

ORIGINAL RESEARCH



# Paediatric nephrectomy: Patterns, indications and outcome in a developing country

Uchekukwu Obiora Ezomike<sup>1</sup>, Victor Ifeanyichukwu Modekwe<sup>2</sup>, Sebastian Okwuchukwu Ekenze<sup>1</sup>

1. Sub-Department of Paediatric Surgery, Faculty of Medical Sciences, College of Medicine, University of Nigeria, Ituku/Ozalla Campus, Enugu, Nigeria  
 2. Paediatric Surgery Unit, Department of Surgery, Nnamdi Azikiwe University Teaching Hospital, Nnewi, Nigeria

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 Correspondence: Uchekukwu Obiora Ezomike (ezomikeuche@yahoo.com)  
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## Abstract

### Background

Patterns of and indications for nephrectomy vary in different age groups, geographical locations and time periods. In some series nephrectomies were mainly for malignant conditions while in others they were predominantly for non-malignant conditions. Such data on patterns, indications, and outcomes of nephrectomy in children is limited in our environment.

### Objectives

To evaluate nephrectomy in childhood at the Sub-Department of Pediatric Surgery University of Nigeria Teaching Hospital, Ituku/Ozalla Enugu with a focus on pattern, indications, and outcome.

### Materials And Methods

Medical records of all patients aged ≤16years who had nephrectomy from January 2007 to December 2016 were studied with emphasis on age, sex, side of nephrectomy, duration of symptoms before presentation, indication for nephrectomy, in-hospital complications, length of hospital stay, in-hospital mortality. SPSS version 15 was used for data entry and analysis.

### Results

There were 52 nephrectomies in 32 males and 20 females. They were for 35 malignant and 17 non-malignant conditions. Most of the malignancies were Wilms tumour (34/35) while non-malignant conditions were late-presenting pelvi-ureteric junction obstruction (9), large multi-cystic dysplastic kidneys (4), renal trauma with pedicle avulsion (1), posterior urethral valve with atrophic kidney (1), duplex system with nonfunctioning upper pole moiety (2). Mean age at nephrectomy was 5.10±3.66 years (range 7 weeks to 16 years); 59% of the nephrectomies were on the left and 41% on the right. Mean duration of hospital stay was 31.78±16.59 days (range 7-66 days). In-hospital mortality rate was 5.8%.

### Conclusions

In our unit, nephroblastoma is the main indication for pediatric nephrectomy and were the only indications in females; neglected pelvi-ureteric junction obstruction was the major non-malignant indication and occurred only in males; most nephrectomies were done in the age range of 1-5 years; nephron-sparing nephrectomy, major morbidity, re-operation are uncommon and in-hospital mortality from nephrectomy is still high at 5.8%.

## Introduction

Surgical removal of the kidney in children is a major undertaking. The procedure may be for non-malignant conditions of the kidney and upper urinary tract causing poorly functioning nephroureteral units<sup>1</sup>. These non-malignant conditions may include chronic destructive infections, chronic obstructions with obstructive uropathy or nephropathy and severe trauma. Nephrectomy in children may also be for malignant conditions of the kidney and adrenal glands. The major indications for nephrectomy vary in different parts of the world and in different age groups and sexes<sup>2</sup> with some recording more benign conditions<sup>3</sup> and others more of malignancies<sup>4,5</sup>.

Traditionally, nephrectomy is undertaken by open surgery and this is still the practice in our unit as well as other centers in Low and Middle Income Countries (LMICs)<sup>6,7</sup>. More recently, however minimally invasive laparoscopic surgical techniques have been increasingly applied for nephrectomy in High Income Countries (HICs)<sup>8,9,10,11</sup>. There has also been a growing interest in the use of nephron-sparing surgery for selected patients. However, late presentation in our environment, especially for malignant diseases of the kidney,

is still a daunting challenge, making such renal conserving surgeries uncommon.

