# Management of Wilm's Tumour – Our Experience.

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**Eleven patients with Wilm's Tumour were jointly dealt in the Departments of Paediatric Surgery and Paediatrics, King Edward Medical College & Mayo Hospital, Lahore. There were eight males and three females. All the eleven patients had a palpable mass. One had gross haematuria. No appreciable associated abnormalities were noticed. Pre-op chemotherapy was offered to two patients that had a significant reduction in the size of tumour. Surgery was performed in all. Follow up period ranges up to 2 years and 2 months. One died, all of the rest are disease free till now.**

**Key words. Wilm's tumour, management, chemotherapy,**

Wilm's tumour accounts for 6% of all childhood malignanices and up to one third of embryonal tumors in children[1]. It is a curable childhood cancer. The management of Wilm's tumour is a model for multimodal treatment of paediatric malignant solid tumour

The majority of Wilm's tumour presents between 2 – 4 year of age, although it can present at birth. Abdominal swelling and a palpable mass are the most common presenting features. Other findings include hematuria, vomiting, weight loss, hypertension, etc. Wilm's tumour may be associated with aniridia, genitourinary anomalies, hemihypertrophy and Beewith-Wiedmann Syndrome[2].

In this study we reviewed the clinical features, pathological findings and management of children suffering from Wilm's tumour admitted in our departments during the year 2000.

## Patients and methods

This prospective study was carried out, in the Departments of Paediatric Surgery and Paediatrics. All patients who presented with intra abdominal palpable mass were evaluated and a reasonably sure diagnosis of Wilms Tumour was arrived at. Abdominal ultrasonography (USG) was done in all cases as a base line investigation. CT scan of abdomen and intravenous urography (IVU) were performed in 6 cases respectively. Other routine investigations included complete blood count, urine analysis, blood urea, serum creatinine, and serum calcium.

Surgery was performed in all cases. Adjuvant chemotherapy administered in all cases was based on SIOP (International Society of Paediatric Oncology) protocol for Wilm's tumour with the modification that preoperative chemotherapy was not given in all cases.Only two patients were offered preoperative chemotherapy prior to surgery. For stages I & II (negative nodes) favourable histology (FH) daily injections of Actinomycin D x 3 along with weekly injections of Vincristine x 4 followed by 6 weekly pulses consisting of 5 injections of daily Actinomycin D and 2 of Vincristine at weekly intervals were given. Five such pulses were given for stage II (positive nodes) and stage III FH. The treatment consisted of weekly injections of Vincristine x 4 then every 3 weeks x 5 doses; Adriamycin x 5 doses at 3 weekly interval; after this five courses of Actinomycin D and Vincristine at 3 weekly intervals as for stage I & II (negative nodes). Radiation was given to involved fields. For stage IV FH Cyclophosphamide was added and radiation to lungs was given when needed. For unfavourable histology of all stages except stage I protocol similar to stage IV FH with a higher dose of Cyclophosphamide was used.

## Results

A total of 11 patients were dealt in the year 2000. The age ranged between 15 months and eight years. Seven patients were between 2 – 4 years (Fig. 1). Only two were under two years. Mean age was 37.6 months in our series. Males out numbered females in a ratio of 8:3 (73% and 27% respectively). Presenting clinical features are shown in Table 1.. Palpable mass was present in all eleven patients 10 were noticed by the parents, only one tumour was diagnosed by the examining paediatrician while treating unexplained weight loss and pallor. Gross haematuria, which is considered an ominous sign, was encountered in only one patient while microscopic haematuria was present in four.

