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Overall treatment outcome and recurrence pattern of olfactory neuroblastoma from a single medical center

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Abstract

Introduction

In this study we tried to evaluate a long-term treatment outcome of olfactory neuroblastoma (ONB).

Method

A retrospective review of patients was performed for those who had definitive treatment for ONB from 1992 to 2021. Tumor stage, treatment modalities, and survivals using Kaplan Meier and Cox's-proportional hazard model were analyzed.

Result

Ninety-four patients were analyzed. Mean age was 41.4±14.1 years and mean duration of follow up was 89.8±78.6 months. In total, 75.5% underwent surgical resection with or without adjuvant therapy and the other 24.5% underwent definitive radiotherapy. The 10-year cancer specific survival (CSS) was 79.2%. Univariate cox-regression analysis revealed that Kadish Stage D (HR=6.66, 95% CI: 2.34-18.96) compared to others, Hyams grade III/IV (HR=13.11, 95% CI:1.58-109.04) compared to others, and definitive RT only (HR 5.76, 95% CI:2.04-16.26) compared to surgical resection were associated with poor cancer specific survival. Overall relapse rate was 42.6%. The 5-year CSS after initial relapse was significantly different among recur site; 82.5% for local, 85.7% for regional, 41.7% for meningeal, and 0% for distant (p<0.001). The 5-year CSS after initial relapse was also affected by the following salvage treatment modality; 79.1% for surgery, 50.0% for RT, and 0% chemotherapy (p<0.001). Kadish D (0<0.001), Hyams high grade (III/IV) (p=0.042), and definitive RT only (p=0.001) was associated with lower distant free survival.

Conclusion

In ONB, definitive RT without surgical resection, higher tumor stage and grade are associated with poor distant control and affect overall survival. Despite high relapse rate, surgery or RT as a salvage therapy significantly improved overall survival.

Introduction

Olfactory neuroblastoma (ONB) is a rare malignancy that develops from the olfactory mucosa, mostly arising in the superior portion of the nasal cavity. Complete surgical resection with adequate margins combined with adjuvant radiotherapy (RT) is the treatment of choice for patients with ONB. In this study we tried to evaluate a long-term treatment of olfactory neuroblastoma (ONB).

Methods and Materials

Retrospective review

- Seoul National University Hospital and Seoul National University Bundang Hospital
- From 1992-2021 (30 years)

Exclusion criteria

- M1 disease
- Did not have definite therapy (either CCRT/RT or Surgery)
- Lack of medical record

→ Evaluated tumor stage/grade, treatment modality, disease specific survival, recurrence pattern (local, regional, distant, and meningeal)

Results

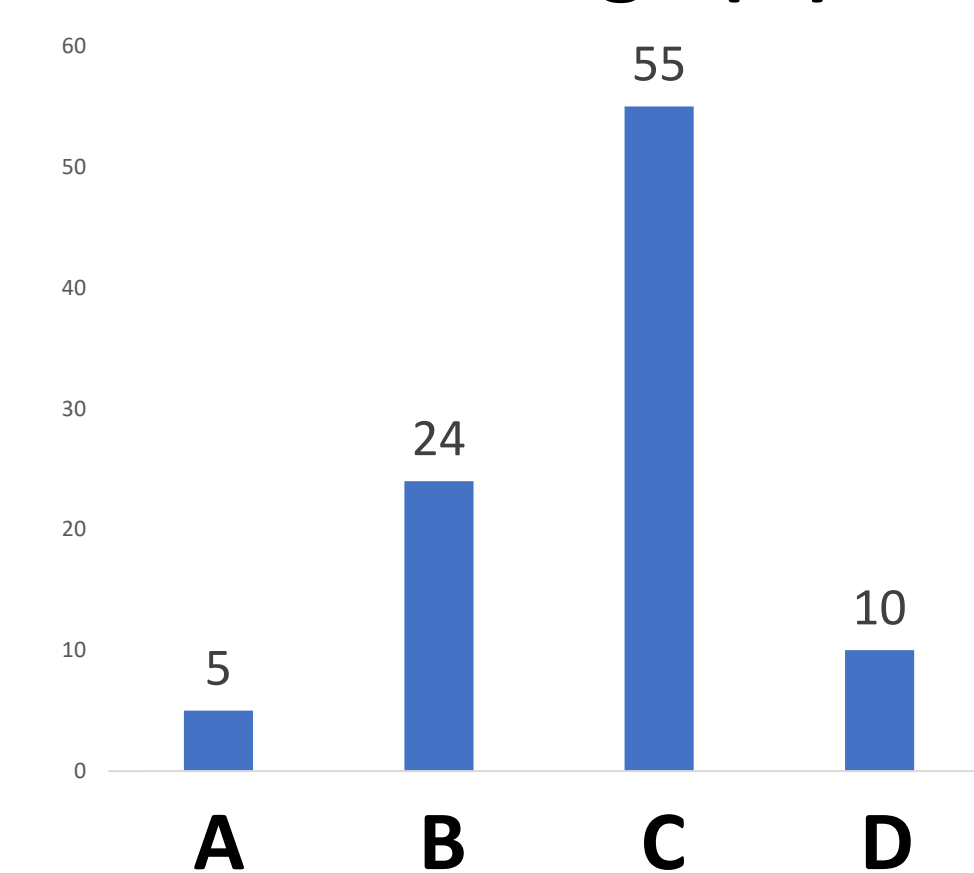
N=94

M:F ratio = 58:36

Mean age = 41.4±14.1

Mean duration of follow up : 89.8± 78.6 (mo)