## Materials and Methods

This is a retrospective review of all children aged ≤ 16 years who had nephrectomy from January 2007 to December 2016. We requested the medical records department to permit us to have access to patients data from the theatre records and data. The medical records were reviewed and data retrieved with emphasis on age, sex, side of nephrectomy, duration of symptoms before presentation, indication for nephrectomy, post-operative complications during admission, length of hospital stay, mortality during admission. The indications were broadly classified into malignant and non-malignant conditions. Data entry and analysis were done with Statistical Package for Social Sciences (SPSS version 15.0 Chicago Illinois, USA). The results are expressed as means ± standard deviation, ratios, percentages, charts and tables

## Results

There were 52 nephrectomies (35 for malignant conditions and 17 for non-malignant conditions) carried out in 32 males and 20 females with a male to female ratio of 1.6:1. Mean age at nephrectomy was 5.1 ± 3.66 years. Mean duration of

symptoms before presentation was 7.6 months (4hours–42 months). There were no bilateral cases in malignancy. 34 of the malignancies were Wilms tumor while 1 was renal rhabdomyosarcoma. The non-malignant conditions were neglected pelvi-ureteric junction obstruction with loss of renal function on intravenous urography (9), large multicystic dysplastic kidneys with non-function on intravenous urography (4), renal trauma with pedicle avulsion (1), posterior urethral valve with no function of one kidney on renal scintigraphy (1), duplex system with non-functioning upper pole moiety (2). In the first five year period 27 nephrectomies were done while 25 nephrectomies were done in the second five year period. Fifty-nine percent (59%) of the nephrectomies were on the left and 41% on the right. Mean duration of hospital stay was 31.78 ± 16.32 days (range 7-66 days).

Male to Female ratio for malignancy was 0.75:1. Malignancy was an indication for all nephrectomies in females (20/35). 60% (21/35) of all malignancies and 54% (28/52) of all nephrectomies were in children aged 1-5 years.

Mean follow-up duration was 25.82 ± 34.12 weeks (range 0-156 weeks). There was only 1 nephron-sparing surgery in a patient with duplex renal system and nonfunctioning upper moiety (1.9%). All others had total nephrectomy. Complications of treatment included stitch reaction (2), small bowel volvulus and bowel gangrene (1), hypertrophic scar (1).

Three (3) mortalities in 2 males and 1 female were noted while on initial hospital admission (mortality rate of 5.8%): 2 with Wilms tumor died intra-operatively on the operating table and one neglected PUJ obstruction died of an undetermined cause in the early postoperative period.

Table 1: Age and sex distribution of nephrectomy patients

	Male	Female	Total	Percentage
< 1year	6	2	8	15.4
1-5 years	16	12	28	53.8
6-10years	4	4	8	15.4
11-16years	6	2	8	14.4
Total	32(61.6%)	20(38.4%)	52(100%)	100

Table 2: Sex and indications for nephrectomy

	Male	Female	Total	Percentage
Wilms Tumor	15	19	34	65.5
Renal rhabdomyosarcoma	-	1	1	1.9
Pelvi-ureteric Junction (PUJ) Obstruction	9	-	9	17.4
Multicystic Dysplastic Kidney (MCDK) Disease	4	-	4	7.6
Renal trauma	1	-	1	1.9
Posterior Urethral valve with no function in one kidney	1	-	1	1.9
Duplex system	2	-	2	3.8
Total	32(61.6%)	20(38.4%)	52(100%)	100

Table 3: Age and indications for nephrectomy

	< 1year	1-5years	6-10 years	11-16years	Total
Wilms Tumor	5	21	6	2	34
Renal rhabdomyosarcoma	-	-	1	-	1
Pelvi-ureteric Junction(PUJ) Obstruction	3	1	-	5	9
Multicystic Dysplastic Kidney(MCDK) Disease	-	4	-	-	4
Renal Trauma	-	-	-	1	1
Posterior Urethral valve with no function in one kidney	-	1	-	-	1
Duplex system	-	1	1	-	2
Total	8	28	8	8	52

