Role of nephrectomy in unilateral multicystic dysplastic kidneys

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Abstract. **Objective:** The objective of our study was to assess role of nephrectomy in patients having unilateral multicystic dysplastic kidneys (MCDK). **Methods:** All patients with the diagnosis of MCDK from January 2000 to December 2010 were retrospectively analysed. **Results:** Out of a total of 27 patients, 10(37%) were diagnosed antenatally. Of the 10 patients diagnosed antenatally, 2(20%) had involution on postnatal ultrasound (USG) and 6(60%) patients showed subsequent involution on USG at average age of 4.3 years. Out of these, 5 underwent magnetic resonance imaging (MRI) to confirm involution. Of the five, 2(40%) showed complete involution but 3(60%) had some identifiable structure on MRI (average size of 2.2 cm). Two patients underwent retroperitoneoscopic excision of the remnant tissue. Out of remaining 17(63%) patients detected postnatally, 14(51.8%) were incidentally detected on USG. Of them, 9(64%) showed little or no change in size and underwent nephrectomy. Only 5(35%) patients showed resolution in the size of the kidney on USG. Two of these five patients showed residual tissue on MRI and both refused to undergo any surgical exploration. Of the remaining 3 patients, 1(7%) had persistent hypertension, underwent removal of dysplastic kidney following which patient is normotensive and other 2 patients had no remnant on MRI; one of them underwent retroperitoneoscopic exploration and remnant tissue was removed. Another 3(11%) patients presented with lump and pain underwent nephrectomy at 2 years of age for average size of dysplastic kidney of 9.2 cm. **Conclusion:** Surgical excision of MCDK should be offered as a safe alternative.

**Key words:** Antenatal, Multicystic dysplastic kidney, Nephrectomy

**Introduction**

The end point of treatment of multicystic dysplastic kidney (MCDK) is not well defined. The present day protocol is conservative management with regular blood pressure monitoring, serial ultrasounds (USG) and clinical assessment. Various protocols have been suggested; most of them advocate vigorous follow up for the first two years followed by annual USG for next 3-5 years.\(^1\)\(^-\)\(^3\) Uniformly all studies report an involution in about 30-50% cases and regression in 30-40% cases.\(^4\)\(^-\)\(^6\) However, most studies do not address the case of the patients, who persist to have a remnant tissue after this period. In the absence of complications, they are increasingly managed conservatively, but because of the perceived long term risk of hypertension and renal neoplasms, mainly Wilms' tumour, they are also followed up throughout childhood.\(^7\)\(^-\)\(^9\) The length of follow up in these patients is unclear. Indications for operating such patients have not been uniformly defined.

Antenatal ultrasounds have completely altered the profile of the patients presenting to the physician. Now-a-days most cases of MCDK are diagnosed antenatally. However this is not the case in the developing nations and it will be still some time before antenatal ultrasounds become universally available. So most patients still present with symptoms or larger sized dysplastic kidney.

The rigorous follow up and cost of repeated ultrasounds, have to be taken into account. Access to medical care may not be a concern in the Western world but is a serious issue in most of the developing nations. As a result lost to follow up and treatment drop-out rates are fairly high in our country. In this scenario, conservative management of MCDK may not always be the best option.

All patients of MCDK were retrospectively analysed for assessing the role of nephrectomy in such patients and need for alternative management protocol to the current observation only policy in vogue.

**Materials and Methods**

All patients registered in the paediatric urology clinic from
January 2000 to December 2010 with the diagnosis of MCDK were analysed retrospectively. The diagnosis of MCDK was confirmed with USG and Tc99m DMSA isotope scan. All the patients underwent a voiding cystourethrogram (VCUG) to diagnose associated anomalies. All symptomatic patients were offered surgery while the others were kept on conservative management. The follow up protocol included 3 monthly ultrasounds in the first year of life, 6 monthly in the second year and annually following that. Patients showing significant remnant tissue at 5 years of age or after 3 years of observation or the ones developing symptoms were offered surgery. The ones showing involution on USG were offered to undergo Magnetic Resonance Imaging (MRI) and laparoscopic exploration to confirm complete involution.

Results

A total of 37 patients including 20 boys and 17 girls, were registered in the clinic. Ten patients had stopped follow up. Of these ten patients, 7 patients could be contacted. At the time of analysis, 5 had stopped visiting due distance from the centre and/or financial reasons. Two patients had undergone nephroureterectomy elsewhere.

Associated urologic anomalies were seen in 20 patients of the remaining 27 children. Contralateral vesicoureteric reflux was seen in 14 patients while hydronephrosis was seen in 6 patients. One patient had hypertension and altered renal parameters, but this patient had severe contralateral reflux. No other patient had hypertension or renal failure. No patient showed any evidence of malignancy during the course of follow up.

Of the 27 patients, 10 patients were diagnosed prenatally (Table 1). Postnatally, two of the babies had completely involuted kidneys on the first USG itself (done at 2 months and 3 months respectively). Another 6 patients, with average initial size 3.8 cm, showed involution at an average age of 4.3 years. Five of these patients underwent MRI. Two (40%) of these children showed complete resolution while 3(60%) showed some remnant dysplastic tissue (average size 2.2 cm) (Table 2). One of them underwent retroperitoneoscopic excision of the dysplastic remnant (size 2.5 cm). One patient is awaiting surgery while another refused surgery. Two (20%) patients had average initial size of 5.3 cm and both of them persisted to have remnant tissue at the end of five years. Both underwent nephrectomy.

