



**42<sup>nd</sup> SIOP Congress**

**IPSO Meeting and Conferences**

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# **DOES SURGERY HAVE A ROLE IN THE TREATMENT OF LOCAL RELAPSED RMS?**

**Experience of the Italian STS Committee**

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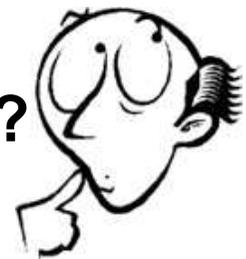
# Introduction

- ✓ Outcome for localized RMS improved during last decades due to successful advances in multidisciplinary treatment
- ✓ Currently 5-years OS ~ 70%
- ✓ 1<sup>st</sup> CR is reached in > 90%
- ✓ Relapses occur in 1/3, mostly local

# Introduction

- ✓ 1<sup>st</sup> line treatment: CT + surgery and/or XRT depending on a risk-factor analysis
- ✓ 2<sup>nd</sup> line treatment is not standardized as 1<sup>st</sup> line
- ✓ Essential role of surgery in 1<sup>st</sup> line treatment

**Is it important in local recurrences treatment too?**







# Clinical Features

## 70 patients

Sex	43 M, 27 F
Age at 1 <sup>st</sup> diagnosis	1 month – 16 years (med 4 years)
Age at relapse	11 months – 20 years (med 7 years)
Time free from disease	3 weeks – 134 months (med 11 months)

# Clinical Features

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<b>SITE</b>	<b>70 pts</b>
Orbit	12
HNnPM	10
HN PM	9
GU BP	7
GUnBP	7
Extremities	7
Other Sites	18

<b>HISTOLOGY</b>	<b>70 pts</b>
Embryonal	41
Alveolar	17
Botryoid	8
Leiomyomatous	2
NOS	2

# Initial Treatment

## 70 patients

Sex 43 M, 27 F  
Age at 1<sup>st</sup> diagnosis 1 month – 16 years (med 4 years)  
Age at relapse 11 months – 20 years (med 7 years)  
Time free from disease 3 weeks – 134 months (med 11 months)

<b>IRS Stage</b>	<b>70 pts</b>
IRS I	9
IRS II	10
IRS III	51

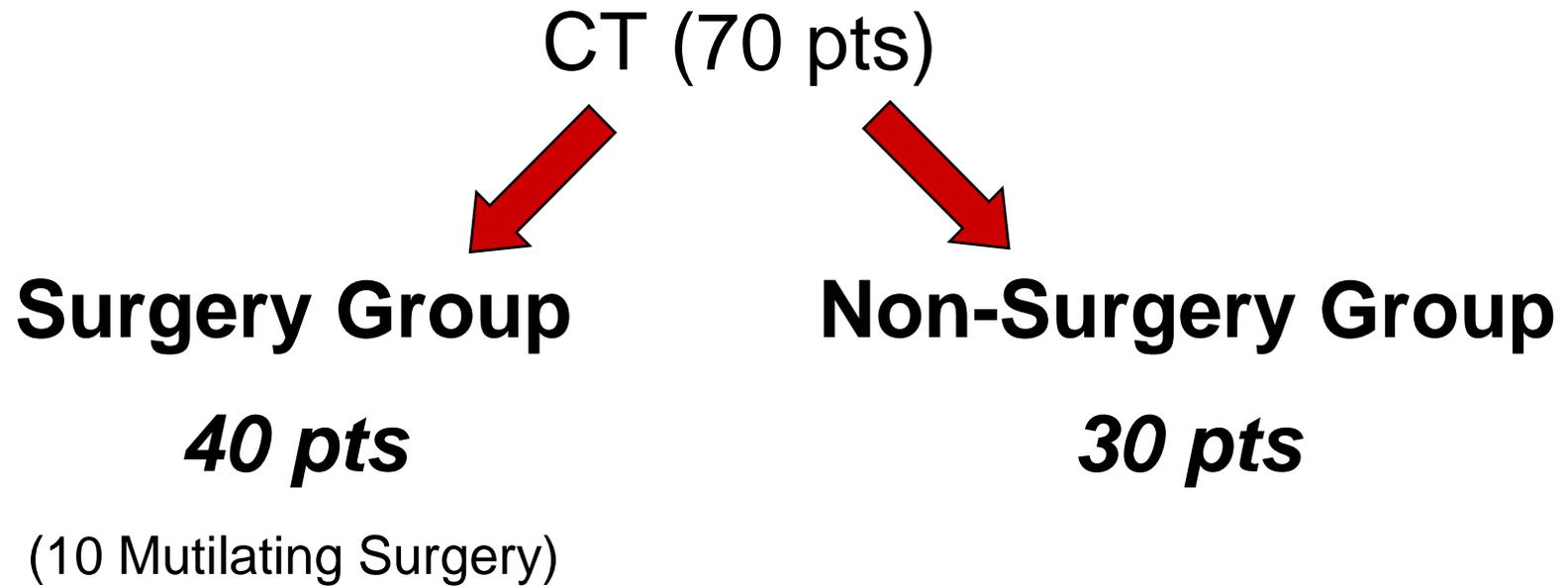
<b>1<sup>st</sup> Line Local Treatm.</b>	<b>70 pts</b>
Surgery	28
Surgery + XRT	16
XRT	17
None	9

