Desmoid Fibromatosis Tumors: The Role of Surgery

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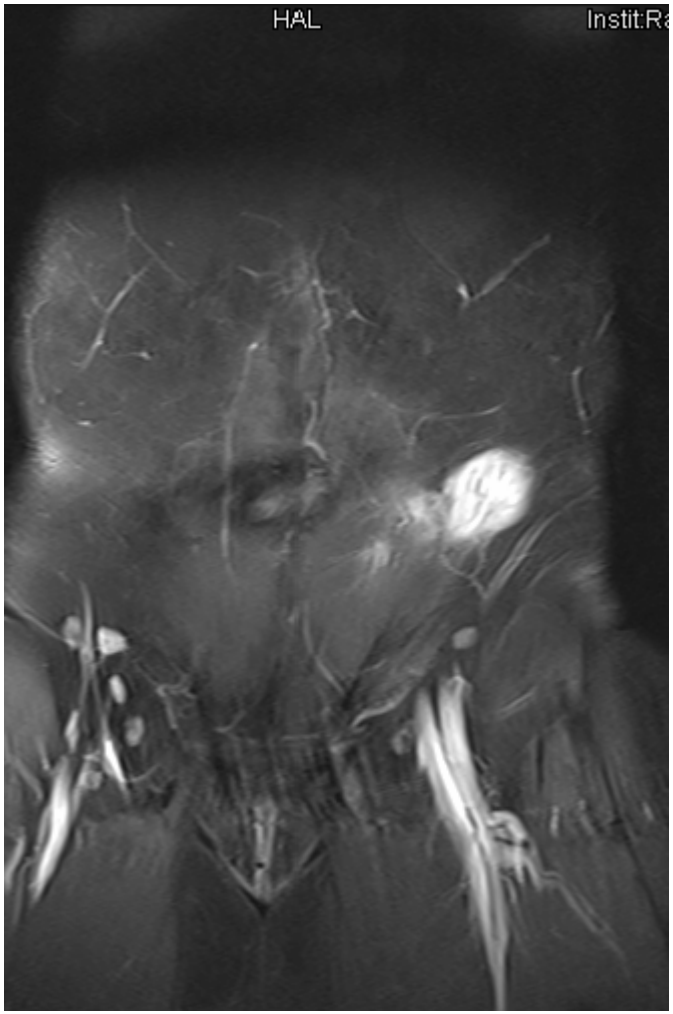

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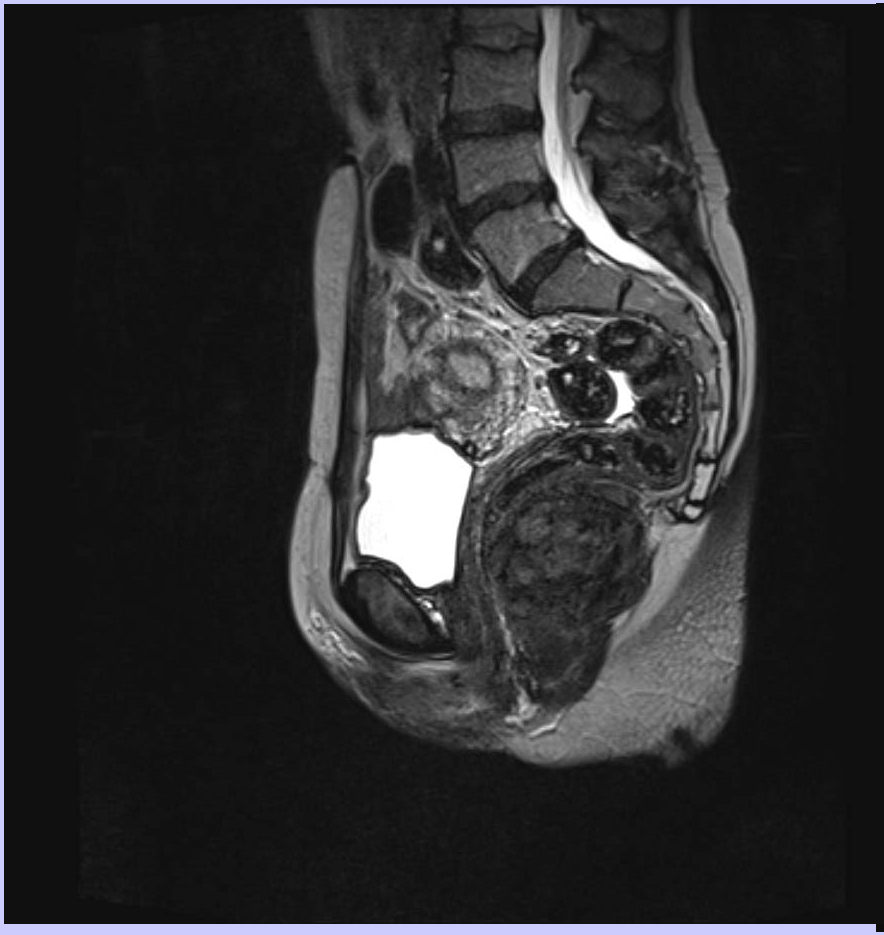

