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Original Article

Paediatrics Section

Clinical Profile and Outcome of Paediatric Wilms Tumour: A Single-Centre Retrospective Cohort Study from Northwest India

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ABSTRACT

Introduction: Wilms Tumour (WT) is the most common paediatric renal tumour affecting children younger than five years of age. The clinical outcome has vastly improved over the years with advancements in treatment protocols, with survival reaching around 90% in the developed world. However, survival remains poor in the developing world. In India, where paediatric oncology still faces multiple challenges, wide variation in outcomes exists, ranging from 48 to 89%.

Aim: To assess the demographic, clinical profile and outcome of WT patients treated at a tertiary-care government medical college in Northwest India.

Materials and Methods: The present retrospective cohort study was conducted in the Department of Paediatrics, Sawai Man Singh Medical College, Jaipur, Rajasthan, India and included 21 patients with WT treated from January 2022 to December 2024. Demographic details, clinical profile, and outcome data were retrieved from hospital records. Follow-up data were collected by telephone and from the follow-up clinic until December 2025. Statistical analysis was done using IBM Statistical Package for Social Sciences (SPSS) version 25.0. Descriptive statistics were used for demographic details. Overall Survival (OS) was analysed by the Kaplan-Meier method.

Results: Median age of diagnosis was 5 years (range 1.5 years-15 years), with a male predominance (M:F=2.5:1). The commonest presentation was abdominal mass 21 (100%), followed by abdominal pain 4 (19%), hypertension and haematuria 3 (14.2%), fever 1 (4.7%) and breathing difficulty 1 (4.7%). One child had a syndromic presentation. 11 (52.4%) patients were in stage I-II, while 10 (47.6%) had advanced disease. The most common cause of death was treatment abandonment 4 (19%), followed by febrile neutropaenia 2 (9.5%) and postoperative complications 1 (4.7%), respectively. OS was 14 (66.7%) across the entire cohort, with a median follow-up of 24 months.

Conclusion: While OS for paediatric WT is encouraging, reaching 66.7% at a government-run tertiary care centre in the present study, owing to specialist care and extensive financial support, treatment abandonment remains a major issue to address. Strengthening patient and caregiver counselling, along with community education, is essential to address misconceptions and fears surrounding surgical procedures that continue to prevail in society, thereby improving treatment adherence and outcomes.

Keywords: Compliance, Epidemiology, Survival, Treatment adherence

INTRODUCTION

Malignant tumours of the kidney account for 5% of all cancers diagnosed in children under 15 [1], with WT being the most common type. Most cases present with a mean age of three to four years [2]. The male-to-female ratio with unilateral and bilateral WT is 0.92:1 and 0.60:1, respectively [3]. The majority of patients present with an asymptomatic abdominal lump found incidentally; however, abdominal pain, haematuria, and fever may be found in some. WT patients frequently have underlying congenital anomalies linked to aniridia, genitourinary malformations, and mental retardation (W- Wilms tumour, A- Aniridia (absence or severe underdevelopment of the iris), G- Genitourinary abnormalities, R- Retardation of development/intellectual disability syndrome), Denys-Drash syndrome, and Beckwith-Wiedemann syndrome [4].

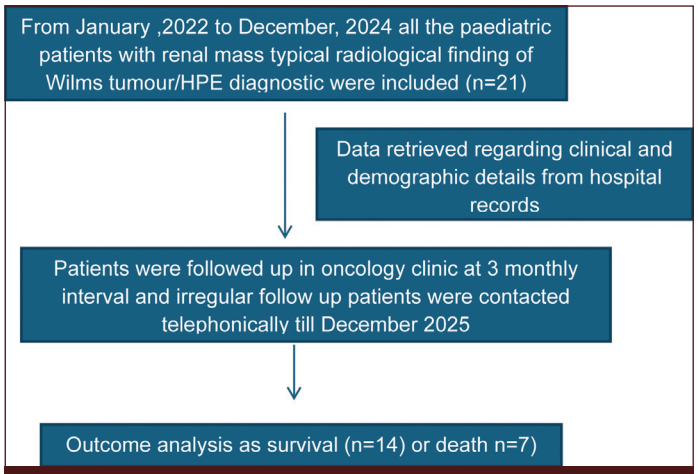
Most of the data pertaining to WT has emerged from two major cooperative group trials, SIOP (International Society of Paediatric Oncology)- neoadjuvant chemotherapy first, and Children's Oncology Group (COG) surgery first, both showing excellent survival [5]. OS ranges from 70 to 97% in high-income countries, 61-94% in upper-middle-income countries, 0%-85% in Lower-Middle-Income Countries (LMIC), and 25-53% in low-income countries [6].

