

SON Update 2017

Diagnosis and management of retroperitoneal sarcoma

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BC Cancer Agency

CARE + RESEARCH

An agency of the Provincial Health Services Authority



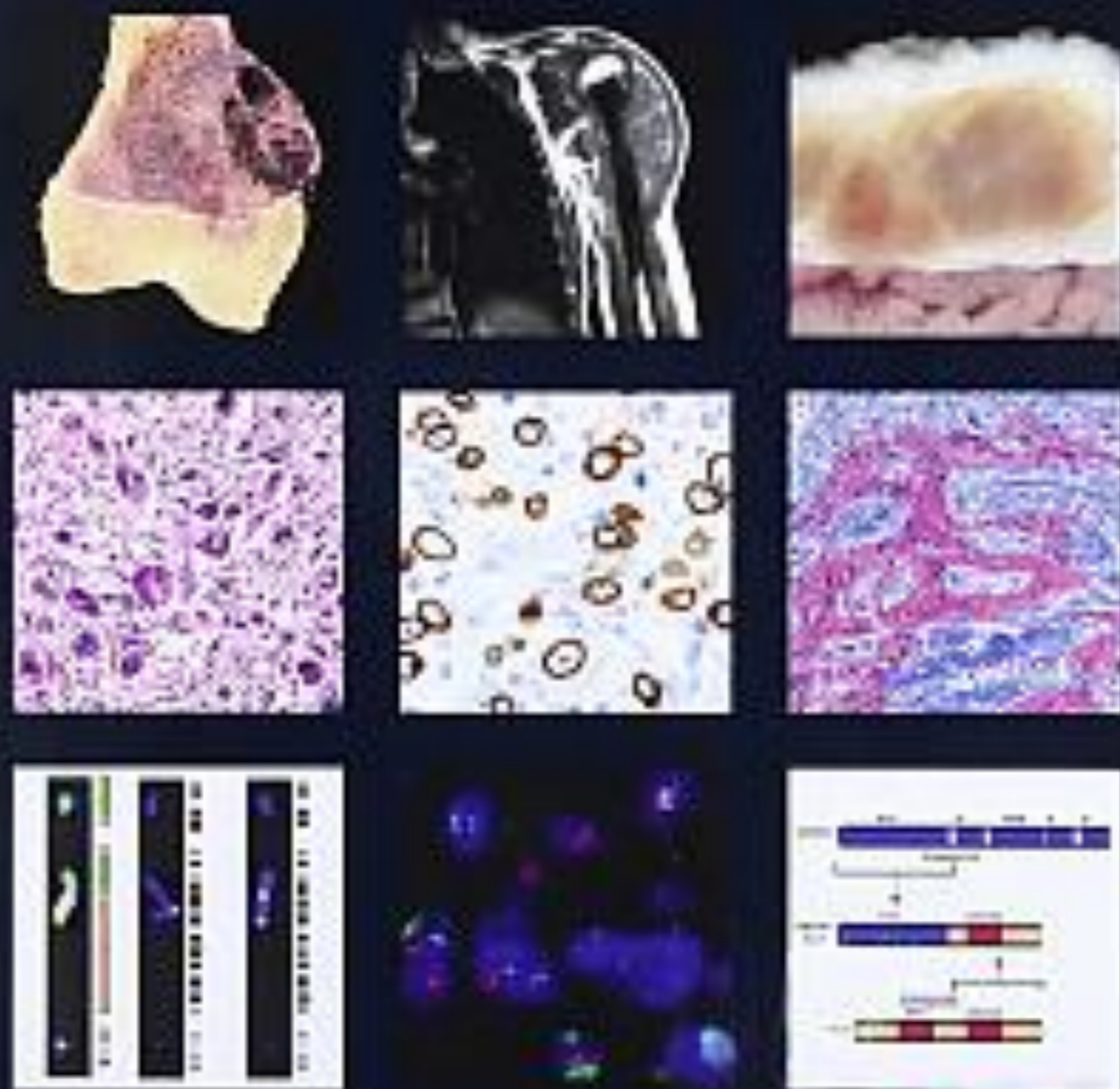
a place of mind

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Vancouver Coastal Health
Promoting wellness. Ensuring care.

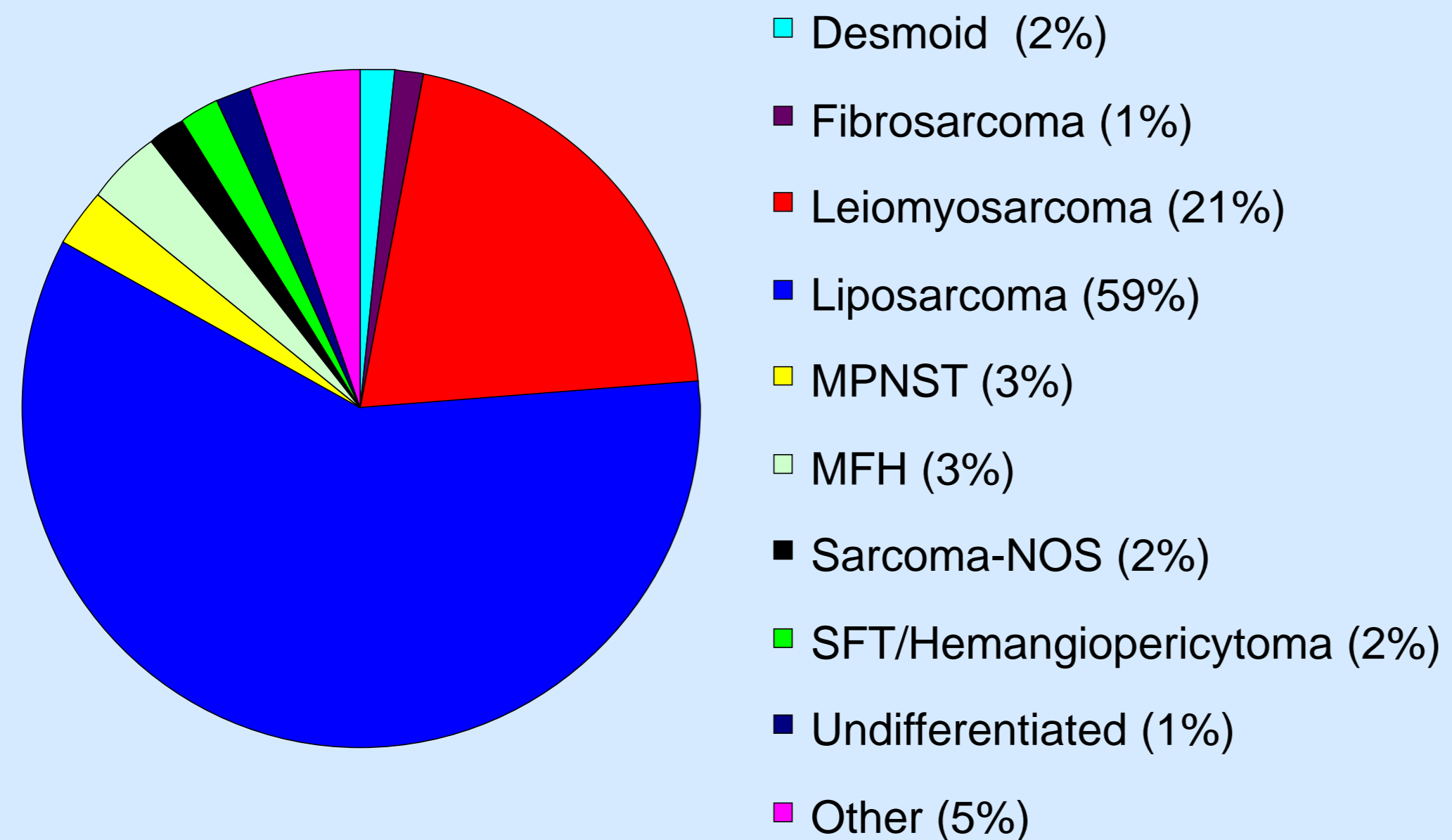
WHO Classification of Tumours of Soft Tissue and Bone