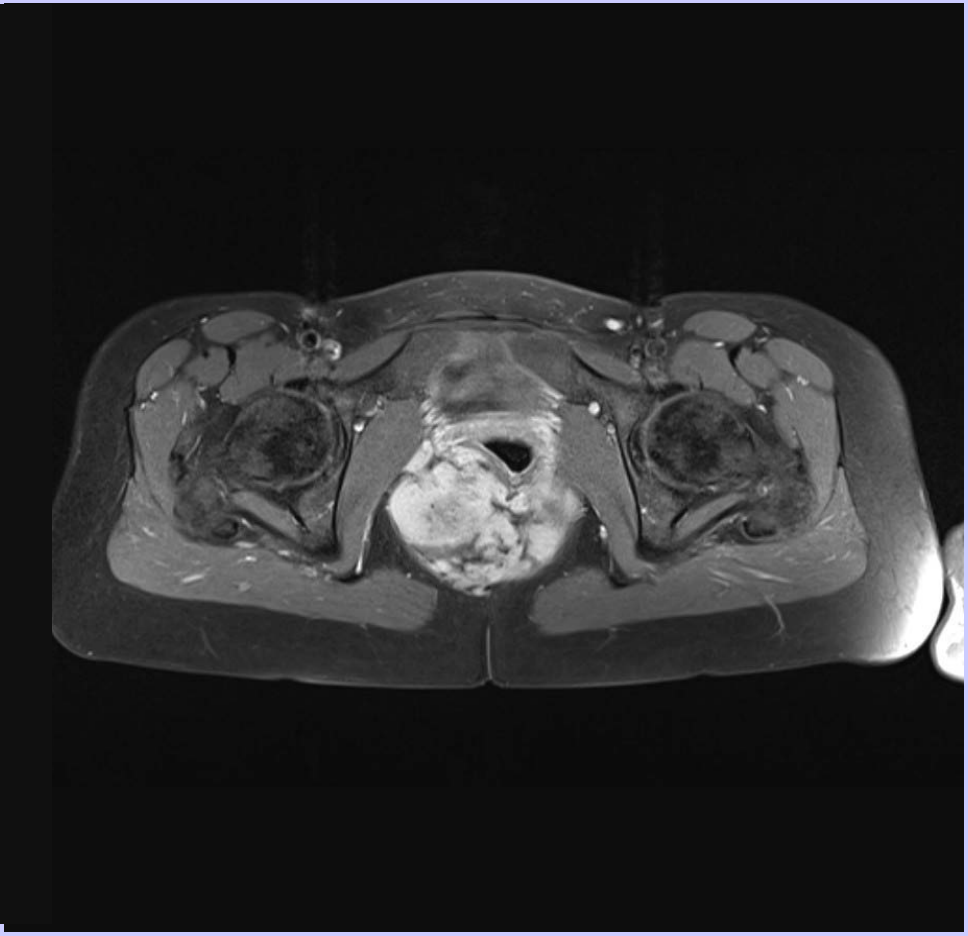

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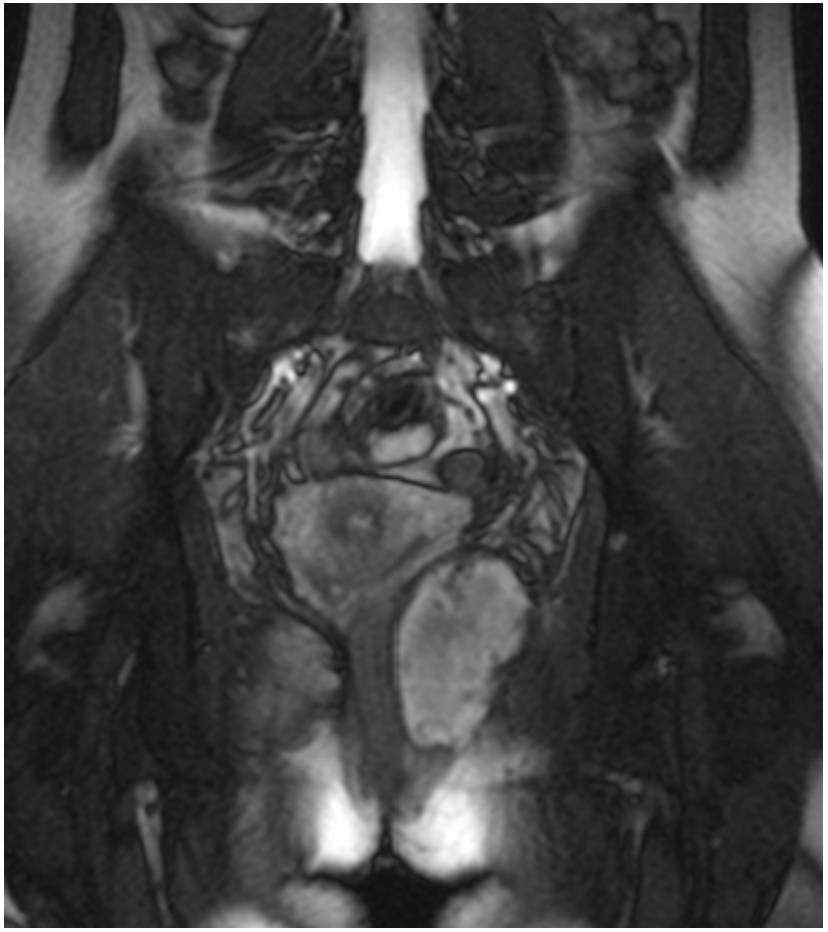



OOB:18.01.1964
AcqDt:04.07.2011
AcqTm:14:25:32
nNo:7
1.279
1
2
2 Länge : 3.34 cm
2 Min/Max : 77/184
2 Pixelzahl : 32
SO:IR/SAT2
Type:ORIGINAL/PRIMARY/M/NORM/...
E:29.000
R:8021.000

Analysis of treatment situation

- Most often patients start at their general practitioner
- Growing lump is excised, then desmoid is detected
- Large (sarcoma-like) tumors or those located in the rectus abdominis muscle show up at centers more often
- After receiving the diagnosis > internet

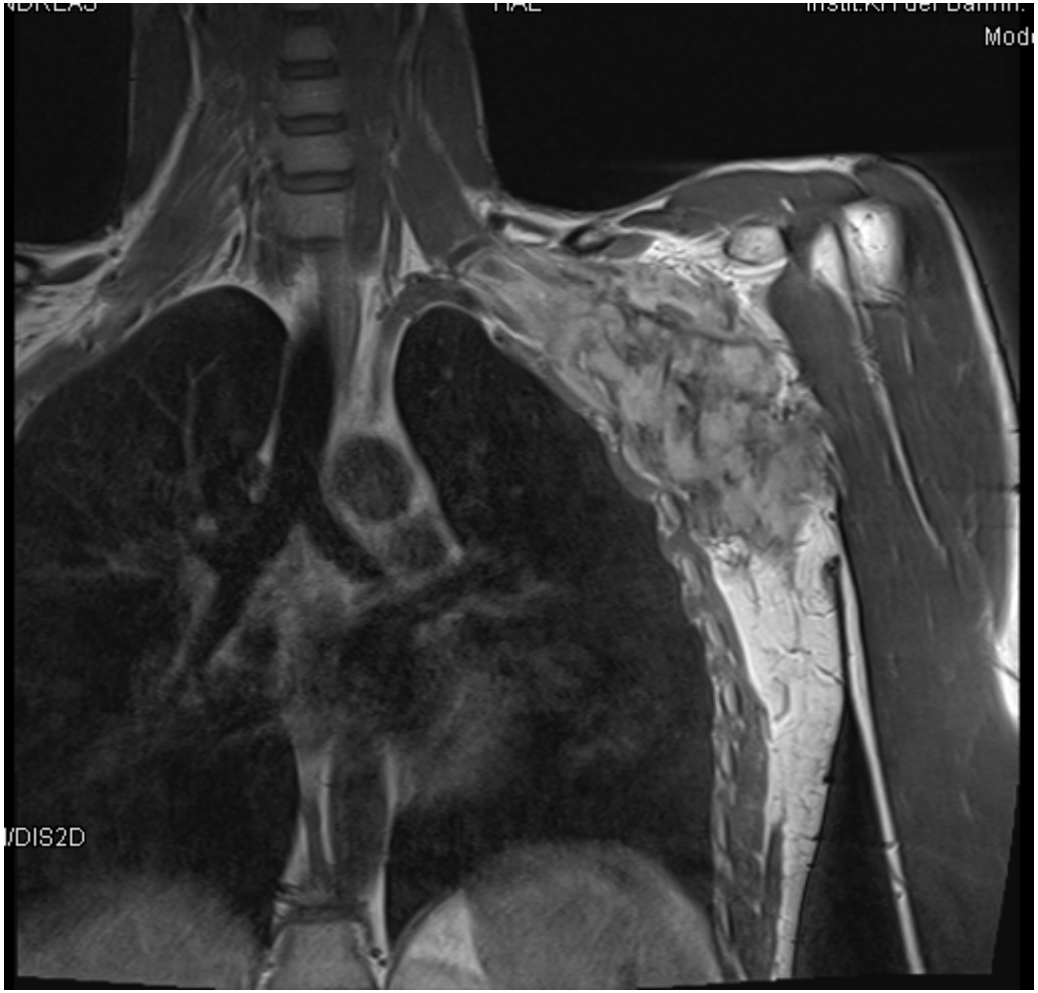
Analysis of treatment situation in Germany

- 237 patients (160 females, 76 males, ratio 2.11)
- Median age: 37.4 years
- Delay until first doctor consultation: 77 days
- Remember trauma at desmoid site: 56%
- Primary diagnosis desmoid suspected: 24%
- Histologically correct diagnosis after 1st operation: 88%

Analysis of treatment situation in Germany

- Primary therapy : 157 patients
- Surgery 82%
- 1.7 operations/patient
- R0 resection rate : 29.5%
- R1 resections : 48%
- R2 resections : 13.5%
- Recurrence rate: 63%
- Radiation therapy: 42%

