The clinical history of STS & GIST

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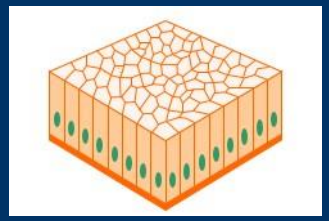
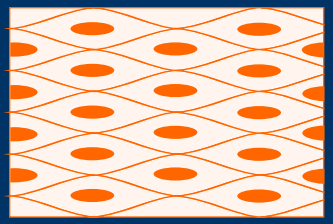
Potential conflicts of interest

	Empl	Cons	Stocks	Honor	Res (inst.)	Test	Other
Amgen Dompé		●			●		
ARIAD		●					
Bayer		●			●		
Blueprint Medicines		●					
Eisai					●		
Glaxo SK		●			●		
Lilly		●					
Merck SD		●					
Merck Serono		●					
Molmed					●		
Novartis		●		●	●		●
Pfizer		●		●	●		
PharmaMar		●		●	●		●

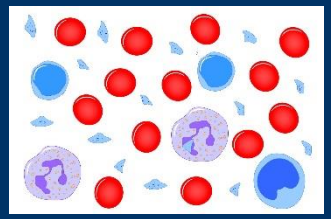


epithelial t.

connective t.

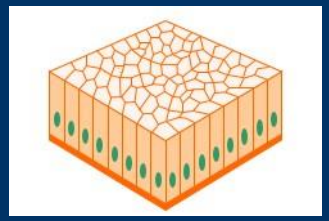


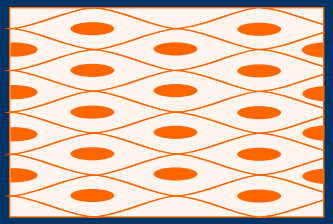
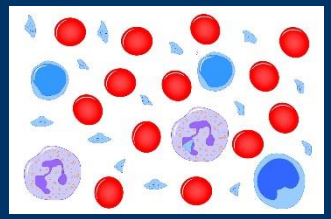
muscle



blood

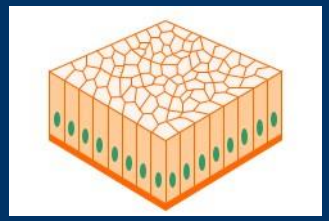
nervous t.

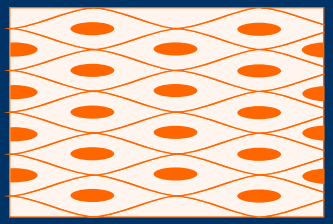


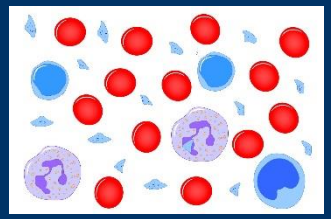




epithelial t. | connective t. | muscle | blood | nervous t.

carcinomas | sarcomas | leukemia, lymphoma, myeloma, ... | glioma, ...

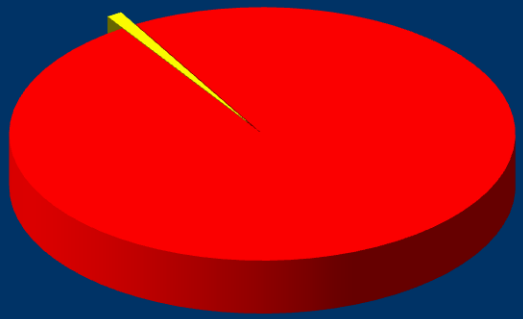






epithelial t. | connective t. | muscle | blood | nervous t.

carcinomas | sarcomas | leukemia, lymphoma, myeloma, ... | glioma, ...



incidence

benign t.	**300**	**/100.000/year**

incidence	soft tissue sa	5	/100,000/year
	benign t.	300	/100,000/year

incidence	soft tissue sa	5	/100,000/year
	benign t.	300	/100,000/year
sex		M ≥ F	
median age	soft tissue sa	60 yrs	

incidence	**soft tissue sa**	**5**	**/100,000/year**
	GIST	**1.5**	**/100,000/year**
	benign t.	**300**	**/100,000/year**
sex		**M ≥ F**	
median age	**soft tissue sa**	**60 yrs**	
	GIST	**60 yrs**	

incidence	**soft tissue sa**	**5**	**/100,000/year**
	GIST	**1.5**	**/100,000/year**
	osteosa	**0.3**	**/100,000/year**
	Ewing	**0.2**	**/100,000/year**
	rhbdomyosa	**0.1**	**/100,000/year**
	benign t.	**300**	**/100,000/year**
sex		**M ≥ F**	
median age	**soft tissue sa**	**60 yrs**	
	GIST	**60 yrs**	
	osteosa	**15 yrs**	
	Ewing	**15 yrs**	
	rhabdomyosa	**5 yrs**	

Study Quantifies Cancer Risk of Morcellation

CME & CE

Published: Jul 23, 2014 | Updated: Jul 24, 2014

By Charles Bankhead, Staff Writer, MedPage Today
Reviewed by Robert Jasmer, MD; Associate Clinical Professor of Medicine, University of California, San Francisco and Dorothy Caputo, MA, BSN, RN, Nurse Planner

save | A A

One of every 368 women treated with a power morcellator had unsuspected uterine cancer identified during or after their procedures, a review of more than 200,000 patients showed.

Medical records showed that morcellation, or the fragmentation of the uterus into smaller pieces, was performed in 36,470 cases and 99 of the women subsequently had uterine cancer diagnoses. In addition, 26 other gynecologic malignancies were identified, along with 39 uterine lesions of uncertain malignant potential and 368 cases of endometrial hyperplasia.

A review of potentially predictive factors showed that older age was the only factor associated with underlying uterine malignancy or endometrial hyperplasia, as reported in a research letter published

For best viewing, click the bottom right corner for full screen.

Action Points

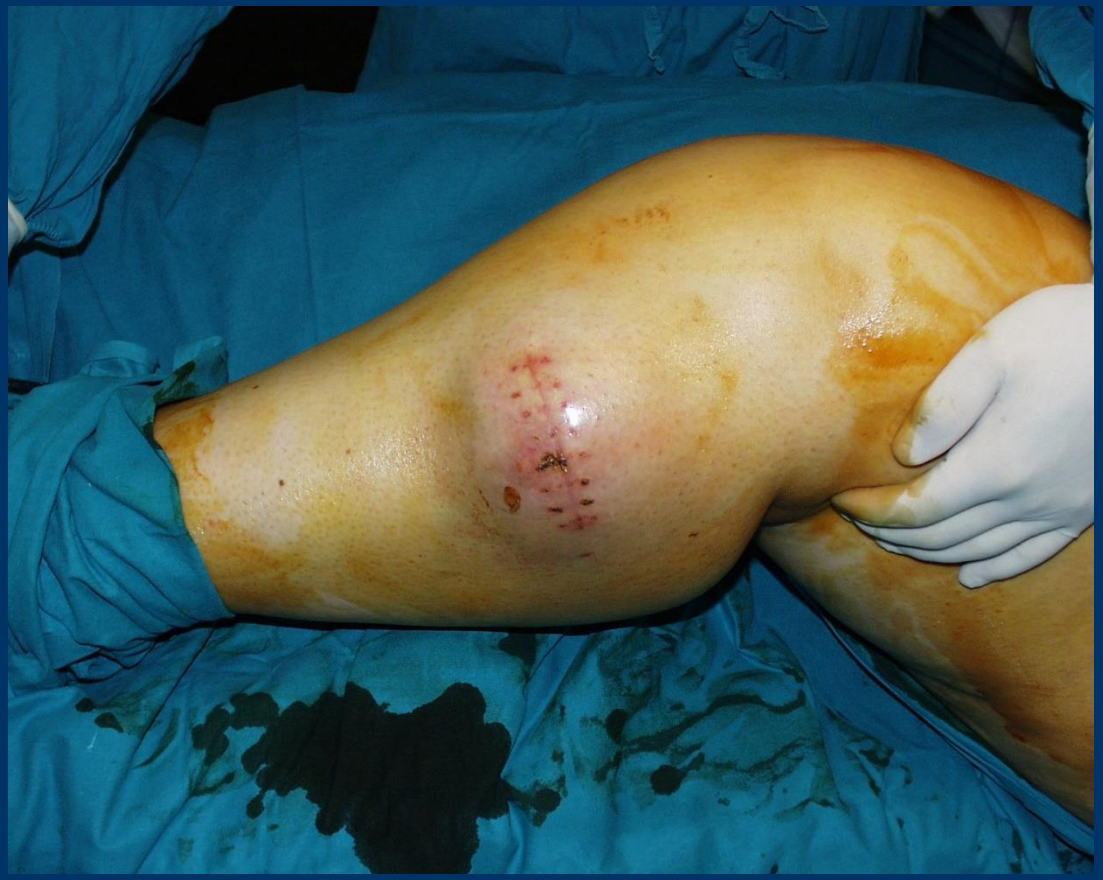
- One of every 368 women treated with a power morcellator -- a device that fragments the uterus into smaller pieces -- had unsuspected uterine cancer identified during or after their procedures.

FDA NEWS RELEASE

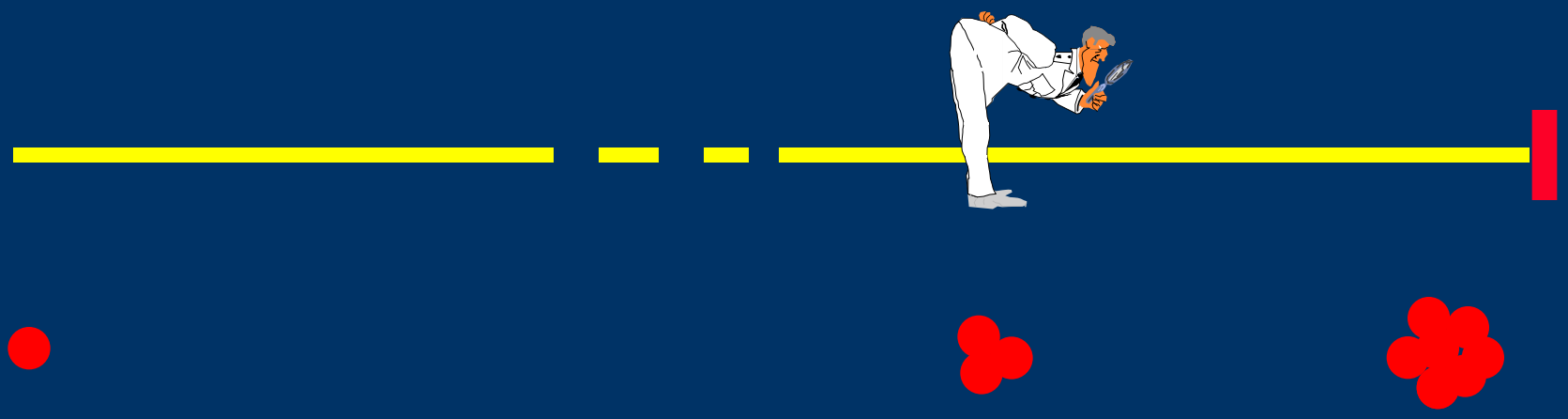
For Immediate Release: April 17, 2014
Media Inquiries: Jennifer Rodriguez, 301-796-8232, jennifer.rodriguez@fda.hhs.gov
Consumer Inquiries: 888-INFO-FDA, dice@fda.hhs.gov

FDA discourages use of laparoscopic power morcellation for removal of uterus or uterine fibroids
Procedure poses risk of spreading undetected cancerous tissue in women with unsuspected cancer

In a safety communication notice issued today, the U.S. Food and Drug Administration discouraged the use of laparoscopic power morcellation for the removal of the uterus (hysterectomy) or uterine fibroids (myomectomy) in women because, based on an analysis of currently available data, it poses a risk of spreading unsuspected cancerous tissue, notably uterine sarcomas, beyond the uterus.

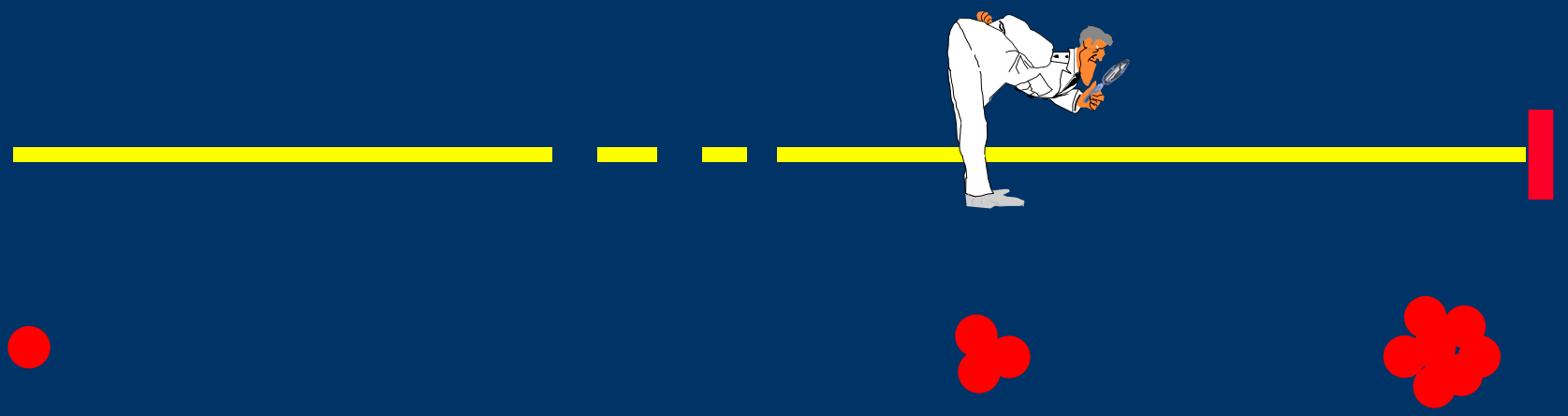


Diagnosis



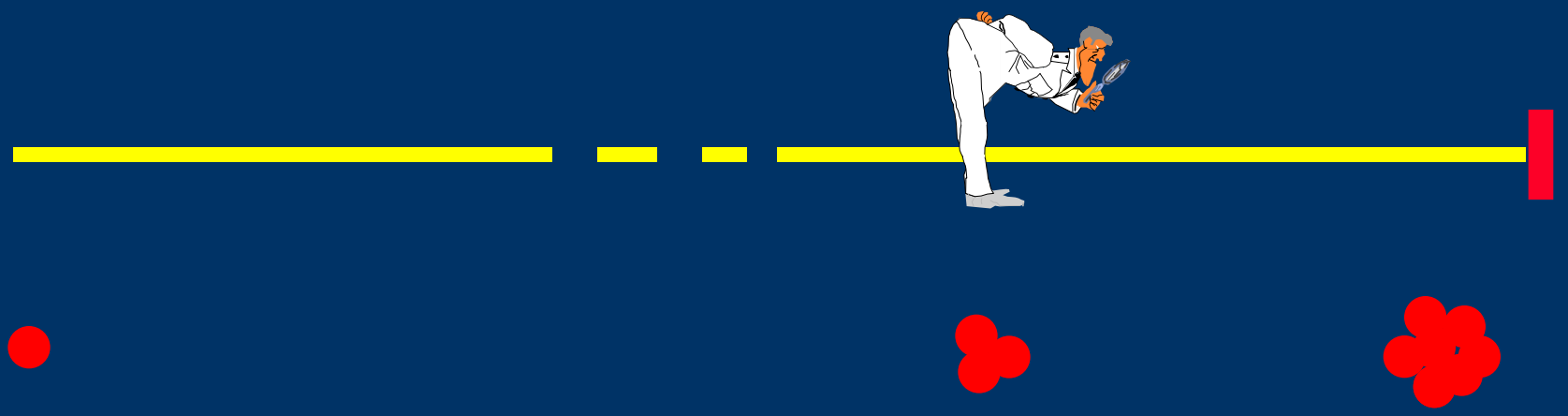
Phase: **clinical**

Timely diagnosis



Phase: **clinical**

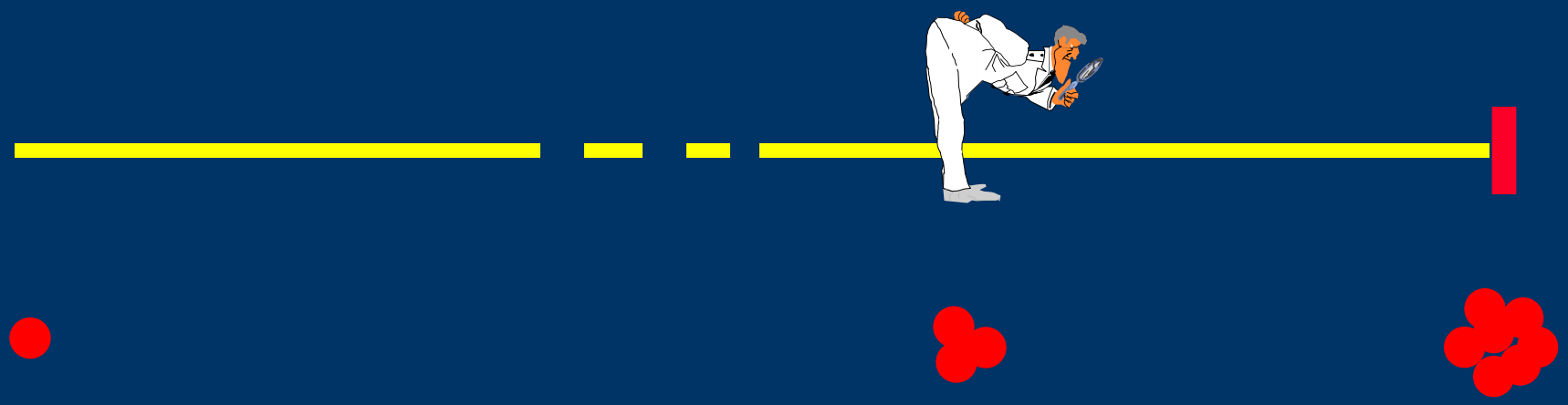
Late diagnosis



Phase: clinical

diagnostic delay

Early diagnosis



Phase: **preclinical** **clinical**
detectable


Is it
SARCOMA
?

https://sarcoma.org.uk/

New Medical Terms

Whoops! procedure

SURGERY

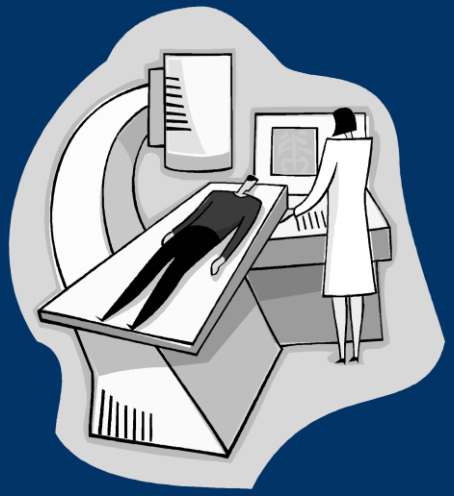
A UK term of art for a surgical procedure in which an inexperienced surgeon (or one inexperienced in the type of procedure on which he* is so boldly embarking) **realises, after getting elbows deep in muck, that he's in tiger country**, guileless, guideless and gunless.

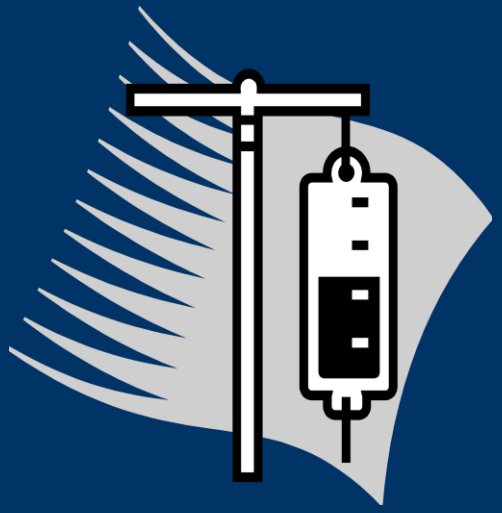
*I would also say she, but I'm told women don't make mistakes…ever. The classic whoops surgery is that in which the operator expects–and plans for that and no more–a banal lesion that he can shell out or excise with a minimal rim of normal tissue and he finds a soft tissue sarcoma in which his impetuosity ("whoops") just screwed up the margins, making complete excision by a specialised sarcoma surgeon more difficult, while worsening the prognosis.