Edited by Christopher D.M. Fletcher, Julia A. Bridge, Pancras C.W. Hogendoorn, Fredrik Mertens

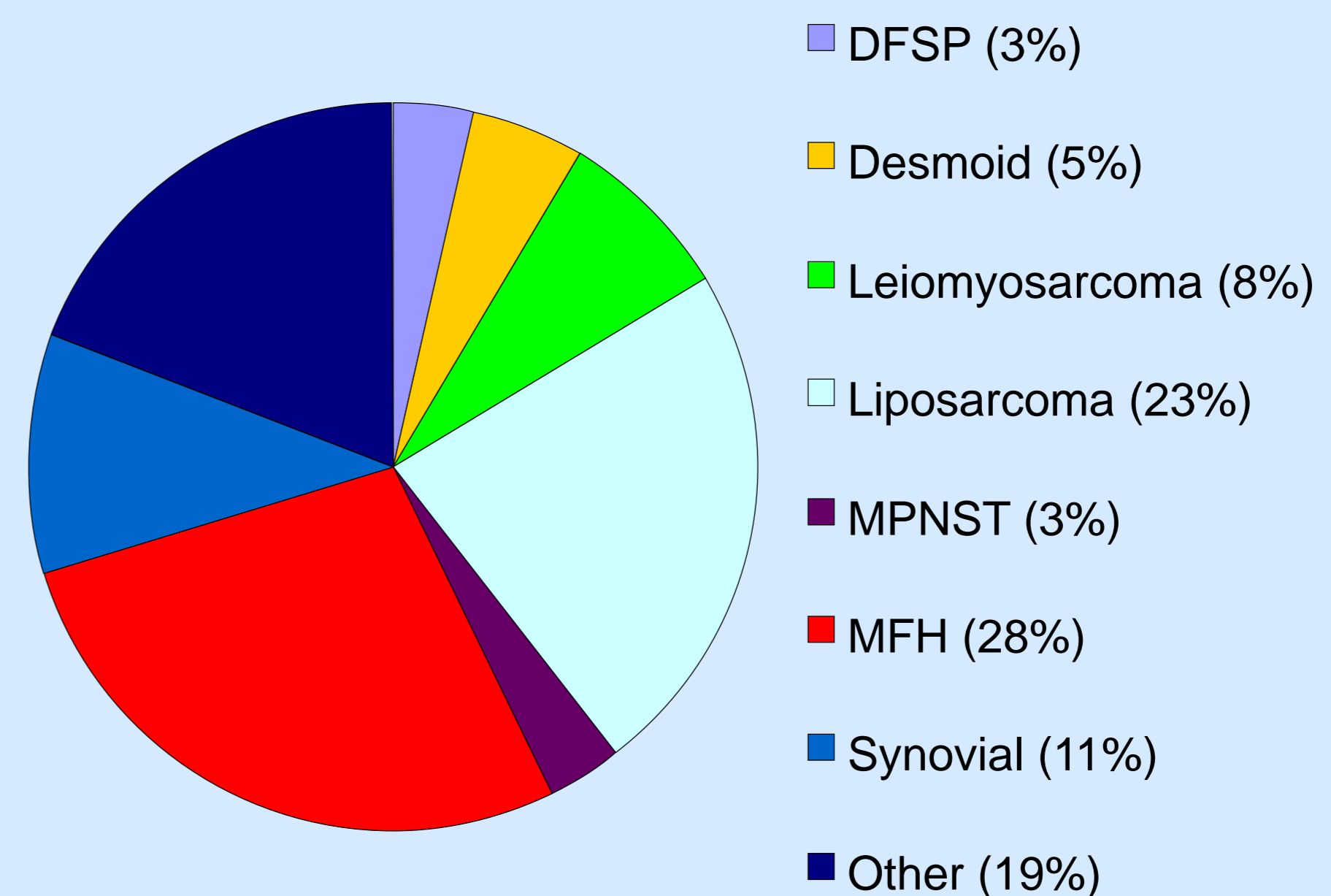


Histologic Subtypes of STS

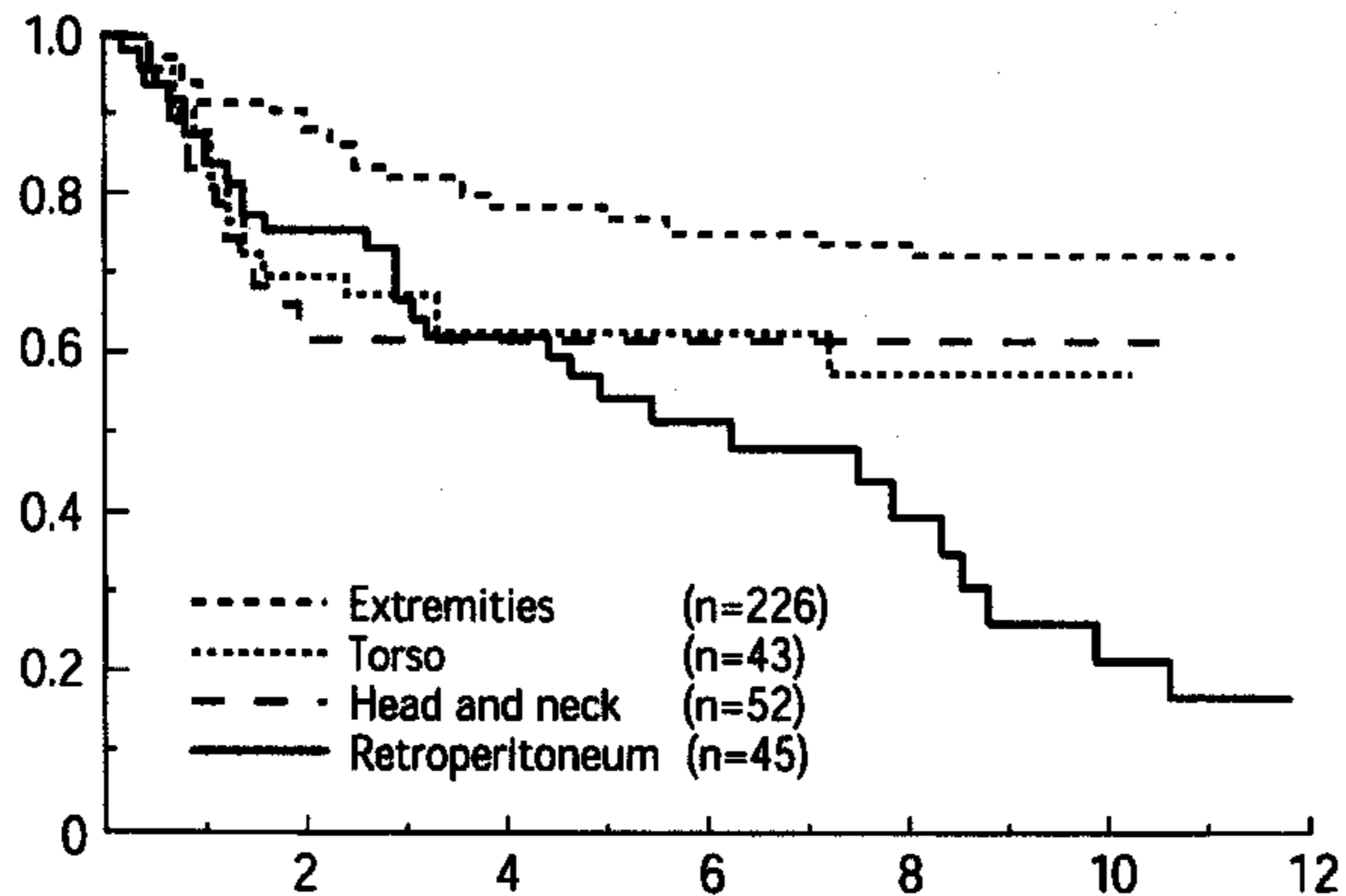
RP Subtypes (n=684)



