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Wilms' Tumor in Children: An Experience of SIOP Protocol from Tertiary Centre

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Abstract

Background: *Wilms' tumor is the commonest renal .tumor. Survival depends on stage and biologic behavior and economic settings. International Society of Pediatric Oncology (SIOP) has laid down protocol for developing country.*

Methods: *Prospective observational study. aged 1-12 year with wilms' tumor department of Pediatrics IPGMER, Kolkata .from February 2009- August 2012.. All cases are managed according to SIOP 2001protocol. Follow-up for 2 to 4 years Prognosis is noted in terms of local recurrence or metastasis and overall survival.*

Results: *Out of 8 children male 5 and female 3with age of presentation ranging from 3-12 year (median-5 year). Abdominal lump is the main clinical presentation along with hematuria in 2 and hypertension in 1 child. Clinically and histologically 6 of the children was on stage 1 and 2 were on stage 2. . Out of 8 children, 1 child died soon after nephrectomy due to postoperative complications. Remaining 7 children are disease free till date with no evidence of local recurrence or metastasis.*

Conclusions: *SIOP protocol can be used in management of pediatric Wilms' tumor Stage I and II with excellent prognosis in our settings. However more studies are needed to bring SIOP protocol in treatment of Wilms tumor in India or develop own protocol for our country.*

INTRODUCTION

Wilms' tumor (WT) or nephroblastoma is the most common genitourinary malignancy in children and is considered as embryonal neoplasm.[1] It usually presents with unilateral abdominal swelling but there may be atypical presentation like vomiting, hematuria or hypertension.[2] Multimodality treatment has improved the outcome markedly. Currently two major group contributed management of it- National Wilms' Tumor Study Group (NWTSG) the International Society of Pediatric Oncology

(SIOP) protocol. [3,4] The main difference between NWTSG and SIOP is SIOP advocates prenepherectomy Chemotherapy. The advantage of preoperative chemotherapy is reduces the tumor mass, prevent spillage of malignant cell which reduces chance of local and distance metastasis and nephron-sparing surgery is possible. Three is no published literature available regarding the management by SIOP and overall survival (OS) of Wilms' tumor in India.[5] Furthermore , The outcome not only depends on the stage of disease or biologic behavior of disease but also economic setting of countries.[6] Moreover, the main cause behind the treatment failure of Pediatric Malignancy in lower income setting is refusal or abandonment of therapy.[7]
Therefore, SIOP would be more suitable protocol to manage Wilms' tumor in developing countries as infrastructure is not comparable with western countries.[5]

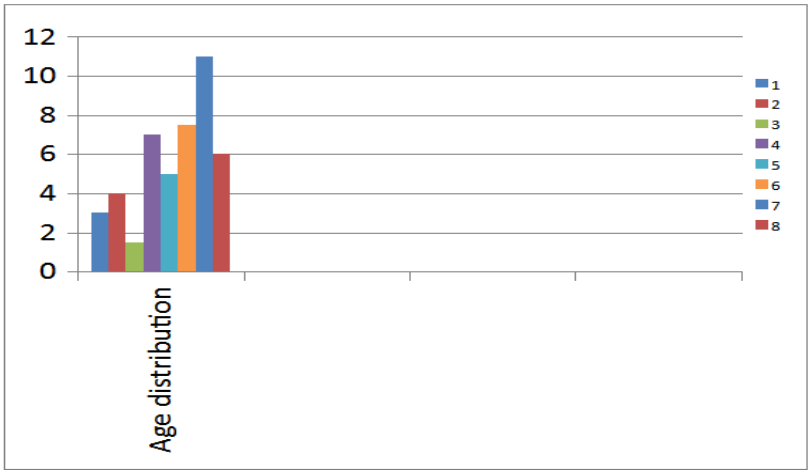
AIMS & OBJECTIVES-

1. To study epidemiological profile, clinical presentation, management and prognosis of pediatric wilms tumor
2. To experience the outcome of SIOP protocol in pediatric wilms' tumor in Tertiary care setting in India

