

**SARCOMA & GIST CONFERENCE 2016** 

# RETROPERITONEAL SARCOMA

Local Control

**Dirk Strauss** 

Royal Marsden Hospital, London, UK

ESMO Sarcoma & GIST events, Feb 2016

esmo.org

# **DISCLOSURE SLIDE**

None



# Conclusion

The current standard of care for retroperitoneal sarcoma:

• SURGERY

extended en bloc complete resection of the tumour and surrounding viscera (which may be adherent to but not necessarily invaded by the tumour)

 in an attempt to include a surrounding cuff of normal tissue around the tumour to minimise the marginality of the resection –



safely performed in high-volume centres.

#### But...

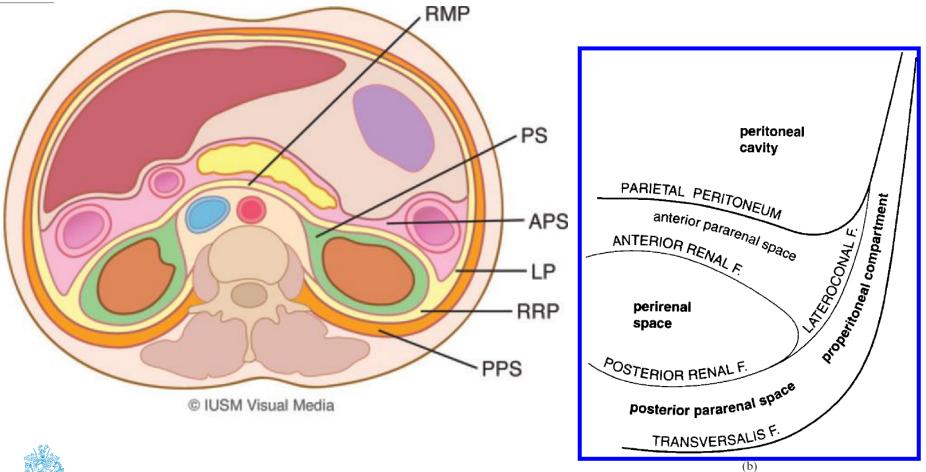
Retroperitoneal sarcomas is not a single disease!

- a group of heterogeneous neoplasms
- different places/organs/structures.

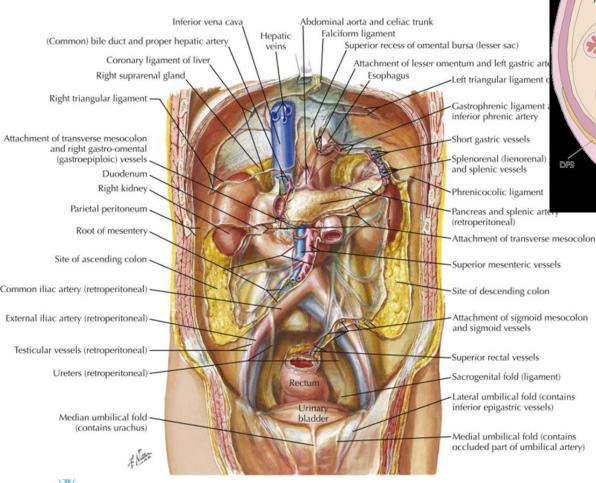
Biologic behaviour, response to treatment and clinical outcomes vary by histological subtype/grade.

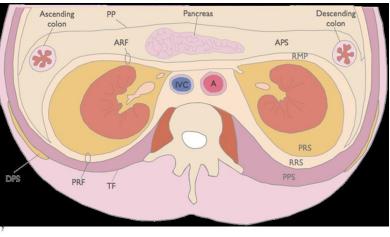
The management plan, including extent of resection and neoadjuvant strategies, should be formulated accordingly.







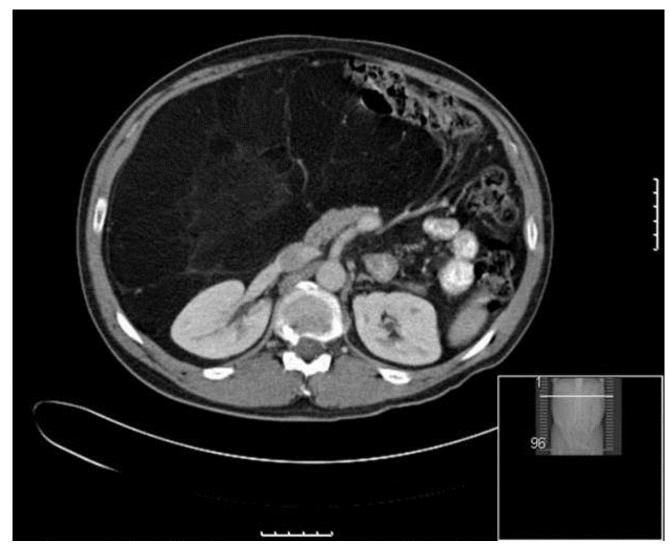


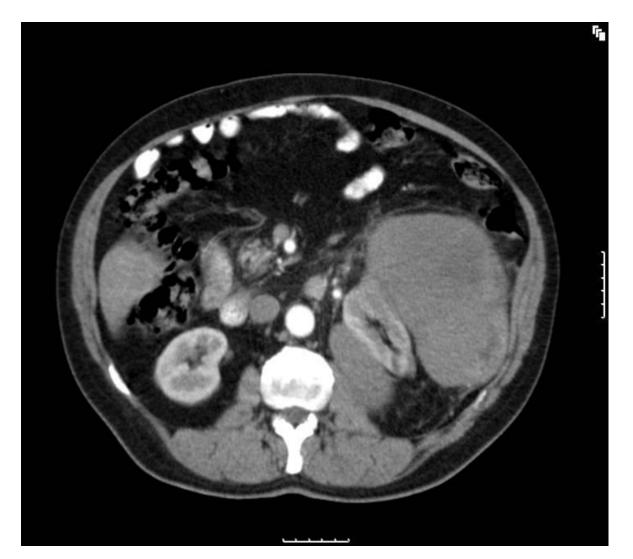


A = Aorta IVC = Inferior vena cava PP = Parietal peritoneum RMP = Retromesenteric plane RRS = Retrorenal space APS = Anterior pararenal space PRS = Perirenal space PPS = Posterior pararenal space TF = Transversalis fascia PRF = Retrorenal fascia ARF = Anterior renal fascia

## Retroperitoneal margins

RETROPERITONEAL COMPARTMENT MARGIN	
ANTERIOR	– posterior parietal peritoneum, ipsilateral colon and mesocolon, tail of pancreas + spleen or head of pancreas, duodenum
POSTERIOR	– psoas, iliacus, quadratus lumborum, transverses abdominis, muscles, diaphragm
MEDIAL	<ul> <li>IVC (right-sided tumours), duodenum, head of pancreas.</li> <li>Aorta + branches (left-sided tumours), spine, paraspinous muscles</li> </ul>
LATERAL	– lateral abdominal musculature, kidney and colon (depending on tumour location)
SUPERIOR	<ul> <li>transverse colon/mesocolon, tail of pancreas or spleen</li> <li>diaphragm, the right lobe of the liver, the duodenum</li> </ul>
INFERIOR	<ul><li>– iliopsoas muscle</li><li>femoral nerve, iliac vessels or pelvic sidewall, bladder/rectum</li></ul>

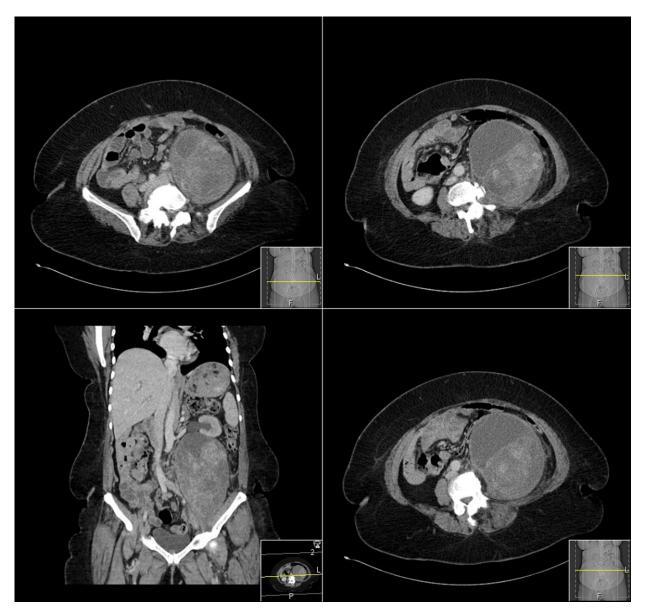








#### Psoas/posterior abdominal wall sarcoma





## Subtypes

#### **Primary retroperitoneal sarcoma**

- Liposarcomas (well-diff, dedifferentiated)
- Leiomyosarcoma
- Solitary fibrous tumours
- Malignant PNST
- Synovial sarcoma, Ewing's sarcoma, Pleomorphic sarcoma,
   Fibrosarcoma, Spindle cell sarcoma NOS, Pleomorphic
   rhabdomyosarcoma, Undifferentiated round cell sarcoma,
   Epithelioid smooth muscle tumour, Desmoplastic small round
   cell tumour



