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Retroperitoneal sarcomas

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Royal Marsden Hospital, London

Sarcoma Webinar, 27 Feb 2021

Introduction

There are more than 60 different subtypes of STS

Within the retroperitoneum, there are five main histologic subtypes that account for 90% of these tumours:

- Dedifferentiated liposarcoma (DDLPS)
- Well-differentiated liposarcoma(WDLPS)
- Leiomyosarcoma (LMS)
- Solitary fibrous tumour (SFT)
- Malignant peripheral nerve sheath tumour (MPNST)

(UPS, synovial, Ewing's, PEComa, DSRCT, IMT)

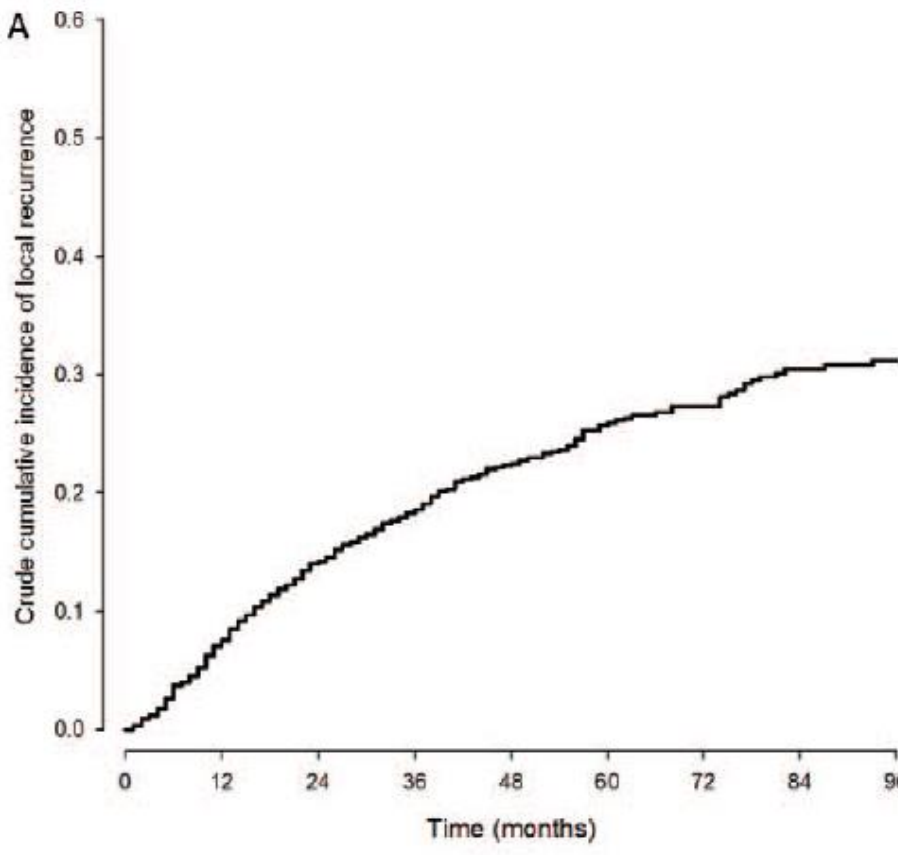
Background

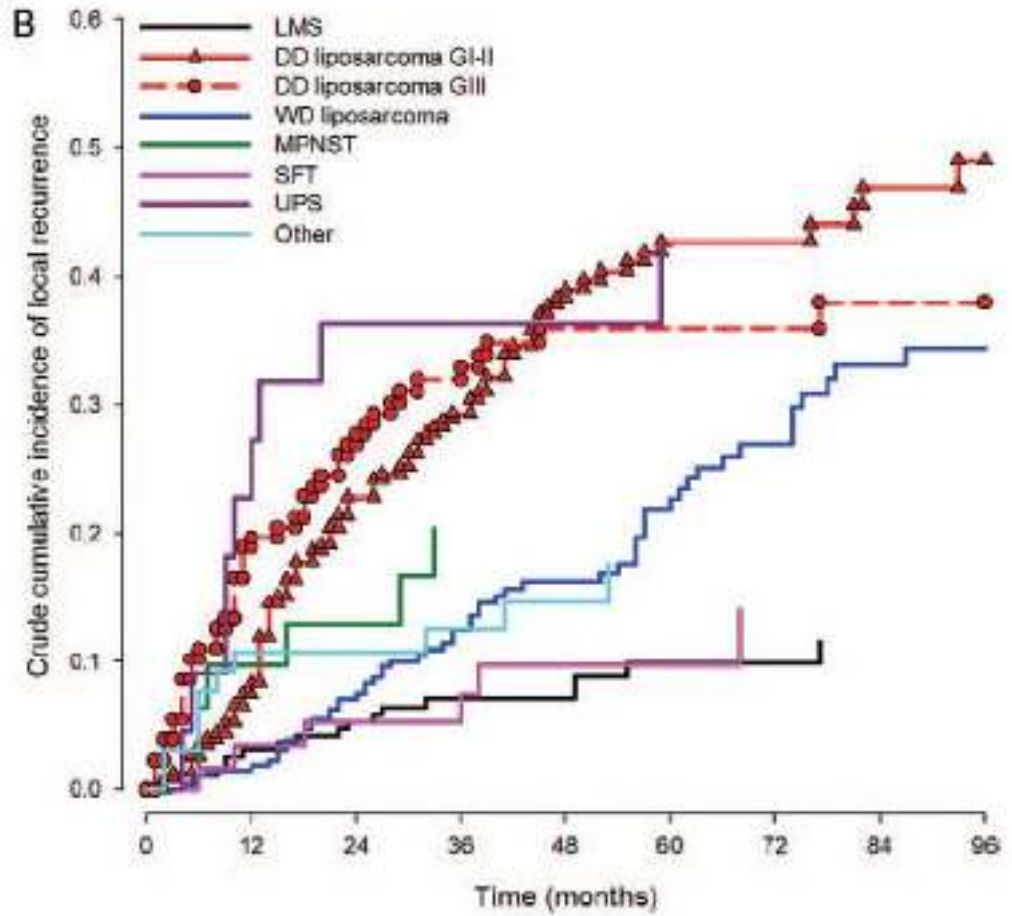
Tumour biology and completeness of surgical resection are two of the most important factors for consideration when treating RPS.

- Tumour biology, (determined by the grade and the histologic subtype) remains outside the control of the surgeon.

- While surgical margins and extent of resection, surgeons can influence to impact the patient outcome.

Cumulative incidence of local relapse





DD LPS Gr2 ~ 50%

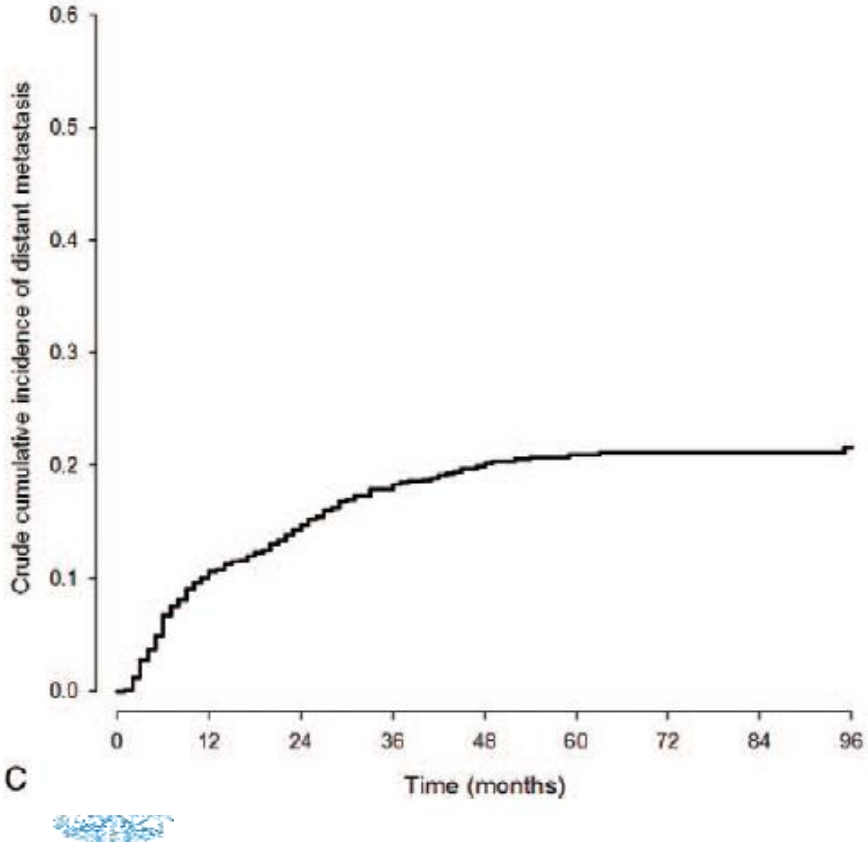
DD LPS Gr3 ~ 40%

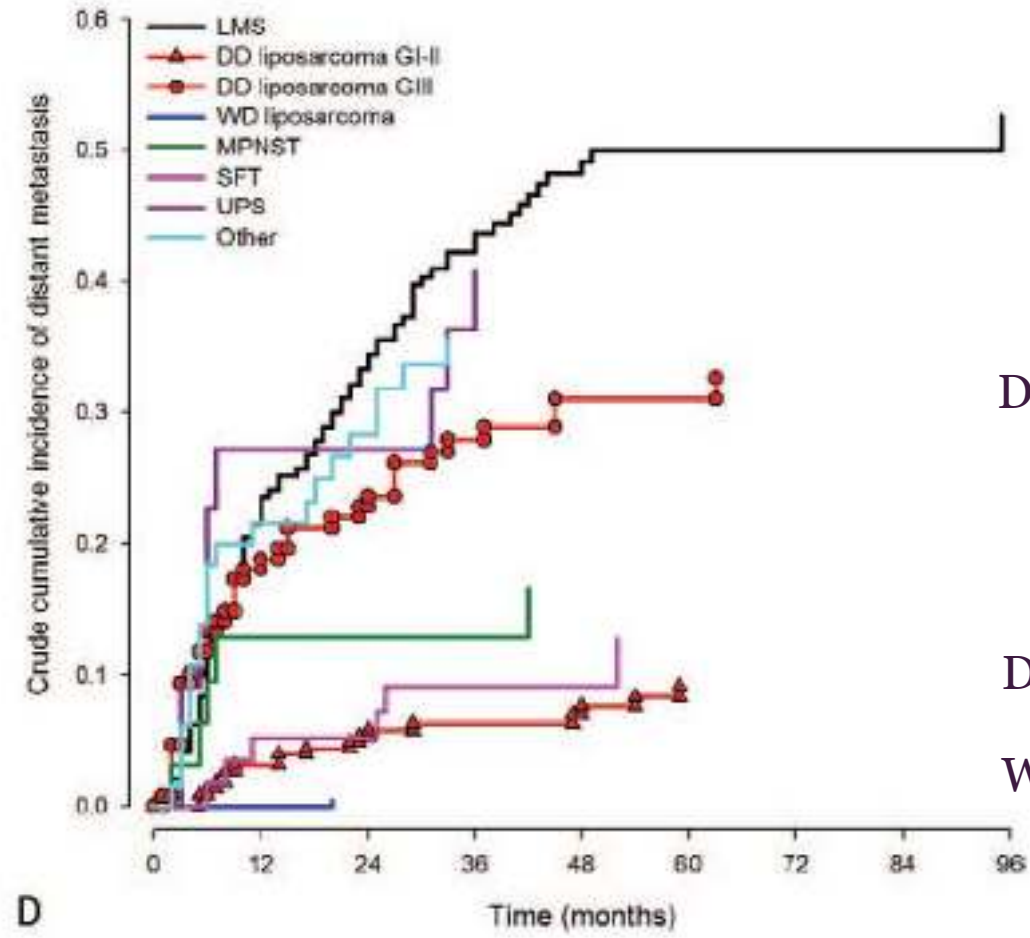
WD LPS ~ 35%

LeiMS ~ 10%

A Report on 1007 Patients From the Multi-institutional Collaborative RPS Working Group.
Ann Surg. May 2016 - Volume 263 - Issue 5 - p 1002–1009

Cumulative incidence of distant metastasis





LeiMS ~ 50%

DD LPS Gr3 ~ 30%

DD LPS Gr2 ~ 6%

WD LPS =0%

A Report on 1007 Patients From the Multi-institutional Collaborative RPS Working Group. Ann Surg. May 2016 - Volume 263 - Issue 5 - p 1002–1009

General approach to RPS…

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.

Core needle biopsy

Image-guided percutaneous coaxial core needle biopsy (14 -18 gauge) is strongly recommended as the standard of care.

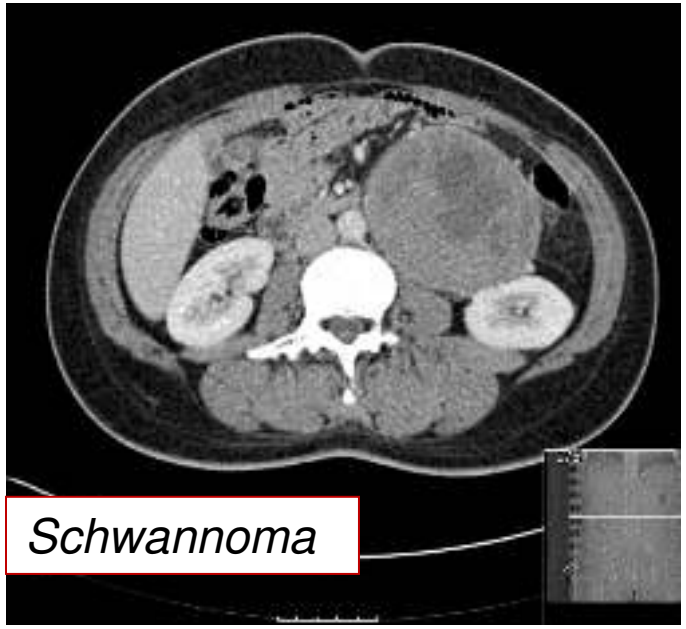
Core needle biopsy

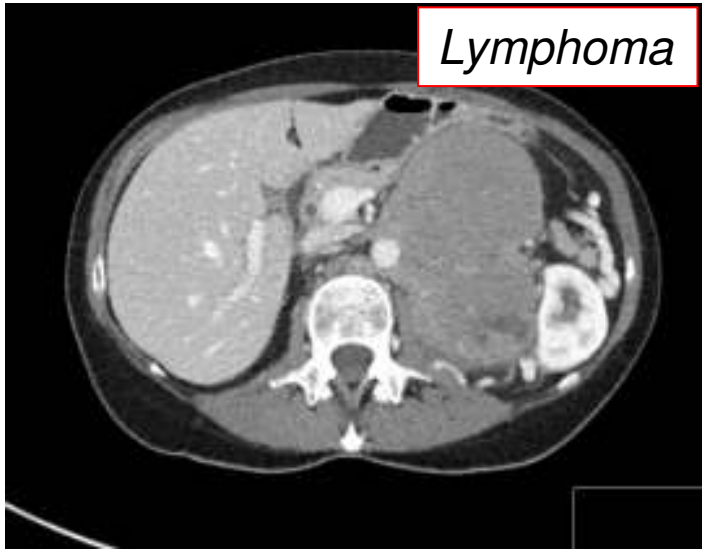
Pre-treatment core needle biopsy of a retroperitoneal mass can identify:

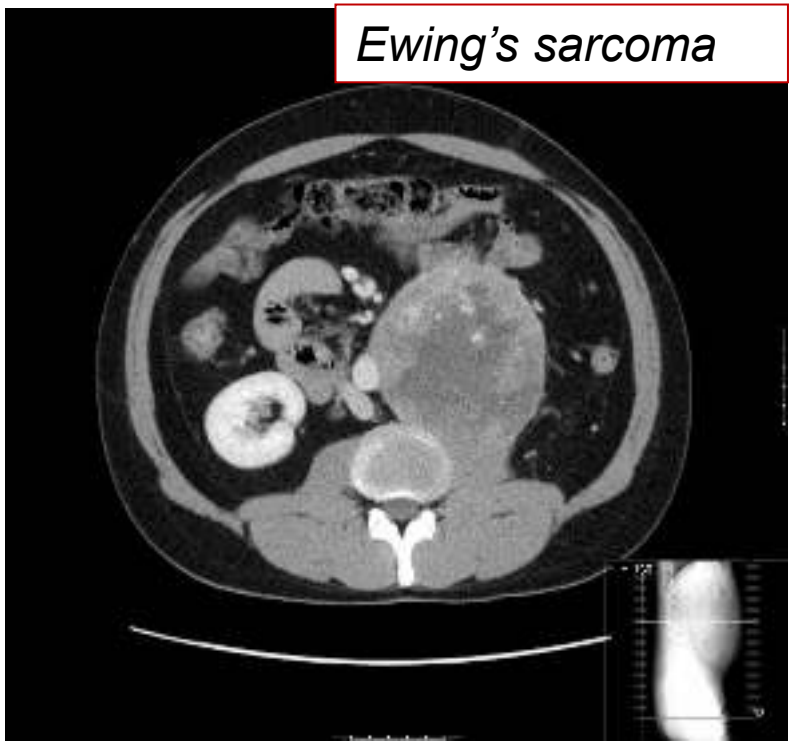
- patients who do not require surgery (benign from malignant tumours)
- enables appropriate multidisciplinary treatment planning (neoadjuvant chemo/radiotherapy)
- histologic subtype- and grade- adapted surgical planning

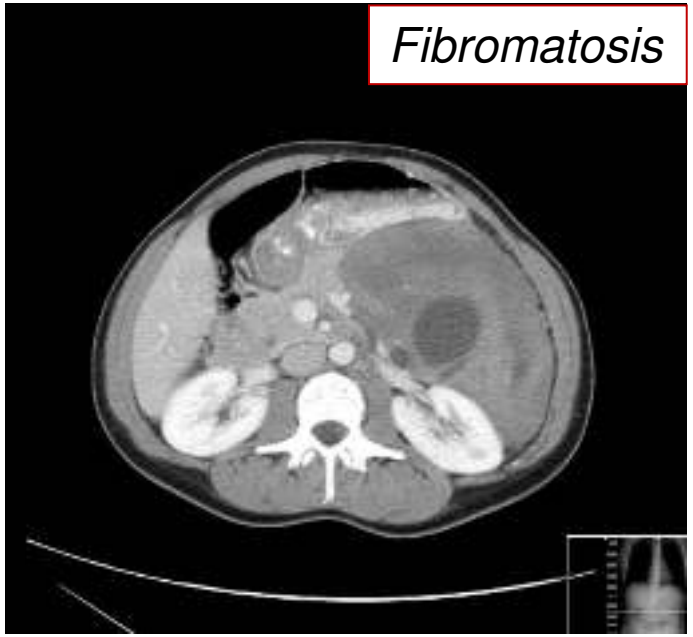
– soft tissue tumours from non-mesenchymal tumours (primary or metastatic epithelial malignancy i.e lymphoma, carcinoma, melanoma etc)

Core needle biopsy

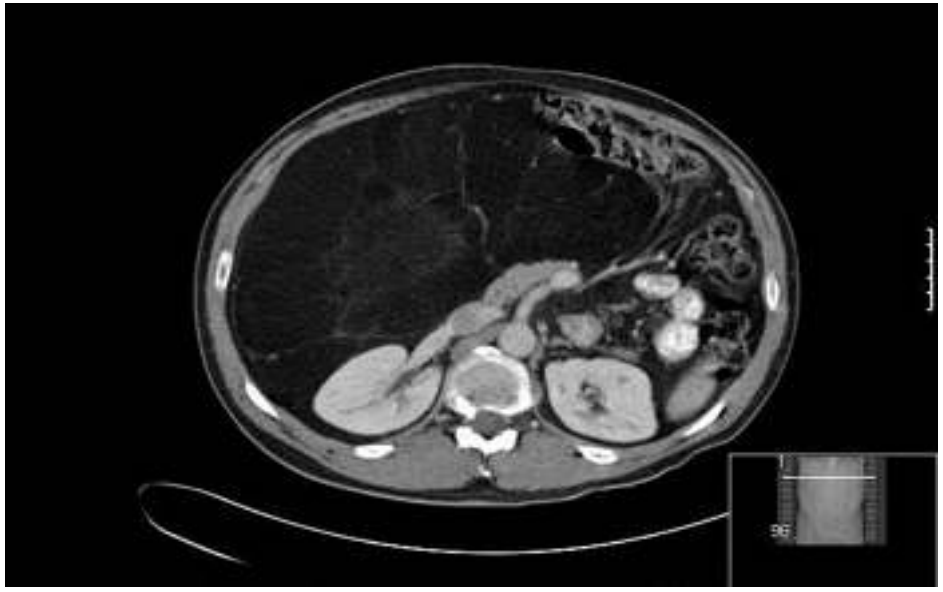






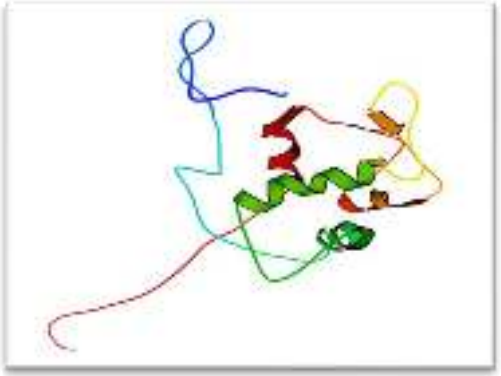


Role of core needle biopsy - lipomatous lesions

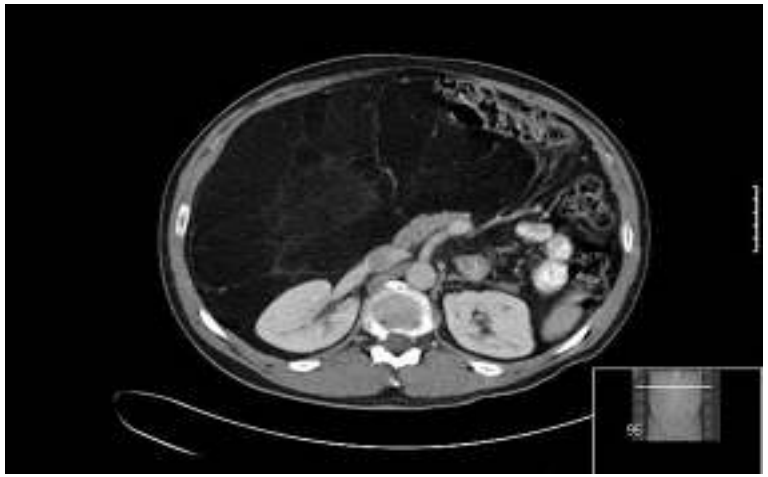


Role of core needle biopsy - lipomatous lesions

– WDLPS and DDLPS harbour supernumeray ring and/or giant chromosomes due to amplification gene MDM2, CDK4 in chromosome 12q13-15



- Determines whether surgery required
- Extent surgery



Role of core needle biopsy

Ann Surg Oncol (2015) 22:853–858
DOI 10.1245/s10434-014-4059-x

Annals of
SURGICAL ONCOLOGY
OFFICIAL JOURNAL OF THE SOCIETY OF SURGICAL ONCOLOGY

ORIGINAL ARTICLE – BONE AND SOFT TISSUE SARCOMAS

Percutaneous Core Needle Biopsy in Retroperitoneal Sarcomas Does Not Influence Local Recurrence or Overall Survival

M. J. Wilkinson, MRCS[1], J. L. Martin, MCRS[1], A. A. Khan, MRCS, MPH[1], A. J. Hayes, PhD, FRCS[1], J. M. Thomas, FRCP, FRCS[1], and D. C. Strauss, MMed, FRCS[1,2]

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Ann Surg Oncol (2019) 26:4692–4698
https://doi.org/10.1245/s10434-019-07656-6

Annals of
SURGICAL ONCOLOGY
OFFICIAL JOURNAL OF THE SOCIETY OF SURGICAL ONCOLOGY

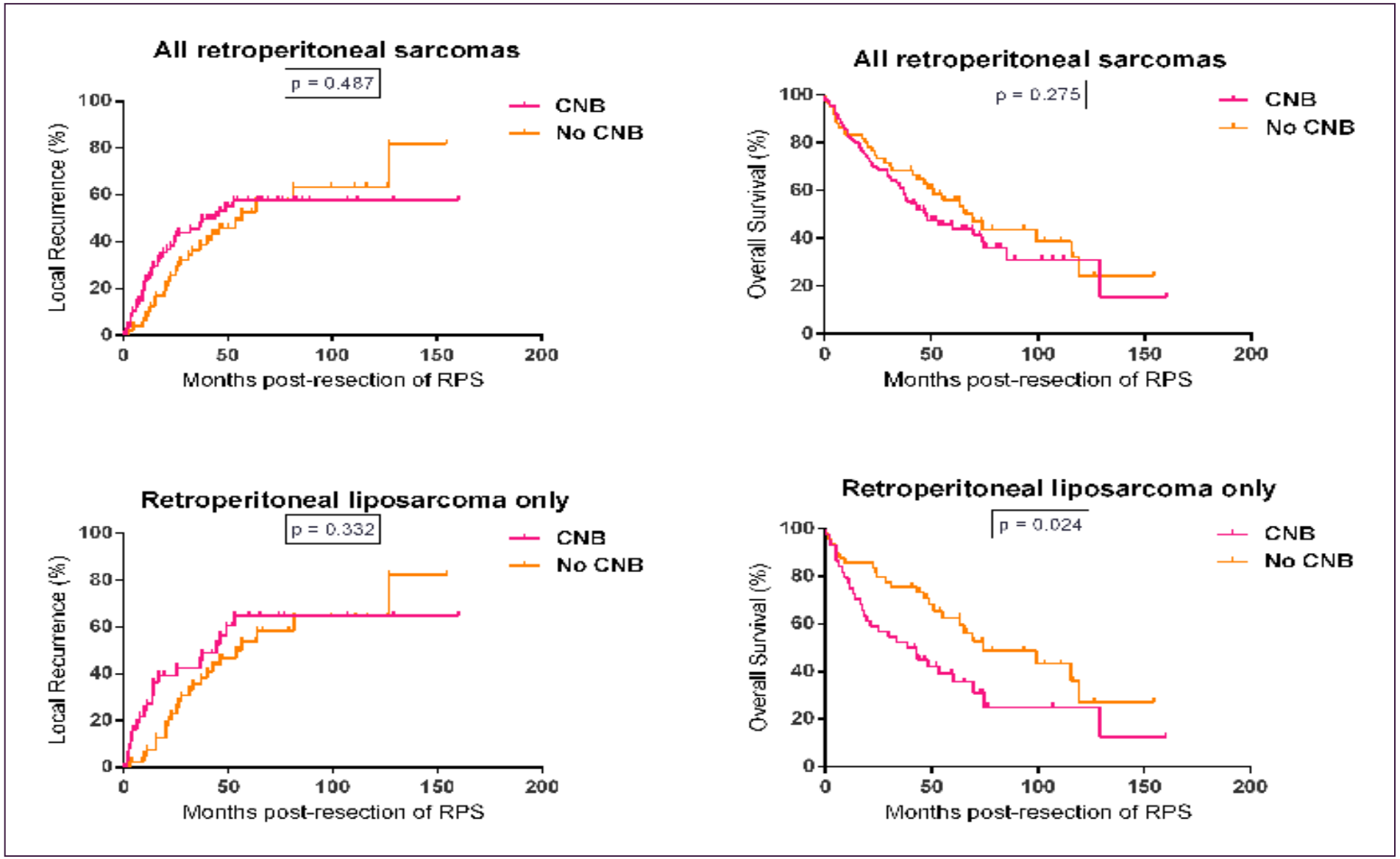
ORIGINAL ARTICLE – SARCOMA

Early and Late Complications of Percutaneous Core Needle Biopsy of Retroperitoneal Tumors at Two Tertiary Sarcoma Centers

David Berger-Richardson, MD[1,2], Sally M. Burtenshaw, MSc[3,4], Andrea M. Ibrahim, MSc[5], Rebecca A. Gladdy, MD[1,2,3,4], Rebecca Auer, MD[5,6,7], Rob Beecroft, MD[8], Brendan C. Dickson, MD[9], Bibianna Purgina, MD[10], Kristin Ambacher, MD[5], Carolyn Nessim, MD[5,6,7], and Carol J. Swallow, MD[1,2,3,4]

Role of core needle biopsy

Safe!!



Initial step...

