

# Wilm's Tumor: Ten Year Experience at Kanti Children's Hospital

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## Abstract

**Introduction:** Wilm's tumor is the second most common abdominal tumor in children. It arises from the kidney. The survival of children with Wilm's tumor has improved over the past 25 years. **Objectives:** To study the clinical presentation of Wilm's tumor and evaluate the ten year survival. **Materials and Methods:** A retrospective hospital based study was conducted at Kanti Children's Hospital from March 1998 to February 2008. A total of 60 histopathologically diagnosed children below 14 yr of age were included in the study. **Results:** About 2/3rd (66.5%) presented with abdominal swelling followed by abdominal pain (16.5%) and fever (13.5%). A few children manifested with red colored urine (3.5%). The age of children ranged from one month to 13 years with the mean age of 36 months. Males were affected more than the females (M:F=3:1). Most affected age group was 2 to 5 yrs (41.5%) followed by 1 to 2 yrs (25.0%). Most of the cases were in stage III (36.5%) followed by stage II (33.5%). SIOP protocol was used to treat these children and overall 10 year survival rate was 50.0%. One fifth (20%) of the cases died, 16.5% relapsed and 13.5% lost to follow up. **Conclusion:** Despite severe resource limitations, paediatric oncology unit at Kanti Children's Hospital has been successfully treating Wilm's tumor with the success rate of 50.0%.

**Key words:** Chemotherapy, Outcome, Remission, Wilm's tumor, SIOP.

## Introduction

Wilm's tumor is the second most common abdominal tumor in children. It constitutes about 6.0% of all childhood cancers<sup>1,2</sup>. Its incidence varies worldwide ranging from 5.1 to 10 per million among under 15 children<sup>3</sup>. It arises from the kidneys and it is thought that it develops as a result of abnormalities in the development of metanephric blastema.

The survival of children with Wilm's tumor has improved over the past 2 decades<sup>4</sup>. It is mainly due to the development of multimodal treatment regimen namely surgery, chemotherapy and radiotherapy<sup>5</sup>. It is expected that more than 80.0% of all children with Wilm's tumor have long term relapse free survival with this treatment modality<sup>6</sup>. However, survival still remains poor among high risk groups needing intensive treatment regimen.

Abdominal swelling or abdominal mass is the main complaints that bring most children with Wilm's tumor to hospital. Other clinical features include abdominal pain, gross hematuria, fever, hypertension and intra-abdominal emergency<sup>3</sup>. Wilm's tumor is also believed to be associated with various anomalies. For instance, it is associated with anomalies of kidneys, WAGR syndrome, Beckwith-Weidemann syndrome, hemihypertrophy etc<sup>1</sup>.

The study on Wilm's tumor is in scarce in our country. The report from Kanti Children's hospital showed that Wilm's tumor constituted about 11% of all childhood malignancies<sup>7</sup>. Therefore this study was conducted to study the clinical features of children with Wilm's tumor and to evaluate the ten year survival.

## Materials and Methods

This was a retrospective hospital based study conducted in oncology unit of Kanti Children's hospital, Kathmandu, Nepal from March 1998 to February 2008. A total of 60 children were included in the study.

All children with childhood cancers are registered in the special case record forms at Kanti Children's Hospital. All children with Wilm's tumor were analyzed from the case records according to age, gender, clinical presentation at the time of diagnosis, stage of the disease, histological classification for risk assessment and outcome for this study.

Children with abdominal masses with the suspicion of Wilm's tumor underwent USG abdomen to differentiate from other abdominal masses. CT scan and MRI of abdomen were done when the USG findings were non conclusive. Serum  $\alpha$ -fetoprotein,  $\beta$ -HCG and urinary catecholamine metabolites like vanillyl mandelic acid and homovanillic acid were done to rule out other abdominal tumors like teratoma, neuroblastoma, germinoma, hepatoblastoma, ovarian embryonal carcinoma etc. Chest-rays were performed in all cases to rule out lung metastases and CT scan of brain was done on suspected brain metastases.

All children with Wilm's tumor underwent operation for removal of the mass followed by biopsy for confirmation of diagnosis and biopsy of the removed mass was considered to be the gold standard for diagnosis of Wilm's tumor in this study. Risk groups were classified based on histopathological findings of the biopsied materials as per SIOP classification. Histopathological finding suggestive of Mesoblastic nephroma was considered as having low risk; epithelial, stromal and mixed nephroblastomas as intermediate risk and blastemal, clear cell sarcoma and rhabdoid tumor as high risk.

Histopathologically confirmed cases were treated according to SIOP protocol and outcome was recorded as remission, death, relapse and lost to follow up. A few cases with suspected inoperable Wilm's tumor were given neoadjuvant chemotherapy prior to operation to shrink the tumor mass.