#### Original Article

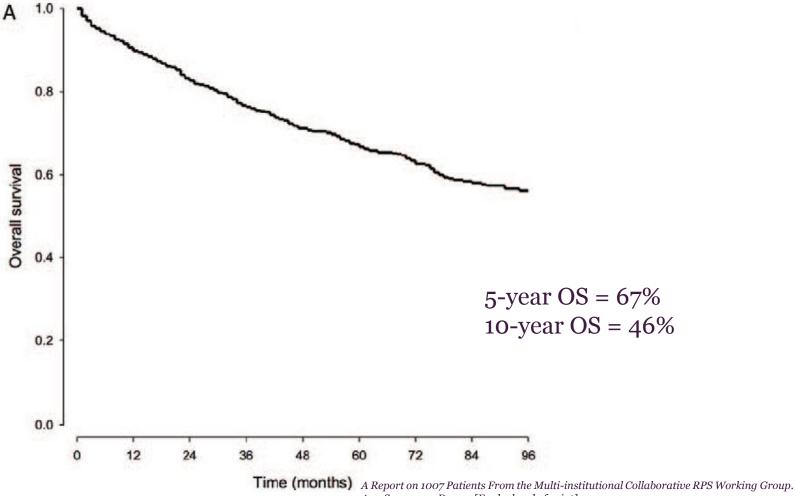
#### Variability in Patterns of Recurrence After Resection of Primary Retroperitoneal Sarcoma (RPS)

#### A Report on 1007 Patients From the Multi-institutional Collaborative RPS Working Group

Alessandro Gronchi, MD,\* Dirk C. Strauss, MD,† Rosalba Miceli, MD, PhD,‡ Sylvie Bonvalot, MD, PhD,§
Carol J. Swallow, MD,¶ Peter Hohenberger, MD,|| Frits Van Coevorden, MD,\*\* Piotr Rutkowsky, MD,††
Dario Callegaro, MD,\* Andrew J. Hayes, MD, PhD,† Charles Honoré, MD,§ Mark Fairweather, MD,‡‡
Amanda Cannell, MD,¶ Jens Jakob, MD,|| Rick L. Haas, MD,§§ Milena Szacht, MD,†† Marco Fiore, MD,\*
Paolo G. Casali, MD,¶¶ Raphael E. Pollock, MD, PhD,|||| and Chandrajit P. Raut, MD‡‡

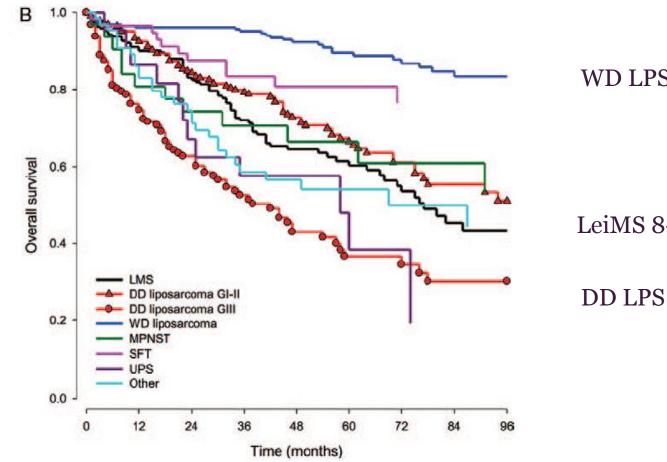


#### Overall survival curve in the whole series



Ann Surg. 2015 Dec 31. [Epub ahead of print]

#### Overall survival curve in the whole series



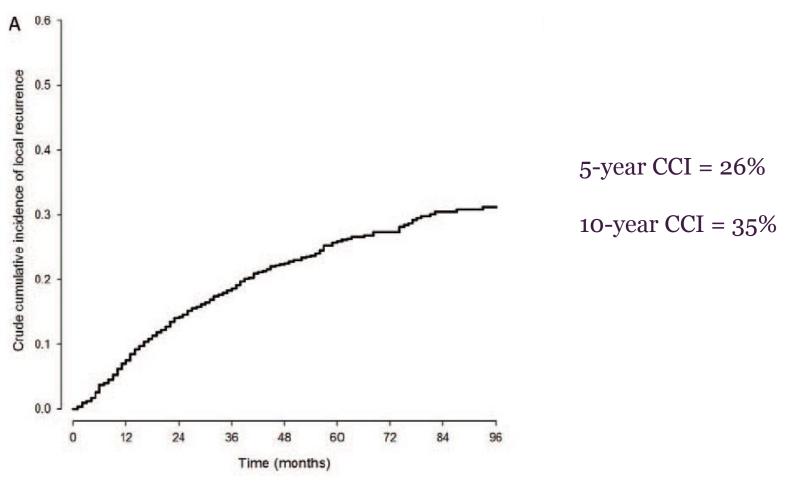
WD LPS 8-yr OS ~ 80%

LeiMS 8-yr OS  $\sim 40\%$ 

DD LPS Gr<br/>3 $8\mathchar`-yr$  OS ~ 30%

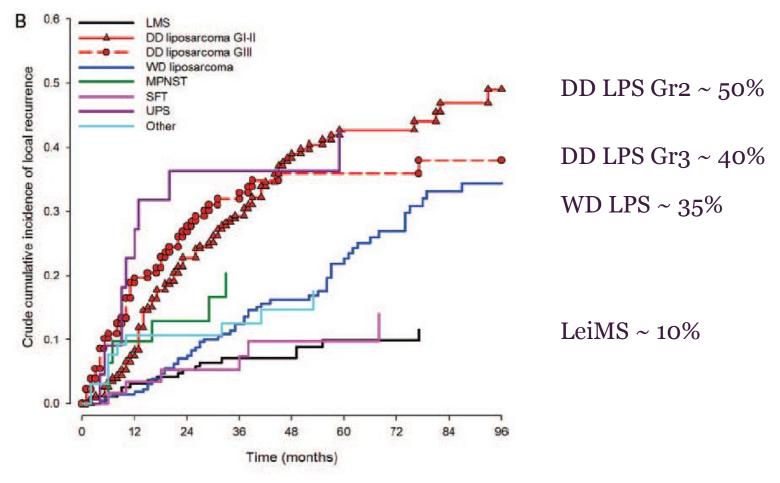
A Report on 1007 Patients From the Multi-institutional Collaborative RPS Working Group. Ann Surg. 2015 Dec 31. [Epub ahead of print]

#### Cumulative incidence of local relapse



A Report on 1007 Patients From the Multi-institutional Collaborative RPS Working Group. Ann Surg. 2015 Dec 31. [Epub ahead of print]

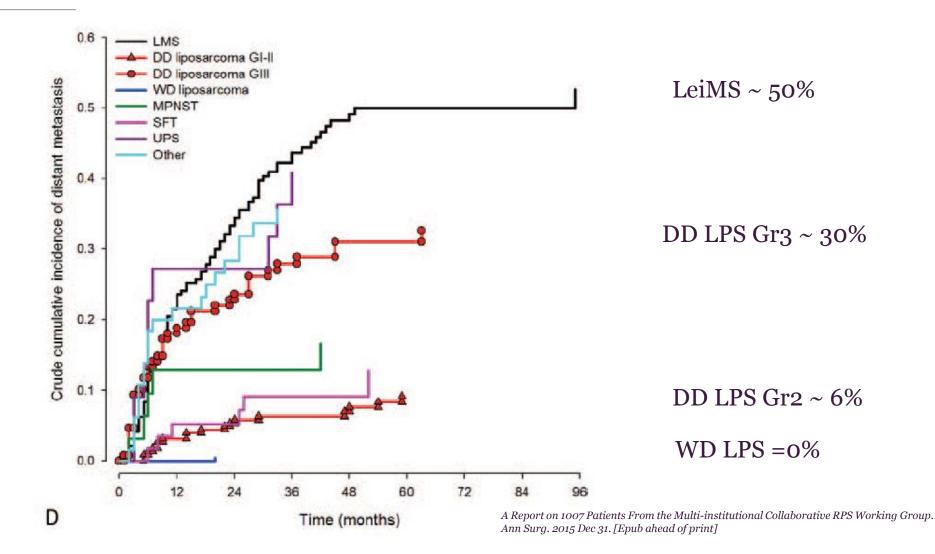
#### Cumulative incidence of local relapse