- Core needle biopsy is simple and safe and provides accurate diagnostic information which is essential to determine the appropriate treatment plan

- The benefits of pre-treatment biopsy in patients with soft tissue tumours outweigh the potential risks of needle tract seeding

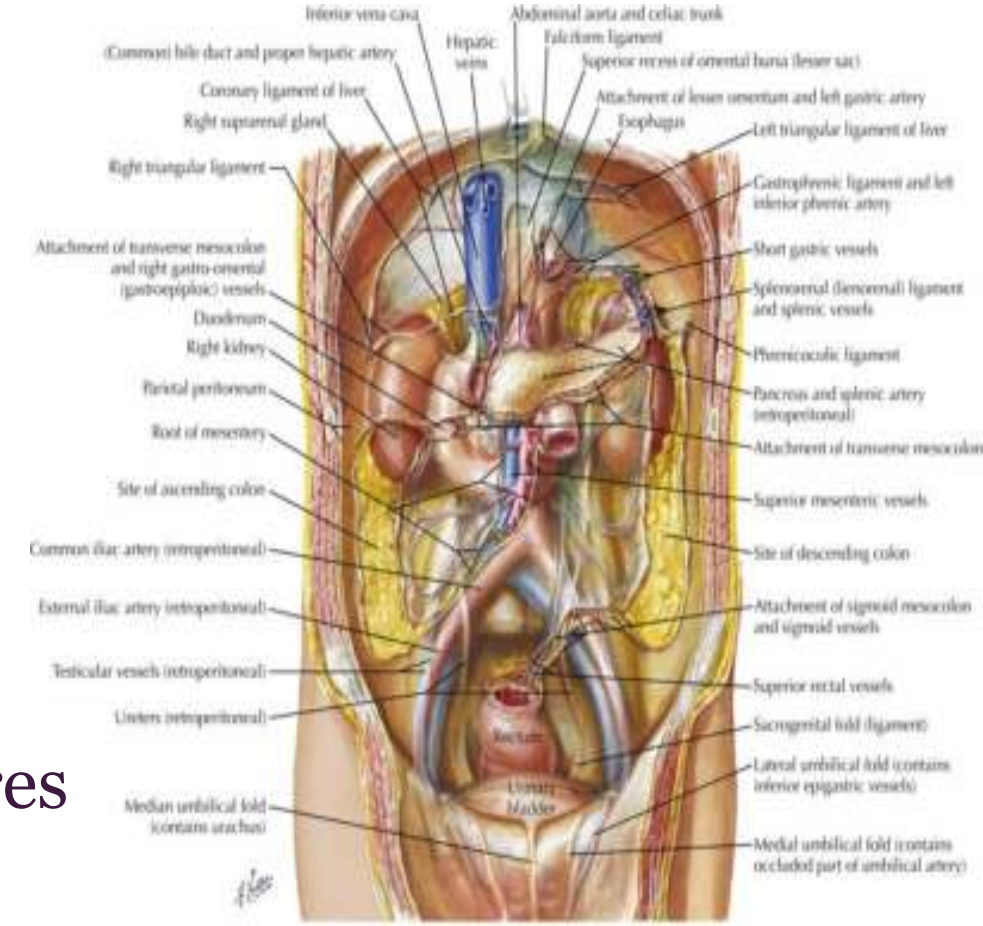
- Core needle biopsy should be the method of choice to obtain an accurate pathological diagnosis in suspected soft tissue tumours in RPS and facilitate planned personalised treatment

Background

Surgery for RPS constitute a therapeutic challenge

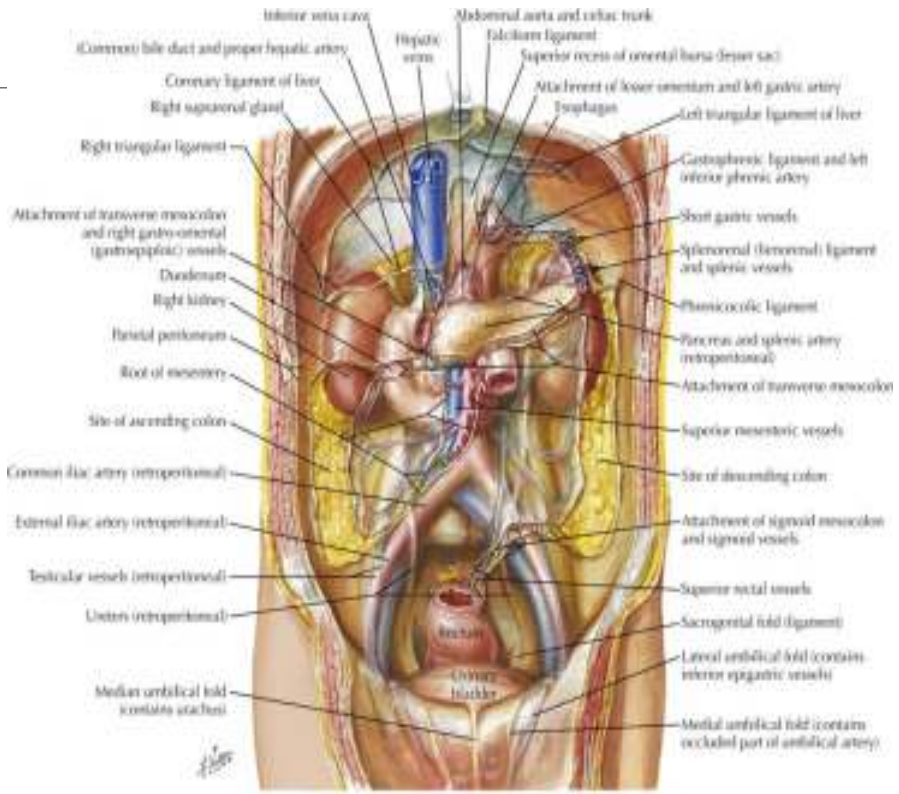
– late presentation, large tumours

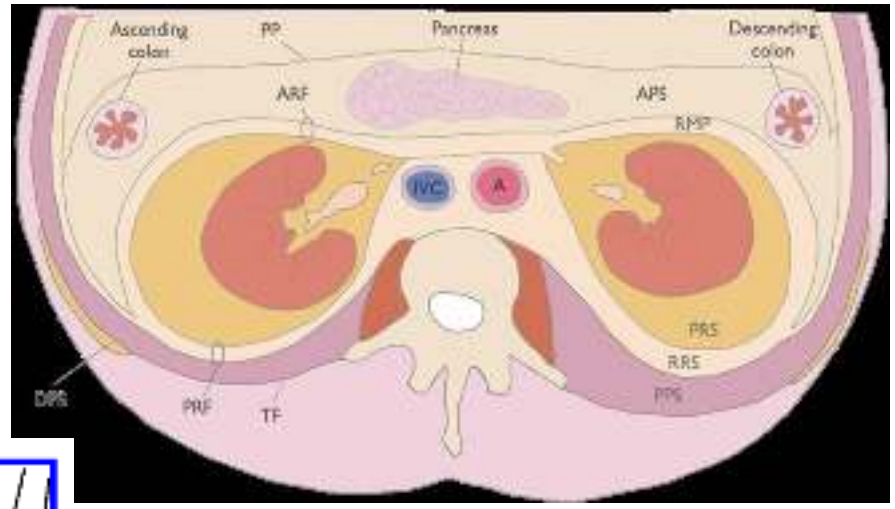
- complex space
- in close relationship to multiple vital structures

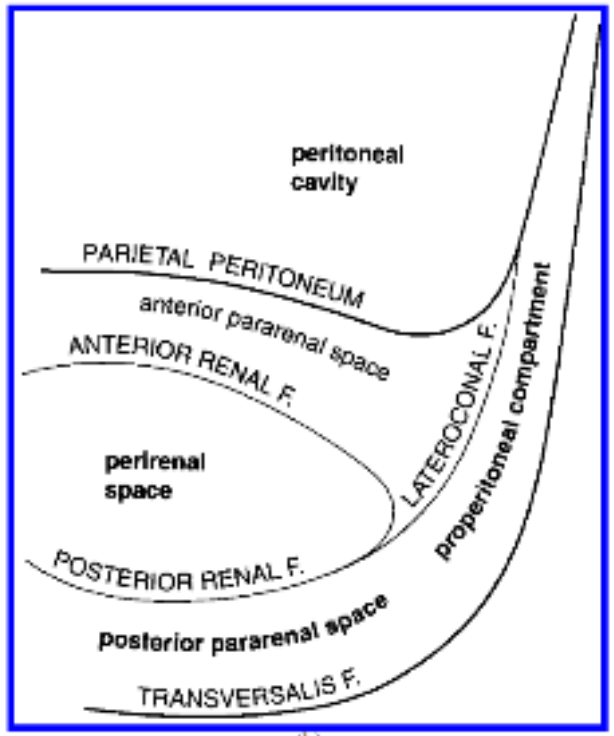


– close relationship to vital structures impacts on the ability to perform a radical wide resection

Anatomy

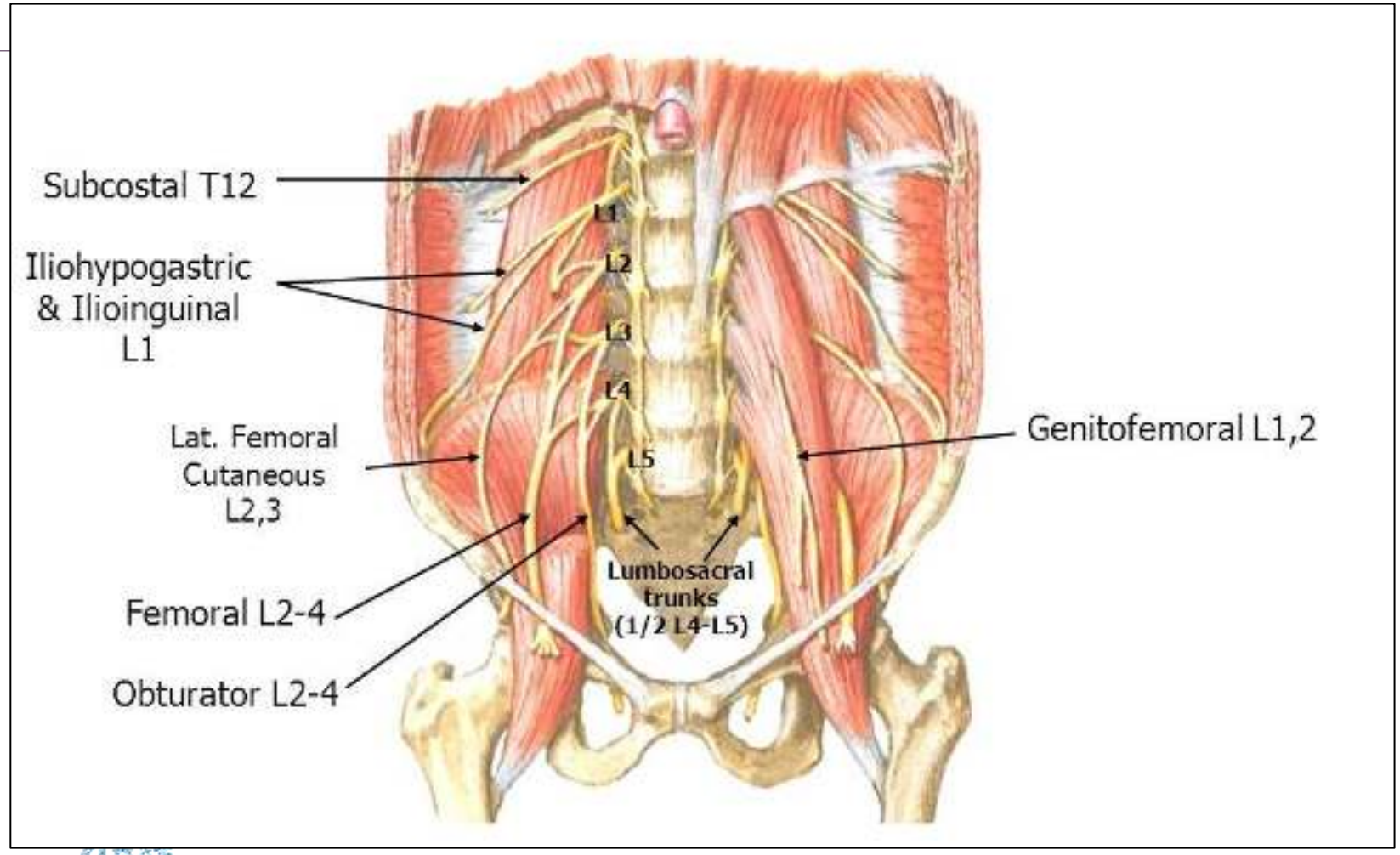






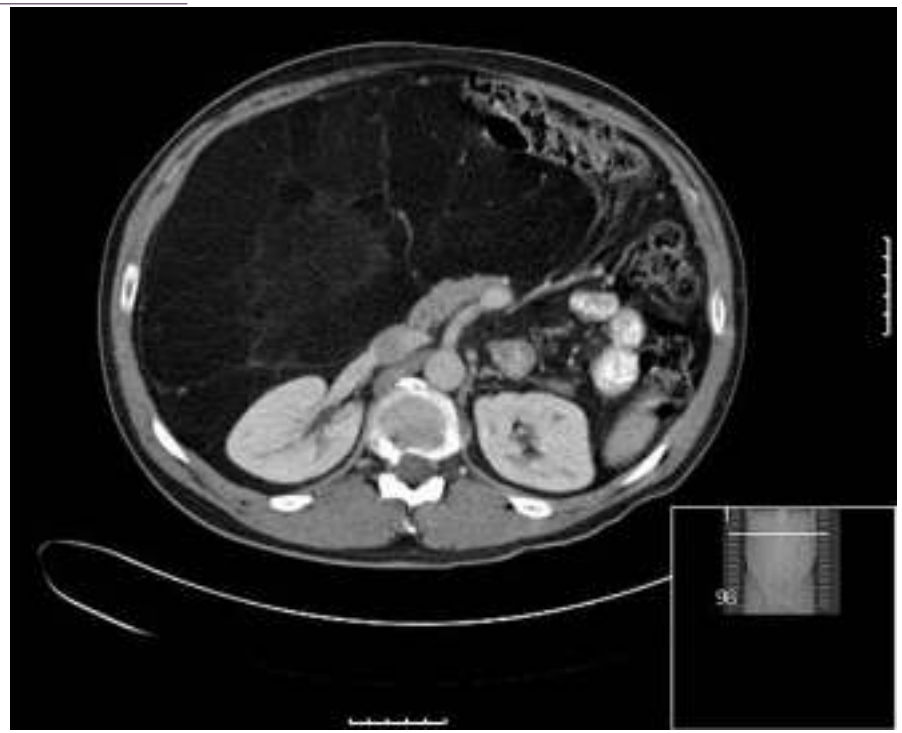
A = Aorta
IVC = Inferior vena cava
PP = Parietal peritoneum
RMP = Retromesenteric plane
RRS = Retrorenal space
APS = Anterior pararenal space
PRS = Perirenal space
PPS = Posterior pararenal space
TF = Transversalis fascia
PRF = Retrorenal fascia
ARF = Anterior renal fascia

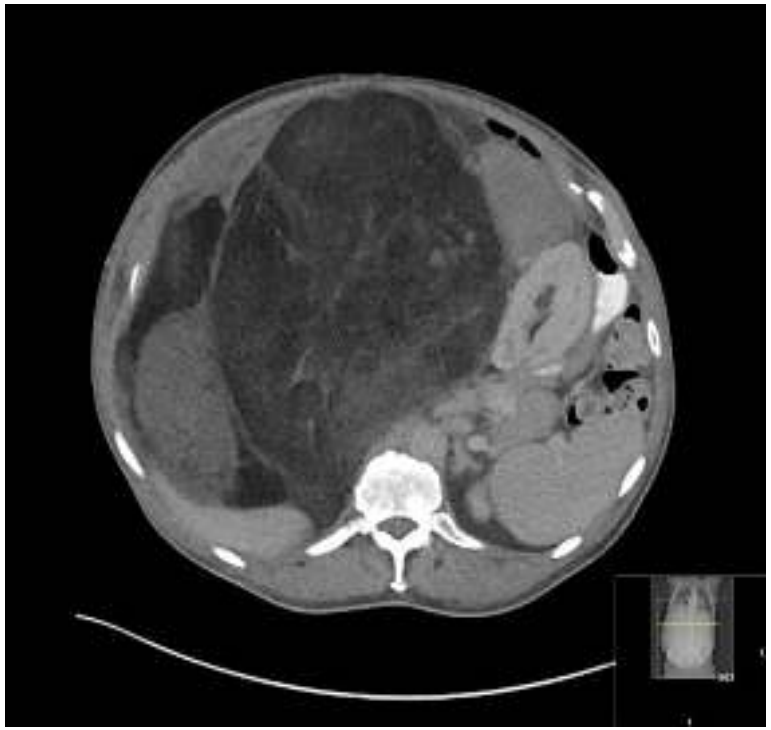
Anatomy



- Genitofemoral n
- Ilioinguinal n
- Iliohypogastric n
- Lateral femoral cutaneous n
- Femoral n
- Obturator n
- Lumbosacral trunks

Anatomy – distorted by tumour

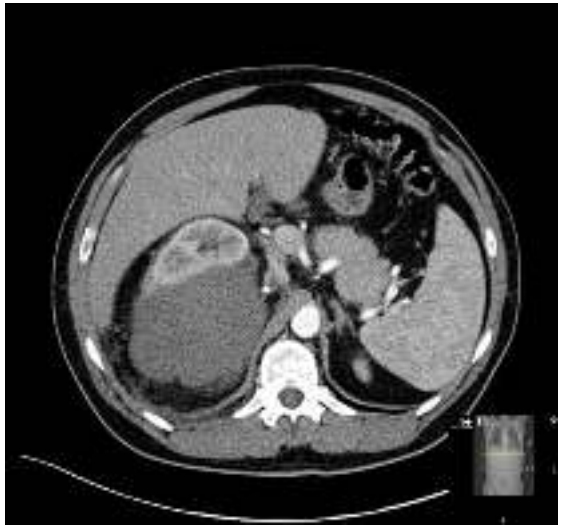


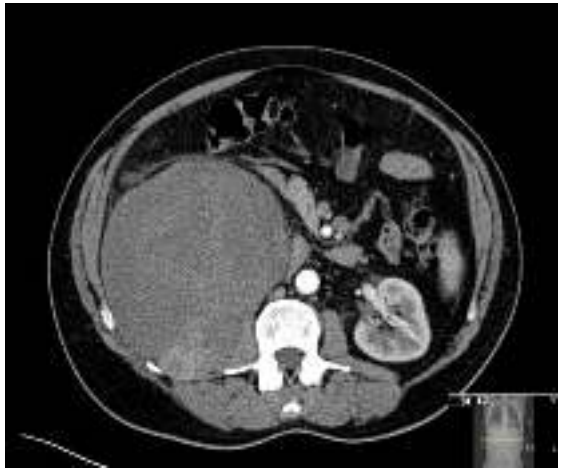


Psoas/posterior abdominal wall sarcoma

Differential diagnosis

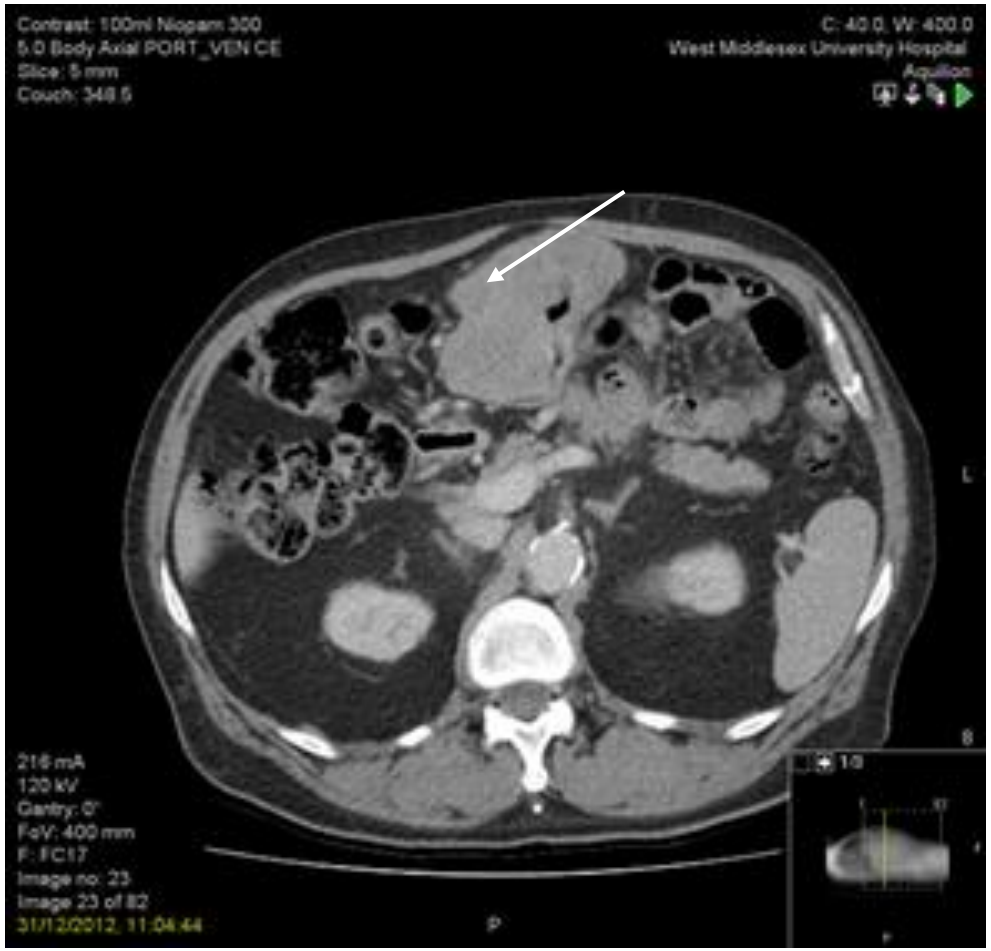
Retroperitoneal v intraperitoneal





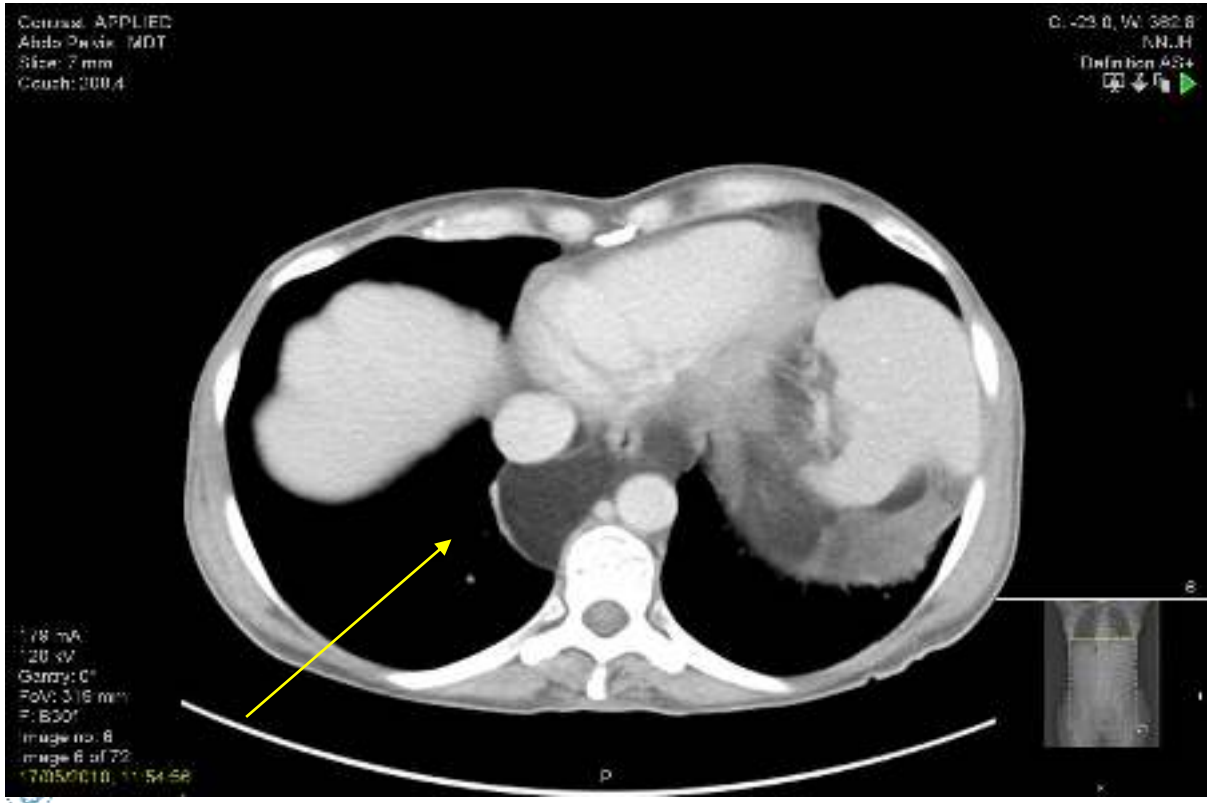
Differential diagnosis

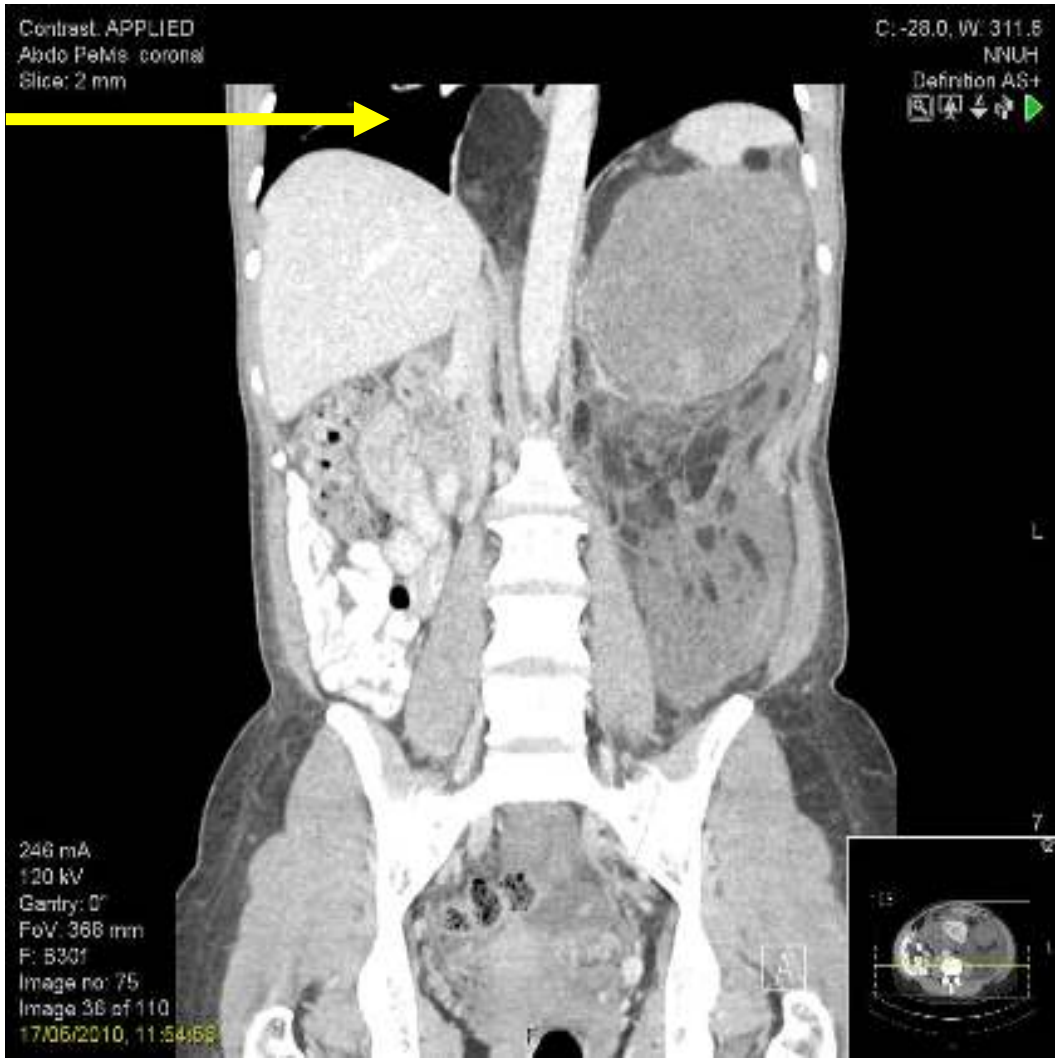
Visceral: renal, adrenal, pancreas, stomach, duodenum, colon, ovary, uterus