India is a large and demographically diverse country, with substantial regional disparities in healthcare access and outcomes. In a recent systematic review, OS of WT in India is found to be ranging between 48 and 89% [7]. Inferior outcomes are attributable not only to disease-related factors but also to significant socioeconomic and healthcare system challenges, such as a heavy disease load, treatment-related mortality, scarcity of surgical skills, and treatment abandonment due to financial constraints, poor literacy and seeking alternative medication [8,9].

There is no data in the literature on the demographic and clinical profiles and outcomes of WT from Rajasthan, the largest state in India with distinct demographics. The present retrospective study was conducted to fill this gap and assess the demographic and clinical profiles and outcomes of patients with WT using an International Society of Paediatric Oncology based approach [5] at the largest tertiary care centre in Rajasthan, India.

MATERIALS AND METHODS

This single-centre, retrospective cohort study was conducted in the Department of Paediatrics at Sawai Man Singh Medical College, Jaipur, Rajasthan, India. The present study was planned in January 2025 and included all consecutive patients with newly diagnosed WT diagnosed between January 2022 and December 2024. Patients were followed-up till December 2025 for outcome analysis. Anonymised data were collected in accordance with standard Institutional guideline to guarantee patient confidentiality and privacy [Table/Fig-1].



[Table/Fig-1]: STROBE flow diagram.

Inclusion and Exclusion criteria: All newly diagnosed paediatric patients with WT managed during the study period were eligible for inclusion. Patients whose treatment started before the study period, who were lost to follow-up before completion of treatment or for whom outcome data were unavailable were excluded from the final analysis.

Sample size: A total of 21 consecutive eligible patients fulfilling the inclusion criteria during the study period were included.

Study Procedure

Data collected included demographic characteristics, clinical presentation, laboratory investigations, imaging findings, histopathological diagnosis, treatment details (including surgery and chemotherapy), treatment-related complications, and clinical outcomes. Diagnosis was established by typical radiological findings and confirmed by Fine-Needle Aspiration Cytology (FNAC) or trucut biopsy in cases with atypical imaging findings. Staging workup included Ultrasonography (USG), whole-abdomen Contrast-Enhanced Computed Tomography (CECT), and Computed Tomography (CT) of the thorax. Staging was done as per SIOP staging for paediatric WT [5]. Informed consent was obtained prior to the start of chemotherapy. All patients received preoperative (neoadjuvant) chemotherapy according to the SIOP protocol, consisting of injection vincristine 1.5 mg/m^2 (maximum dose 2 mg) weekly for four weeks and injection actinomycin D 45 µg/kg (maximum dose 2 mg) at weeks one and three in localised disease. The duration of neoadjuvant chemotherapy was 6-9 weeks, including the AVD regimen (actinomycin D, vincristine and doxorubicin). All the drugs were administered intravenously. A reassessment CT scan was done after completion of neoadjuvant chemotherapy. Surgeons were informed of the potential timing of surgery, per standard protocol, to avoid treatment abandonment at the time of surgery. The surgical procedure for a unilateral WT was a transperitoneal radical nephroureterectomy with ipsilateral lymph node sampling, while bilateral partial nephrectomy was done in a stage V case. Histopathological findings and initial imaging were evaluated to stratify patients for chemotherapy and to determine indications for radiotherapy. Patients were stratified into low, intermediate, and high-risk groups depending on staging and histopathology [5]. Risk-stratified postoperative chemotherapy was delivered based on the SIOP protocol. After completion of therapy, patients were regularly followed up at 3-month intervals by Ultrasonography (USG) abdomen in Outpatient Department (OPD), and those who were not on regular follow-up were contacted telephonically.