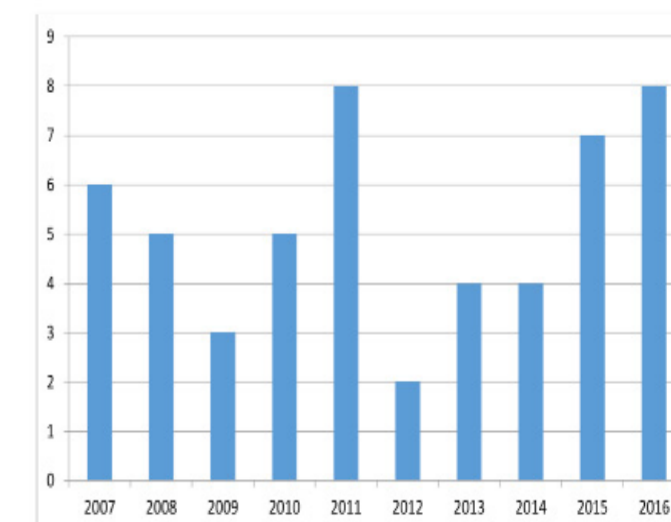


Figure 1 : Bar chart showing number of nephrectomies performed per year from January 2007 to December 2016

## Discussion

In this series, 67% of the nephrectomies were for malignant renal conditions and 33% for benign conditions. This was similar to findings by Bouhafs et al<sup>7</sup> in Morocco where 62.5% of the nephrectomies were for nephroblastoma. This finding, however, is contrary to many other studies on pediatric nephrectomy from different parts of the world, where there were more benign than malignant indications recorded. From Sammon et al<sup>8</sup> in USA, 73.8% of nephrectomies were for benign indications, Daradka<sup>3</sup> in Jordan 59%, Hammad et al<sup>2</sup> in New Zealand 76%, Adamson et al<sup>13</sup> in England 70.5%, Chabchoub et al<sup>6</sup> in Tunisia 78.7%, Featherstone et al<sup>14</sup> in London, 67.5%, Nggada et al<sup>15</sup> in Nigeria 60.3%. However, in some studies on indications for nephrectomy in the adult population in both developed<sup>16,17</sup> and Low and Medium Income Countries (LMICs)<sup>4,5</sup> more malignancies were noted as indication for nephrectomy in adults. This may be due to the higher incidence of malignancy in older patients and the higher proportion of children who present with congenital malformations<sup>12</sup>. One may not readily explain why malignancy was the major indication for nephrectomy in our pediatric population just like adults in same environment. However, this may be related to absence, in this study, of key non-malignant indications like complications of vesicoureteric reflux (VUR) and renal stone

disease which were major indications in some other studies<sup>3,13</sup>. In this study, the malignant cases were mainly Wilms tumour (96.5%). Wilms tumour was also the commonest malignant indication for nephrectomy in some other studies<sup>3,15,18</sup>. Neuroblastoma featured prominently as an indication for nephrectomy in another study<sup>3</sup>, but in our study there were no neuroblastomas as indications for nephrectomy. This may be due to late presentation of neuroblastoma cases in our environment when they are no longer resectable. There was one renal rhabdomyosarcoma in this study and this agrees with the rarity of such renal tumour which exists in literature as mainly case reports<sup>19,20</sup>.

In this study, non-malignant cases were mainly late-presenting pelvi-ureteric junction (PUJ) obstruction with loss of function on intravenous urography. Neglected PUJ obstruction is also most common non-malignant indication in some other series<sup>2,21</sup>. However, in some others complications of vesicoureteric reflux (VUR) were the most common benign indications<sup>1,3,13</sup>. Other non-malignant indications were enlarging multicystic dysplastic kidney (MCDK) disease, Posterior Urethral Valve (PUV) with unilateral non-functioning kidney, duplex renal system with non-functioning upper moiety, renal trauma with pedicle avulsion. It is important to note that complicated VUR, which is generally more common in females<sup>3</sup>, was not an indication for nephrectomy in this study. This may be related to the lower incidence of VUR and hence its associated complications in blacks when compared to Caucasians<sup>22</sup>. Since VUR is the most common benign condition in some series and also more common in females, this finding may explain why benign indications were less likely in females in this study. In the current study, however, non-malignant indications were seen only in males and these were mainly complicated PUJ obstruction. It must be noted that PUJ obstruction is generally seen more in males than females<sup>3,23</sup>.