Those who received treatment outside of this centre and who refused treatment were excluded from the study.

## Results

Sixty (60) children with Wilm's tumor who received chemotherapy over 10 year period (between March 1998 to February 2008 i.e. 2055 Magh -2065 Paush) were included in the study. The male to female ratio was 3:1. The age of children at presentation ranged from 1 month to 13 years with the mean age of 36 months.

The distribution of children by age group (Fig 1) as seen shows that the most affected age group was 2 to 5 years followed by 1 to 2 years. Children above 5 years constituted only 17.0% of the total cases.

Clinical features of children with Wilm's tumor at presentation is as shown in Table 1. It is clear from the table that 2/3<sup>rd</sup> of children presented with abdominal swelling followed by abdominal pain and fever. Only a few children manifested with red colored urine. Only 2 cases had lung and one had brain metastases.

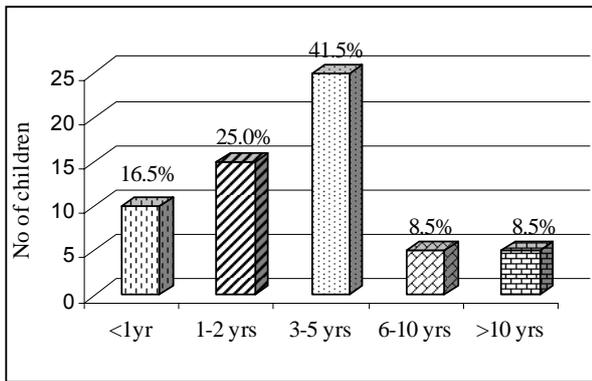
**Table 1:** Showing clinical presentation of Wilm's tumor

Symptoms	Percentage (%)
Abdominal swelling	66.5
Abdominal pain	16.5
Fever	13.5
Red colored urine	3.5

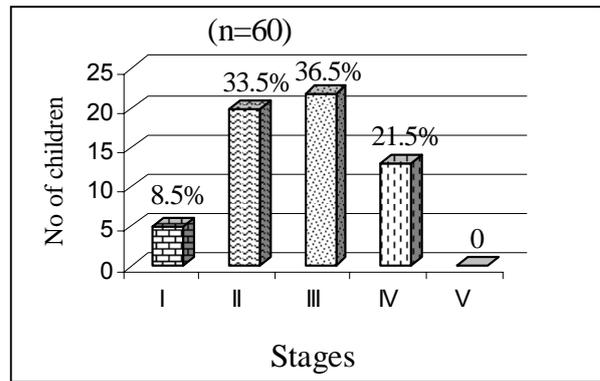
The distribution of children with Wilms tumor as per the staging system is shown in Fig 2. The most common stage at presentation was stage III followed by stage II. No one was in the stage V.

The distribution of children by risk based on histopathological finding is as shown in Fig 3. It is seen that half of the children were classified as intermediate risk tumor followed by low risk tumor. One fifth of the children were classified as high risk tumor.

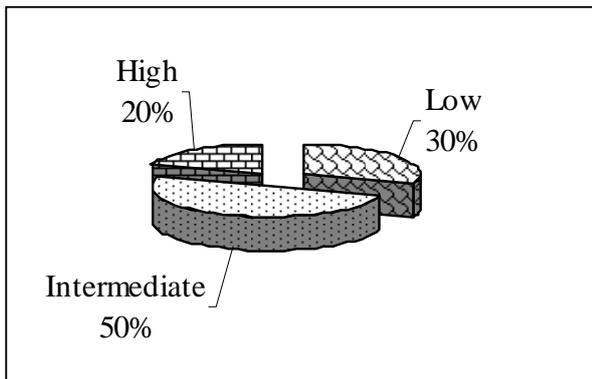
As seen in the Fig 4, an overall 10 year survival rate was 50.0% and that one fifth of the cases died during the treatment. Quite significant numbers of children relapsed and were lost to follow up.



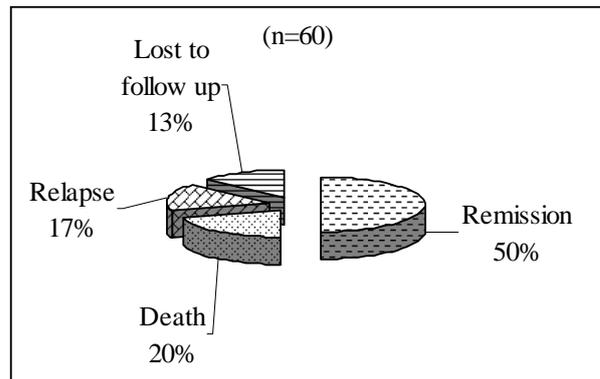
**Fig.1:** Distribution of children with Wilm's tumor by age (n=60)