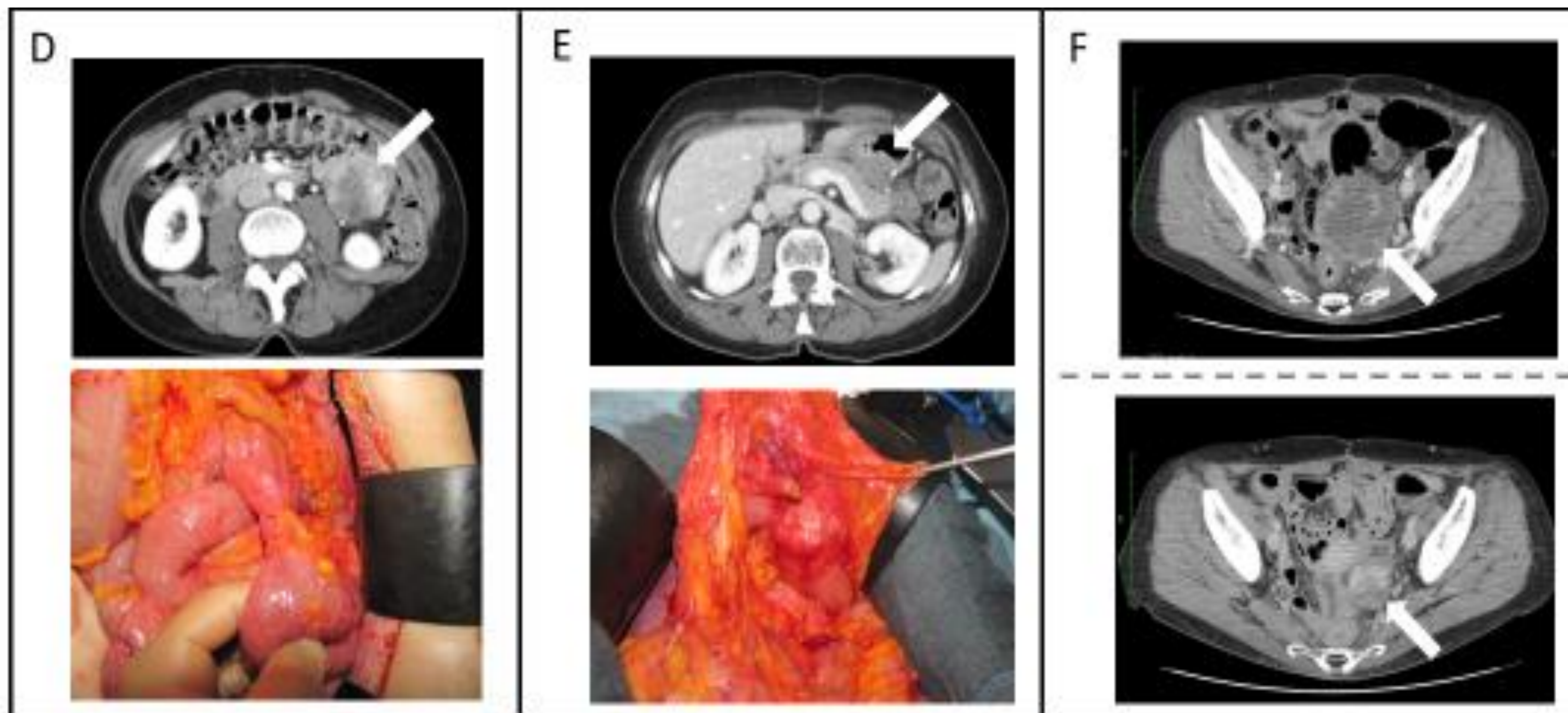
Extremity Subtypes (n=3,039)



Outcomes in STS



Local failure kills



- Local failure leading cause of disease-specific mortality
- Anatomic constraints often preclude R0 resection
 - ✓ *Complete (R0/1) vs Incomplete (R2)*

Evolution of Surgical Approach to RPS

VOLUME 27 · NUMBER 1 · JANUARY 1 2009

JOURNAL OF CLINICAL ONCOLOGY

ORIGINAL REPORT

Aggressive Surgical Policies in a Retrospectively Reviewed Single-Institution Case Series of Retroperitoneal Soft Tissue Sarcoma Patients

Alessandro Gronchi, Salvatore Lo Vullo, Marco Fiore, Chiara Mussi, Silvia Stacchiotti, Paola Collini, Laura Lozza, Elisabetta Pennacchioli, Luigi Mariani, and Paolo Giovanni Casali

From the Departments of Surgery, Biostatistics, Pathology, and Radiotherapy, Fondazione IRCCS Istituto Nazion-

A B S T R A C T

VOLUME 27 · NUMBER 1 · JANUARY 1 2009

JOURNAL OF CLINICAL ONCOLOGY

ORIGINAL REPORT

Primary Retroperitoneal Sarcomas: A Multivariate Analysis of Surgical Factors Associated With Local Control

Sylvie Bonvalot, Michel Rivoire, Marine Castaing, Eberhard Stoeckle, Axel Le Cesne, Jean Yves Blay, and Agnès Laplanche

From the Departments of Surgery, Public Health, and Medical Oncology, Institut Gustave-Roussy, Villejuif

