



Malignant peripheral nerve sheath tumours

AN OUTCOME ANALYSIS OF 32 PATIENTS

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Background

- ▶ Def: Malignant nerve sheath tumour arising from a peripheral nerve, from a pre-existing benign nerve sheath tumour (usually neurofibroma) or in a patient with neurofibromatosis type 1 (WHO, 2013)
- ▶ MPNST – rare, 5% of all STS
- ▶ Biologically aggressive, rapid progression and high mortality rate
- ▶ Malignant Triton tumour - MPNST with rhabdomyosarcomatous differentiation, very aggressive
- ▶ Poor prognostic factors – NF1 associated, truncal location, size > 5 cm, local recurrence, high grade

Significant prognostic factors

Author/ Centre	Year	N	5YOS	NF1	Size	Site	Margin	RTX	CTX
Sordillo/ MSKCC	1981	165	23- 47%	+	NR	+	NR	NR	+
Ducatman/ Mayo	1986	120	34%	+	+	-	NR	-	-
Wong/ Mayo	1998	134	52%	-	+	-	+	+	-
Carli/ Italy	2005	167	37%	+	+	+	-	+	+
Madhabana nda/ India	2006	24	58%	-	NR	NR	NR	+	NR
Okada, Japan	2007	56	43%	-	+	-	-	-	-
Anghileri/ INT-MI	2007	205	NR	-	+	+	-	+	-
Longhi/ Rizzoli	2010	62	38%	-	+	+	+	+	-
Stucky/ Mayo	2012	175	60%	+	+	+	+	-	-

Objectives

- ▶ Identify the prognostic factors for local recurrence and distant metastasis
- ▶ Analyze the survival outcome
- ▶ Identify the prognostic factors related to patients' survival

Materials & Method

Restrospective study in Lerdsin Hospital
from 2009-2017



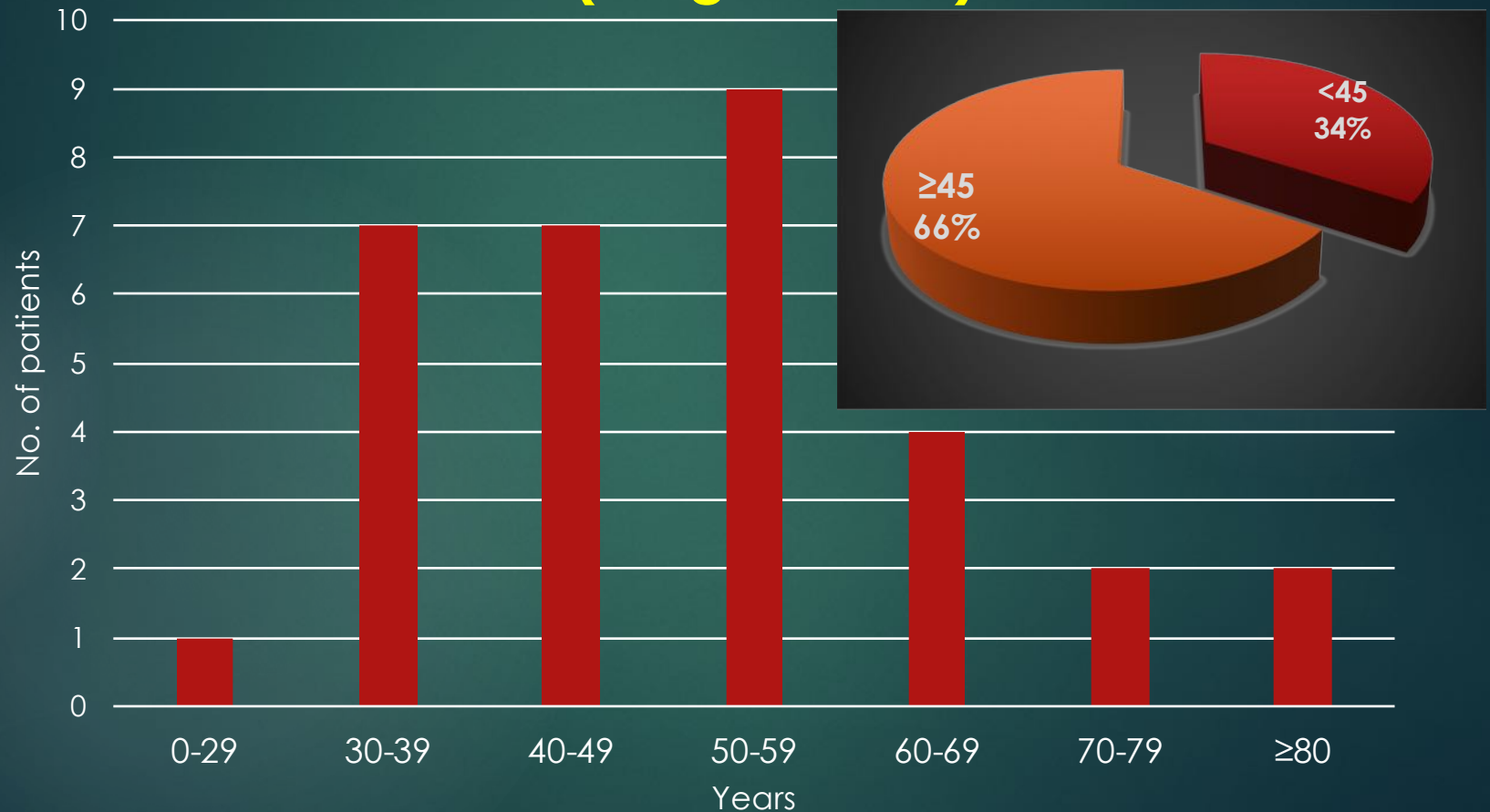
32 patients (mean follow-
up 28 months)



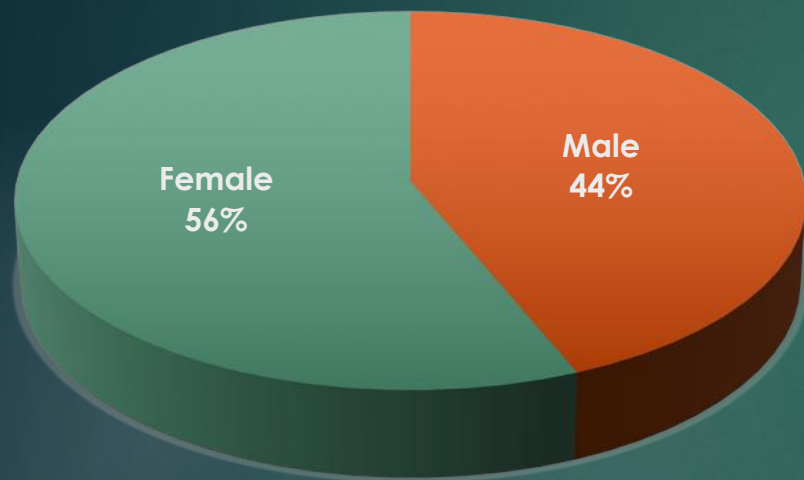
Data collection from
database & patients'
clinical records

