OUTCOME AND PREDICTORS OF SURVIVAL AMONG CHILDREN MANAGED FOR ORBITAL RHABDOMYOSARCOMA AT UGANDA CANCER INSTITUTE.

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INTRODUCTION.

- Most common rapidly growing soft tissue sarcoma in children in the orbit having an occurrence of 10%(Bouutrox et al 2015)

- In a study at Makerere University in 1967, ORMS accounted for 7.6% of all tumors of the eye and adnexa.(Templeton, 1967).

- Highly curable, with about 90% survival in the developed world this is attributed to the multimodality approach.

- However little is reported about ORMS outcomes in developing countries like Uganda.

OBJECTIVES.

General objective;

To assess the outcome and predictors of survival of children with orbital Rhabdomyosarcoma at UCI from January 2008 to December 2019.

Primary objectives;

- To determine the survival at 3 years, 5 years of children at UCI from January 2008 to December 2019.

- To determine the predictors of survival of children at UCI from January 2008 to December 2019.

Secondary objective;

- To describe the clinical presentation of children at UCI from January 2018 to December 2019.

METHODS.

- **Study design**; Retrospective cohort study.

- **Study period;** 1st May 2021 to 31st May 2021.

- **Study population;**

o All children with orbital rhabdomyosarcoma

 managed at Uganda Cancer Institute between January 2008 and December 2019

- **Inclusion Criteria**;

All patients with histological confirmation of ORMS.
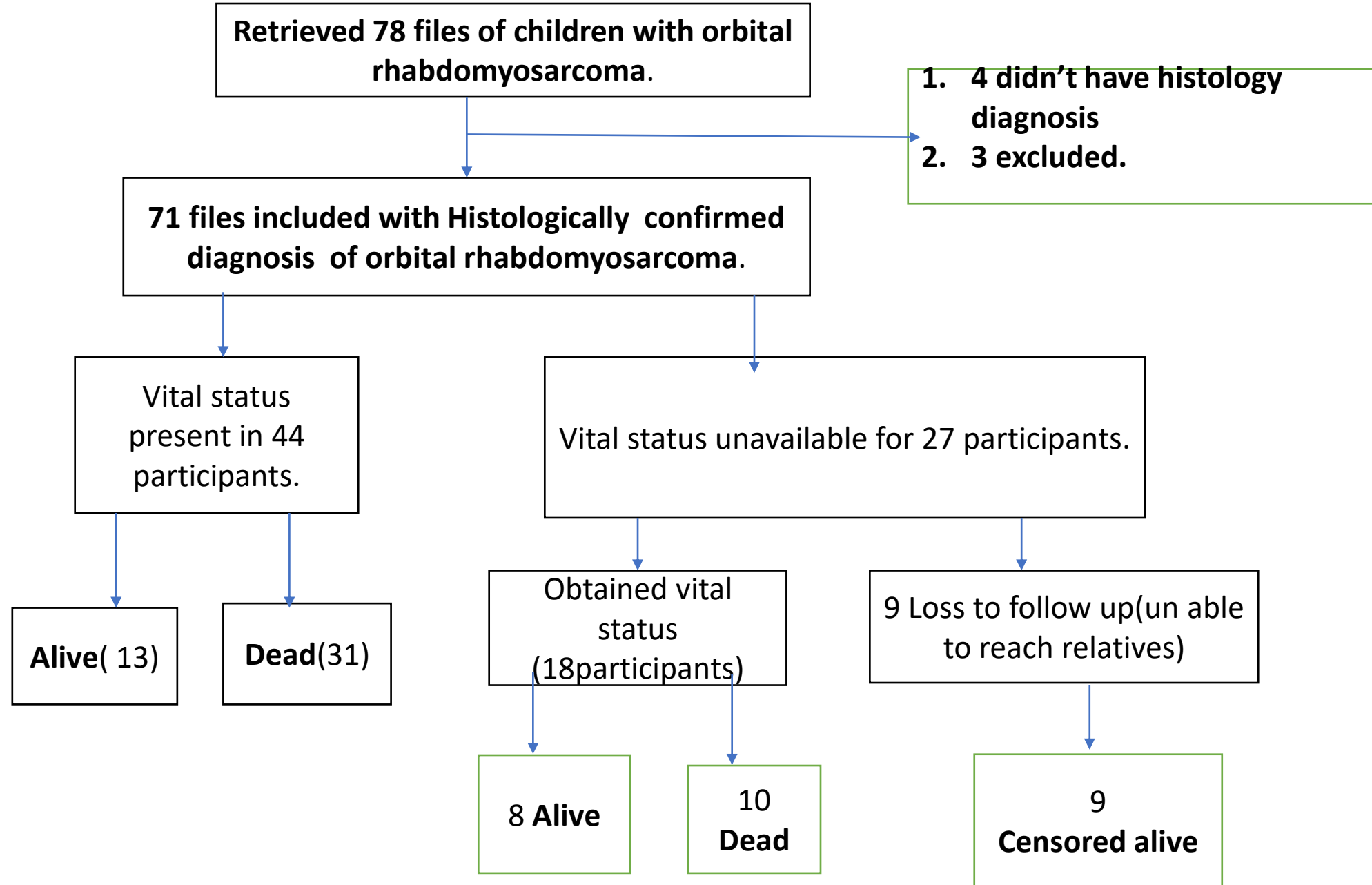
- **Exclusion Criteria;**