A horizontal banner image showing a sunset over a mountain range. The sun is low on the horizon, casting a warm orange glow across the sky and illuminating the silhouettes of the mountains. The text 'Treatment after LR' is overlaid in the center of the banner.

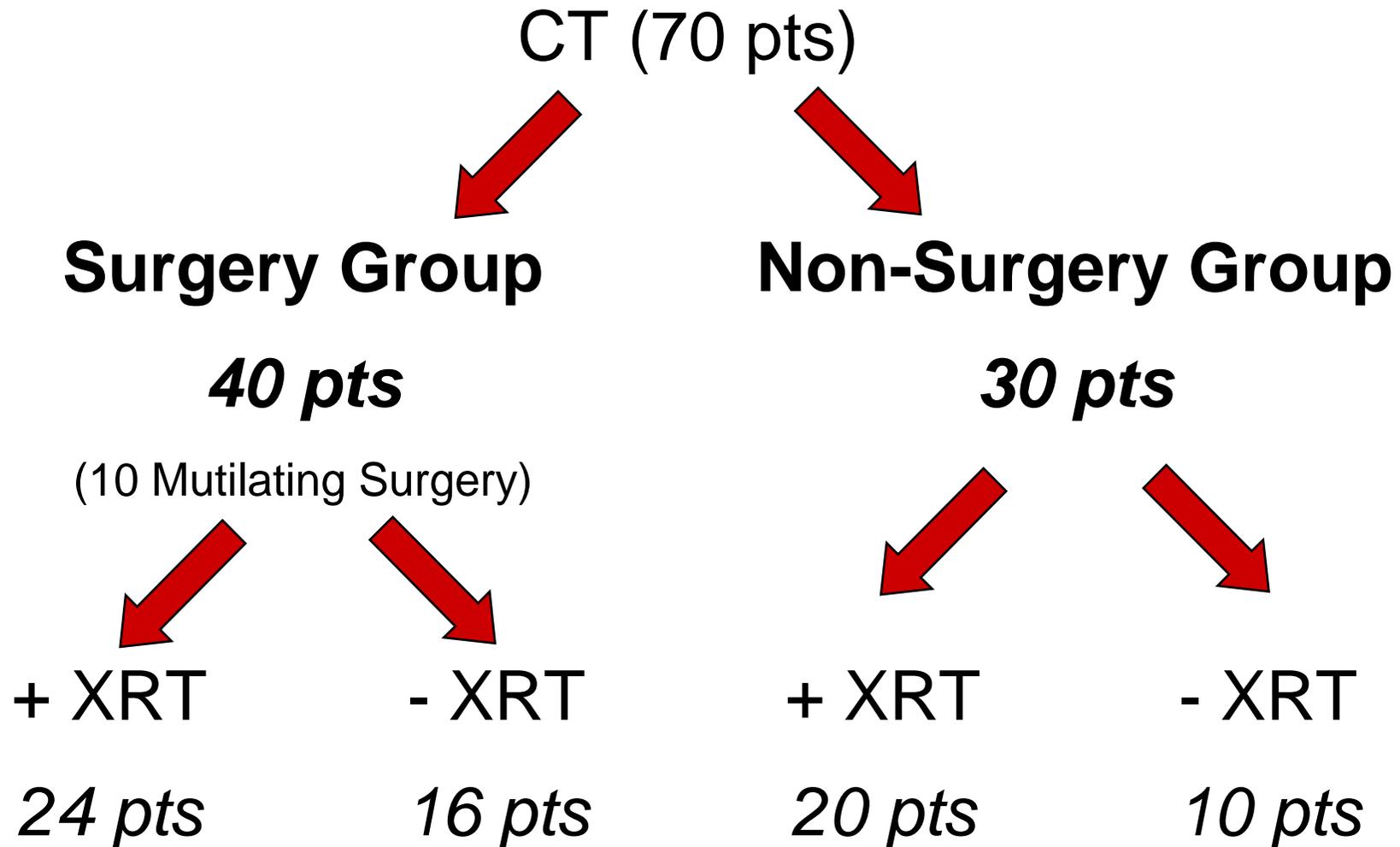
# Treatment after LR

CT (70 pts)