Demographic factors

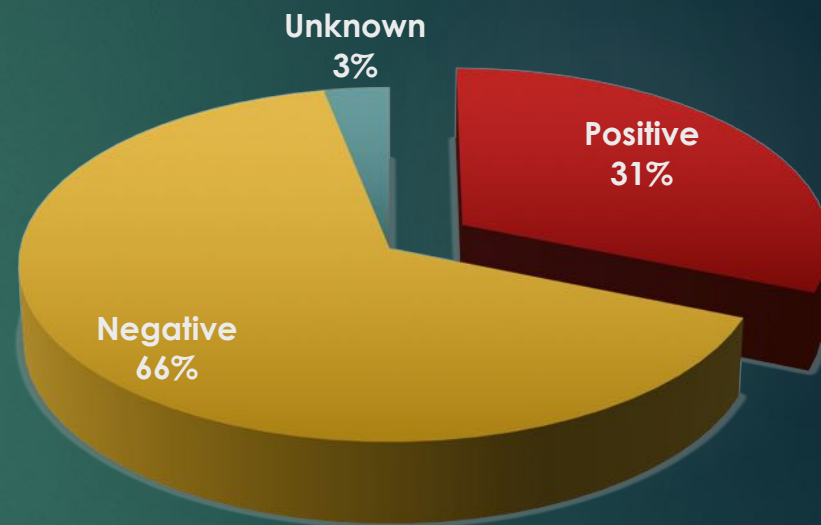
**Mean age, 51 years old
(range, 26-84)**