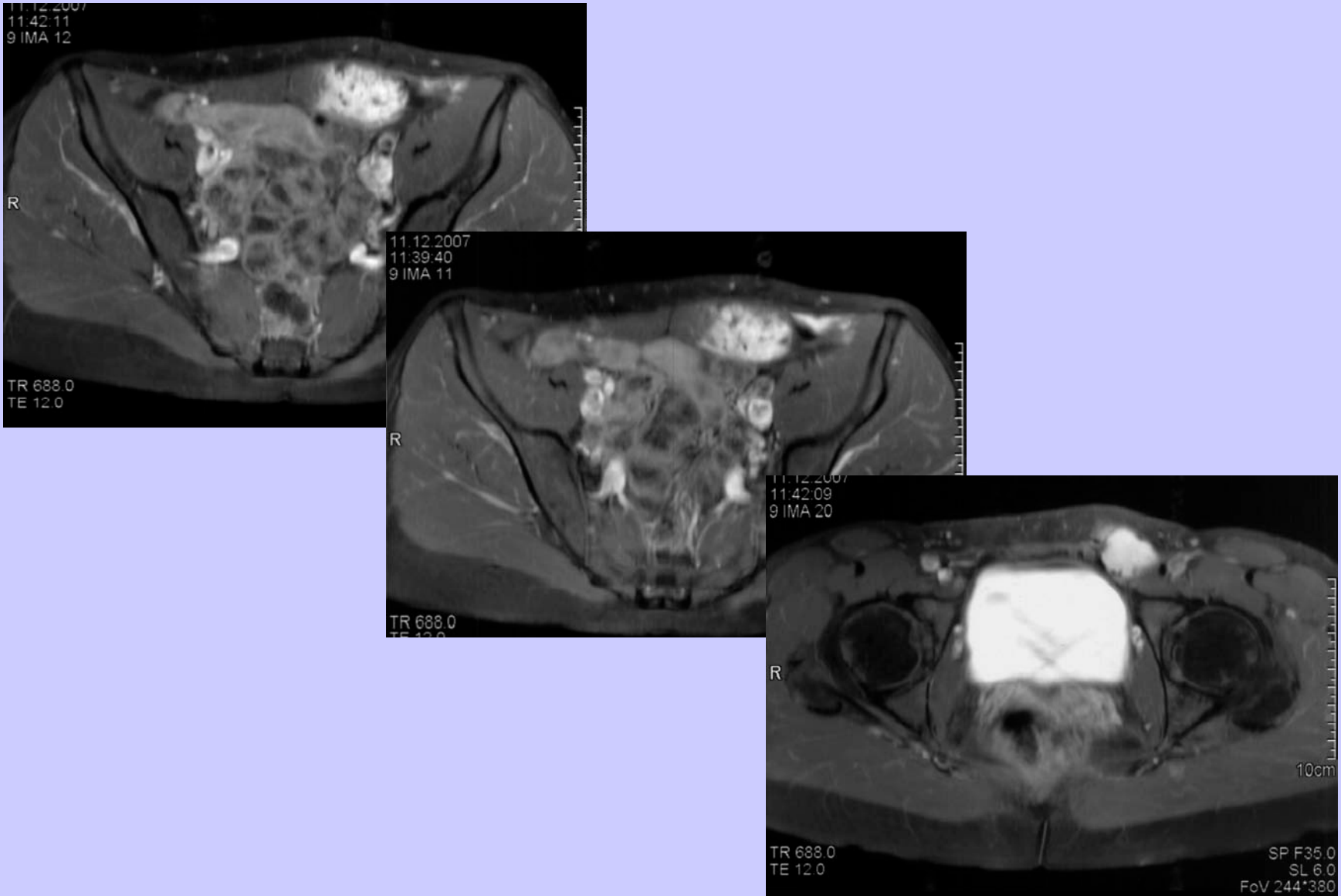
Sometimes : Does operation make sense ?



Analysis of treatment situation

- 2.4 to 4.3 new cases per 10^6 inhabitants/year
- Star shape tumors, devoid of a capsule, invade surrounding structures
- Wide range of local failure rates >> great variability of accrual, treatments and follow-up
- Impact of surgical margins after excision remains unclear
- Biology of the different desmoid types not well characterized
- Discrepancies between the results of the impact of the quality of surgery

Rectus abdomins : resectable



Standard chest wall replacements

Goretex membrane

Vicryl - mesh

Marlex-mesh (Prolene)

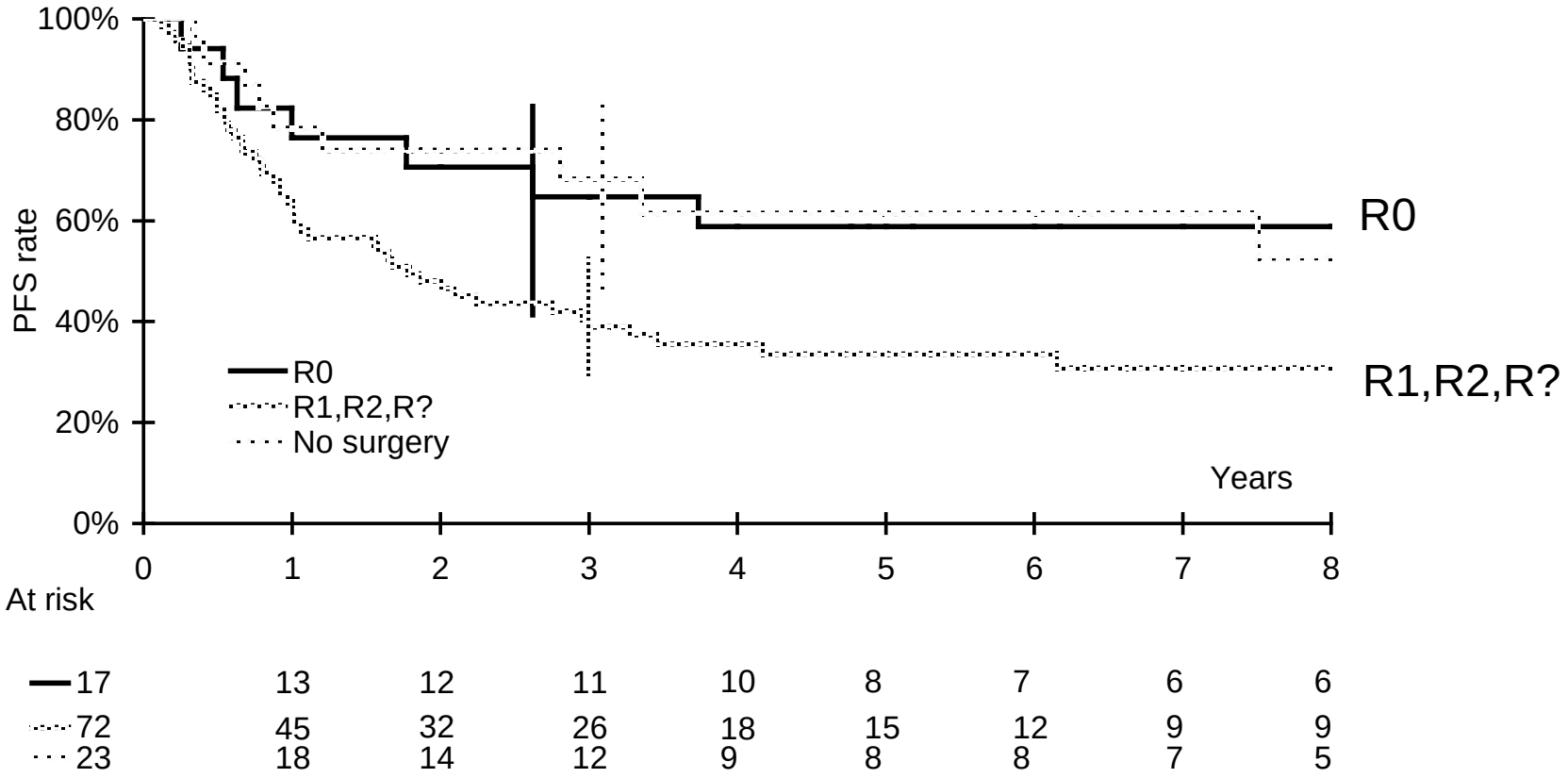
(Lyodura)

