

## Indications for Nephrectomy in children : What has changed?

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La Néphrectomie chez l'enfant. Les indications ont-elles changé ?

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### R É S U M É

**Prérequis :** Des progrès remarquables ont été faits ces dernières années dans la prise en charge des uropathies chez l'enfant sur le plan investigations, indications et techniques opératoires. Plusieurs uropathies congénitales ont bénéficié d'une meilleure connaissance de leur physiopathologie et de leur génie évolutif.

**But :** Revoir les indications de la néphrectomie chez les enfants opérés entre 1996 et 2008 au service de chirurgie pédiatrique de l'Hôpital d'Enfant de Tunis.

**Méthodes :** Il s'agit d'une étude rétrospective concernant 80 enfants ayant eu une néphrectomie. Les deux périodes allant de (1996-2000) et (2001-2008) ont été comparées.

**Résultats :** Le nombre total de néphrectomie par an a augmenté (4,6 et 8 néphrectomies /an pour 1996-2000 et 2001-2008, respectivement;  $P < 0.05$ ), ainsi que pour la dysplasie rénale multikystique (zéro à 5 pour 1996-2000 et 2001-2008, respectivement) et les néphroblastomes (8,3% et 29,16% pour 1996-2000 et 2001-2008, respectivement). Les néphroblastomes, le reflux vésico-urétéral et l'anomalie de la jonction pyélo-urétérale représentent plus que la moitié des néphrectomies (80% et 58% pour 1996-2000 et 2001-2008, respectivement). La proportion des néphrectomies secondaires à un reflux vésico-urétéral n'a pas changé (15% et 12% pour 1996-2000 et 2001-2008, respectivement). Par contre les anomalies de la jonction pyélo-urétérale sont de moins en moins justiciables d'une néphrectomie (44% à 4,16% respectivement pendant 1996-2000 et 2001-2006 ;  $p < 0.05$ ).

**Conclusion :** Le nombre des néphrectomies y compris les néphrectomies polaires ont significativement augmenté ces dernières années. Les anomalies de la jonction pyélo-urétérale sont de moins en moins justiciables d'une néphrectomie grâce au diagnostic anténatal et la prise en charge précoce de cette uropathie. La proportion des néphrectomies secondaires à un reflux vésico-urétéral n'a pas changé par contre cette proportion a augmenté pour les néphroblastomes et les dysplasies multikystiques du rein.

### S U M M A R Y

**Background:** The last decade has witnessed significant refinements in preoperative diagnostic evaluation and an improvement in surgical techniques and postoperative management for paediatric patients. There has been an improvement in our understanding of the natural history of some congenital renal anomalies which has caused some changes in management approach.

**Aim :** To review the indications for nephrectomy in children between 1996 and 2008, at the department of paediatric surgery, children's hospital in Tunis.

**Methods:** There were 80 nephrectomies. A retrospective review of the patients' notes was performed. The 13-year period was divided into two halves (1996-2000 and 2001-2008) which were then compared.

**Results:** The total number of nephrectomies per year significantly increased over the period of the study (4,6 and 8 nephrectomies per year for 1996-2000 and 2001-2008, respectively ;  $P < 0.05$ ), as did the number of nephrectomies for Multicystic dysplastic kidney (MCDK) (zero and 5 for 1996-2000 and 2001-2008, respectively) and wilms'tumour (8,3% and 29,16% for 1996 - 2000 and 2001 - 2008, respectively). Wilms' tumour, vesico-ureteric reflux (VUR) and pelvi-ureteric junction (PUJ) obstruction accounted for more than half of the nephrectomies (80% and 58% for 1996-2000 and 2001-2008, respectively). The proportion of nephrectomies performed for VUR did not change (15% and 12% for 1996-2000 and 2001-2008, respectively) but fewer nephrectomies were performed for pelvi-ureteric junction (PUJ) obstruction in the second half of the study period (44% and 4,16% for 1996-2000 and 2001-2008, respectively ;  $P < 0.05$ ).

**Conclusion:** The total number of nephrectomies, including partial nephrectomies, has increased significantly. The decrease in nephrectomies for PUJ obstruction could be accounted for by a more aggressive approach in the management and follow up of prenatally diagnosed hydronephrosis. Of note is that there was no significant change in the proportion of nephrectomies performed for VUR. On the contrary, the proportion of nephrectomies increased for neoplastic lesions and MCDK.