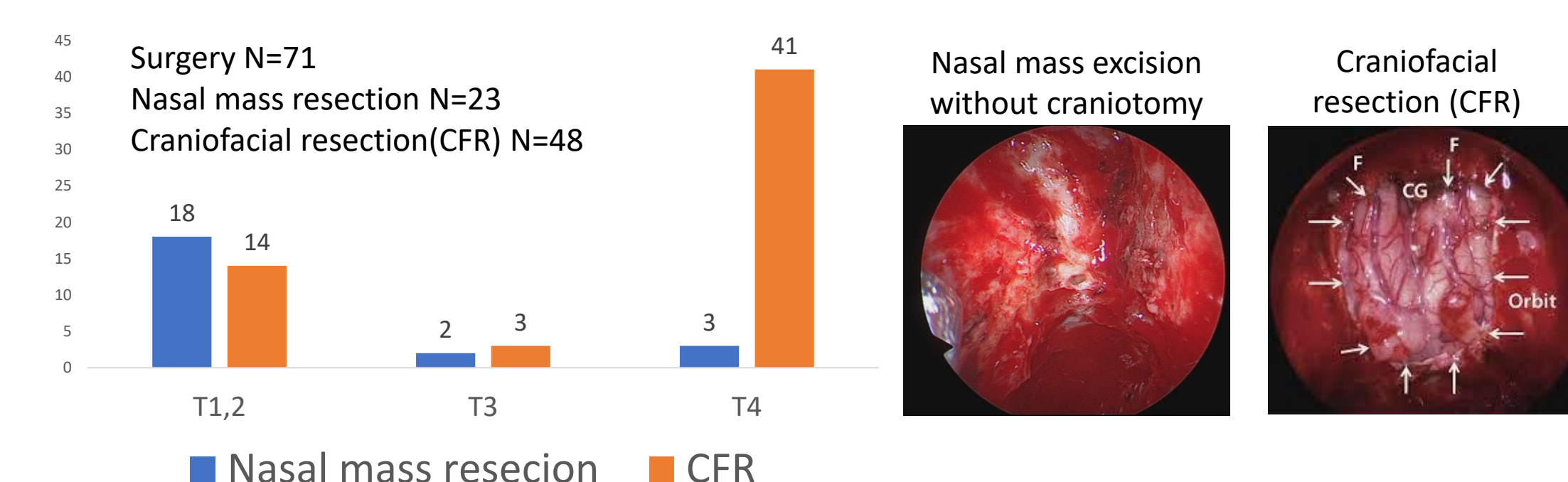
Kadish Stage (N)



Treatment Modality

Modality	Neoadjuvant	Definitive	Adjuvant	N
1	Chemotherapy	Surgery	±Radiotherapy	32 (34.0%)
2	Chemotherapy	RT/CCRT		19 (20.2%)
3		Surgery	±Radiotherapy	39 (41.5%)
4		RT/CCRT		4 (4.3%)