Patients who were diagnosed but ran away from ward before commencement of treatment.

- **Sample size;**

71 children

Study flow diagram.

Retrieved 78 files of children with orbital rhabdomyosarcoma.

1. 4 didn't have histology diagnosis
2. 3 excluded.

71 files included with Histologically confirmed diagnosis of orbital rhabdomyosarcoma.

Vital status present in 44 participants.

Vital status unavailable for 27 participants.

Alive(13)

Dead(31)

Obtained vital status (18participants)

9 Loss to follow up(un able to reach relatives)

8 Alive

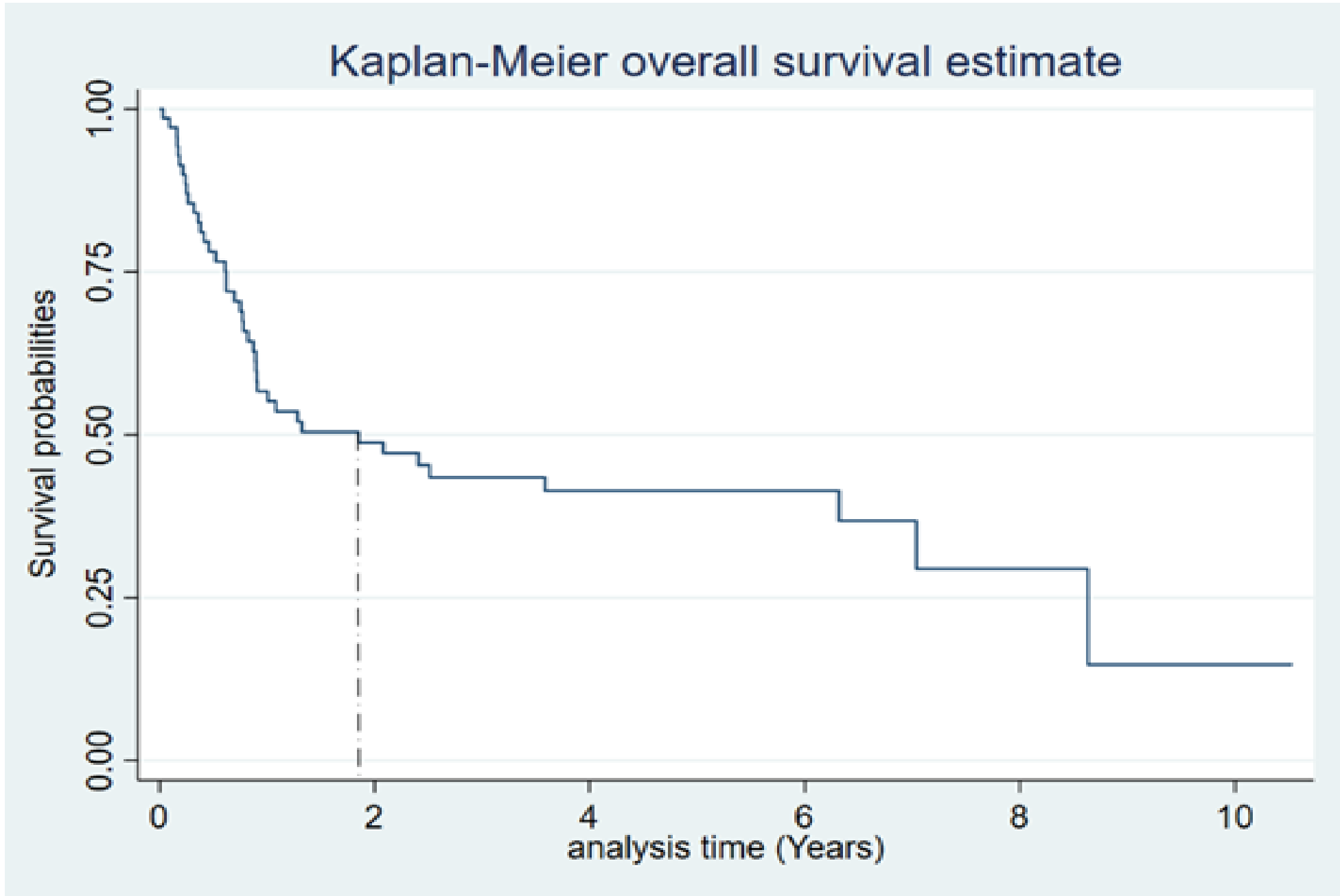10 Dead

9 Censored alive

RESULTS.

Population characteristics (N=71)

- More than half of the study participants were male, 54.9%(n=39)

- The median age at diagnosis was 6 years (IQR: 4-12). The youngest participant was 5 months, oldest was 18 years and majority of the participants, n=31(43.7%) were aged between 7-14years.

- Total of 177.3 person-years.

- Most of the participants, n=46 (64.8%) came from rural areas.

Outcome of children with ORMS

Outcome		Frequency (%)
Vital status	Alive	30(42.2)
	Dead	41(57.8)
Declared cured	Yes	11(15.5)
	No	60(84.5)
Lost to follow up	Yes	9(12.7)
	No	62(87.3)
Disease progression	Yes	42(59.2)
	No	29(40.8)
Abandoned treatment* (defaulted)	Yes	49(69.0)
	No	22(31.0)

Kaplan Meir survival curve(OBJECTIVE 1)