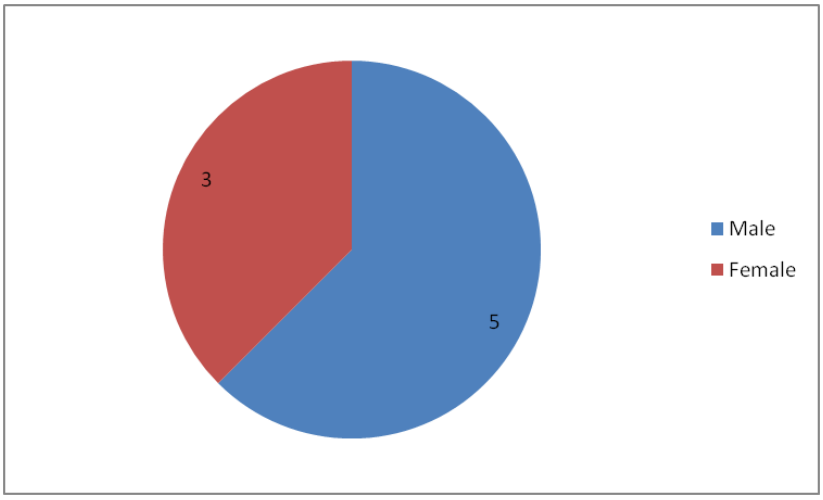
MATERIALS AND METHODS-(STUDY DESIGN)

Type of study- Prospective observational study.
Study population- All patient aged 1-12 year with wilms' tumor attended the inpatient and outpatient department of pediatrics department, IPGMER & SSKM hospital.
INCLUSION CRITERIA – Children aged 1-12 yrs with wilms' tumor, diagnosed clinically and imaging as per SIOP protocol between February 2009 to Dec 2010.
EXCLUSION CRITERIA - post nephrectomy histology was not considered for diagnosing wilms' tumor
Study period- February 2009- August 2012. With follow-up 2to 4 years
Study Technique (METHOD)
Epidemiological and clinical information are collected to all children aged 1-12 years diagnosed as wilms' tumor, who attended the pediatric medicine department of IPGMER & SSKM hospital. Blood counts, USG abdomen, Chest X ray were done. All cases are managed according to SIOP2001 protocol. They are closely observed during management. Prognosis is noted in terms mortality , local recurrence or metastasis of the disease and Event free survival.

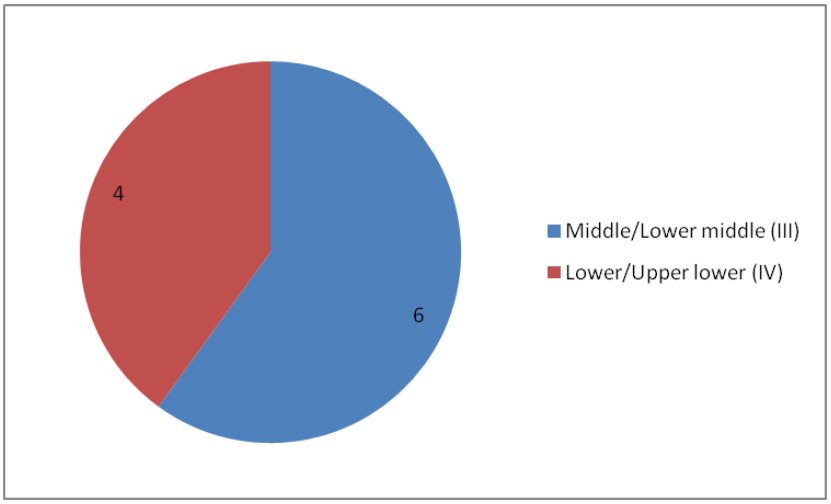
RESULTS
EPIDEMIOLOGY
A. AGE
Out of 8 children male and female is almost equal with age of presentation ranging from 3-12 year (median-5 year)