Omentum + Meshgraft

Chestwall & abdominal wall reconstruction Principles

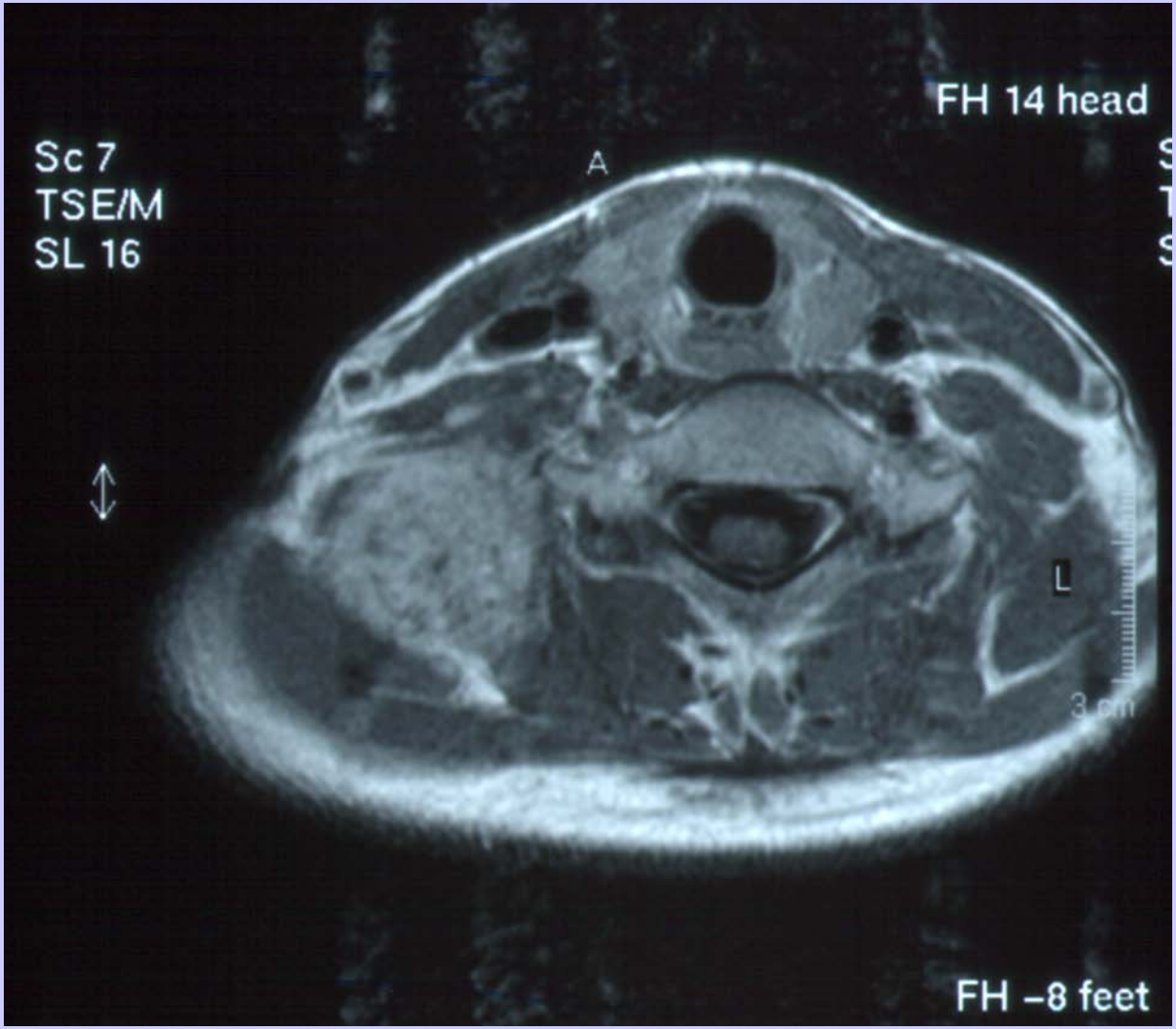
- Function > aesthetics
- Wound closure mandatory
- Avoid fluid accumulation
- Wound infection detrimental if allografts implanted

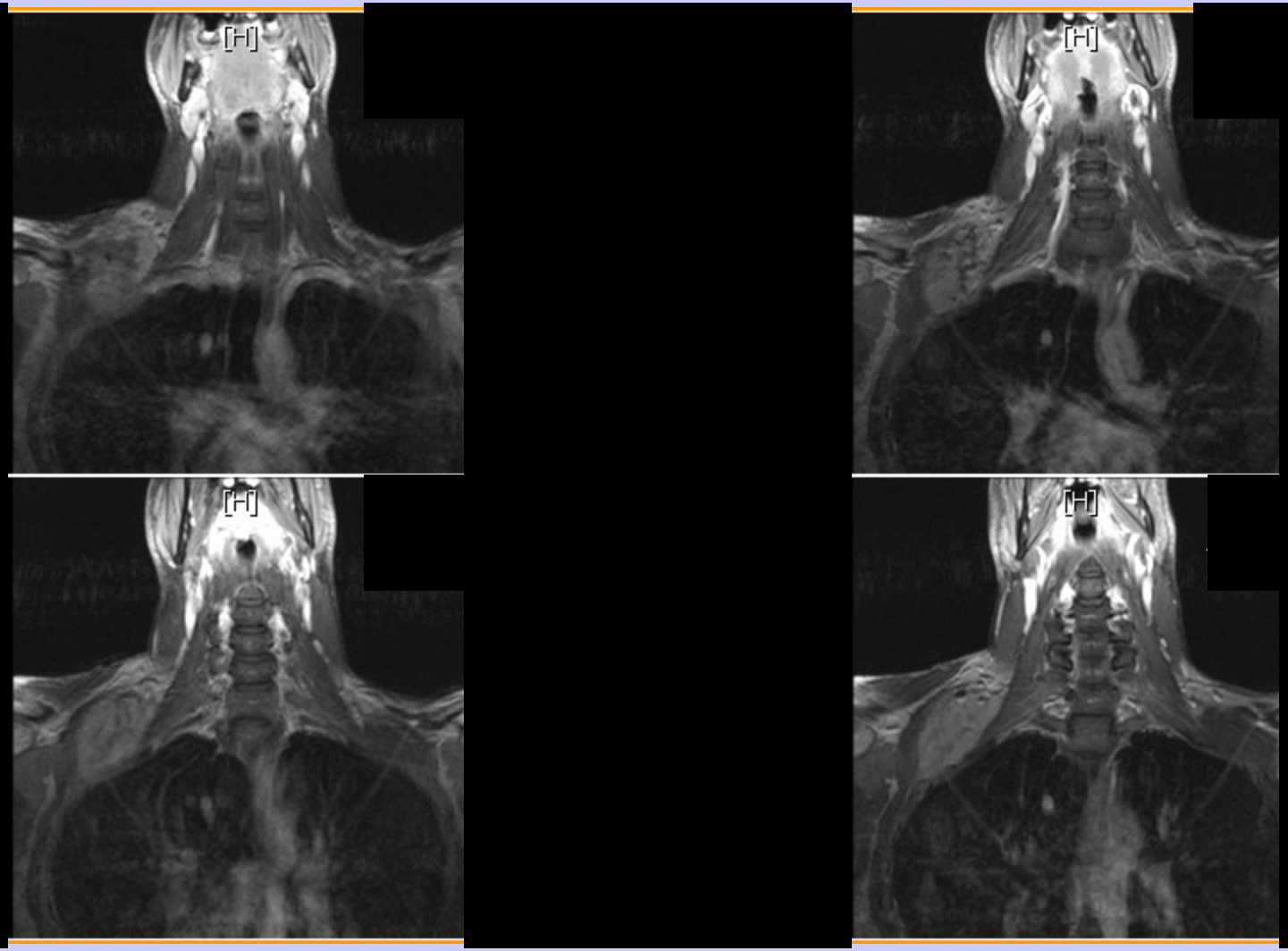
Disease-free survival according to the quality of surgery



