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CLEAR CELL SARCOMA OF KIDNEY: A SINGLE CENTER EXPERIENCE

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CCSK: Introduction

- ◎ Clear cell sarcoma of the kidney- rare renal neoplasm
 - › second most common renal tumor of childhood.
- ◎ Peak age : 1-3 years
- ◎ Predominantly males : M:F=2:1
- ◎ High recurrence rate
- ◎ Poor overall outcome

CCSK: Introduction

- ◎ No bilateral cases
- ◎ 4% present with distant metastasis.
 - › Bone metastases occur in 15–60% of patients with metastatic CCSK,
 - › metastases to other organs (lymph nodes, lungs, brain, and liver) also reported.

CCSK: Aims

- ◎ To evaluate the overall outcome, recurrence rate and event free survival in children treated for CCSK at a single center over a period of ten years.

CCSK: Materials and Methods

- ◎ Prospective study
- ◎ August 1999-November 2009
- ◎ All cases of CCSK registered at Pediatric Surgery Cancer Clinic, AIIMS
- ◎ Staging as per NWTS 5
- ◎ Statistical analysis – Kaplan Meier curve
 - > Overall survival (OS)
 - > Event free survival (EFS)

CCSK: Treatment protocol

- ◎ Pre-operative diagnosis of CCSK (FNAC)*

Resection



Chemotherapy + Radiotherapy

*Venkateswaran K, Sandeep Agarwala et al,
Diagnostic Cytopathology, 2005 Vol 33, No 2

CCSK: Treatment protocol

- ◉ No diagnosis pre-operatively

Neoadjuvant chemotherapy (as for Wilms Tumour)

↓
Reassessment and Resection

↓
Chemotherapy (Regime I) + Radiotherapy

CCSK: Treatment protocol

○ Surgery

- > Nephroureterectomy + lymph node sampling

○ Chemotherapy

- > Regime I (NWTS 5)
- > Vincristine, doxorubicin, cyclophosphamide and etoposide for 24 weeks

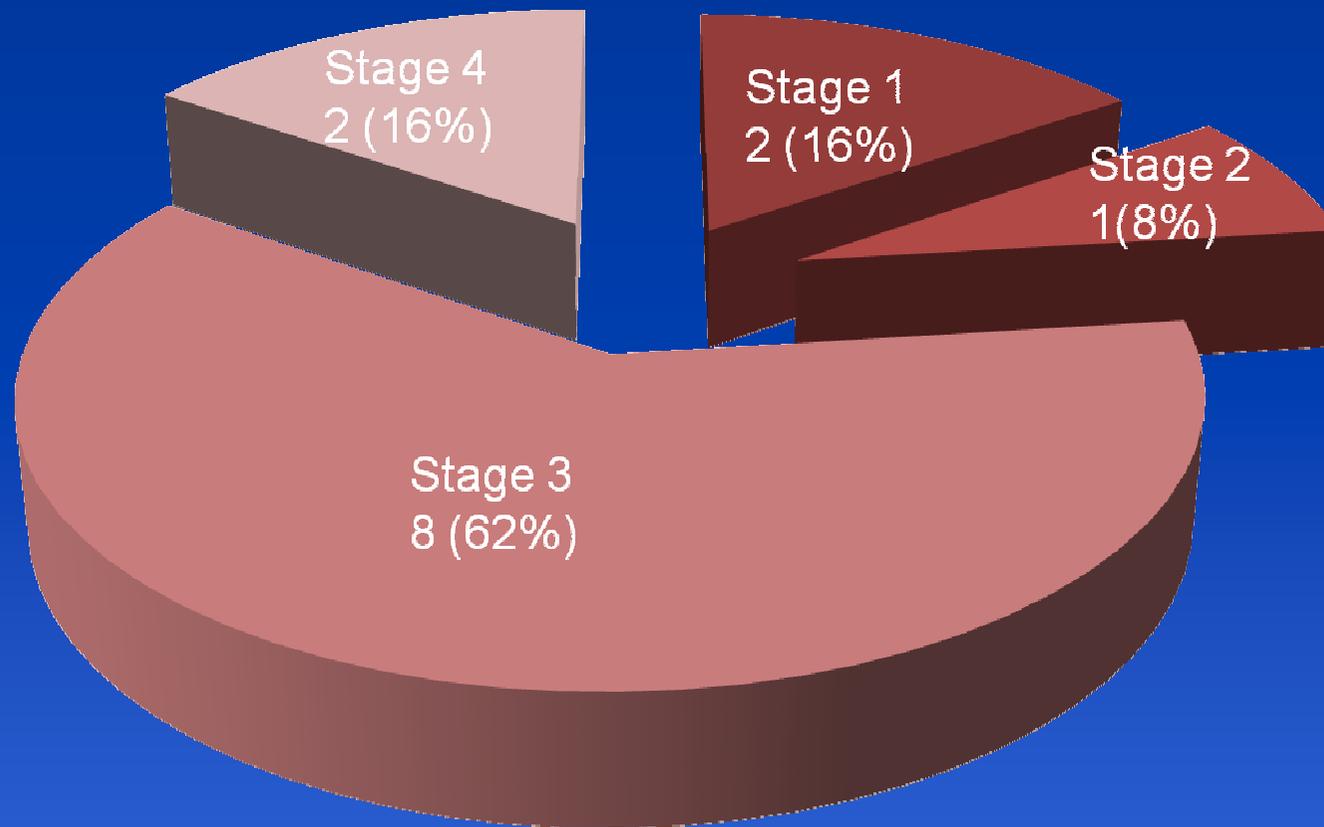
Guidelines and road map:

	Weeks																			
	0	1	2	3	4	5	6	7	8	9	10	11	12	13	15	18	21	24		
V		■	■		■	■	■	■	■		■	■	■*	■*		■*		■*		
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CCSK: Results

- ◎ Total - Renal tumors = 222
 - > CCSK = 13 (1.4%)
- ◎ Age
 - > Range 12 – 120 months
 - > Median 24 months
- ◎ Sex
 - > Boys - 11
 - > Girls - 2

CCSK: Results- Stage distribution



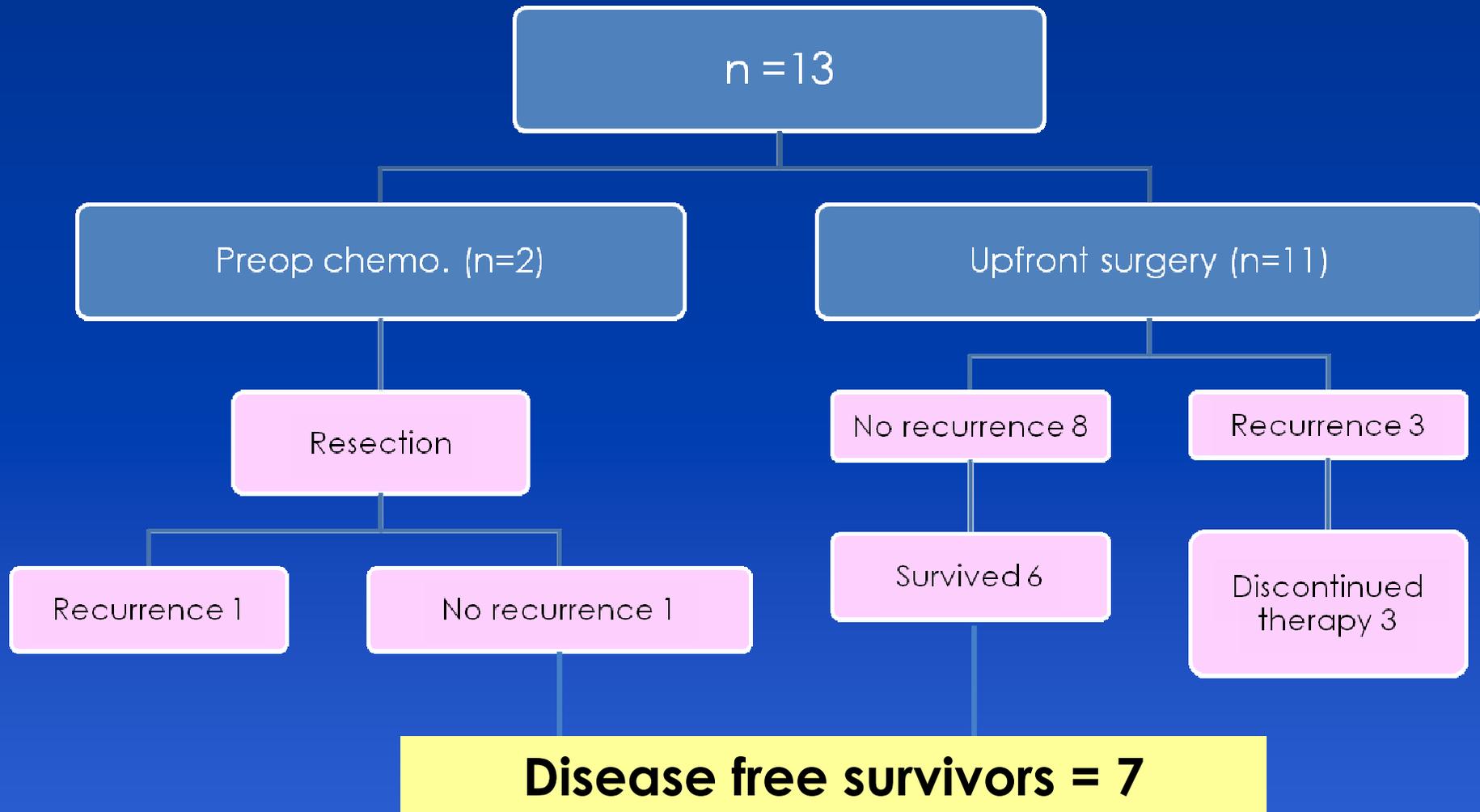
CCSK: Results

◎ Presentation