Sex



NF1 status

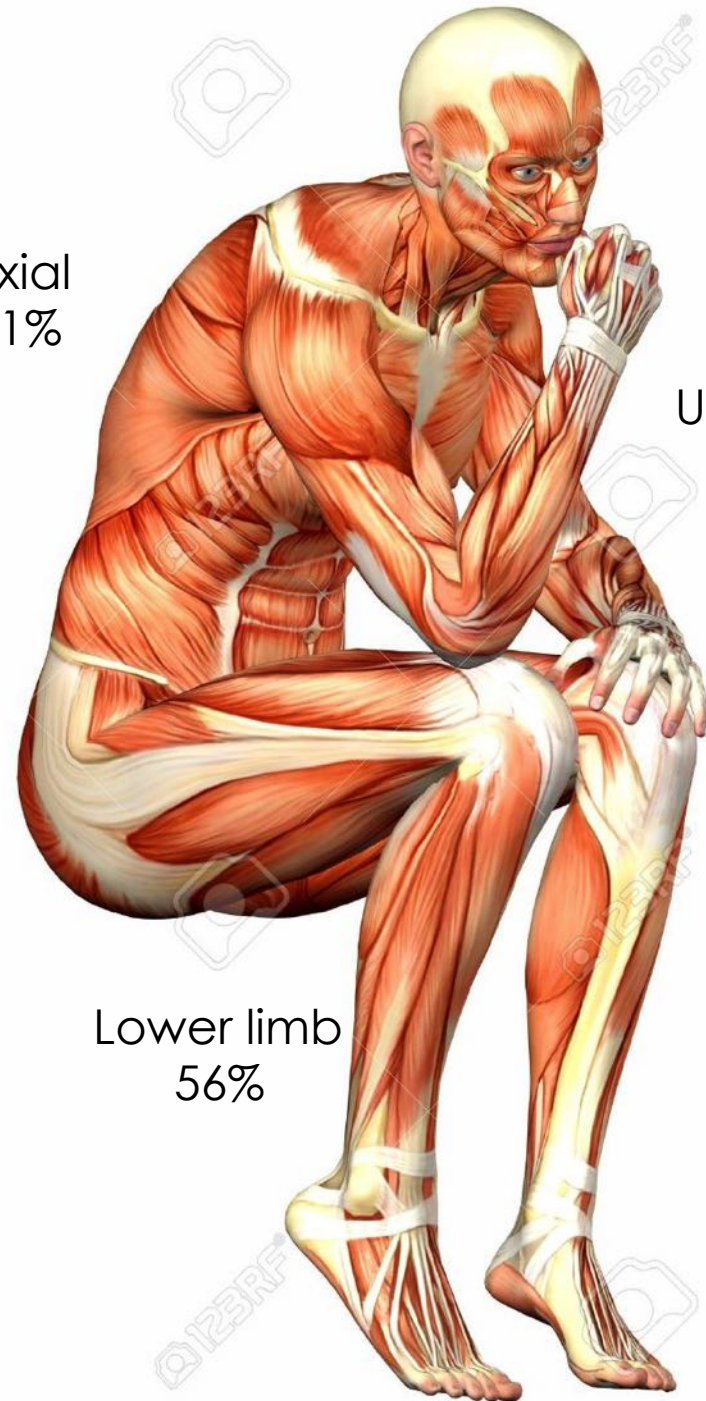


Location

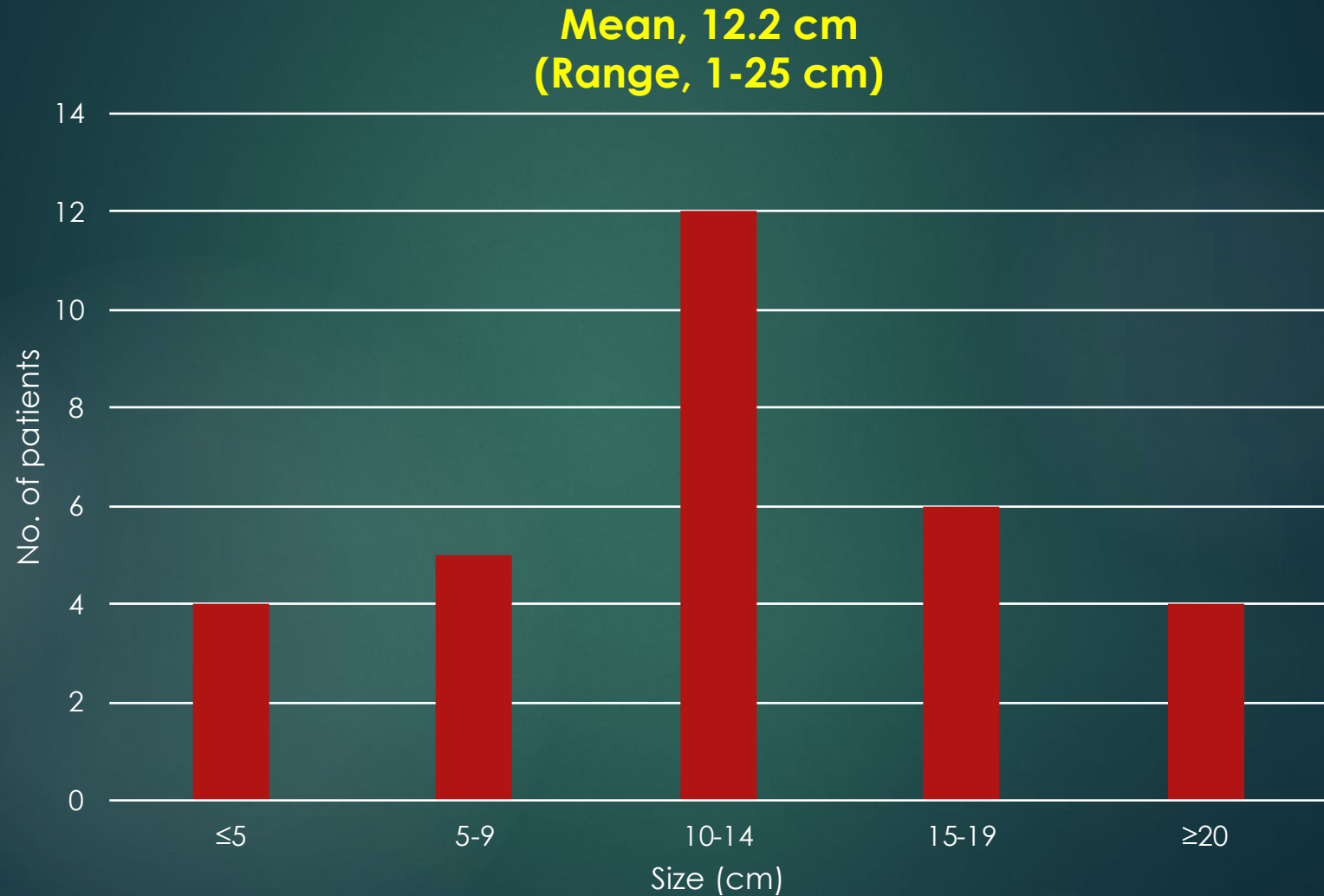
Axial
31%

Upper limb
13%

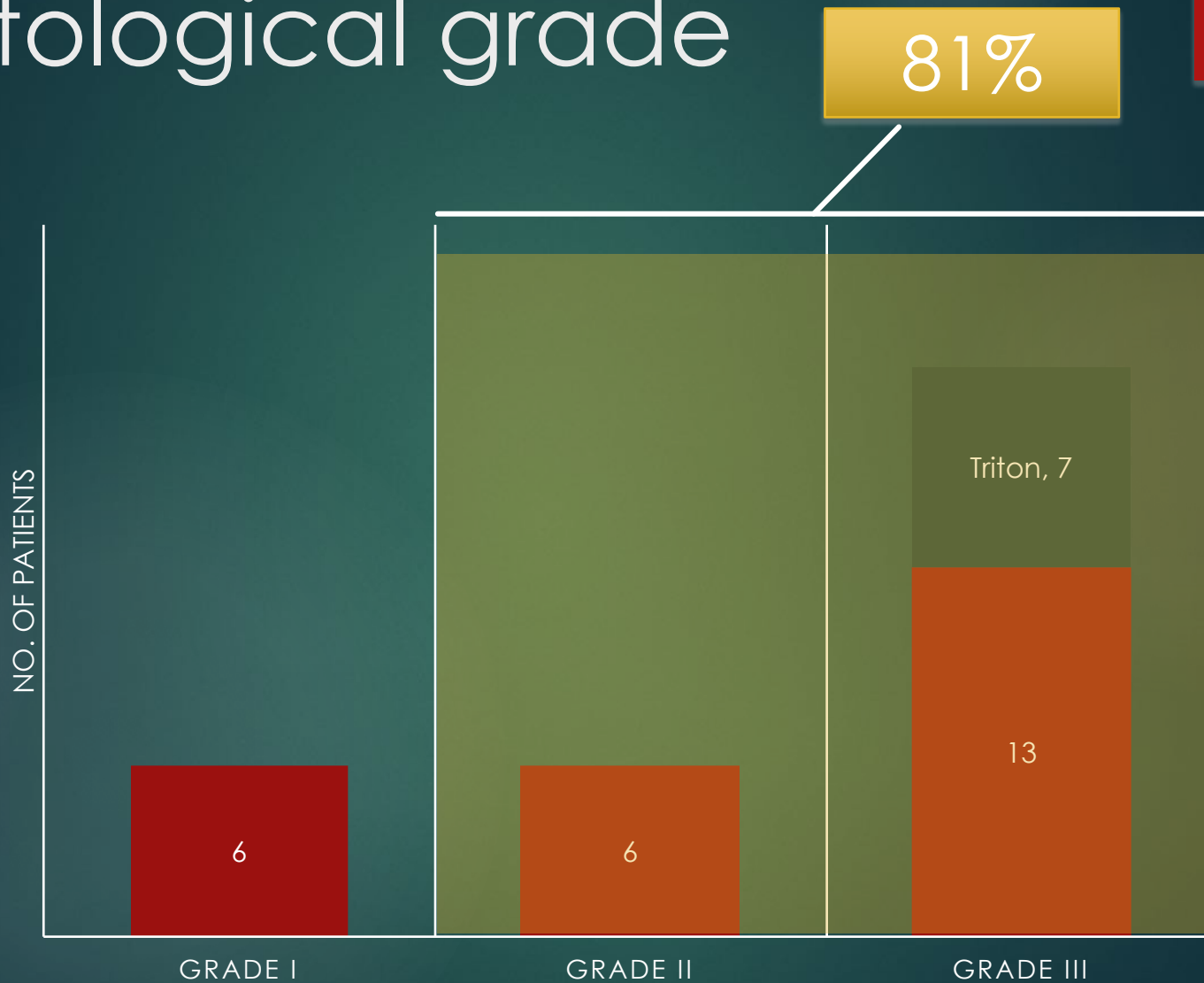
Lower limb
56%



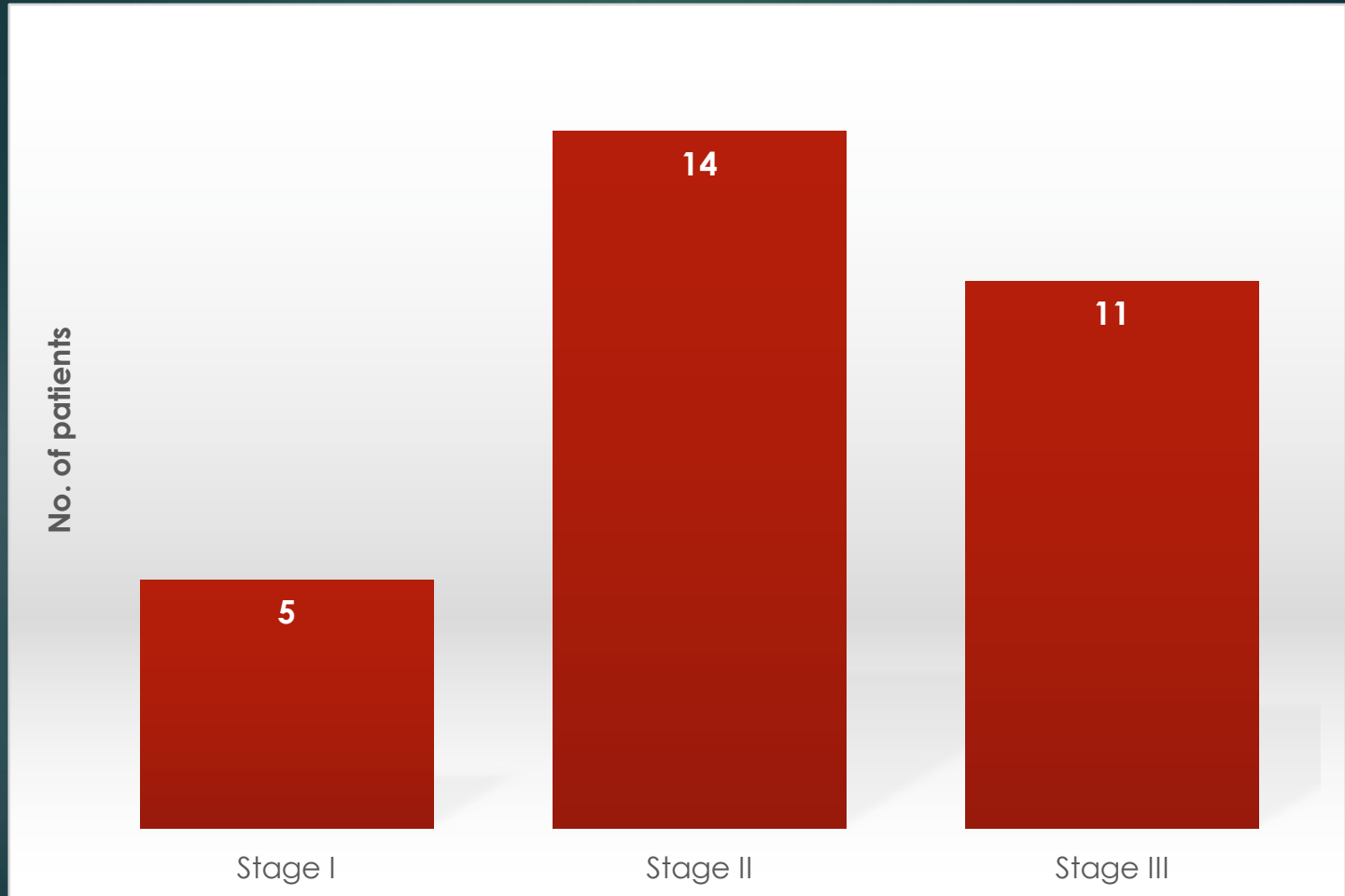