# Treatment after LR



# Treatment after LR



# Results

	SG		NS	
	+XRT	-XRT	+XRT	-XRT
<b>SITE</b>				
Orbit	3**	3***	6	--
HNnPM	4	1	4	1
HN PM	1*	2	1	5
GU BP	2**	4**	--	1
GUnBP	3 (2M,1F)	1 (1M)	3 (1M,2F)	--
Extremities	4	2	--	1
Other Sites	6	4	6	2

\* = demolitive surgery

	SG		NS	
	+XRT	-XRT	+XRT	-XRT
<b>HISTOLOGY</b>				
Embryonal	14***	9**	12	12
Alveolar	5	4*	4	4
Botryoid	4**	2*	2	--
Leiomyom.	--	1*	1	--
NOS	1	--	1	--

# Results

	SG		NS	
	+XRT	-XRT	+XRT	-XRT
<b>IRS St.</b>				
IRS I	5	4*	--	--
IRS II	2*	2*	5	1
IRS III	16****	11***	15	9

	SG		NS	
	+XRT	-XRT	+XRT	-XRT
<b>1°Line Local T.</b>				
Surgery	11*	6*	10	1
Surgery+XRT	7**	6**	3	--
XRT	2	4**	2	9
None	3**	1	5	--

\* = demolitive surgery



# Demolitive Surgery

**10 patients**

- **5 exenteratio orbitae** (2 +XRT): in 1<sup>st</sup> line 1 IRS I, 4 IRS III
- **4 cystoprostatectomy** (2 +XRT): in 1<sup>st</sup> line 1 IRS II, 3 IRS III (1 underwent cystoprostatectomy and reconstructive surgery during 1<sup>st</sup> line delayed surgery, 1 received XRT)
- **1 mastoidectomy** (+XRT): in 1<sup>o</sup>line IRS II and XRT



# Outcome

**Follow-up (after LR): 2 weeks – 19 years (med 29 months)**

- 27 pts reached 2<sup>nd</sup> CR
- 1 pt presented further recurrences, but is actually in CR
- 42 pts DOD (after 2 weeks-65 months from the LR, med 17 months)