A B S T R A C T

Extended resection leads to improved **local control**

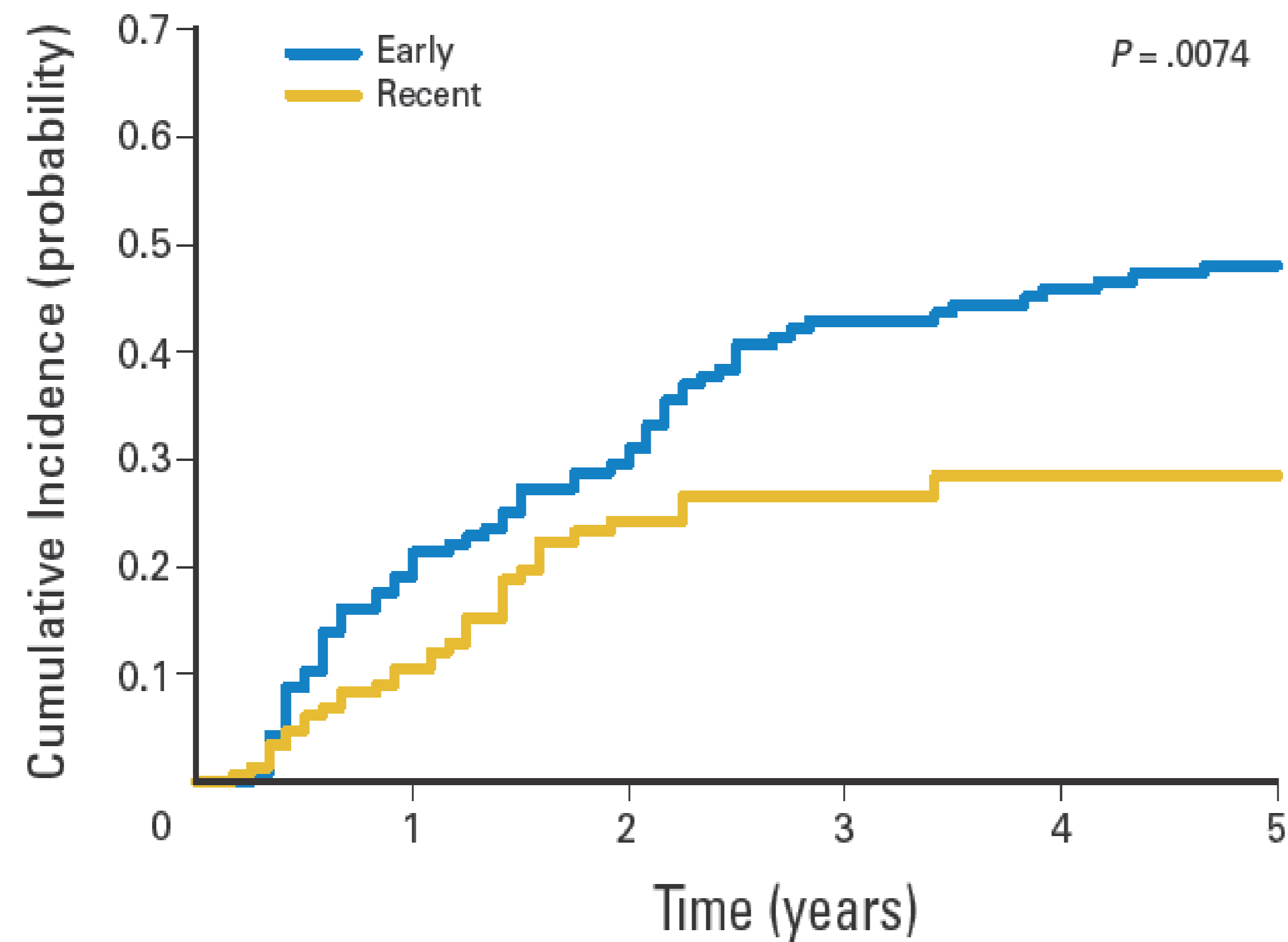
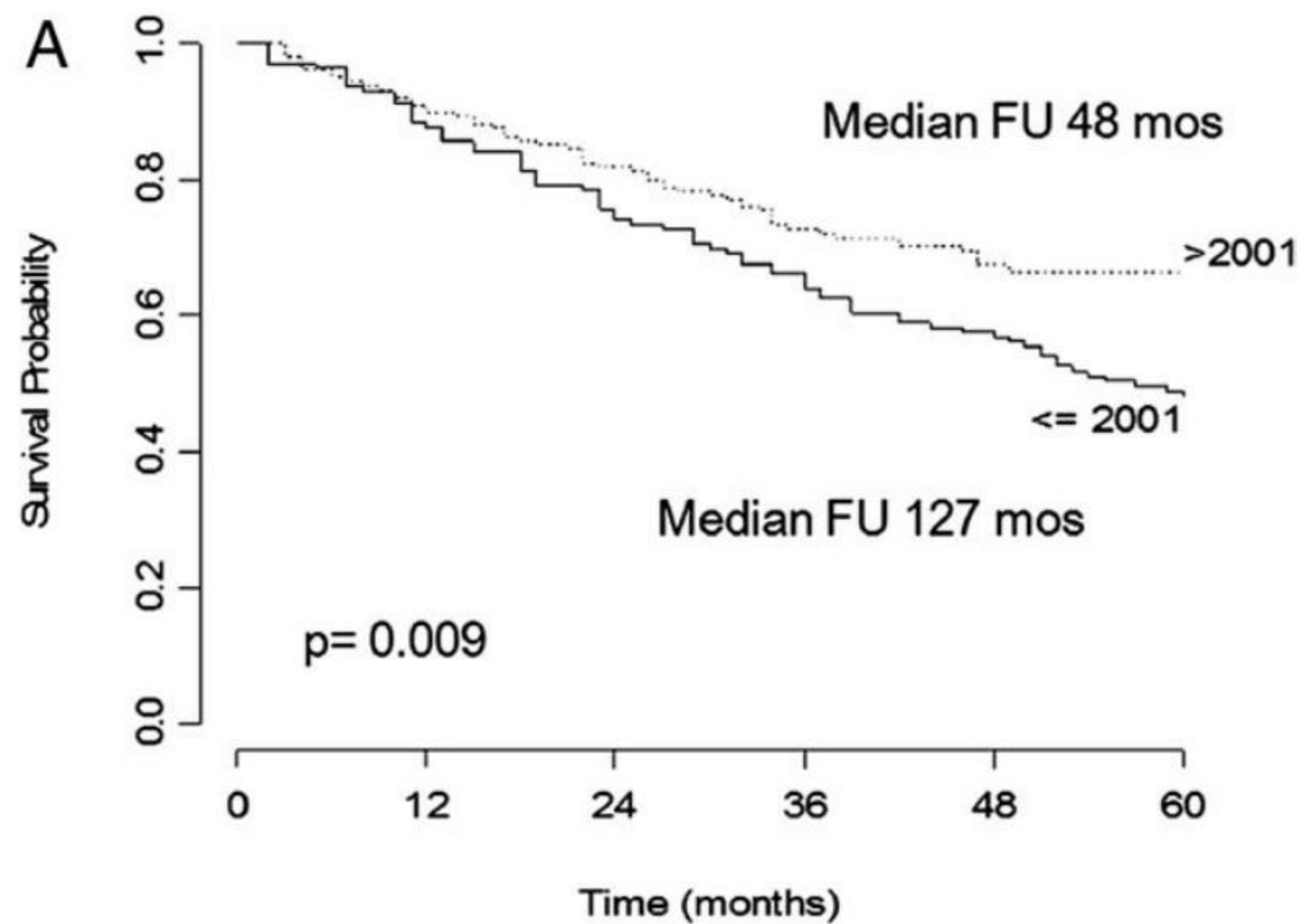


fig 1. Crude cumulative incidence of local recurrence by period of surgical resection at a single institution.