Tumour size



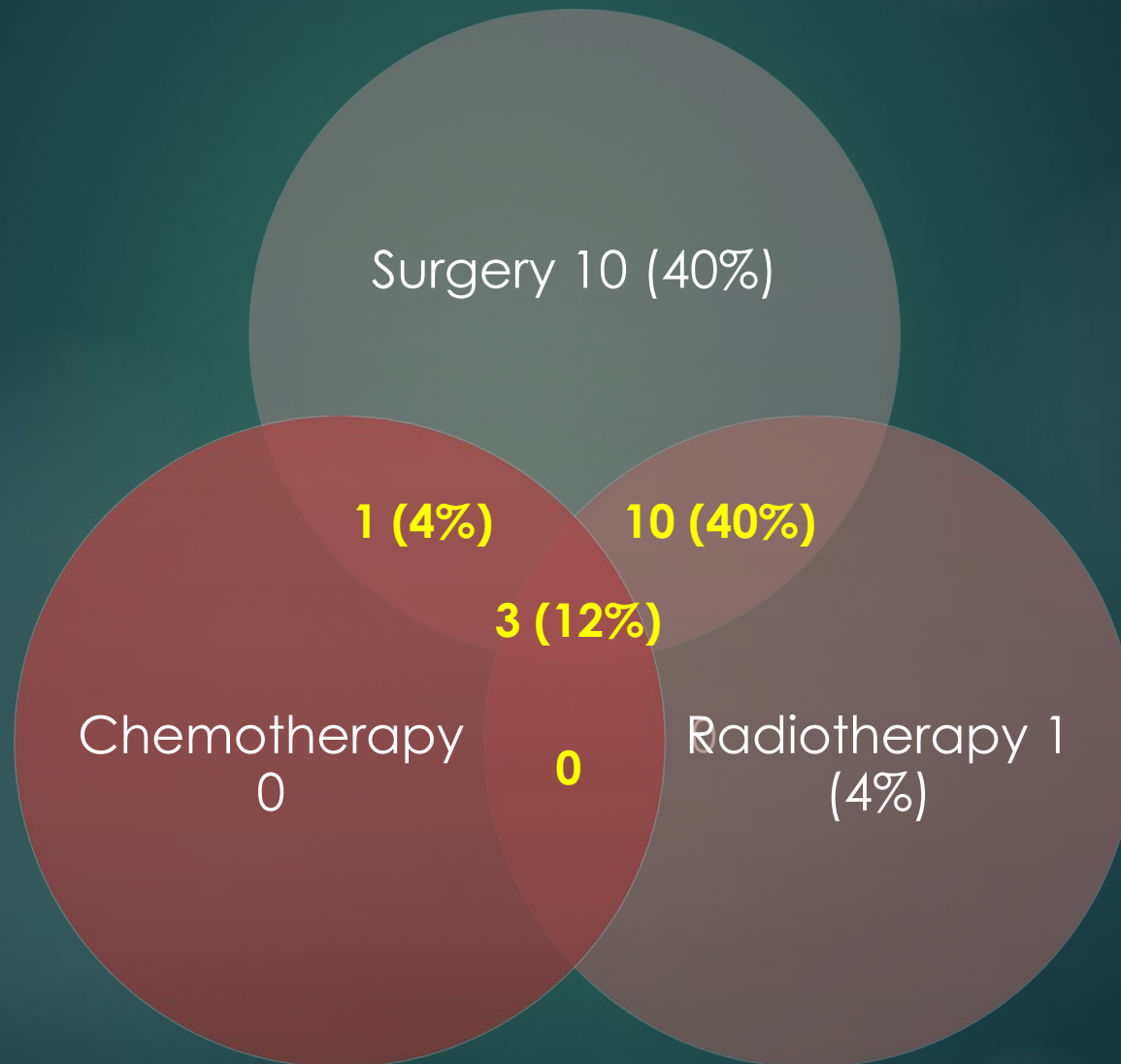
Histological grade



Stage of disease



Treatment



Surgery

Type	N	%
Wide resection	17	57
Sacrectomy/ hemipelvectomy	5	17
Marginal resection	4	13
Amputation	4	13