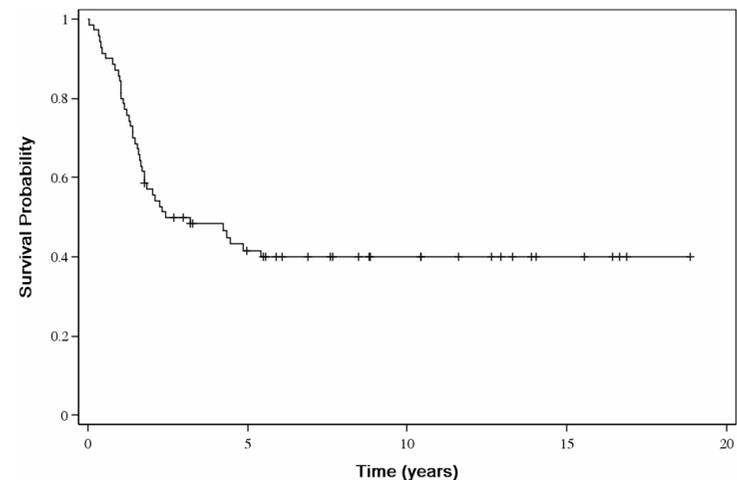
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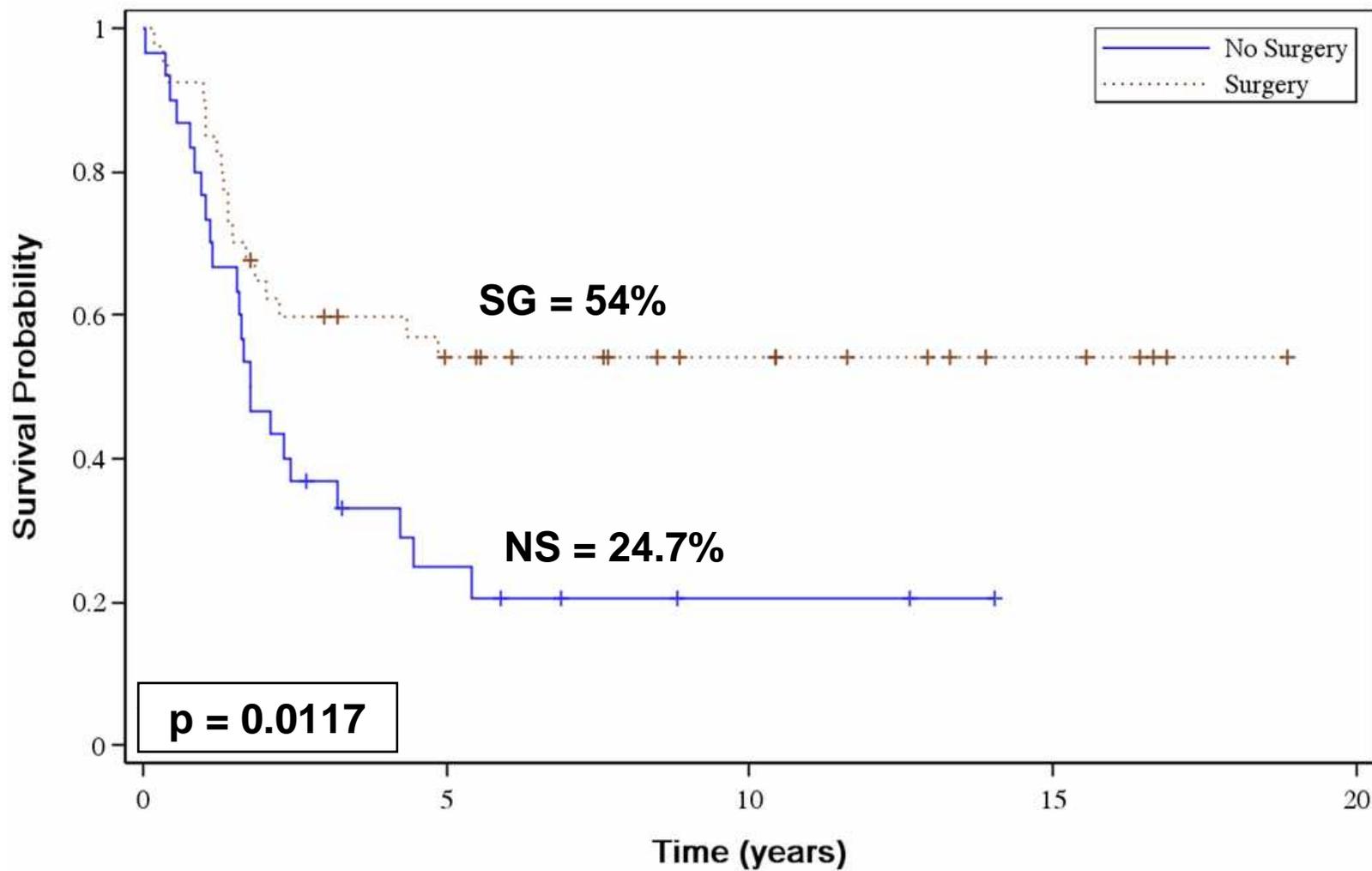
- 27 pts reached 2°CR
- 1 pt presented further recurrence, but is actually in CR
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**5-years OS = 41.6% (95%CL 29.7-53%)**