Synonym ***Oh my God!*** procedure

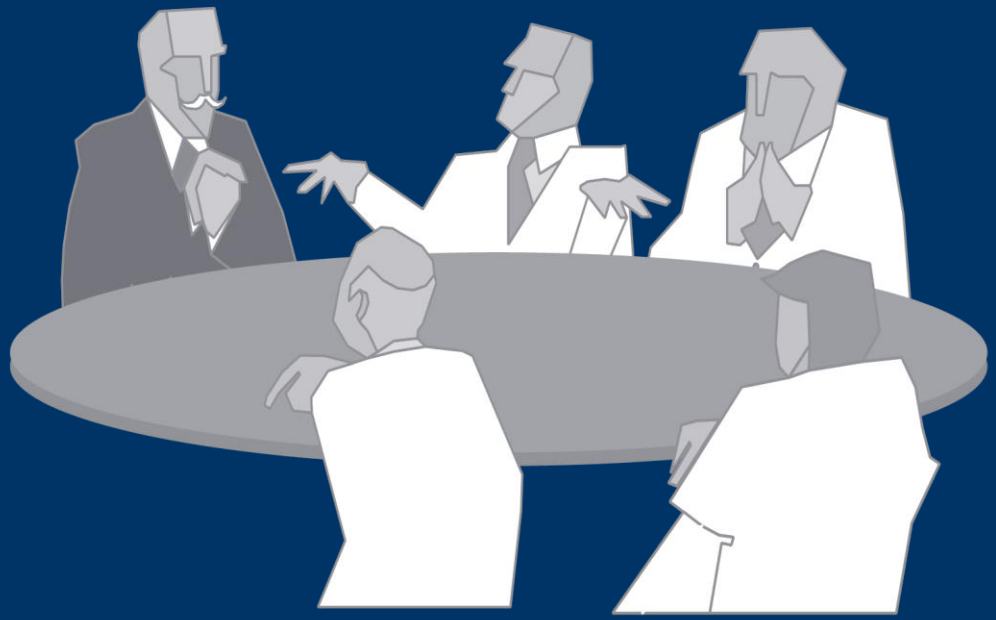
Reference Professor Neil Shepherd, 25.01.2011

Multidisciplinarity



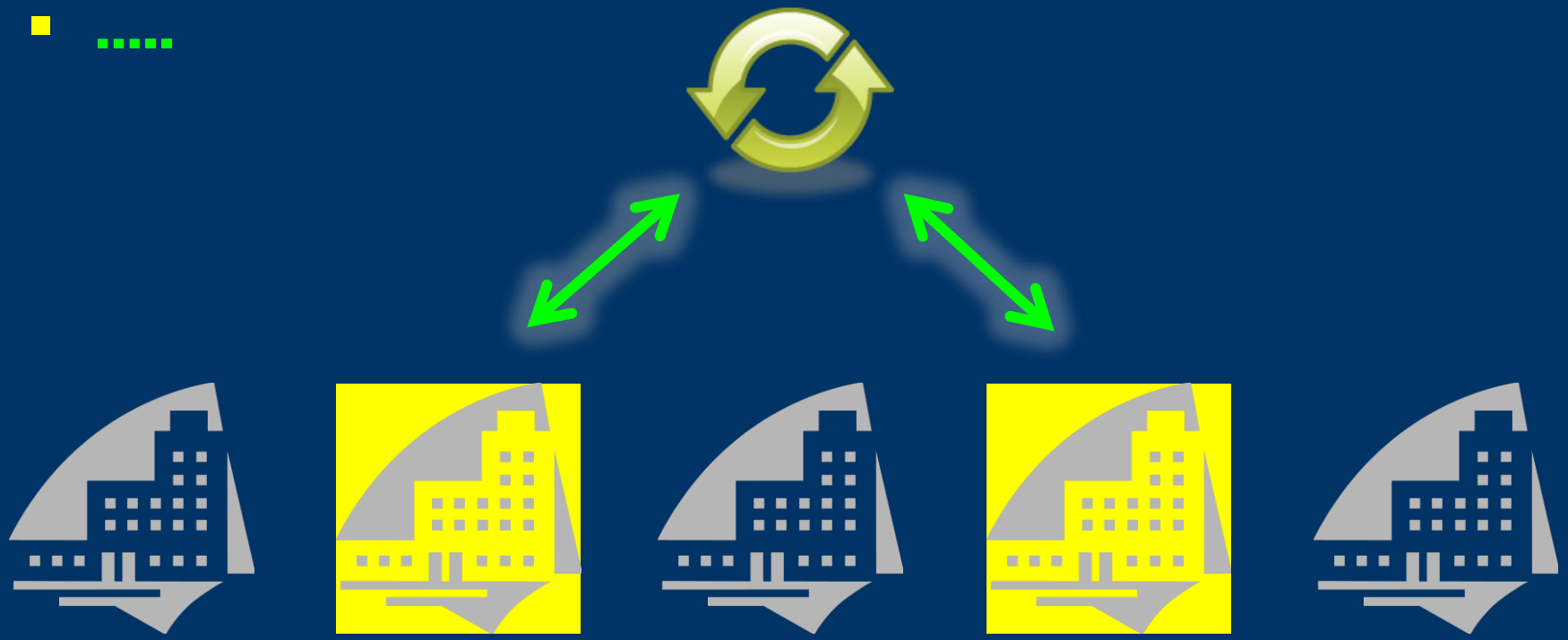


Multidisciplinarity

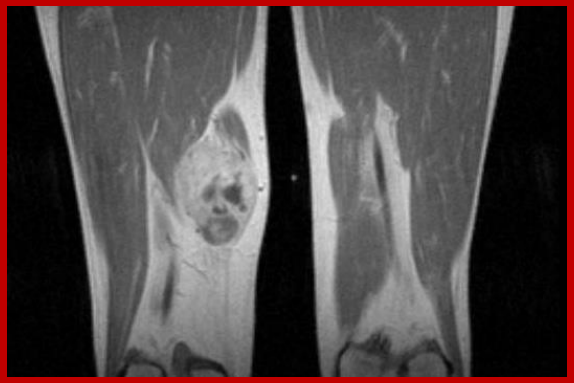


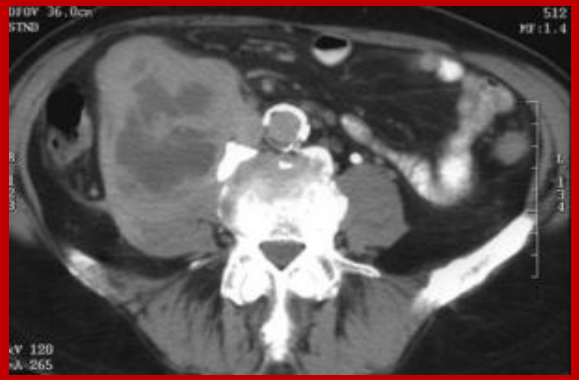
Referral & Networking…

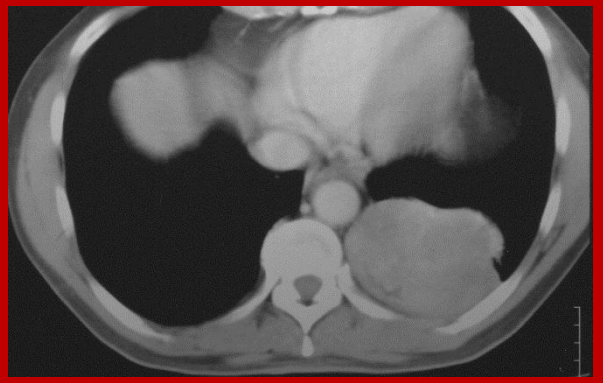
- pathologic diagnosis
- strategic clinical decision-making
- local treatment
- …..

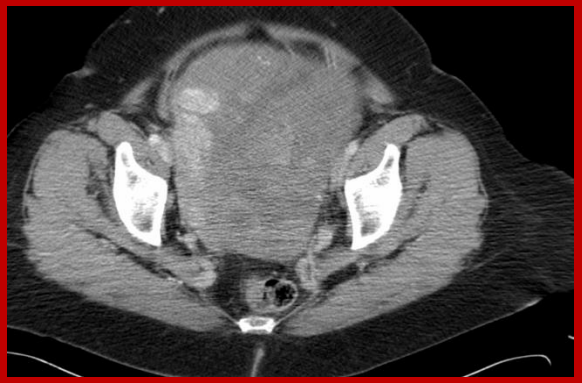


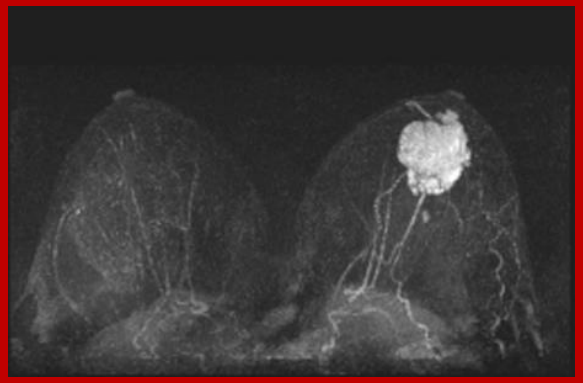
Adult soft tissue sarcomas

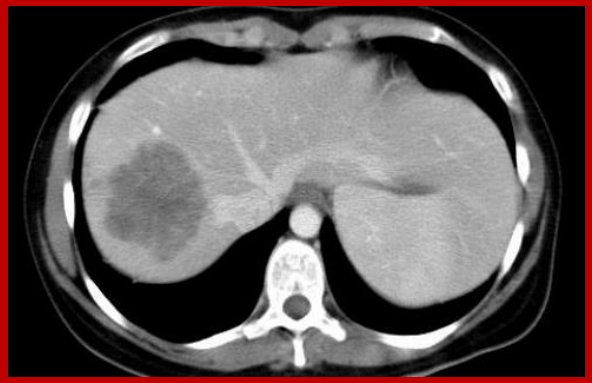


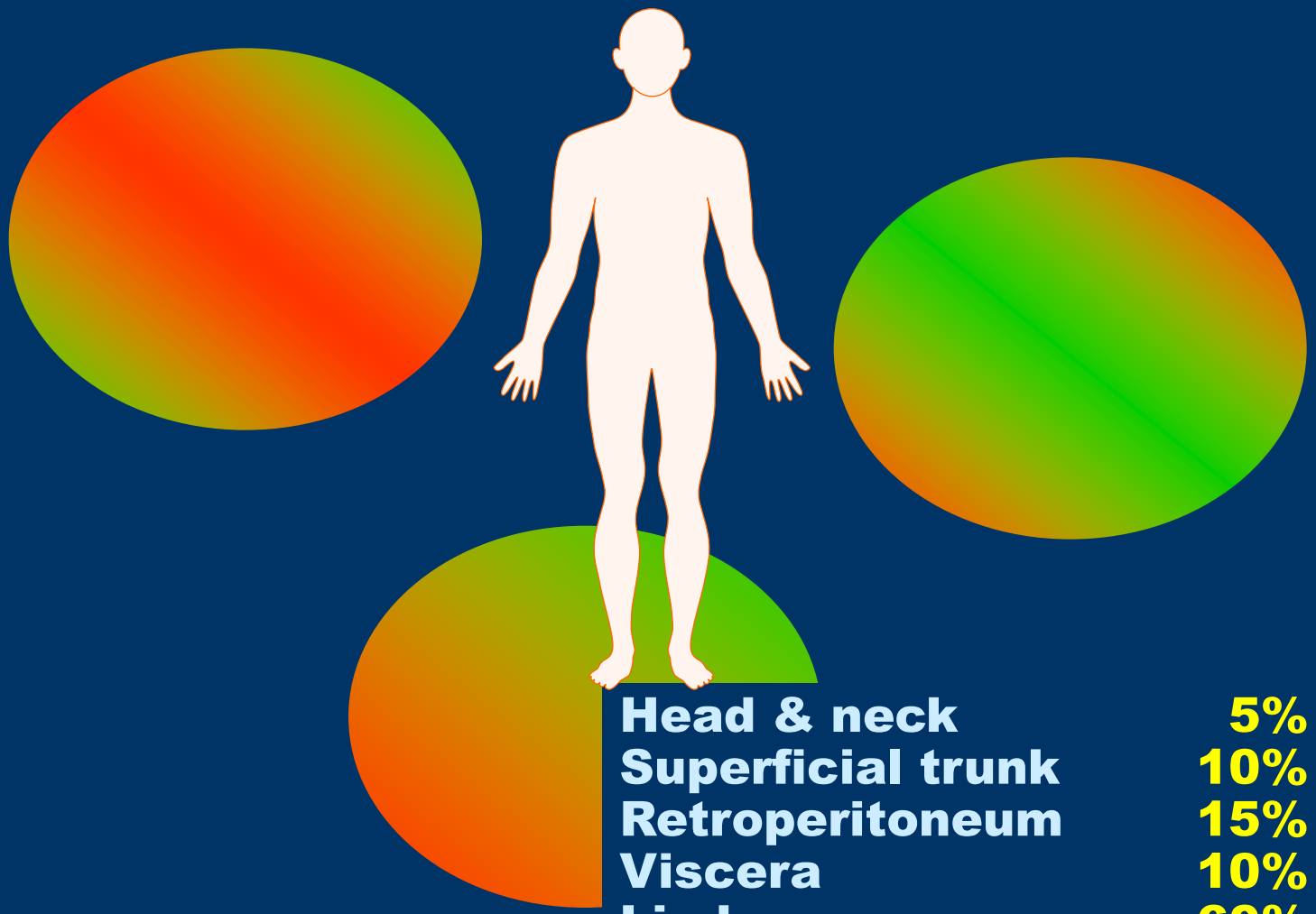




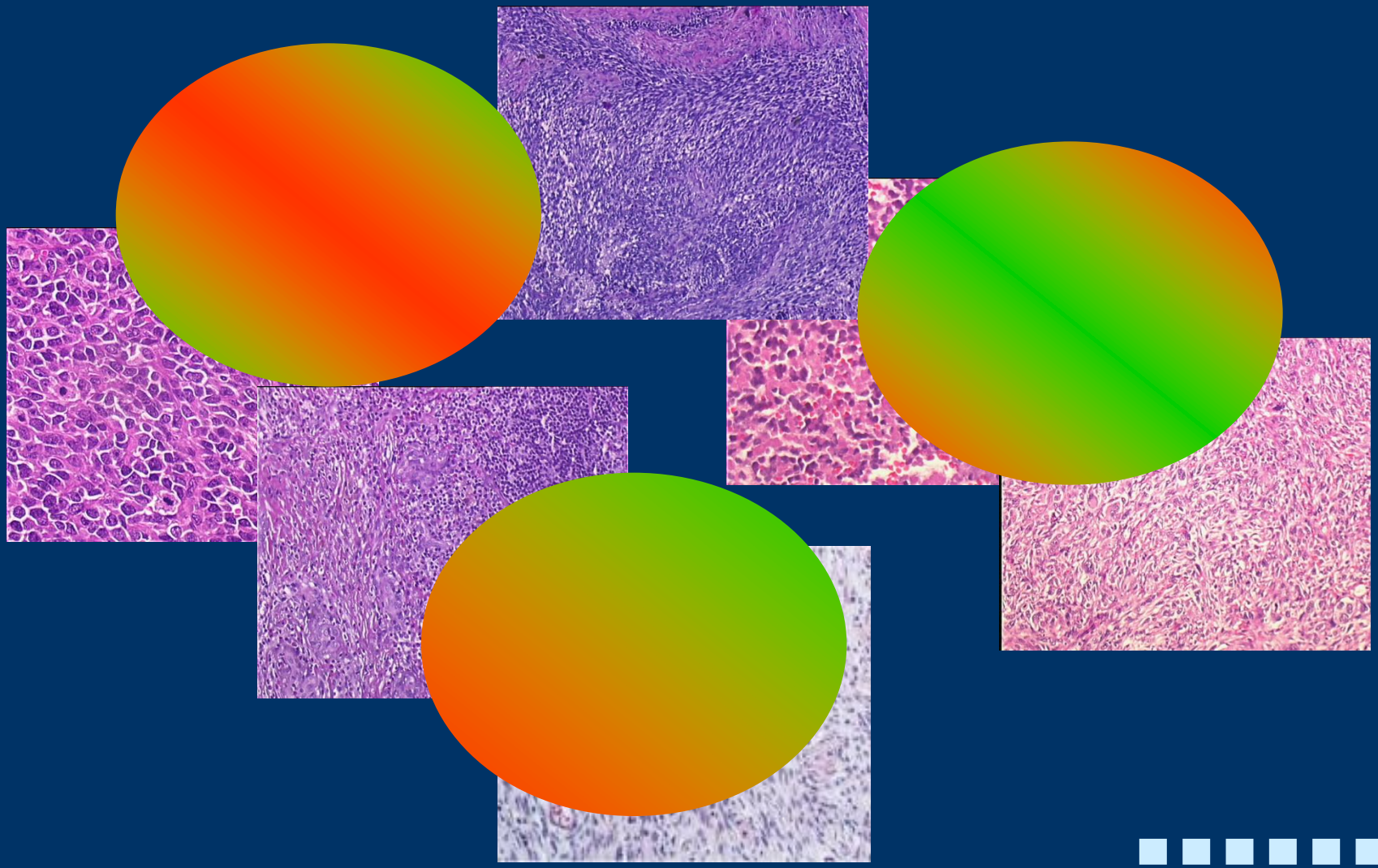


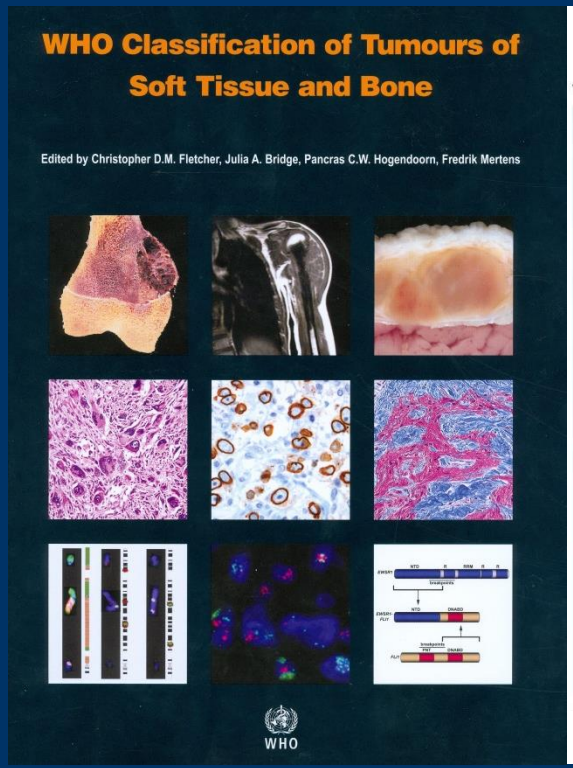






Head & neck	5%
Superficial trunk	10%
Retroperitoneum	15%
Viscera	10%
Limbs	60%





WHO classification of tumours of soft tissue[a,b]

ADIPOCYTIC TUMOURS

Benign

Lipoma	8850/0
Lipomatosis	8850/0
Lipomatosis of nerve	8850/0
Lipoblastoma/lipoblastomatosis	8881/0
Angiolipoma	8861/0
Myolipoma	8890/0
Chondroid lipoma	8862/0
Extra-renal angiomyolipoma	8860/0
Extra-adrenal myelolipoma	8870/0
Spindle cell/pleomorphic lipoma	8857/0
Hibernoma	8880/0

Intermediate (locally aggressive)

Atypical lipomatous tumour/ well differentiated liposarcoma	8850/1 8850/3

Malignant

Dedifferentiated liposarcoma	8858/3
Myxoid liposarcoma	8852/3
Pleomorphic liposarcoma	8854/3
Liposarcoma, not otherwise specified	8850/3

FIBROBLASTIC / MYOFIBROBLASTIC TUMOURS

Benign

Nodular fasciitis	8828/0*
Proliferative fasciitis	8828/0*
Proliferative myositis	8828/0*
Myositis ossificans	
Fibro-osseous pseudotumour of digits	
Ischaemic fasciitis	
Elastofibroma	8820/0
Fibrous hamartoma of infancy	
Fibromatosis colli	
Juvenile hyaline fibromatosis	
Inclusion body fibromatosis	
Fibroma of tendon sheath	8813/0
Desmoplastic fibroblastoma	8810/0
Mammary-type myofibroblastoma	8825/0
Calcifying aponeurotic fibroma	8816/0*
Angiomyofibroblastoma	8826/0
Cellular angiofibroma	9160/0
Nuchal-type fibroma	8810/0
Gardner fibroma	8810/0
Calcifying fibrous tumour	8817/0*

Intermediate (locally aggressive)

Palmar/plantar fibromatosis	8813/1*
Desmoid-type fibromatosis	8821/1
Lipofibromatosis	8851/1*
Giant cell fibroblastoma	8834/1

Intermediate (rarely metastasizing)

Dermatofibrosarcoma protuberans	8832/1*
Fibrosarcomatous dermatofibrosarcoma protuberans	8832/3*
Pigmented dermatofibrosarcoma protuberans	8833/1*
Solitary fibrous tumour	8815/1*
Solitary fibrous tumour, malignant	8815/3
Inflammatory myofibroblastic tumour	8825/1
Low-grade myofibroblastic sarcoma	8825/3*
Myxoinflammatory fibroblastic sarcoma/ Atypical myxoinflammatory fibroblastic tumour	8811/1*
Infantile fibrosarcoma	8814/3

Malignant

Adult fibrosarcoma	8810/3
Myxofibrosarcoma	8811/3
Low-grade fibromyxoid sarcoma	8840/3*
Sclerosing epithelioid fibrosarcoma	8840/3*

SO-CALLED FIBROHISTIOCYTIC TUMOURS

Benign

Tenosynovial giant cell tumour localized type	9252/0
diffuse type	9252/1*
malignant	9252/3
Deep benign fibrous histiocytoma	8831/0

Intermediate (rarely metastasizing)

Plexiform fibrohistiocytic tumour	8835/1
Giant cell tumour of soft tissues	9251/1