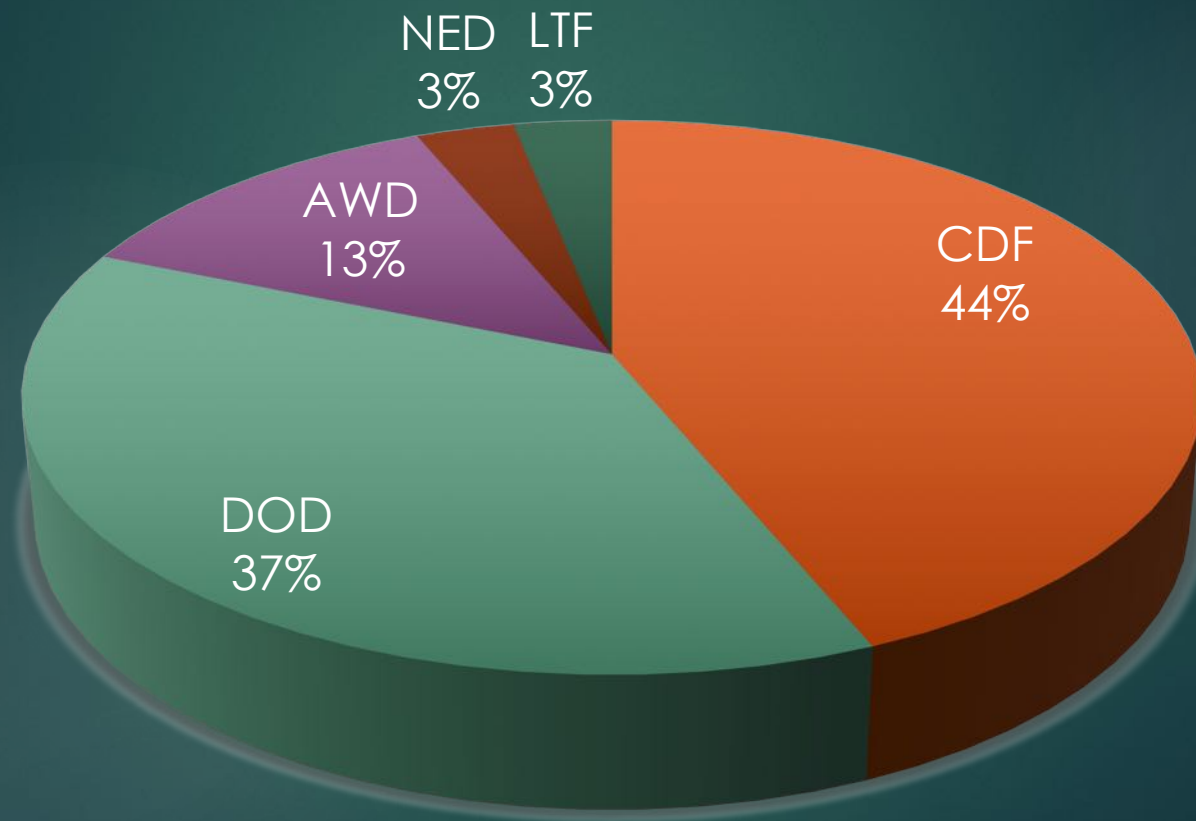
Median survival after surgery
= 56 months



Margin status

Positive	31%
Negative	66%
Unknown	3%

Status at final follow-up



Statistical analysis

$P < 0.05$

1

Univariate analysis

Chi-square test,
Fisher exact test
(expected cell
sizes < 5)