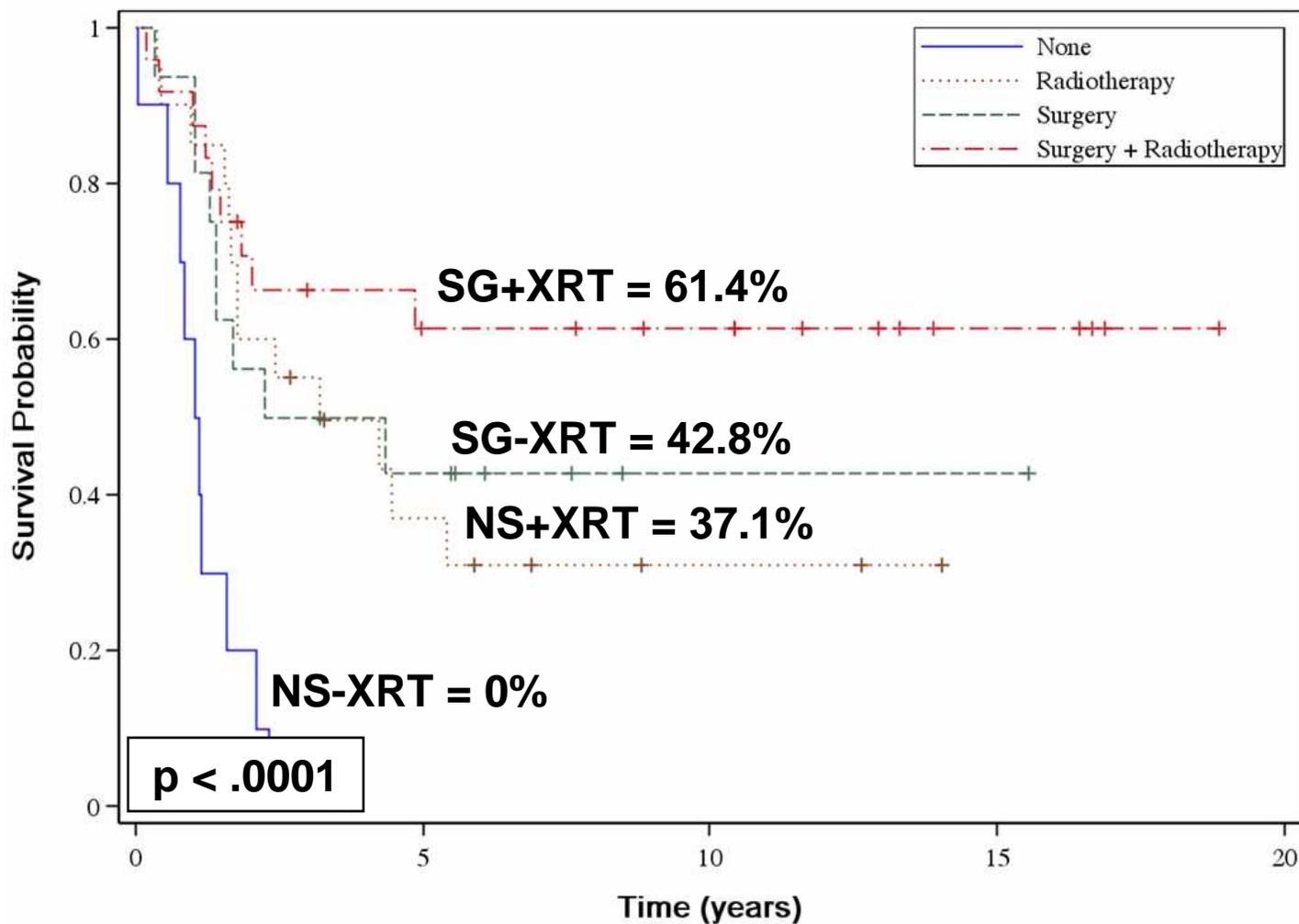
**10-years OS = 39.8% (95%CL 28.1-51.3%)**