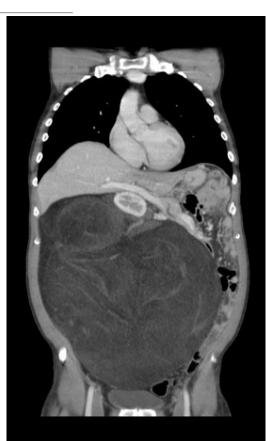


A Report on 1007 Patients From the Multi-institutional Collaborative RPS Working Group. Ann Surg. 2015 Dec 31. [Epub ahead of print]

#### Cumulative incidence of distant metastasis

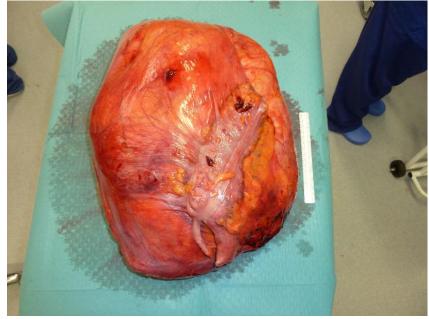


#### Surgical extent according to biologic behaviour



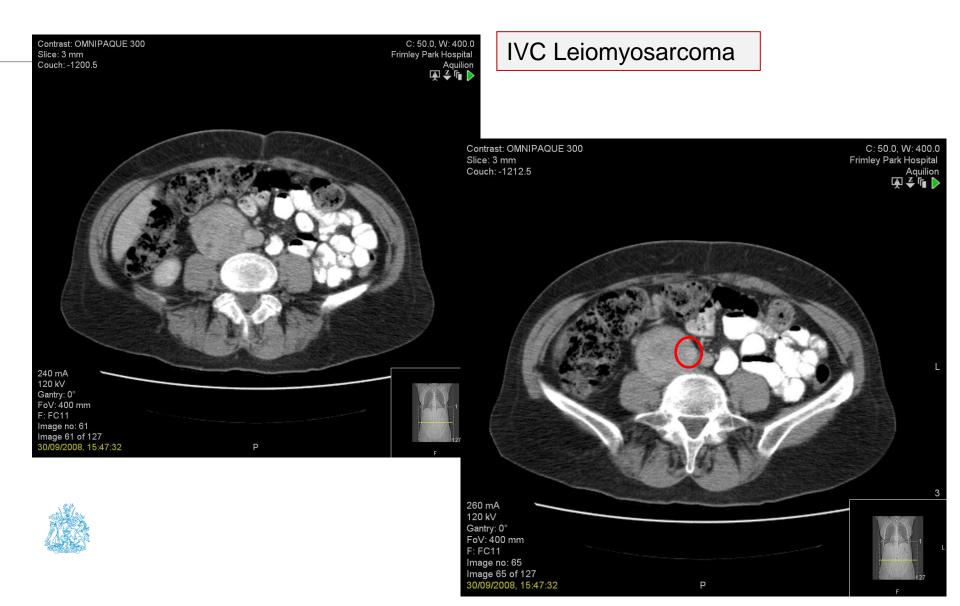
Well-differentiated Liposarcoma



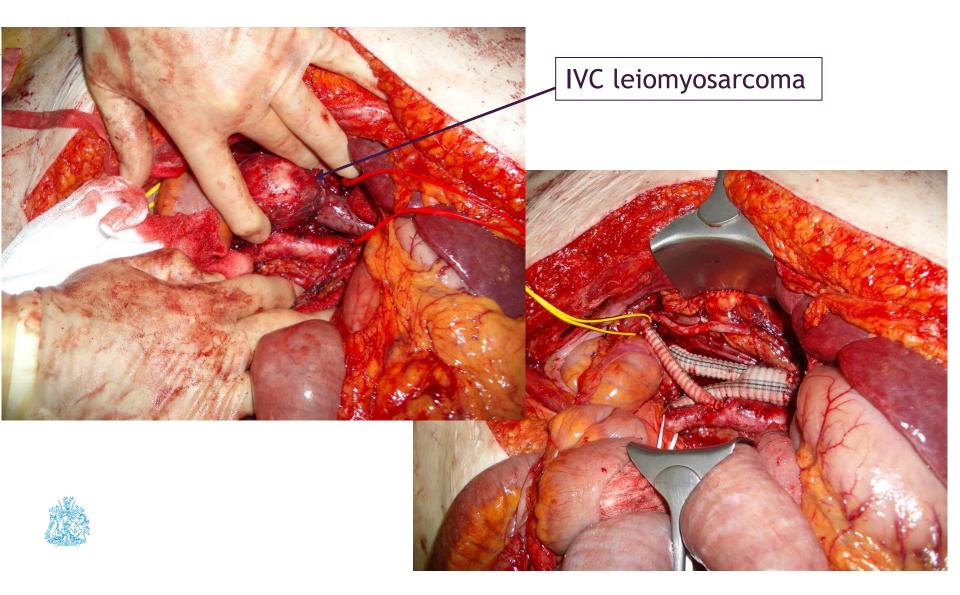




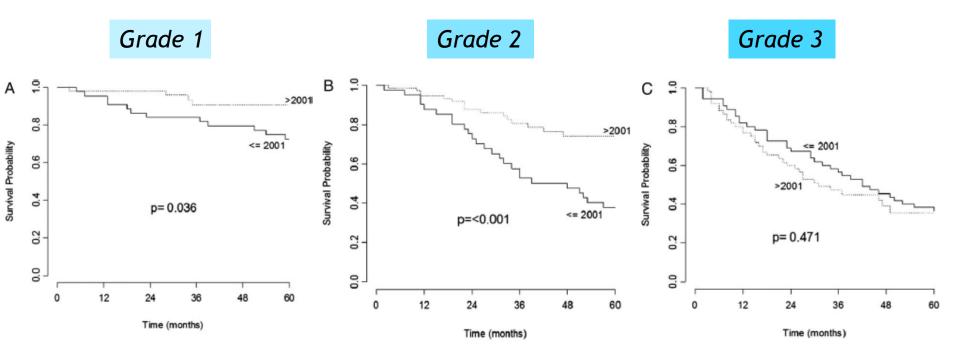
## Surgical extent according to biologic behaviour



#### Surgical extent according to biologic behaviour



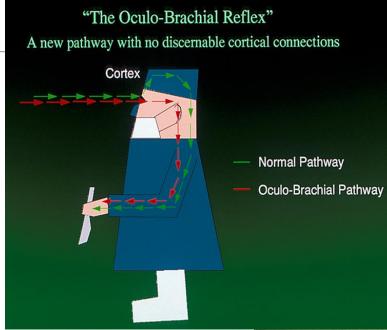
#### Frontline extended surgery



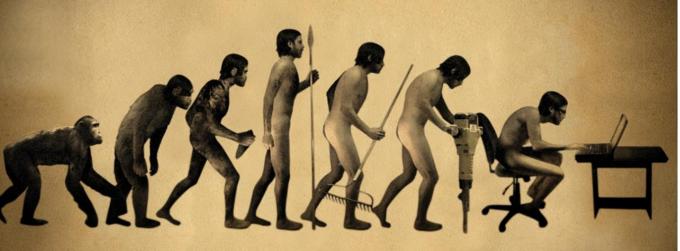
Gronchi A et al, Frontline extended surgery is associated with improved survival in retroperitoneal low- to intermediate-grade soft tissue sarcomas. Ann Oncol. 2011 Jul 16. [Epub ahead of print]

The Royal Marsden

# Initial approach to RP mass



- 1. Diagnosis?
- 2. Should it be taken out?
- 3. Should I be taking it out?
- 4. What will it involve taking it out?
- 5. Is it possible/beneficial/risks to resect?