STATISTICAL ANALYSIS

The IBM SPSS for Windows, version 25.0 was used for the statistical analysis. Descriptive statistics were used to describe patient demographics and clinical features. Continuous variables were expressed as median and range, as appropriate. Categorical variables were shown as frequencies or percentages. The median follow-up duration was calculated from the date of diagnosis to the date of death or the last follow-up, whichever occurred first. OS was defined as the interval from diagnosis to death from any cause. Patients who were alive at the last follow-up were censored on the date of their last documented follow-up. OS was estimated using the Kaplan-Meier method. Survival probability was plotted over time from the date of diagnosis, and censored observations were indicated on the Kaplan-Meier survival curve.

RESULTS

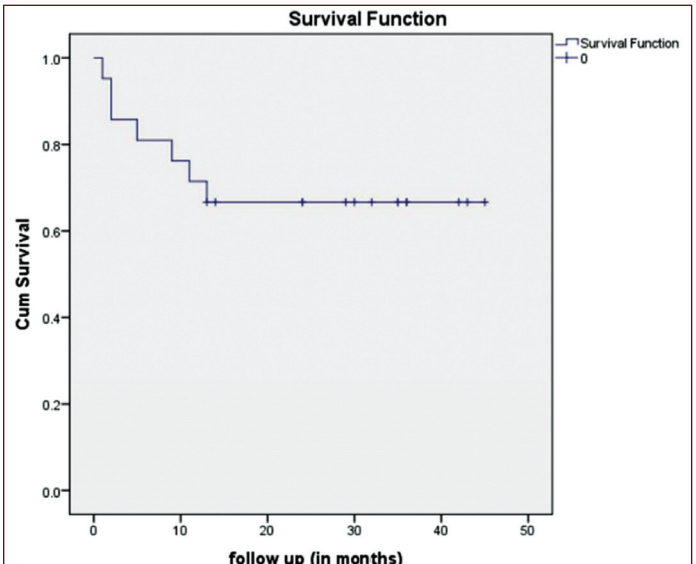
A total of 21 patients were analysed in the present study. The median age at presentation was five years (age range 1.5-15 years). The gender distribution showed male predominance with a sex ratio of 2.5:1. One child had syndromic features including hypospadias and chordee; the rest were non syndromic. Abdominal mass was the most common presenting feature (100%), followed by abdominal pain 4 (19%), hypertension and haematuria 3 (14.2%), fever 1 (4.7%) and breathing difficulty 1 (4.7%). The diagnosis was based mainly on clinical, histopathological, and radiological evidence. Average tumour volume was 779 mm^3 (range 220-2190 mm^3). The most common stage at diagnosis was stage II 9 (42.8%), while 47.62% of patients presented with advanced disease. On histopathology, triphasic WT was most common 7/21 (33.3%), while anaplasia was seen in three cases (14.3%). On histopathological risk stratification, 14 (66.7%) of cases were intermediate risk, and 2 (9.5%) cases were high risk [Table/Fig-2].

Parameters		n (%)
Age	1.5 years to 15 years	Median age 5 years
Male and Female ratio		2.5:1
Syndromic association	Syndromic	1
	Non syndromic	20
Presenting complaints	Abdominal mass	21 (100%)
	Abdominal pain	4 (19%)
	Hypertension and haematuria	3 (14.2%)
	Fever	1 (4.7%)
	Breathing difficulty	1 (4.7%)
Side	Right kidney	11 (52.4%)
	Left kidney	9 (42.9%)
	Bilateral	1 (4.7%)