SMOOTH MUSCLE TUMOURS

Benign

Deep leiomyoma	8890/0

Malignant

Leiomyosarcoma (excluding skin)	8890/3

PERICYTIC (PERIVASCULAR) TUMOURS

Glomus tumour (and variants)	8711/0
Glomangiomatosis	8711/1*
Malignant glomus tumour	8711/3
Myopericytoma	8824/0
Myofibroma	8824/0
Myofibromatosis	8824/1
Angioleiomyoma	8894/0

SKELETAL MUSCLE TUMOURS

Benign

Rhabdomyoma	8900/0
Adult type	8904/0
Fetal type	8903/0
Genital type	8905/0

Malignant

Embryonal rhabdomyosarcoma (including botryoid, anaplastic)	8910/3
Alveolar rhabdomyosarcoma (including solid, anaplastic)	8920/3
Pleomorphic rhabdomyosarcoma	8901/3
Spindle cell/sclerosing rhabdomyosarcoma	8912/3

VASCULAR TUMOURS OF SOFT TISSUE

Benign

Haemangioma	9120/0
Synovial	
Venous	9122/0
Arteriovenous haemangioma/malformation	9123/0
Intramuscular	9133/0
Epithelioid haemangioma	9125/0
Angiomatosis	
Lymphangioma	9170/0

Intermediate (locally aggressive)

Kaposiform haemangioendothelioma	9130/1

Intermediate (rarely metastasizing)

Retiform haemangioendothelioma	9136/1*
Papillary intralymphatic angioendothelioma	9135/1
Composite haemangioendothelioma	9136/1
Pseudomyogenic (epithelioid sarcoma-like) haemangioendothelioma	9136/1
Kaposi sarcoma	9140/3

Malignant

Epithelioid haemangioendothelioma	9133/3
Angiosarcoma of soft tissue	9120/3

CHONDRO-OSSEOUS TUMOURS

Soft tissue chondroma	9220/0
Extraskeletal mesenchymal chondrosarcoma	9240/3
Extraskeletal osteosarcoma	9180/3

GASTROINTESTINAL STROMAL TUMOURS

Benign gastrointestinal stromal tumour	8936/0
Gastrointestinal stromal tumour, uncertain malignant potential	8936/1
Gastrointestinal stromal tumour, malignant	8936/3

NERVE SHEATH TUMOURS

Benign

Schwannoma (including variants)	9560/0
Melanotic schwannoma	9560/1*
Neurofibroma (incl. variants)	9540/0
Plexiform neurofibroma	9550/0
Perineurioma	9671/0
Malignant perineurioma	9671/3
Granular cell tumour	9580/0
Dermal nerve sheath myxoma	9562/0
Solitary circumscribed neuroma	9570/0
Ectopic meningioma	9530/0
Nasal glial heterotopia	
Benign Triton tumour	
Hybrid nerve sheath tumours	9563/0*

Malignant

Malignant peripheral nerve sheath tumour	9540/3
Epithelioid malignant peripheral nerve sheath tumour	9542/3*
Malignant Triton tumour	9561/3
Malignant granular cell tumour	9580/3
Ectomesenchymoma	8921/3

TUMOURS OF UNCERTAIN DIFFERENTIATION

Benign

Acral fibromyxoma	8811/0
Intramuscular myxoma (including cellular variant)	8840/0
Juxta-articular myxoma	8840/0
Deep ("aggressive") angiomyxoma	8841/0*
Pleomorphic hyalinizing angiectatic tumour	8822/1*
Ectopic hamartomatous thymoma	8587/0

Intermediate (locally aggressive)

Haemosiderotic fibrolipomatous tumour	8811/1*

Intermediate (rarely metastasizing)

Atypical fibroxanthoma	8830/1
Angiomatoid fibrous histiocytoma	8836/1
Ossifying fibromyxoid tumour	8842/0
Ossifying fibromyxoid tumour, malignant	8842/3*
Mixed tumour NOS	8940/0
Mixed tumour NOS, malignant	8940/3
Myoepithelioma	8982/0
Myoepithelial carcinoma	8982/3
Phosphaturic mesenchymal tumour, benign	8990/0
Phosphaturic mesenchymal tumour, malignant	8990/3

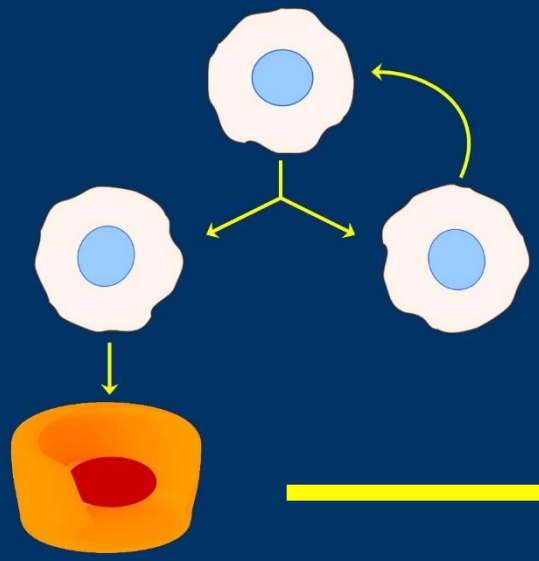
Malignant

Synovial sarcoma NOS	9040/3
Synovial sarcoma, spindle cell	9041/3
Synovial sarcoma, biphasic	9043/3
Epithelioid sarcoma	8804/3
Alveolar soft-part sarcoma	9581/3
Clear cell sarcoma of soft tissue	9044/3
Extraskeletal myxoid chondrosarcoma	9231/3
Extraskeletal Ewing sarcoma	9364/3
Desmoplastic small round cell tumour	8806/3
Extra-renal rhabdoid tumour	8963/3
Neoplasms with perivascular epithelioid cell differentiation (PEComa)	
PEComa NOS, benign	8714/0*
PEComa NOS, malignant	8714/3*
Intimal sarcoma	9137/3*

UNDIFFERENTIATED/UNCLASSIFIED SARCOMAS

Undifferentiated spindle cell sarcoma	8801/3
Undifferentiated pleomorphic sarcoma	8802/3
Undifferentiated round cell sarcoma	8803/3
Undifferentiated epithelioid sarcoma	8804/3
Undifferentiated sarcoma NOS	8805/3

[a] The morphology codes are from the International Classification of Diseases for Oncology (ICD-O) (916A). Behaviour is coded /0 for benign tumours, /1 for unspecified, borderline or uncertain behaviour, /2 for carcinoma in situ and grade III intraepithelial neoplasia, and /3 for malignant tumours. [b] The classification is modified from the previous WHO histological classification of tumours (870A) taking into account changes in understanding of these lesions. * These new codes were approved by the IARC/WHO Committee for ICD-O in 2012.



differentiation towards...

- **connective t.**
  (***Fibrosarcoma, Liposarcoma, ...***)
- **muscle**
  (***Leiomyosarcoma, Rhabdomiosarcoma***)
- **endothelium**
  (***Hemangioendotelioma, Angiosarcoma***)
- **nerve sheaths**
  (***M. peripheral nerve sheaths tumor***)

Rhabdomyosarcoma in Adults

A Retrospective Analysis of 171 Patients Treated at a Single Institution

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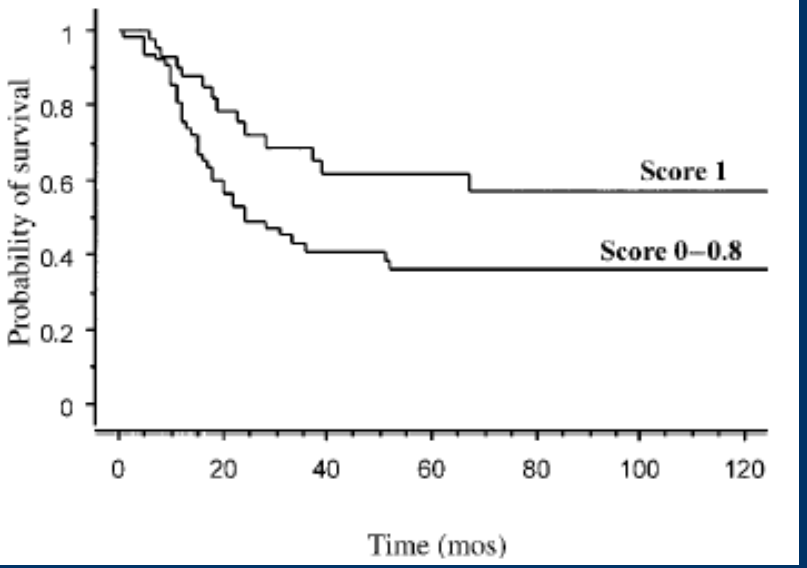
BACKGROUND. The goal of the current study was to clarify treatment outcomes for adult patients with rhabdomyosarcoma (RMS). Published series have reported definitively worse results for adults with RMS compared with children with RMS. This finding casts doubt on whether RMS is the same disease in adults as it is in children.

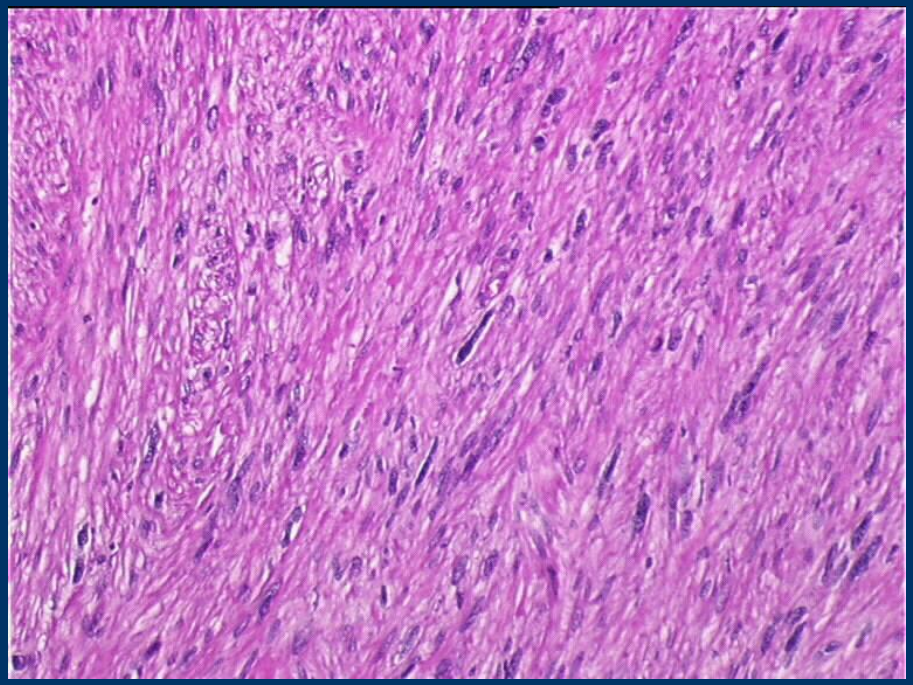
METHODS. Of 190 patients with RMS who were age 18 years or older and whose cases were recorded over a 25-year span in the pathology database of the Istituto Nazionale Tumori (Milan, Italy), 171 could be analyzed retrospectively for treatment outcome. The authors attempted to stratify patients according to the degree to which they had been treated appropriately, based on current treatment guidelines for childhood RMS.

RESULTS. The overall rate of response to chemotherapy was 85%. For the entire series, 5-year event-free survival and 5-year overall survival (OS) were 28% and 40%, respectively. Among the 110 patients with embryonal, alveolar, or 'not otherwise specified' RMS, 5-year OS was 46%; however, 5-year OS was 61% for patients within this group (39% of the total) who had high scores for appropriate treatment.

CONCLUSIONS. The current series parallels other published series in that it confirms the finding of a relatively poor long-term outcome for adult patients with RMS. However, for patients whose treatment adhered to the current guidelines for treatment of children, outcome was similar to what has been reported in pediatric series. In addition, the rate of response to chemotherapy for the entire series was similar to the rate typically observed among children. These findings suggest that adults and children with RMS should receive similar treatment. Treatment protocols adopted from pediatric programs but tailored to adults could increase adults' chances of receiving appropriate treatment; prospective studies are needed to test this idea.

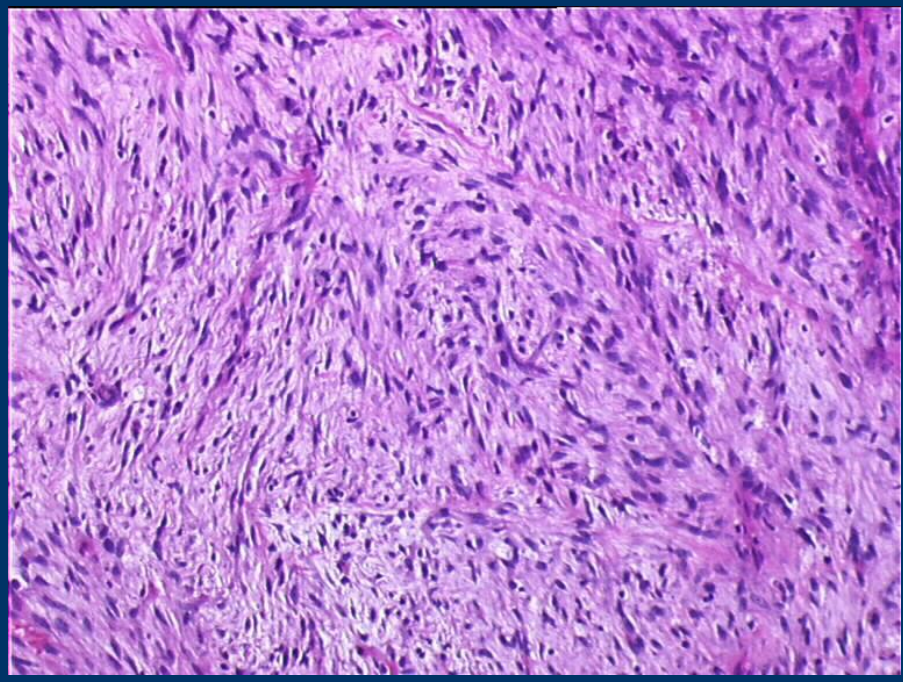
Cancer **2003;98:571–80.**

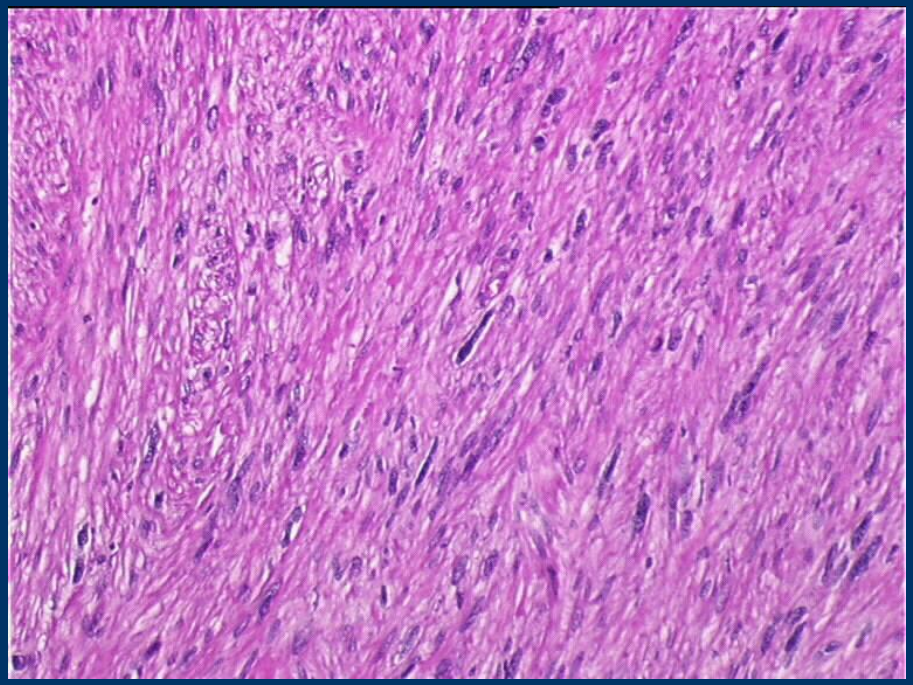




leiomyosarcoma G1

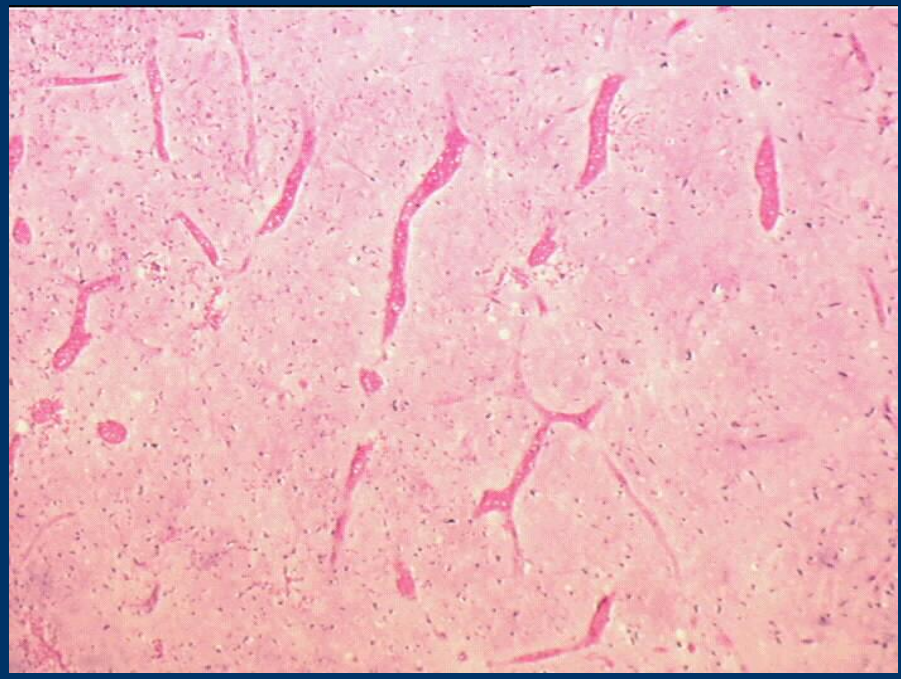
leiomyosarcoma G3





leiomyosarcoma

liposarcoma



The Grading of Soft Tissue Sarcomas

Results of a Clinicohistopathologic Correlation in a Series of 163 Cases

JOSE COSTA,* R. A. WESLEY,† E. GLATSTEIN,‡ AND S. A. ROSENBERG§

A multidisciplinary study of 163 patients treated at the NCI for soft tissue sarcomas allowed the correlation of a number of histologic features (histologic type, mitosis, necrosis, pleomorphism, cellularity, and matrix) of the primary lesion to time to recurrence and overall survival of the patients. The results of the stratified analyses show that necrosis is the single best histopathologic parameter to predict the time to recurrence ($P = 0.025$) and the overall survival of the patients ($P = 0.002$). Necrosis in the primary lesion is also of value in predicting survival after the first recurrence has taken place ($P = 0.001$). The value of necrosis in the primary lesions predicting the clinical course after recurrence appears to be independent of age, sex, location, and size of the tumor. The authors propose a grading system based on histologic typing and histologic parameters to identify a group of lesions with minimal metastatic potential (Grade 1), and on the use of necrosis to distinguish between aggressive lesions with good patient survival (Grade 2) and aggressive lesions with poor patient survival (Grade 3).

Cancer 53:530–541, 1984.

Int. J. Cancer: **33,** 37-42 (1984)

SOFT-TISSUE SARCOMAS OF ADULTS; STUDY OF PATHOLOGICAL PROGNOSTIC VARIABLES AND DEFINITION OF A HISTOPATHOLOGICAL GRADING SYSTEM

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The pathological features of 155 adult patients with soft-tissue sarcomas were studied retrospectively, in an attempt to set up a grading system for these tumors. As the first step, seven histological criteria (tumor differentiation, cellularity, importance of nuclear atypia, presence of malignant giant cells, mitosis count, pattern of tumor necrosis and presence of vascular emboli) were evaluated in a monofactorial analysis. Five of these (tumor differentiation, cellularity, mitosis count, tumor necrosis, and vascular emboli) were correlated with the advent of metastases and with survival. A multivariate analysis, using a Cox model, selected a minimal set of three factors (tumor differentiation, mitosis count, and tumor necrosis) the combination of which was necessary and sufficient to retain all the prognostic information. A grading system was elaborated, which turned out to be correlated with the advent of metastasis and with patients' survival. A second multivariate analysis introducing clinical prognostic features showed that the histological grade was the most important prognostic factor for soft-tissue sarcomas. Thus, this grading system appears to be highly interesting because of its prognostic value and the facility of its elaboration. However, its reproducibility should be tested.