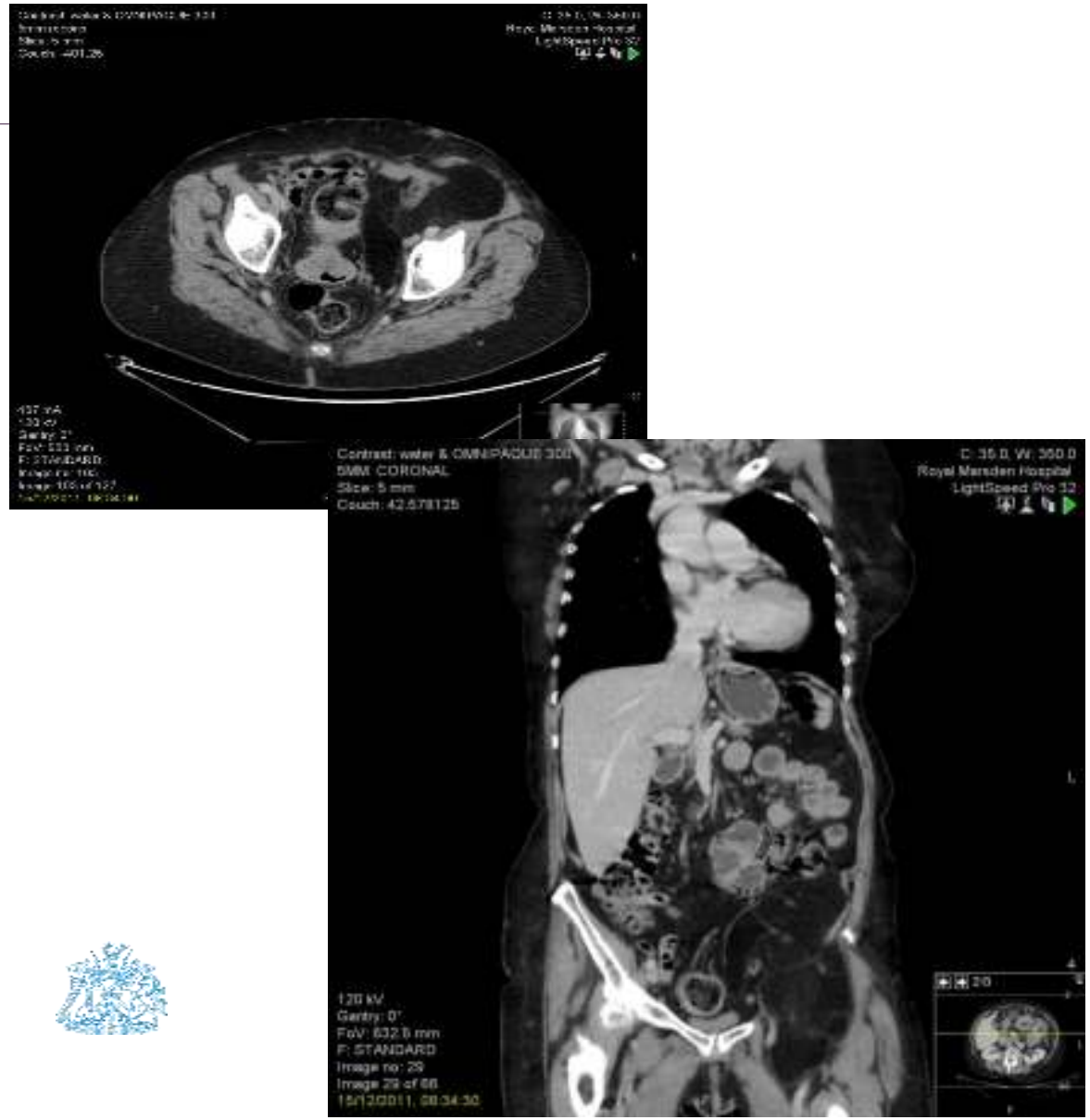
Extra-abdominal extension

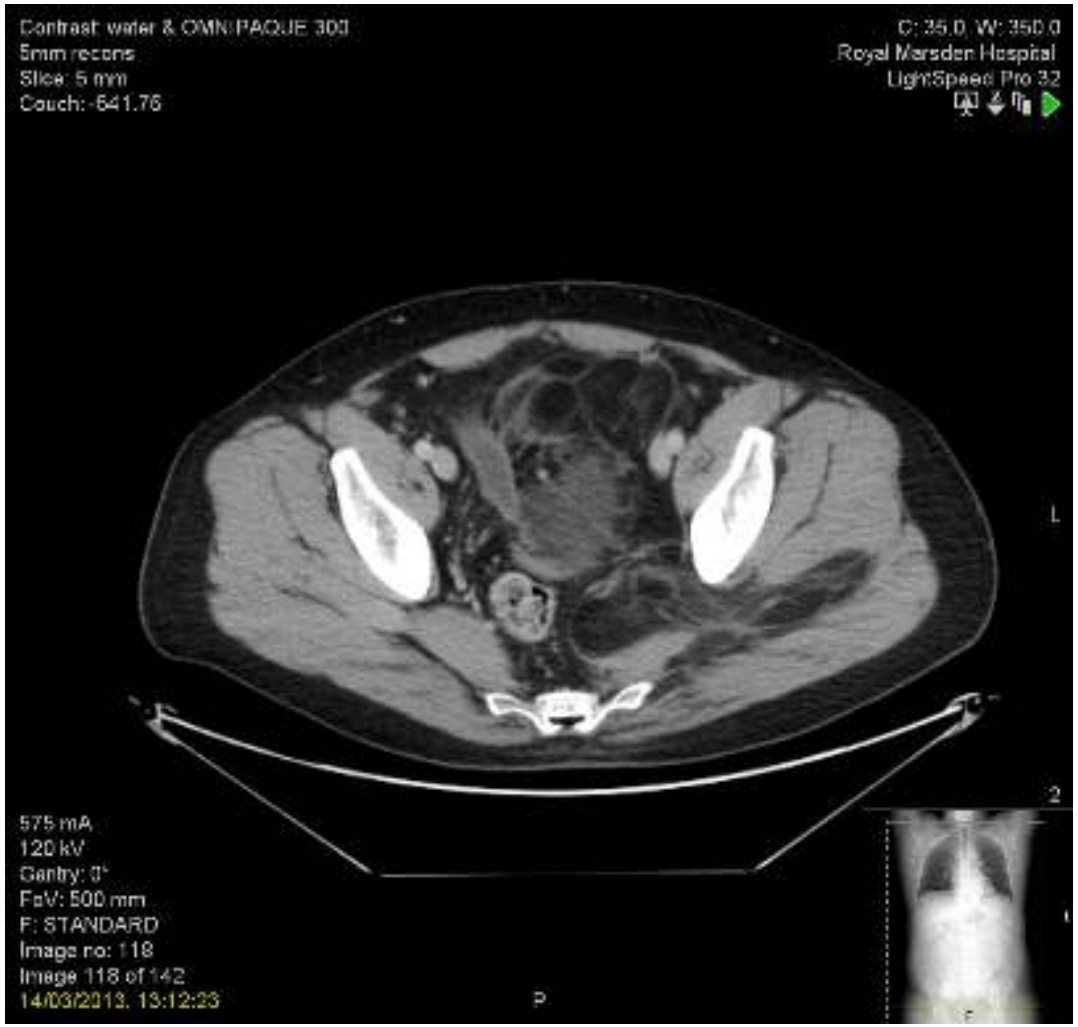
Surgical extent



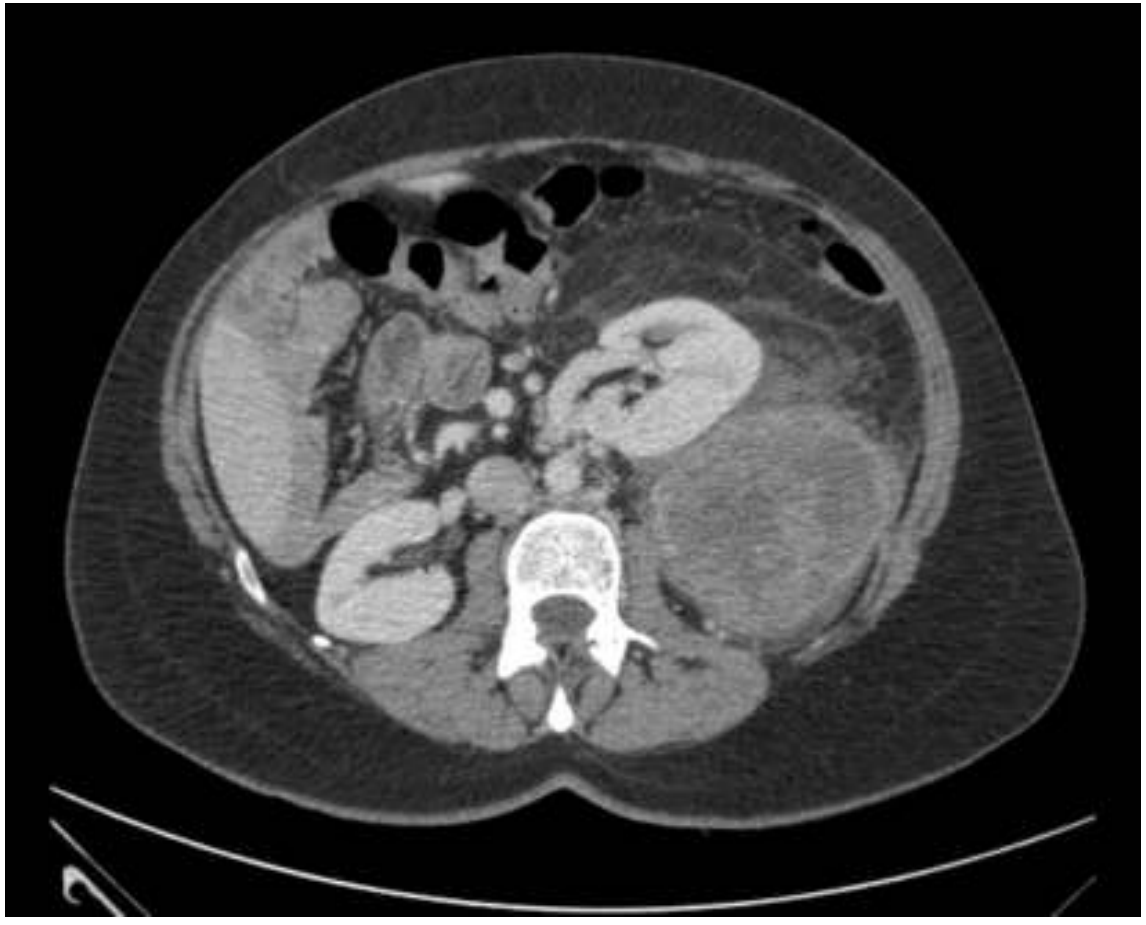


Extra-abdominal extension





Preoperative evaluation



Conclusion…

- Surgical expertise in RPS resection requires specific anatomic knowledge of the retroperitoneal space to minimize the risk of intra- and perioperative morbidity

- The standard method for staging is contrast tomography (CT) scan of the chest/abdomen/ pelvis with IV contrast.

- Large tumours distort anatomy

Conclusion…

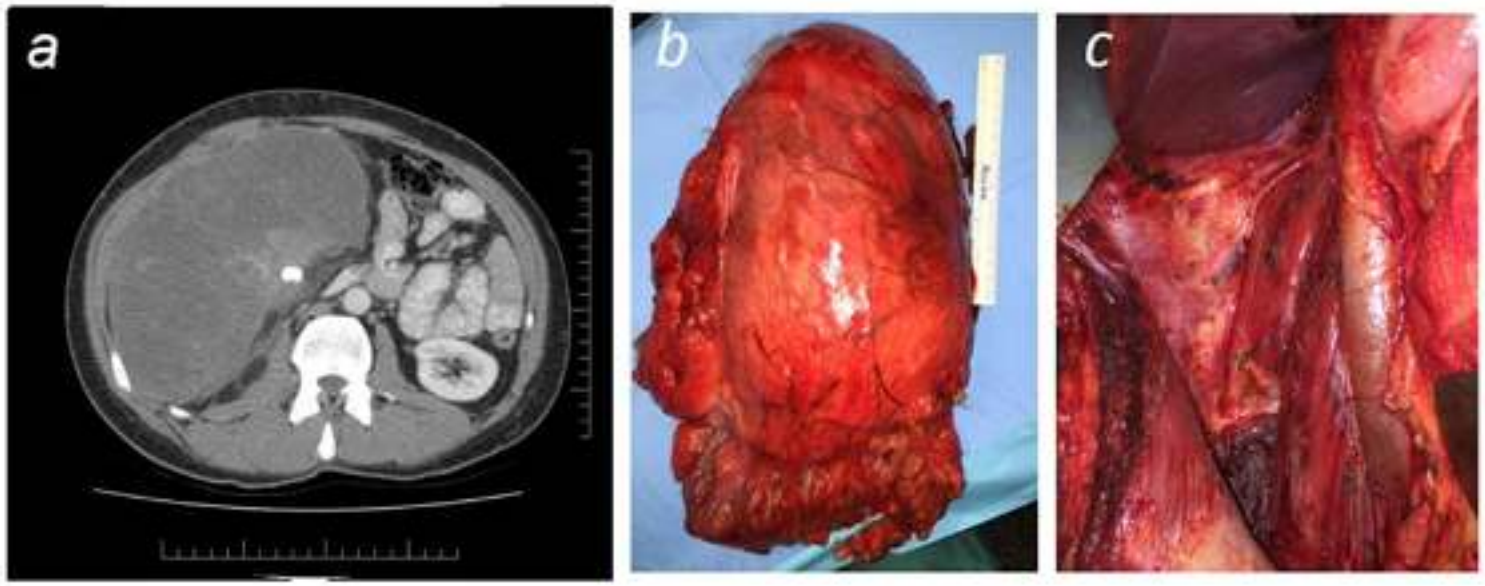
– Improper planning may lead to inadequate incisions, tumour rupture, incomplete resections, and risk of catastrophic bleeding.

– Tumour extent and anatomic relationship with intra-abdominal organs, vessels, parietal wall, and bone must be carefully analysed.

Operative strategy

Complete en bloc gross resection is the cornerstone of management

- In the case of primary RPS, surgery should be aimed at achieving macroscopically complete resection, with a single specimen encompassing the tumour and involved contiguous organs



Operative strategy

The concept of adapting the operative approach according to the histologic subtype has recently gained increased recognition.

Operative strategy for liposarcoma

Intraoperative macroscopic assessment of appropriate resection margins in RP liposarcoma can be challenging, particularly for WD LPS where tumour tissue is difficult to distinguish from normal fat.

Given this uncertainty regarding margin definition, an extended approach to systematically resect adherent viscera, irrespective of expected microscopic infiltration, should be considered for RP liposarcomas.

A policy of resecting only structures/viscera which are clearly invaded by liposarcoma is more likely to result in residual disease being left in the operative bed.

Radicality of surgery – “completeness of resection”

Extended resection

Frontline resection

Aggressive resection

Radical en bloc multivisceral resection

Radicality of surgery – "completeness of resection"

Topic of significant debate

Published series (2009) from Italy + France:

–"the adoption of a policy of more liberal visceral en bloc resections is paralleled by greater local control"

Clinical evidence to support

Pathological evidence to support

Bonvalot S, et al Primary retroperitoneal sarcomas: A multivariate analysis of surgical factors associated with local control. J Clin Oncol 2009;27:31–37.

Gronchi A, et al Aggressive surgical policies in a retrospectively reviewed single-institution case series of retroperitoneal soft tissue sarcoma patients. J Clin Oncol 2009;27:24–30.

Compartmental 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

- Macroscopically complete resection (R0/R1) of the tumour mass with en bloc resection of organs adjacent to the tumour, based on preoperative assessment and intraoperative findings.

- Organs to be resected when in proximity to the tumour surface: kidney, colon, psoas muscle or its aponeurosis.

- Organs or structures should be resected only if directly infiltrated: liver, stomach, major abdominal vessels, nerves, duodenum, head of pancreas, bone.

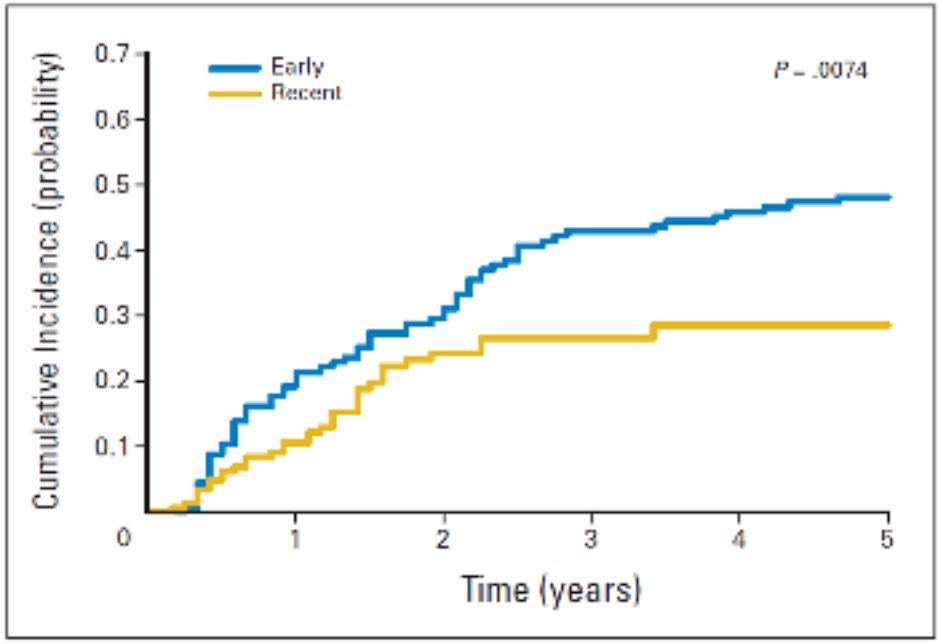
Compartmental resection

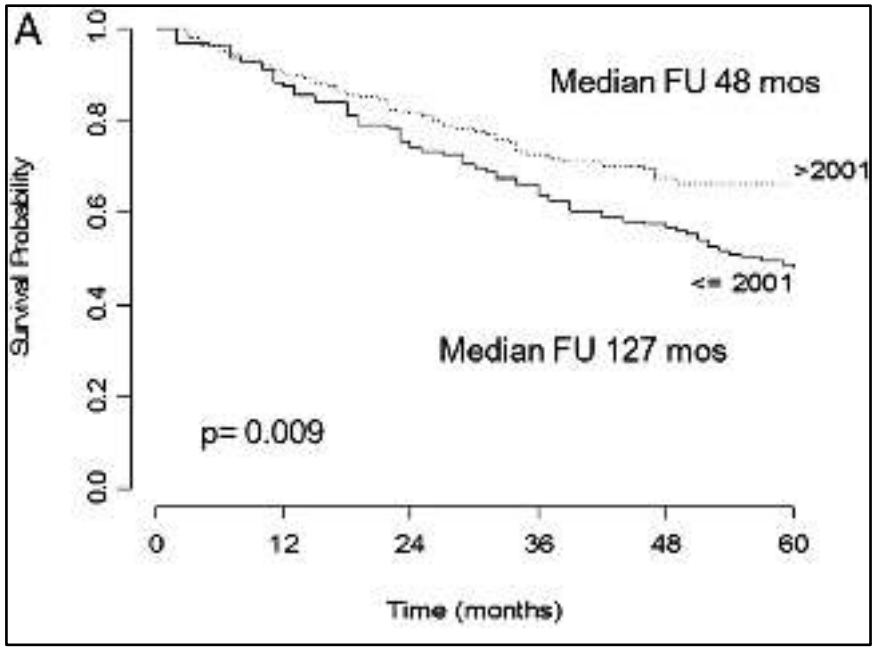
The objective is to achieve a wide microscopic margin along some surfaces by removing adjacent, easily disposable organs

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

Radicality of surgery – "completeness of resection"

Clinical evidence



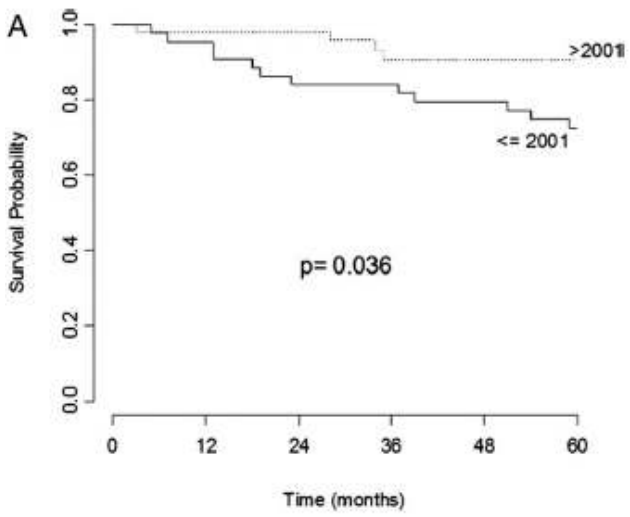


5yr local recurrence
Early – 48%
Late – 28%

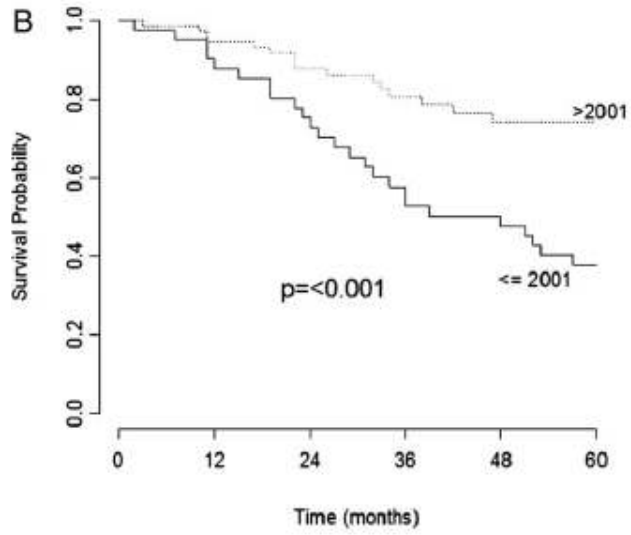
5yr overall survival
Early – 48%
Late – 66%

Radicality of surgery – “completeness of resection”

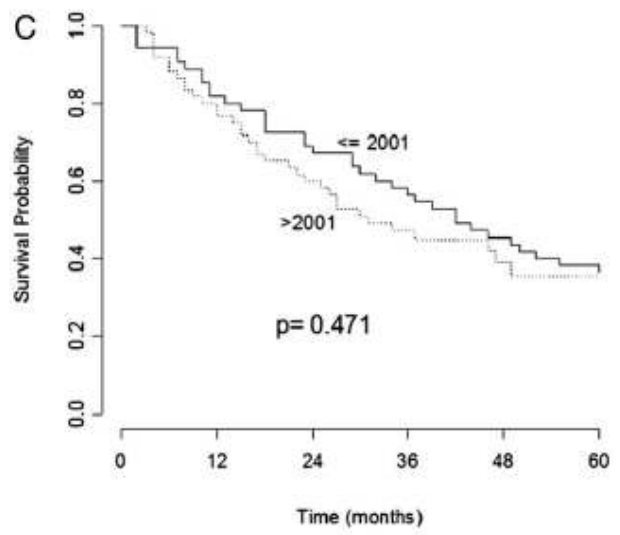
Grade 1



Grade 2



Grade 3



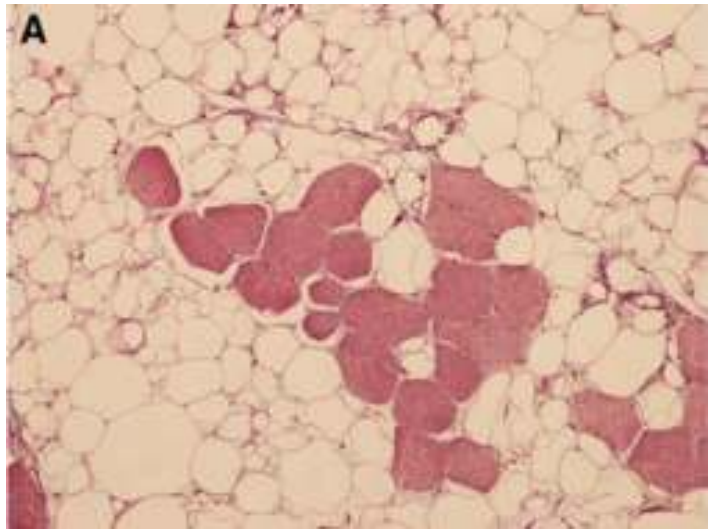
Gronchi A et al, Frontline extended surgery is associated with improved survival in retroperitoneal low- to intermediate-grade soft tissue sarcomas. Ann Oncol. 2011 Jul 16.

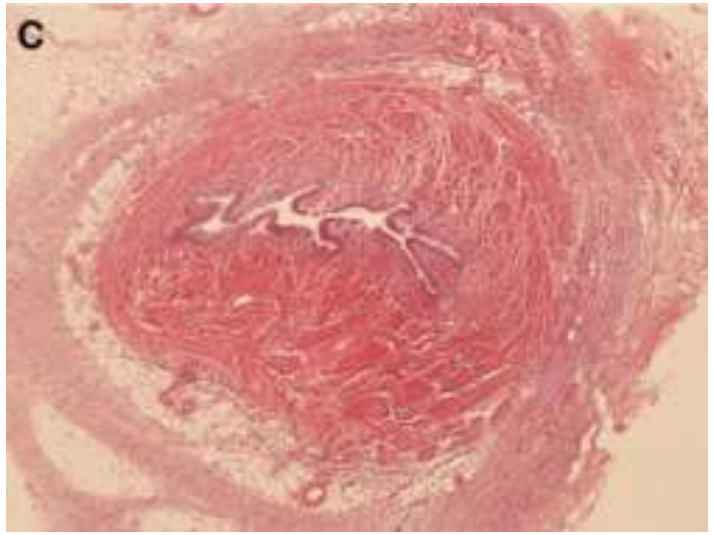
Radicality of surgery – "completeness of resection"

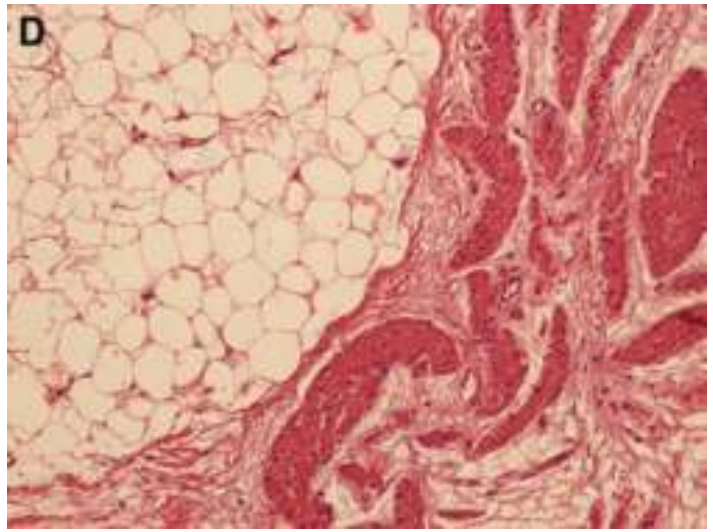
– Pathology evidence

– Meticulous pathological protocol:

- Infiltrative pattern of tumour involvement in 39/92 resected viscera (42%)
- Expansive pattern of tumour involvement in 53/92 resected viscera (58%)







Mussi C et al, Ann Surg Oncol. 2011 Aug;18(8):2136-42

Compartmental resection

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

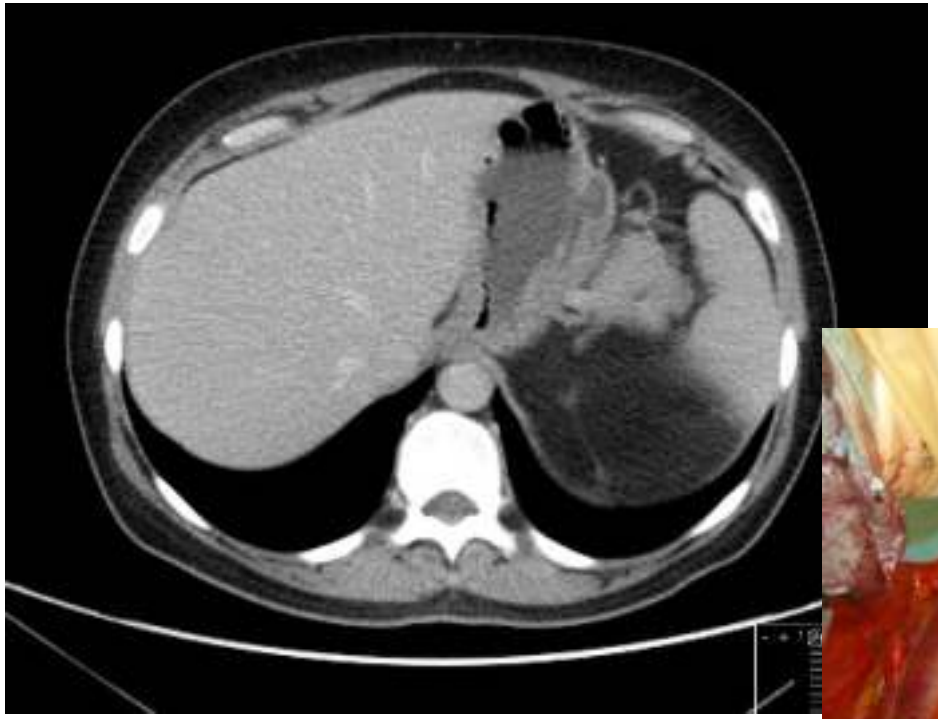
Compartmental resection

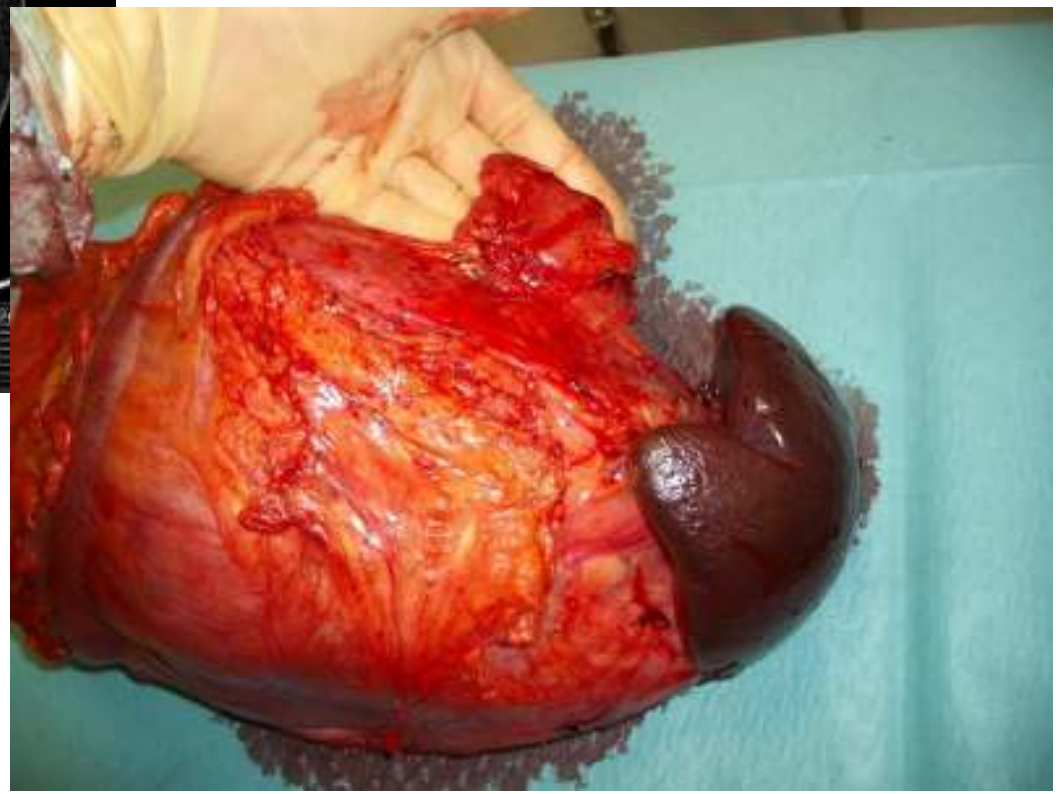
- The same quality of margins in the limb may be difficult to achieve in the retroperitoneum
- The quality of some margins may be improved in certain dimensions
- Carefully balance the oncological improvement with morbidity related to multivisceral resection