B. Gender

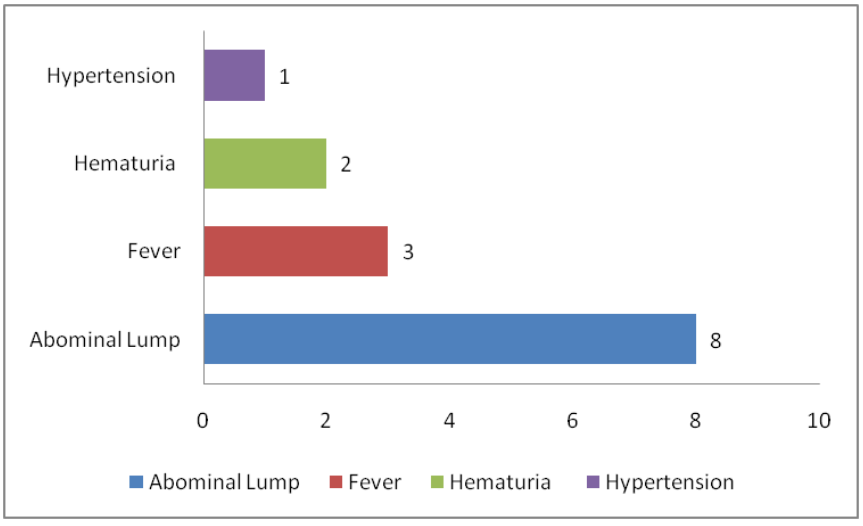


C.Kuppuswamy's Socioeconomic Status Scale



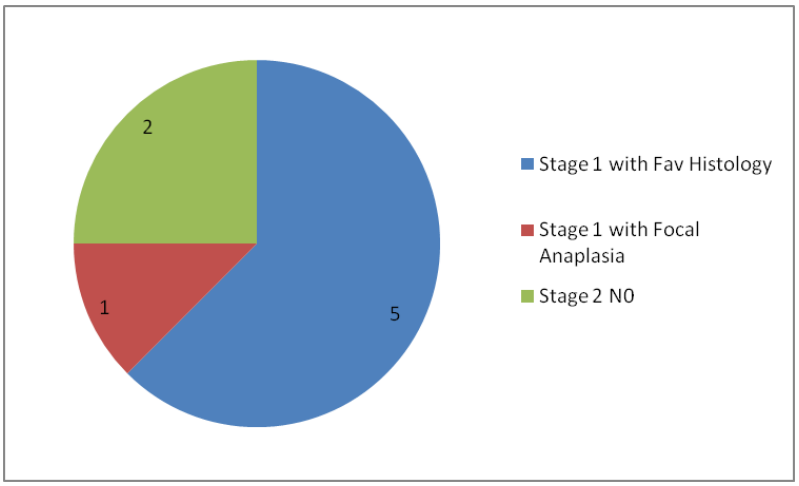
CLINICAL MANIFESTATION

Abdominal lump is the main clinical presentation along with fever 3, hematuria in 2 and hypertension in 1 child.



Staging

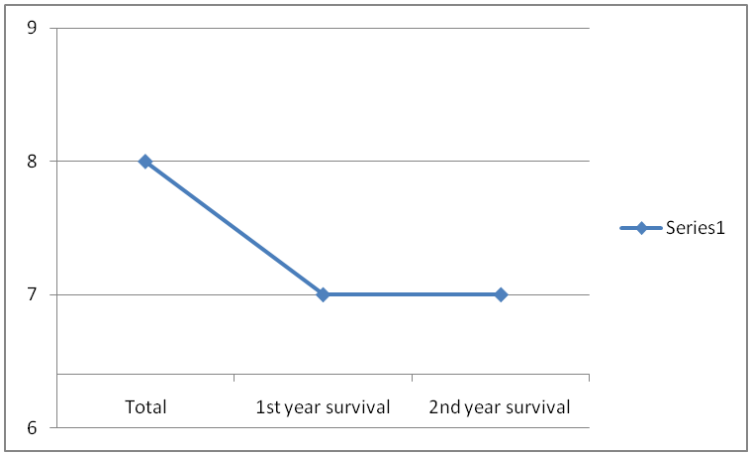
Clinically and histologically 6 of the children was on stage 1 and 2 were on stage 2.