- > Massive abdominal tumours- all patients
- > Metastasis at presentation- 2 (15%)
 - Bony metastases

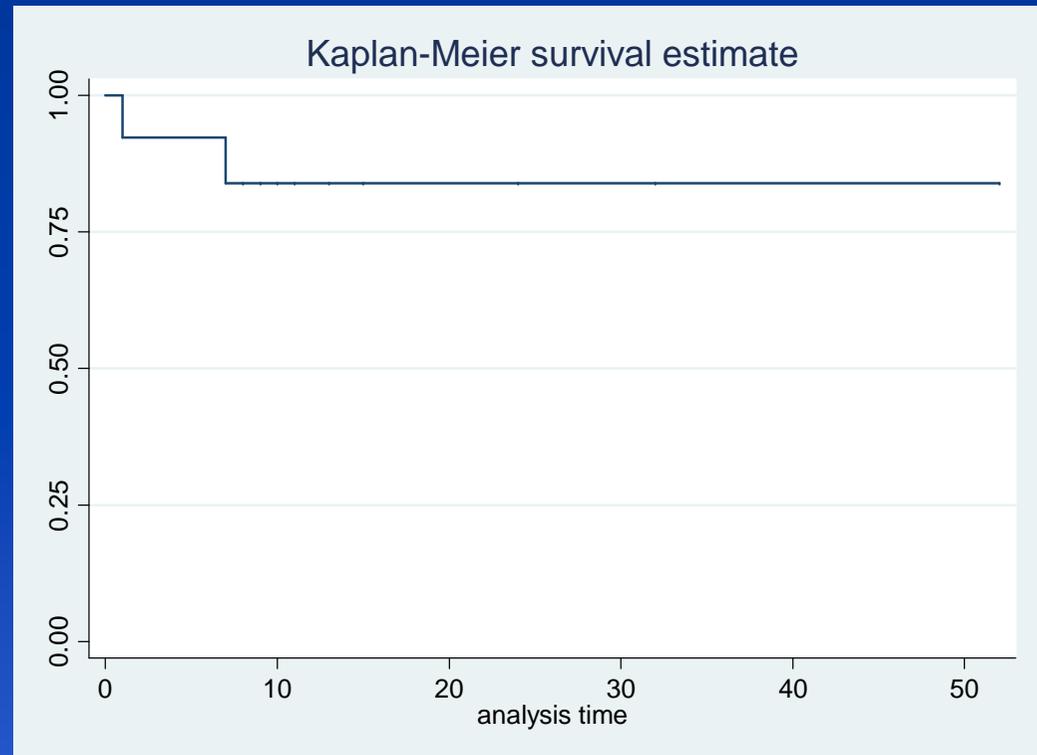


CCSK: Results- management



CCSSK: Overall survival (OS)

- ◎ 2 years
 - > OS – 83.9%
 - (95CI 49.4-95.7)
- ◎ 2 patients expired
 - > 1 chemotherapy related
 - > 1 disease related



Disease free survival DFS at the end of 1 year 65%

Slide 14

A2

What is thjis surgery related death??