Anterior colon/mesentery

Anterior colon/mesentery

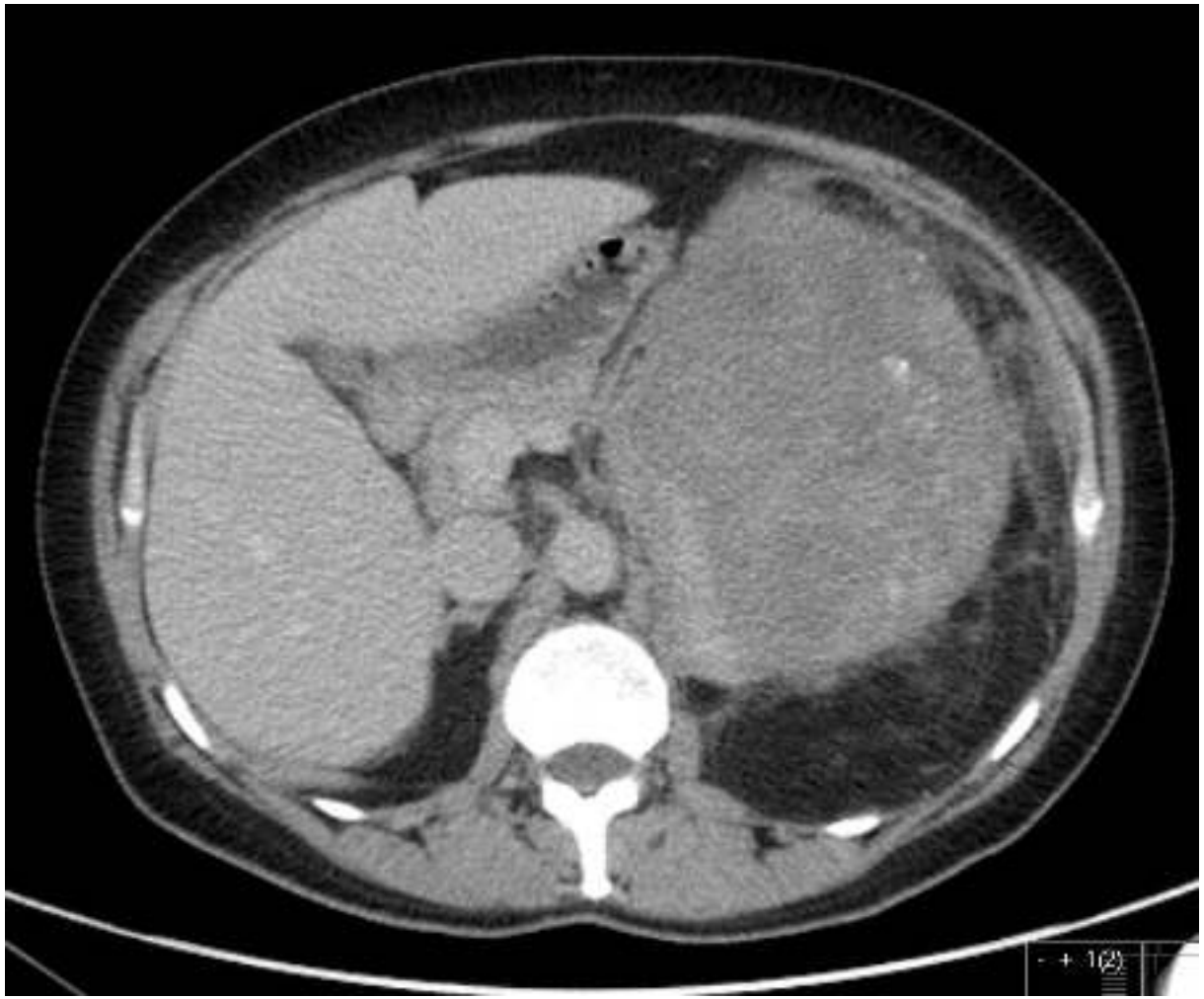
Superior margin –spleen



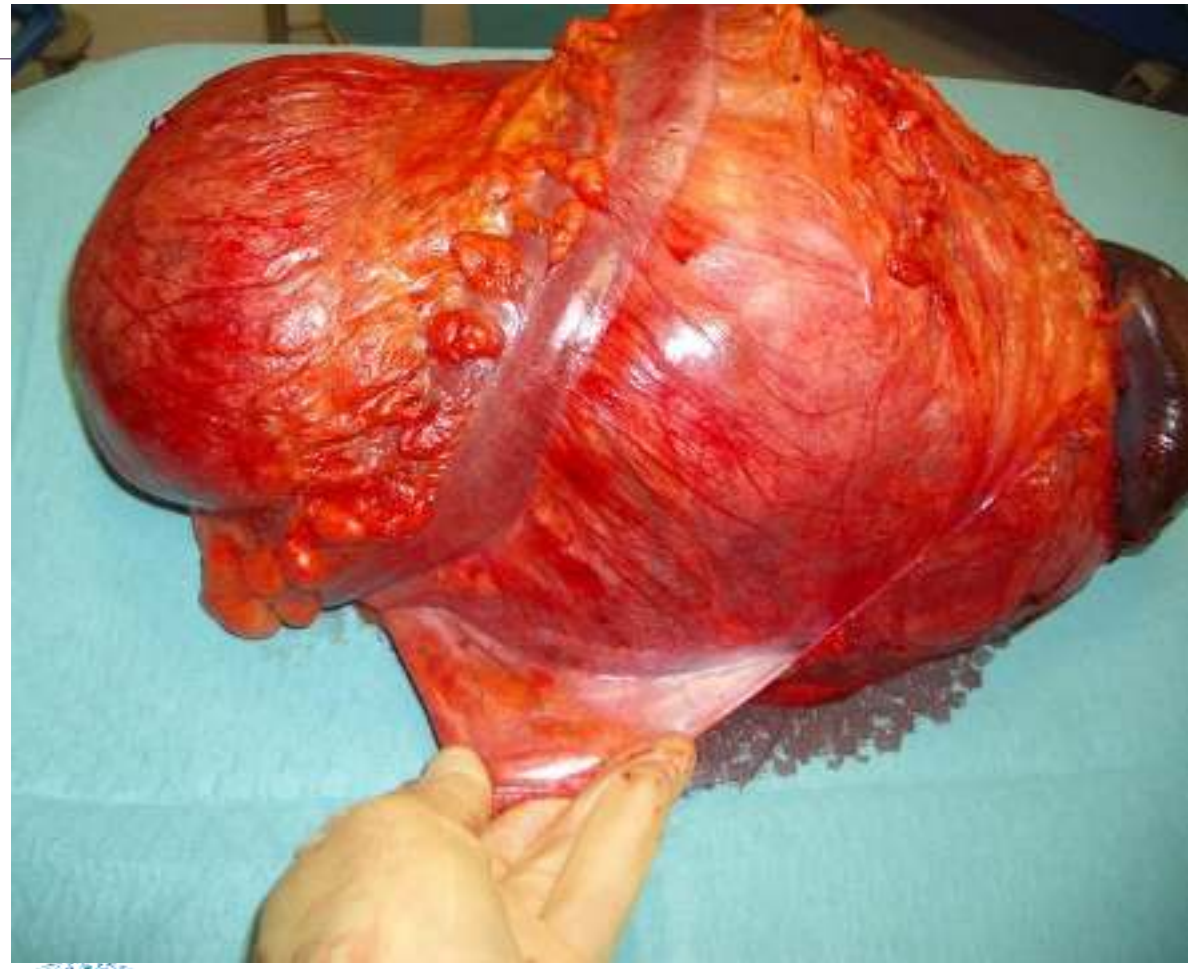


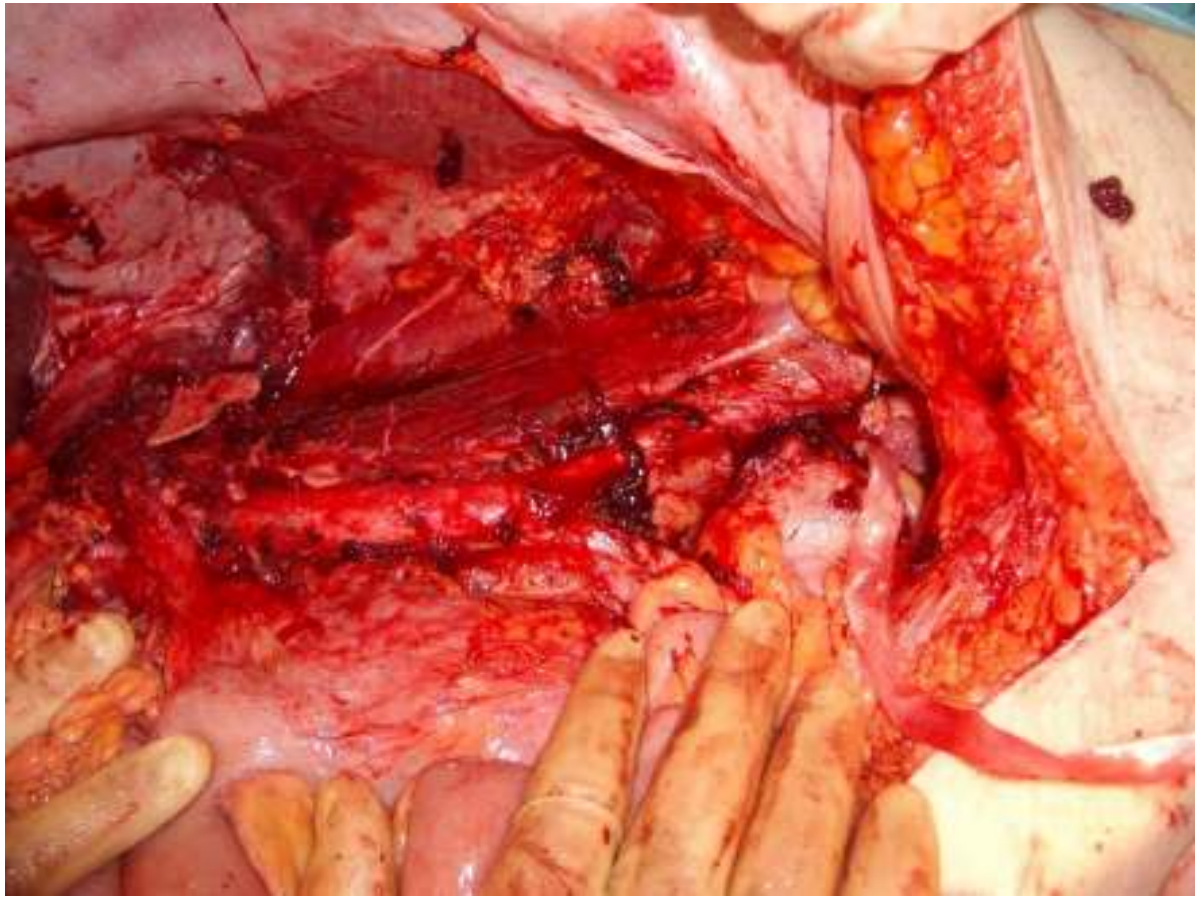
Superio-medial margin –pancreas

Lateral margin – peritoneal

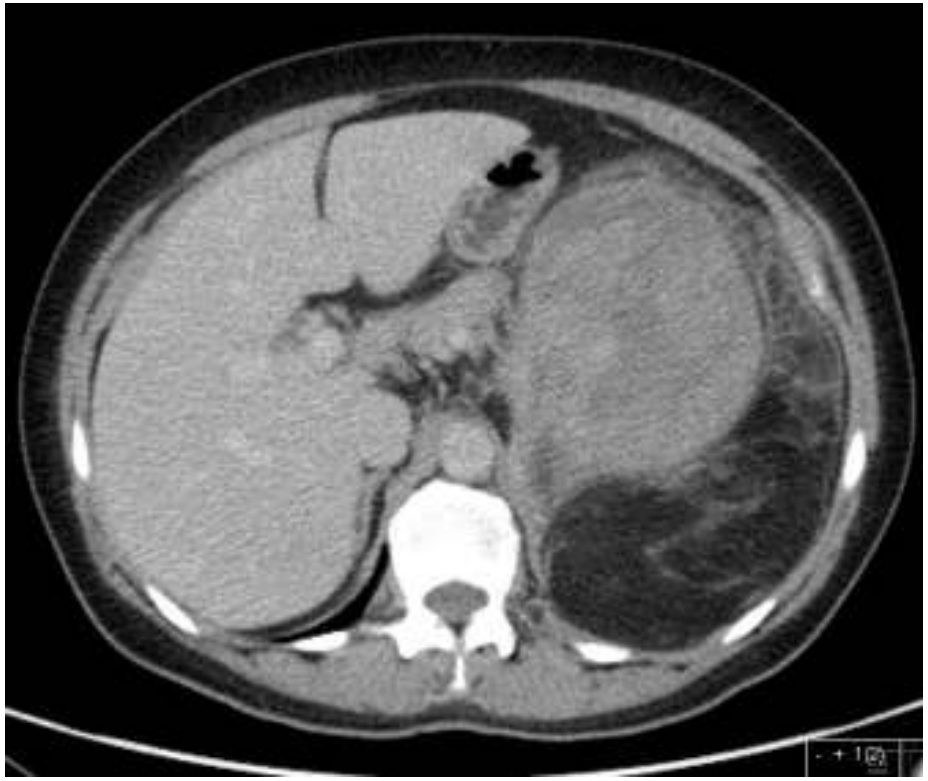


Lateral margin – peritoneal



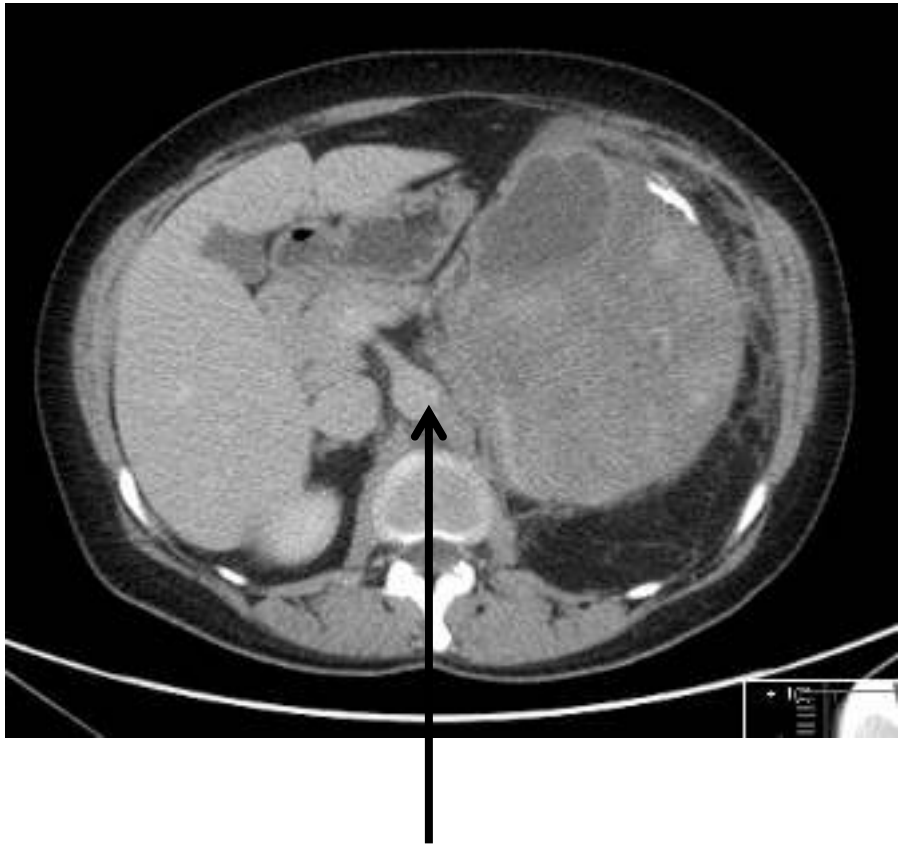


Medial margin – diaphragm crus

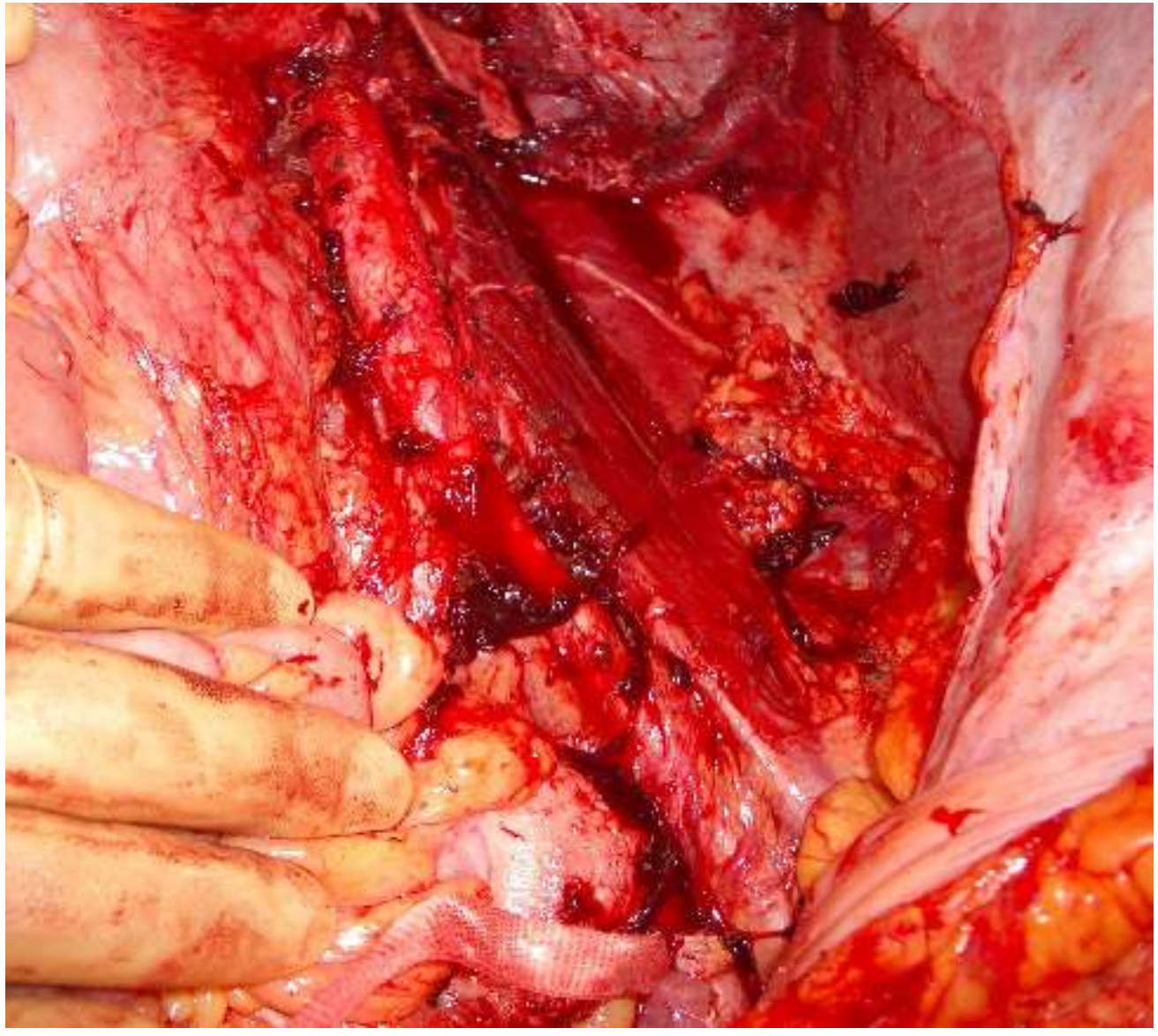


Medial margin – diaphragm crus

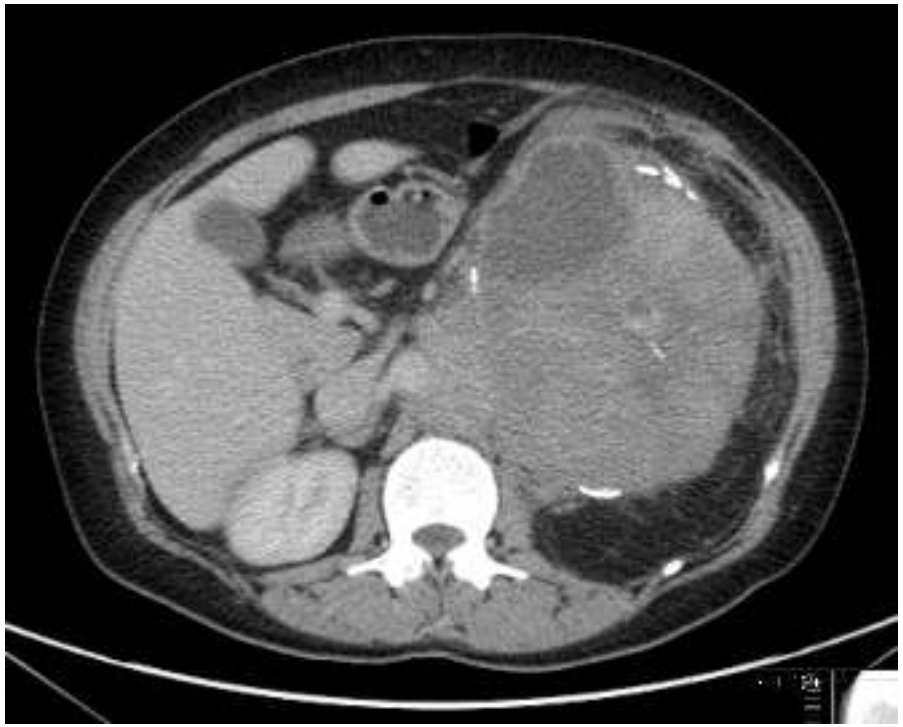
Medial margin – aorta

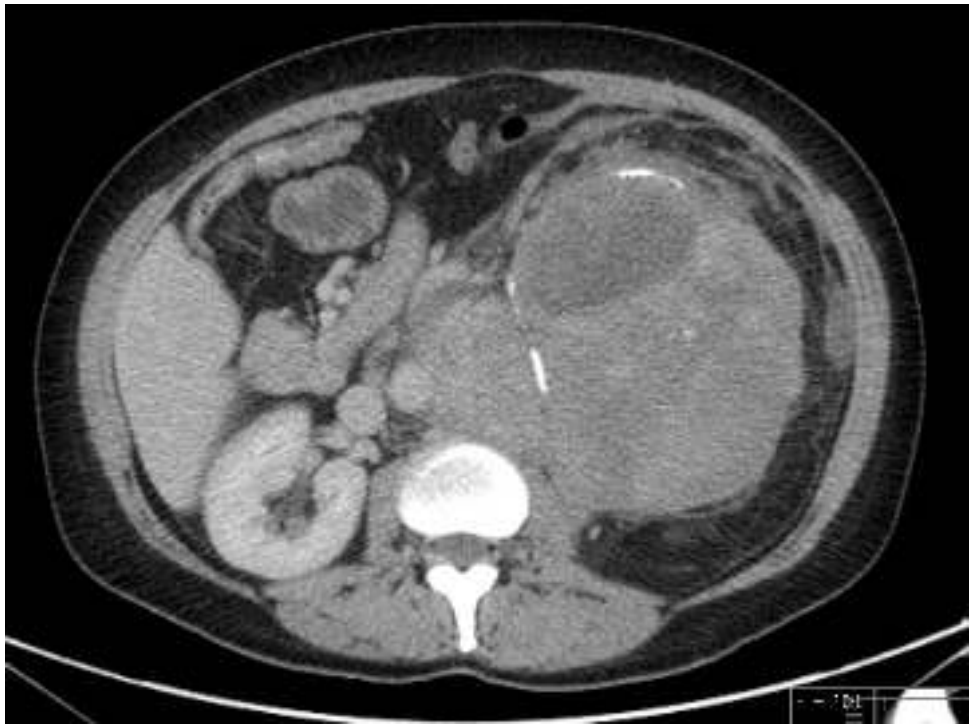


Medial margin – aorta



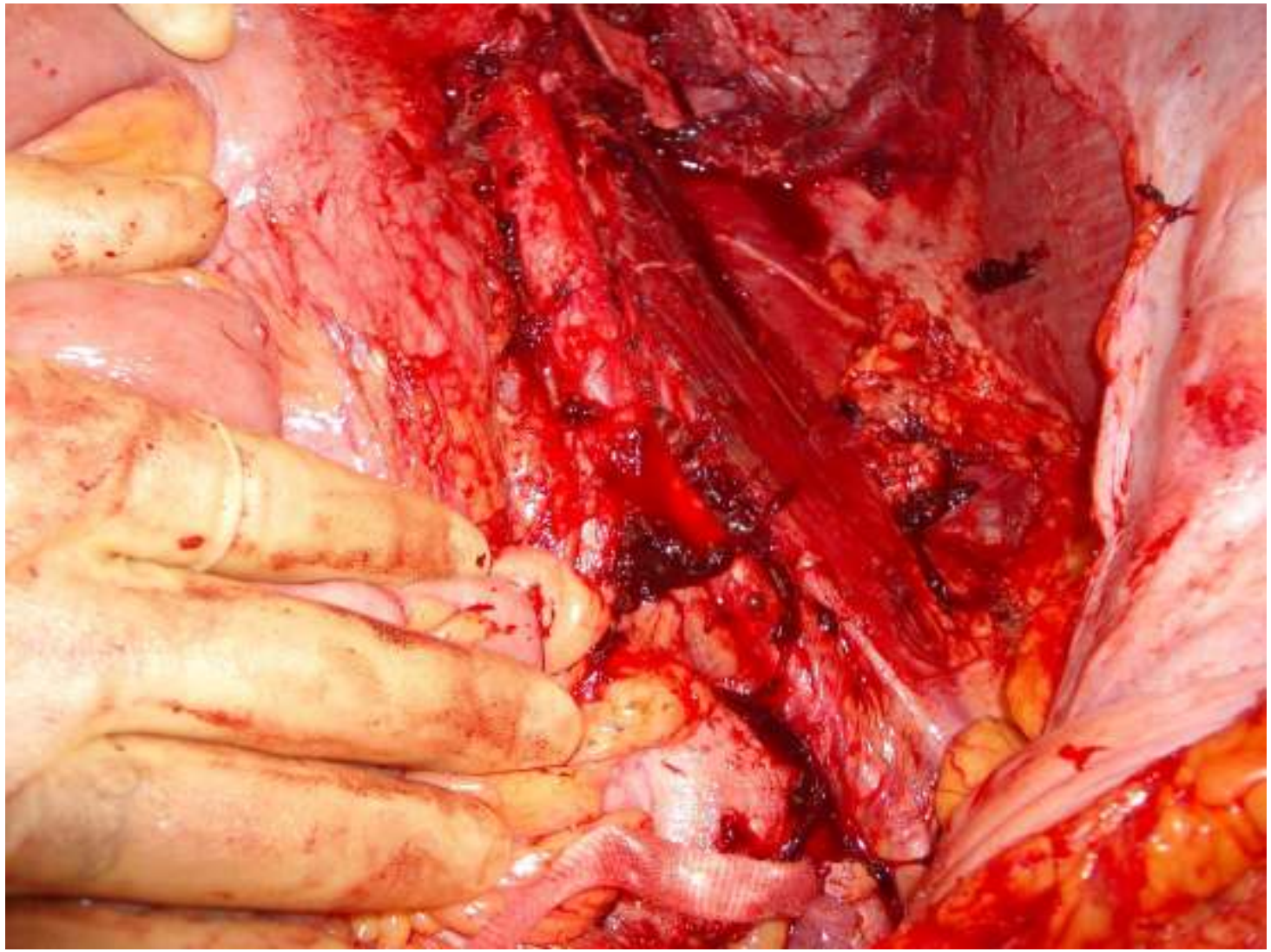
Posterior margin –psoas muscle



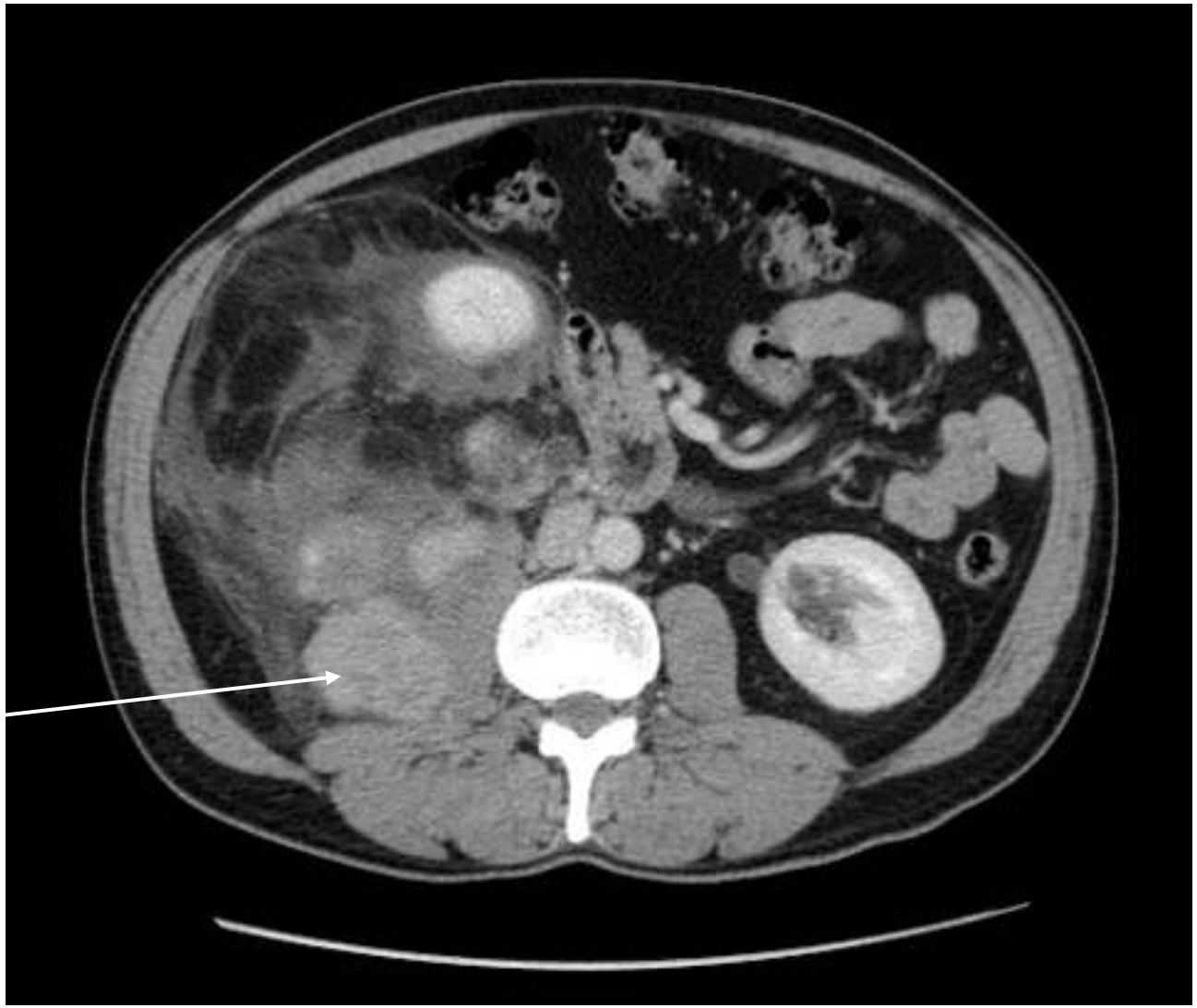


Psoas muscle or psoas aponeurosis?

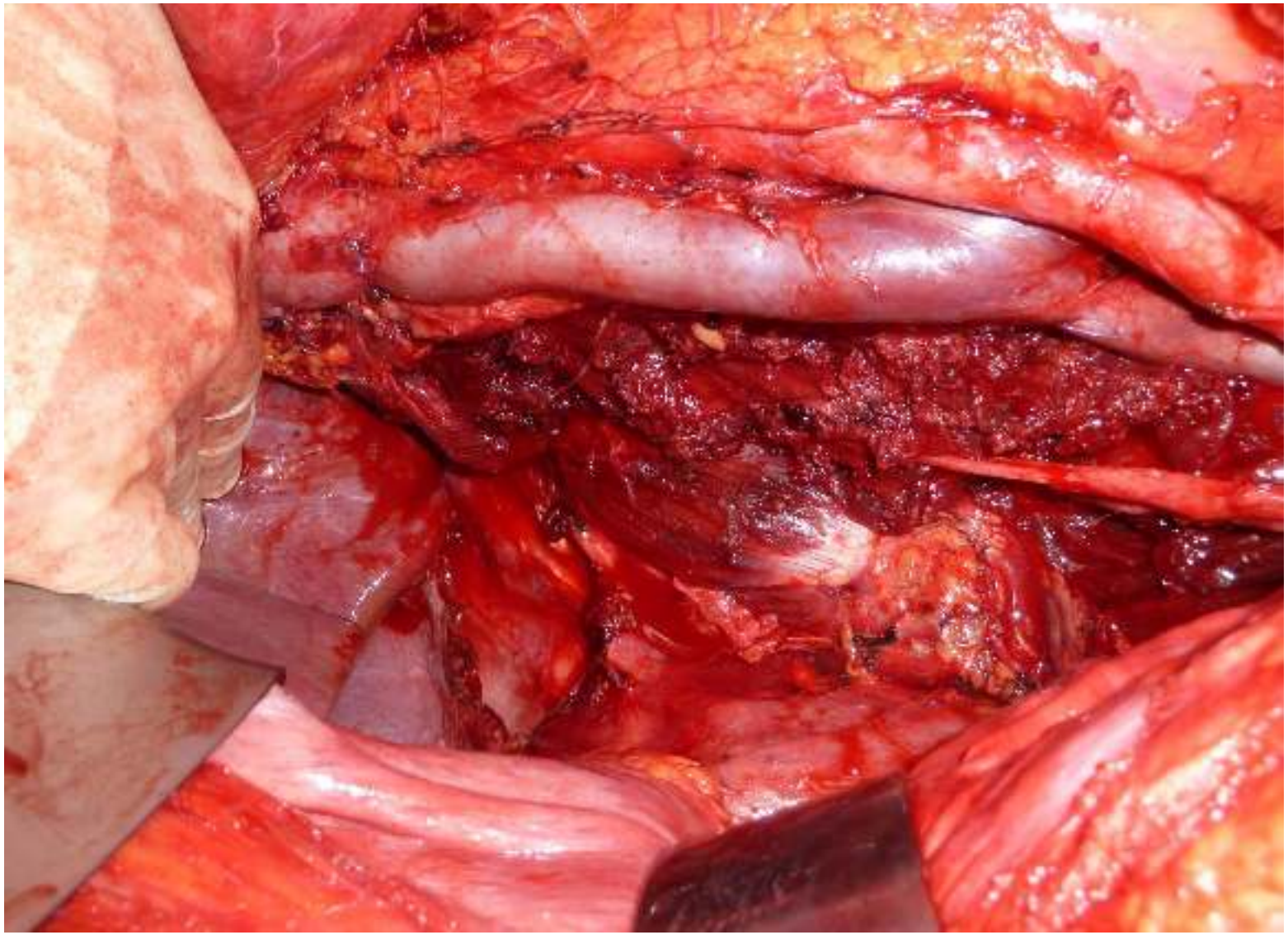
Posterior margin –psoas muscle



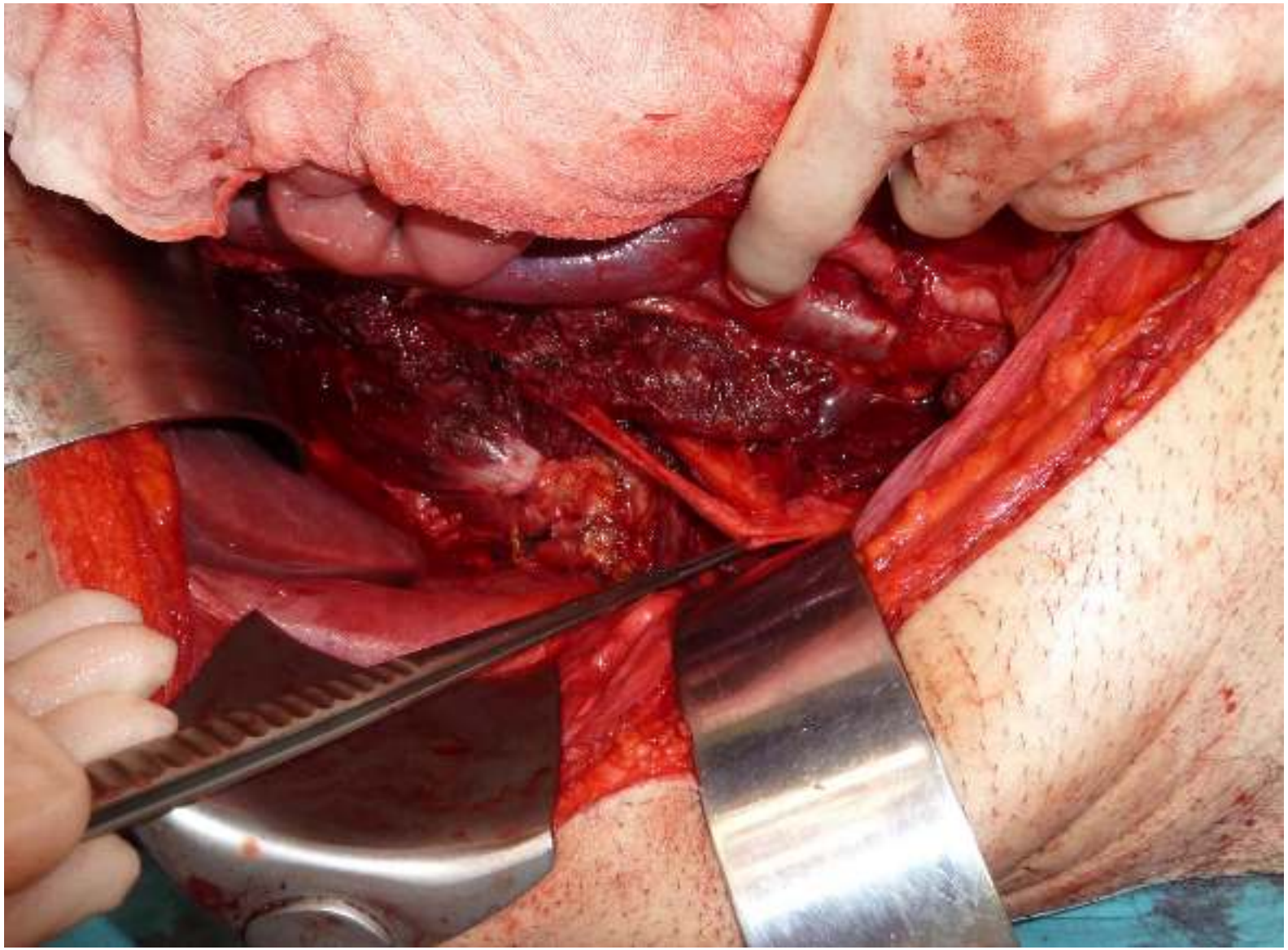
Posterior margin –psoas muscle



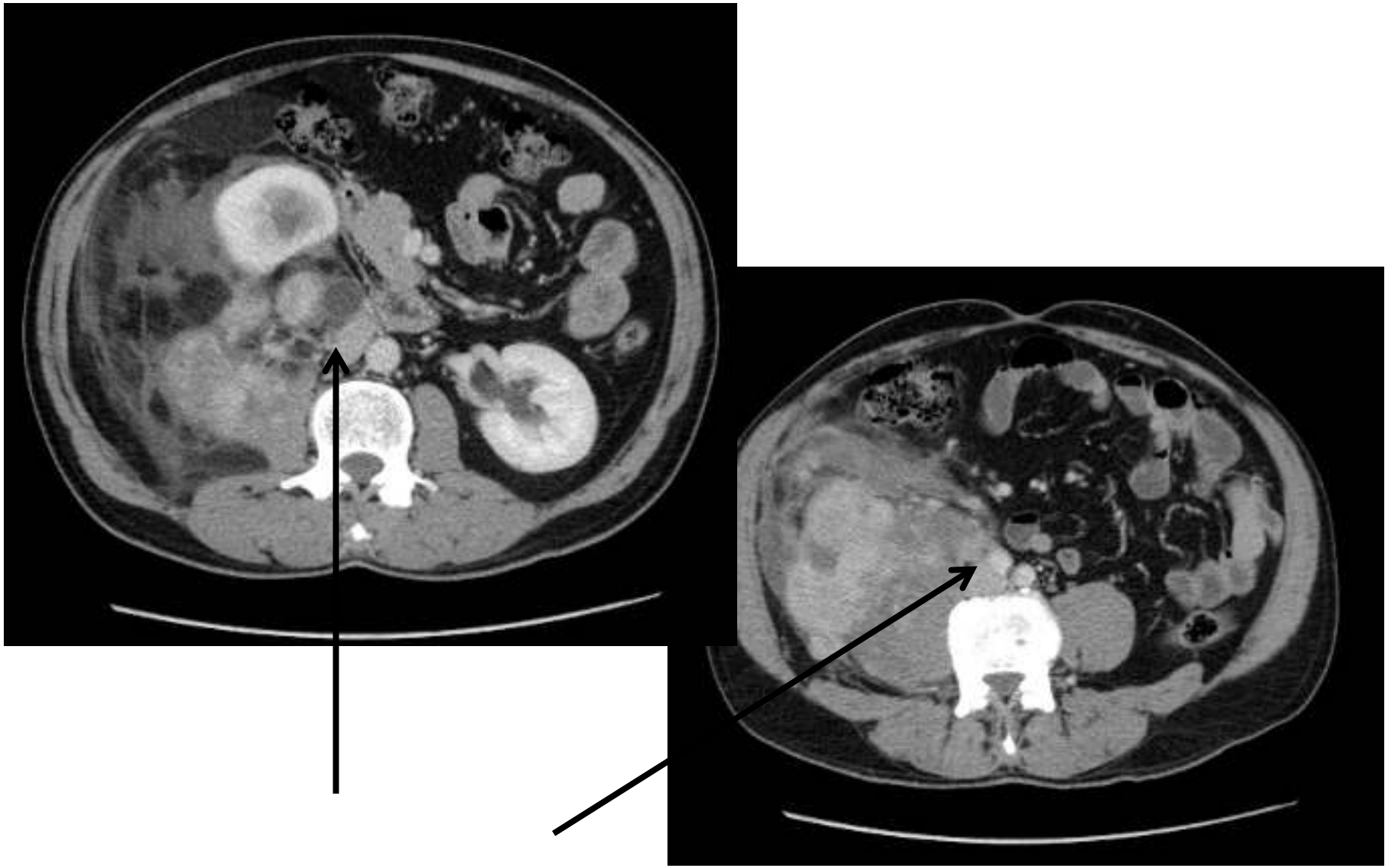
Posterior margin –psoas muscle