Bonvalot et al. EJSO 2008

Neck region : not R0 resectable

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Radiation therapy postoperatively mandatory !

Authors	Institution/ Town	Period	N patients P/R	Local recurrences Progressions	Impact margins
Philipps (BJS 2004)	Royal Marsden	86-03	109 P+R	20% 3 years	No
Posner (Arch surg 89)	MSKCC	65-84	138 P+R	20% 2 years	Yes
Merchant (Cancer 99)	MSKCC	82-97	189 P+R	20% 2 years	No
Sorensen (acta Orth Scand 2002)	Arhus	70-98	72 P	27% 5 years	Yes
Pignatti (Clin Orth Res 2000)	Bologne	70-96	83 P+R	44% 2 years	Yes
Spear (I J R O B P 1998)	Harvard	71-92	107 P	S RT S+RT 30 7 22% 5 years	Yes
Goy (I J R O B P 97)	UCLA	65- 92	61 P	R0 R1 S+RT 15 70 20% 6 years	Yes
Gronchi (JCO 2003)	Milan	65-00	203 P+R	Primary: 25% 10 years	No
Lev (JCO 2007)	MDA	95-05	189 P+R	20% 5 years	Yes
Bonvalot (EJSO 2008)	IGR	88-03	112 P	R0 35% 3 years No surgery/no RT 30% 3 years	Yes

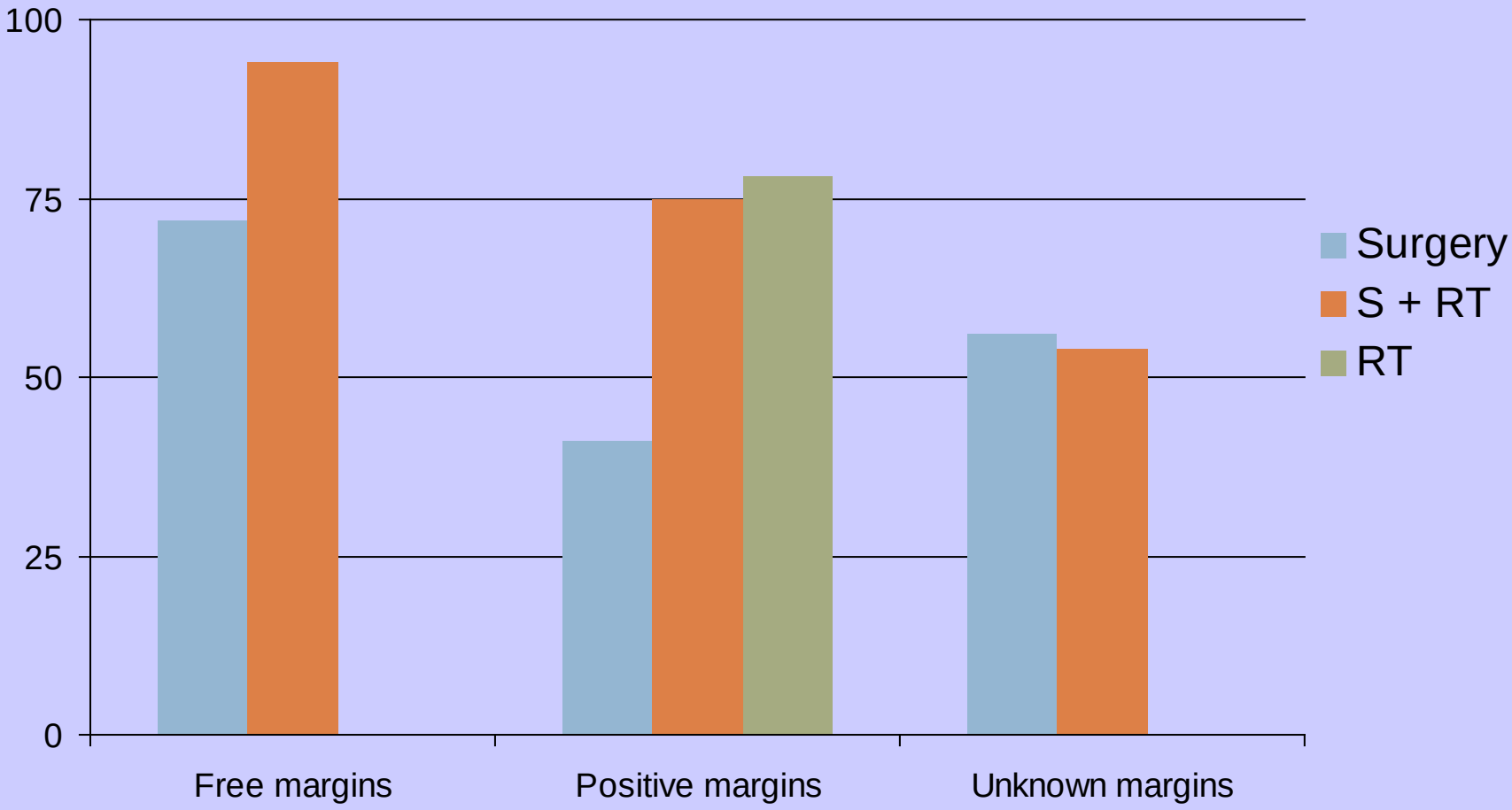
Surgery versus radiation therapy for patients with aggressive fibromatosis or desmoid tumors: Review of 22 articles

1983 1998	Surgery alone			Surgery + RT			RT
	Margins			Margins			
	-	+	overall	-	+	overall	
Local Control rate	72%	41%	61%	94%	75%	75%	78%