Comparative Study of the National Cancer Institute and French Federation of Cancer Centers Sarcoma Group Grading Systems in a Population of 410 Adult Patients With Soft Tissue Sarcoma

By Louis Guillou, Jean-Michel Coindre, Françoise Bonichon, Nguyen Binh Bui, Philippe Terrier, Françoise Collin, Marie-Odile Vilain, Anne-Marie Mandard, Viviane Le Doussal, Agnès Leroux, Jocelyne Jacquemier, Huguette Duplay, Xavier Sastre-Garau, and José Costa

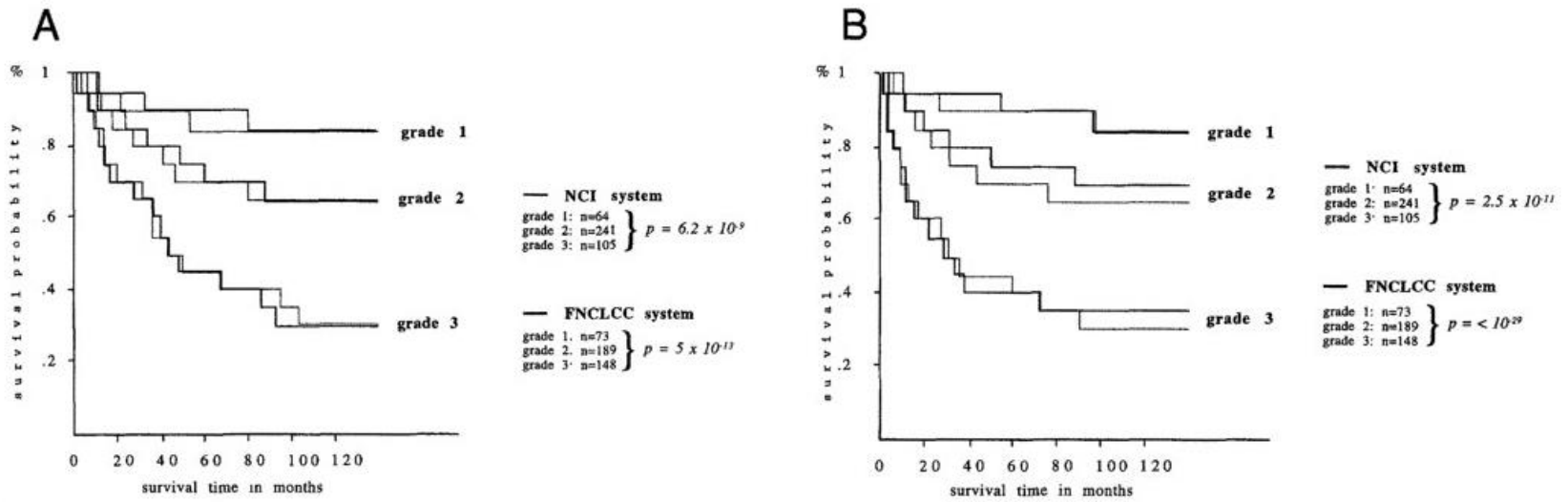
Purpose: **Several histologic grading systems have been validated in soft tissue sarcomas (STS), but no system is currently accepted worldwide. The National Cancer Institute (NCI) and French Federation of Cancer Centers Sarcoma Group (FNCLCC) systems were examined comparatively in the same population of patients with STS to determine which system is the best prognosticator with regard to metastasis development and tumor mortality.**

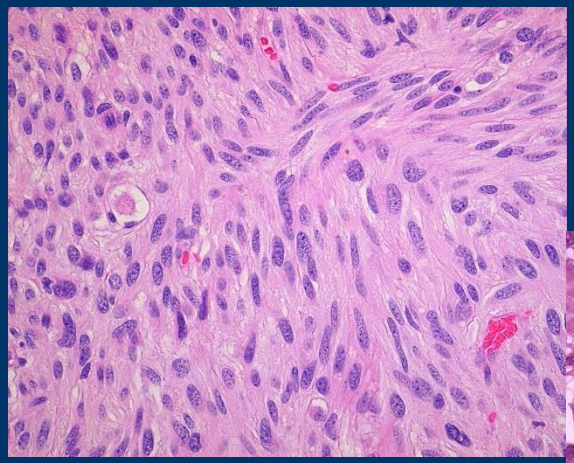
Patients and Methods: **Four hundred ten adult patients with nonmetastatic STS were examined. Histologic grade was established according to the NCI and FNCLCC systems in each case. The prognostic value of both systems was examined using univariate and multivariate (Cox's model) analyses, and special attention was devoted to tumors with discordant grades.**

Results: **In univariate analysis, both the NCI and FNCLCC systems were of prognostic value to predict metastasis development and tumor mortality. In multivariate analysis, high-grade tumors, irrespective of the system used, size ≥ 10 cm, and deep location were found to be independent prognostic factors for the advent of metastases. Tumor grade had a higher predictive value than size or depth, and higher prognostic weight was assigned to the FNCLCC grading system in Cox models. Grade discrepancies were observed in 34.6% of the cases. An increased number of grade 3 STS, a reduced number of grade 2 STS, and a better correlation with overall and metastasis-free survival within subpopulations with discordant grades were observed in favor of the FNCLCC system.**

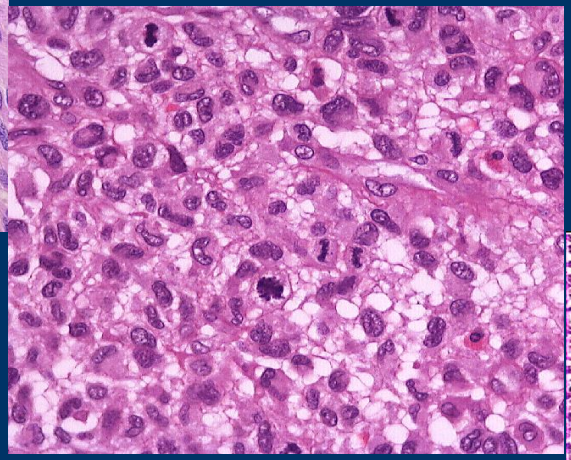
Conclusion: **The FNCLCC system showed slightly increased ability to predict distant metastasis development and tumor mortality. The use of this system to evaluate STS aggressiveness might be favored.**

J Clin Oncol 15:350-362. © 1997 by American Society of Clinical Oncology.

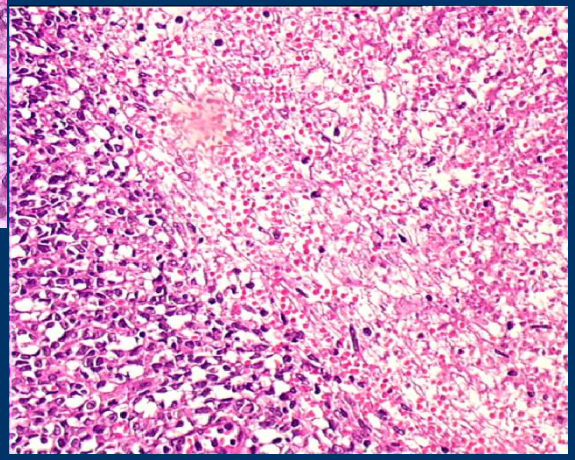




differentiation
1-2-3



mitoses
1-2-3



necrosis
0-1-2

G1 = 2-3
G2 = 4-5
G3 = 6-8

Primary Tumor (T)

TX	Primary tumor cannot be assessed
T0	No evidence of primary tumor
T1	Tumor 5 cm or less in greatest dimension*
T1a	Superficial tumor (Figure 28.3)
T1b	Deep tumor (Figure 28.4)
T2	Tumor more than 5 cm in greatest dimension*
T2a	Superficial tumor (Figure 28.3)
T2b	Deep tumor (Figure 28.5)

**Note*: Superficial tumor is located exclusively above the superficial fascia without invasion of the fascia; deep tumor is located either exclusively beneath the superficial fascia, superficial to the fascia with invasion of or through the fascia, or both superficial yet beneath the fascia.

Regional Lymph Nodes (N)

NX	Regional lymph nodes cannot be assessed
N0	No regional lymph node metastasis
N1*	Regional lymph node metastasis

**Note*: Presence of positive nodes (N1) in M0 tumors is considered Stage III.

Distant Metastasis (M)

M0	No distant metastasis
M1	Distant metastasis

ANATOMIC STAGE/PROGNOSTIC GROUPS

Stage IA	T1a	N0	M0	G1, GX
	T1b	N0	M0	G1, GX
Stage IB	T2a	N0	M0	G1, GX
	T2b	N0	M0	G1, GX
Stage IIA	T1a	N0	M0	G2, G3
	T1b	N0	M0	G2, G3
Stage IIB	T2a	N0	M0	G2
	T2b	N0	M0	G2
Stage III	T2a, T2b	N0	M0	G3
	Any T	N1	M0	Any G
Stage IV	Any T	Any N	M1	Any G

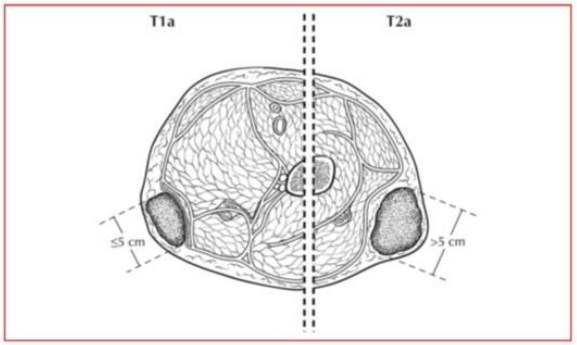


FIGURE 28.3. *T1a is defined as a superficial tumor 5 cm or less in greatest dimension, and T2a is defined as a superficial tumor more than 5 cm in greatest dimension.*

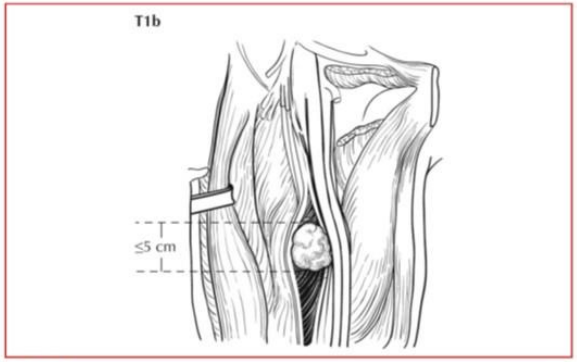


FIGURE 28.4. *T1b is defined as deep tumor 5 cm or less in greatest dimension.*

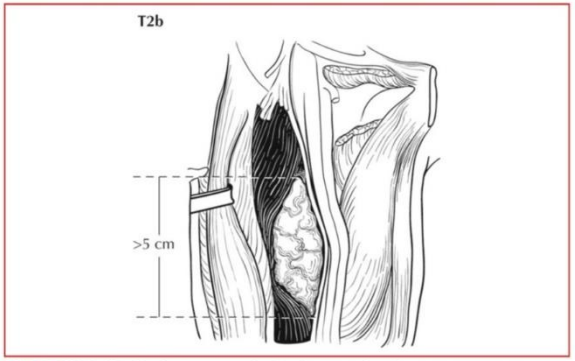


FIGURE 28.5. *T2b is defined as deep tumor more than 5 cm in greatest dimension.*

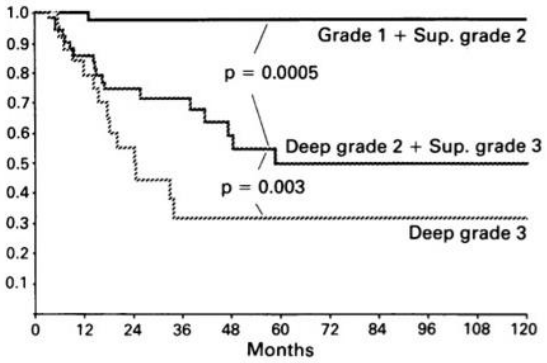


Figure 9 Overall survival according to prognostic groups. Grade 1 + superficial grade 2 (49 pts), deep grade 2 + superficial grade 3 (52 pts), deep grade 3 (30 pts).

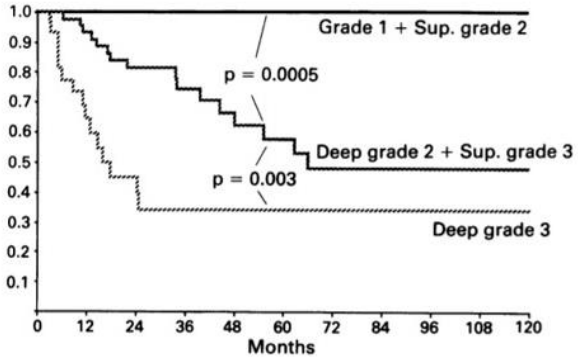


Figure 10 Metastasis-free survival according to prognostic groups. Grade 1 + superficial grade 2 (49 pts), deep grade 2 + superficial grade 3 (52 pts), deep grade 3 (30 pts).

Ravaud A et al. Br J Cancer 1992;66:961

Overall Survival Probability Following Surgery

Our uterine leiomyosarcoma nomogram is a tool designed to predict the likelihood of survival at five years after undergoing surgery to remove the uterine leiomyosarcoma, a type of uterine cancer. It is not appropriate for patients who have not had surgical treatment for uterine leiomyosarcoma. more...

Enter Your Information

Clear Calculate

All fields are required unless noted optional

How old were you when you were diagnosed with uterine leiomyosarcoma?

years (20 to 80)

What was the size of the primary uterine leiomyosarcoma tumor?

cm (0 to 28)

What was the grade of the tumor?

Select tumor grade

What is grade?

Was there cervical involvement at the time of your surgery?

Yes No

What is cervical involvement?

Were there loco-regional metastases at the time of your surgery?

Yes No

Note: Loco-regional metastases were present if, at the time of surgery, the leiomyosarcoma was found in the structures near the uterus, including the bladder, nearby bowel, nearby lymph nodes, parametria, ovaries, or fallopian tubes.

Were there distant metastases either before or soon after your surgery?

Yes No

Note: Distant metastases were present if imaging studies done either before or soon after surgery showed that the leiomyosarcoma had spread to distant areas, such as the lung, liver, or bone.

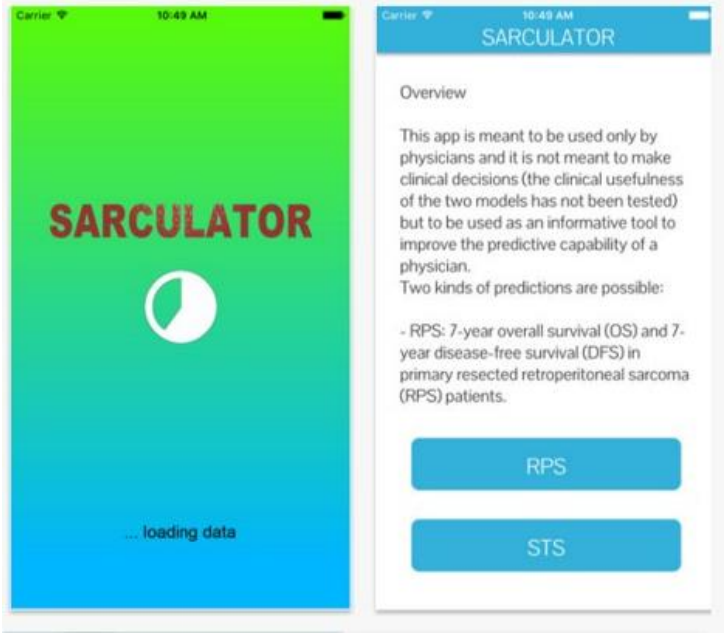
What was the mitotic index?

mitoses/10 HPF (1 to 245)

Note: This is expressed as the number of mitotic figures per 10 high-powered fields (HPF) seen under the microscope. If the pathology report gives a range (such as 8 to 20 mitoses/10 HPF), use the higher number (20 mitoses/10 HPF).

What is mitotic index?

Calculate Clear



Local treatment

amputation

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compartimental resection

reactive zone

pseudo capsule

T

wide excision

marginal excision

intralesional excision

The Effect of the Anatomic Setting on the Results of Surgical Procedures for Soft Parts Sarcoma of the Thigh

WILLIAM F. ENNEKING, MD,* SUZANNE S. SPANIER, MD,† AND MARTIN M. MALAWER, MD‡

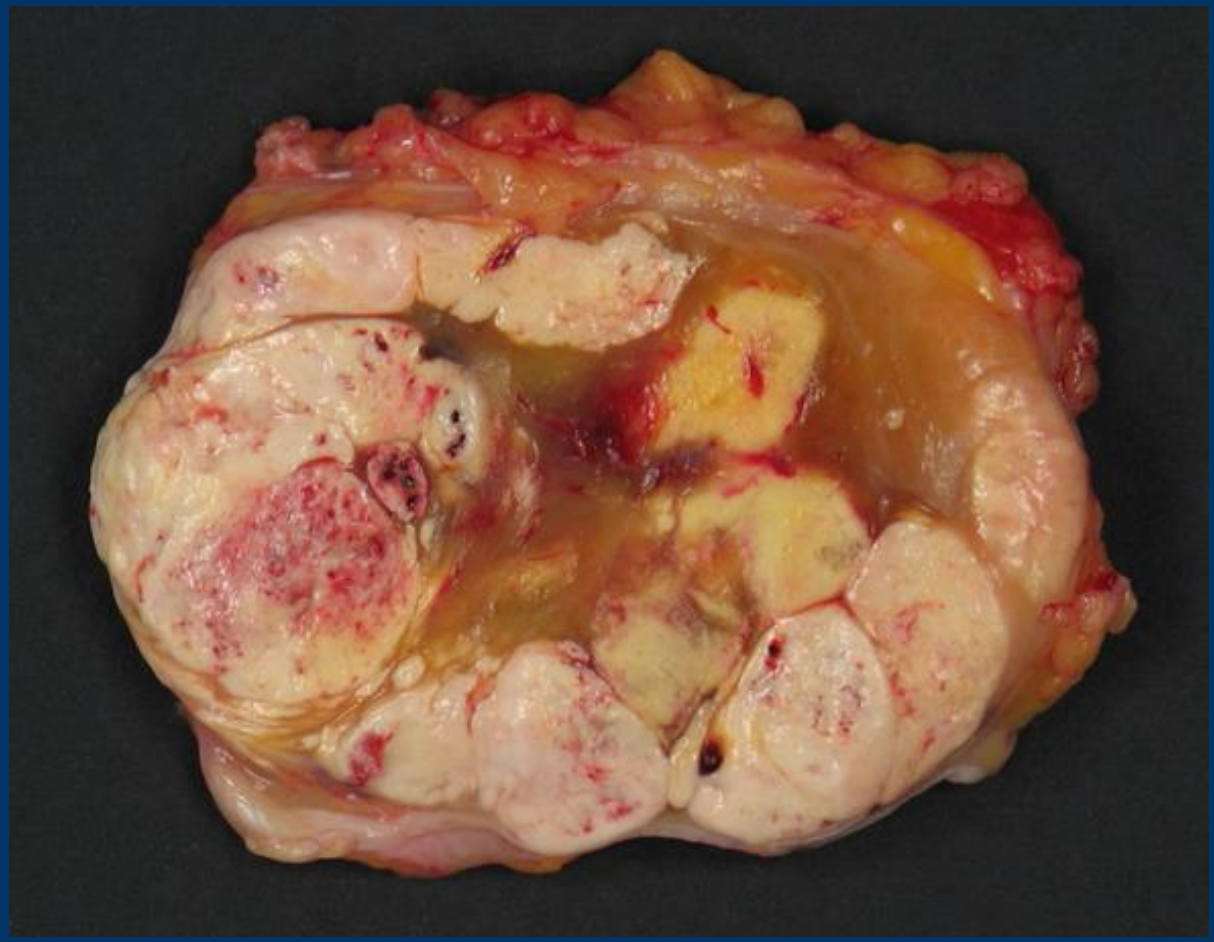
The results of surgical treatment in 40 patients with a soft tissue sarcoma of the thigh were analyzed to determine the influence of the anatomic setting on the effectiveness of the procedure. The anatomic setting, based on functional anatomic compartments, was defined as either intra- or extracompartmental. The lesions were graded for aggressiveness as either high or low. The lesions were staged by biologic aggressiveness, anatomic setting, and metastases. The procedures, whether amputations or local resections, were classified by the relationship of the surgical margin to the pseudocapsule and reactive zone about the lesion as marginal, wide, or radical.

Marginal procedures were done four times with two recurrences. Wide margins were achieved 12 times. When done for low grade lesions, there were no recurrences (0/2), but when done for high grade lesions, the recurrence rate was 30% (3/10). Radical margins were obtained 24 times. There was one recurrence after a radical procedure. Recurrence rates did not depend upon whether the procedure was a resection or amputation but upon the margin achieved.

The anatomic setting of the lesion was intracompartmental in 13 cases and extracompartmental in 27. Not only were surgically adequate margins achieved more often for intracompartmental lesions (10/13) than for extracompartmental lesions (17/27), but there was a significant difference in the manner required to achieve an adequate margin. Although 9 of the 13 intracompartmental lesions were amenable to nonablative resection, only 3 of 27 extracompartmental lesions were resectable.

The margin required for local control (wide vs. radical) was dictated by the biologic aggressiveness (grade) of the lesion. How the necessary margin was most satisfactorily achieved (resection vs. amputation) was determined by the anatomic setting (intra- vs. extracompartmental).

Cancer **47:1005–1022, 1981.**



Is There No Influence of Local Control on the Rate of Metastases in High-Grade Soft Tissue Sarcoma?