Table 1: Presenting clinical featues (n=11)

| Clinical features | n= |
|---|---|
| Palpable Mass: | 11 |
| a) Noticed by parents: | 10 |
| b) Noticed by doctors: | 01 |
| Abdominal Distension | 07 |
| Weight loss | 04 |
| Pallor | 05 |
| Gross haematuria | 01 |
| Microscopic haematuria | 04 |
| Fever | 03 |
| Abdominal pain | 02 |
| Hypertension | 00 |

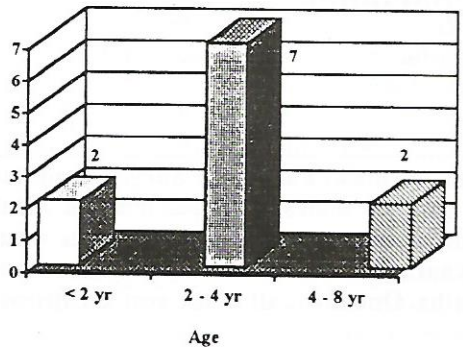



Fig. 1: Age distribution

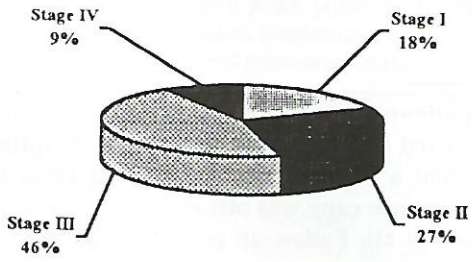



Fig. 2: Staging of Wilm's tumour.

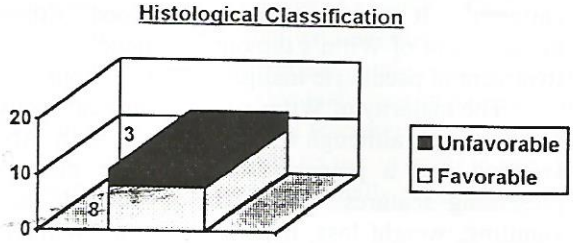



Figure. 3: Histological classification of Wilm's tumour in our patients.

No bilateral tumour was encountered. Seven children had involvement of the right kidney while left sided tumour contributed the rest. One tumour was huge enough to cross the midline. Hepatomegaly was present in one and splenomegaly in two. Intra abdominal lymph nodes were palpable in only one patient.

No associated abnormalities like varicocele, overgrowth syndrome (hemihypertrophy), aniridia or other genitourinary abnormality was noted except hypospadias in one case. Haemoglobin levels ranged from 8.4 – 11.5 gm/dl. There was no other significant haematological abnormality. All patients had X-ray chest to rule out metastases. Abdominal ultrasonography was done in all cases. CT scan was obtained in 8 patients and IVU in six. Right sided lesions were seven in number. No bilateral disease was encountered.

Ten out of eleven patients had unifocal origin. One was diagnosed to be having multiple focal origin of the tumour. Upper pole was involved in seven and lowers pole in four. Pre-op chemotherapy was instituted in two patients (Table 2) which showed a considerable reduction in size.

Table 2: Effect of preoperative chemotherapy.

| Tumour size on USG | Before | After |
|---|---|---|
| 1 | 15.6x9.8x6cm | 12.4x8.1x5cm |
| 2 | 18.2x12.1x8.6cm | 15.6x8.9x6.1cm |

*Operative Results*

Abdominal approach was the standard for all surgeries. Nephrectomy and gross tumour resection was performed in all. Contralateral kidney was examined in all. One patient had inferior vena cava (IVC) involvement. The tumour extension in IVC was removed through a low suction pressure sucker which was applied after venacotomy; the IVC was repaired after wards. Two patients had renal vein involvement. One patient had an intra operative rupture of tumour. The operative staging of the disease is shown in Fig. 2. The pathologist reported two metastatic tensions. One had stage III and the other stage IV disease. Favourable pathology was reported in eight. Three had unfavourable histology (Fig. 3)

*Operative complications*:

Two had post operative chest complications. One had a minor wound infection. One patient presented six month later with post operative adhesive bowel disease which responded to conservative management. Ten patients have survived so far and all of them are disease free. One stage IV patient died of septicaemia during chemotherapy 3 months later table.

**Discussion**

Wilm's tumour is the most common primary renal tumour of children. Developments in chemotherapy, surgery, and radiotherapy have led to dramatic change in the prognosis of most of the patients with this once uniformly lethal malignancy.