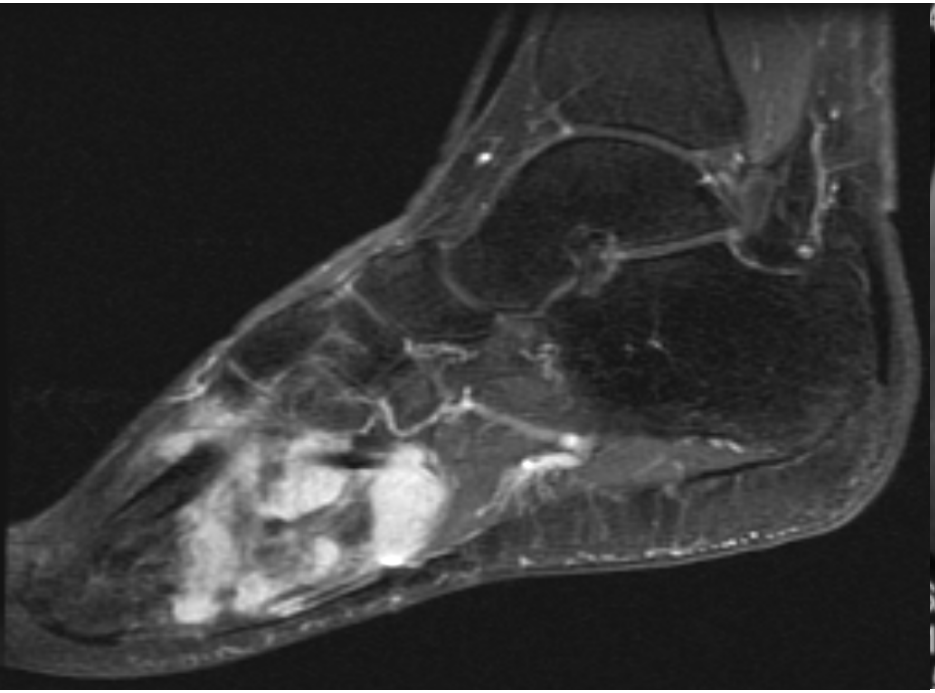
- RT or S + RT results in significantly better local control than S
- Even after dividing the groups into cases with +/-margins and primary and recurrent T: the best local control is achieved with RT or S + RT

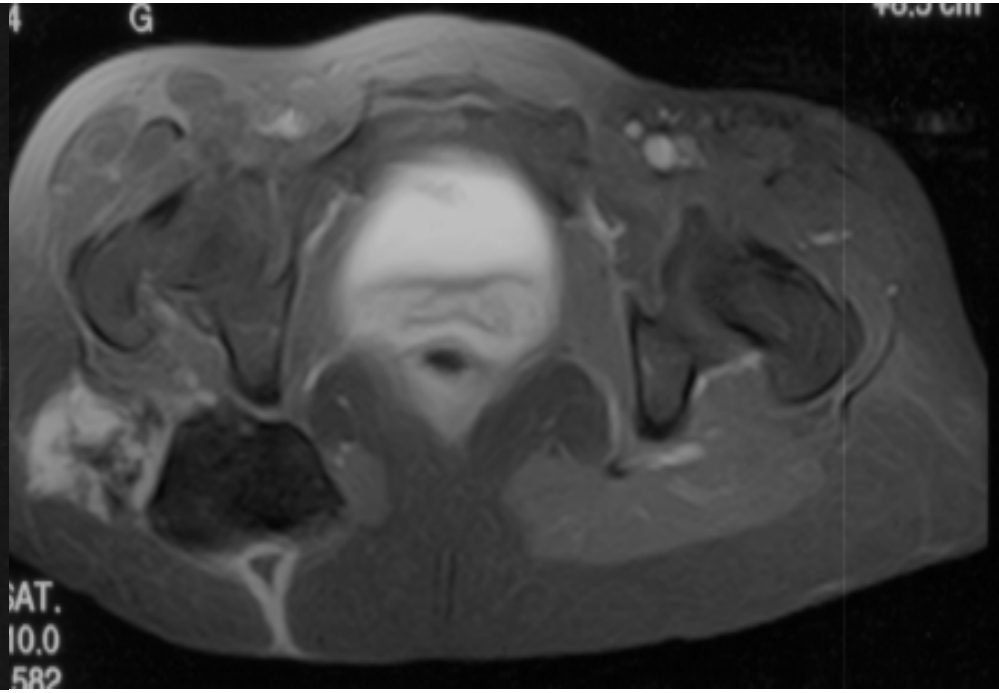
Nuyttens JJ et al Cancer. 2000

LOCAL CONTROL
Fibromatosis meta-analysis



R0 surgery hardly feasible without major morbidity
Does it really need therapy ?





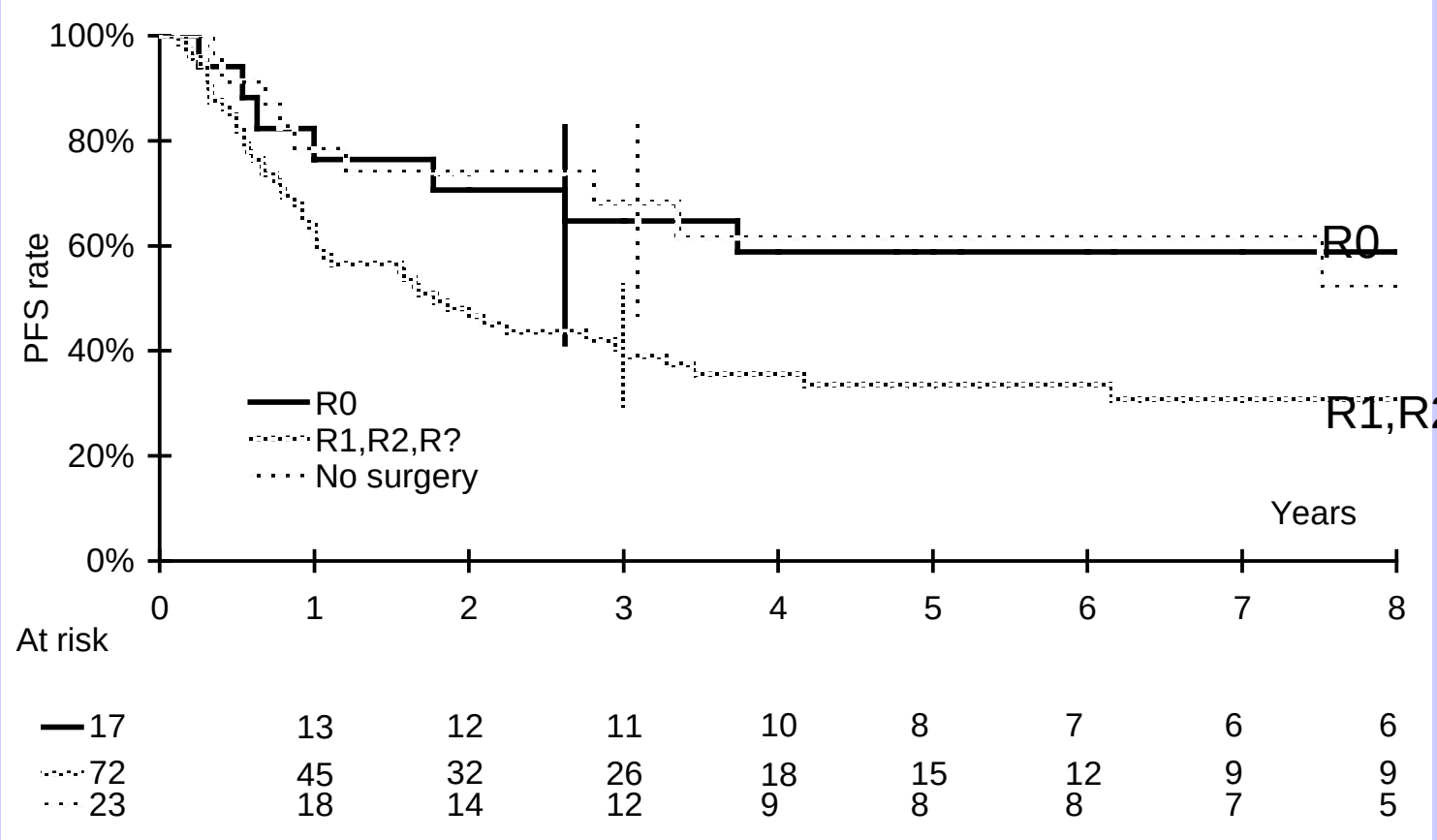
Recurrent fibromatosis after surgery
No change 5 years later
If progressive : isolated limb perfusion

Primary fibromatosis (surgical biopsy)
No change 6 years later

A possible role for isolated limb perfusion with tumor necrosis factor-alpha and melphalan

	N patients	RR	CR	PR	Local progression
Lev- chelouche (Surgery 1999C	6	83%	33%	50%	2 (Follow up 45 months)
Grunhagen (EJSO 2005)	12	75%	17%	58%	?