# **Retroperitoneal mass**

Biopsy not essential	<ul> <li>imaging reviewed in Sarcoma MDT and is diagnostic and absolutely certain of a resectable retroperitoneal liposarcoma</li> <li>and no neoadjuvant treatment planned</li> </ul>
Biopsy	<ul> <li>lipomatous lesion when radiological uncertainty</li> <li>include non-lipomatous soft tissue sarcomas, GIST, benign soft tissue tumours, suspected lymphomas, germ-cell tumours or primary/metastatic carcinoma.</li> </ul>



#### <u>Advantage</u>

1. Accurate

Differentiate malignant and benign STT (n = 530)

- -Sensitivity = 96.3%
- -Specificity = 99.4%
- –Positive predictive value = 99.5%
- –Negative predictive value = 95.1%

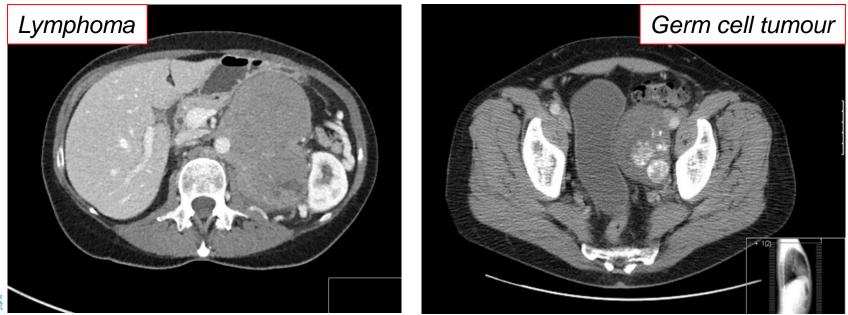
#### **Accuracy = 98%**





#### <u>Advantage</u>

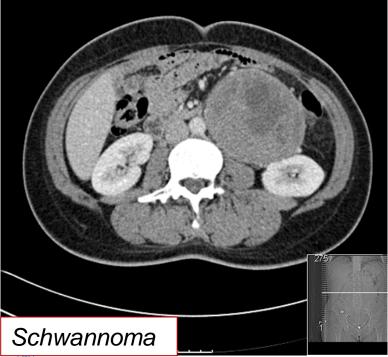
2. To identify the non-STS malignancies (lymphoma, metastatic carcinoma, germ cell tumours)





#### <u>Advantage</u>

#### 3. To differentiate benign from malignant







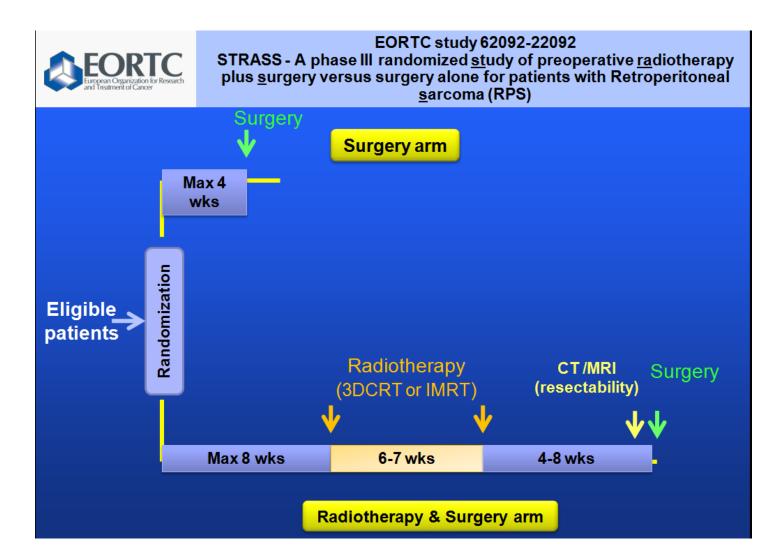
#### Advantage

4. To subtype and grade soft tissue sarcomas

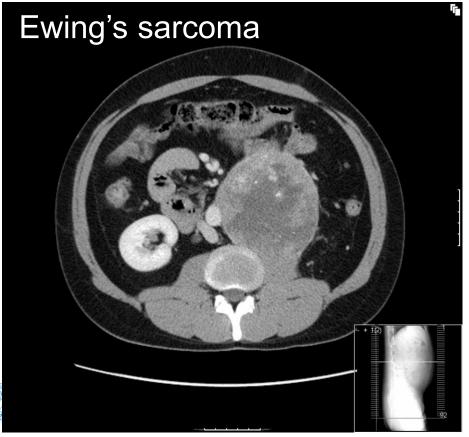
Preoperative biopsy allows to plan the optimal surgical + neoadjuvant treatment for each patient and to tailor treatment according to the individual sarcoma subtype



#### Prognostic factors predicting outcome

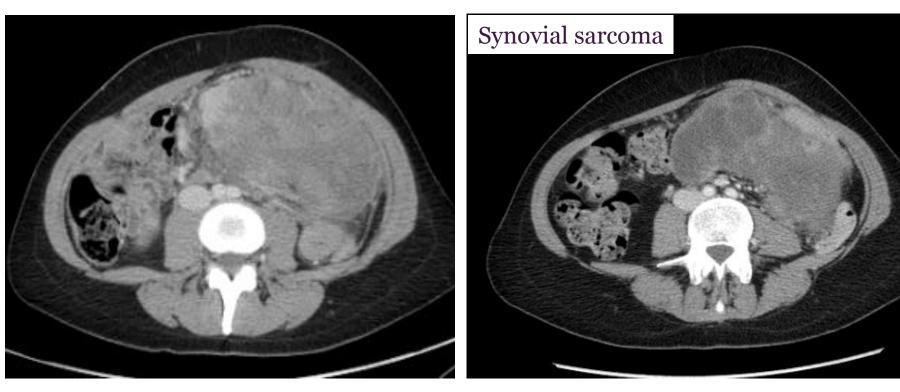


#### <u>Advantage</u> To identify chemo-sensitive sarcoma subtypes



- Core needle biopsy:
- Extraskeletal Ewing's sarcoma
- Preoperative chemotherapy (vincristine,doxorubicin, cyclophosphamide, ifosfamide, and etoposide )
- Resection specimen:
- marked treatment-related changes and <10% viable tumour.

### Chemo-sensitive subtypes



Aug 2012

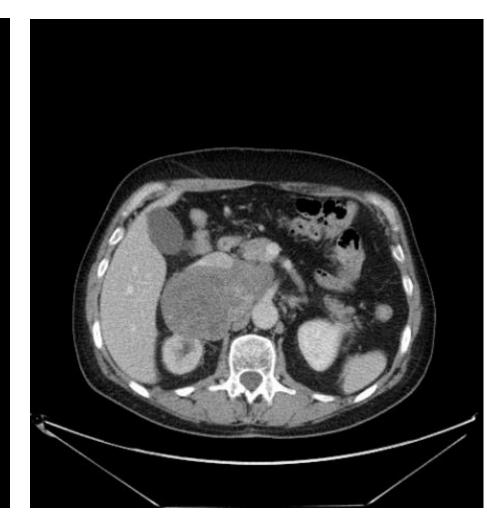


Nov 2012 – post chemotherapy (4 cycles ifosfamide and doxorubicin)