Most worldwide registries report a sex ratio of close to one except in USA where this tumour is more common in girls by 22% [3]. Our results showed a marked predominance of boys. Paul et al [4] also reports similar gender distribution in Karachi. This indicates that incidence of Wilm's tumour is more in boys in our population. However the age incidence in our patients corresponds with that reported in the literature. We observed a mean age of 37.6 months as compared to 37 months in white registrants in United States [3].

Majority of children suffering from Wilm's tumour come to attention because of abdominal swelling or

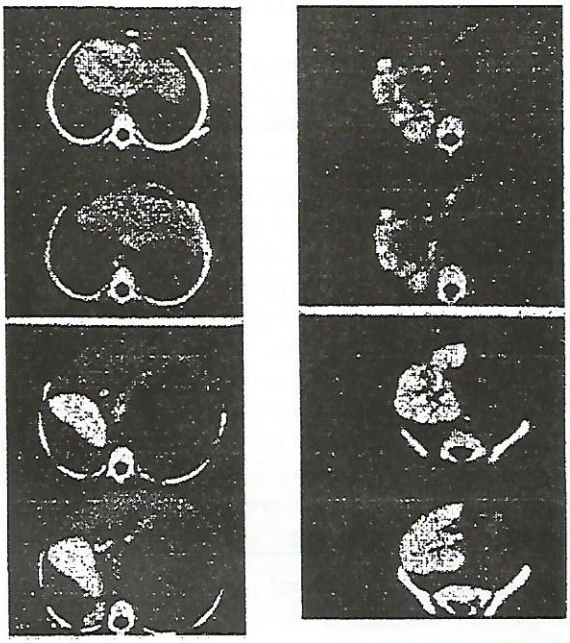
because of a palpable abdominal mass which is most often felt by the mother while changing clothes or diapers or bathing the child. It is reported in 75% of these case [5]. 100% of children in our series had abdominal distension / palpable mass at presentation. None of the patients had hypertension inspite of renal vein involvement. This is in contrast with international literature where 25% of the patients have hypertension [5]. Gross and microspcopic hematuria in our patients was about 25% which correlates well with the reported prevalene of 18 – 25%[6].

Our ultrasonography reports co-relates well with international literature; it is used as the main tool of investigation to delineate and pinpoint the organ of origin [7]. Apart form the solid and cystic nature of the tumour ultrasonography was able to pick up one multifocal lesion which was confirmed on histopathology. The increased number of stage II(27%) and stage III(46%) indicate late presentation of the disease. Ignorance and illiteracy and fear of surgery and seeking medical advice after the visits of hakeems and quacks, play an important role. Paul et al [4] also report similar picture of Wilm's tumour cases from Karachi who presented in higher stages.

Classical pattern of presentation of Wilm's tumour is favourable histology. It comprises of persistent blastema, dysplastic tubules and supporting mesenchyma or stroma. Anaplasia is present in approximately 5% of Wilm's tumour [1]. We saw anaplastic pattern in 27.3% cases which is significantly higher than the reported incidence ($p<0.001$). It is probable that many of our patients may have a poorer long term outcome due to higher stage and unfavourable histology.

We administered pre-operative chemotherapy to 2 patients. A 4-week pre-operative chemotherapy pulse with Vincristine and Actinomycin D helped to reduce the tumour size and probably down staged both the tumours which seemed large and difficult to remove. With pre-operative chemotherapy there was no rupture and these were completely excised.

The demographic, clinical and pathological aspects of children suffering form Wilm's tumour were different from those reported in the literature in the following aspect. There was a predominance of male sex; the abdominal distension / mass was universal initial finding; hypertension was absent in all of the cases; all patients had unilateral disease; and, many patients had advanced and more aggressive (anaplastic) disease. Perioperative chemotherapy also needs further tiral as indicated in European literature and encouraging results in two of our patient.



**Preoperative and postoperative chemotherapy**

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