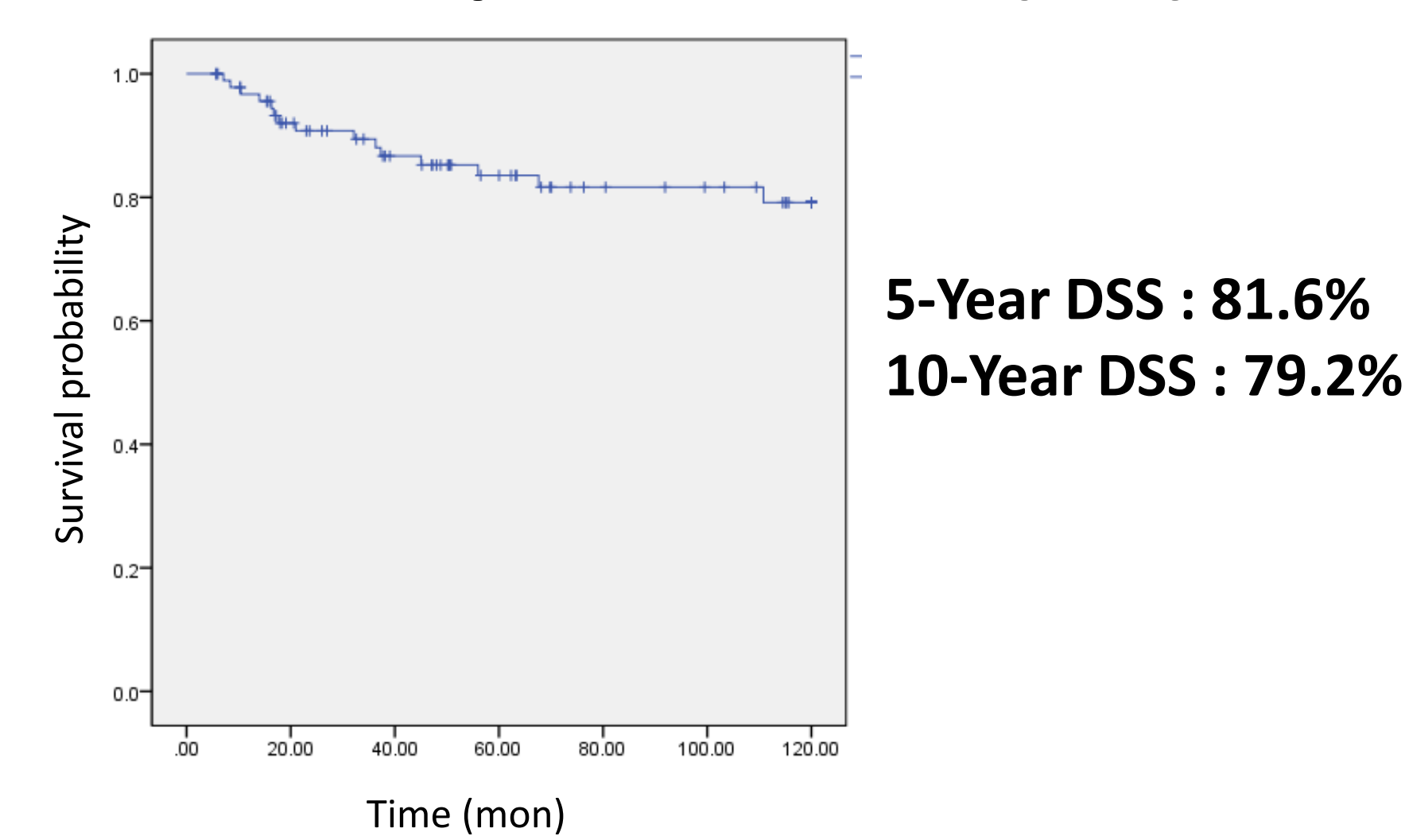
Extent of resection



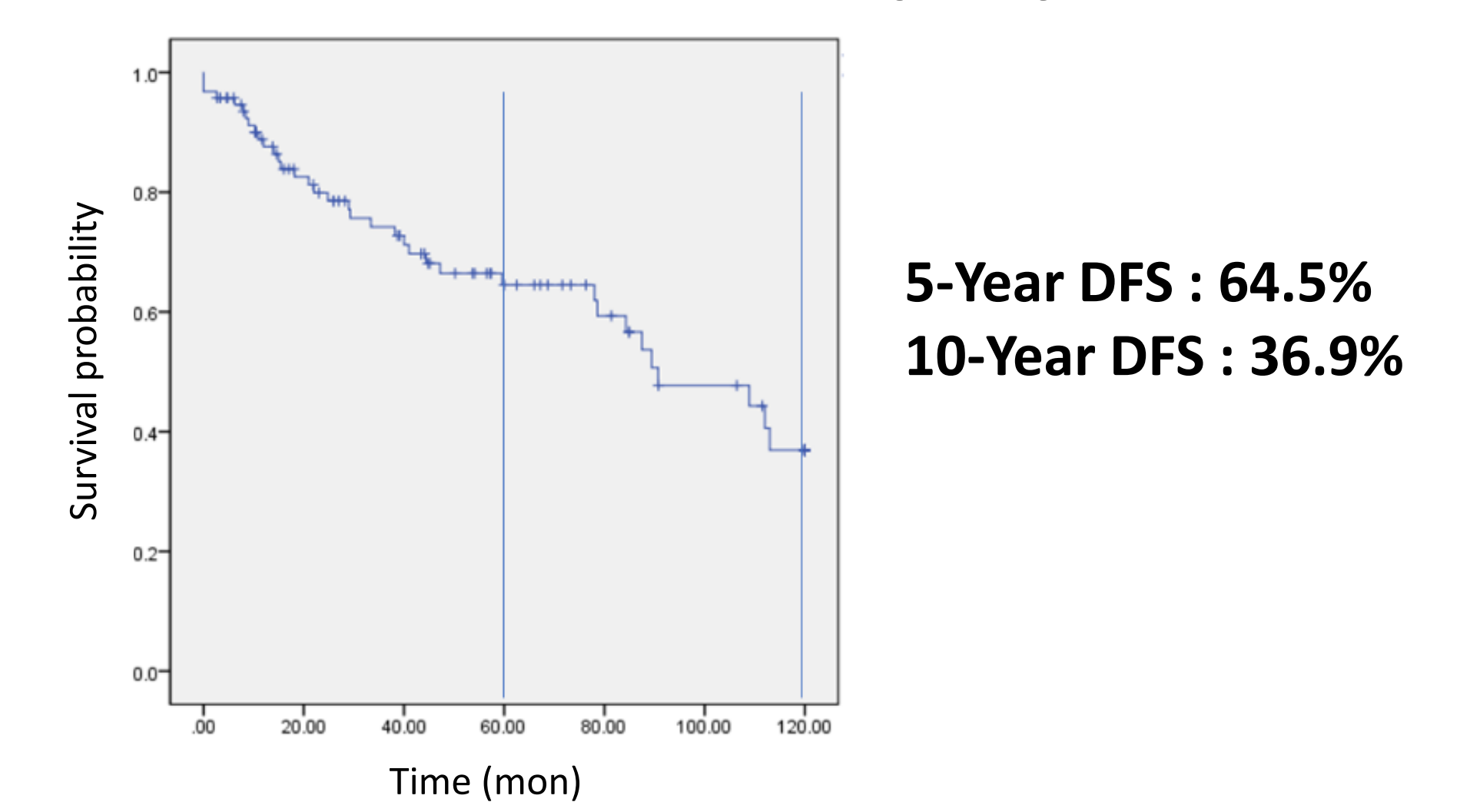
Prognostic factor : Univariate cox regression for 10-year disease specific survival