Bo Rööser, MD, PhD, Pelle Gustafson, MD,
and Anders Rydholm, MD, PhD

Primary host and tumor-related prognostic factors, the occurrence of local recurrence, and the time interval between diagnosis of the primary tumor and metastatic disease were analysed in 39 patients with metastatic Grade 4 soft tissue sarcoma. The pattern of prognostic factors and the timing of metastases were the same in patients with and without local recurrence. Thus, primary prognostic factors alone may determine the risk of metastases in high-grade soft tissue sarcoma; local recurrence *per se* may not influence the prognosis in these cases.
***Cancer* 65:1727–1729, 1990.**

Extremity Soft Tissue Sarcoma in a Series of Patients Treated at a Single Institution

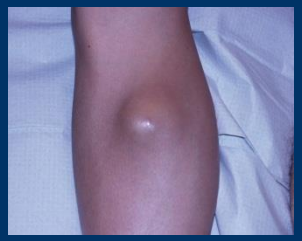
Local Control Directly Impacts Survival

Alessandro Gronchi, MD, Salvatore Lo Vullo, BSc,† Chiara Colombo, MD,* Paola Collini, MD,‡ Silvia Stacchiotti, MD,§ Luigi Mariani, MD,† Marco Fiore, MD,* and Paolo Giovanni Casali, MD§*

Purpose: To improve understanding of what is adequate in local treatment of extremity soft tissue sarcomas (ESTS), to maximize the ratio between local control, limb preservation and prognosis.
Patient and Methods: Nine hundred ninety-seven consecutive patients affected by primary ESTS were reviewed. Size, depth, histotype and grade of the tumor, margin status (R0, R1, R2) of surgical resection, and adjuvant treatments were analyzed. Univariable and multivariable analysis were carried out. For the subgroup of R1 resection the presence/absence of the tumor at the inked surface and the presence/absence of an anatomic barrier were also considered.
Results: Five- and 10-year mortality estimates (95% confidence interval) were 0.29 (0.20–0.38) and 0.38 (0.28–0.49) in R1 cases, and 0.16 (0.13–0.19) and 0.19 (0.16–0.23) in R0 cases ($P = 0.0003$). Size, grade, depth, and histologic subtype were also significant predictor of mortality. Significant determinants for local relapse were surgical margins, radiation therapy, and histologic subtype. In the subset of R1 resections trends towards a better local control for R1 negative cases and histology other than myxofibrosarcoma were identified. Significant determinants for distant metastases were size, grade and histologic subtype of the tumor but not surgical margins.
Conclusions: Quality of surgical margins independently predicted local control and survival. The effect on survival was directly mediated by local recurrence to proximal sites invading the abdomen/thorax, and this may indeed be the main way by which quality of surgery directly impacts the final prognosis of ESTS patients.

(*Ann Surg* 2010;251: 506–511)

local relapse



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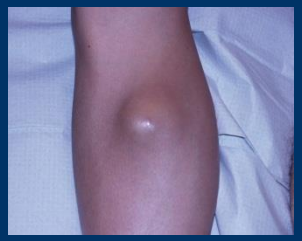
systemic relapse

local relapse



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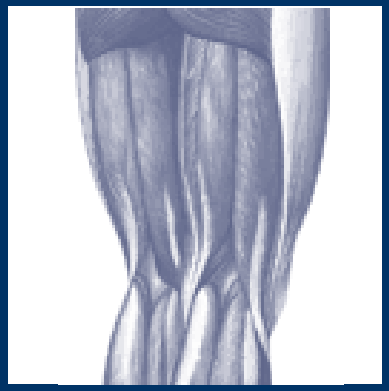
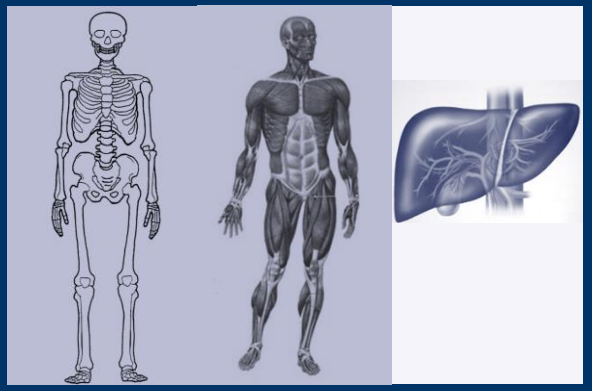
- **re-excision** → **costs!**
- **mutilations** → **QoL!**
- **up-staging(grading)** → **OS!**

Natural history of disease

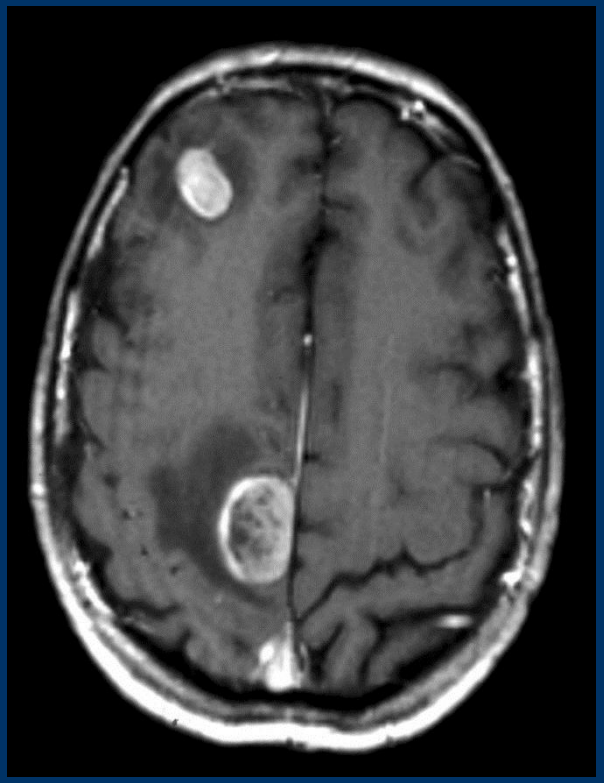


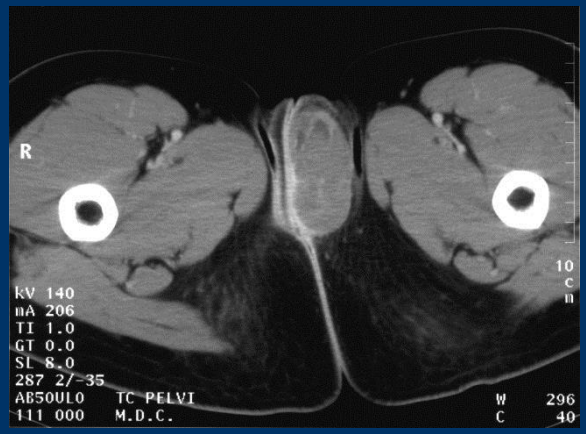


localized

isolated pulmonary

extra pulmonary

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kV 140
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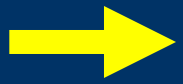

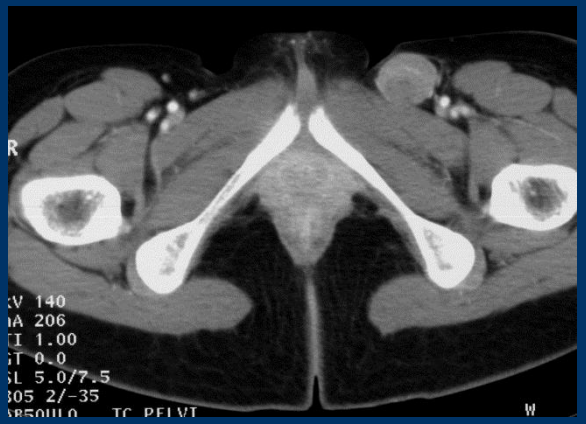
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Lymph Nodes as Sites of Metastases From Sarcomas of Soft Tissue

JEAN-JACQUES MAZERON, MD,* AND HERMAN D. SUIT, MD

Records of 323 patients with TNM Stage M0 sarcoma of soft tissue treated by the Radiation Medicine Service of the Massachusetts General Hospital over a 14-year period were reviewed to study the incidence and the implication of regional lymph node involvement. Nineteen patients (5.9%) had evidence of sarcoma metastatic to draining lymph nodes, zero of 63 (0%) were Grade 1 sarcomas, two of 118 (2%) were Grade 2, and 17 of 142 (12%) were Grade 3 sarcomas. Among patients with Grade 3 sarcomas, rhabdomyosarcoma (five of 14), vascular sarcoma (two of five), and epithelioid sarcoma (four of five) were associated with a higher incidence of lymph node involvement than synovial sarcomas (zero of four), fibrosarcomas (zero of 16), malignant fibrohistiocytomas (one of 29), neurofibrosarcomas (one of eight), liposarcomas (one of 14), and leiomyosarcomas (one of ten). From the 19 patients who had evidence of metastatic nodes, six (32%) were alive more than 58 months after the treatment of the nodes; four of six patients were without further tumor. The data of this study are compared with those cited in a review of the literature.

Cancer **60:1800–1808, 1987.**

TABLE 5. Pooled Data from Published Reports on Regional Lymph Node Involvement in 5257 Patients Treated for Sarcoma of Soft Tissue

Histologic type	Proportion of patients with spread to lymph nodes	
	No./no. of patients	Percent
Liposarcoma	16/504	3.2
Fibrosarcoma	54/215	4.4
Synovial sarcoma	117/851	13.7
Rhabdomyosarcoma	201/1354	14.8
Leiomyosarcoma	21/524	4
Malignant fibrous histiocytoma	84/823	10.2
Alveolar soft part sarcomas	3/24	12.5
Neurofibrosarcoma	3/476	0.6
Epithelioid sarcoma	14/70	20
Vascular sarcoma	43/376	11.4
Clear cell sarcoma	11/40	27.5

World J Surg (2007) 31:1433–1437
DOI 10.1007/s00268-007-9078-3

World Journal of Surgery

The Impact of Lymph Node Metastases on Survival in Extremity Soft Tissue Sarcomas

Can Atalay · Mehmet Altinok · Besim Seref

Abstract

Background The impact of lymph node metastases on survival in extremity soft tissue sarcomas has been studied for a long time with controversial results. The purpose of this study was to compare survival of patients with initial lymph node metastases with those having lymph node or distant metastases or both after initial curative surgery.

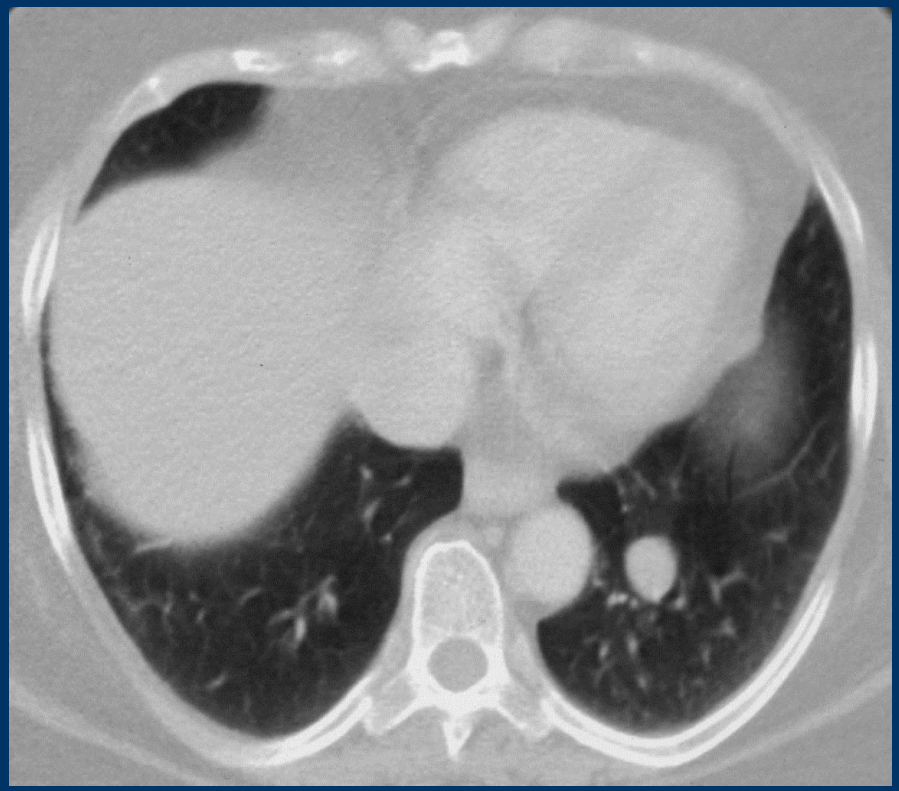
Methods Patients treated between 1995 and 2000 for extremity soft tissue sarcoma were retrospectively studied in four groups: those with metastatic regional lymph nodes at the time of diagnosis, those with only regional lymph node recurrences, those with only distant metastatic relapses, and those with both regional lymph node recurrences and distant metastatic relapses, all of the last three groups after initial curative surgery. The impact of timing of lymph node metastases on disease-free and overall survival was evaluated.

Results A total of 110 patients (73 men) with a median age of 45 years were eligible for the study. Three-year disease-free survival was significantly longer in patients with initial regional lymph node metastases than in patients with only lymph node recurrences after curative surgery ($p = 0.04$) and patients with initial ($p = 0.0002$) and recurrent ($p = 0.0004$) regional lymph node metastases had longer disease-free survival than patients with distant metastases. Overall survival difference between patients with initial regional lymph node metastases and patients with only lymph node recurrences after curative surgery was significant at 5 years ($p = 0.01$).

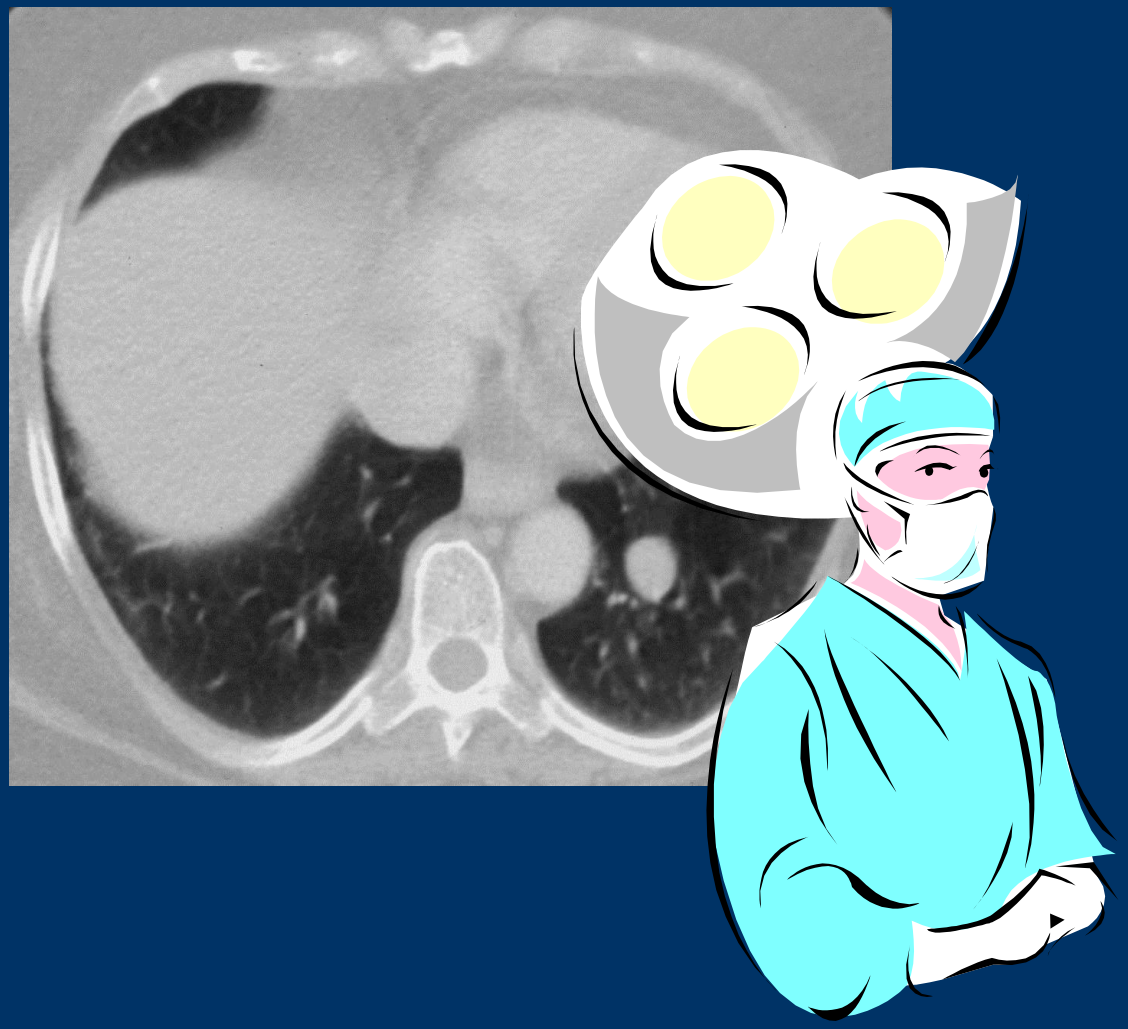
Conclusions It is logical to separate patients with initial lymph node metastases from those with distant metastases in staging and to treat patients with initial lymph node metastases with radical surgical interventions if complete tumor resection seems feasible.