#### Chemo-sensitive subtypes

#### Leiomyosarcoma 04/2013 – 11/2013 6 cycles gemcitabine

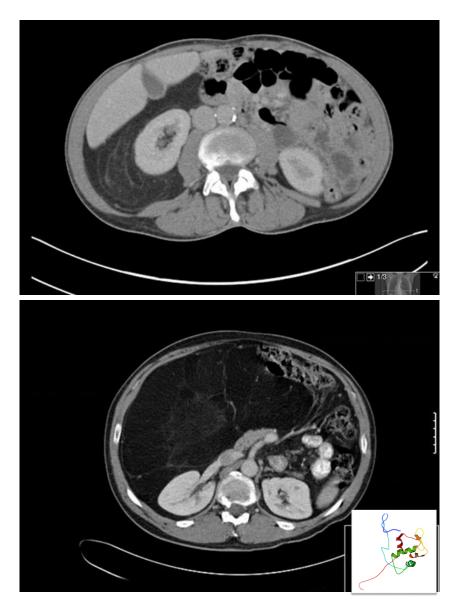


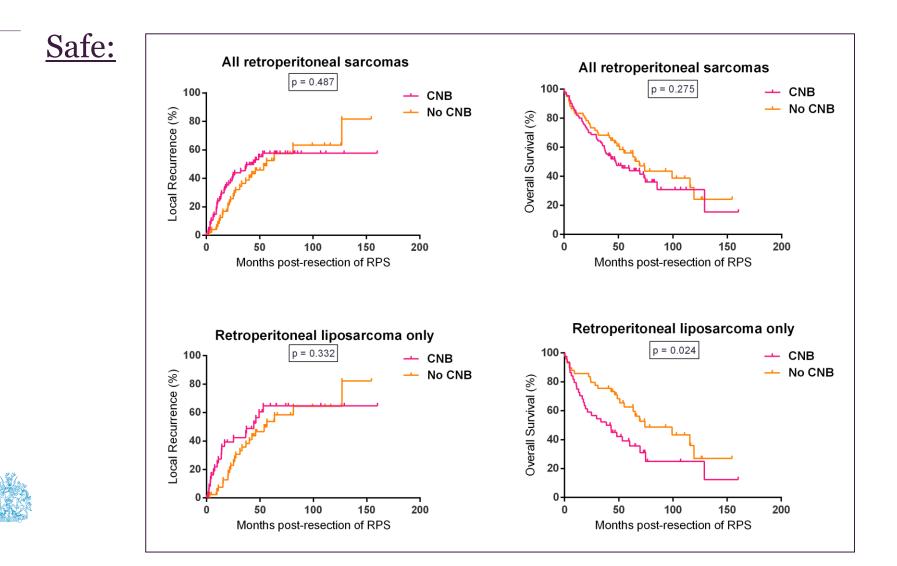


#### Role of core needle biopsy - lipomatous lesions









#### Resectability

Is it beneficial/oncologically sensible to resect the tumour?

Patient benefit v treatment morbidity

"In the field of surgical oncology: **tumour biology** is king, **patient selection** is queen,



and technical manoeuvres are the prince and princess.

Occasionally the prince and princess try to overthrow the powerful forces of the King and Queen, sometimes with temporary apparent victories, usually to no long term avail."



#### Resectability

Is it beneficial/oncologically sensible to resect the tumour?

- Systemic metastasis
- Incomplete resection of high-grade tumour
- Tumour rupture
- Aggressive tumour biology



#### **Encasement of vessels** \_



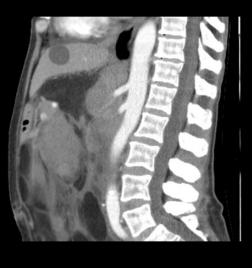


Contrast: 100mls Xenetix 300 Body 5.0 Arterial/Abdo CE Slice: 5 mm Couch: -80 BHRUT NHS TRUST Aquilion 194 mA 120 kV Gantry: 0° FoV: 356.25 mm F: FC13 1/3 Image no: 17 Image 17 of 50 05/12/2012, 09:42:57

Contrast: 100mls Xenetix 300 Body 5.0 Arterial/Abdo/Sagittal CE Slice: 5 mm Couch: -1.9

C: 40.0, W: 400.0 BHRUT NHS TRUST Aquilion

C: 40.0, W: 400.0

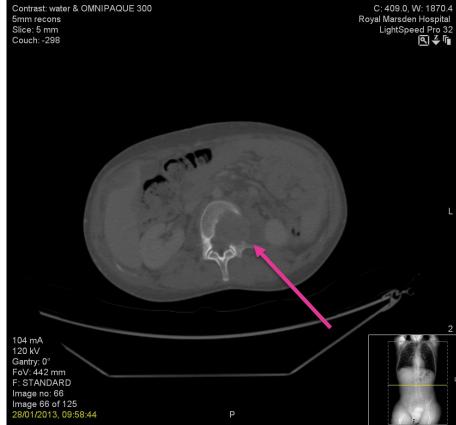


1/3

100 mA 120 kV Gantry: 0° FoV: 356.25 mm F: FC03 Image no: 33 mage 32 of 64 12/2012. 09:42:57

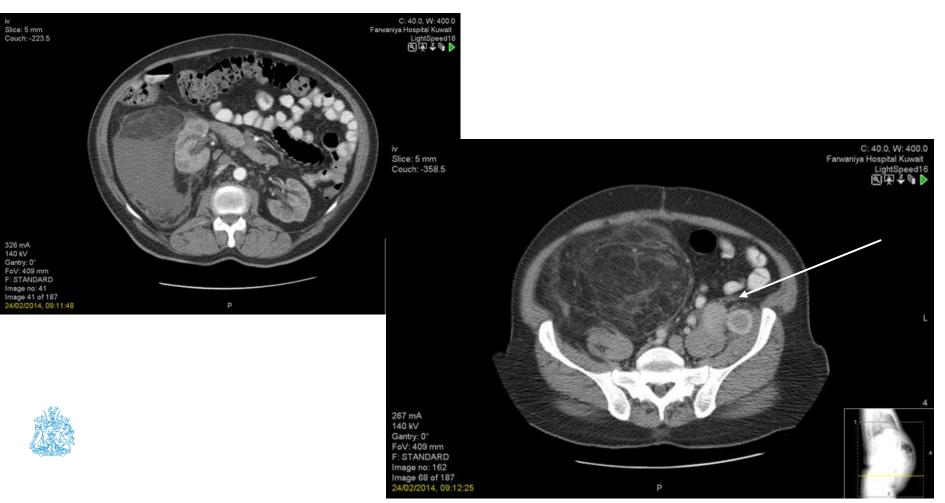
#### Spinal involvement



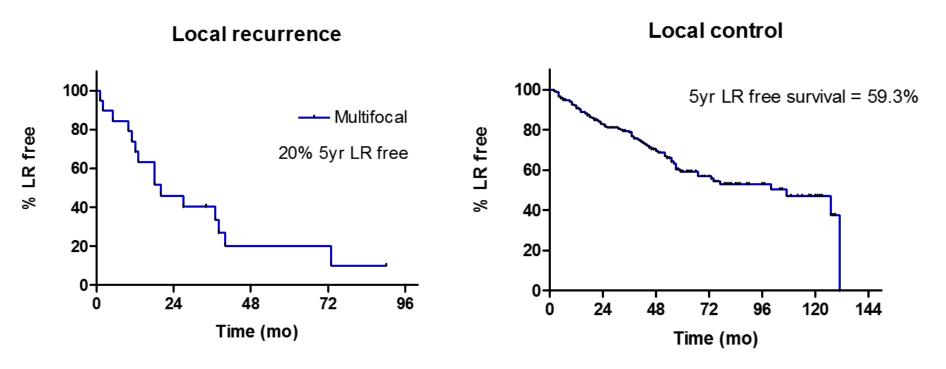




#### Multifocality



#### Multifocality





## Surgical principle of resection

Liberal visceral en bloc resection in an attempt to

include an envelope of normal tissue around the tumour

to minimise the marginality of the resection - in the

hope of improving outcome.



#### Compartmental resection

The objective is to achieve an envelope of normal tissue along some tumour surfaces by removing adjacent, easily disposable organs

while performing what is essentially a marginal excision along critical structures.