2

Multivariate analysis

Cox proportional
hazards regression
model

3

Survival

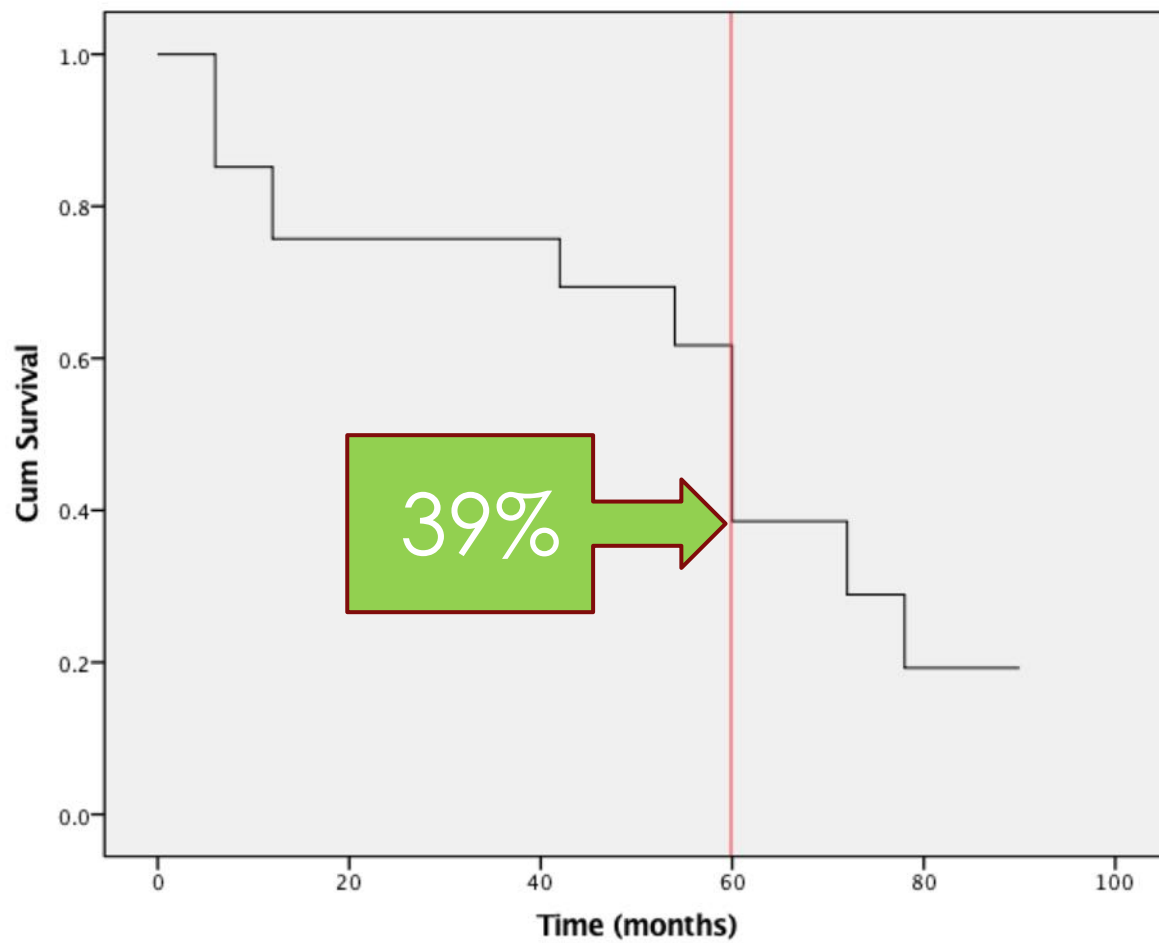
Kaplan-Meier
survival curve

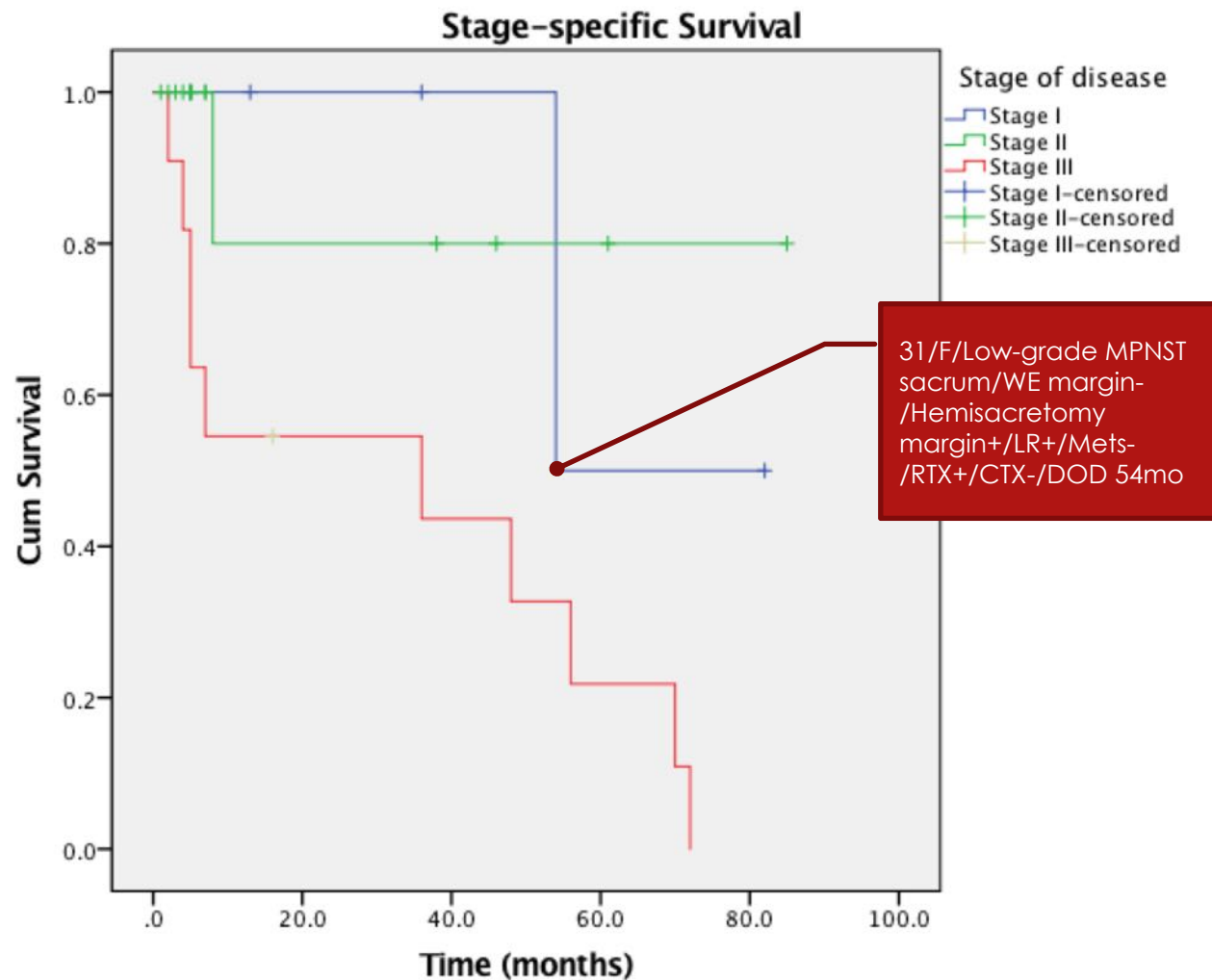
Results

	Local recurrence, N (%)	Mets at presentation, N (%)	Mets, N (%)
Stage I	1 (3.7%)	0	0
Stage II	1 (3.7%)	0	0
Stage III	5 (18.5%)	7 (24.1%)	12 (41.4%)
Overall	7 (25.9%)	7 (24.1%)	12 (41.4%)

Variables		N	LR, n=7 (%)	P-value	N	DM, n=12 (%)	P-value
Age	<45	9	2 (22)	0.571	10	6 (60)	0.140
	≥45	18	5 (28)		19	6 (32)	
Sex	Male	10	2 (20)	0.475	12	8 (67)	0.026
	Female	17	5 (29)		17	4 (24)	
NF-1 status	Positive	8	1 (13)	0.301	9	5 (56)	0.263
	Negative	19	6 (32)		20	7 (35)	
Size	<5 cm	4	1 (25)	0.731	4	0 (0)	0.100
	≥5 cm	23	6 (26)		25	12 (48)	
Location	Upper limb	4	1 (25)	0.273	4	1 (25)	0.732
	Lower limb	14	2 (14)		15	7 (47)	
	Axial	9	4 (44)		10	4 (40)	
Tumour grade	Low	6	1 (33)	0.498	6	0 (0)	0.026
	High	21	6 (29)		23	12 (52)	
Margin status	Negative	17	4 (24)	0.525	18	8 (44)	0.368
	Positive	10	3 (30)		10	3 (30)	
Radiotherapy	Yes	13	5 (38)	0.118	14	7 (50)	0.231
	No	11	1 (9)		11	3 (27)	
	Unknown	8			7		
Chemotherapy	Yes	4	1 (25)	0.731	4	2 (50)	0.477
	No	23	6 (26)		23	8 (35)	
	Unknown	5			5		