Stage at diagnosis	I	2 (9.5%)
	II	9 (42.8%)
	III	2 (9.5%)
	IV	7 (33.3%)
	V	1 (4.7%)
Histopathology	A. High-risk	
	Diffuse anaplasia	2
	B. Intermediate-risk	
	Focal anaplasia	1
	Triphasic	7
	Epithelial predominant	4
	Stromal predominant	1
	Foetal rhabdomyomatous	1
	C. Not available*	5
Overall Survival (OS) as per stage	I	2/2 (100%)
	II	6/9 (66.7%)
	III	1/2 (50%)
	IV	4/7 (57%)
	V	1/1 (100%)
	Treatment abandonment	4 (19%)
Cause of death	Febrile neutropaenia	2 (9.5%)
	Post-op complications	1 (4.7%)
	Survived	14 (66.7%)
Overall outcome	Expired	7 (33.3%)

[Table/Fig-2]: Demographic, clinical and outcome data of Wilms Tumour (WT).
* Not available for patients who refused surgery or expired.

Survival probability was plotted over time from diagnosis, with censored observations indicated on the curve. An initial decline in survival during the first year after diagnosis was primarily attributable to treatment abandonment and therapy-related complications. Thereafter, the survival curve plateaued, with an OS of 66.7% maintained throughout the follow-up period, demonstrating favourable long-term survival among patients who successfully completed treatment [Table/Fig-3]. The most common cause of



[Table/Fig-3]: Kaplan-Meier survival curve showing Overall Survival (OS) among paediatric patients with Wilms Tumour (WT) included in the retrospective cohort. Overall survival in the study population was 66.7 % (n=14) and remained unchanged over a median follow-up of 24 months.

early mortality was treatment abandonment 4 (19%), followed by febrile neutropaenia and postoperative complications in 2 (9.5%) and 1 (4.7%) of cases, respectively.

DISCUSSION

The WT is the most common renal malignancy in children. Despite significant improvement in survival rates in high-income countries, outcomes in LMICs, including India, continue to lag. These challenges arise from limited rural healthcare access, resulting in delayed diagnosis and malnutrition; socioeconomic factors hinder access to specialised paediatric cancer care and treatment abandonment. The demographic and social factors are major contributors to mortality due to non medical reasons [6]. For the aforementioned reasons, the authors analysed the study data to look for the demographic and clinical profile of the study patients and clinical outcomes.

In the present study, the median age at diagnosis was comparable to the commonly reported age range of 3-5 years in the literature. Breslow N et al., reported that WT predominantly affects young children with peak incidence around 3-4 years of age [2]. Similar findings were reported by Saha S et al., from north India and John R et al., from south India, where most children presented before five years of age [8,10]. The predominance of younger age at diagnosis reflects the embryonal origin of the tumour and is consistent across most Indian and international studies.

Study demonstrated male predominance with a male-to-female ratio of 2.5:1. This is higher than that reported in international literature, where the gender distribution is usually equal or shows slight female predominance. Breslow N et al., observed a male-to-female ratio close to 1:1 in unilateral tumours [2]. Similar Indian studies have shown slight male predominance [8,10]; however, the higher M:F ratio in this study may reflect gender bias in healthcare-seeking behaviour and preferential treatment access for male children [11].

Abdominal mass was the most common presenting complaint in the study cohort, seen in 100% of patients, which shows a late presentation of the disease. The present study finding is comparable to studies by Wani SQ et al., and Saha S et al., [3,8]. A study by Guruprasad B et al., reported that 90% of their patients presented with abdominal swelling or mass [12], while Singh P et al., reported 50% [13]. Additional symptoms such as abdominal pain, haematuria, hypertension, and fever were observed less frequently in the present study patients, which is also consistent with previous reports [8,12]. Hypertension and haematuria are known manifestations of renal vascular compression and renin secretion by the tumour.

Right-sided renal involvement (52.4%) was slightly more common than left-sided disease (42.9%) in the present study, while bilateral involvement was observed in one patient, similar to another Indian study [12]. Similar laterality patterns have been reported previously, though no significant predilection for either side has been consistently established in the literature.

Only one child in the present cohort had associated syndromic features. Syndromic associations such as WAGR syndrome, Denys-Drash syndrome, and Beckwith-Wiedemann syndrome are well documented in WT. Dome and Huff reported that approximately 10% of WT patients may have associated congenital anomalies or genetic syndromes [4]. The low prevalence in the present study may be due to the small sample size and limited genetic evaluation facilities.