## **Compartmental resection**



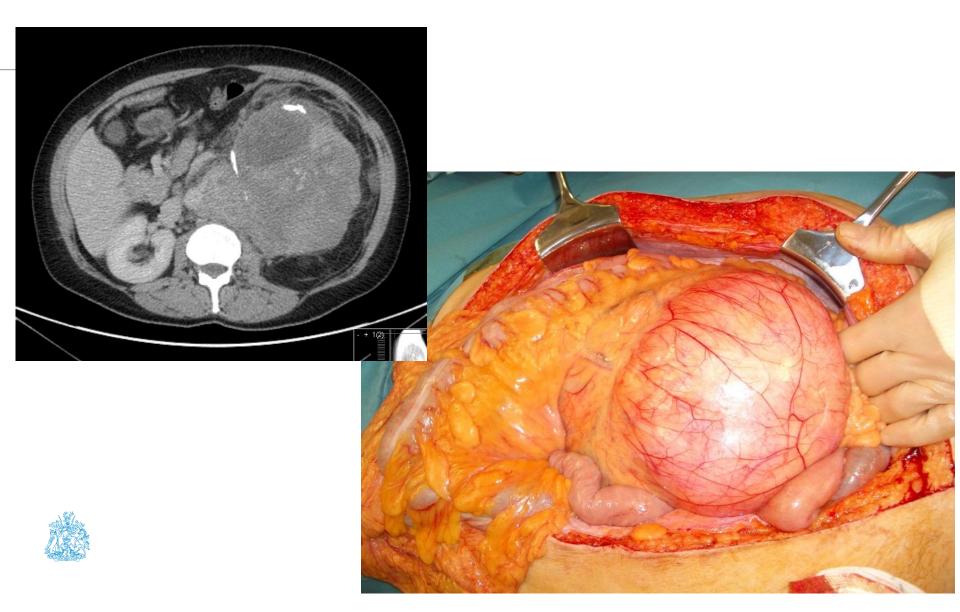


## **Compartmental resection**

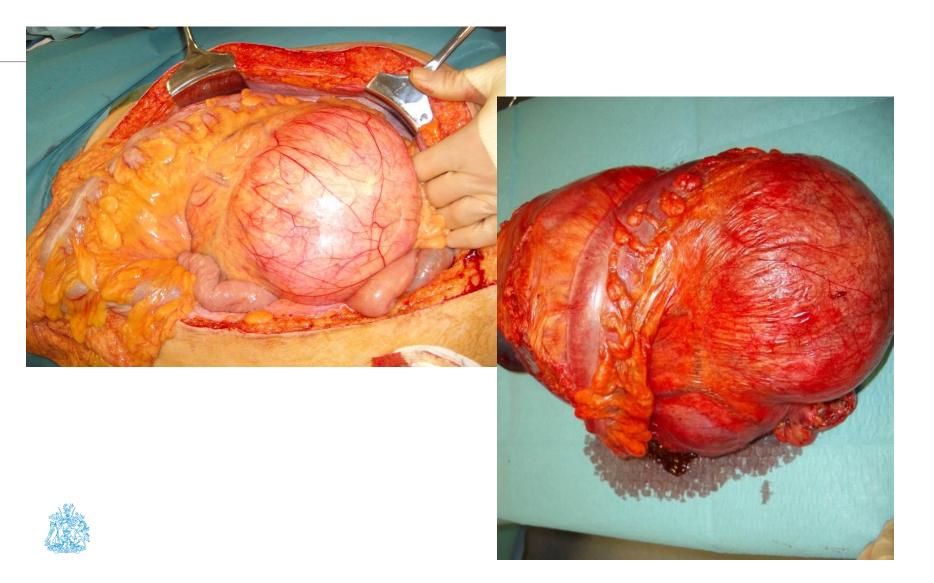




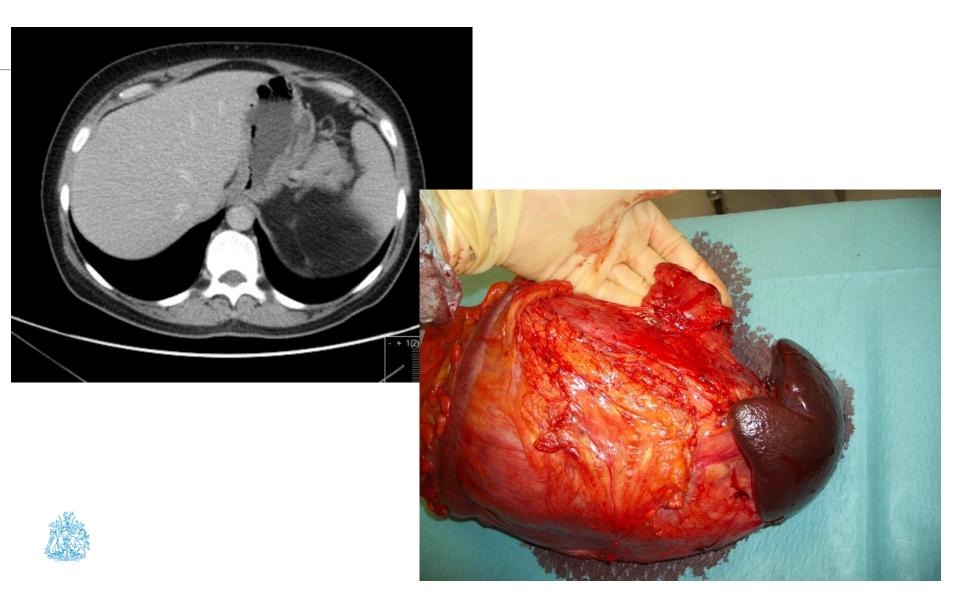
# Anterior colon/mesentery



# Anterior colon/mesentery

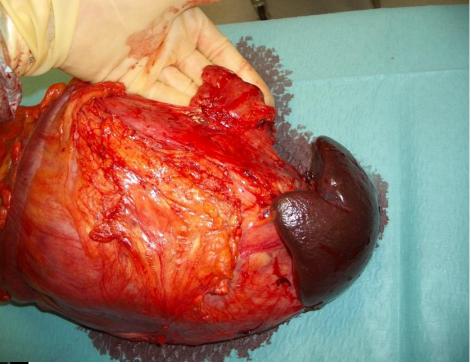


# Superior margin -spleen

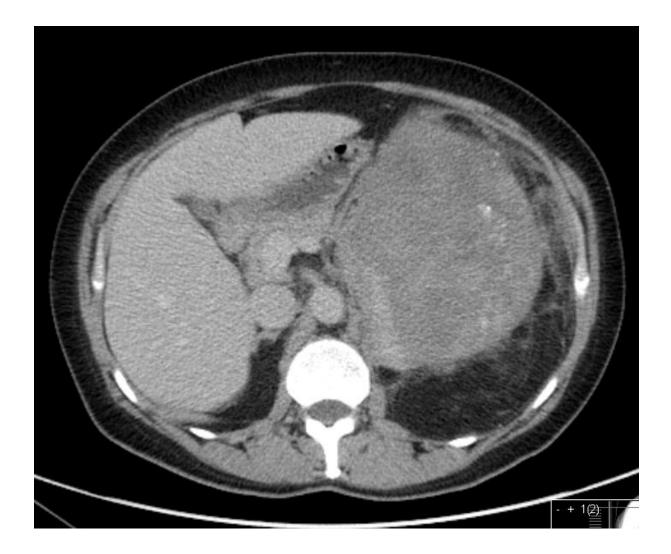


## Superio-medial margin -pancreas



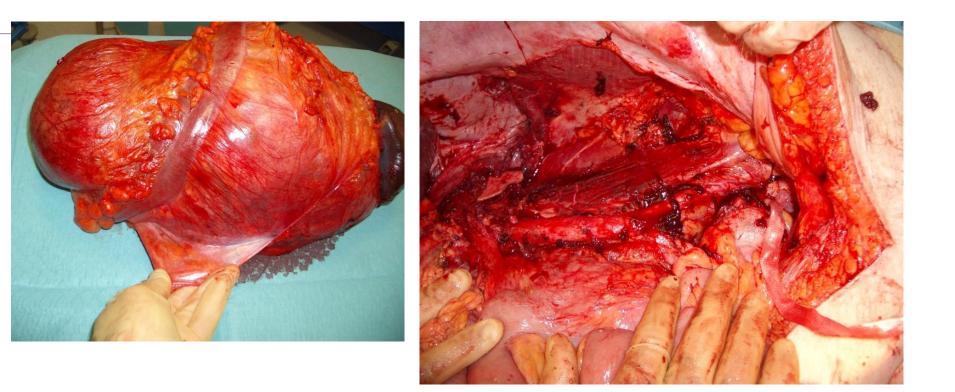