Overall Survival

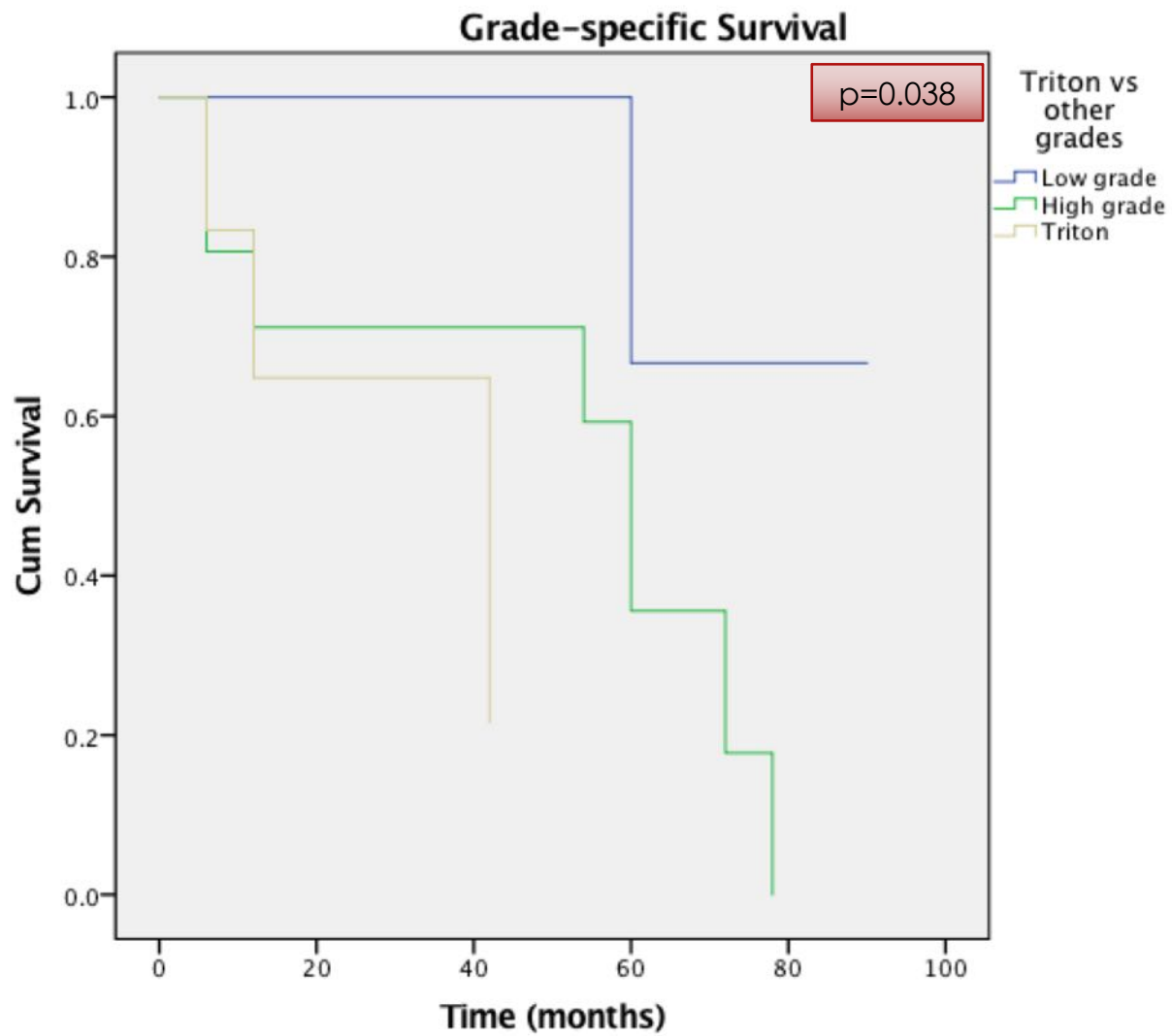


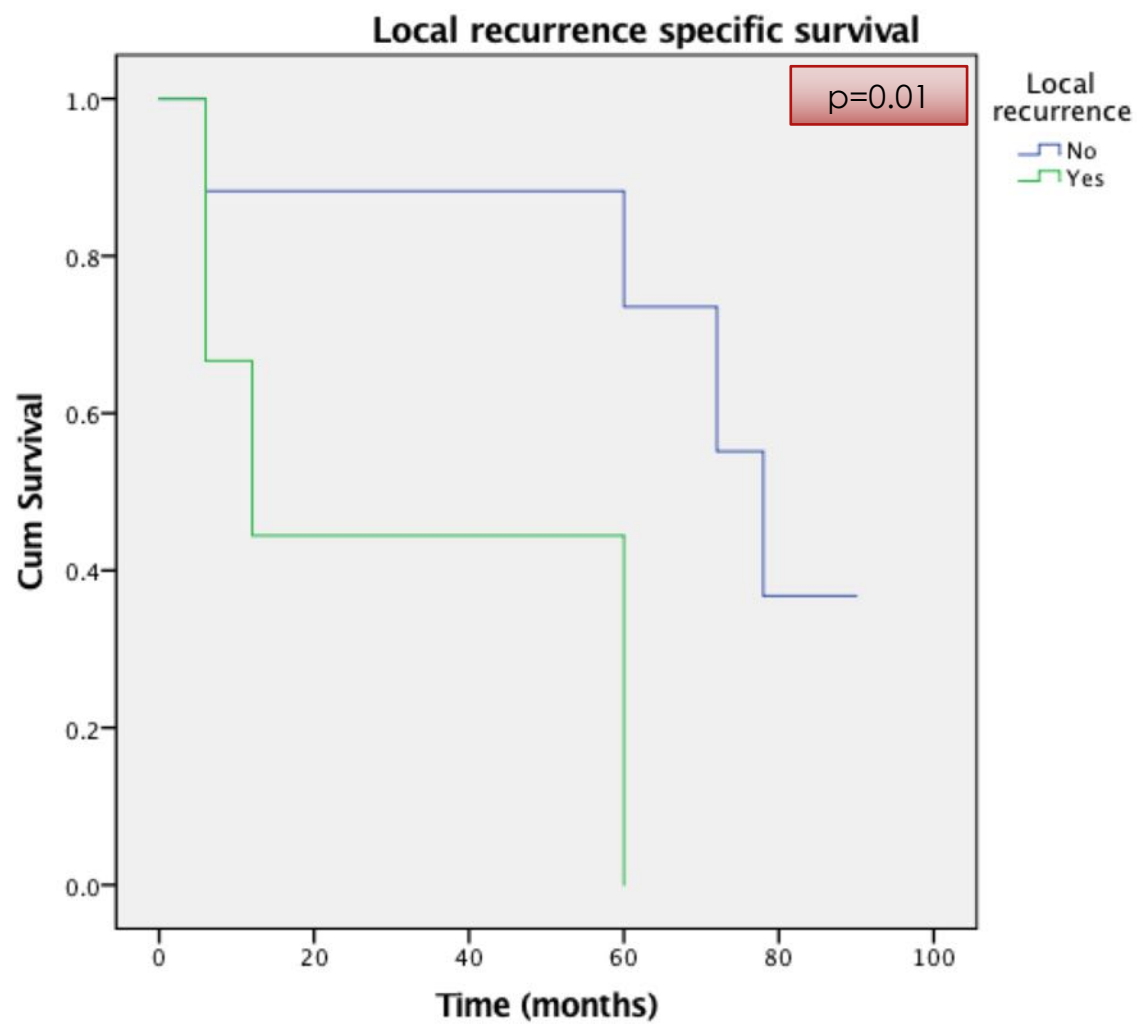


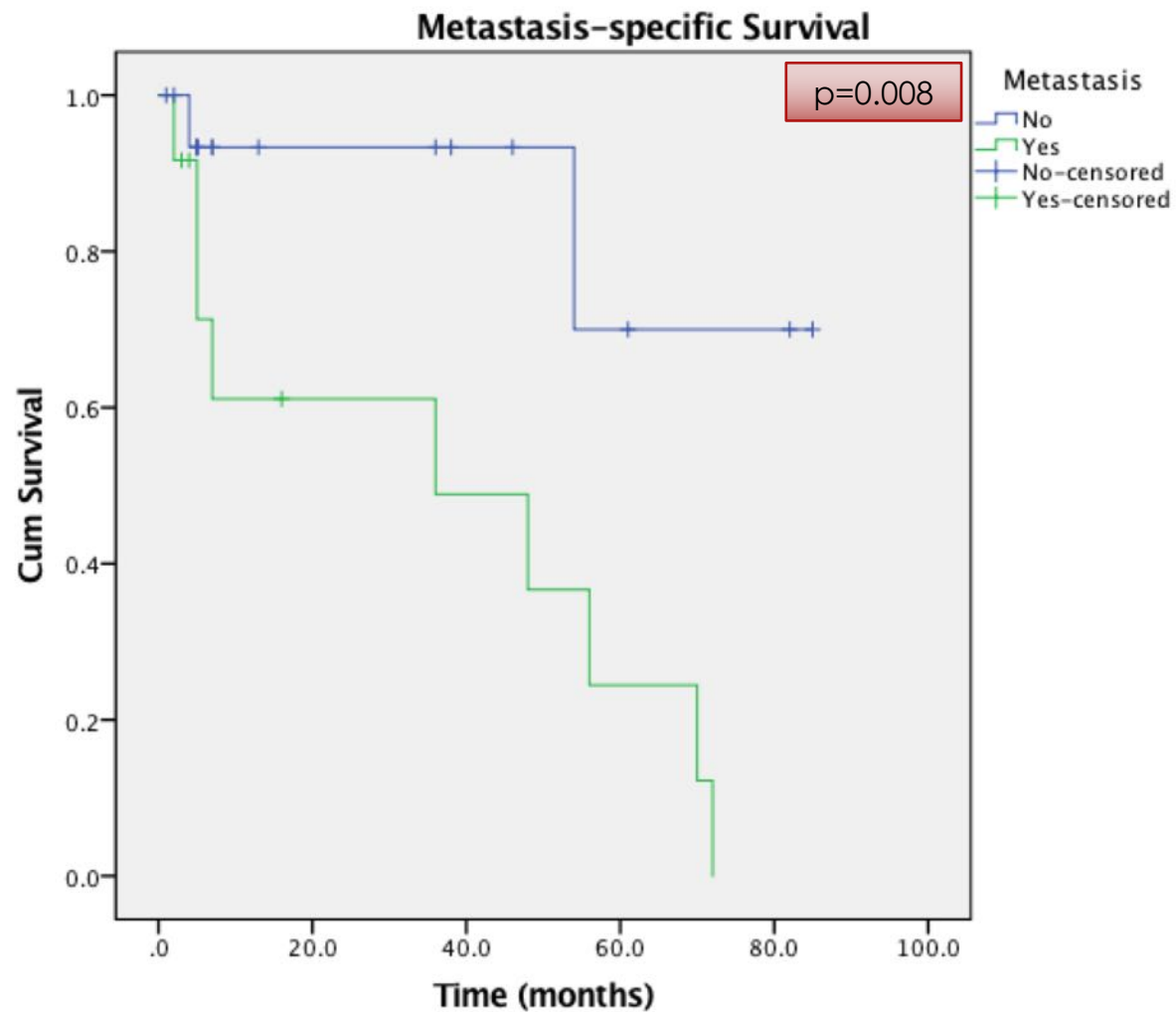
Survival analysis

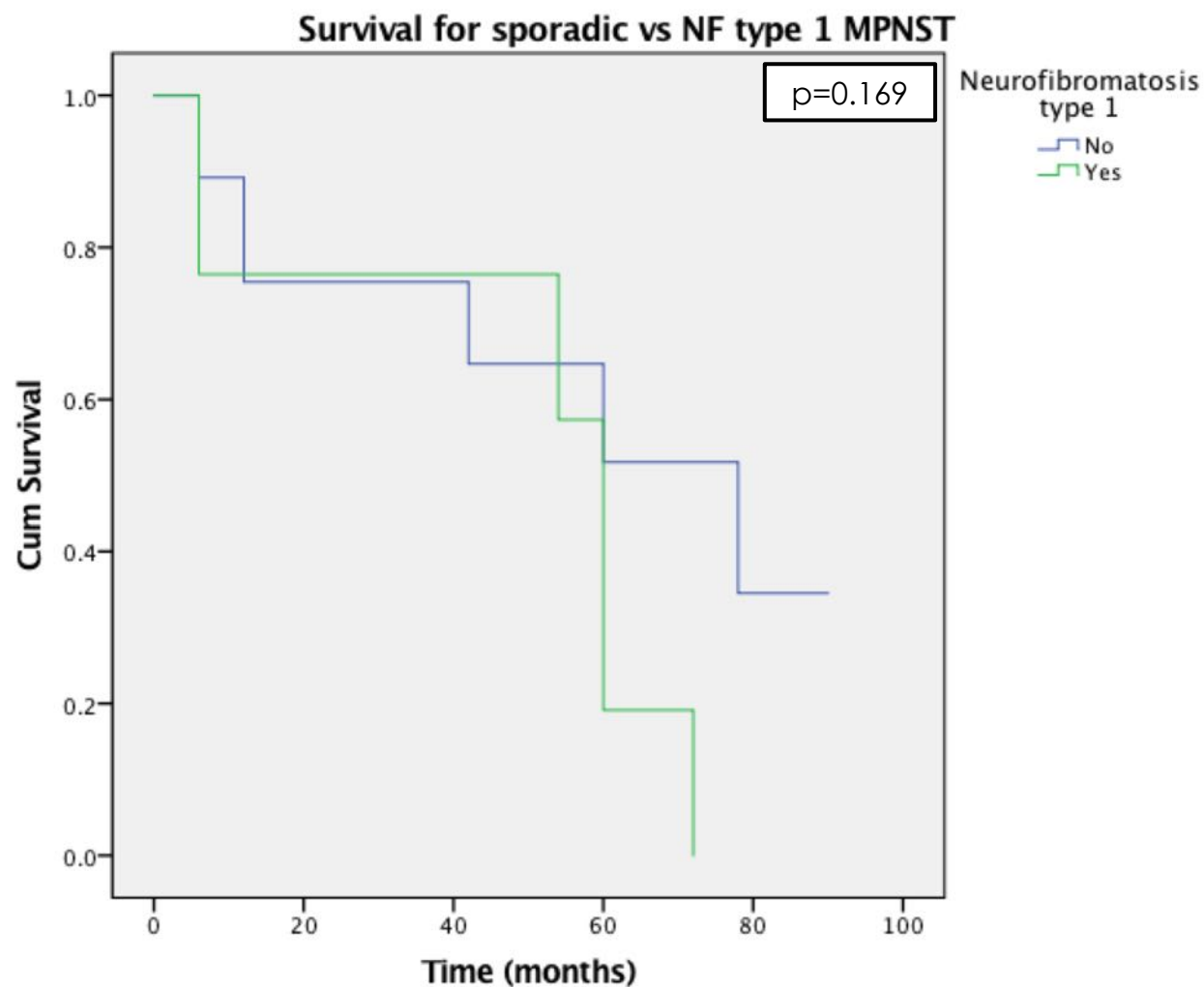
Factors	Univariate analysis	
	5-year DSS	P-value ^a
Tumour grade Low High	67% 30%	0.038
Local recurrence Yes No	0% 74%	0.01
Distant metastasis Yes No	25% 70%	0.008

^a Log-rank analysis









Cox-regression analysis

Factors	Crude HR	Adjusted HR	95% CI	P-value
Age (<45 vs ≥ 45)	0.464			
NF1	2.166	7.962	0.9-69.9	0.061
Grade (low vs high)	6.747	16.945	0.9-321.8	0.06
Triton tumour	2.617			
Local recurrence	6.884	20.035	1.8-222.8	0.015
Metastasis	6.131			
Chemotherapy	2.603			

Discussion

	Longhi/ Rizzoli	Stucky/ Mayo	Lerdsin
Age	Median, 39	Median, 44	Mean, 51
NF1	35%	32%	31%
Site – extremity axial	77% 23%	45% 34%	69% 31%
Size	Median, 9cm (2-30)	Median, 6cm	Mean, 12 cm (1-25)
Grade – high Triton	NR 2%	61% 3%	81% 22%
Margin - positive	44%	11%	31%
Adjuvant Rx – RTX CTX	48% 29%	63% 28%	56% 16%
Local recurrence	34%	22%	26%
Metastasis	40%	21%	41%
5YOS	38%	60%	39%

Conclusion

- ▶ High grade tumour, local recurrence and distant metastasis – poor prognostic factors for survival
- ▶ Tumour size, surgical margin and adjuvant therapy – no prognostic influence on the survival outcome
- ▶ Malignant triton tumour – highest cases compared to other studies and has lowest survival
- ▶ Need larger multi-institutional study due to rarity of MPNST