the study group. MCDK was defined on the basis of antenatal and postnatal ultrasound appearance of non-communicating cysts and dysplasia with no function on a dimercaptosuccinic acid (DMSA) radionuclide scan usually performed within three months of birth. A kidney with cystic dysplasia and function of any degree on the DMSA was excluded from the study.

Infants in this series were not routinely prescribed prophylactic antibiotics from birth. Trimethoprim (2 mg/kg/dose) twice a day for 2 days was prescribed to cover the micturating cystourethrogram (MCUG). If VUR Grade II or greater was detected on the MCUG, children were commenced on prophylactic trimethoprim for 2 years (2 mg/kg nocte). VUR was graded Grades I–V according to the international classification.

Clinical assessment was carried out at 3 months at the time of the DMSA scan and at 6 and 12 months of age with clinic visits. Annual reviews were performed thereafter until 5 years of age and then biannually until 10 years of age. At each clinic visit there was enquiry of clinical symptoms, especially urinary tract infection, with measurement of growth parameters and urinalysis by dipstick. Blood pressure was assessed by Doppler for systolic pressure in young infants or standard mercury or aneroid sphygmomanometer in older children with reference to standard blood pressure centile charts for age and sex.

Ultrasound examinations (USS) were performed at 2, 5, and 10 years of age. The USS measurements included an assessment of size of the MCDK and contralateral kidneys in relation to standard deviations of the mean value of normal kidneys.

The involution rate was calculated using life table analysis and Kaplan–Meier statistics. χ² analysis was used for comparison of urinary tract infection rates in those with or without vesicoureteric reflux.

Abbreviations:
GFR, glomerular filtration rate; MCDK, multicystic dysplastic kidney; MCUG, micturating cystourethrogram; PUJ, pelviureteric junction obstruction; USS, ultrasound scan; UTI, urinary tract infection; VUR, vesicoureteric reflux

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RESULTS

Between 1985 and 2004, 202 cases of unilateral MCDK were registered. The right kidney was involved in 99 cases (49%). In six children the unilateral MCDK was associated with other congenital abnormalities (radial aplasia, Ehlers–Danlos syndrome, bladder diverticulum, hypospadias, CHARGE association, and DiGeorge syndrome). In four families there was a history of a single kidney (two grandparents, one mother, one elder sibling who died prematurely at 26 weeks). The mean age of the patients under follow up at the time of the study was 8.1 years (range 0.1–19.7 years); 165 had been followed to 2 years, 117 to 5 years, and 43 to 10 years.

A total of 143 children had undergone MCUG; 27 patients (19%) had reflux into the contralateral kidney, with VUR occurring into the atretic ureter of the MCDK in 23 (16%). The grade of VUR into the contralateral kidney was mild (Grade I–II) in 74%, moderate (Grade III) in 22%, and gross (Grade V) in one child where the kidney was small and “globally scarred”. The initial USS in this child showed abnormal kidney structure and ureteric dilatation. Thirteen children with no documented VUR on MCUG had a UTI compared to seven children who also had UTIs while on prophylactic antibiotics for VUR under 2 years of age (not significant). The urinary tract symptoms were those of cystitis; no cases of pyelonephritis or hospitalisation for antibiotic treatment were documented.

Repeat MCUGs and DMSA scans were not performed after the initial investigations. A detailed USS at 2 and 5 years showed no evidence of renal scar formation in children with or without VUR or UTI.14 No child has had any evidence of proteinuria on dipstick urinalysis to date. No children with hypertension were recorded. No children had symptoms of vomiting or mass effect and no cases of malignancy have been noted.

Two children developed acute renal failure with PUJ obstruction in the contralateral kidney which required pyeloplasty at 9 and 14 months respectively.15 The initial USS in both showed renal pelvic dilatation in the good kidney which merited closer follow up. Nephroureterectomy of the MCDK was performed in 11 children before 2 years of age. Indications included parental anxiety in two, suspicion of benign nephroma in one (histopathology confirmed MCDK only), and failure to reduce in size by 1 year of age in one patient. No indications were clearly specified in the remainder. No patient has undergone nephrectomy in Nottingham regional centre since 1988.

The rate of involution of the MCDKs on serial ultrasound is shown in fig 1. In 11 infants the initial postnatal USS showed disappearance of the MCDK which had been clearly defined antenatally. At 2 years, 34% had completely involuted; this percentage had risen to 47% at 5 years and 59% at 10 years. Forty three patients have reached 10 years’ follow up, with 35 of 43 (81%) demonstrating compensatory hypertrophy of the contralateral kidney. In five patients the kidney was above average size, and in three, size was not recorded. The mean estimated GFR (eGFR) of 31 patients based on height and plasma creatinine16 was 86.1 ml/min/1.73 m² (range 48–125), with 13 patients having eGFR >90, 16 eGFR 60–90, and two an eGFR <60 ml/min/1.73 m². All children had compensatory hypertrophy (>2 SD) of the contralateral kidney, with abnormal echogenicity suggestive of dysplasia being evident in the two patients with eGFRs of 48 and 59 ml/min/1.73 m² respectively.

DISCUSSION

This study is the largest reported series to date documenting the natural history of patients with MCDK who have been followed with a standard protocol. Paediatric surgeons and physicians have contributed to the registry, and apart from 11 children who underwent nephrectomy during the early years of the study, the follow up has documented the natural history of progressive involution. The lack of any clinical problems justifies conservative management.

Various reports have documented an involution rate of the MCDK of 25–52% between 2.5 and 6.5 years.17 18 Rabelo et al, in a recent paper based on follow up of 43 children from a single centre, suggested a median time of 122 months for the MCDK to become undetectable on USS.19 The current series suggests that 40% of MCDK kidneys completely involute by 5 years and 59% by 10 years. Those that remain visible at 10 years are considerably smaller, with few remaining cysts. We therefore continue to follow these patients into their teenage years to confirm our suspicion that all will eventually involute with time. It is strongly suspected that MCDK is the major contributor to one of the commonest abnormalities in the general population, that of possessing only one visible kidney. Discussion with adult urological colleagues suggests that an MCDK kidney is rarely, if ever, recognised in adult practice.

The contrary view is that nephrectomy for all non-functioning kidneys can easily be performed on an outpatient basis with improvement in the child's insurability.20 21 The reasons for advocating nephrectomy of the MCDK are the rare problems of mass effect, hypertension, and potential for malignancy. The MCDK kidney can cause a large mass effect antenatally. However, we are unaware of any patient in our series who had delivery by caesarean section or who underwent postnatal operation for vomiting due to the mass effect.

Hypertension has been reported in children with MCDK and a recent systematic review of 29 studies revealed only six cases in 1115 eligible patients.22 Resolution of hypertension by removal of the affected kidney at 3 months, 23 months, and in a 14 year old patient has been reported,2 23 but hypertension may persist even after operation. Hivert and in a 14 year old patient has been reported,2 23 but hypertension may persist even after operation.21 Reports have suggested that the hypertension may be renin mediated24 and may be transient in infants where there are difficulties in measurement of blood pressure.25 26 It is still uncertain how often and for how long children with a unilateral MCDK need to be assessed for hypertension. Further reports such as ours increase the denominator of patients at risk. Hypertension must be a very rare event when one considers all the other


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