### Mots-clés

Néphrectomie ; Indication ; Enfant.

### Key- words

Nephrectomy; Indications; Children

ادواحي إستئصال الكلية عند الأطفال  
الباحثون : نويرة. ف - سراي. ن - غربال. س - ولد محمد صغير. ي - خماخم. ر - شريق. ع - جليدي. س - شواشي. ب.  
الهدف من هذا العمل هو دراسة ادواحي إستئصال الكلى عند الأطفال اللذين خضعوا لذلك في الفترة ما بين 1996 إلى 2008 بقسم الجراحة ب "مستشفى الأطفال باب سعدون".  
هذه دراسة مؤخرة 80 طفل خضعوا لعملية إستئصال الكلية، وقد وزعوا على حقيبتين الأولى من (2008 - 1996) والثانية من (2008 - 2001) وتمت المقارنة بينهما.  
الكلمات الأساسية : إستئصال الكلى، الطفل.

The last decade has witnessed significant refinements in preoperative diagnostic evaluation and an improvement in surgical techniques and postoperative management for paediatric patients. Laparoscopic nephrectomy in children has become a routine practice in several centres (1,2). There has been an improvement in our understanding of the natural history of some congenital renal anomalies, such as multicystic dysplastic kidney (MCDK), which has caused some changes in management approach. We thought it pertinent to assess the effects of such changes on the indications for nephrectomy in the paediatric age group. A literature review revealed a limited number of publications on the indications for nephrectomy in children. A few reports reviewed the indications in various age groups without any special mention of paediatric patients (3,4). Others reported on a small number of nephrectomies in children (5,6). Here, we report on the series of nephrectomies in paediatric patients, their indications, and the clinical presentation of this particular group of patients with various renal abnormalities that required nephrectomy.

## PATIENTS AND METHODS

All nephrectomies performed in the unit of pediatric surgery « B » at children's hospital of Tunis were reviewed retrospectively. The study covered the period between 1996 and 2008 inclusive. Partial nephrectomies were included. Data were collected regarding age of patient, indication for nephrectomy, clinical presentation, and radiological, laboratory and histological investigations.

The indication for nephrectomy was determined by clinical presentation, investigations and pathological diagnosis when applicable. A total of 89 nephrectomies were performed in the study period. The clinical records related to 80 nephrectomies were available for analysis. The number of nephrectomies each year was calculated. To study the change in indications, the 13 years period was divided into two periods (1996-2000 and 2001-2008) that were then compared. The  $\chi^2$  (Chi-squared) test was used for statistical analysis. A P value of  $<0.05$  was considered statistically significant.

## RESULTS

Table 1 shows the indications for nephrectomy in the period of study. All nephrectomies were performed using an open technique and no laparoscopic nephrectomies were identified. The mean age at the time of the operation was 50 months (range 0.3-110). There were 46 males (57,8%). Patient had 70 total nephrectomies (right : 29; left : 41), 6 partial nephrectomies and four cases that had simultaneous total and partial nephrectomies. The number of nephrectomies per year has significantly increased over the period of the study (4,6 and 8 nephrectomies per year for 1996-2000 and 2001-2008, respectively;  $P < 0.05$ ). Neoplastic lesions accounted for 43,6% ( $n = 35$ ) of the nephrectomies (Table 1). Twenty seven

nephrectomies (33%) were performed for Wilms' tumour. Out of these, there were four simultaneous total and partial nephrectomies for synchronous bilateral tumours. The main clinical presentations in patients with Wilms' tumours were abdominal swelling ( $n = 18$ ) and haematuria ( $n = 4$ ) (Table 2). Using the National Wilms' Tumour Study staging system, the distribution of stages is: stage I ( $n = 11$ ), stage II ( $n = 7$ ), stage III ( $n = 4$ ), stage IV ( $n = 1$ ), stage V ( $n = 4$ ). The proportion of nephrectomies performed for Wilms' tumour had changed between the two halves of the study period (8,3% and 29,16% for 1996 - 2000 and 2001 - 2008, respectively).