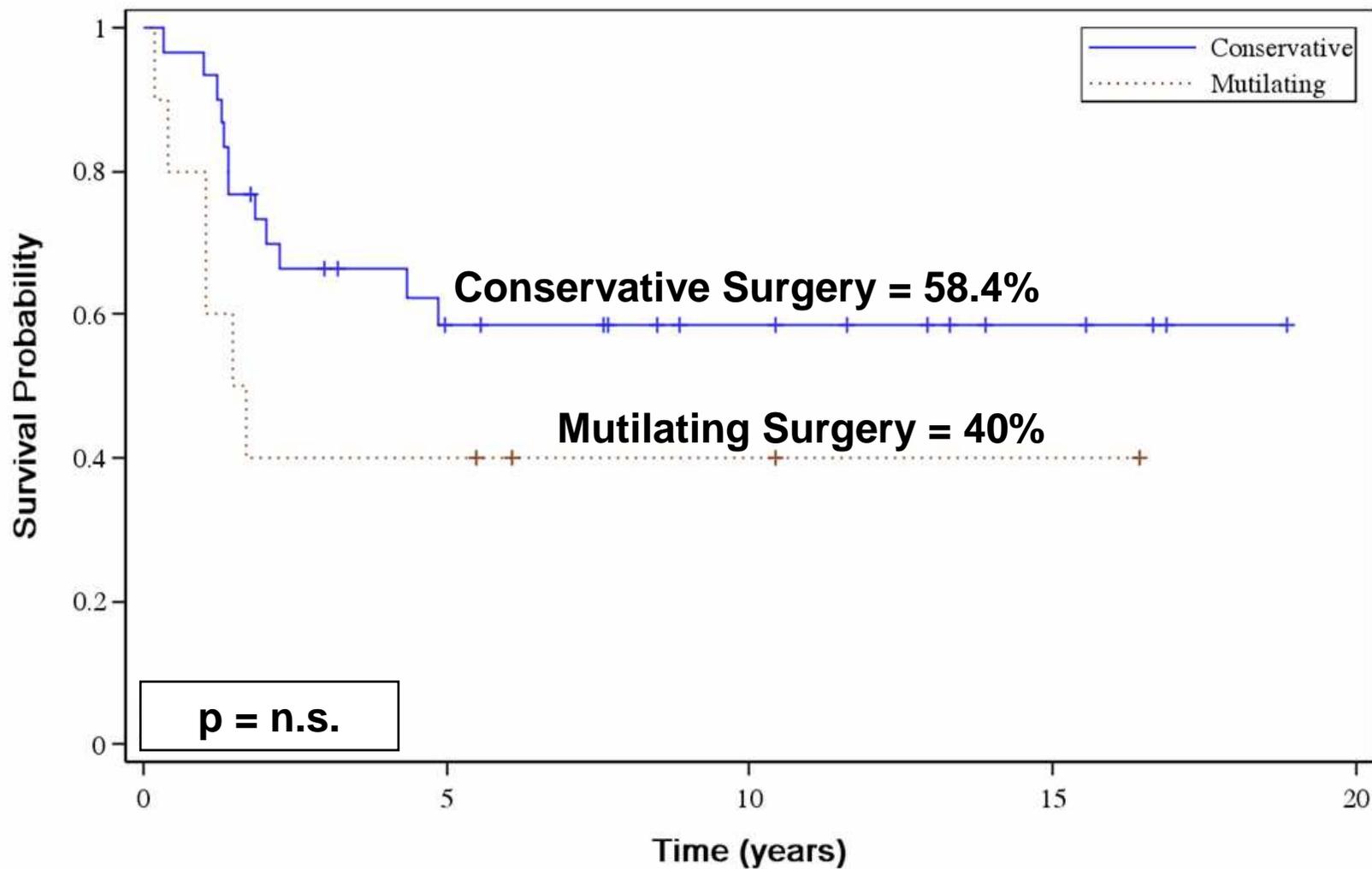
# 5-years OS: Surgery



# 5-years OS: Surgery and/or XRT



# 5-years OS: Demolitive Surgery





# Conclusions

- ✓ Patients with LR have a severe prognosis (OS~40%)
- ✓ The treatment of recurred RMS represents a challenge
- ✓ Multimodal treatment is the best choice, as in 1<sup>st</sup> Line Treatment

**... but ...**

- ✓ SG patients had a better outcome than NS patients ( $p=0.0117$ )
- ✓ Those treated with resection + XRT had the best outcome ( $p<.0001$ )
- ✓ Those without any local treatment had unfavourable outcome (OS~0%)
- ✓ Demolitive Surgery seems to not improve survival (40% vs 60%)



# Take Home Message

- ✓ Several factors are important in determining the survival after RMS LR
- ✓ The efficacy of surgery is influenced by other variables
- ✓ Surgery, whether feasible, offers a better chance to cure
- ✓ **Surgeons should be encouraged to evaluate the feasibility of surgical excision of RMS LR as part of a multimodal treatment**

A scenic view of a mountain range seen through a dark, rocky opening, with the text "Thank you for your attention" overlaid in a cursive font. The image shows a dark, shadowed interior space on the left and right, framing a bright, clear view of a mountain landscape. The mountains are covered in green vegetation and have rugged, rocky peaks. The sky is blue with some white clouds. The text is written in a black, cursive font and is centered in the lower half of the image.

*Thank you for your attention*

# Surgery and/or XRT

	<b>Surgery</b>	<b>No Surgery</b>	<b>Total</b>
<b>XRT</b>	15/24 <b>(63%)</b>	6/20 <b>(30%)</b>	21/44 <b>(48%)</b>
<b>No XRT</b>	7/16 <b>(44%)</b>	0/10 <b>(0%)</b>	7/26 <b>(27%)</b>
<b>Total</b>	22/40 <b>(55%)</b>	6/30 <b>(20%)</b>	28/70 <b>(40%)</b>