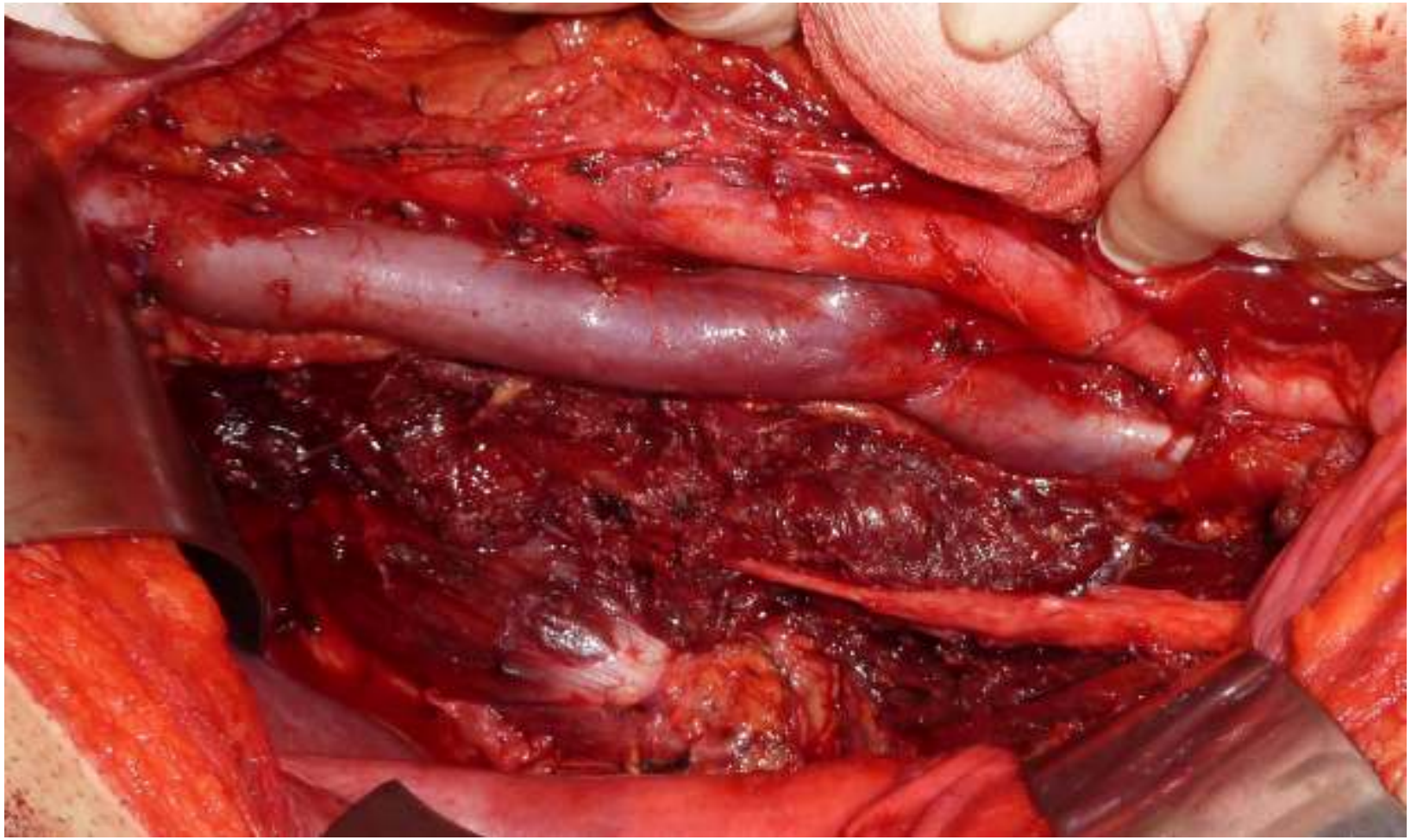
Posterior margin –psoas muscle



Medial margin - Inferior vena cava, aorta



Medial margin - Inferior vena cava

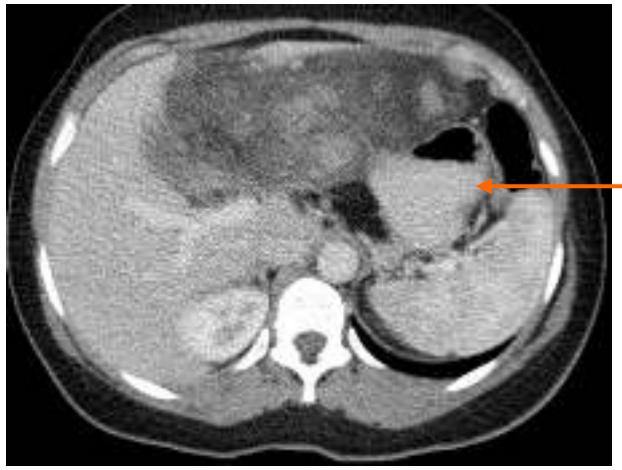


Direct infiltration

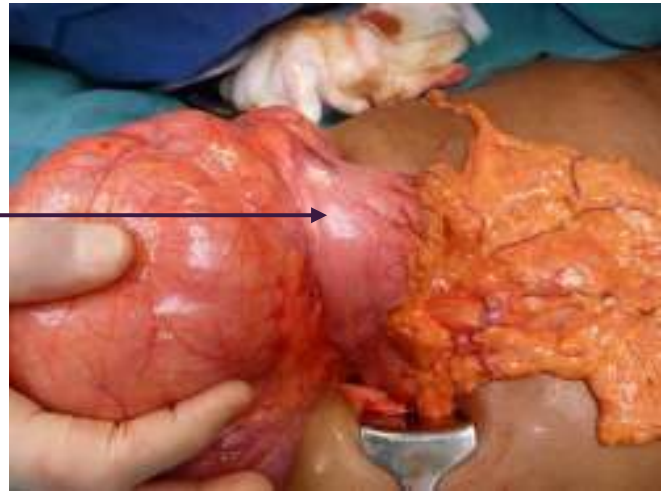
Organs or structures should be resected only if directly infiltrated:

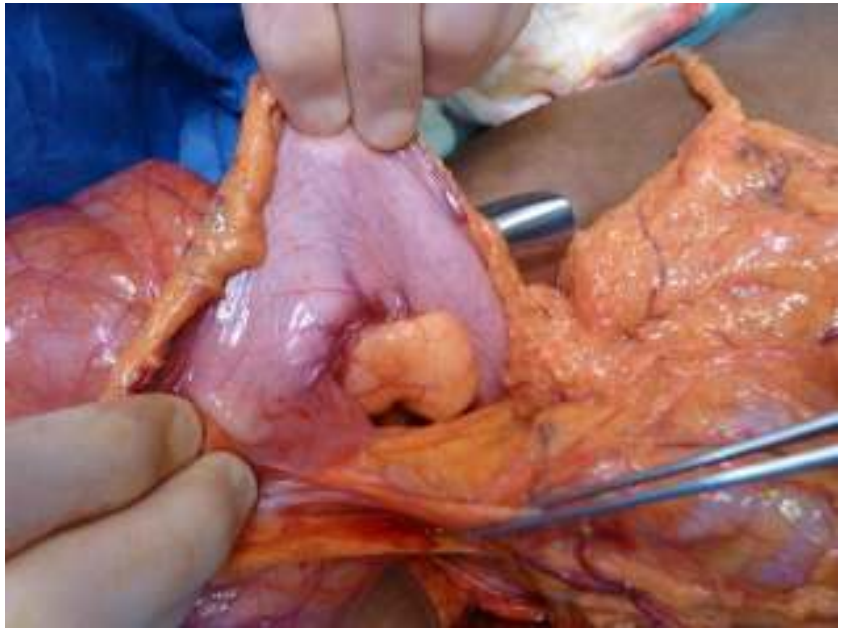
duodenum, head of pancreas, liver, stomach, diaphragm, major abdominal vessels, bladder/rectum and nerves, bone.

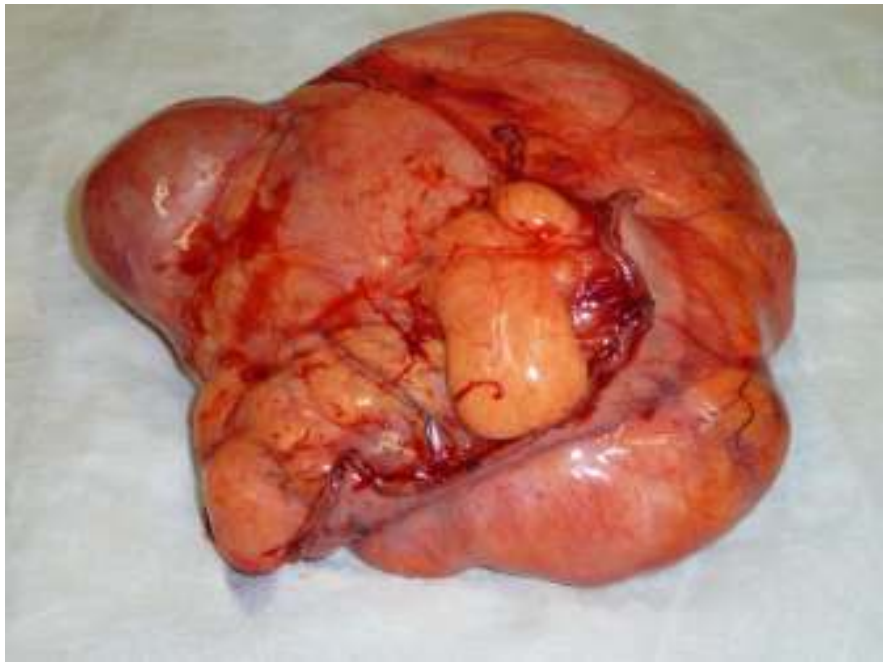
Direct infiltration



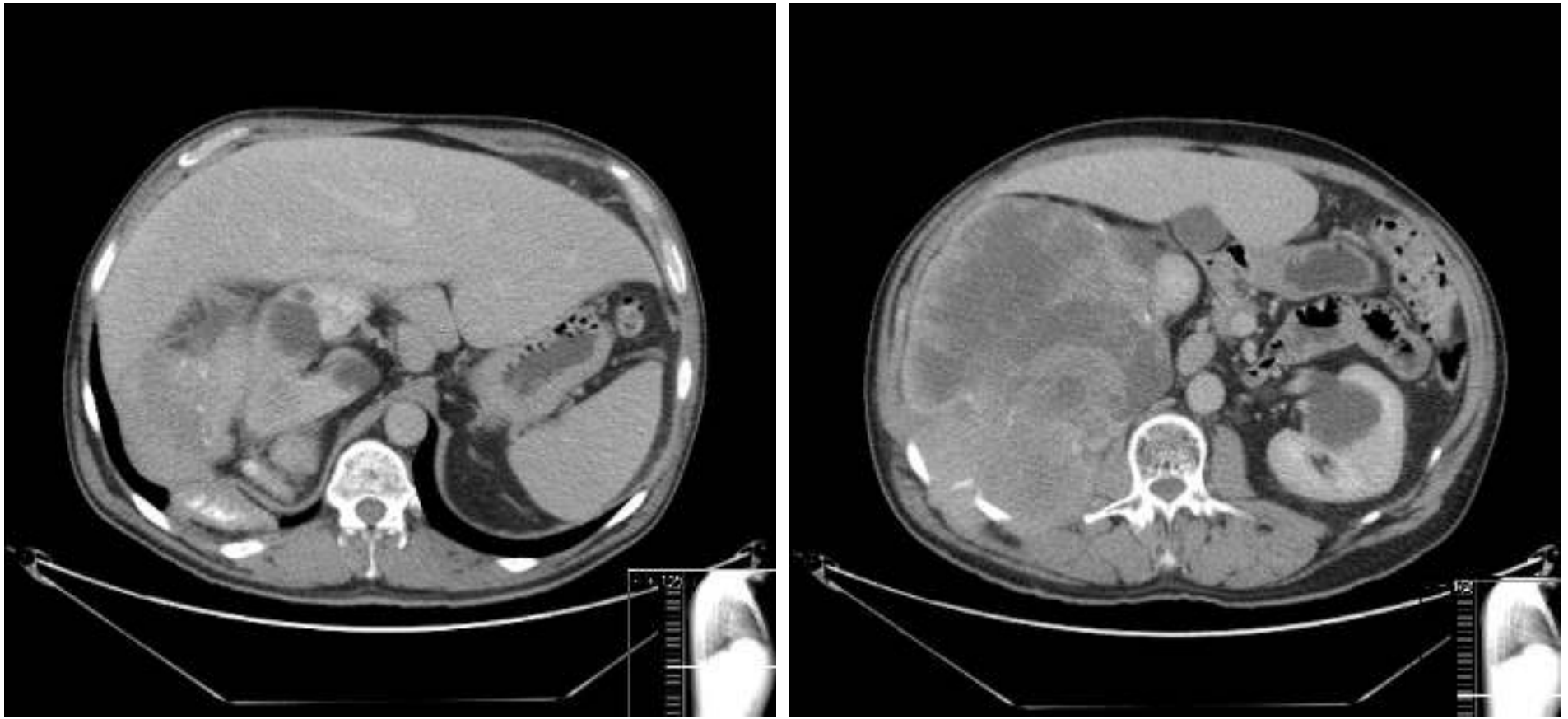
Stomach







Dedifferentiated liposarcoma



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

Surgical extent according to biologic behaviour

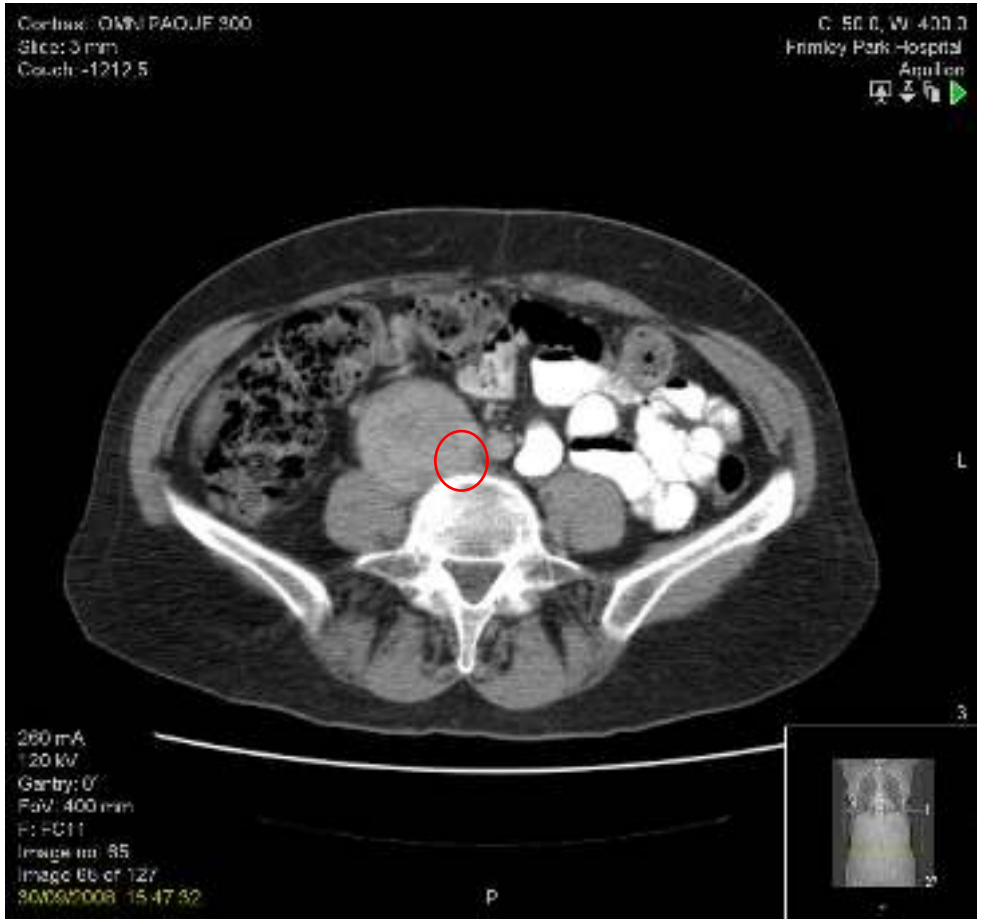
For leiomyosarcomas, with more clearly defined borders,

- organs that are closely adjacent, but not directly adherent to/invaded by the tumour can potentially be preserved, provided this does not result in an avoidable positive margin.