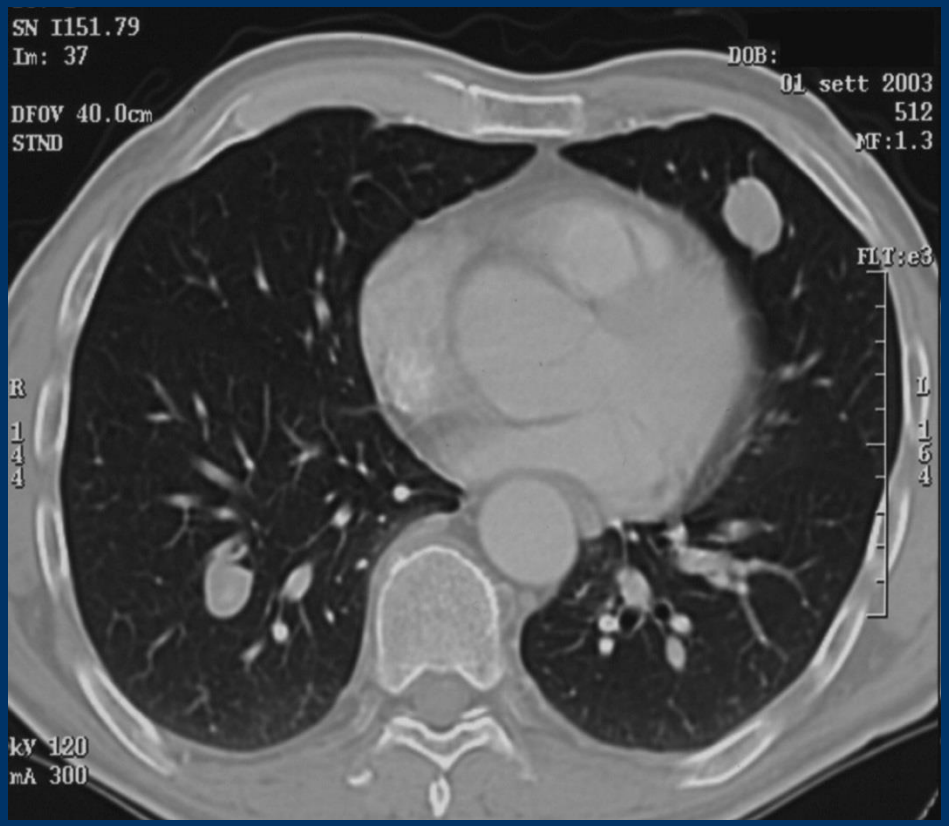
First isolated pulmonary metastases

Osteosa	>70%
Soft tissue sa	>50%
Ewing	>40%
Rhabdomyosa	>20%



Soft tissue sarcoma: isolated pulmonary metastasis

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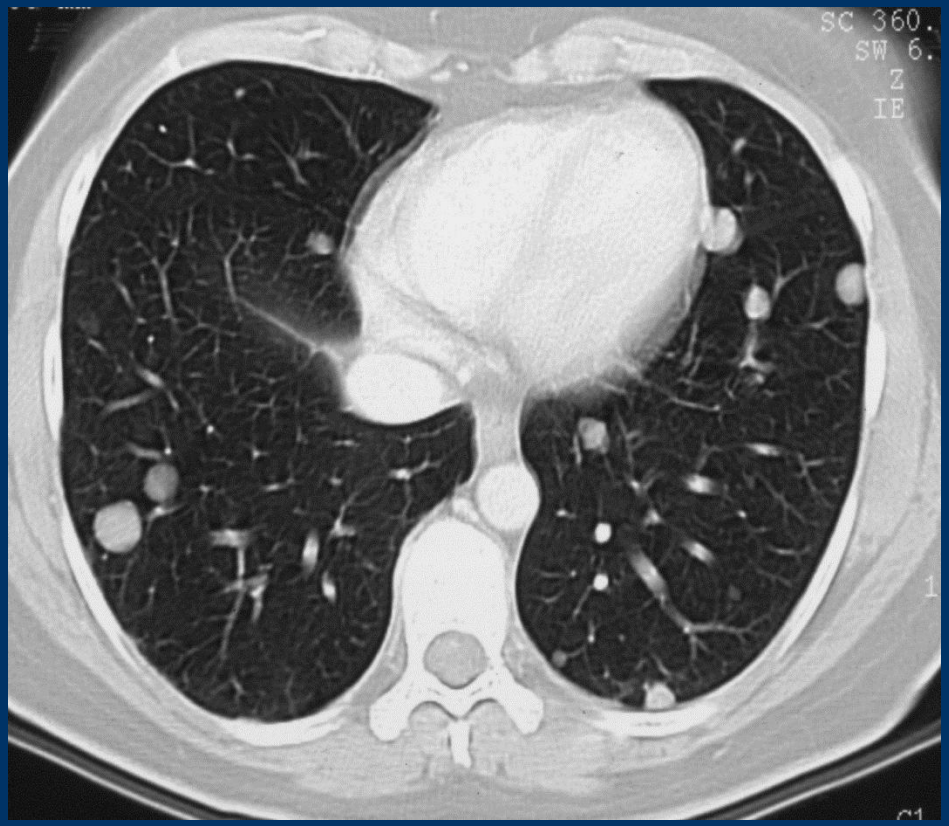
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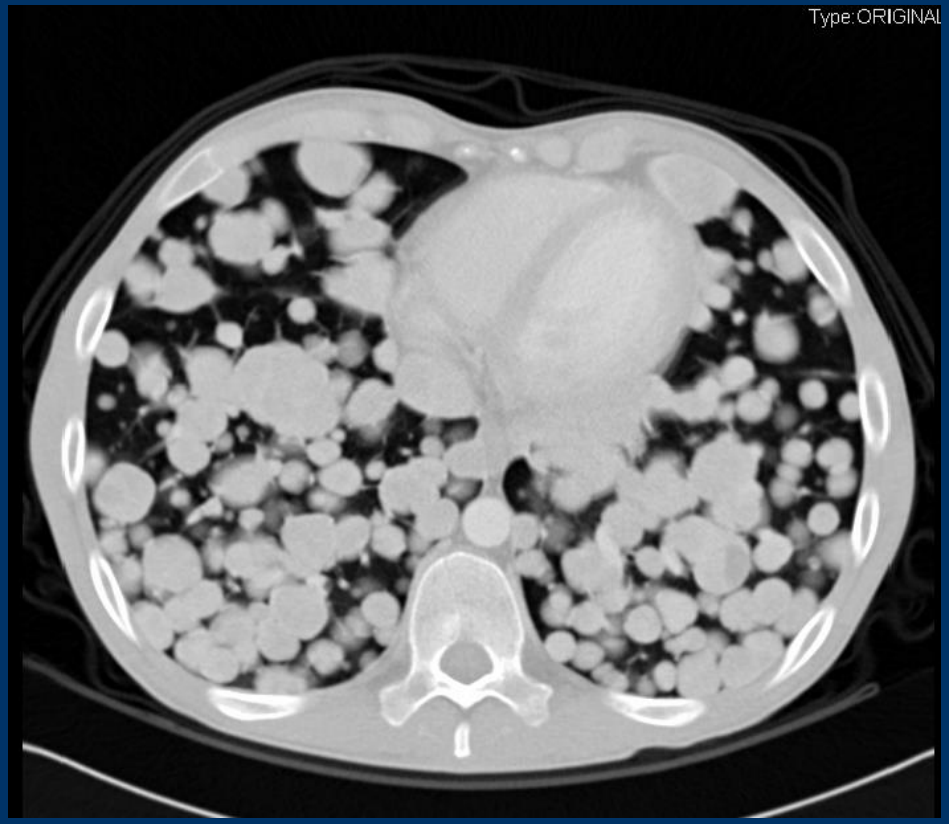
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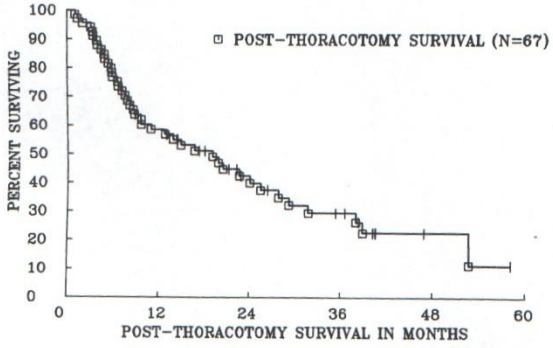
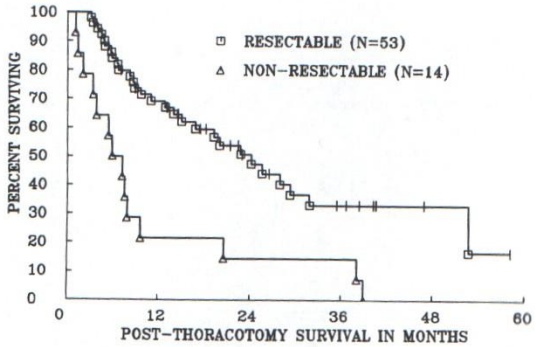
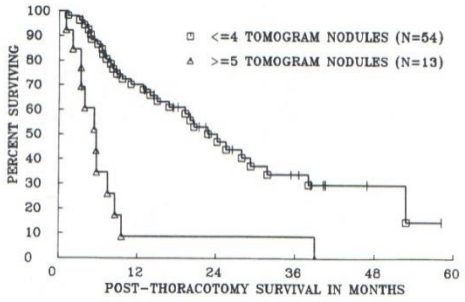
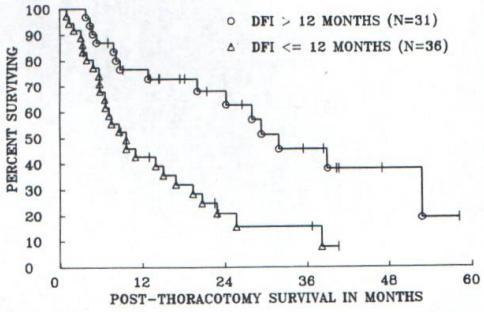
J THORAC CARDIOVASC SURG 87:260-268, 1984

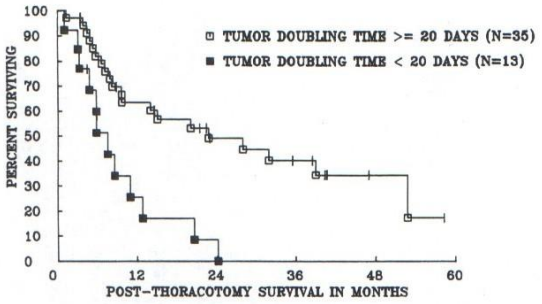
Analysis of prognostic factors in patients undergoing resection of pulmonary metastases from soft tissue sarcomas

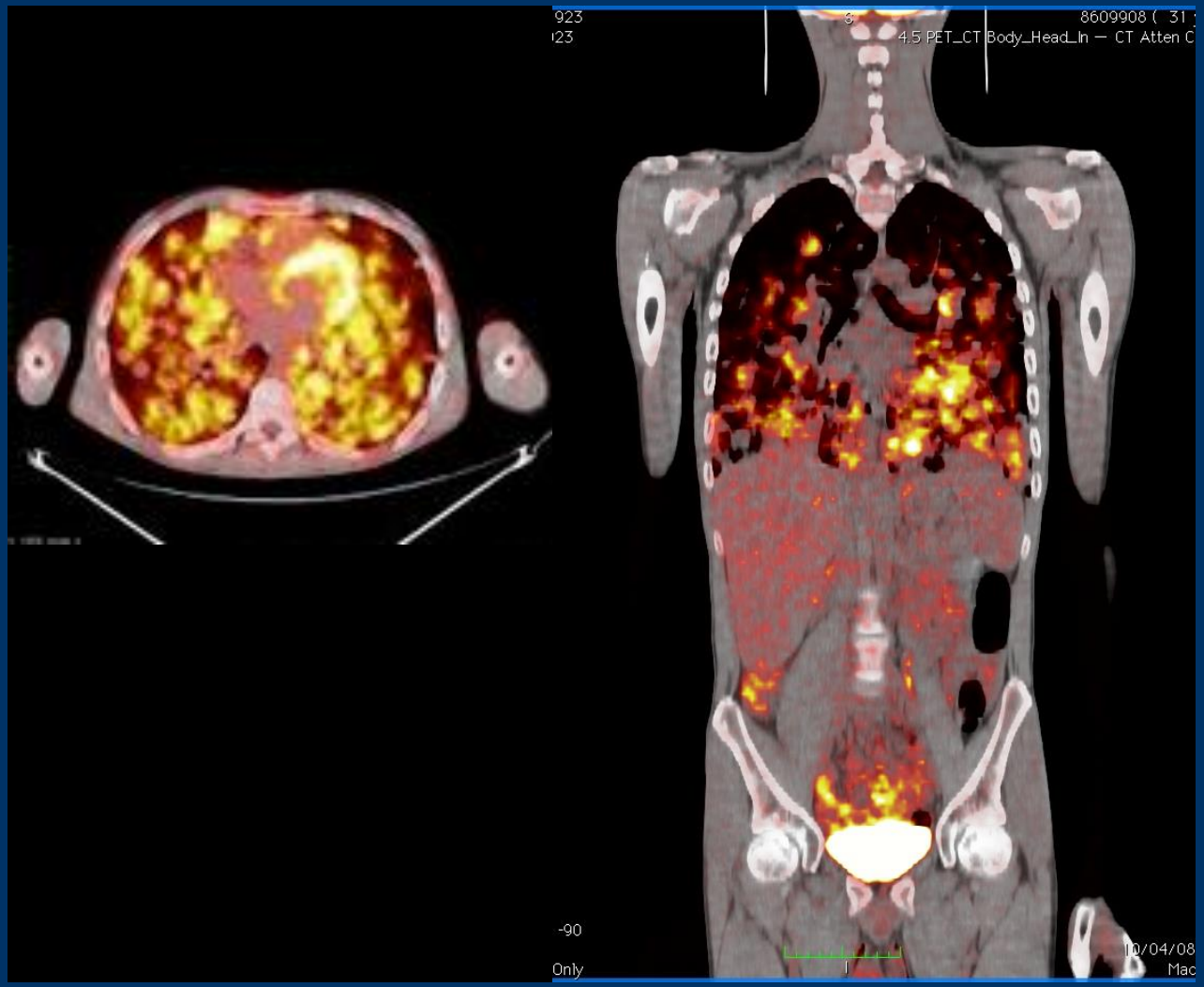








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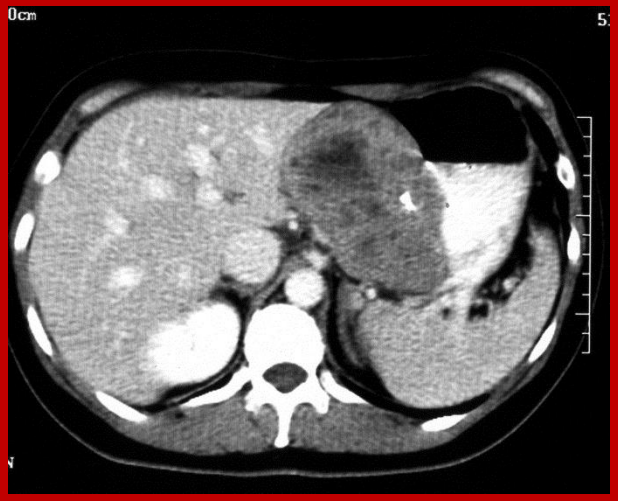
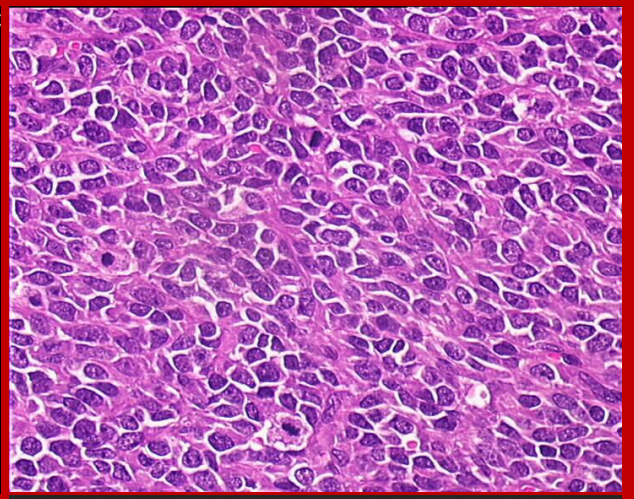
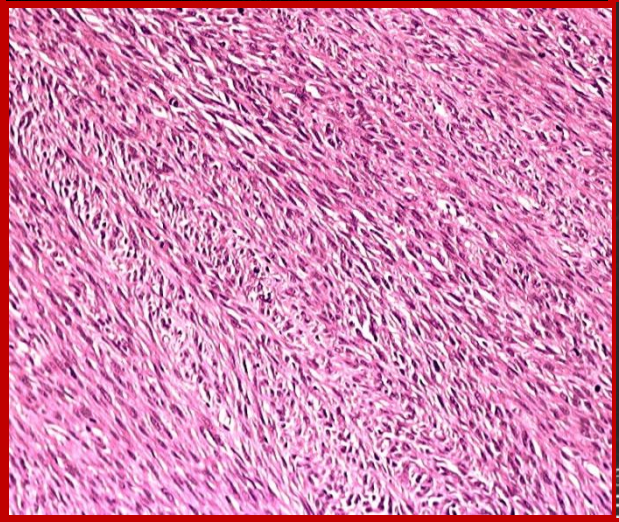
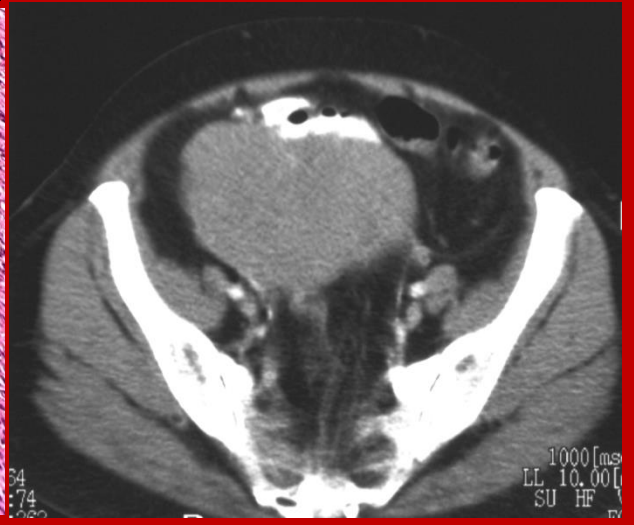
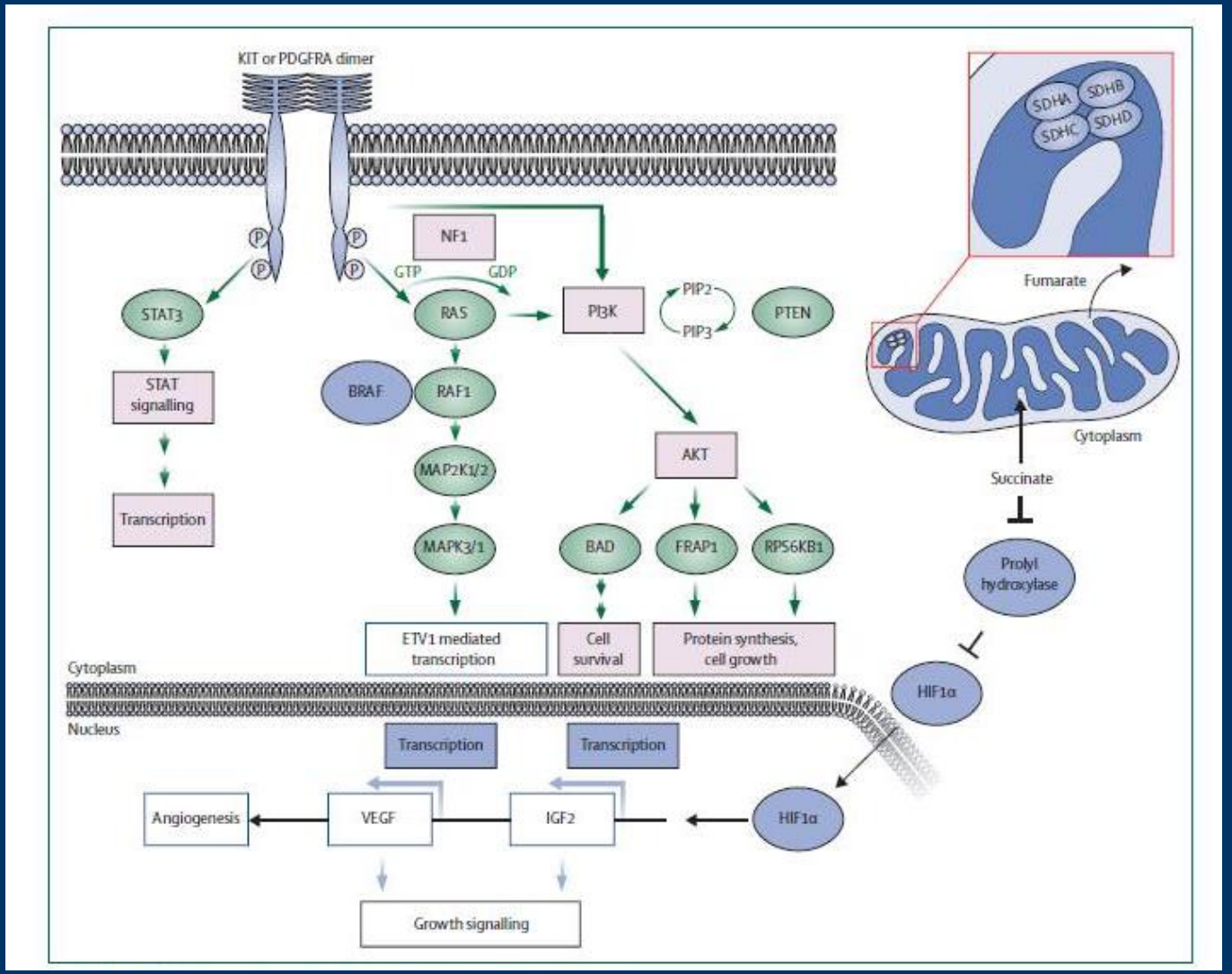
Gastrointestinal stromal tumors (GIST)



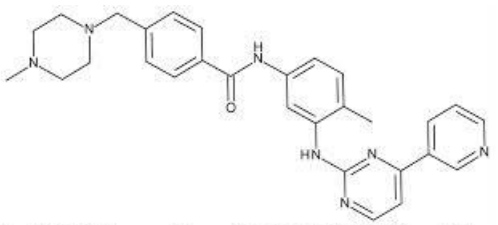




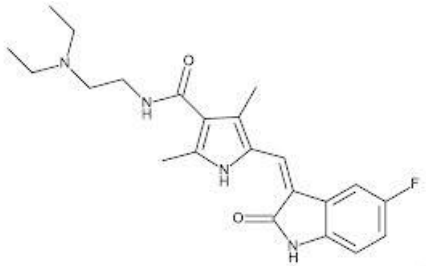




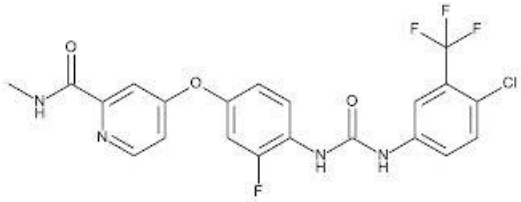
Joensuu H et al. Lancet 2013;382:973



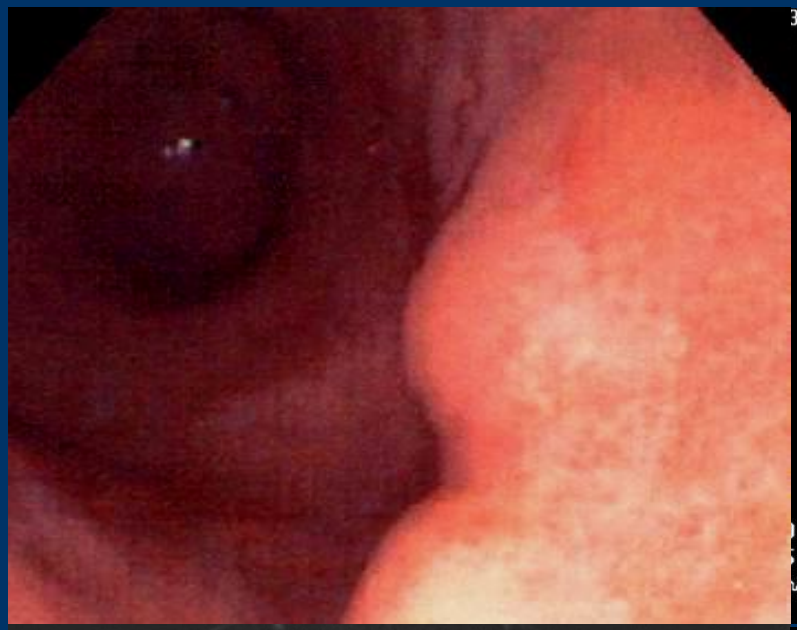
Imatinib (Gleevec, Novartis); $C_{29}H_{31}N_7O$; MW = 494