Extended resection is associated with improved overall survival



66%

48%

Extended resection operative approach

Principle – liberal resection of all “involved” organs

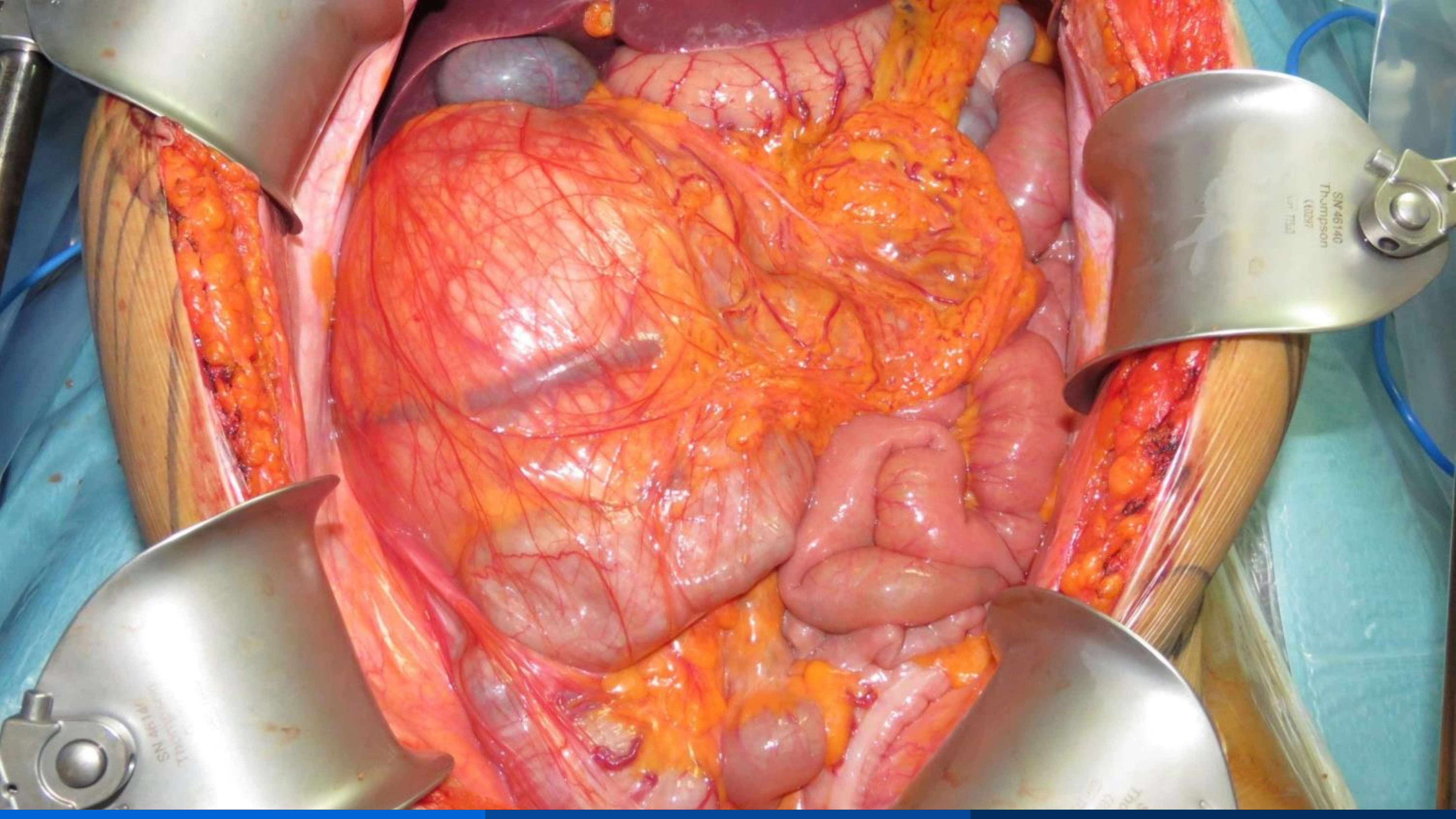
Right side RPS

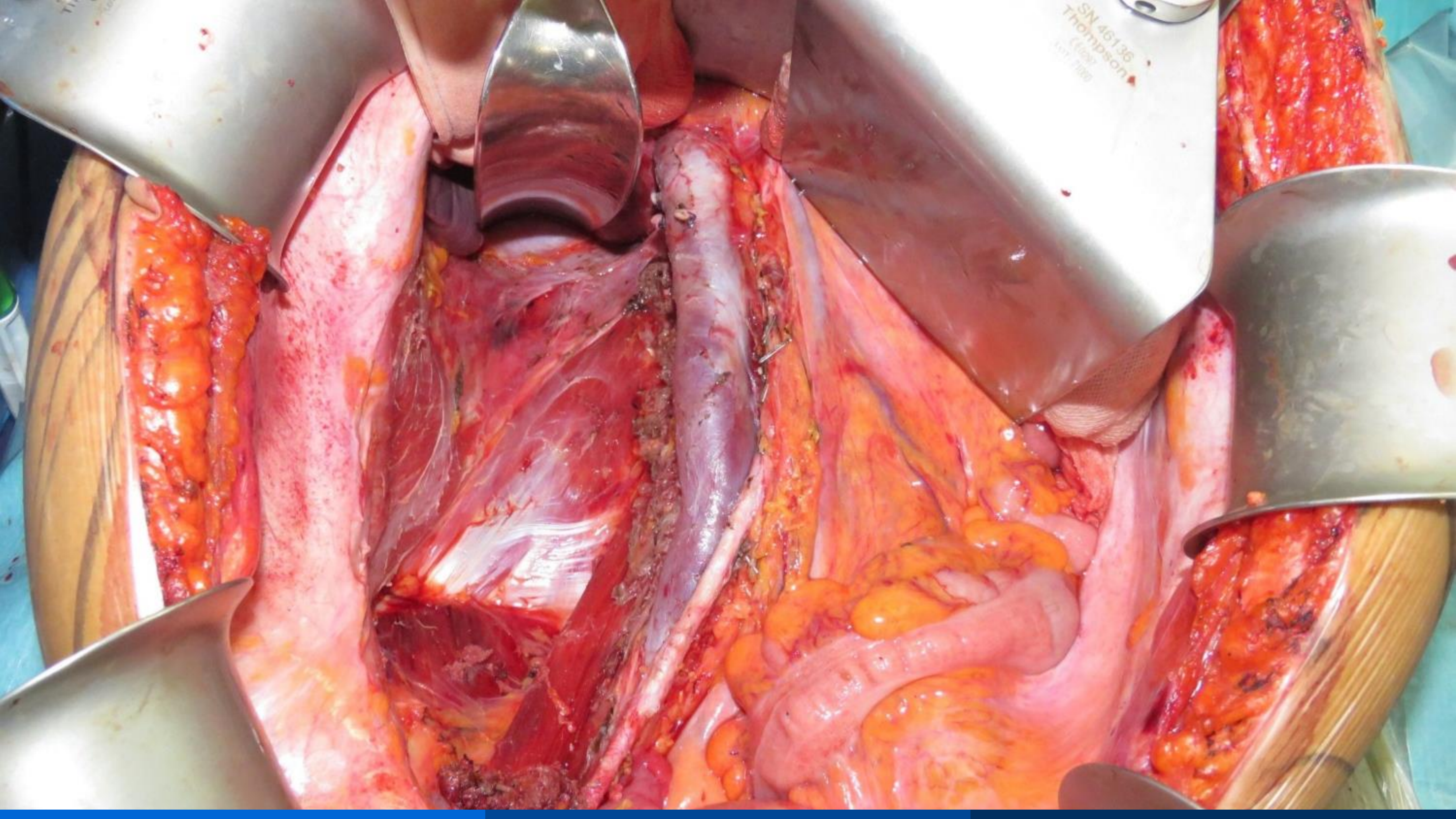
- Colon
- Kidney
- Psoas fascia
- +/- Duodenum
- +/- Liver capsule
- +/- IVC/iliac vessels

