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Pediatric Wilms tumor: Insights from Benghazi children hospital on case characteristics and outcome

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Abstract

Wilms tumor (nephroblastoma) is the most common abdominal and renal pediatric cancer, accounting for over 90% of kidney tumors in children. This study aims to enlighten the local health environment with the characteristics and outcomes of Wilms tumors affecting children treated at Benghazi Children Hospital. This retrospective, descriptive study analyzed the records of patient files between 1995 and 2023. The data included sex and age at diagnosis, associated congenital abnormalities, clinical stage, tumor size, treatment, and vital status. Statistical analysis was performed using SPSS, with categorical data compared using Chi-square or Fisher's exact test. Among the 74 patients included in the study, the mean age was 3.7 years, with male predominance of 59.5%. Most patients presented with advanced stages of the disease, with 32.4% at stage II and 27% at stage IV. The right side was the most frequent site of the tumor lesion representing 43.2%. The overall survival rate was 90.5%, with no significant difference in outcomes based on age, sex, or side of the lesion. However, the stage of the tumor was a significant prognostic factor ($P = 0.025$). Wilms tumor in Benghazi predominantly affects young children under six years old, with a slight male predominance. Advanced stages at presentation make it a crucial prognostic indicator and highlight the need for raising awareness, early detection, and intervention.

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Introduction

Wilms tumor, also known as nephroblastoma, is the most common abdominal and renal pediatric cancer. It accounts for over 90% of all kidney tumors in children and is the fourth most common cancer in children overall (Leslie *et al.*, 2023). Typically, it is detected in children younger than five years old, the median age of diagnosis being 3.5 years (Chu *et al.*, 2010; Ko *et al.*, 2009).

Wilms tumor can also be diagnosed in adolescents or adults, but this is extremely rare, representing less than 1% of all renal tumors (Szychoł *et al.*, 2014). Clinically, Wilms tumor usually manifests as an asymptomatic abdominal lump, often noticed by parents or caretakers.

Up to 25% of patients may present with gross hematuria, abdominal pain, or hypertension (Dénes *et al.*, 2013). Wilms tumors generally appear sporadically, with only 1-2% of patients having a family history of the illness (Ikhuorah *et al.*, 2023). The tumor usually arises in a single kidney, with synchronous bilateral or multifocal tumors occurring in approximately 10% of patients, typically presenting at an earlier age (Szychoł *et al.*, 2014). The standard treatment approach includes a combination of surgery and chemotherapy, with radiotherapy added for high-risk patients (Lopes *et al.*, 2017). Due to the collaborative efforts among surgeons, pediatricians, pathologists, and radiation oncologists, the overall cure rates exceed 85% (Spreatico *et al.*, 2006). Over 50 susceptible gene disorders have been identified and implicated for the tumor development, accounting for about 5% of Wilms tumor cases (Theilen *et al.*, 2022).

This study aims to raise awareness among health decision-makers for improved planning and early detection of Wilms tumor and to assist healthcare providers in making informed clinical management decisions.

Materials and methods

We conducted this descriptive, retrospective study at the Hematology and Oncology unit of Benghazi Children Hospital. We examined all patient files and extracted the

information on patients diagnosed with Wilms tumors from 1995 to 2023. The data recorded from the files included sex, age at diagnosis (in months), associated congenital abnormalities, date of diagnosis, clinical stage of the disease, tumor size, treatment hospital, and vital status. We categorized the clinical stage of the disease into five groups (I, II, III, IV, or bilateral = V).

Analysis

Results were expressed as mean \pm standard deviation (SD) or number and percentage. Categorical data were compared using the chi-square test or Fisher's exact test, with a p -value $<.05$ considered significant. Statistical analysis was performed using SPSS (version 25 for Windows).

Results

Out of all the confirmed 74 patients with Wilms tumors, 44 (59.5%) were males and 30 (40.5%) were females (Fig. 1). The children's ages ranged between 2 months and 12 years, with a mean age of 3.7 years. The most prevalent age was 1 year, and the peak of the affected age was 1 to 5 years, accounting for 49 (66.2%) cases. The second most affected age was 6 to 10 years, having 15 (20.3%) cases (Fig. 2).