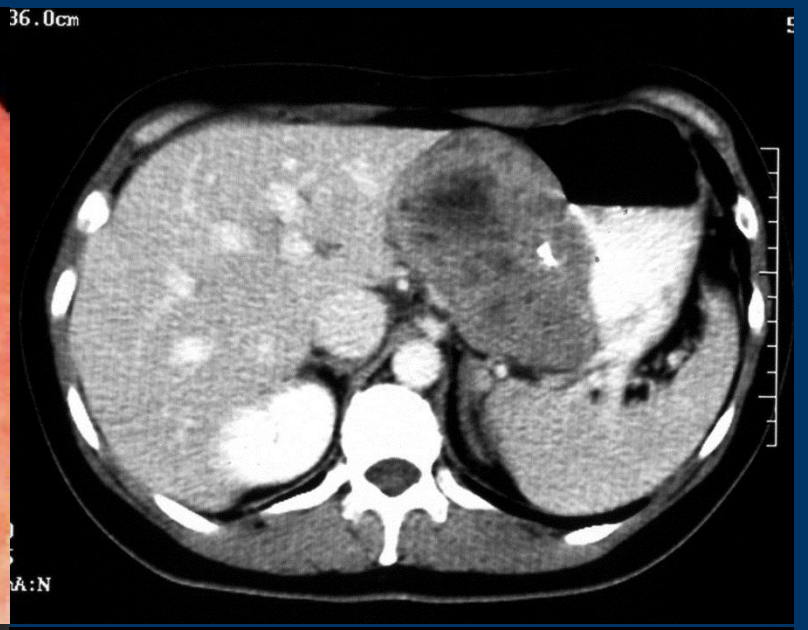


Sunitinib (Sutent, Pfizer); $C_{22}H_{27}FN_4O_2$; MW = 398



Regorafenib (Stivarga, Bayer); $C_{21}H_{15}ClF_4O_3$; MW = 482

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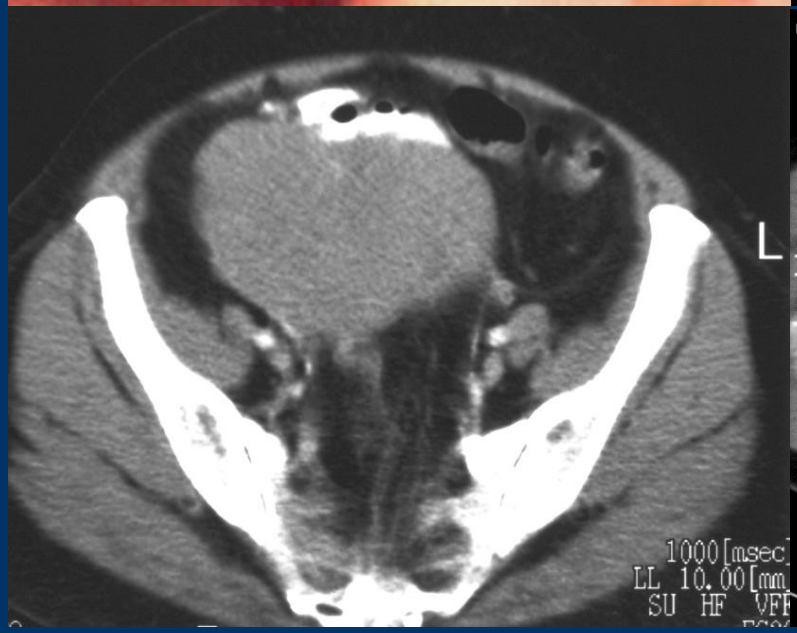
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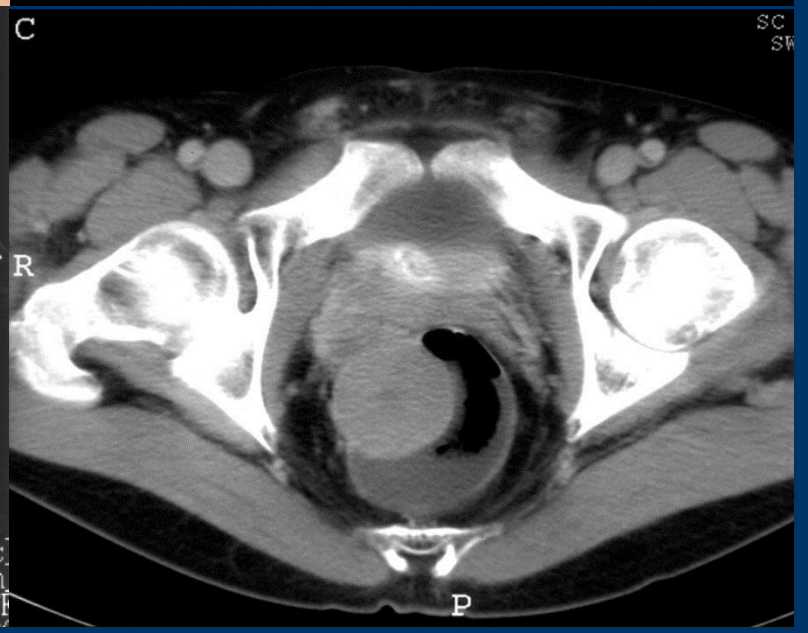
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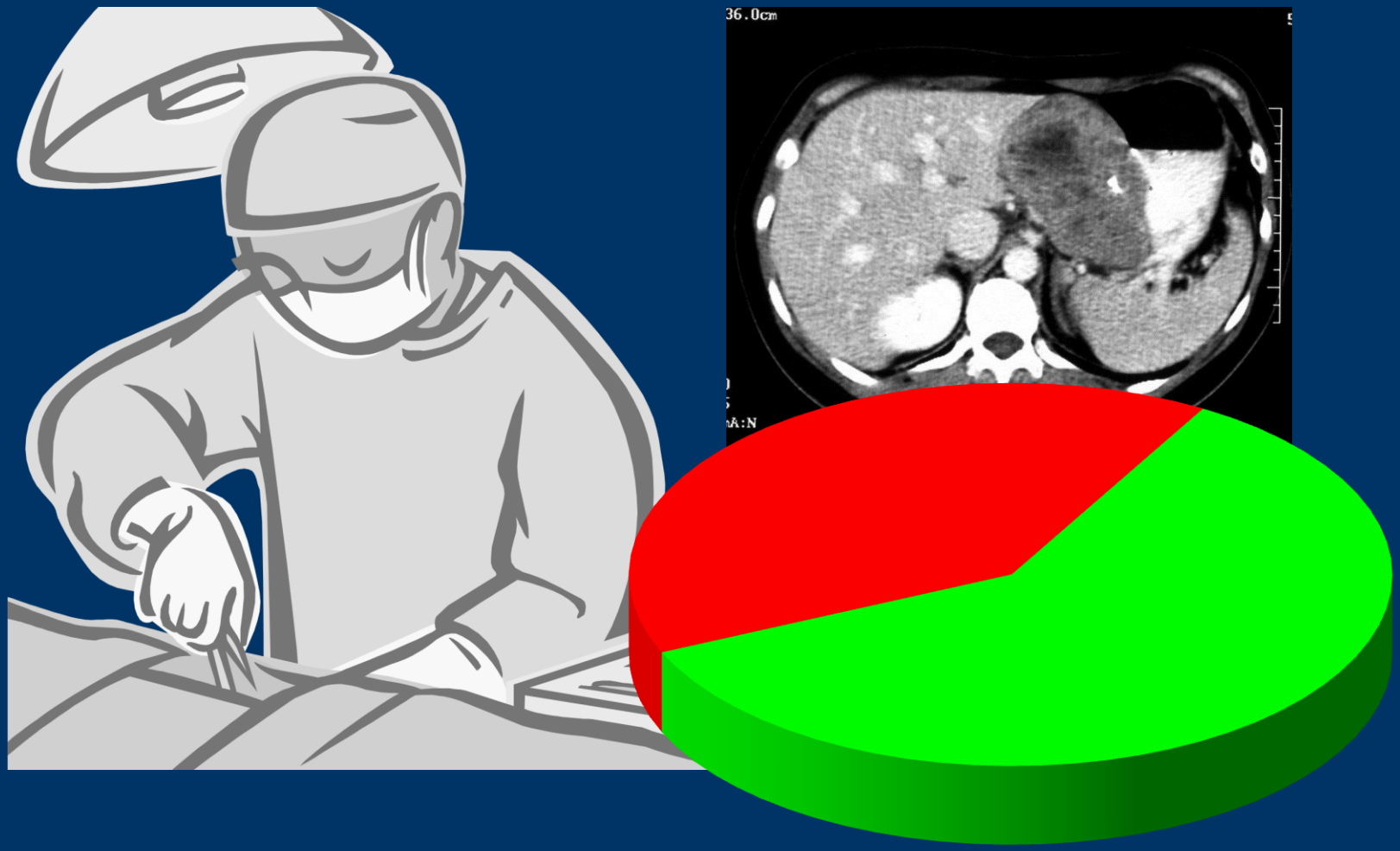
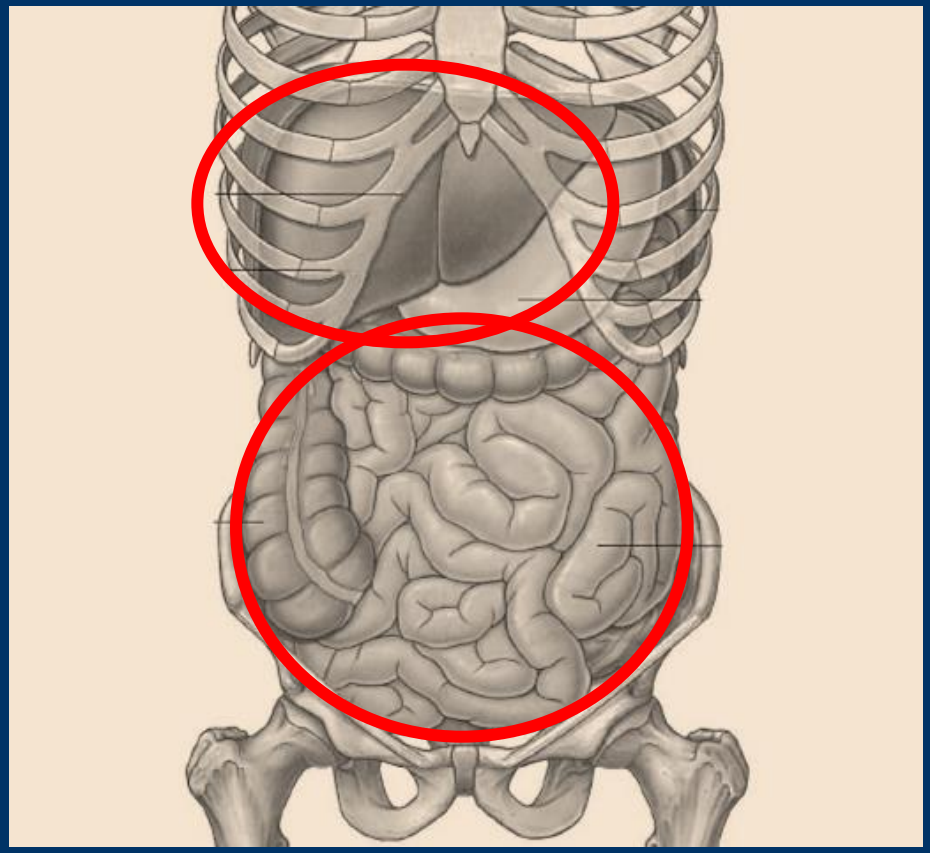
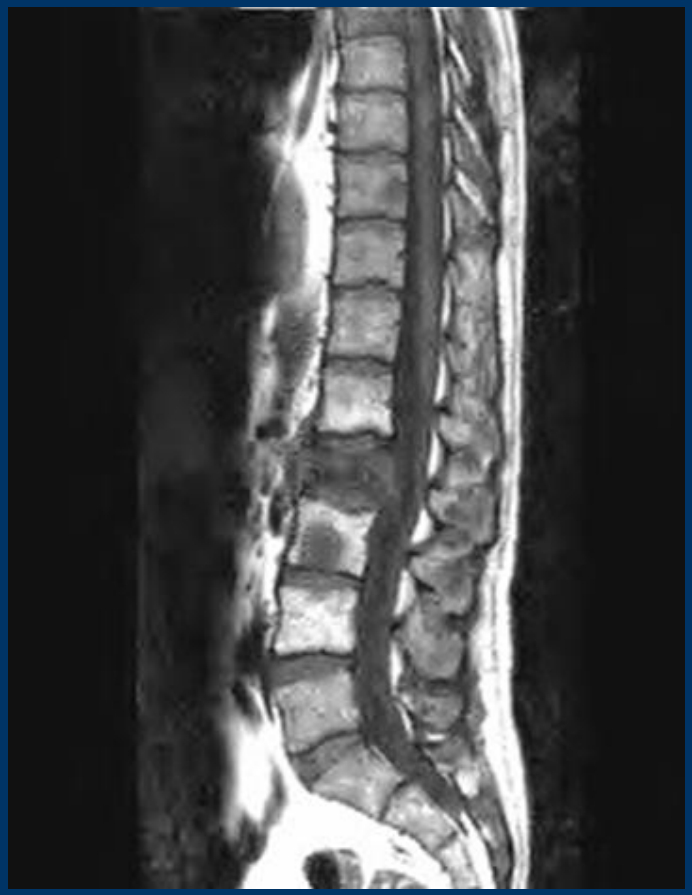
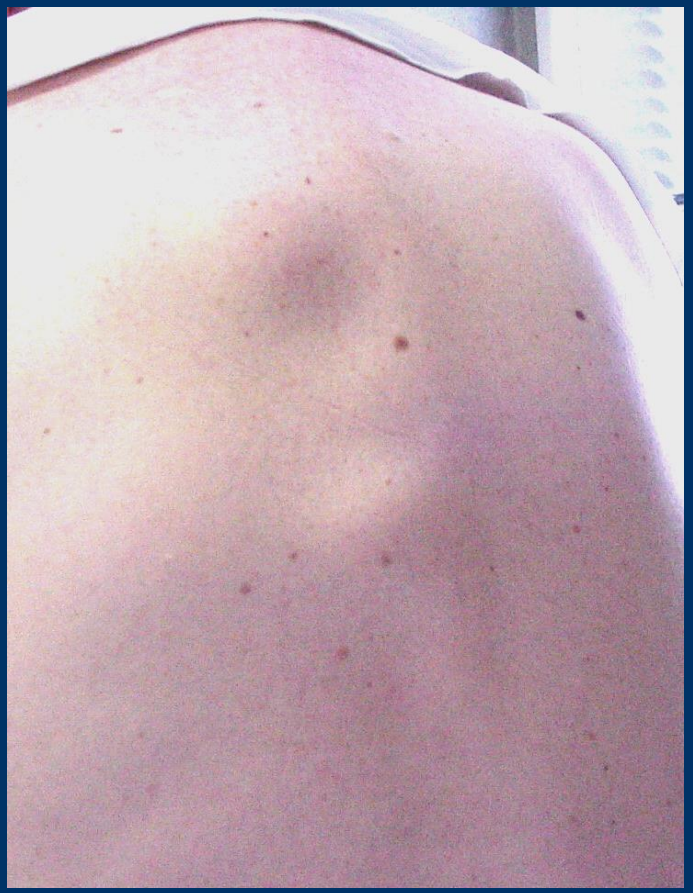
Standard treatment

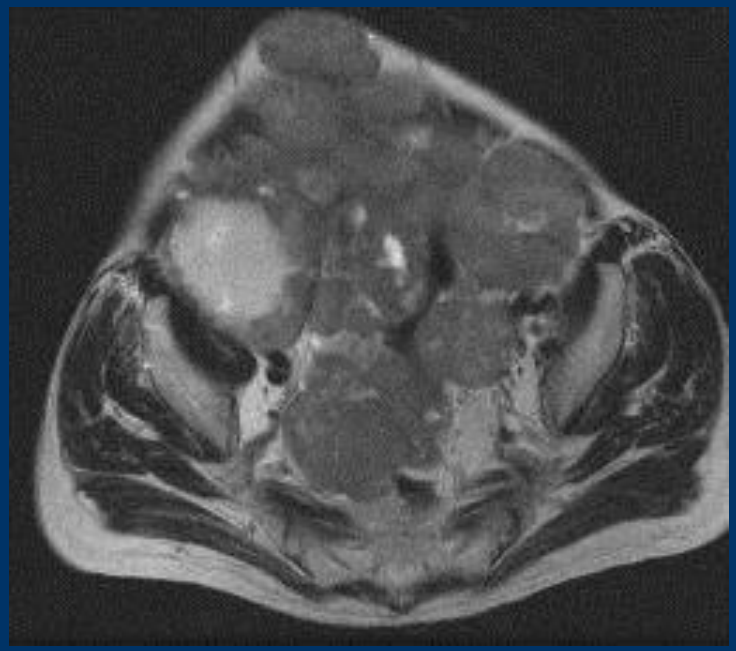


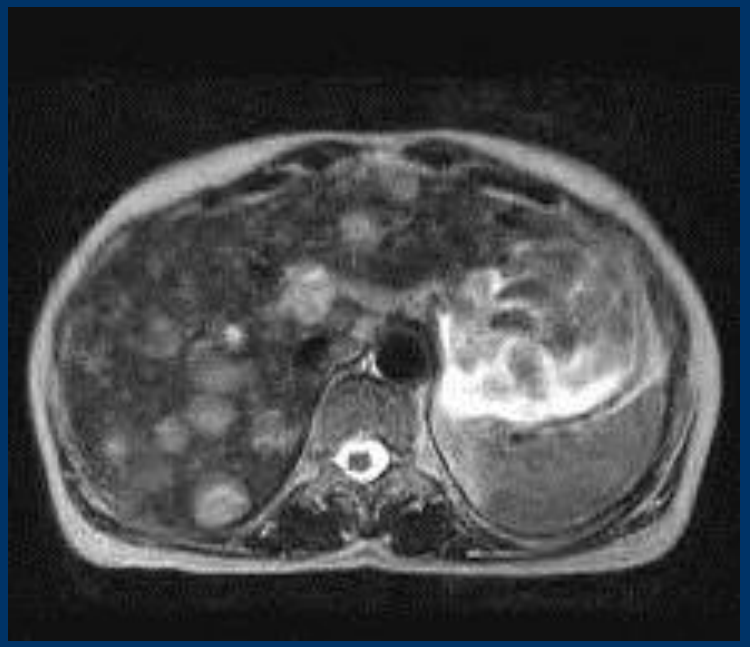
Relapse patterns











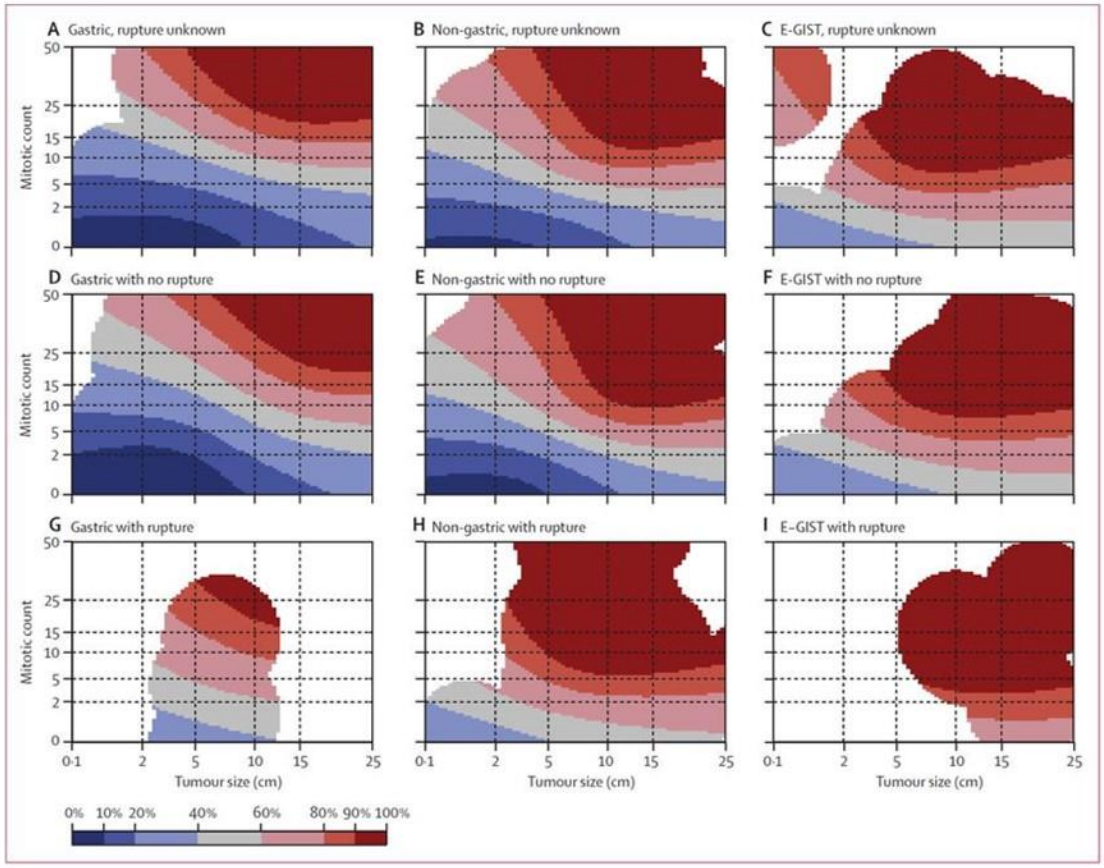
Risk stratification

	cm	M/50HPF	gastric	jejunal/ ileal	duodenal	rectal
1	≤2	≤5	0 none	0 none	0 none	0 none
2	>2≤5	≤5	1.9% very low	4.3% low	8.3% low	8.5% low
3a	>5≤10	≤5	3.6% low	24% moderate		
3b	>10	≤5	12% moderate	52% high	34% high	57% high
4	≤2	>5	0	50%		54% high
5	>2≤5	>5	16% moderate	73% high	50% high	52% high
6a	>5≤10	>5	55% high	85% high		
6b	>10	>5	86% high	90% high	86% high	71% high

Miettinen M. Semin Diagn Pathol 2006; 23: 70

Risk of recurrence of gastrointestinal stromal tumour after surgery: an analysis of pooled population-based cohorts

Heikki Joensuu, Aki Vehtari, Jaakko Riihimäki, Toshirou Nishida, Sonja E Steigen, Peter Brabec, Lukas Plank, Bengt Nilsson, Claudia Cirilli, Chiara Braconi, Andrea Bordoni, Magnus K Magnusson, Zdenek Linke, Jozef Sufliarsky, Massimo Federico, Jon G Jonasson, Angelo Paolo Dei Tos, Piotr Rutkowski



Lancet Oncol 2012;13:265

Assess the Risk: GIST

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Category: Medical
Released: May 05, 2013
Version: 1.0
Size: 1.9 MB
Language: English
Seller: Novartis Pharmaceutical Corporation

Rated 12+ for the following:
Infrequent/Mild Alcohol, Tobacco, or Drug Use or References

Compatibility: Requires iOS 6.0 or later. Compatible with iPad.