Outcome

All children treated according to SIOP2001 protocol i.e chemotherapy followed by nephrectomy and post surgical chemotherapy depending upon pathological risk classification and surgical staging. Out of 8 children, 1 child died soon after nephrectomy due to Postoperative complications. Remaining 7 patients are disease free with no local recurrence and metastasis with follow up 2 to 4 years.

Overall survival 2years



Follow up was done 3 monthly on OPD basis clinically, USG abdomen 6 monthly.

DISCUSSION

WT is 9.30% [8] to 4.94 % [9] of all childhood malignancy in India compared to USA 6.5%.[10] Median age of unilateral WT in our study is 60 months ; in Breslow N [12] et.al study 36months . Male preponderance in our study (M:F 1.5:1) whereas slightly female preponderance found in Gupta DK et.al study.(M:F 0.92:1)[12]

Clinical presentation in our study abdominal lump (100%), fever (37%), hematuria (25%), hypertension (12%) is almost similar to Tongaonkar HB et. al[13] study except incidence of hypertension (25%) was higher in their study.

According to SIOP Paediatric Oncology in Developing Countries (PODC) our set-up is fulfilling the criteria of Setting 1, minimal necessity for curative treatment of WT.[6] Although Pathology and Radiotherapy unit not included in minimal criteria we have those facilities.

The management of WT is multidisciplinary; surgery, chemotherapy and radiotherapy combines depending upon the stage of disease.[14]

Furthermore, outcome not only depends on the stage but economic settings. Economically advanced countries have survival rate as high 85% [6] compare to low economic group as low 11% to 50%[15-19] Postoperative death of one patient in our study might have been because of the scarcity of advanced care.

History and Physical eamination are suggestive for WT and abdominal ultrasonography is diagnostic.[20,21]

SIOP protocol mainly emphasized on preoperative chemotherapy in all stages to prevent intraoperative complications of tumor spillage (Fig 1) and decreasing the dose of radiotherapy if required.

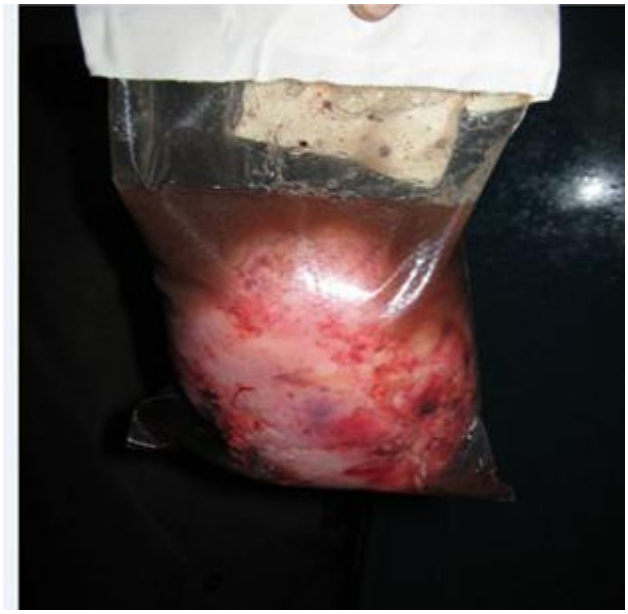


Fig 1: intact tumor, no spillage, after preoperative chemotherapy SIOP protocol

Although most of Wilms' tumor are advanced diseases in India [5], which is not present in our study. Our study consist of Stage I and II is 100% but Gupta DK et.al[12] is 35% . The reason may have been a reflection large number of refusal/abandonment tumor treatment for advanced diseases in developing countries.[7]

Outcome is comparable with other study. Overall Survival rate of our study for Stage I and II is 87.5% which little lower than the study of Gupta DK et.al[12] 95% and Tournade MF et al[22] 93% . Furthermore, 2 year relapse free survival in Tournade MF et al[22] study is 88% and our study is 100%.Good outcome with no evidence of local recurrence and metastasis as showed in our study. The prognosis is excellent in patient treated with SIOP protocol as we found in our study.

CONCLUSION

SIOP protocol can be used in management of pediatric Wilms' tumor with good prognosis and outcome especially in our country with limited resources and financial constraints. However more studies from INDIA is needed to bring SIOP protocol in treatment of pediatric wilms tumor in our country

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