Disease-free survival according to the quality of surgery



Bonvalot et al. EJSO 2008

Annals of Oncology 19 (Supplement 2): ii89–ii93, 2008
doi:10.1093/annonc/mdn101

Soft tissue sarcomas: ESMO Clinical Recommendations for diagnosis, treatment and follow-up

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On behalf of the ESMO Guidelines Working Group*

[1]*Department of Cancer Medicine, Istituto Nazionale dei Tumori, Milan, Italy;* [2]*Department of Oncology, Kantonsspital, Bruderholz, Switzerland;* [3,4]*Department of Medical Oncology, Erasmus University Medical Center, Rotterdam, The Netherlands;* [5]*INSERM U590, Claude Bernard University, Lyon, France*

desmoid-type aggressive fibromatosis

Standard treatment for primary disease, if amenable to surgery without significant functional losses, is wide excision [IV, B]. In those cases in which only marginal excision can be performed, postoperative radiation therapy is an option, after sharing the decision with the patient in conditions of uncertainty, considering the possible occurrence of radiation-related high-grade sarcomas in a non-metastasizing disease. Observation is another option in selected cases, after shared decision-making with the patient, taking into account the indolent natural history of some clinical presentations.

For primary disease only amenable to surgery with significant functional losses, wide excision is an option, along with radiation therapy, observation, isolated limb perfusion (if the lesion is confined to an extremity) or systemic therapy (see below) [V, D]. The same applies to recurrent disease.

For the inoperable disease, radiation therapy, ILP (if the lesion is confined to an extremity), and systemic therapies are options, along with observation [V, D]. Systemic therapies include: hormonal therapies (tamoxifen, toremifene, Gn-RH analogs) ± NSAIDs; low-dose chemotherapy, such as methotrexate + vinblastine or methotrexate + vinorelbine; low-dose interferon; imatinib; full-dose chemotherapy (using regimens active in sarcomas). It is reasonable to employ stepwise the less toxic therapies before the more toxic.

Soft Tissue Sarcoma

Desmoid Tumors

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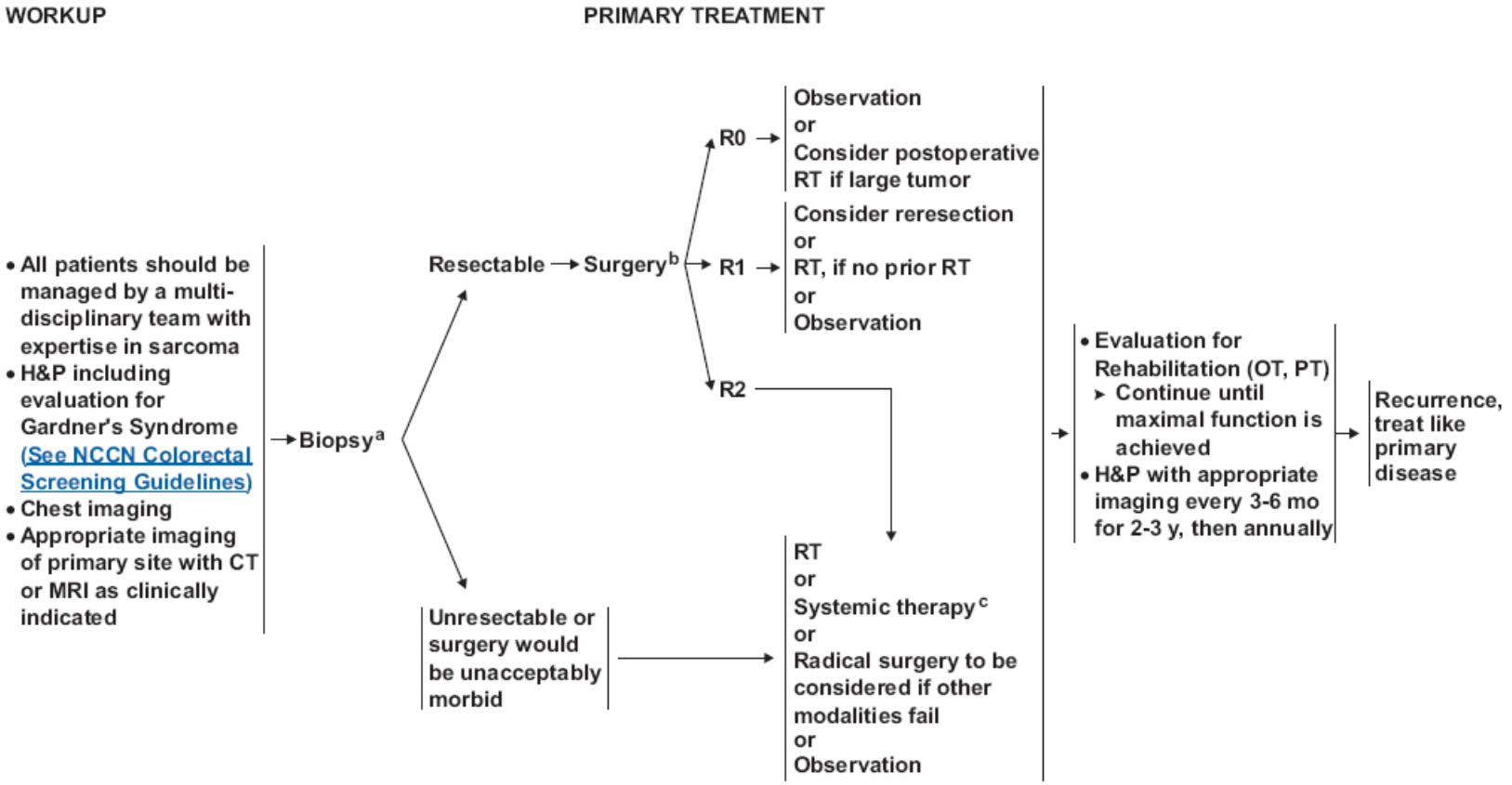
Staging, MS, References



[a] May not be necessary if complete resection planned.
[b] For desmoids, microscopic positive margins are acceptable if achieving negative margins would produce excessive morbidity.
[c] See Principles of Systemic Therapy (SARC-C).

Note: All recommendations are category 2A unless otherwise indicated.
Clinical Trials: NCCN believes that the best management of any cancer patient is in a clinical trial. Participation in clinical trials is especially encouraged.