# Lateral margin - peritoneal



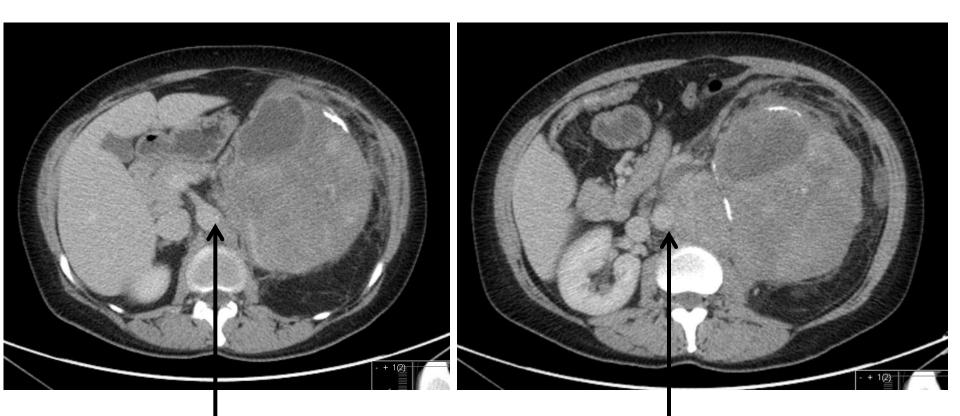


# Lateral margin - peritoneal



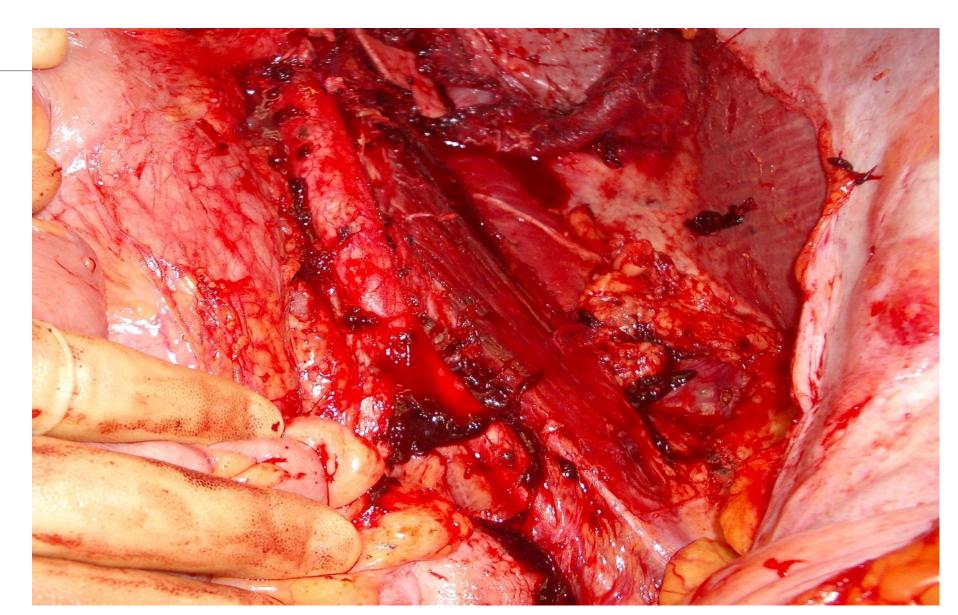


## Medial margin - aorta

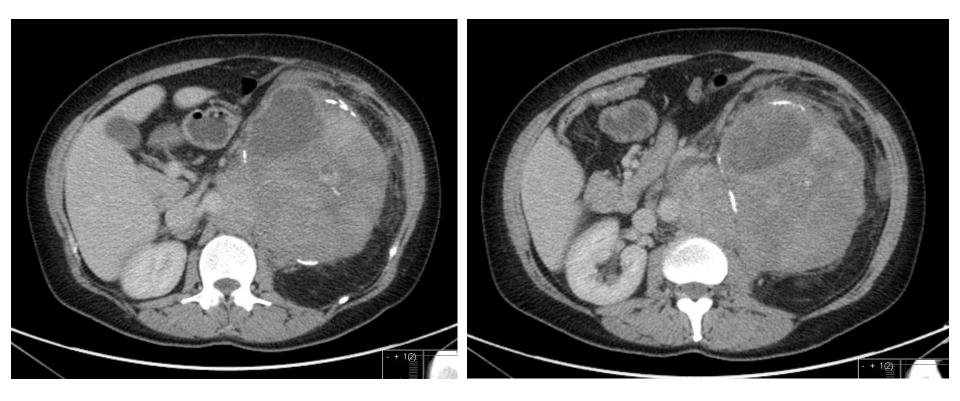




# Medial margin - aorta



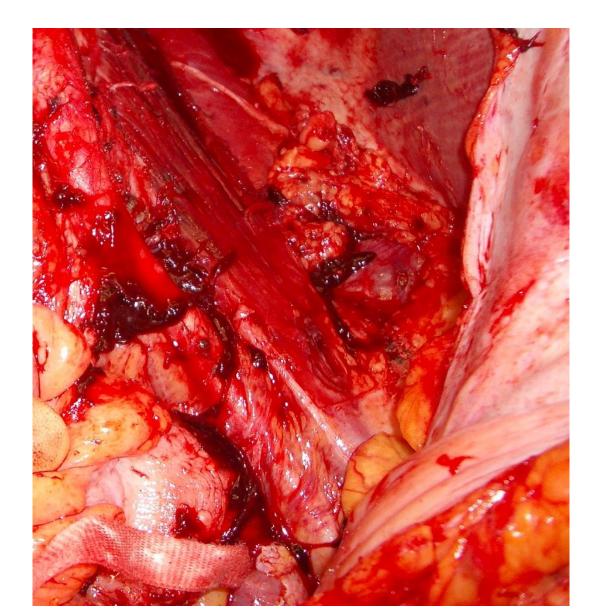
#### Posterior margin -psoas muscle





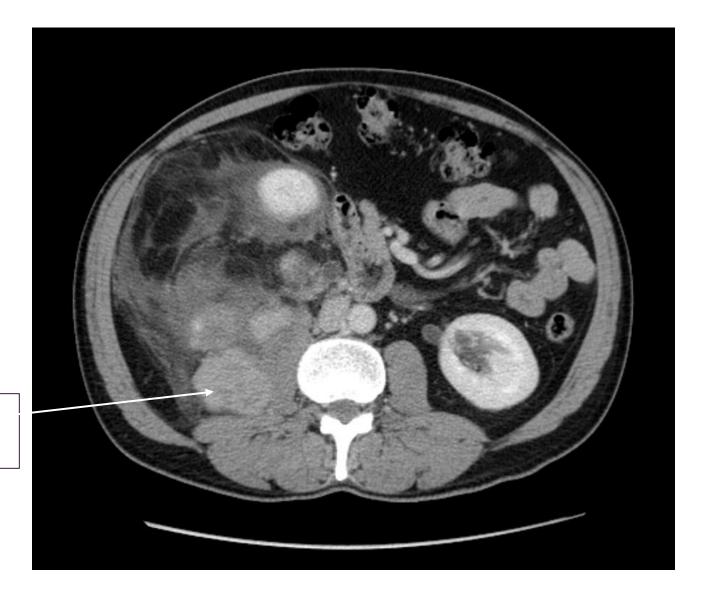
Psoas muscle or psoas aponeurosis?