The second most common non-malignant indication in this study is enlarging multicystic dysplastic kidneys (MCDK) with non-function on IVU. MCDK was the most common indication for nephrectomy in some studies which also recorded reducing incidence of PUJ obstruction as an indication for nephrectomy<sup>12,14</sup>. In another study<sup>15</sup>, chronic pyelonephritis was the most common non-malignant indication, but this was not an indication in the current study.

Though PUV is the most common cause of bladder outlet obstruction in children only one child (1.9%) had nephrectomy due to PUV-associated complications. Similarly another study<sup>3</sup> recorded that 2.5% of nephrectomies were for PUV-associated complications. This may be explained by the fact that most late-presenting cases of PUV may have developed chronic kidney disease and end stage renal disease where various renal substitution strategies, and not nephrectomy, are employed in their treatment. In some other studies, stone disease was the major non-malignant indications for nephrectomy<sup>6,11,24</sup>. It is noteworthy that no nephrectomy due to renal stone disease nor renal tuberculosis was observed in this study.

Early investigation of children suspected to have urological malformations, or who present with urinary tract infection, will encourage early diagnosis and reduce incidence of nephrectomies for non-malignant conditions<sup>25</sup>. In this way most cases of PUJ obstruction will present earlier for monitoring and possible pyeloplasty instead of presenting very late for nephrectomy. In a report by Ocheke et al in

Cape Town no nephrectomy was done for PUJ obstruction despite the substantial number of prenatal diagnoses of PUJ obstruction<sup>23</sup>. Most studies had non-malignant indications for nephrectomy more in females and malignancy more in males<sup>3,11,24</sup>. In this series however all nephrectomies in females were for malignancy and 59% of all malignancies were in females. All non-malignant conditions were in males.

In the current study, we had only one emergency nephrectomy following renal trauma with pedicle avulsion in a 15 year old male who had low-velocity penetrating abdominal trauma. This corroborates with findings in other studies where abdominal trauma was an uncommon indication for nephrectomy<sup>3,4,5,12</sup>. Though our unit is in a teaching hospital, mean age at nephrectomy of 5.1±3.7 years is higher than 3 years Sammon et al recorded for Teaching Hospitals in the USA<sup>8</sup>. This may be attributed to late presentation generally seen in LMIC's. However, this mean age is comparable to 6 years for Non-Teaching Hospitals in The USA<sup>8</sup>, 4.75 years in New Zealand as reported by Hammad et al<sup>12</sup> and 5 years by Bouhafis et al in Morocco<sup>7</sup>. In the current study, peak age range for nephrectomy is 1-5 years and least in 11-16 years (table 1). In another study by Sammon et al in USA, peak age range at nephrectomy was 0-1 years and least in 6-9 years<sup>8</sup>. In Ekenze et al, nephrectomy peaked at 2-5 years age range<sup>26</sup>. Mean age at nephrectomy for malignancy was 3.4 years in this study while in Daradka et al it was 3.6 years<sup>3</sup>. 55% of all nephrectomies and 66% of all nephrectomies for malignancy occurred in the age range 1-5 years. This is opposed to another study where most (36%) nephrectomies were in 0-1 year age range and the indications were mainly benign<sup>8</sup>.

Overall male: female ratio for all nephrectomies is 1.6:1 comparable to 1.5:1 recorded by Hammad et al<sup>12</sup> and 1.6:1 by Nggada et al<sup>15</sup>. However, our study showed more malignancy in females with M:F ratio for malignancy of 0.75:1. Comparable male to female ratios for malignancy were recorded by Featherstone et al<sup>4</sup> (0.8 : 1) and Eke et al<sup>5</sup> (1:1.09). This is contrary to some studies like Ekenze et al<sup>26</sup> (1.1:1) and Ghalayini et al<sup>27</sup> (1.3 : 1) where there were more malignancy in males. On the whole, 60% of the nephrectomies were for Wilms tumor. The contribution of Wilms tumor to nephrectomy in this study is very high when compared to 29% recorded by Daradka et al<sup>3</sup>, 26% by Sammon et al<sup>8</sup>, 29.16% by Noura et al<sup>28</sup> and 33% by Choubchab et al<sup>6</sup>. Renal scintigraphy was done only in one patient who had PUV to assess differential renal function. He showed 1% function on one side and hence ipsilateral nephrectomy was done. In other patients with non-malignant renal conditions, no function or marked reduction in renal function, were assessed using intravenous urography. Facilities for renal scintigraphy were not readily available in our environment.