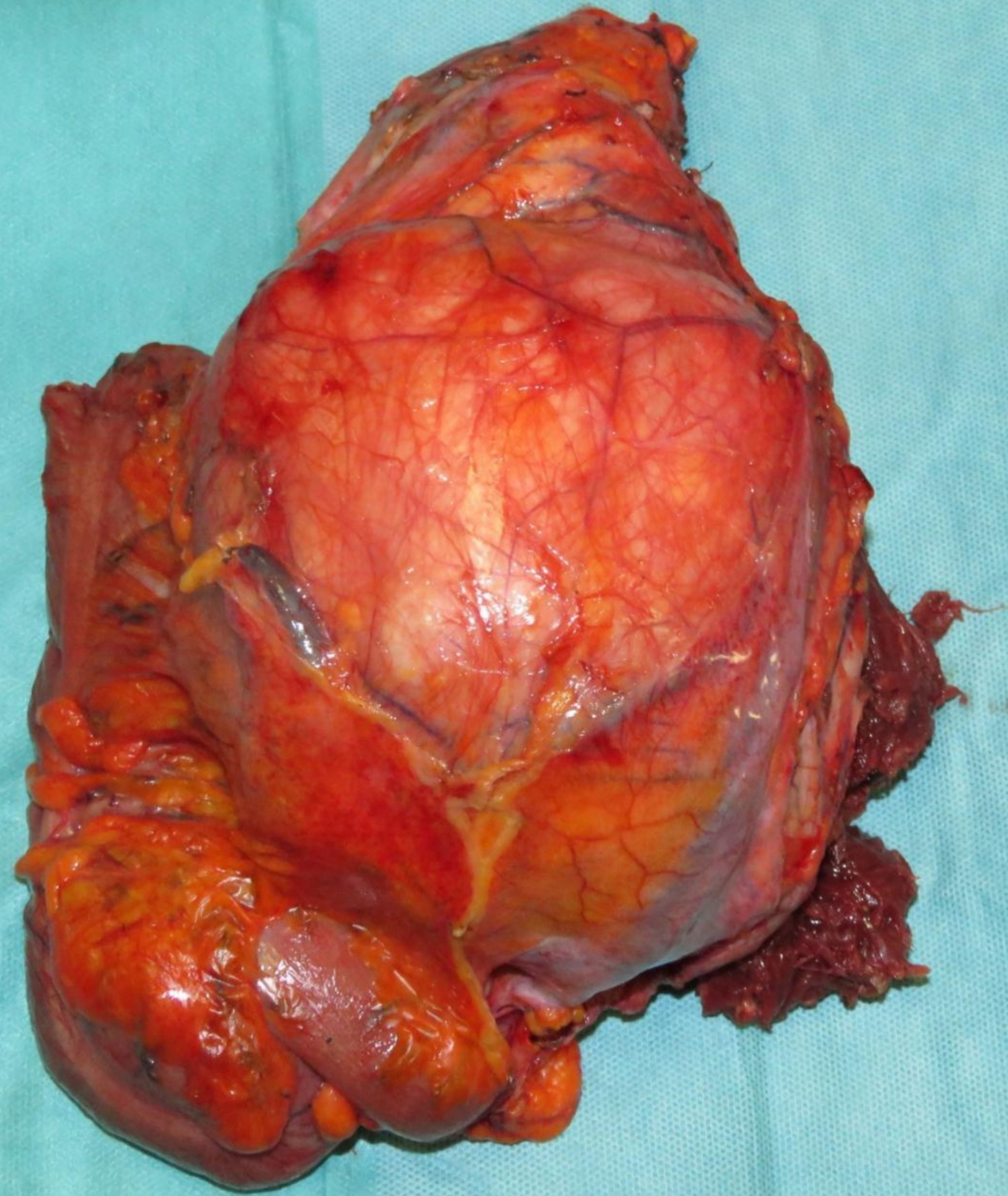
Left side RPS

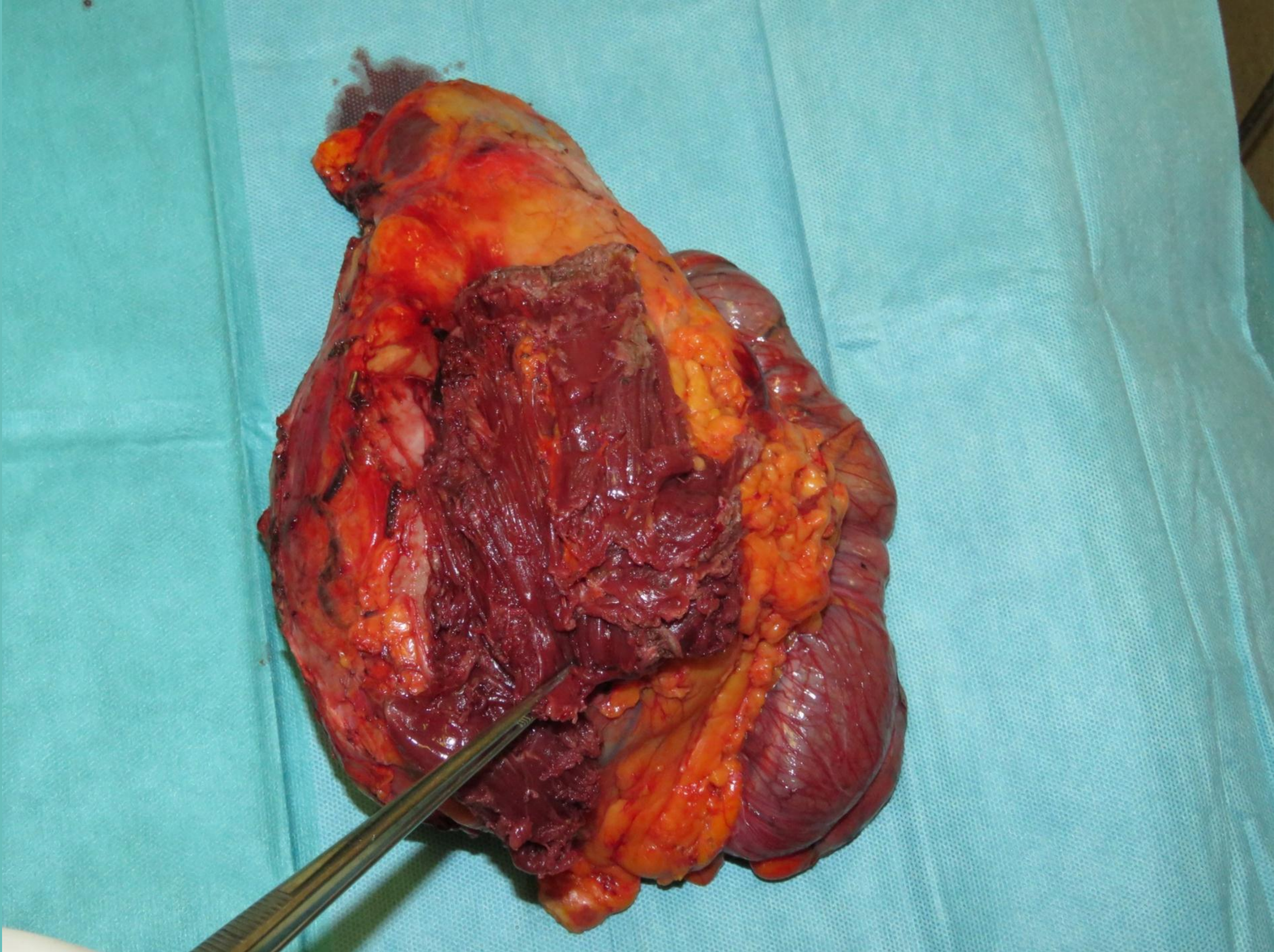
- Colon
- Kidney
- Psoas fascia
- Distal pancreas + spleen
- +/- Aorta/iliac vessels
- +/- Diaphragm