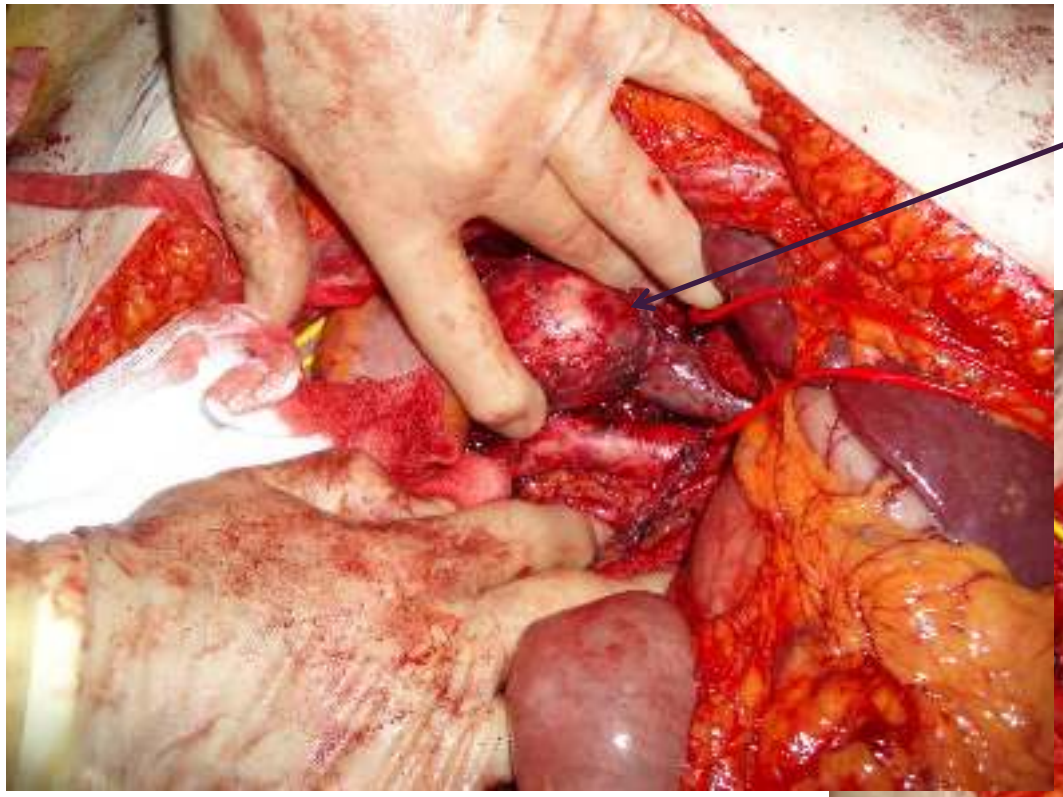
- In the case of LMS arising from a major vein, specific attention should be directed to achieving microscopically negative margins on the vein of origin (longitudinal margins).

Surgical extent according to biologic behaviour

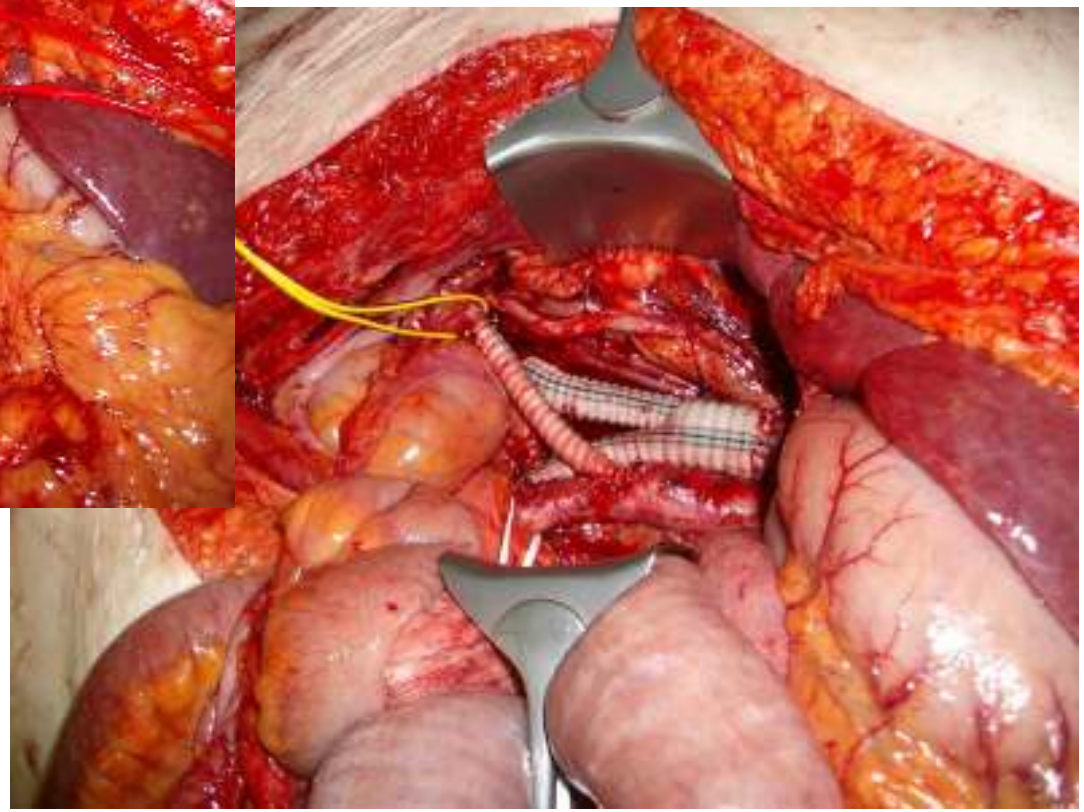
IVC Leiomyosarcoma



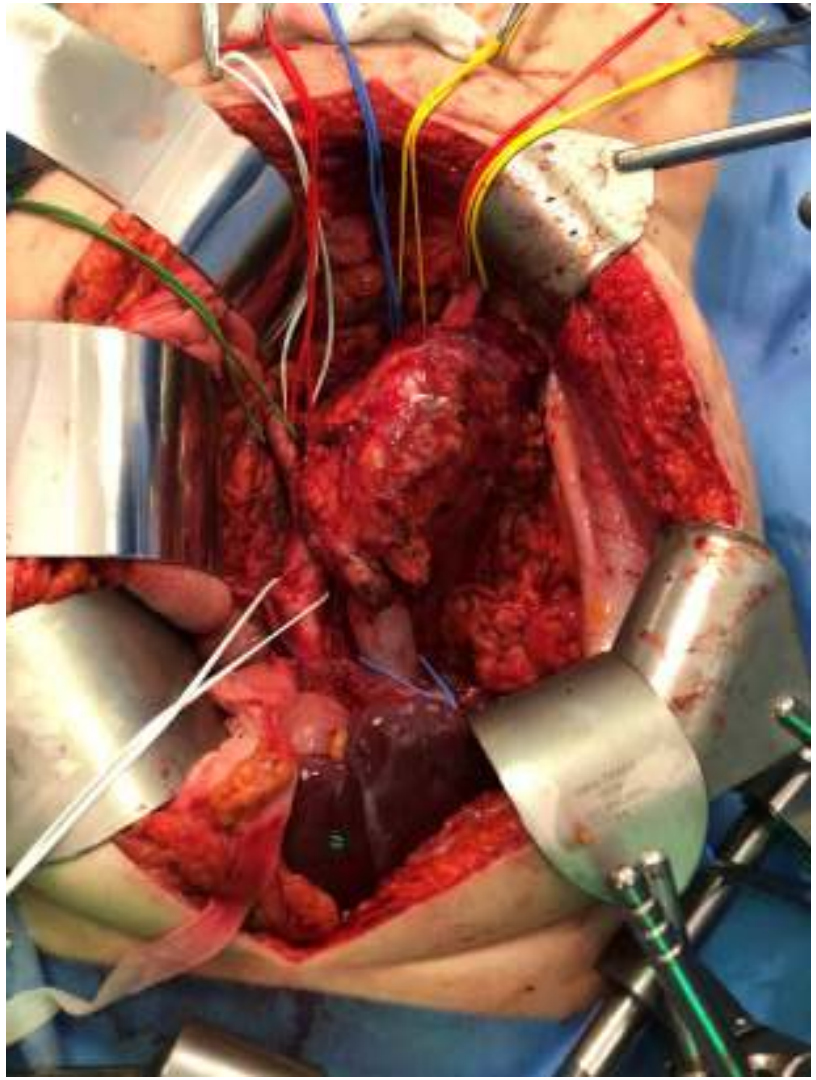
Surgical extent according to biologic behaviour

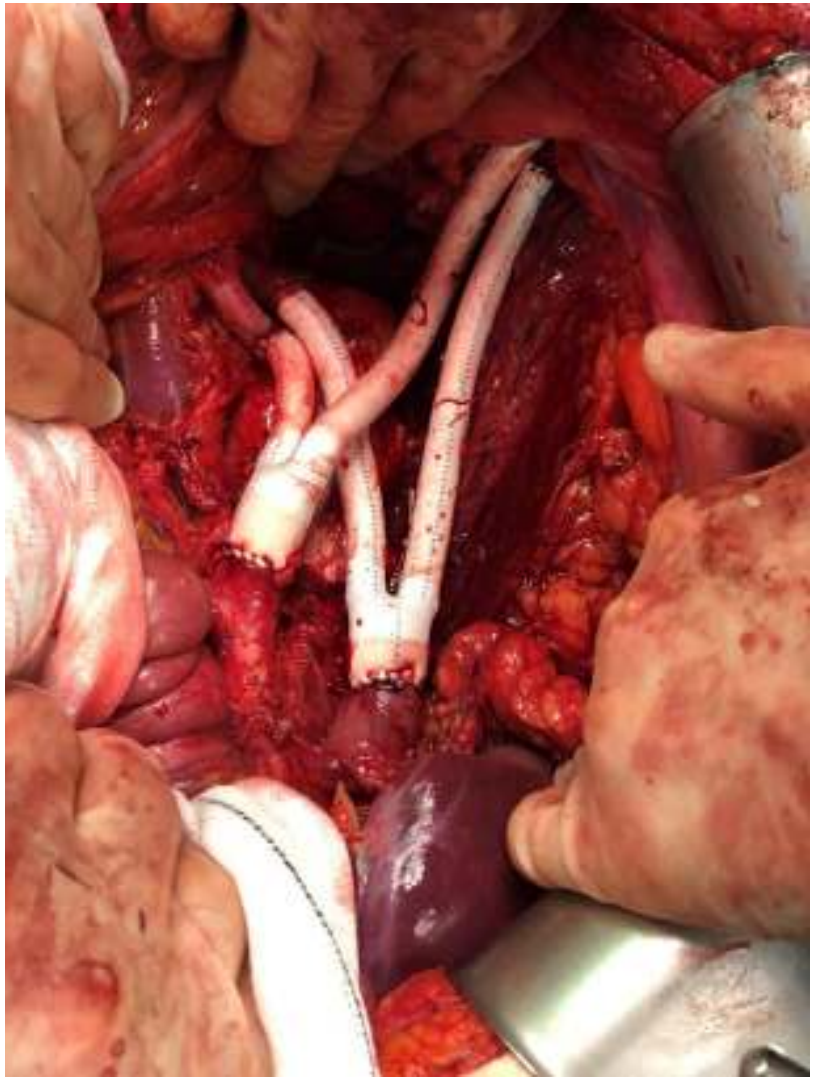


IVC leiomyosarcoma



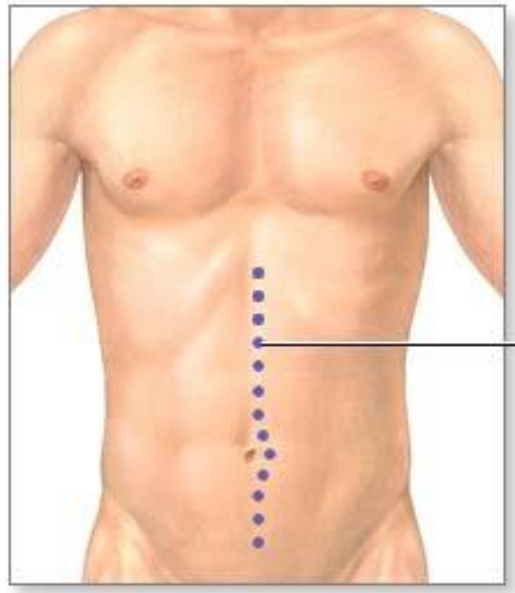
Surgical extent according to biologic behaviour





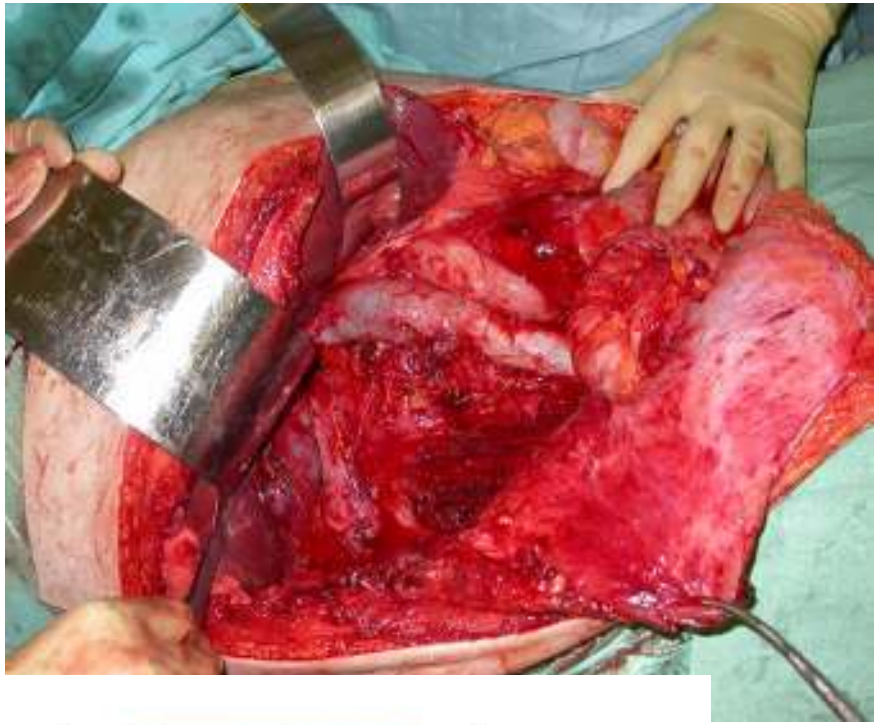
General technical aspects

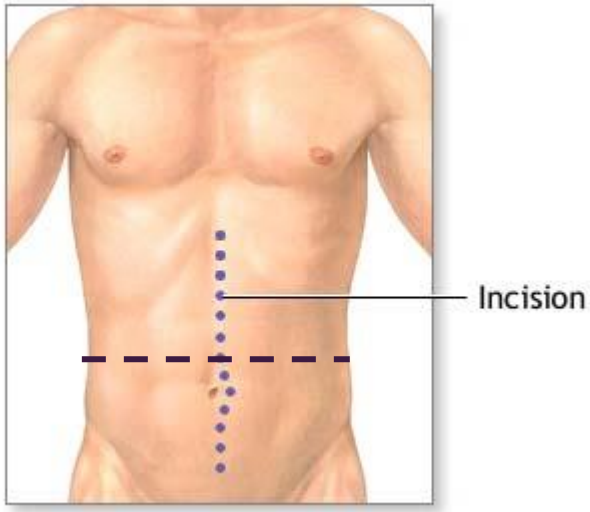
– Incisions



General technical aspects

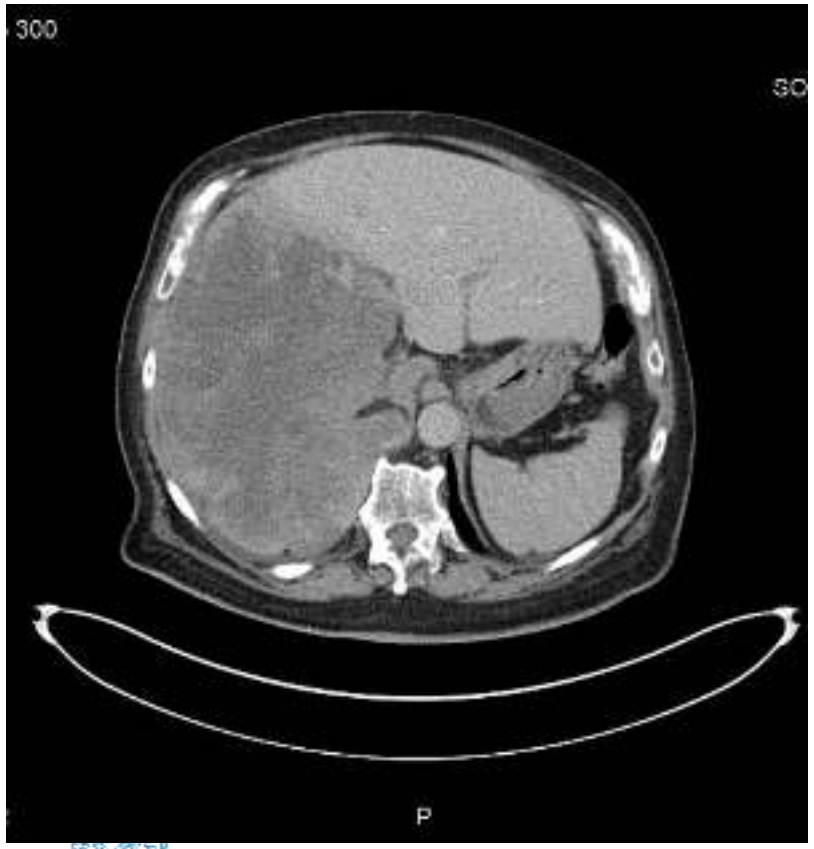
– Incisions

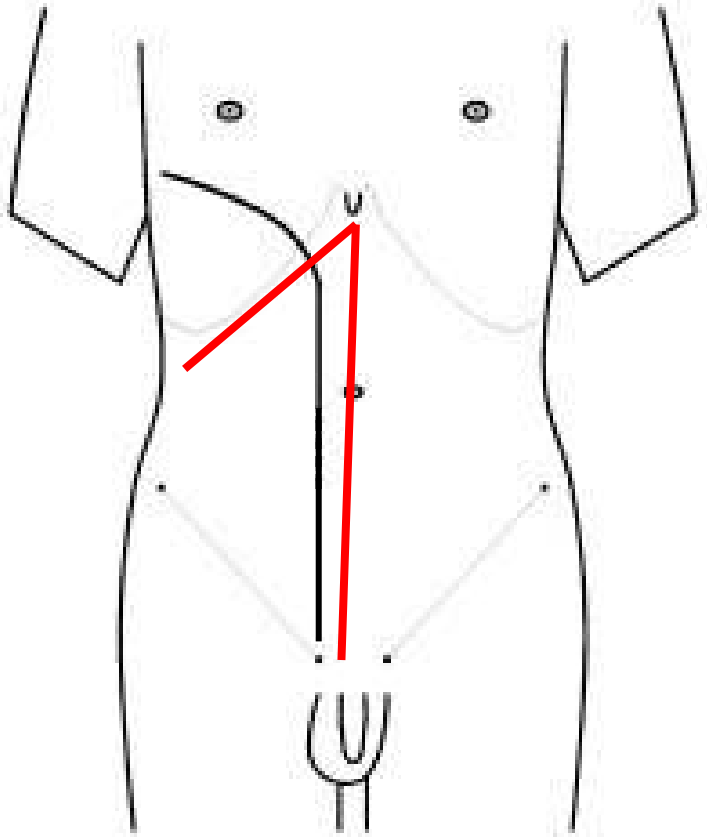




General technical aspects

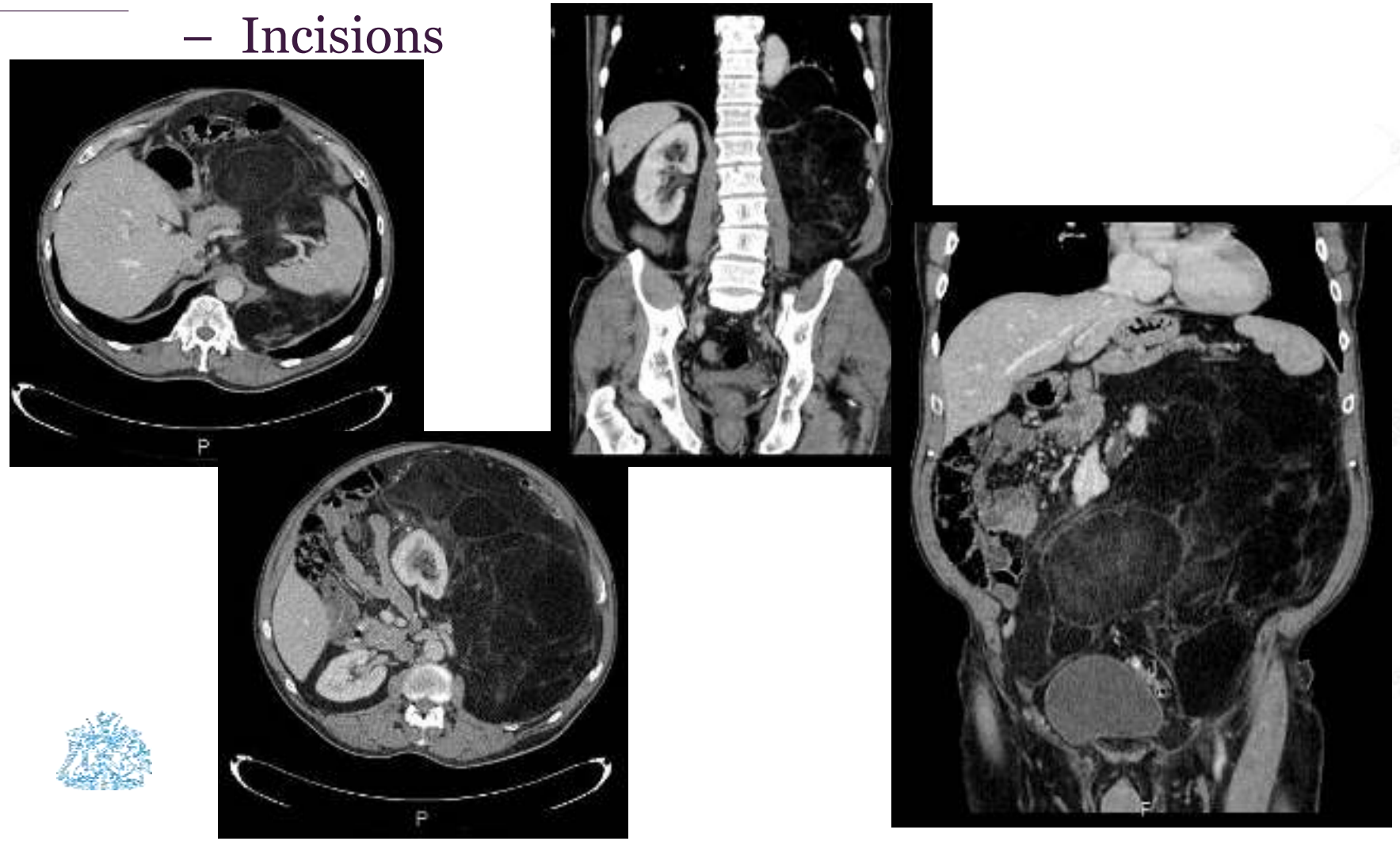
– Incisions

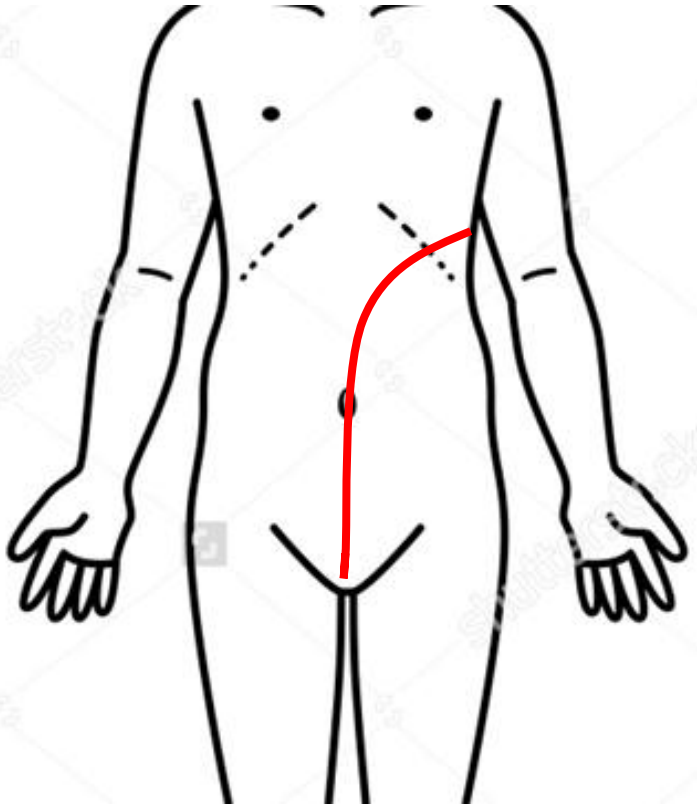




General technical aspects

– Incisions

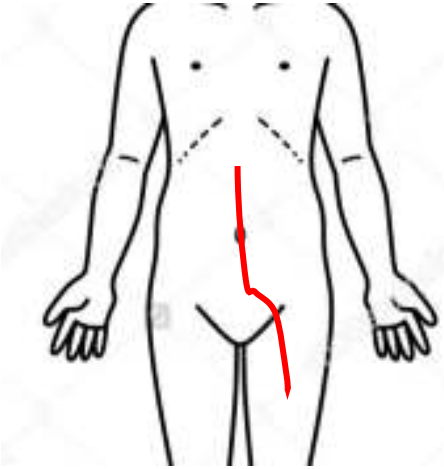


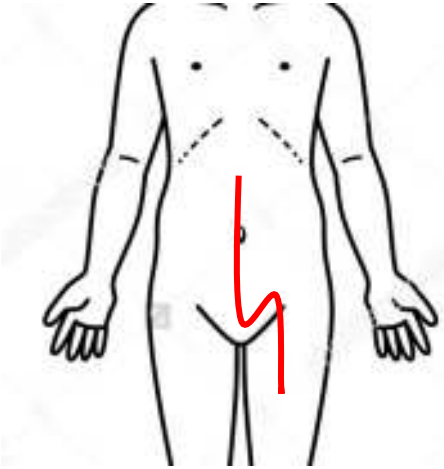


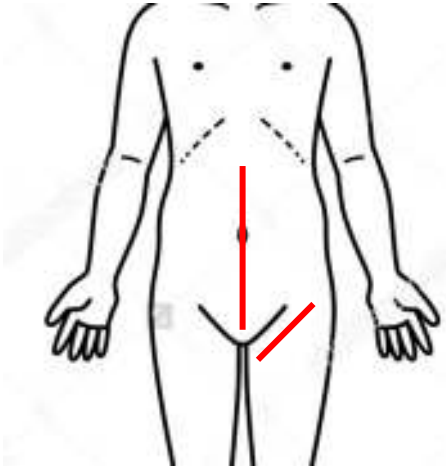
General technical aspects

Incisions through the groin into the proximal thigh:

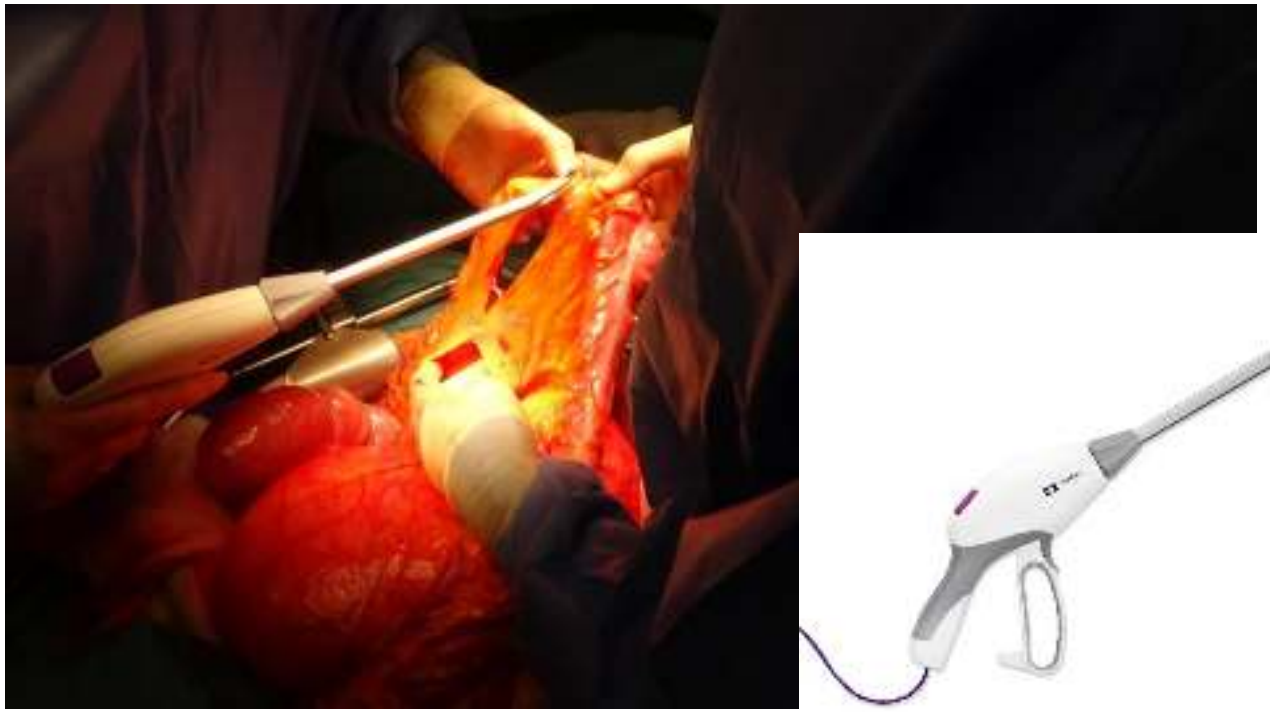
- a midline incision and an S-shaped incision over the iliac vessels
- L- or Z-shaped incisions (lower midline with transverse or oblique division of the rectus abdominis muscle) may be chosen
- midline + groin incision







Toys and tools

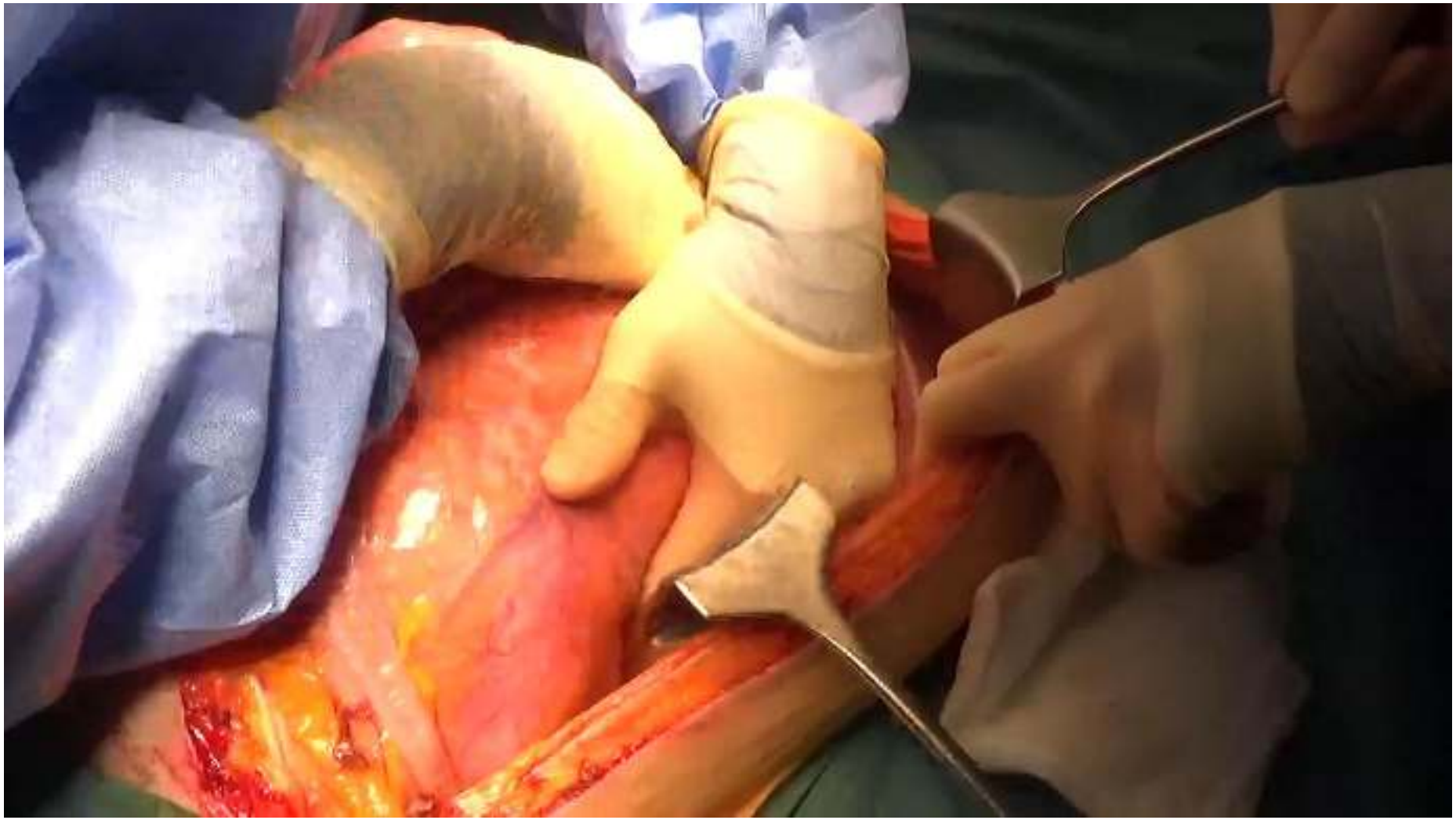


Toys and tools

Stepwise approach

Exploration – decision based on intra-operative findings and preoperative scans which organs to include in the resection

Stepwise approach



Stepwise approach

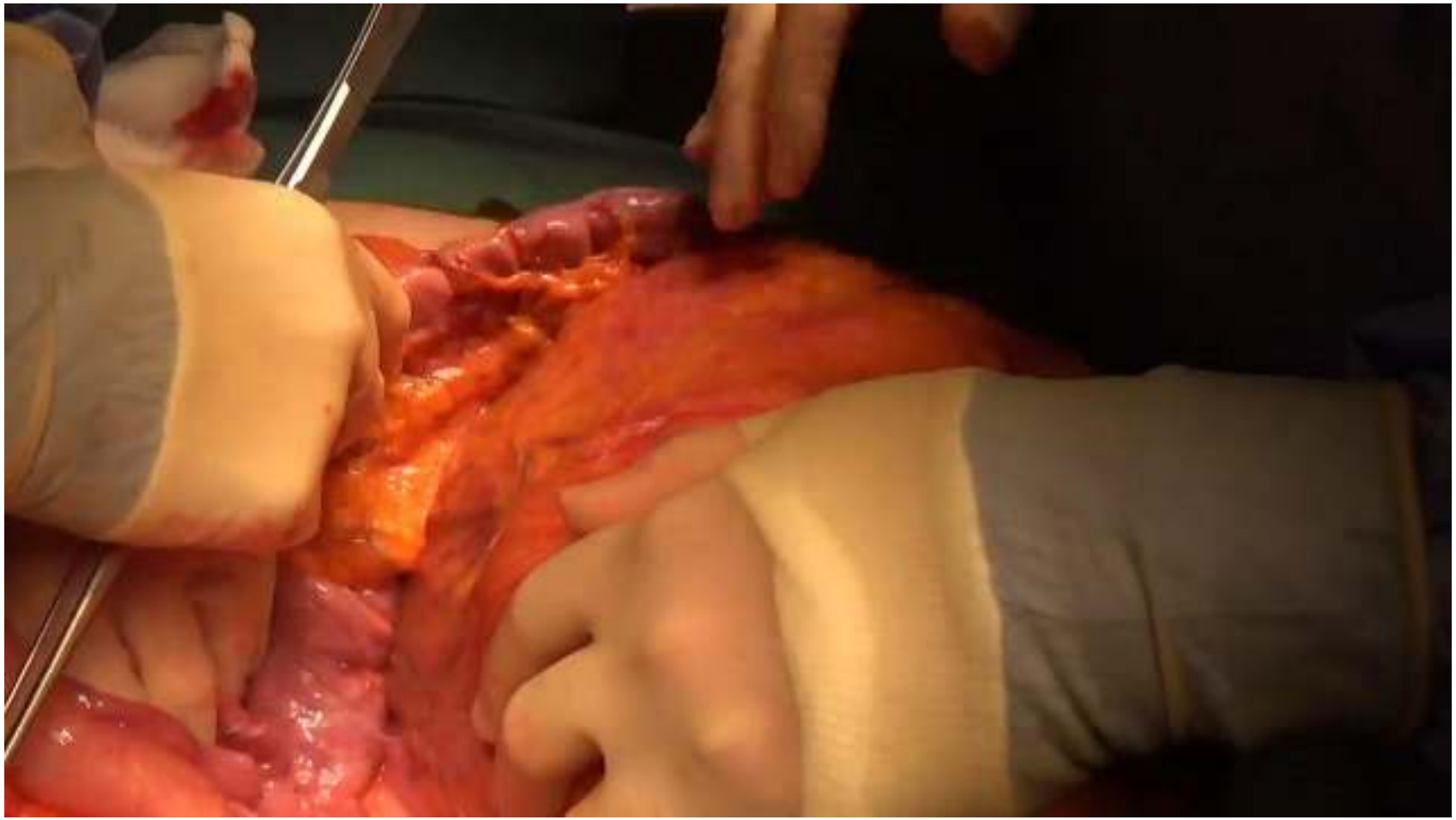
Exploration – decision based on intra-operative findings and preoperative scans which organs to include in the resection

The first step is liberation of the root of mesentery.

- Colo-epiploic separation is performed and transverse colon divided usually with preservation of middle colic artery

Division of rectosigmoid junction (left) or terminal ileum (right)

Stepwise approach



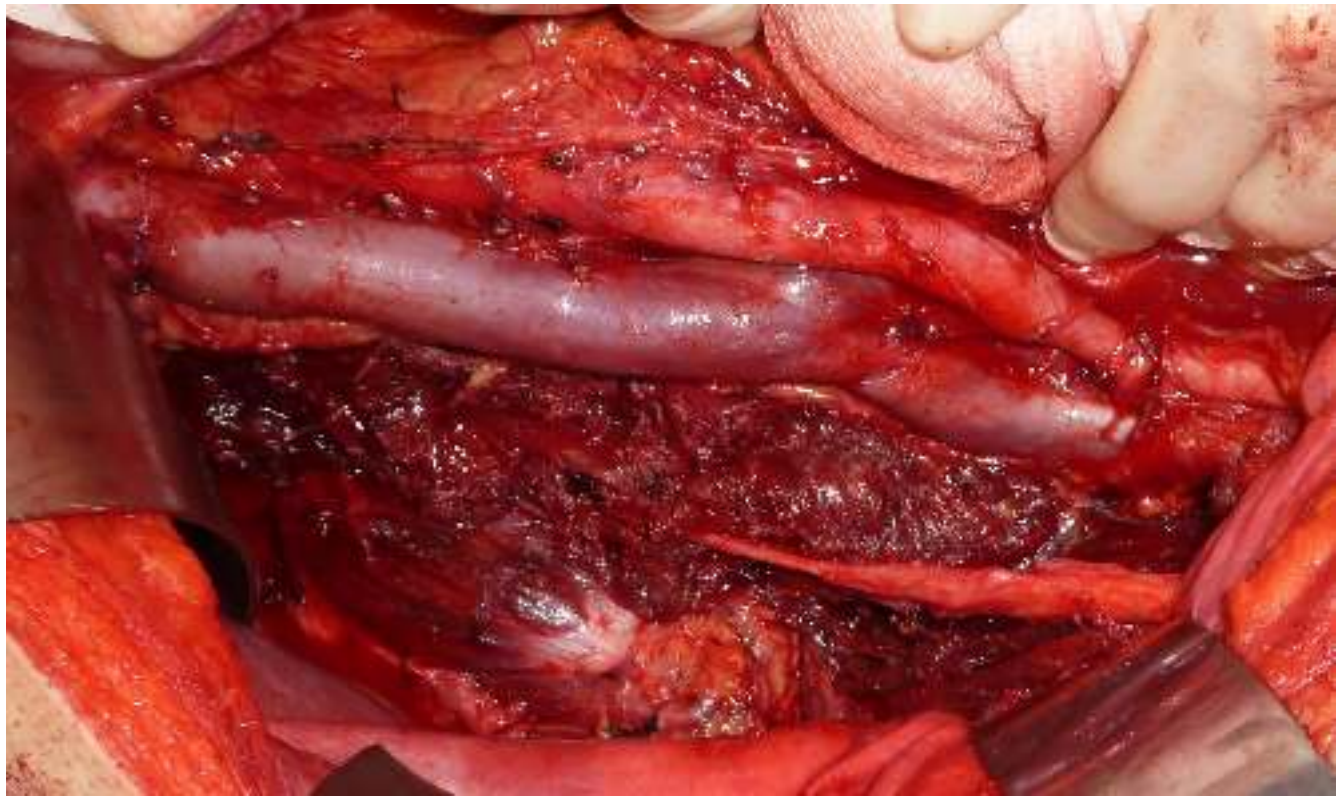
Stepwise approach

- The major vessels are then accessible and can be prepared on the medial side of the tumour. This should be freed from midline to periphery in order to have major vascular structures prepared and avoid undesirable tension at the end of surgery with the risk of vascular tears.

- It is often easier to start dissection from the iliac vessels up to the proximal aorta/vena cava with a sub adventitial dissection. This adventitia is left on the side of the tumour and serves an anatomical barrier.

Stepwise approach

Stepwise approach



Stepwise approach

Duodenum

- Left: The duodenojejunal junction may be displaced and is seldom invaded, and it may be detached from the tumour surface which usually remains covered by the root or medial edge of left mesocolon.
- If invaded or tightly adherent to the tumour, the third/fourth portion of the duodenum and the proximal jejunum just distal to the ligament of Treitz are divided, leaving this duodenojejunal junction attached to the surface of the tumour.
- At the time of reconstruction, a side-to-side anastomosis between the second portion of the duodenum and the proximal remaining jejunum is performed.

Stepwise approach

Duodenum

- Right: A Kocher manoeuvre is performed to free the duodenum and head of the pancreas, providing complete access to the inferior vena cava.
- This is frequently a critical point for right-sided tumours.
- The duodenum and head of the pancreas are often dissected off the surface of the tumour, leaving a very thin layer on the tumour, if any margin at all.
- One could argue that performing a pancreaticoduodenectomy would provide a better margin. Nevertheless, pancreaticoduodenectomy is not routinely performed unless there is a clear infiltration, given the concerns on morbidity this procedure may add.

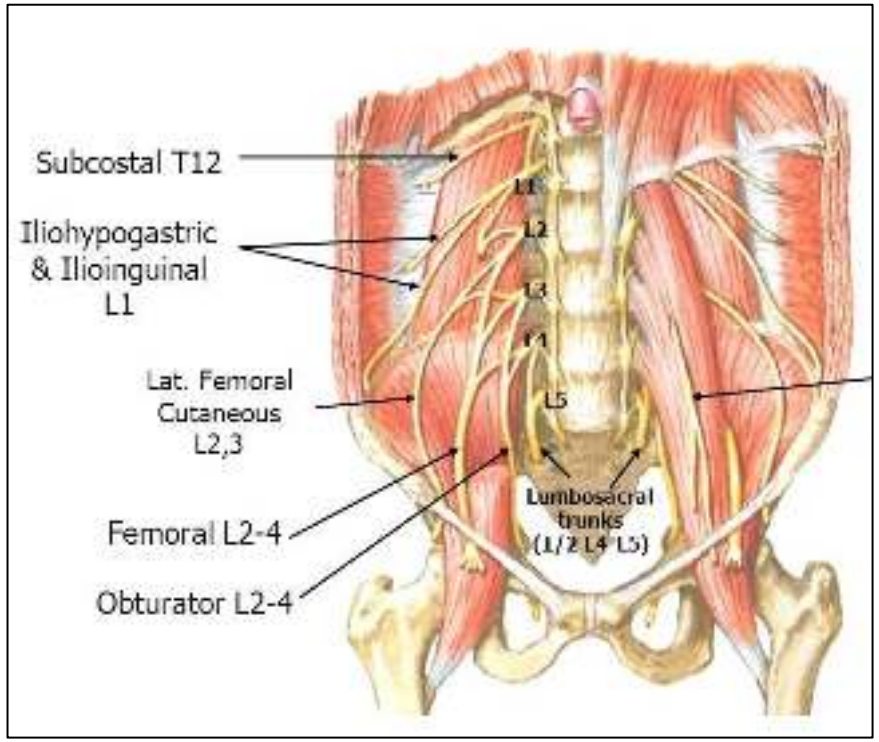
Stepwise approach

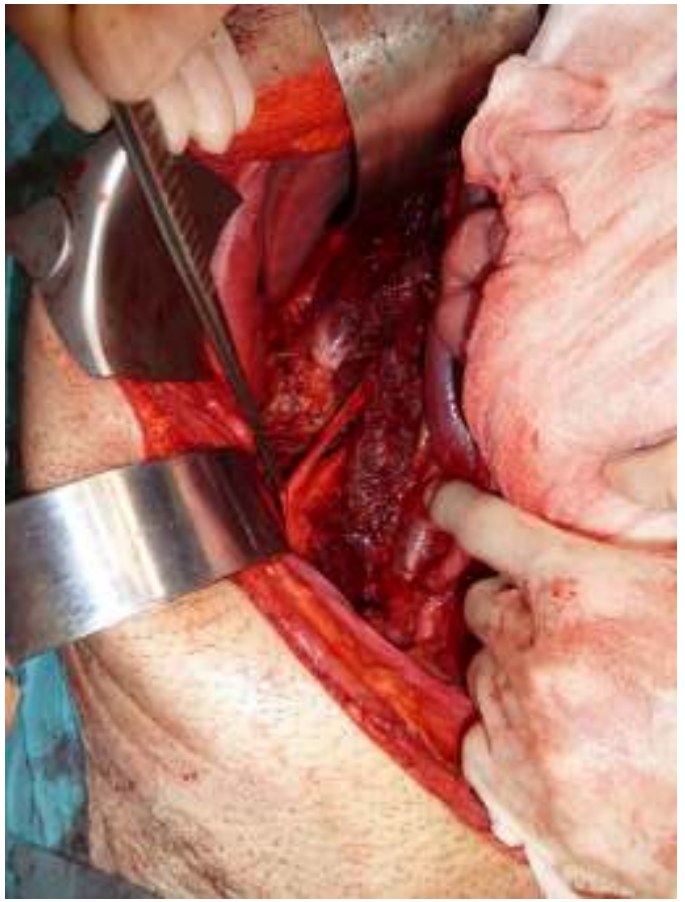
Ligation of all branches from the major vessels is performed close to their edge, on the side of the tumour.

- Gonadal vessels, renal vessels, adrenal, lumbar, IMA are ligated and divided as needed.

Stepwise approach – femoral nerve

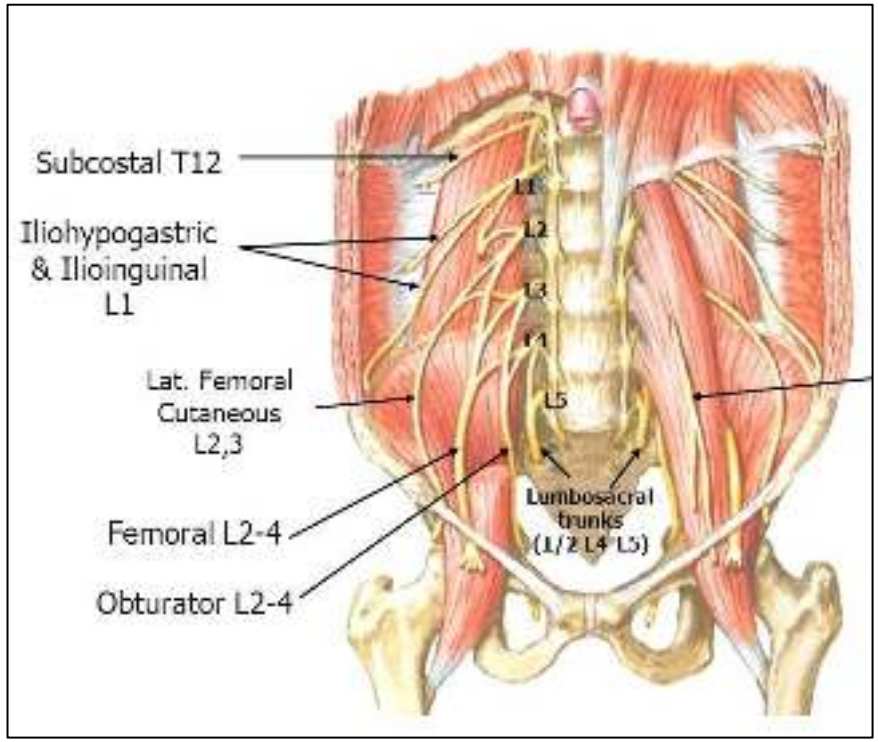
– The femoral nerve is identified just above the inguinal ligament: the fascia of the psoas muscle is opened, and the nerve may be identified posterolaterally.

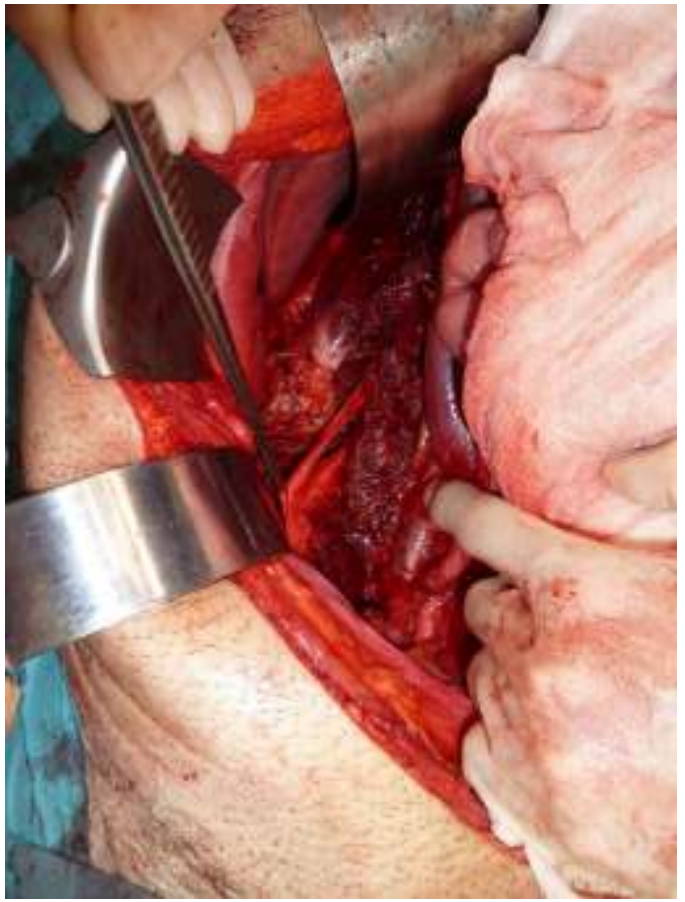




Stepwise approach - psoas

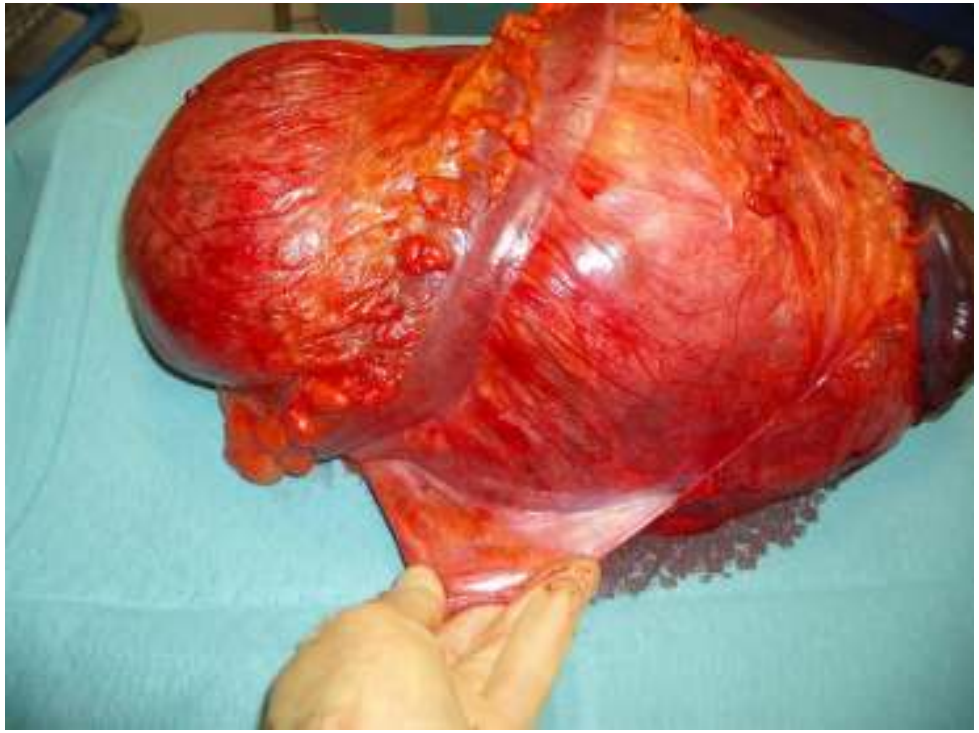
– When the psoas is involved (either directly invaded as evidenced by CT or MRI or tightly adherent, as noted intraoperatively), it should be resected from just superior to the inguinal ligament and detached from vertebral bodies.