Left nephrectomies were done in 55.2% of cases and 44.8% were on the right. The corroborates findings in some other studies<sup>2,10</sup>. Most nephrectomies in PUJ obstruction which is the most common benign condition for nephrectomy were done in the age range 11-16 years(5/9) as shown in table 3.

There was a marginal reduction in total number of nephrectomies from 27 in the first 5 years of the study period to 25 in the second 5-year period (figure 1). In some other studies<sup>2,3,12,28</sup> increases in number of nephrectomies were noted over various time periods while others<sup>29</sup> noted no change. Woldrich et al, studying an adult population, revealed

a significant increase in annual incidence attributable to rise in incidental discovery of renal masses on cross-sectional imaging<sup>30</sup>. The median annual hospital volume in childhood nephrectomy in this study is 8 while in Sammon et al<sup>8</sup> it was 12. Mean number per year was 5.

The mean follow-up duration of 26±37 weeks (range 0—156 weeks) is short. It is encouraged that children with a solitary functioning kidney have long-term follow-up until they reach adulthood because they could experience subtle renal deterioration at that time or later in adult life<sup>31</sup>. Long term follow up is important in early detection of subtle renal deterioration<sup>31</sup>. In 2010, Sammon et al in USA noted that 5.8% of their pediatric nephrectomies had minimally invasive nephrectomy, though open trans-abdominal approach was still the most common route for nephrectomy in their series. In the current study, however, there were no facilities and manpower in our hospital for minimally invasive nephrectomy, during the study period and hence, as in some other studies,<sup>4,6,7,12</sup> all nephrectomies were done via the transabdominal route. In Hammad et al<sup>12</sup>, most partial nephrectomies were for duplex renal systems and there was increase in number of partial nephrectomies done over their study period. In the current study, however, only one partial upper pole nephrectomy was done in a patient with duplex system and non-functional upper pole moiety. The lower pole was preserved in this patient. There was no partial nephrectomy for Wilms tumour in the current study. This may be due to the fact that many patients present late as already documented in a previous study from the same environment<sup>26</sup>. Furthermore, there were no cases of bilateral renal tumors in this study as well as other studies from the same environment<sup>26,33</sup>. Some other authors had cases of bilateral Wilms tumor in their series and the patients subsequently had nephron-sparing nephrectomy<sup>3</sup>.

Major post-operative complications were not common as we recorded stitch reaction in (2) patients, hypertrophic scar in (1) patient. Only one patient who had emergency left nephrectomy following low velocity penetrating abdominal injury had small bowel volvulus with gangrene post-operatively and required re-exploration and bowel resection. Mortality during the period of hospitalization for nephrectomy was 5.8%. This is high when compared to lower 30-day mortality rates of 0.9% in Jordan<sup>27</sup>, 0.8% in India<sup>2</sup>, 1.29% in Pakistan<sup>24</sup> and 0.8% in Spain<sup>34</sup> found in other series. The mortalities were in two Wilms tumor patients (one male and one female) who died on the operating table from intraoperative complications. 1 male who had PUJ obstruction died in the early post-operative period from an undetermined cause.

This study is limited by the fact that number of nephrectomies per year is low, follow-up is short and study is retrospective and done in single pediatric surgery unit. We hereby recommend larger multi-centre prospective studies with long term follow-up periods in assessing indications, pattern and outcome of nephrectomies in our environment.

## Conclusions

In our unit, nephroblastoma is the main indication for pediatric nephrectomy and were the only indications in females. Neglected pelviureteric junction obstruction was the major non-malignant indication and occurred only in males. Most nephrectomies were done within the age range of 1-5

years. Nephron-sparing nephrectomy, major morbidity and re-operation are uncommon. In-hospital mortality from nephrectomy is still high at 5.8%.

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