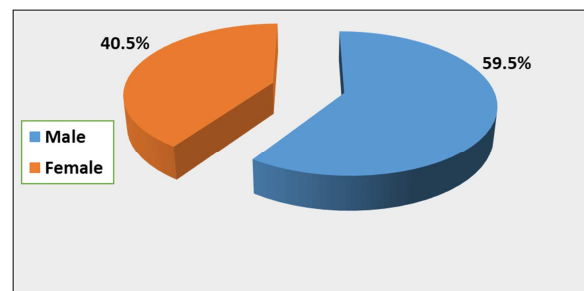


Fig. 1. Distribution of patients according to sex

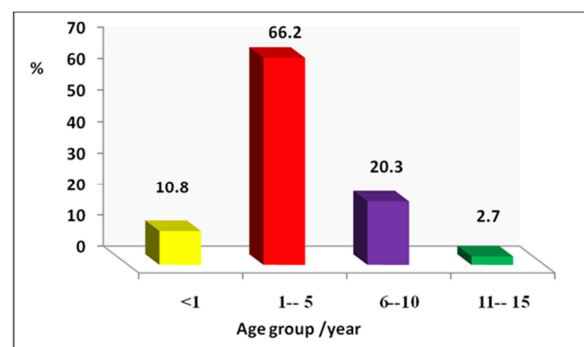


Fig. 2. Distribution of patients according to age

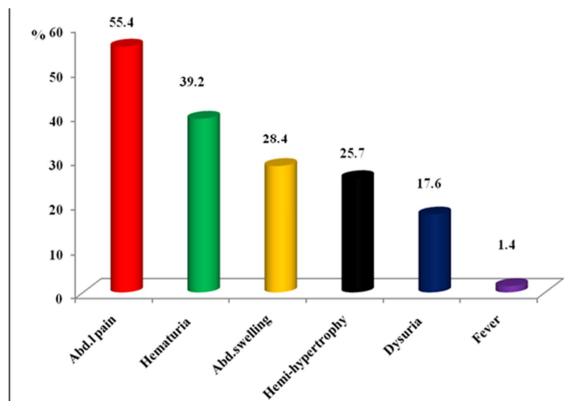


Fig. 3. Distribution of patients according to presentation

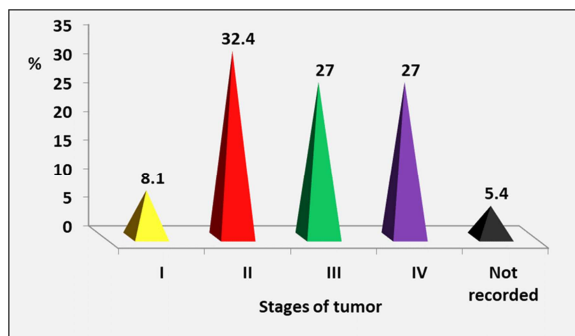


Fig. 4. Distribution of patients according to the stage of the tumor

Table 1. Distribution of patients according to the side of lesion

Side of lesion	No.	%
Right side kidney	32	43.2
Left side kidney	28	37.8
Bilateral	4	5.4
Not recorded	10	13.5
Total	74	100

Table 2. Distribution of patients according to presentation

Presentation	No.	%
Abdominal pain	41	55.4
Fever	1	1.4
Hematuria	29	39.2
Dysuria	13	17.6
Abdominal swelling	21	28.4
Hemi-hypertrophy	19	25.7

The most frequent site of the lesion was the right side (43.2%), followed by the left side (37.8%), and the least was the bilateral lesion, accounting for 5.4% (Table 1). Abdominal pain was the most common symptom, observed in 41 (55.4%) cases, while fever was the least common, seen in 1 (1.4%) case (Fig. 3).

Table 3. Distribution of patients according to outcome

Outcome	No.	%
Died	7	9.5
survive	67	90.5
Total	74	100

Table 4. Comparison of cases outcome in relation to age groups

Age /years	Outcome			
	Died No. %		Survived No. %	
<1	1	12.5	7	87.5
1 - 5	5	10.2	44	89.8
6 - 10	0	0	15	100
11 - 15	1	50	1	50
Total	7	9.5	67	90.5

Fisher's Exact Test= 4.945 P= .134 (Not significant)

Table 5. Comparison of cases outcome in relation to sex groups

Sex	Outcome			
	Died No. %		Survived No. %	
Male	4	9.1	40	90.9
Female	3	10	27	90
Total	7	9.5	67	90.5