Stepwise approach - lateral dissection:

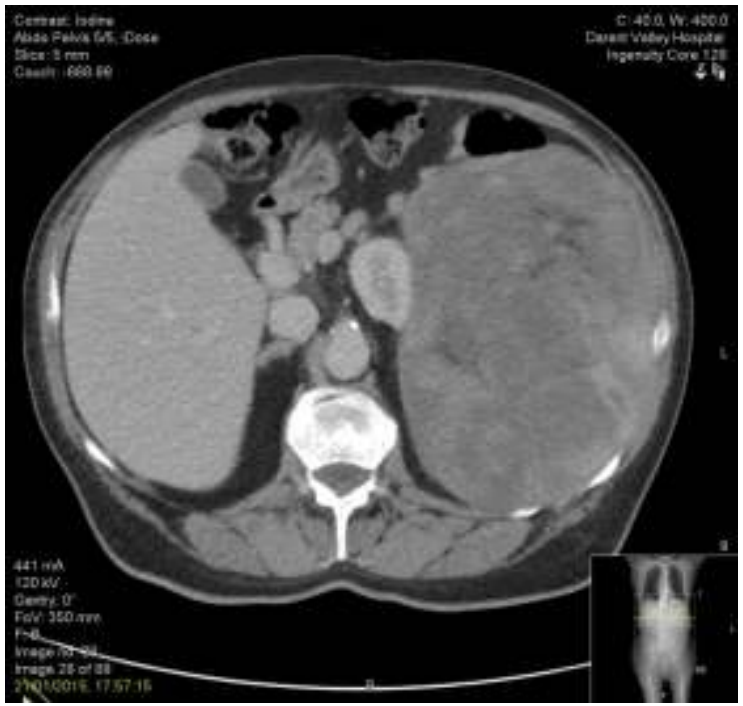
On the external side of the tumour, the parietal wall is incised at the level of the lateral muscles and the peritoneum, or the peritoneum and the internal layer of abdominal side wall are left on the tumour side.



Stepwise approach

– Lateral dissection:

On the external side of the tumour, the parietal wall is incised at the level of the lateral muscles and the peritoneum, or the peritoneum and the internal layer of abdominal side wall are left on the tumour side.



Stepwise approach

Pancreas/spleen

- For tumours confined to the lower left retroperitoneum — below the transverse mesocolon—the distal pancreas and spleen are detached from the upper extend of the tumour, which remains covered by transverse mesocolon, and are rotated medially to obtain good exposure.

- For tumours extending into the upper left retroperitoneum, the distal pancreas is also divided, the splenic artery and vein ligated, and aorta dissected up to the diaphragmatic hiatus.

Technical nonresectability

For the purposes of being considered for enrolment onto the international trial STRASS2, which is currently open, the patient's RPS must be deemed technically resectable as judged by CT scan.

The criteria for technical nonresectability are defined in the trial protocol as:

- Involvement of the superior mesenteric artery, aorta, coeliac trunk and/or portal vein
- Involvement of bone
- Growth into the spinal canal
- Progression of retro-hepatic inferior vena cava leiomyosarcoma towards the right atrium
- Infiltration of multiple major organs such as liver, pancreas and or major vessels

– It should be emphasized that these criteria have been established for the purposes of defining the study population in a prospective trial, and that outside of this setting, each individual patient should be evaluated for technical resectability by experienced RPS surgeons, who will consider technical issues in context with tumour- and patient- related variables.

Prognostic factors predicting outcome

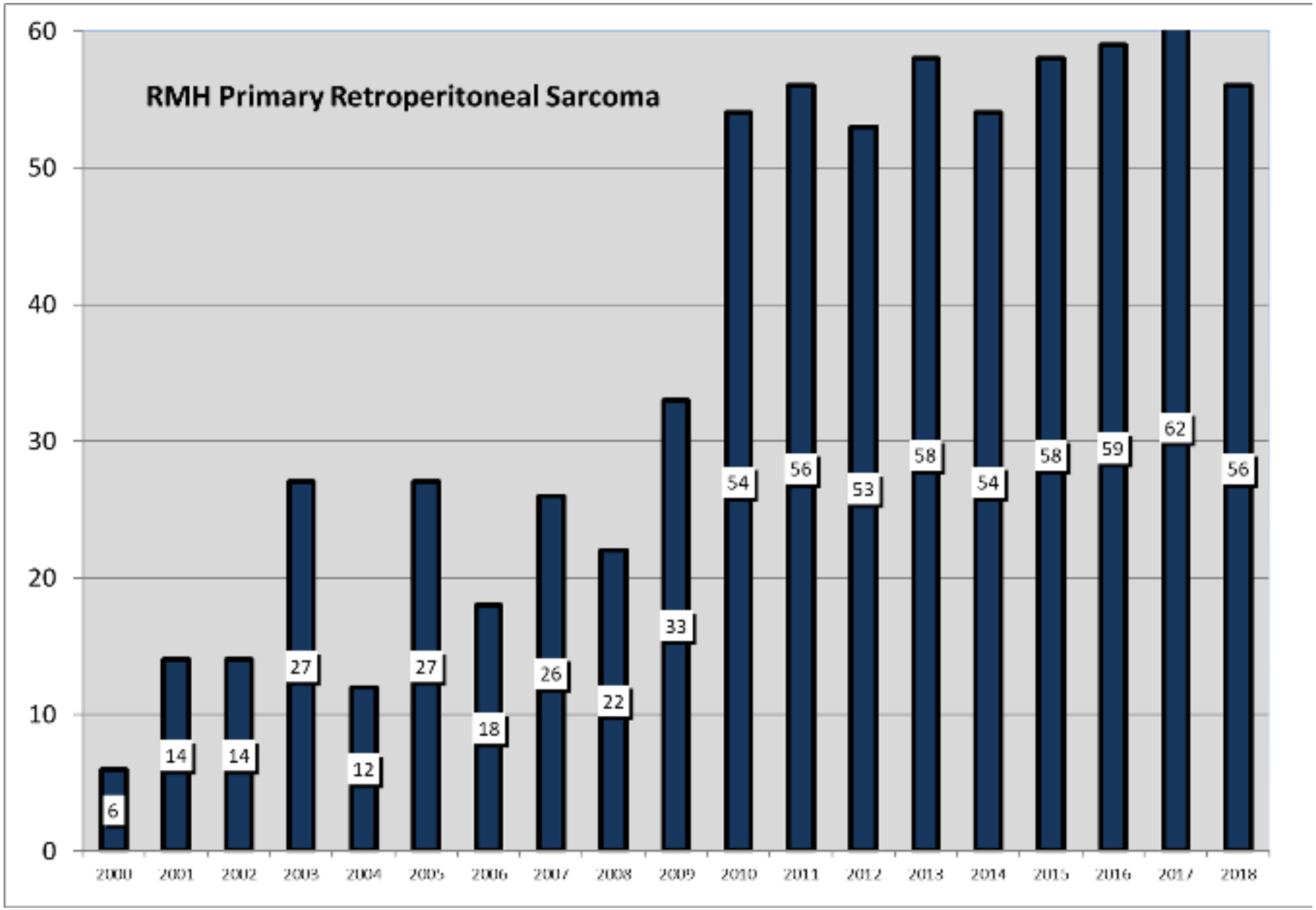
High-volume vs. low volume centres

- Volume-outcome relationships in the surgical care of RPS support the regionalization of care to high-volume specialist sarcoma units.

- The minimum annual volume of primary RPS resections that should be used to confer the status of specialist centre will be area-specific. A recent quantitative analysis of data derived from the NCDB identified 13 as the minimum annual institutional volume of RPS resections that was associated with improved long-term overall survival in the USA

- Members of TARPSWG who were surveyed regarding these results concurred that a minimum annual institutional surgical volume of 10 – 20 RPS cases was appropriate for a centre to be considered one of RPS expertise

Annual case load



Prognostic factors predicting outcome

High-volume vs. low volume centres

TABLE 3. Outcomes for Patients Following Surgical Resection for Soft Tissue Sarcoma According to Volume of Medical Facility

Outcome	Low-Volume Center (n = 2865; 68.1%)	High-Volume Center (n = 1340; 31.9%)	*P**
30-day mortality (%)	1.5	0.7	0.028
90-day mortality (%)	3.6	1.6	<0.001

On multivariate analysis, treatment at a high-volume centre was a significant independent predictor of improved median survival, 5 year and 10 year survival for all ***high-grade or RPS***

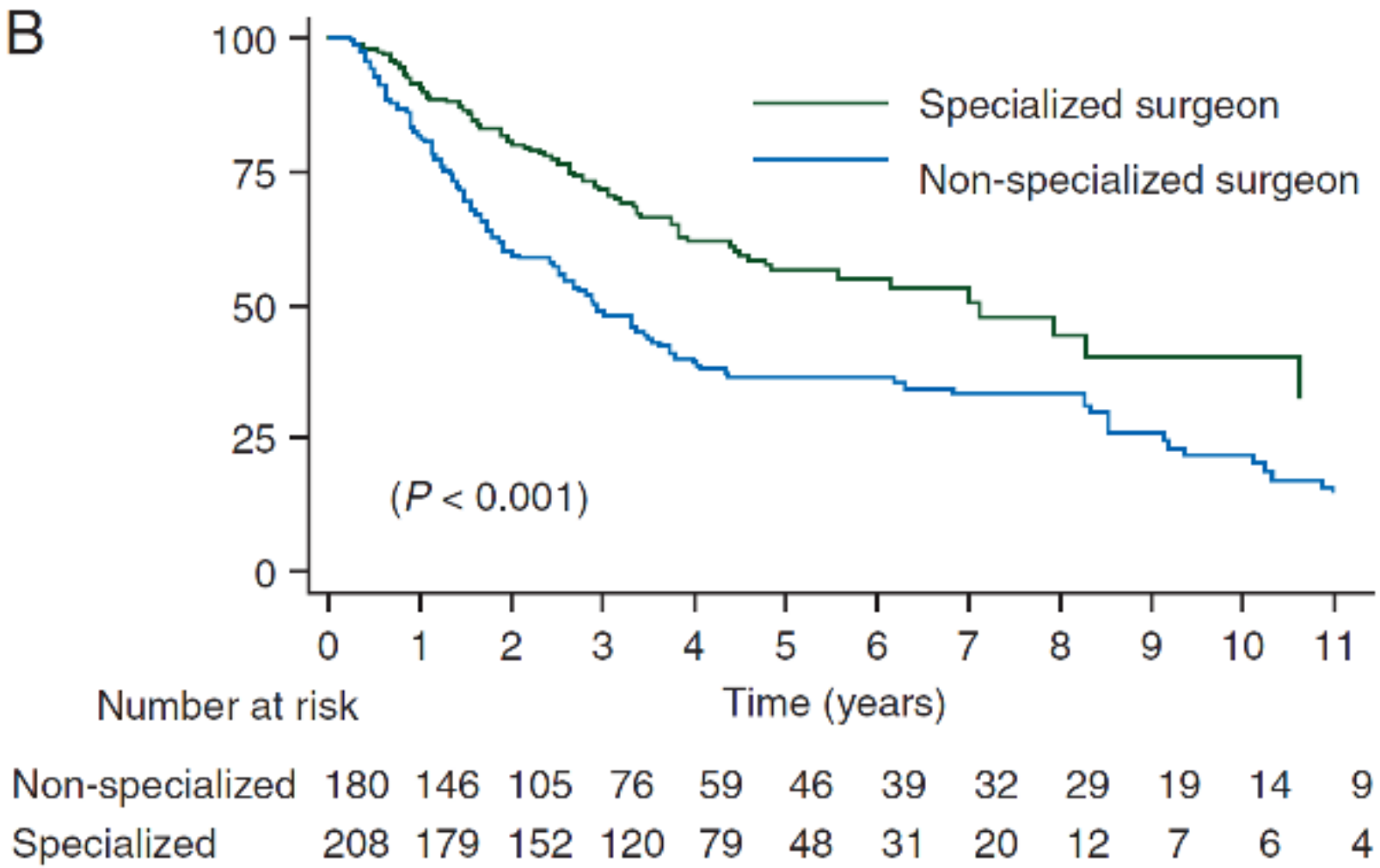
Prognostic factors predicting outcome

High-volume vs. low volume RPS centres

- Early surgical outcomes (morbidity/mortality) are improved
- Oncology outcome improved
  - Institutional volume was a significant risk factor for local recurrence
  - Higher rate of incomplete resections in low volume centres (38% vs. 18%, P = 0.002)
  - Intra-operative tumour rupture rate is inversely related to institutional volume in RPS

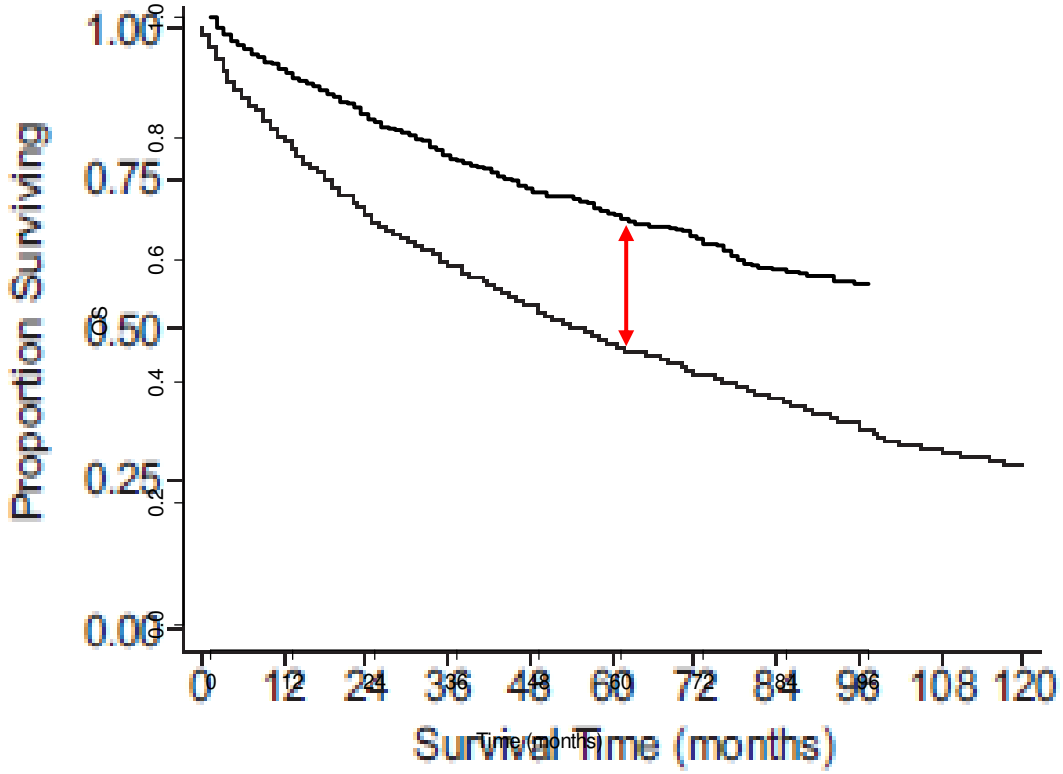
1. *Learn PA, Bach PB. Med Care. 48(12) 1041-9 (2010).*
2. *Gutierrez JC, Perez EA, Moffat FL, Livingstone AS, Franceschi D, Koniaris LG. Ann Surg. 245(6) 952-8 (2007).*
3. *Bonvalot S, Rivoire M, Castaing M, et al. J Clin Oncol. 27(1) 31-7 (2009).*
4. *Van Dalen T, Hennipman A, Van Coevorden F, et al. Ann Surg Oncol. 11(5) 483-90 (2004).*

French Sarcoma Group



Toulemond et al. Ann Oncol 2014; 25: 735-742

High-volume specialist centres

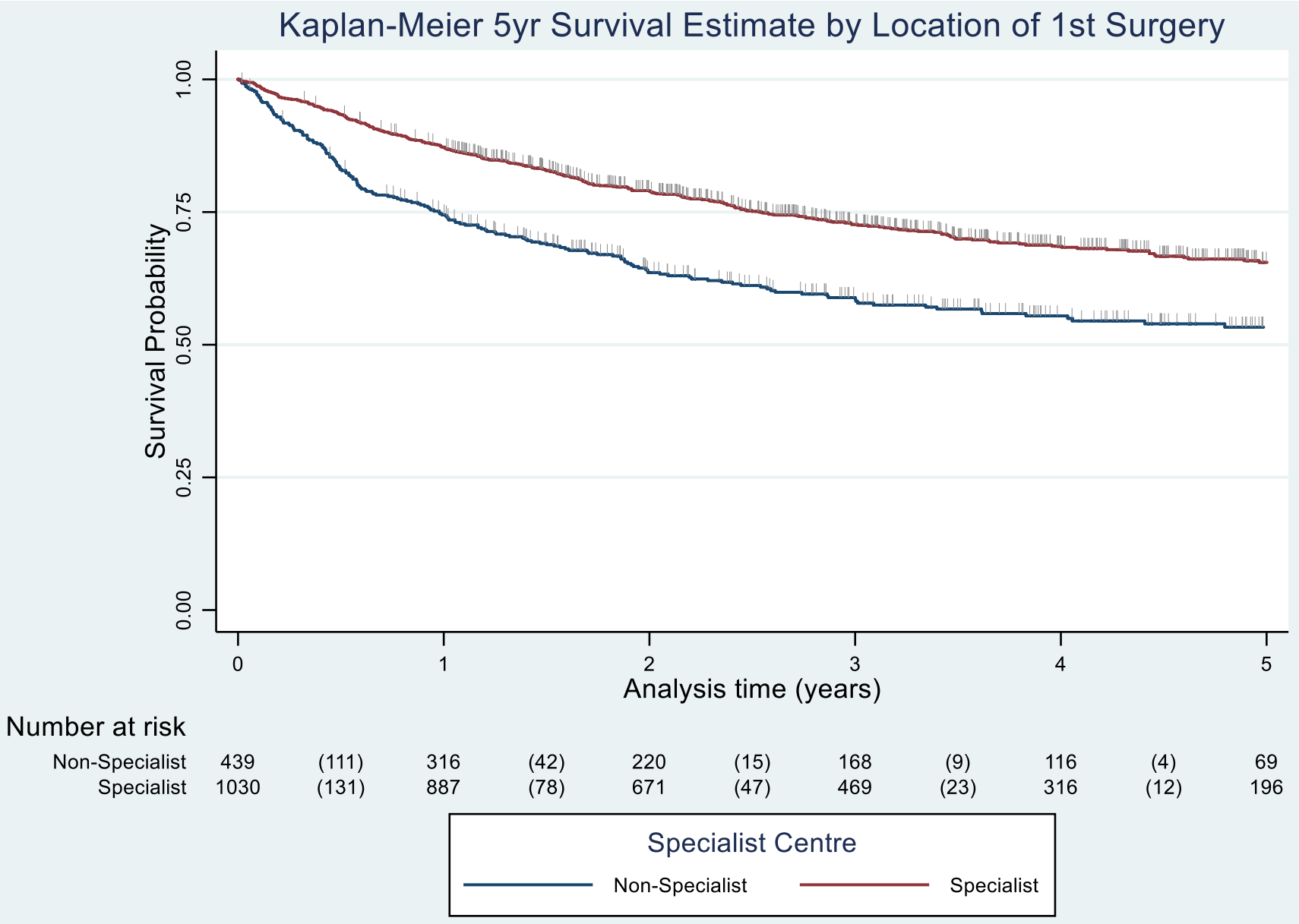


Transatlantic Sarcoma Group
>1000 Primary RPS
5yr OS – 67% *(2013)*

SEER: Primary 1365 RPS
5yr OS – 47% *(2010)*

First Surgery at a Non-Specialist Centre Results in Poorer Survival Outcomes

UK 2013-18 data



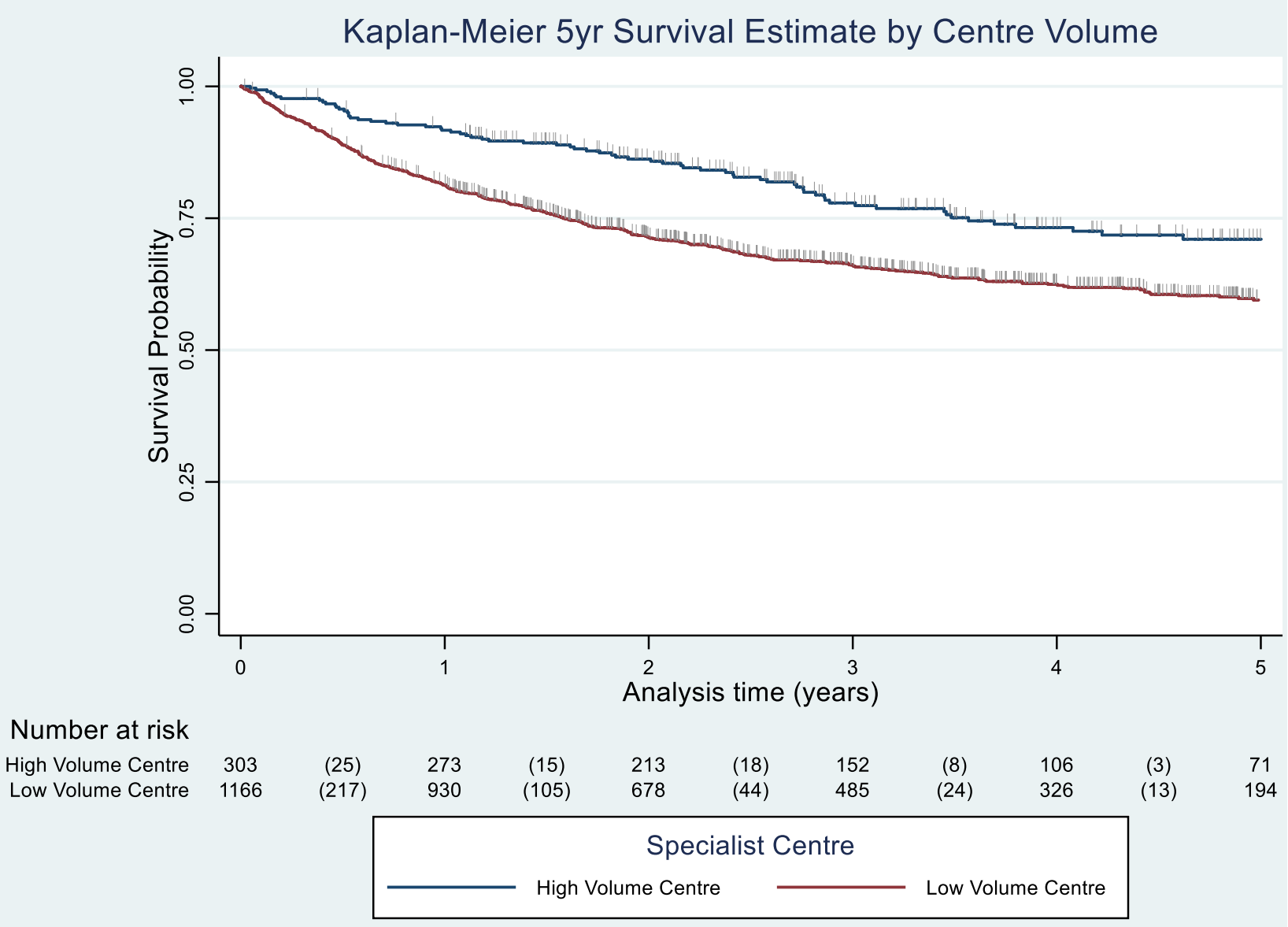
- Specialist centre is a Trust which holds a sarcoma MDT. This definition is subject to update.
- 70% undergo their first surgery at a 'specialist centre'.

- 5yr Survival
  - Specialist Centre: 66% (CI: 62-69%)
  - Non-Specialist Centre: 53% (CI: 48-58%)

The Royal Marsden

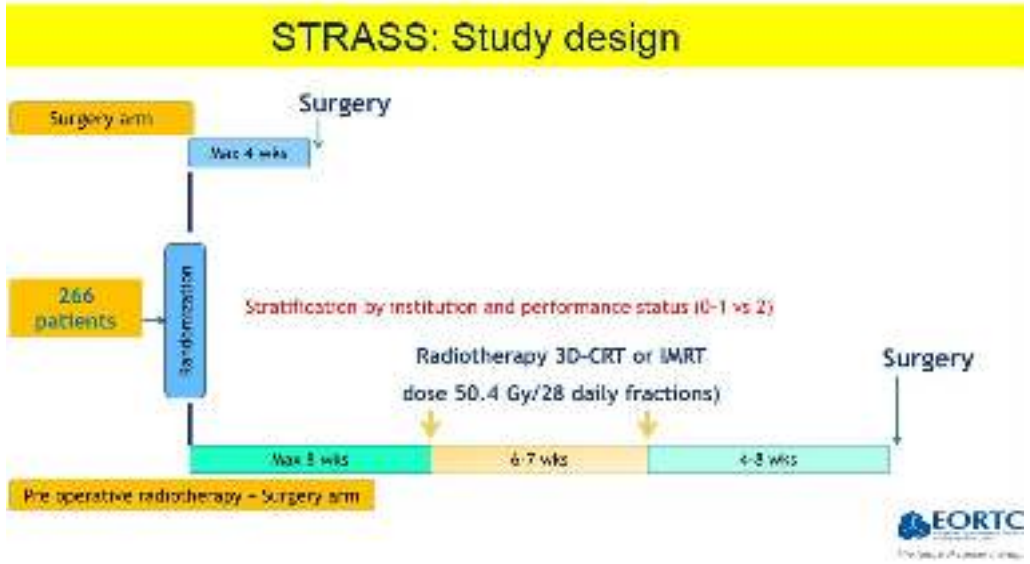
High Volume Centres have Superior Survival Outcomes

UK 2013-18 data



- A high volume centre was defined as a trust which *meets* the sarcoma service specification carrying out at least 24 operations per year.
- 5yr Survival
  - High Volume: 71% (CI: 64-77%)
  - Low Volume: 59% (CI: 56-63%)
- Those who undergo their first surgery at a high volume centre have superior survival outcomes

Neoadjuvant radiotherapy

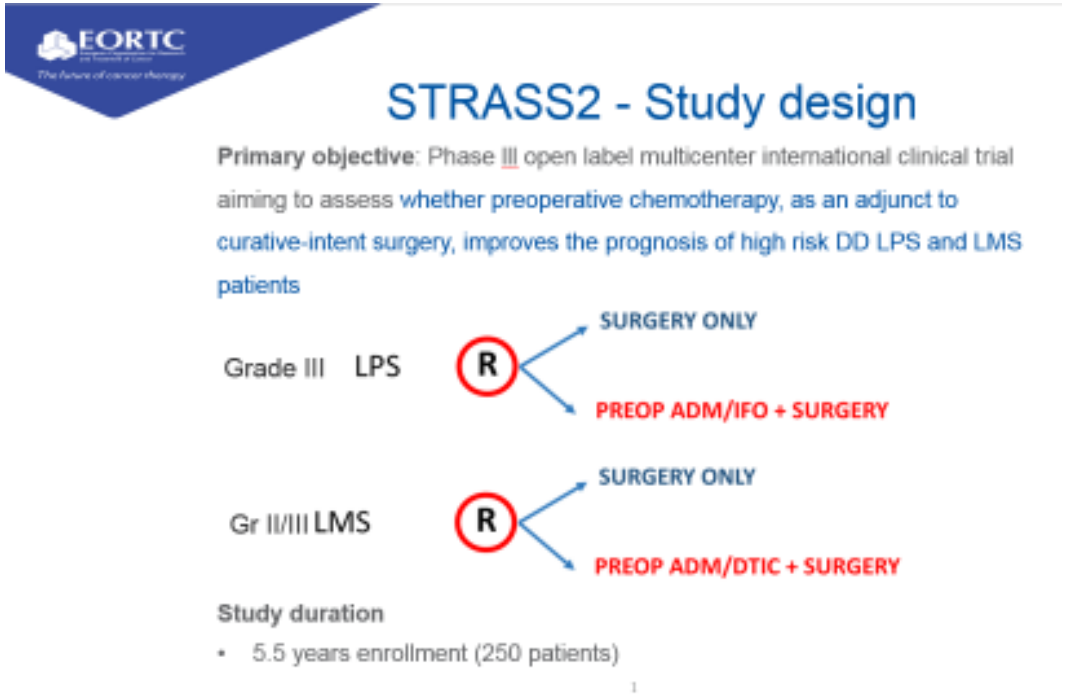


- A phase III randomized study of preoperative radiotherapy plus surgery versus surgery alone for patients with retroperitoneal sarcoma (STRASS) has been performed

- Based on recent analysis of the primary endpoint, Abdominal Recurrence Free Survival (ARFS), routine use of neoadjuvant RT is not recommended in patients with high-grade RPS

- May be considered in those with high risk of local (abdominal)-only recurrence, i.e. well-differentiated liposarcoma and low-grade dedifferentiated liposarcoma.

Neoadjuvant chemotherapy.



- Neoadjuvant chemotherapy can be discussed for use in individual patients with chemo-sensitive histologies such as synovial sarcoma and high-grade LMS, among others, or within prospective clinical studies

- Consideration of preoperative chemotherapy or chemoradiation with cytoreductive intent is particularly relevant in the case of technically unresectable or borderline resectable RPS that could potentially be rendered more amenable to safe grossly complete resection through tumour downsizing

- Patients with high-grade dedifferentiated liposarcoma or high-grade leiomyosarcoma should be considered for entry into the STRASS2 trial of preoperative chemotherapy vs. resection alone for potentially curable primary RPS

Conclusion

- 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.

Conclusion

- 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.

Conclusion

- 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.

Conclusion

- 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 high-volume centres.

The ROYAL MARSDEN
NHS Foundation Trust

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Thank you