KM survival estimated for secondary orbital tumors

Kaplan-Meier survival estimates for secondary orbital tumors

P = 0.001

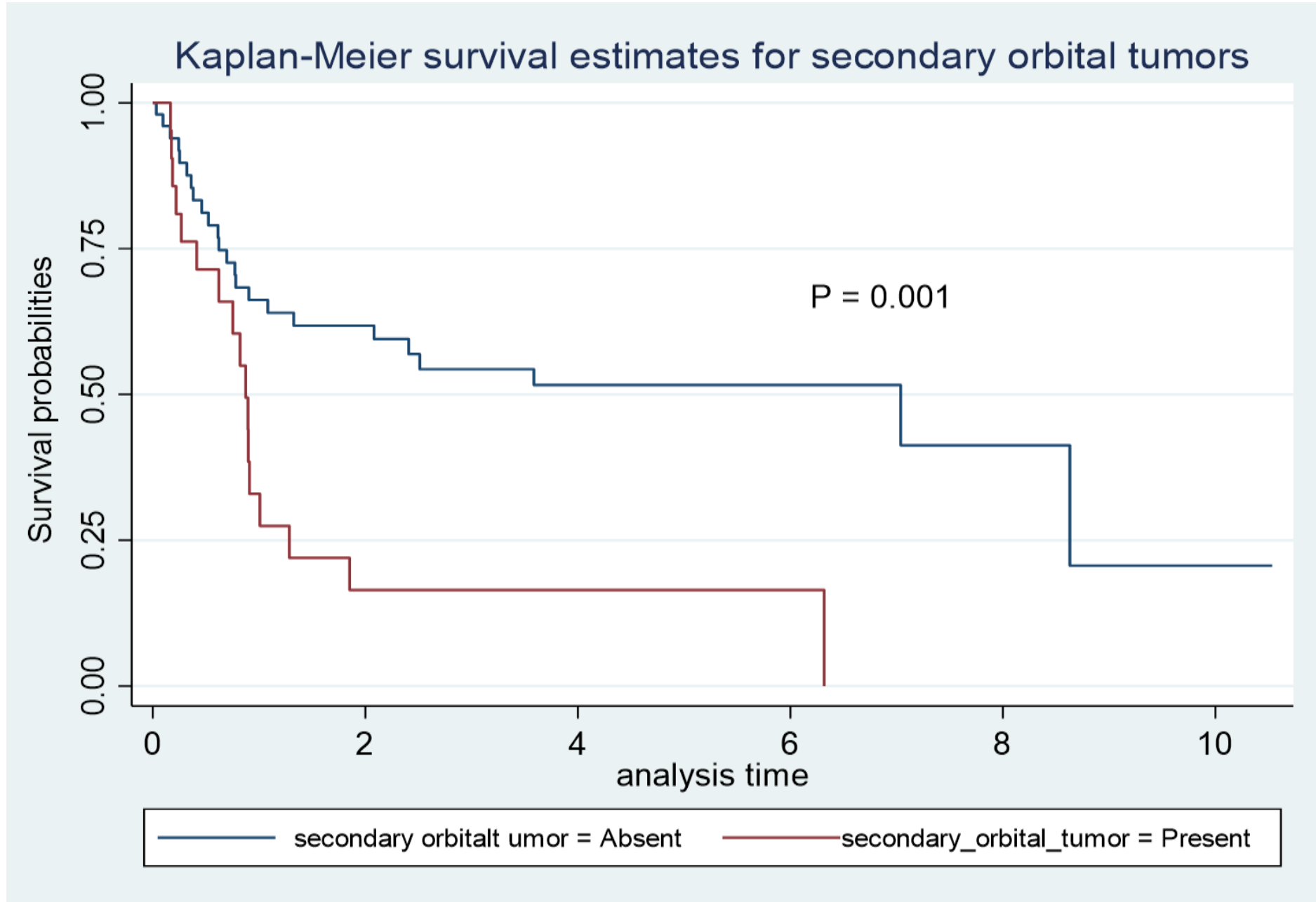secondary orbitalt umor = Absent   secondary_orbital_tumor = Present

Table 7: Multivariate analysis for predictors of survival for the 71 study participants OBJECTIVE 2

characteristic	Crude HR[95%CI]	Adjusted HR[95%CI]	P value
Age groups			
Below 3ears	1.00	1.00	
3 to <7 years	4.42 [0.18-1.01]	**0.30 [0.11-0.81]**	**0.018**
7 to <15 years	**0.42 [0.19-0.95]**	**0.26 [0.10-0.66]**	**0.005**
15 -18 years	0.32 [0.06-1.63]	0.45 [0.05-3.89]	0.466
Duration of symptoms (months)			
0-2	1.00	1.00	
3-6	1.63 [0.81-3.29]	2.05 [0.92-4.57]	0.078
>6	0.54 [0.06-1.84]	**0.24 [0.06-0.92]**	**0.037**
Baseline tumor group			
I	1.00	1.00	
II	0.50 [0.13-1.94]	0.63 [0.13-3.09]	0.567
III	0.74 [0.31-1.76]	**0.21 [0.05-0.95]**	**0.042**
IV	2.60 [0.93-7.29]	0.23 [0.03-2.15]	0.199
Primary orbital tumor			
Present	**0.34 [0.18-0.65]**	**0.37 [0.16-0.83]**	**0.016**
Absent	1.00	1.00	
Disease progression			
Yes	1.00	1.00	
No	**0.22 [0.09-0.49]**	**0.16 [0.06-0.47]**	**0.001**

DISCUSSION.