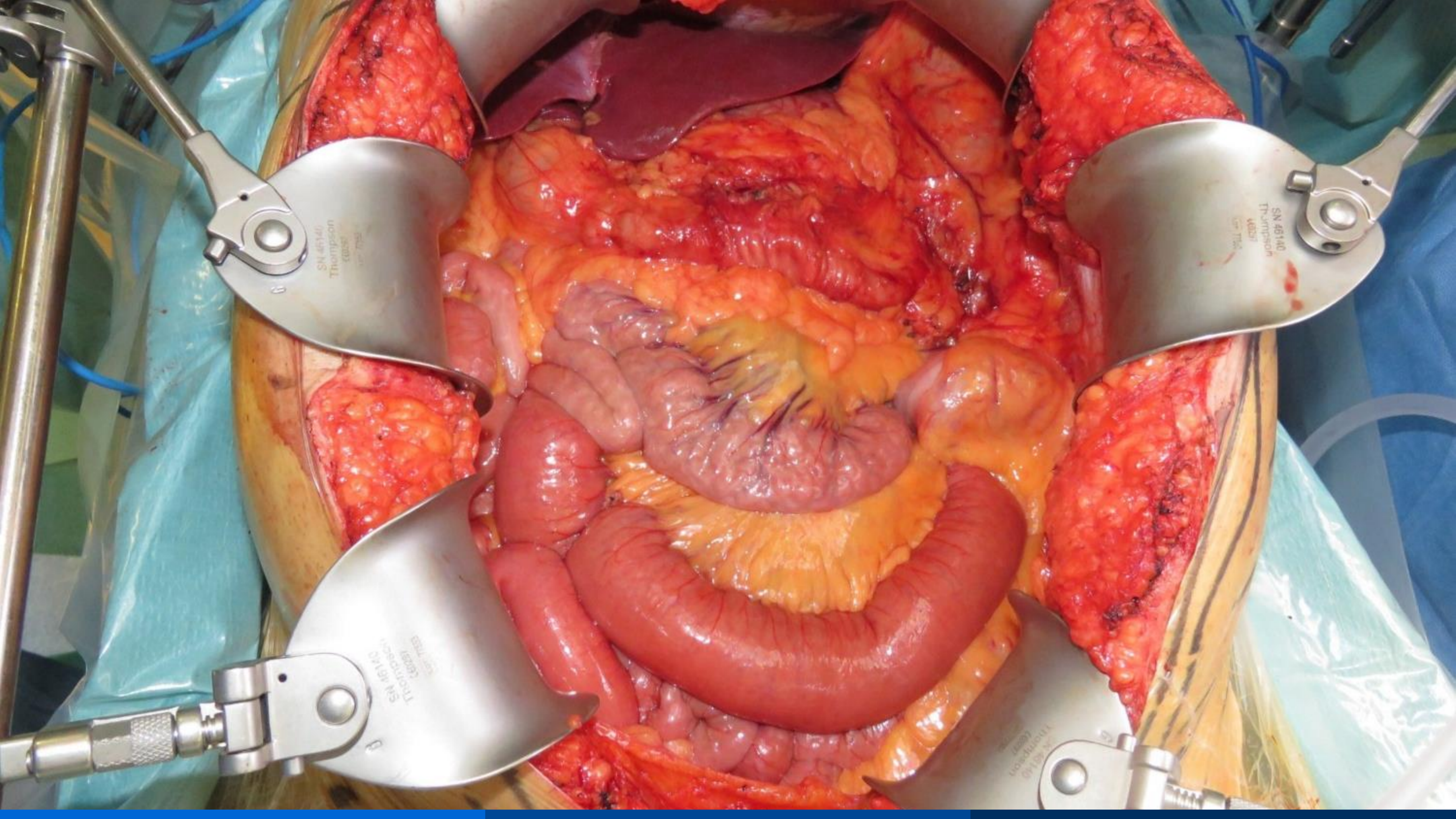


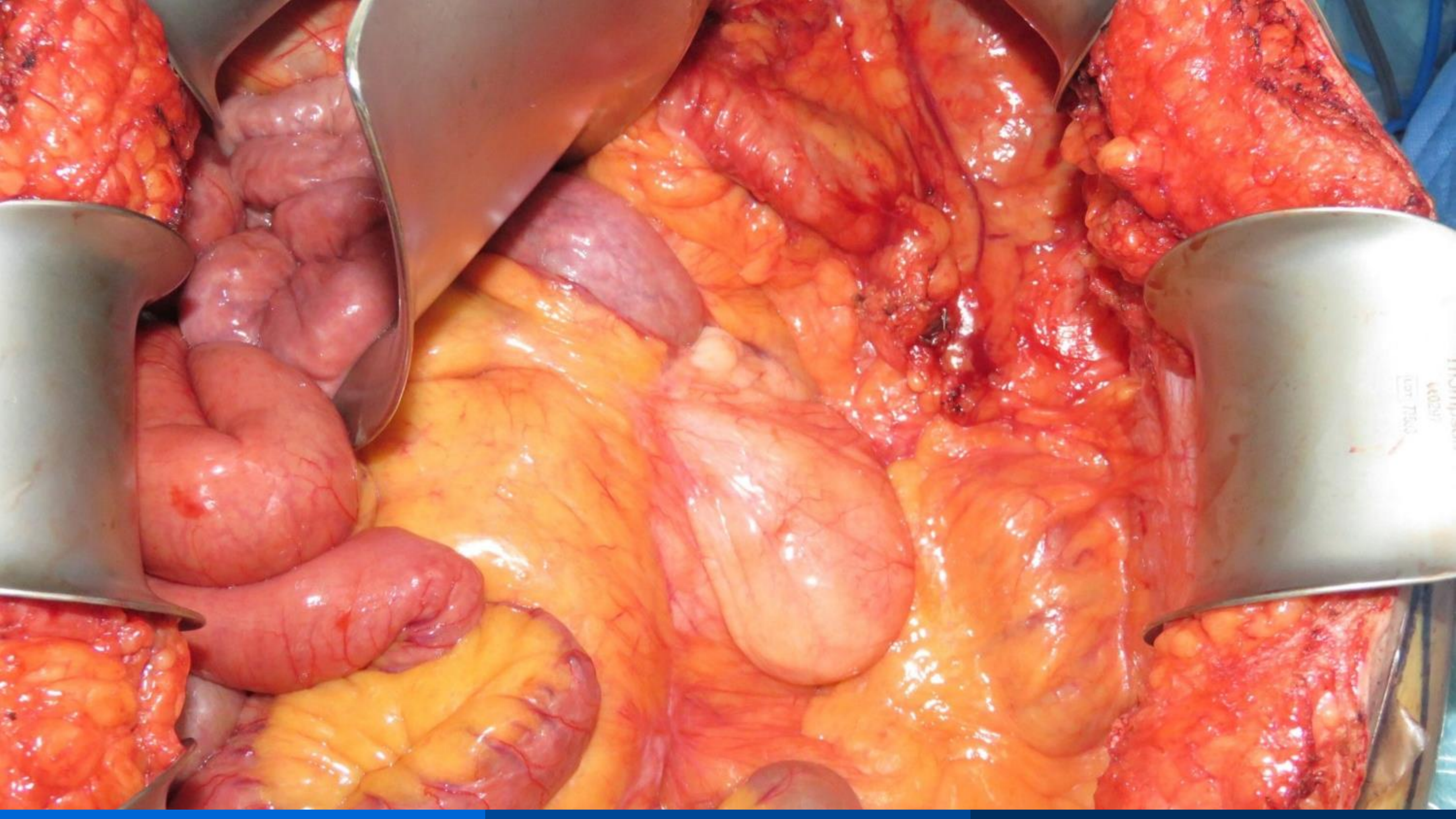