## Posterior margin -psoas muscle





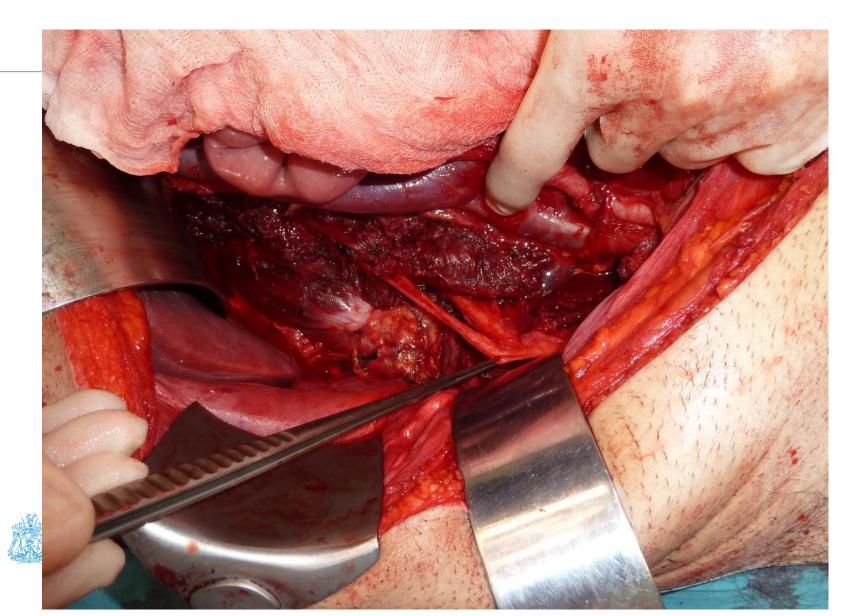
#### Posterior margin -psoas muscle



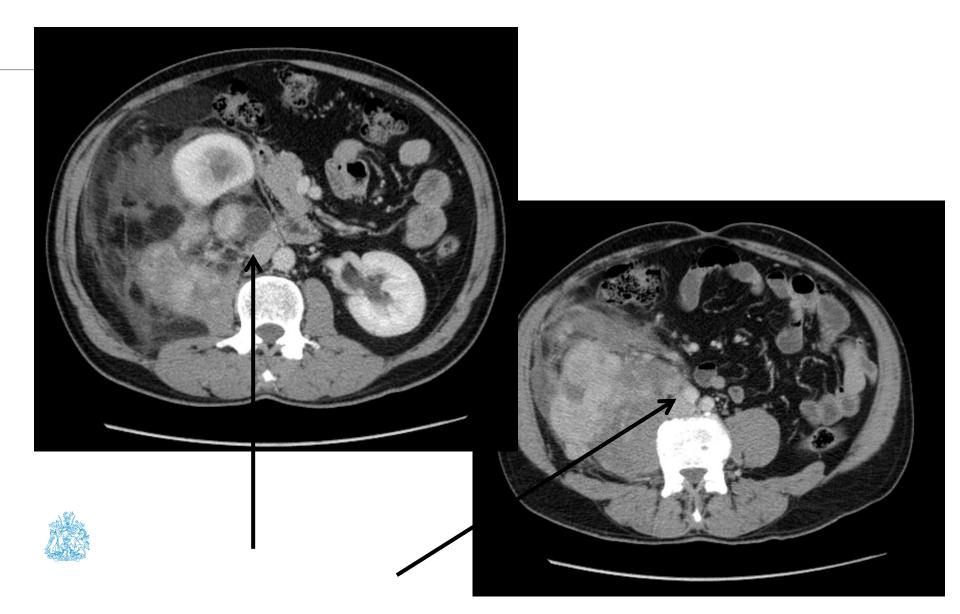
High-grade component



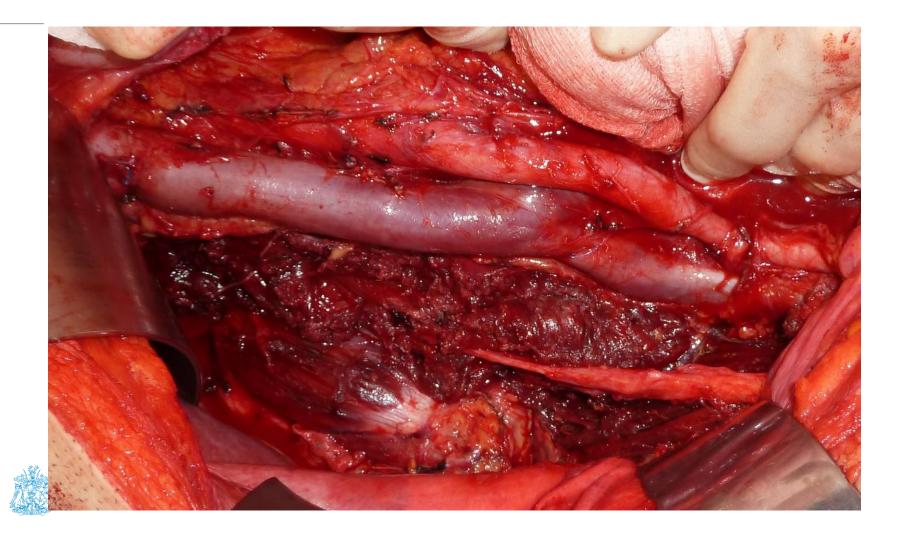
#### Posterior margin -psoas muscle



#### Inferior vena cava, aorta



#### Inferior vena cava, aorta



#### Direct infiltration

Organs or structures should be resected only if directly

infiltrated: duodenum, head of pancreas, liver, stomach,

diaphragm, major abdominal vessels, bladder/rectum

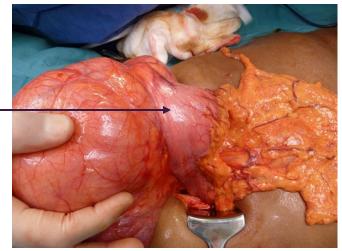
and nerves, bone.

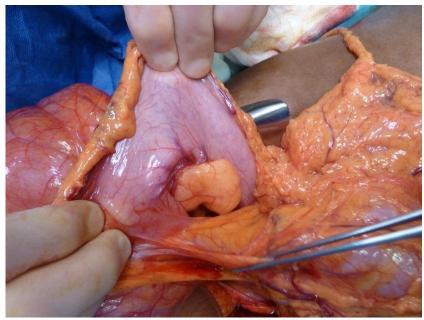


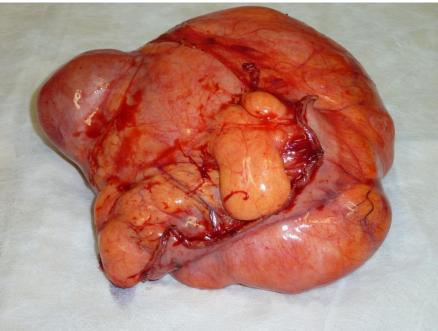
#### **Direct infiltration**



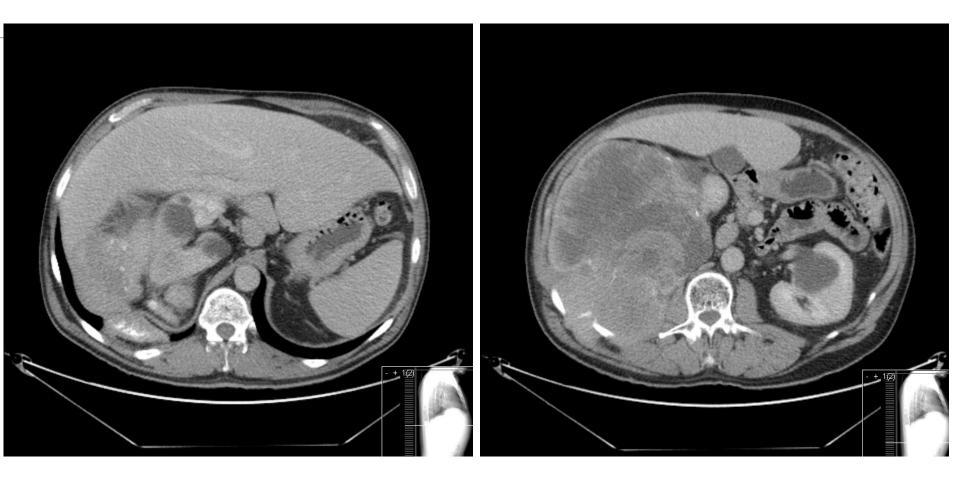
- Stomach -







#### **Direct infiltration**





R kidney, liver, diaphragm, abdominal wall and chest wall

#### Pleomorphic sarcoma

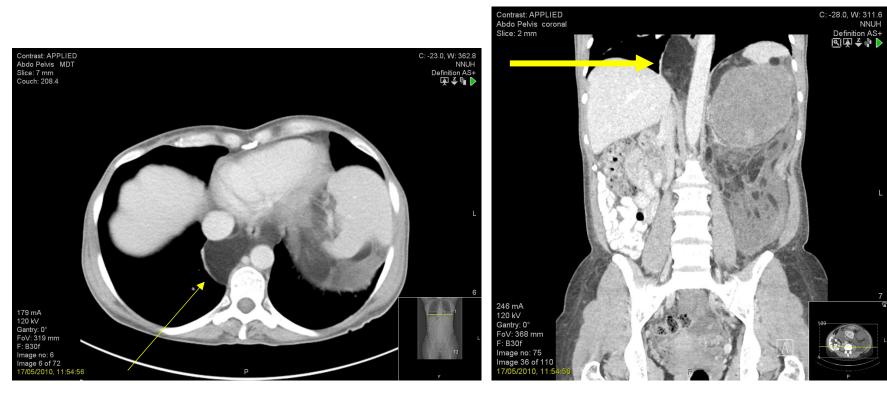


Pelvic exenteration



## Extra-abdominal extension

#### Surgical extent





#### Extra-abdominal extension





- The retroperitoneum can host a wide spectrum of pathologies, including a variety of rare benign tumours and malignant neoplasms which can be primary or metastatic lesions.
- Other diagnoses must be considered when the radiological appearance is not typical of a retroperitoneal liposarcoma.



- The optimal management of retroperitoneal sarcoma (RPS) is facilitated by pre-treatment diagnosis and staging.
- Image-guided percutaneous core needle biopsy of RPS is strongly recommended.
- A preoperative core needle biopsy is safe and does not affect oncological outcome.
- An open or laparoscopic surgical incision biopsy must be strongly discouraged.



 The best chance of a curative resection is at the time of primary presentation.

 The individual management plan should be determined taking into account both imaging and pathologic findings.



- Biologic behaviour, response to treatment, and clinical outcomes vary by histological subtype of RPS. The management plan, including extent of resection and neoadjuvant strategies, should be formulated accordingly.
- The current standard of care for retroperitoneal sarcoma is extended en bloc complete resection of the tumour and surrounding viscera performed in highvolume centres.



#### The ROYAL MARSDEN NHS Foundation Trust



Dirk.Strauss@rmh.nhs.uk