Clinical factors	Hazard Ratio	95% CI	P value
Kadish D	6.663	2.342-18.955	<0.001
Hyams grade 3,4 (high grade)	13.116	1.578-109.042	0.017
RT/CCRT only	5.763	2.042-16.263	0.001
Residual (subtotal resection)	55.562	4.938-627.202	0.001
Grade II orbital invasion	3.239	1.029-10.202	0.045

Disease specific survival (DSS)



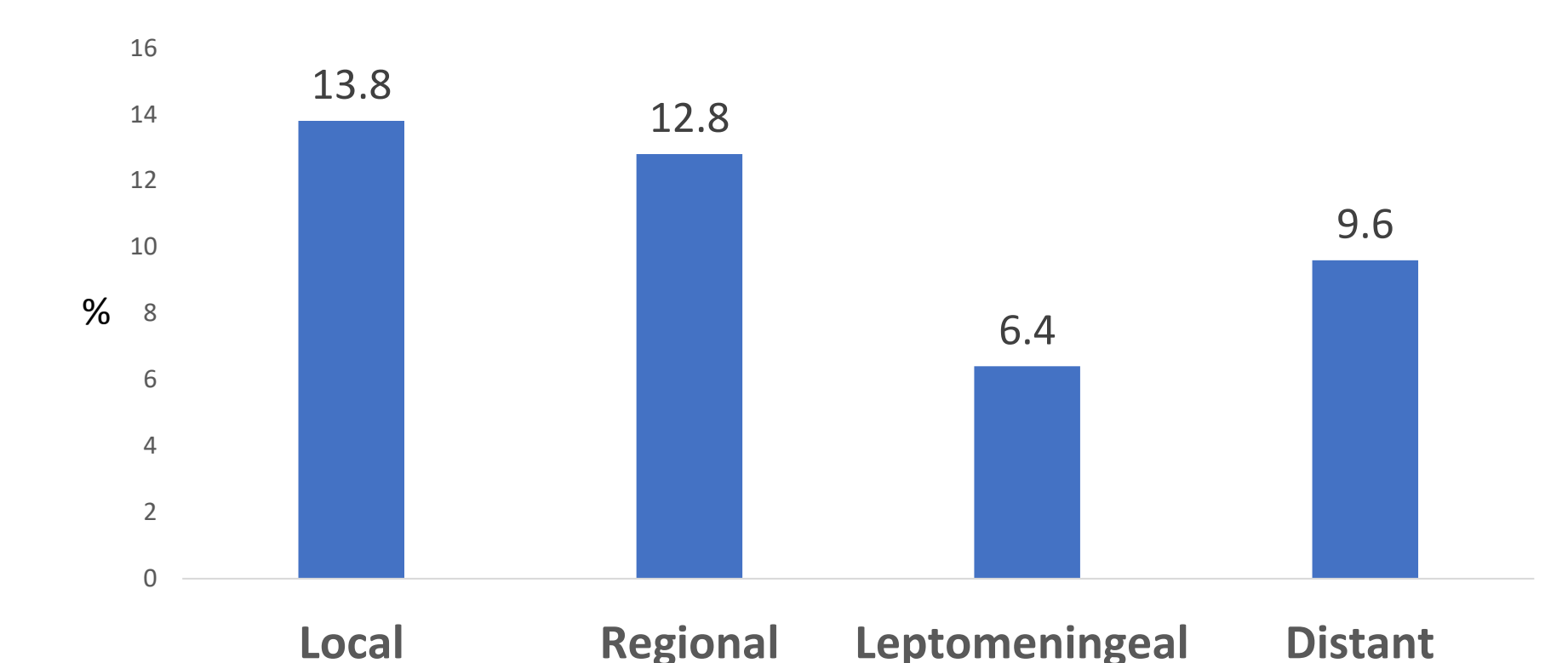
Disease free survival (DFS)