The remaining 17 patients presented without an antenatal diagnosis (Table 1); 3 presented with lump and pain while 14 were detected incidentally on USG done for other indications at an average age of 4.8 years. Three detected with symptoms, underwent nephrectomy and the average size of the dysplastic tissue was 9.2 cm. The remaining 14 patients were kept under observation with serial ultrasounds. Of these, 9 patients showed little or no change in the size of the dysplastic kidney for three years and underwent nephrectomy. The average size of MCDK was 7.7 cm at the time of diagnosis in this group while it was 6.8 cm at the time of nephrectomy (Table 2). Other 5 patients in this group showed resolution in the size of the kidney. The average renal size was 4.1 cm at the time of initial diagnosis. Two of these patients showed some residual tissue on MRI but patient refused to undergo any surgical exploration for confirmation. Two patients could not afford MRI. One patient had persistent hypertension and underwent retroperitoneoscopy and persistent dysplastic tissue was found and excised from the renal fossa (Fig. 1). The patient became normotensive following surgery. Two patients had no residual tissue on MRI and one of them underwent retroperitoneoscopic exploration and remnant tissue of 2.4 cm was removed. The average size of the dysplastic tissue in the patients undergoing nephrectomy was 5.6 cm (n=12). Average size of tissues excised after USG had failed to detect was 2.0 cm (n=3). The patients presenting with symptoms had an average size of 6.6 cm (n=3), while it was 5.3 cm (n=9) in whom the dysplastic tissue persisted after follow up (Table 2).

Discussion

In last few years standard treatment of multicystic dysplastic kidney has changed from nephrectomy to regular follow up for involution. Routine follow up with regular ultrasonography is done to see the decrease in size of cystic structure. Once cysts disappeared it is regarded as involution.

According to some authors it is only cystic component which disappears but dysplastic small component always
Table 2. Management of dysplastic kidneys

<table>
<thead>
<tr>
<th>Group</th>
<th>Symptoms</th>
<th>Fate on USG</th>
<th>MRI</th>
<th>Surgery</th>
<th>Average size at initial detection</th>
<th>Average size at surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Antenatal</td>
<td>Asymptomatic</td>
<td>Resolved at first USG (2)</td>
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<td>Nephrectomy (2)</td>
<td>5.3 cm</td>
<td>4.8 cm</td>
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<tr>
<td></td>
<td>(10)</td>
<td>Static (2)</td>
<td>Remnant tissue not seen (2)</td>
<td>Refused further intervention (2)</td>
<td>3.8 cm</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td>Regressed (6)</td>
<td>Remnant tissue picked up (3)</td>
<td>Nephrectomy (1)</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Awaiting surgery (1)</td>
<td></td>
<td></td>
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<td></td>
<td>Refused surgery (1)</td>
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</tr>
<tr>
<td>Postnatal</td>
<td>Symptomatic (3)</td>
<td>-</td>
<td>Nephrectomy (3)</td>
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<td>9.2 cm</td>
<td>9.2 cm</td>
</tr>
<tr>
<td>Incidental</td>
<td>Static (9)</td>
<td></td>
<td>Nephrectomy (9)</td>
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<td>7.7 cm</td>
<td>6.8 cm</td>
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<tr>
<td></td>
<td>Regressed (5)</td>
<td>Remnant tissue seen (2)</td>
<td>Refused (2)</td>
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<td>2.4 cm</td>
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<td>Not done (1)</td>
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<td></td>
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<tr>
<td></td>
<td></td>
<td>Not seen (2)</td>
<td>Nephrectomy (1)</td>
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Fig. 1. Retroperitoneoscopic view showing multicystic dysplastic kidney (Asterix: Individual cysts)

remained there. This dysplastic component is responsible for which we are worried in MCDK. Most of the times there remain a small sized dysplastic component which cannot be picked up on routine ultrasonography and easily detected by MRI or retroperitoneoscopy. In a study by R Luque-Mialdea et al[11] retroperitoneoscopic approach revealed persistence of dysplastic renal tissue in 100% of the patients. In 86% the renal remnant was located in the renal fossa and in two cases in a pelvic ectopic location. Luis m. Perez et al[12] in 1998 concluded that early nephrectomy is more cost-effective than observation in neonates with multicystic dysplastic kidney. Our study has a number of limitations firstly it is retrospective study, secondly period of follow up is not defined, thirdly lost to follow up is high. All purpose of this study is to state that in country like ours where health institute serves a very large area, access to health care is very limited and most of patient strata is from the group where it is very difficult to spend money on regular follow ups; nephrectomy is a good option.

Conclusion

Till endpoints of observation are well defined, surgical excision of multicystic dysplastic kidneys should be offered as a safe alternative to parents. It avoids repeat ultrasounds, which in itself does not correctly identify complete involution. It also allays the anxiety of the parents and is
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important in developing nations where repeat visits to health centres is not feasible for all. Antenatal ultrasounds are still not widely available to all and most of the patients present with larger and symptomatic dysplastic kidneys and are less likely to regress. Nephrectomy has bigger role in multicystic dysplastic kidneys in our nation.

References