A, 20/10/2011

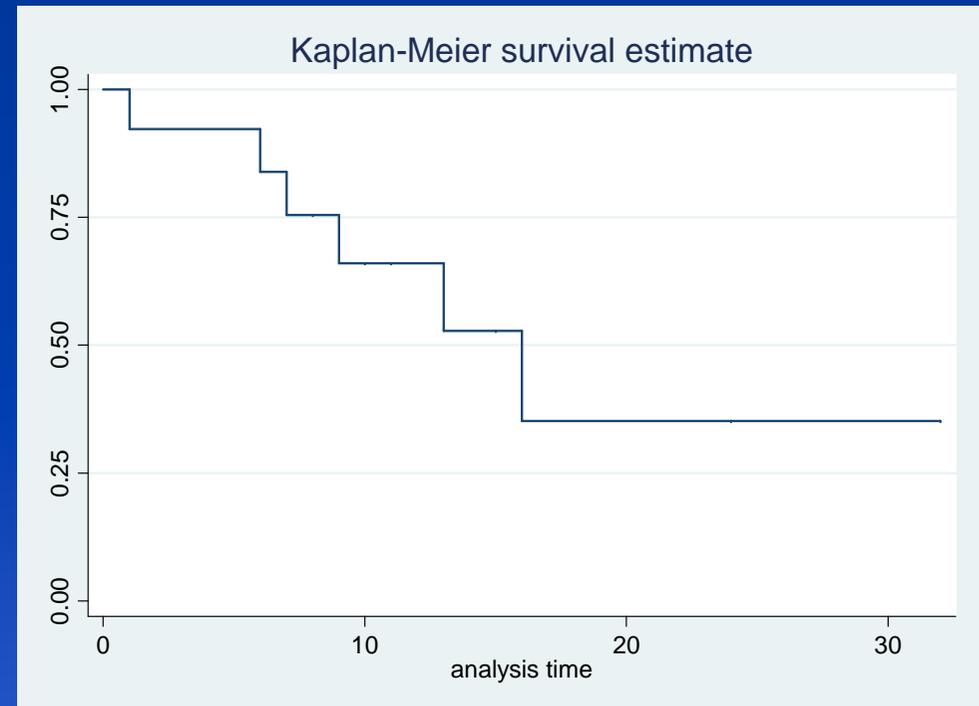
CCSK: Recurrence

- ◎ Recurrences – 4 (33%)
(6,9,13 and 16 months)
 - > 3 -Progressive disease
 - Discontinued treatment
 - > 1- re-resection, alternate chemotherapy (ICE regime) and radiotherapy
 - Survival



CCSK: Event free survival (Death, recurrence, progression)

- ◎ Events – 6
 - > Death: 2
 - > Recurrence : 4
 - Progression: 3
- ◎ 1 year EFS-66.8%
(95CI 32.5-85.8)
- ◎ 2 year EFS-35.2%
(95CI 6.5 -67.1)



CCSK: Discussion

- ◎ Bilateral cases not reported
- ◎ 30% relapses beyond 2 years
 - > WT most relapses within 2 years of diagnosis
 - > Longer intensive surveillance for recurrences
- ◎ Sites of metastatic recurrence
 - > Bone- 50%
 - (compared to 2% in WT)
 - > lungs, abdomen, brain and liver
 - > Soft tissues
 - Scalp, epidural, nasopharynx, neck, paraspinal, abdominal wall and axilla

CCSK: Outcomes

◎ Prognostic factors

- > Age > 4 years
- > Stage 2,3,4
- > Necrosis
- > No treatment

with Doxorubicin

Increased relative risk for death and relapse

(Argani P et al. Am J Surg Pathol 24:4, 2000)

CCSK: Outcomes

◎ Overall 4 year survival:

- > Stage 1 : 98%
- > Other stages : 68%

◎ Relapse free survival 60%

Series	Year	N	OS %	DFS%
# Kusumkumary P et al	1999	12	64	56
*Nita L. Seibel et al NWTS 4	2004	86	95	65.2
@ Hadley GP et al,	2010	14	57	-

#Pediatr Hematol Oncol 1999 Mar-Apr; 16(2); 169-74

*J Clin Oncol 2004 22:468-473.

@ Pediatr Surg Int 2010 Apr26(4):345-8

CCSK: Conclusions

- ⊙ All patients presented with massive tumors
- ⊙ Pre-operative diagnosis of CCSK was made in 85%
 - ⊙ Good cytopathology back up
 - ⊙ Underwent upfront resection
- ⊙ 15% received initial therapy as for Wilms tumor
- ⊙ Recurrences were early and in 31%
 - ⊙ Recurrence occurred at a median of 11 months.
 - ⊙ Only one could be salvaged
 - ⊙ 75% progressed
 - ⊙ Recurrence in CCSK represents a poor outcome
- ⊙ 2 yr Overall survival (83.9%) was good but the event free survival was 35.2%.