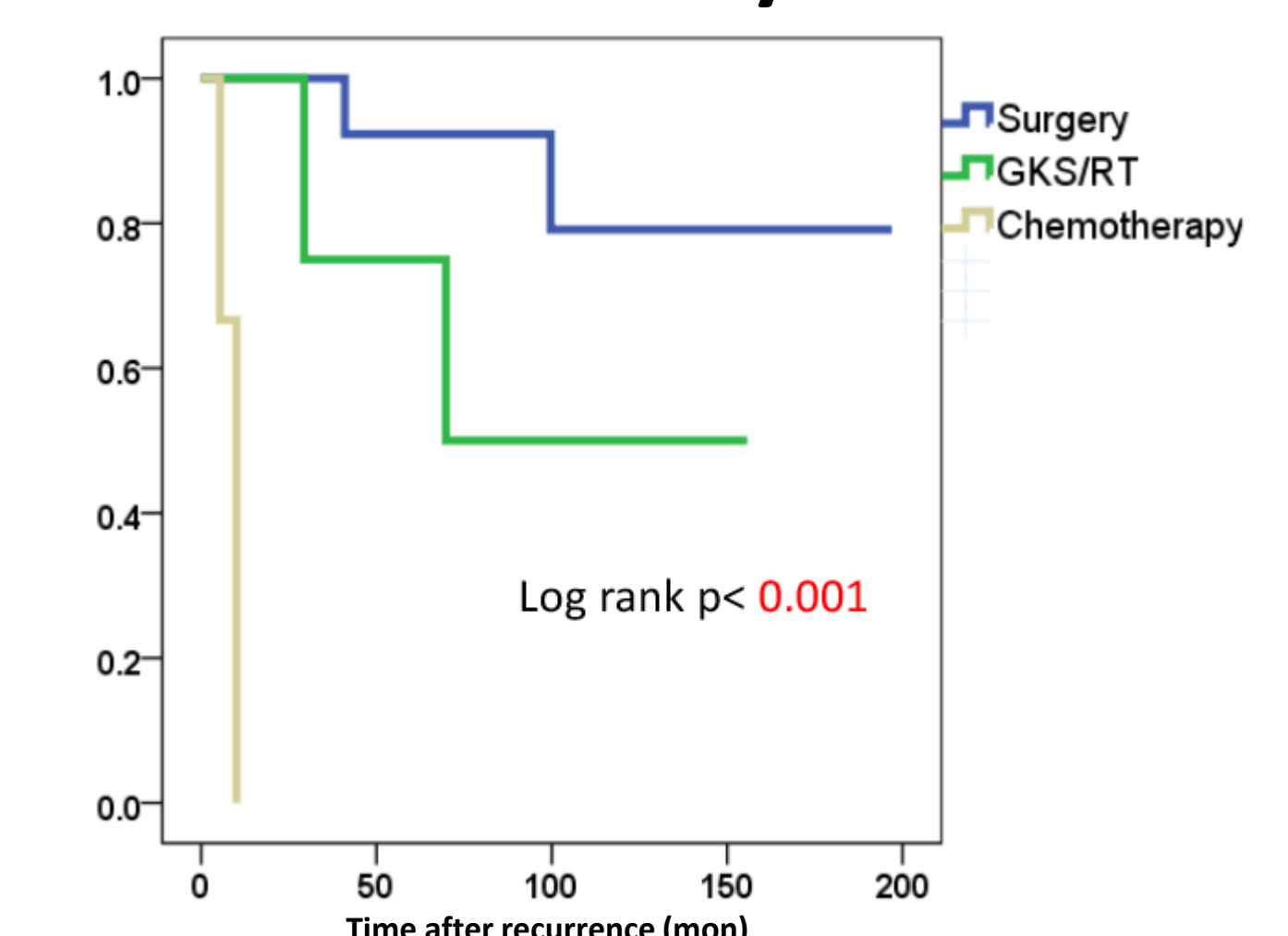