**Fig. 2:** Staging of children with Wilm's tumor (n=60)



**Fig. 3:** Distribution of children with Wilm's tumor by risk (n=60)



**Fig. 4:** Outcome of children with Wilm's tumor (n=60)

## Discussion

This was a retrospective study to evaluate the clinical features of Wilm's tumor and to assess the outcome. Such study has not been conducted in Nepal before, therefore it is hoped that this study would be beneficial for further studies in this field. Kanti Children's hospital is the only centre where both surgical and chemotherapy is being given to children with Wilm's tumor because of lack of paediatric oncologist in other hospitals treating childhood cancer cases.

The most affected age group was found to be 3 to 5 yrs in this study. This is similar to the study conducted in Lithuania<sup>6</sup>. On the contrary, much younger age group was observed affected most by other studies (18 months and 30 months)<sup>3,4</sup>. It is believed that renal tumors diagnosed in the first 7 months of life have an excellent prognosis<sup>8</sup>.

Wilm's tumor has been reported less frequent in boys than in girls<sup>9,10,11</sup>. However, three fourth of children with Wilm's tumor were male children in our study. Similar observations were made by Rathi et al<sup>4</sup> in India and Peculyte et al<sup>6</sup> in Lithuania. The male to female

ratio observed by Peculyte et al<sup>6</sup> was much lower (28:19) than in our study. In the contrary, Weirich et al<sup>12</sup> found females to be affected more than males. The male predominance in our study could be due to the fact that more attention and care is given to male children in our part of the world.

Triad of flank pain, abdominal mass and haematuria has been considered the basis of clinical diagnosis<sup>13</sup>. However, only 2/3<sup>rd</sup> of children with Wilm's tumor presented with abdominal mass in our study. This is in very much contrast to the study by Peculyte et al<sup>6</sup> who observed that all children presented with abdominal mass. Pain abdomen was also complained by lower number of children in our study than the study done by Peculyte et al<sup>6</sup>. (16.5% vs 27.0%). Fever was complained by even lower number of children. Hematuria was observed in a very small percentage of children in the study conducted by Peculyte et al<sup>6</sup> which is again higher than the finding of our study. It seems that Wilm's tumor should be considered in any child presenting with abdominal mass and it should be differentiated from other masses like neuroblastoma, teratoma, lymphoma,

rhabdomyosarcoma, hydronephrosis, cystic kidney diseases and splenomegaly.

The most common stage at the time of diagnosis in our study was stage III (36.5%). Similar finding was observed by Rathi et al<sup>4</sup> (38.5%) in India. Much higher percentages of children (44.0% and 57.0%) having stage III at diagnosis were observed by other studies conducted in much more developed countries<sup>3,6</sup>. Perhaps, other factors play more important role than education level and economic status of the community in seeking health care.

It was found by our study that fifty percent of children had intermediate risk (standard) tumors which are much lower than the finding by Peciulyte et al<sup>6</sup> (74.0%). One fifth of children had high risk (unfavorable) tumors in our study which is similar to the finding by Peciulyte et al<sup>6</sup> (23.0%). Only about 1/3<sup>rd</sup> children had low risk (favourable) tumors in our study. This is in contrast to the finding by Mulhim et al<sup>3</sup> who observed that 94.0% had favorable histology. That could be one of the reasons for better outcome in their study.

Mortality rates for Wilm's tumor have decreased in most of the developed countries in the last few years<sup>14</sup>. This is most probably due to improvements in the treatment<sup>15</sup>. We do not have such data for comparison in Nepal and it is hoped that this study would serve as a base line for further studies in this field. Fifty percent of our children had remission which is lower than the finding by Peciulyte et al<sup>6</sup>, Mulhim et al<sup>3</sup> and Davidoff<sup>16,17</sup> (73.0%, 88.8% and 90% respectively). This could be due to the fact that supportive care was better in those countries as the most common stage at diagnosis in those countries were also the same (stage III). Their relapse rate was also much lower than our finding (11.0% vs 17.0%). The death rate was less in those two studies (about 2%) as compared to our study (20.0%). There was no one who was lost to follow up in the study by Peciulyte et al.<sup>6</sup> However, quite significant number of children (13.0%) did not appear on the follow up visits in our study. This could be because of low socio-economic condition and low awareness about the condition. Better counseling program for their parents and care givers may increase the follow up visits in our part of the world.

## Conclusion

Despite severe resource limitations, paediatric oncology unit at Kanti Children's Hospital has been successfully treating Wilm's tumor with the success rate of 50.0%.

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**Conflict of Interest:** None

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