X2= .017, df= 1 P= .896 (Not significant)

Table 6. Comparison of cases outcome in relation to side of lesion

Side of lesion	Outcome			
	Died No. %		Survived No. %	
Right kidney	3	9.4	29	90.6
Left kidney	1	3.6	27	96.4
Bilateral	1	25	3	75
Not recorded	2	20	8	80
Total	7	9.5	67	90.5

Fisher's Exact Test=4.229 P = .172 (Not significant)

Table 7. Comparison of cases outcome in relation to disease stage

Stages	Outcome			
	Died No. %		Survived No. %	
I	0	0	6	100
II	0	0	24	100
III	2	10	18	90
IV	3	15	17	85
Not recorded	2	50	2	50
Total	7	9.5	67	90.5

Fisher's Exact Test =8.902 P= .025 (Significant)

Amongst the cases with available data, 8.1% were stage I, 32.4% stage II, 20% stage III, and 27% were in the metastatic stage IV (Fig. 4). The year 2011 recorded the highest number of cases (9.4%), while 2000, 2004, and

2016 registered the least number of cases (1.4% each). The survival rate was 90.5%.

There was no significant difference in outcomes based on the age at diagnosis ($P = .134$), sex ($P = .896$), or side of the lesion ($P = .172$) (Table 4-6). However, there was a significant difference between stages of the tumor and outcomes ($P = .025$) (Table 7).

Discussion

This study assessed the clinical characteristics and outcomes of patients with Wilms tumor at the Benghazi Children Hospital; our data revealed a slight male predominance, similar to a number of international studies, including those from Taiwan, Turkey, Egypt, Morocco, Iran, and Eastern China (Hun *et al.*, 2004; Abd El-Aal *et al.*, 2005; Rais *et al.*, 2016). On the contrary, studies from Jordan, Saudi Arabia, and Europe reported a higher incidence in females (Maher *et al.*, 2014; Rančelytė *et al.*, 2019; Al Mulhim, 1997). The peak age of incidence in our study was 1 to 5 years, with a median age of 3 years, which aligns with some regional studies but is lower than others (Haddadin and Hazza, 2000; Yildiz *et al.*, 2000).

In our study, most children (32.4%) presented with stage 2 and above. This can probably be due to the rapid progression of WT being unnoticed at the early stage. A similar finding was reported in some neighboring countries, as a higher incidence of earlier stages was reported in Egyptian and Jordanian studies (11, 13). Moreover, an 18-year experience report that appeared in a Turkish study manuscript indicated that stage I of WT constituted only 10% of all diagnosed patients, pointing to the late presentation for diagnosis and treatment (Yildiz *et al.*, 2000).

However, in Europe, the Wilms Tumor Study Group (NWTS) and the Society of Pediatric Oncology (SIOP) reported that stage I represented the highest percentage, accounting for 47% in the NWTS report and 61% in the SIOP report (Green, 2004; Reinhard *et al.*, 2004). Late presentation in

the case of our patients could have been caused by a delay in seeking professional medical help since the tumor exhibits a painless pattern, yet it progressively grows.

A higher incidence of hemi hypertrophy was noted in our study (25.7%), which was the commonest malformation reported by the NWTS group (D'Angio *et al.*, 1989). On the contrary, no associated predispositions, namely Wagar or Andria syndromes were noted.

The most frequent clinical feature in our study was abdominal pain and hematuria. This was contrary to the results reported previously, where abdominal pain was the most common initial presenting symptom, but hypertension was the second most frequent feature, and more common than hematuria (Leslie *et al.*, 2023).

Our data on the site of the tumor was similar to that reported by one Wilms Tumor Study Group (Ritchey *et al.*, 2005).

Most relevant, to our objective is that the age, sex, and side of the lesion did not have an impact on the patient's survival. However, the stage of the tumor was a significant prognostic marker, and this was consistent with the Metzger group report (Metzger and Dome, 2005).

Conclusion

This report revealed that Wilms tumor was more predominant in males. In addition, in our setting, the tumor primarily affected young children under the age of 6 years and was most presenting at advanced stages. Finally, the stage of the tumor was a significant prognostic factor for the outcomes in patients with Wilms tumor.

Limitations

Missing data in the patient files due to an inefficient archiving system, probably due to the past political unrest, was the main limitation in our retrospective study.

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