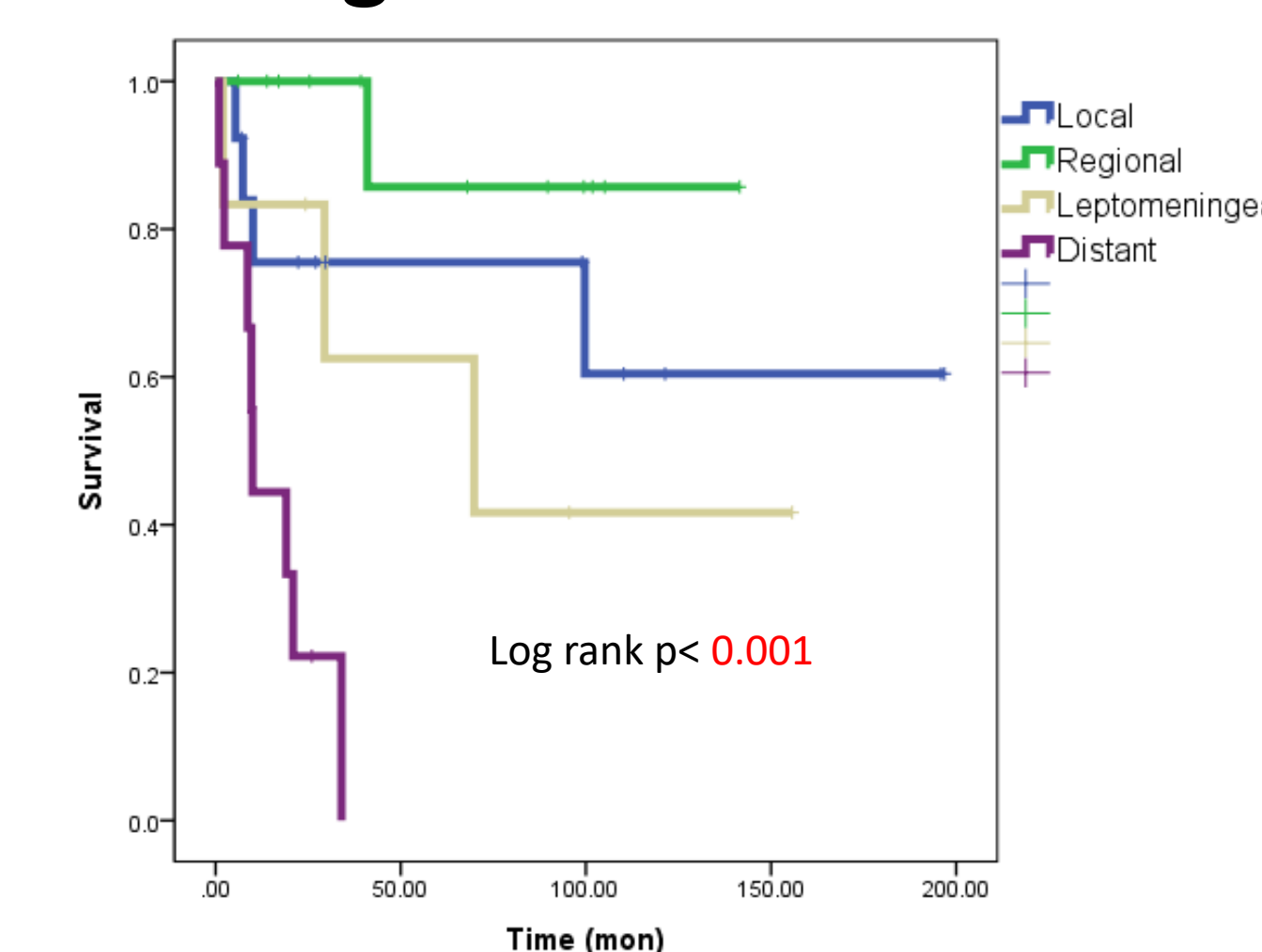
Recur site and mean time to recur