- Survival among children with ORMS in Uganda are quite low compared to the Global Initiative for Childhood Cancer (GICC) 2018 target of 60% by 2030.

- Slightly similar- African countries like South Africa which reported overall 5-year survival rate of 45%,(Van Der Schyff & Stefan, 2010).

- Significantly low-developed countries such as China, USA and France have estimated overall survival at 91.3%, 97% and 98% respectively (Ducrey, Nenadov-Beck, & Spahn, 2002; Tang et al., 2018).

- The low survival rates observed in this study may point to gaps in care provided in Sub-Saharan countries like Uganda when compared to high- income countries.
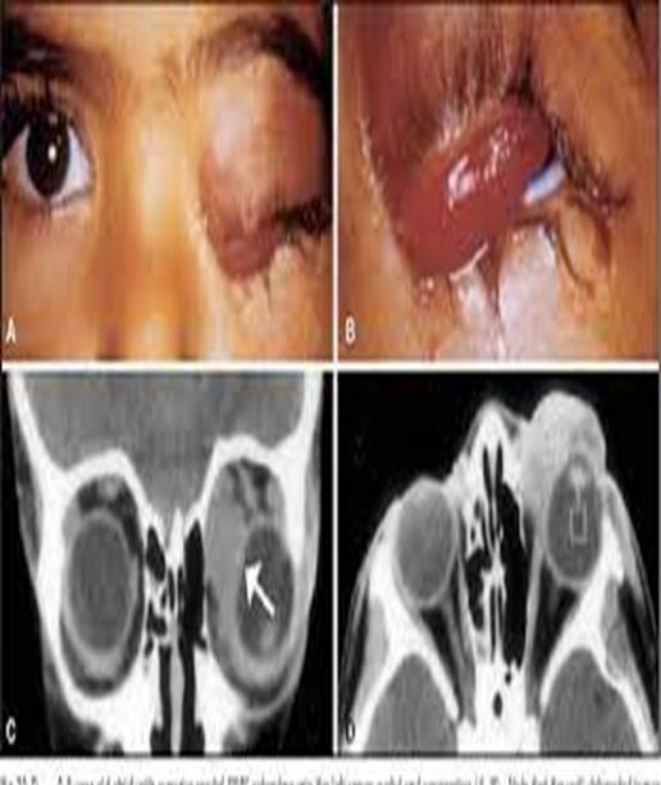
Discussion…..

- Data from our current study shows up to 58% increased chances of survival of children >3-15 years.

- A study done in USA also found infants to have poor prognosis compared to older children(Iyad Sultan et al., 2010).

- This may be due to less tolerance and adverse toxicities to treatment modalities and disease biology.

- Furthermore, probably due to individual participant characteristics like genetics, nutritional status and socio-economic factors.

Discussion………….

- Having ORMS as a primary tumor is a positive predictor of survival. Evidence from the current study points to 66% increased survival when compared to children who had secondary ORMS.(3times risk of death).

- This is possibly because secondary ORMS is due to metastatic disease which is often an indicator of an advanced malignancy in another organ.

- Several studies have reported poor survivals of as low as 20% for secondary ORMS.

- Conversely several studies have reported survival greater than 90% in patients that had localized primary tumor (Marc Hendricks et al., 2017; Shields & Shields, 2003).

conclusion

- The 5-year survival of children with ORMS at UCI is very low at 42%.

- Older age (age groups >3-15years), having a primary tumor and no disease progression positively predict survival.

- Majority presented with advanced disease-group III and IV tumor (60.5% and 19.5%) respectively.

- There is a high rate of treatment abandonment among patients with ORMS(69%).

thank you