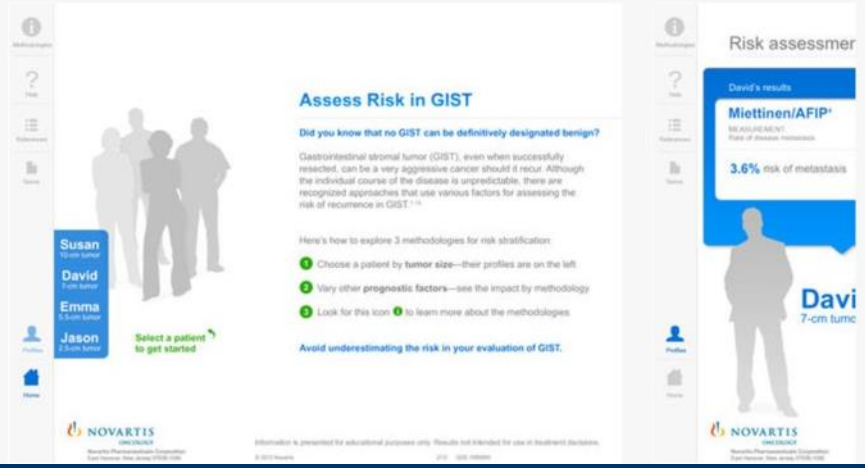
Description

An interactive activity designed for use on an iPad, this program estimates the risk of recurrence in gastrointestinal stromal tumor (GIST)—a GI cancer—using various assessment methodologies. The application features 4 hypothetical GIST patients and allows you to select input for key tumor-related prognostic factors. The recurrence

Assess the Risk: GIST Support ›

...More

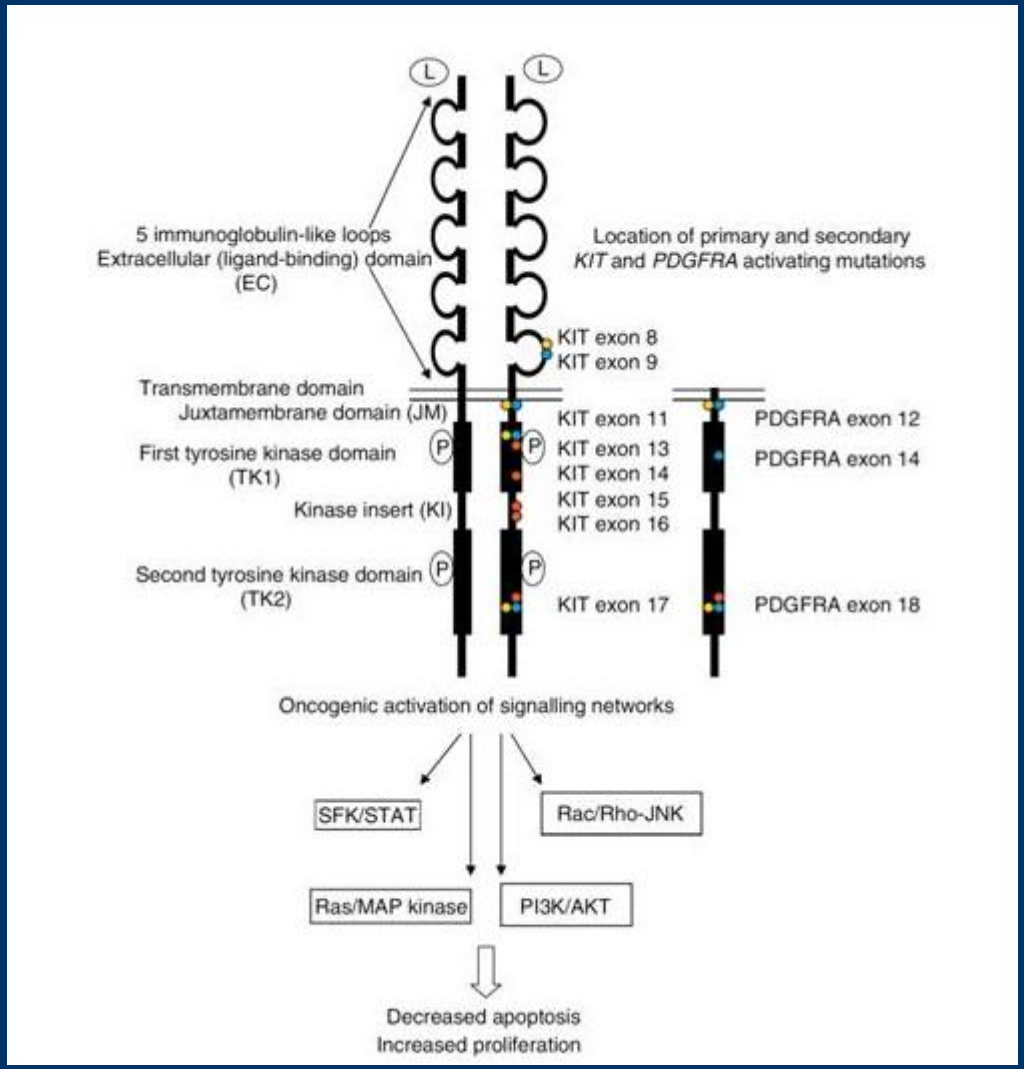
iPad Screenshots



KIT(~80%) **PDGFRA** (~10%) **WT** (~10%)

exon 9 (~10%)
exon 11 (~70%)
exon 13 (~5%)

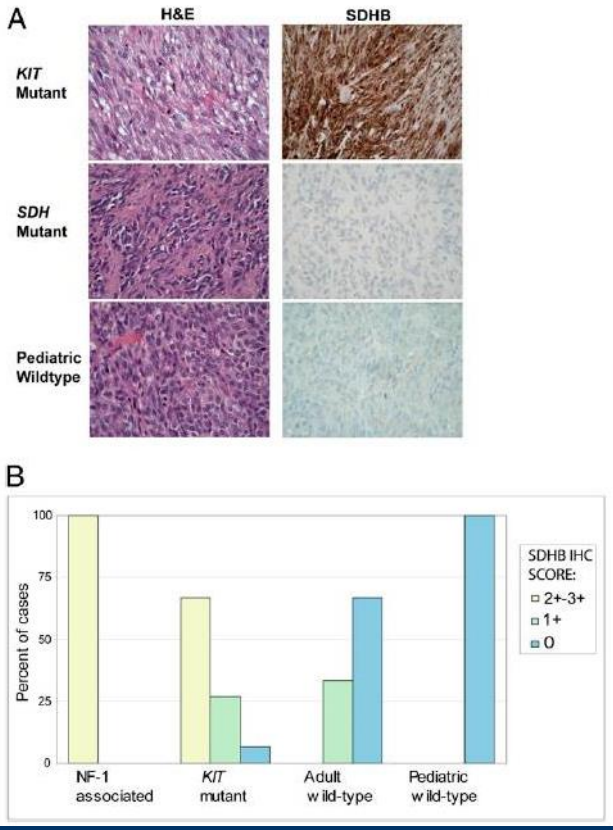
exon 17 (~5 %)



Lasota J et al, Histopathology 2008;53:245

Defects in succinate dehydrogenase in gastrointestinal stromal tumors lacking *KIT* and *PDGFRA* mutations

Katherine A. Janeway[a,1,2], Su Young Kim[b,1], Maya Lodish[c], Vânia Nosé[d], Pierre Rustin[e], José Gaal[f], Patricia L. M. Dahia[g], Bernadette Liegl[h], Evan R. Ball[c], Margarita Raygada[i], Angela H. Lai[a], Lorna Kelly[j], Jason L. Hornick[k], NIH Pediatric and Wild-Type GIST Clinic[l,m,n,o,p,3], Maureen O'Sullivan[j,q], Ronald R. de Krijger[f], Winand N. M. Dinjens[f], George D. Demetri[r], Cristina R. Antonescu[s], Jonathan A. Fletcher[k], Lee Helman[b], and Constantine A. Stratakis[c]



PNAS 2011;108:314

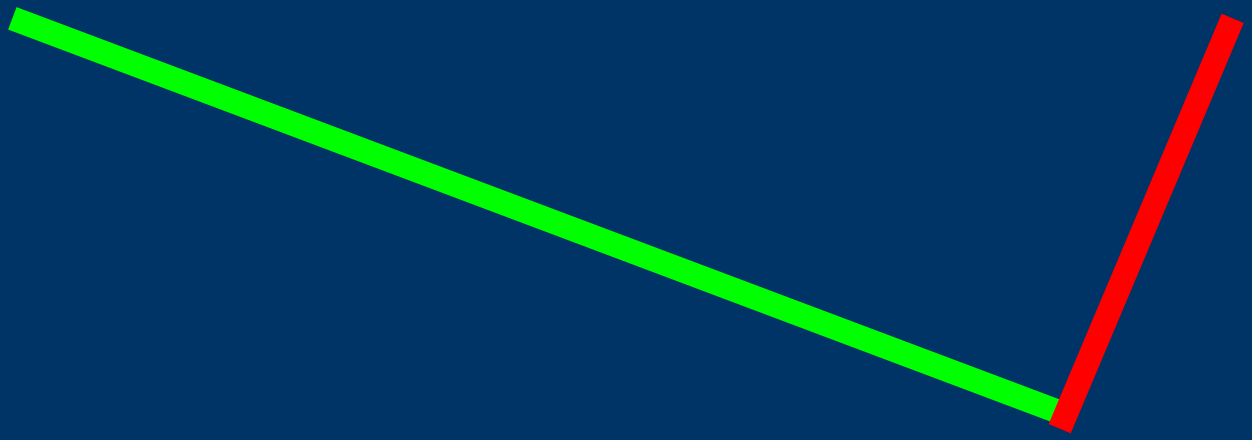
Neurofibromatosis, type 1

Series	No. patients	KIT mutation	PDGFRA mutation	NF1 mutation
Kinoshita, 2004	7	None	None	2 of 3 pts
Cheng, 2004	3	1 ex 11	None	NS
Anderson, 2005	12	None	None	NS
Takazawa, 2005	9	2 ex 11, 1 ex 13	1 ex 12, 1 ex 18	NS
Yantiss, 2005	3	1 ex 11	None	NS
Miettinen, 2006	15	None	None	NS
Maertens, 2006	3	2 polymorphisms	5 silents	3 of 3 pts
Nemoto, 2006	1	None	None	1 of 1 pts
Guillaud, 2006	1	None	None	NS
Lee, 2006	1	None	None	NS
Steward, 2007	2	None	None	1 of 2 pts
Kang, 2007	5	None	None	NS
Present series	25	1 ex 11, 1 ex 9	1 ex 18	NS

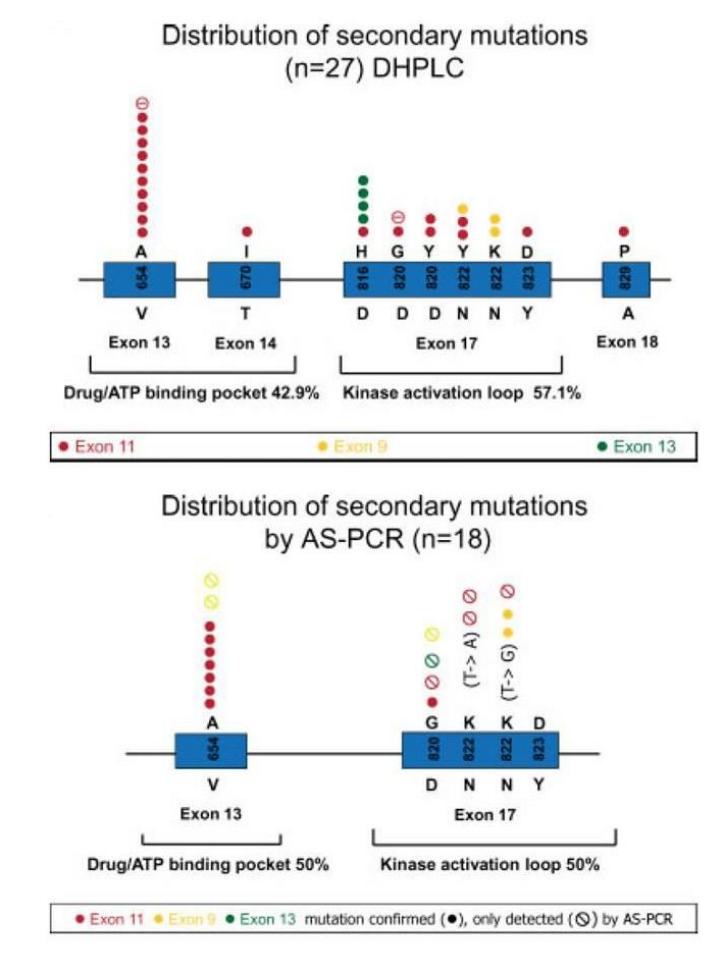
Mussi C, Clin Cancer Res 2008;14:4550

Primary resistance

Secondary resistance



Secondary resistance: molecular heterogeneity



Liegl B, J Pathol, 2008;216:64

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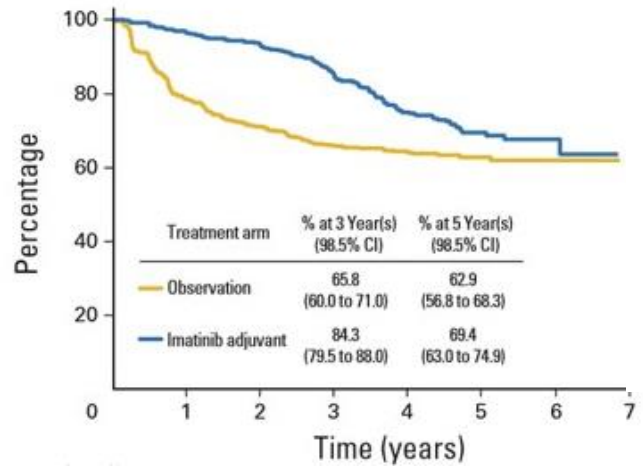
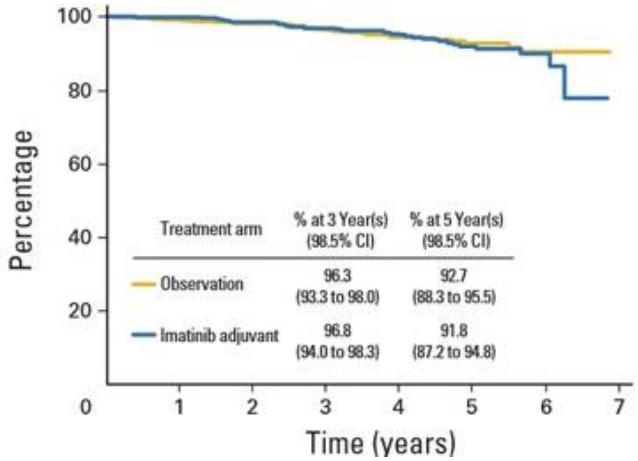


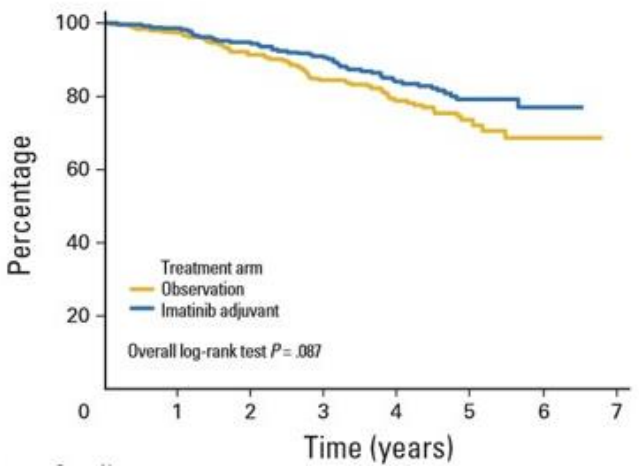
Time to Definitive Failure to the First Tyrosine Kinase Inhibitor in Localized GI Stromal Tumors Treated With Imatinib As an Adjuvant: A European Organisation for Research and Treatment of Cancer Soft Tissue and Bone Sarcoma Group Intergroup Randomized Trial in Collaboration With the Australasian Gastro-Intestinal Trials Group, UNICANCER, French Sarcoma Group, Italian Sarcoma Group, and Spanish Group for Research on Sarcomas

Paolo G. Casali, Axel Le Cesne, Andres Poveda Velasco, Dusan Kotasek, Piotr Rutkowski, Peter Hohenberger, Elena Fumagalli, Ian R. Judson, Antoine Italiano, Hans Gelderblom, Antoine Adenis, Jörg T. Hartmann, Florence Duffaud, David Goldstein, Javier M. Broto, Alessandro Gronchi, Angelo P. Dei Tos, Sandrine Marréaud, Winette T.A. van der Graaf, John R. Zalcberg, Saskia Litière, and Jean-Yves Blay

OS

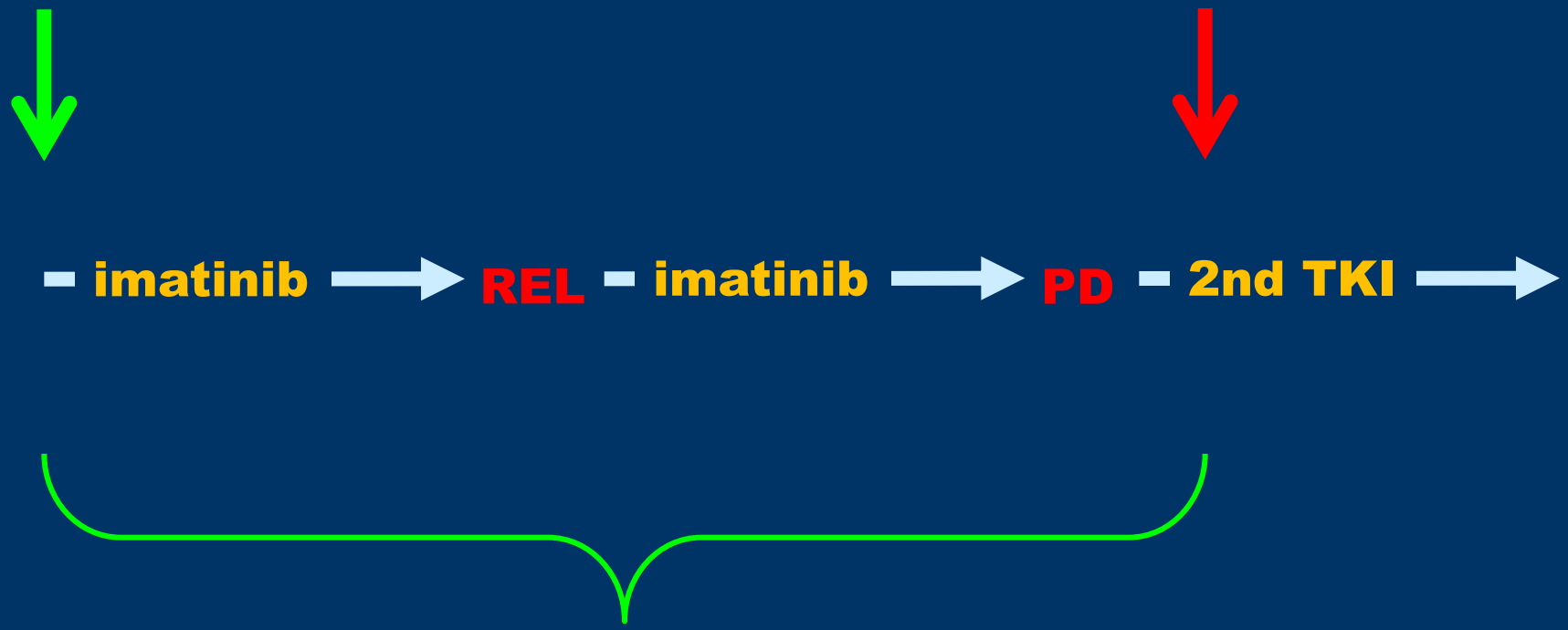


TTF



J Clin Oncol 2015 [Epub]

1st TKI failure-free survival



RARE CANCERS EUROPE

Joining forces for action

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