Overall relapse rate = 42.6% (N=40)

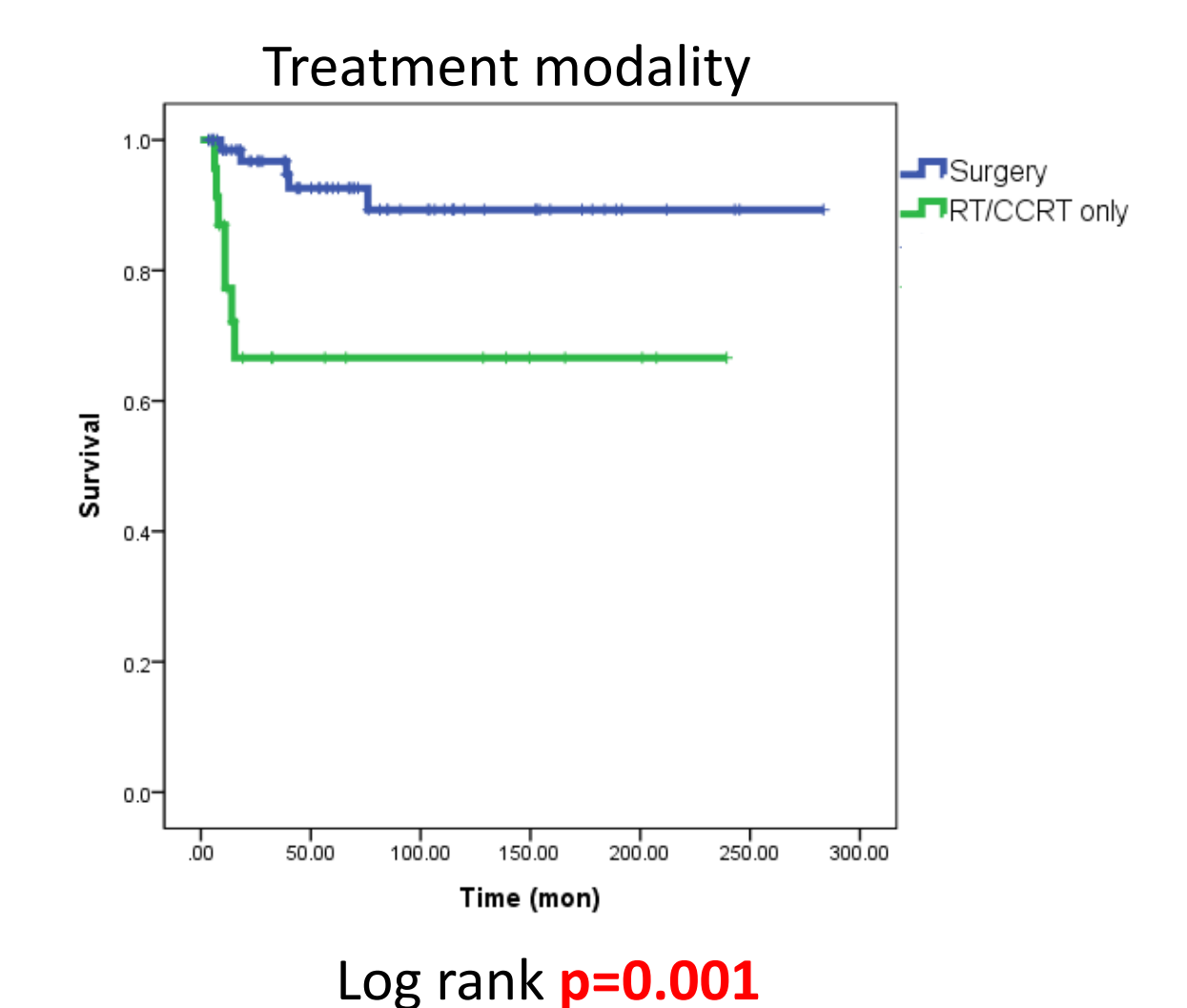
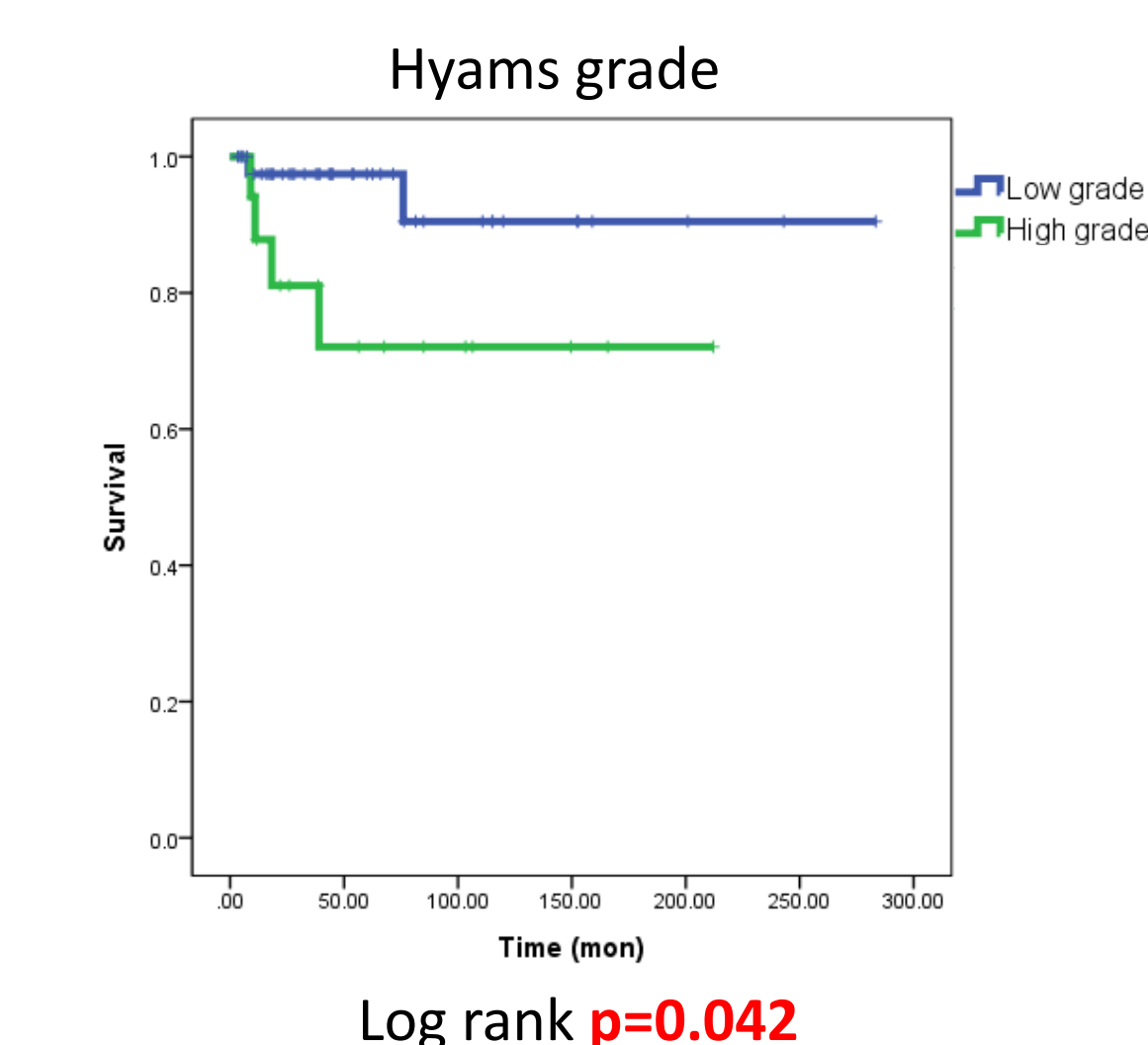
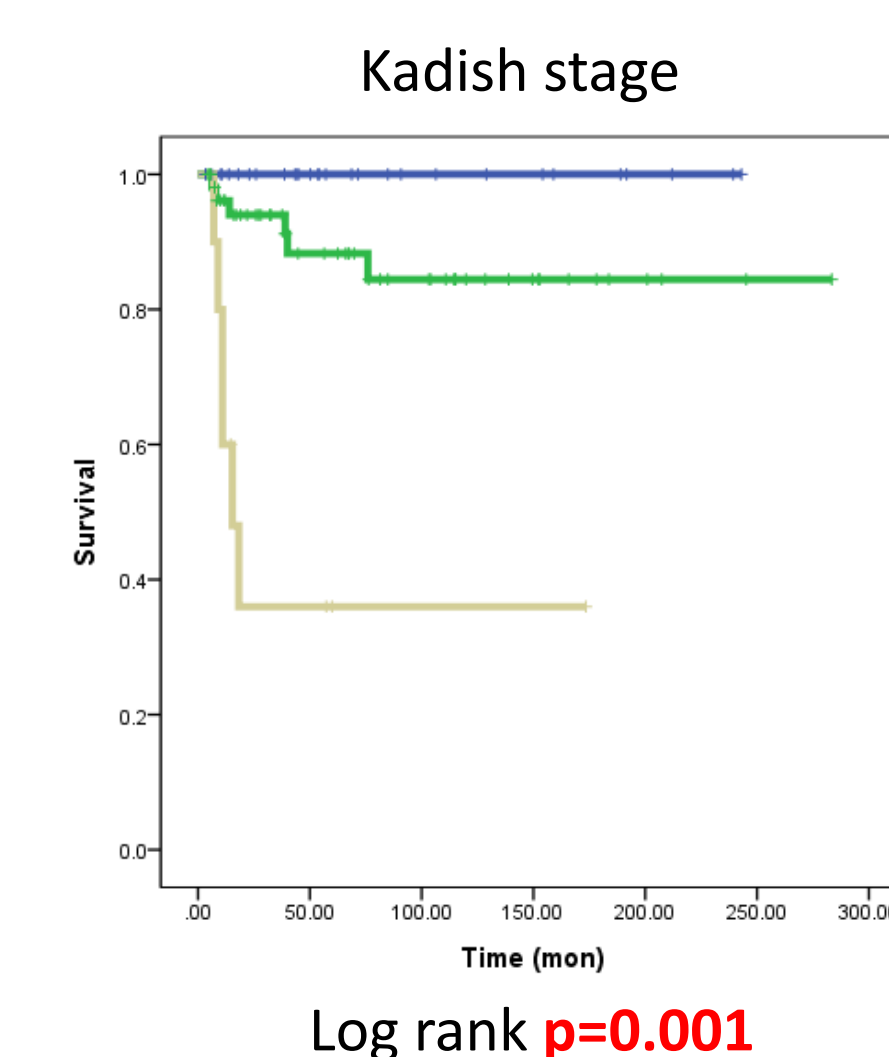
Mean time = 43.7 ± 44.4 (mon)



Survival according to site of recurrence and treatment modality after recurrence



Factors associated with distant recurrence



Conclusion

In ONB, definitive RT without surgical resection, higher tumor stage and grade are associated with poor distant control and affect overall survival. Despite high relapse rate, surgery or RT as a salvage therapy significantly improved overall survival.

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