A major finding in the present study was the high rate of treatment abandonment (19%), which emerged as the leading cause of mortality. Two patients refused surgery, and one patient abandoned

treatment after surgery. The perioperative period was found to be the critical time point where most treatments were interrupted. Abandonment remains a critical challenge in paediatric oncology in developing countries and has been consistently associated with poor survival outcomes. Financial constraints, long travel distances, poor awareness, and social factors contribute significantly to therapy discontinuation. Similar observations were reported by Cunningham ME et al., and Saha S et al., emphasising that non medical causes significantly influence survival in LMICs [6,8]. It is imperative that a multidisciplinary team be included at the inception of the treatment. Strengthening counselling services, improving social support systems, and implementing patient-navigation programs may help reduce abandonment rates. Earlier review on treatment abandonment has shown that maternal education played a significant role in the choices made for child treatment [9].

Febrile neutropaenia accounted for 9.5% of deaths in the present study cohort. Although WT chemotherapy protocols are generally associated with moderate haematological toxicity; malnutrition, delayed presentation, and lack of trained personnel and timely intervention may predispose children to severe infections in resource-constrained settings. Comparable treatment-related mortality due to infections has been reported in a study by Sen S et al., and remains substantially higher than in developed countries [14,15]. Special training of nursing staff in the management of immunocompromised patients could certainly address the lacunae. Postoperative complications contributed to mortality in one patient, highlighting the importance of specialised paediatric surgical expertise and perioperative intensive care support.

The OS in the present study was 66.7% , which is lower than the survival rates reported in developed countries, where OS exceeds 85-90% with multimodal treatment protocols [5]. In the present study, 10 patients (47.6%) were in the advanced stage of the disease (stages III, IV and V). This delayed presentation and poor nutrition may have led to poor tolerance of intensive chemotherapy and radiotherapy in our patients. Cunningham ME et al., highlighted significant global disparities in WT outcomes, particularly in LMICs [6]. However, the present study findings are comparable to several Indian studies. Srinivasan S et al., in a systematic review from India, reported OS ranging from 48% to 89%, reflecting marked heterogeneity in outcomes across centres [7]. Saha S et al., from North India reported survival outcomes close to those observed in the present study [8].

Overall, the present study findings suggest that survival outcomes for WT have improved with standardised, protocol-based multimodal therapy, even in resource-limited settings in India. In view of a different demography and social factors related to our state, which potentially contributed to non medical causes of death, these need to be addressed. Further improvement depends on reducing treatment abandonment, strengthening supportive care services, improving nutritional rehabilitation, and ensuring timely multidisciplinary management.

Limitation(s)

The present study has several limitations. First, it was a retrospective single-centre study with a relatively small sample size, which limits the generalisability of the findings to the broader population. Detailed molecular and genetic profiling could not be performed due to limited resource availability, thereby limiting the correlation between genetic factors and clinical outcomes. In addition, socioeconomic factors contributing to treatment abandonment could not be analysed comprehensively. Variability in supportive care, nutritional status, and follow-up compliance may also have influenced treatment outcomes. Despite these limitations, the study provides valuable insight into the clinical profile and challenges affecting outcomes of paediatric WT in a tertiary care centre in northwest India.

CONCLUSION(S)

In conclusion, the present study highlights that favourable outcomes can be achieved when WT cases are managed with standardised multimodal therapy, especially the SIOP-based protocol, as a substantial proportion of patients present at an advanced stage due to delayed diagnosis and referral, rendering upfront nephrectomy infeasible. Treatment abandonment and infectious complications are the major contributors to mortality in resource-limited settings, which underscores the need to increase awareness among communities and healthcare providers to facilitate early diagnosis and timely referral. Additionally, strengthening supportive care, optimising nutritional rehabilitation, and implementing structured counselling programs are essential for reducing treatment abandonment and improving overall treatment outcomes.

Larger prospective studies, including both private and public institutions from India, are required to better understand regional disease patterns and optimise management strategies for children with WT.

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