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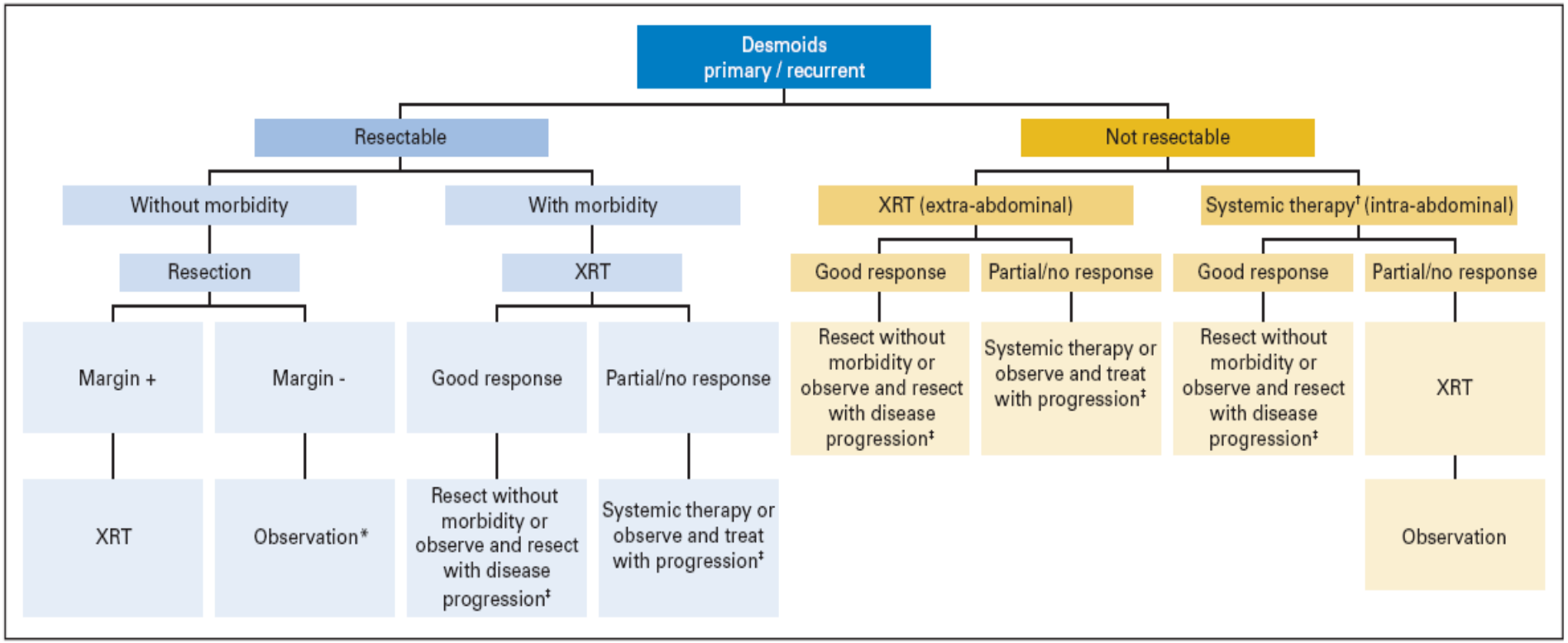
ORIGINAL REPORT

Optimizing Treatment of Desmoid Tumors

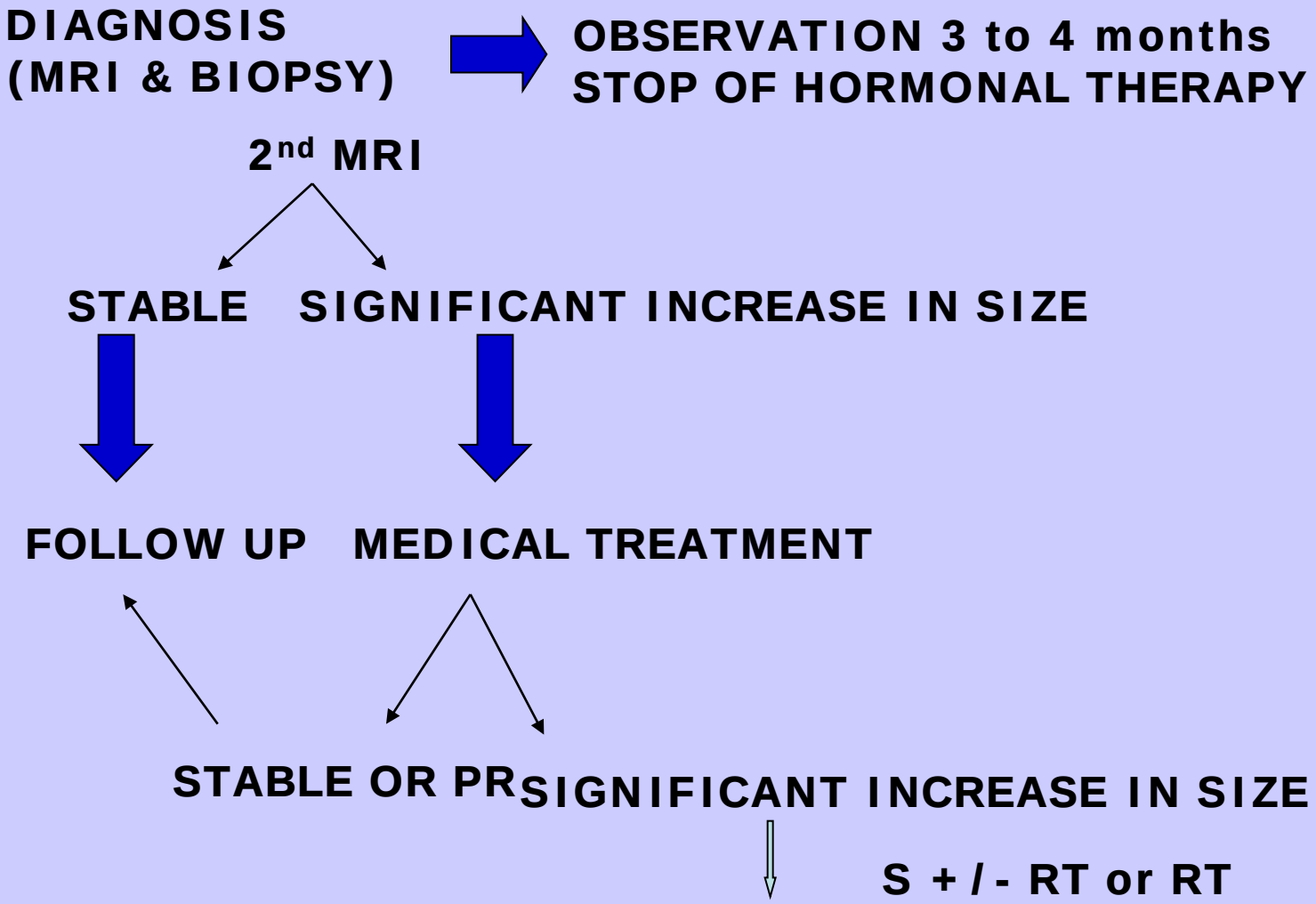
Dina Lev, Dhanasekaran Kotilingam, Caimiao Wei, Matthew T. Ballo, Gunar K. Zagars, Peter W.T. Pisters, Alexander A. Lazar, Shreyaskumar R. Patel, Robert S. Benjamin, and Raphael E. Pollock



Treatment recommendations

- Based on the risk/benefit analysis for the patient
- Dependent on tumor and patient characteristics, location, and evolution
- Observation alone could be considered for primary tumors
  - if the diagnosis is confirmed (biopsy, beta-catenin)
  - located such that progression would not cause significant morbidity

Handling of non-R0 resectable disease



Role of surgery

- If surgery, then look for R0 resection
- Whether surgery at all –
  - progressing lesion
  - can be treated with acceptable morbidity
  - patient wish
- If surgery is not expected to be R0, consider radiation therapy
- IORT
- Of course, postoperative RT if a recurrence would allow re-resection with acceptable morbidity

Recommendations

- Asymptomatic patients with extra-abdominal desmoid may be observed or treated with low morbidity therapy.
- Symptomatic **abdominal** desmoids should be considered for chemotherapy.
- Formal prospective trials may help refine recommendations.