Table 1 : Nephrectomy indications

	Number		
	1996-2000	2001-2008	1996-2008
Neoplastic lesions			
- Wilms' tumour	6	21	27
- Adrenal neuroblastoma	0	2	2
- Renal cell carcinoma	0	2	2
- Bolland tumour	1	1	2
- Immature teratoma	0	1	1
- Multilocular cystic	0	1	1
nephroma			
Hydronephrotic anomalies	4	6	10
- VUR	12	3	15
- PUJ obstruction	2	4	6
- Duplex kidney	0	2	2
- VUJ obstruction	1	4	5
- Posterior urethral	0	5	5
valves	1	1	2
MCDK	27	53	80
Urolithiasis			
Total			

Table 2 : Mode and age at presentation of the most common indications for nephrectomy

	N	Age (months)
Wilms' tumour		
- Abdominal swelling	18	(3-126)
- Haematuria	4	(10-56)
- Abdominal pain	3	(54-167)
- Hypertension	3	(9-24)
- UTI	2	(11,42)
- Anemia	1	(36)
- Coincidental	1	(31)
VUR		
- UTI	6	(35-94)
- Hypertension	1	
- Antenatal	3	
PUJ obstruction		
- Antenatal	4	N/A
- UTI	2	(26-36)
- Abdominal pain	9	(44-88)
Posterior urethral valves		
- UTI	4	(1-123)
- Antenatal	1	N/A
MCDK		
- Antenatal	5	N/A
Duplex kidney		
- Antenatal	2	N/A
- UTI	3	(6-35)
- Enuresis	1	(72)

Pelvi-ureteric junction (PUJ) obstruction accounted for 18% (n =15) of the nephrectomies performed on native kidneys. Assessment of renal function by an isotope renogram was performed in all these cases. The degree of function of the affected kidneys was as follows: 5% (n = 7), 5-10% (n = 5), 10-15% (n = 1), 15-20% (n = 1), and one kidney contributed to 28% of the total renal function. On exploring this kidney with the intention of performing a pyeloplasty, very little parenchyma was noted, and the decision was taken intraoperatively to remove the kidney. The proportion of nephrectomies performed for PUJ obstruction has decreased significantly over the years (44% and 4,16% for 1996-2000 and 2001-2008, respectively ;  $P < 0.05$ ).

Partial nephrectomy was performed in 10 cases: unilateral (n =6), and associated with contralateral total nephrectomy (n =4). The indications for partial nephrectomy were duplex kidney (n =6) and Wilms' tumour (n =4). All cases had upper pole nephrectomy. The number of partial nephrectomies has increased over the years (3 and 7 partial nephrectomies for 1996-2000 and 2001-2008, respectively).

Ten cases (12%) had total nephrectomy due to vesico-ureteric reflux (VUR). All cases had micturition cystourethrogram prior to surgery. The grade of the reflux is as follows: V (n=6), IV (n =3) and III (n = 1). An isotope renogram was performed in all cases to determine the split renal function which ranged from 0 to 15%. UTI (n = 8) was the main clinical presentation in VUR cases needing nephrectomy. The proportion of total nephrectomies performed for VUR did not change between the two halves of the study period (15% and 12% for 1996-2000 and 2001-2008, respectively).

Nephrectomy was performed for MCDK in 6,2% (n=5) at the period 2001-2008. None was realised at the first period. All these cases were first detected during antenatal US examination. An intravenous urogram and an isotope renogram were used to demonstrate the no or poor renal function. Two cases were found to have contralateral urinary abnormalities, mainly VUR reflux. The proportion of nephrectomies performed for MCDK had change between the two halves of the study period (0% and 6,9% for 1996-2000 and 2001-2008, respectively).

## DISCUSSION

A literature review revealed a limited number of publications on the indications for nephrectomy in children. A few reports reviewed the indications in various age groups without any special mention of paediatric patients. This series reported of nephrectomies in the paediatric age group.

Despite the significant increase in the number of nephrectomies in the second half of the study period, it is difficult from this report to conclude that the rate of performance of this procedure in children has risen, owing to the increase in the paediatric population served by our institute.

Neoplasia was the major indication for nephrectomy in this study. In an adult population, neoplasia is the main indication of

nephrectomy. Beisland and colleagues showed that 67% of nephrectomies in adults were due to malignant tumours (4). In children, however, neoplasia only represents 43,6% of indications. This is to be expected in view of the higher incidence of malignancy in older patients and the higher proportion of children who present with congenital malformations.

The decrease in the proportion of nephrectomies performed for PUJ obstruction is probably due to the vigorous use of antenatal scan to detect these cases as early as possible thus allowing proper follow up and intervention when required. In effect the management of PUJ obstruction at the child considerably evolved in the course of last 20 years. Before the epoch of obstetrical ultrasound scan, diagnosis was carried in the waning of symptoms and drove quasi systematically to an operation. The provision of anténatal diagnosis from 1980, augmented the number of noticed cases, profoundly changed the principles of treatment and allowed to individualize new physiological notions. The anténatal monitoring and the neonatal taking care of these children allowed to note that many hydronéphroses (7) regress spontaneously. In effect several series showed that the only 20 % of the watched patients require a surgical treatment (8), provide a regular surveillance over several months, or even several years is given.

This is in agreement with the findings of Capolicchio and colleagues who demonstrated that early diagnosis of hydronephrosis provided by prenatal ultrasonography is associated with less obstructive nephropathy (9).

Reflux nephropathy was the third major indication for nephrectomy in children. The introduction of an active medical and surgical treatment approach for children with VUR in the 1960 did not prevent renal damage and the ultimate need for nephrectomy (10). Whether VUR is associated with congenital dysplasia/hypoplasia that is not amenable to any form of postnatal therapy, or whether this form of treatment is unsuccessful in preventing further renal damage has yet to be clarified. A report from the Australia and New Zealand Dialysis and Transplant Registry concluded that treatment of children with VUR does not appear to reduce the incidence of end-stage renal disease (11).

The fact that the majority of patients with MCDK presented antenatally and the male predominance are in agreement with previous reports (12,13). The findings of contralateral and ipsilateral urological abnormalities were also previously demonstrated by others (14).

The treatment of MCDK has passed through different phases. Historically, nephrectomy was long regarded as the standard treatment and the safest means to avoid the complications of infection, bleeding, flank pain, hypertension and possible malignant transformation until 1980. In the last decade, there has been a shift in the management of MCDK from surgical removal to a conservative approach using serial US examination (12,15). This change in treatment is due to the perception that malignant transformation is a rare occurrence, and sequential US imaging of a large number of infants diagnosed with MCDK has revealed that most of these structures involute over time (12). In recent years, the argument

has emerged again with regard to the management of MCDK; several studies have recommended surgical removal because the natural history of the condition in the long term is still uncertain, and nephrectomy is more cost effective than conservative management (16,17,18). The effect of the different phases of management in the last decade is significantly reflected in the results of our study, as shown by the increase in the proportion of nephrectomies performed due to MCDK in the second half of the study period.

Improvements in surgical techniques have made nephron-sparing surgery an attractive alternative to total nephrectomy in select patients. Partial nephrectomy is an appropriate therapy for patients with localized renal disease (inflammatory or mitotic) in whom preservation of functioning renal parenchyma is a relevant clinical consideration. This concept is reflected in the practice of our institute where there was a significant increase in the proportion of partial nephrectomies performed in the latter part of the study period.

The majority of partial nephrectomies performed in the current study are in patients with duplex kidney. This is consistent with the finding of Privett and associates of a higher percentage of abnormalities in duplex units than in single-system kidneys

(27% vs 3%) (19). All our four cases of partial nephrectomy for Wilms' tumour were performed post-chemotherapy in patients with a synchronous tumour with favourable histology. This is in agreement with the finding of Linni and colleagues that nephron-sparing procedures remain the operative approach of choice in patients with bilateral Wilms' tumor (20). It is of interest to note that none of our partial nephrectomies were performed in patients with unilateral Wilms' tumour. Despite the fact that good results have been reported (21) for partial nephrectomy in patients with small and favourable histology, this treatment approach is still controversial.

## CONCLUSION

Wilms' tumour and hydronephrotic anomalies are the main indications for nephrectomy in children. The proportion of nephrectomies performed for VUR did not change over the study period reported. On the contrary, the proportion of nephrectomies increased for neoplastic lesions and MCDK. The decrease in the proportion of nephrectomies for PUJ obstruction could be due to the increased use of antenatal ultrasonography.

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