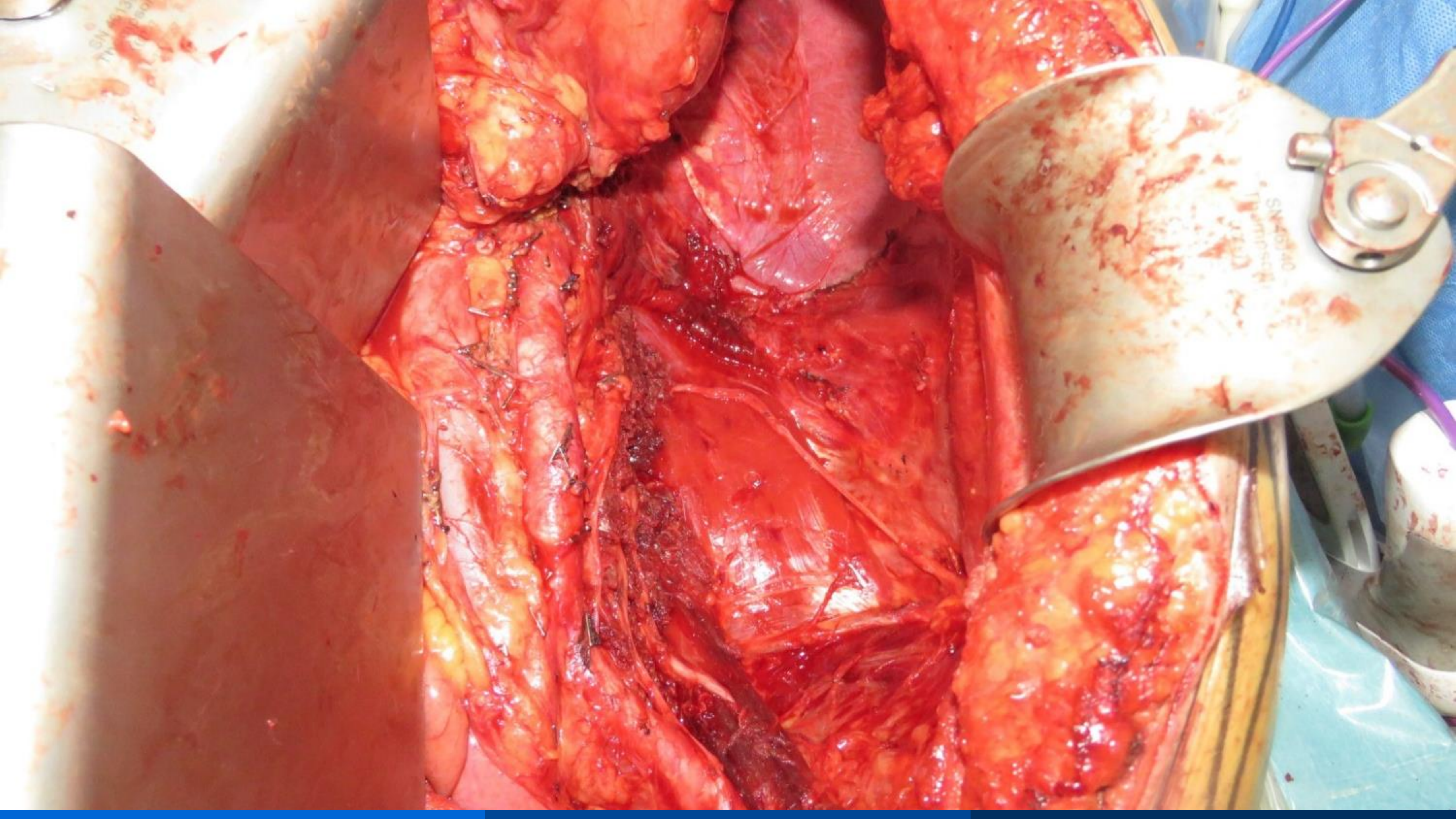




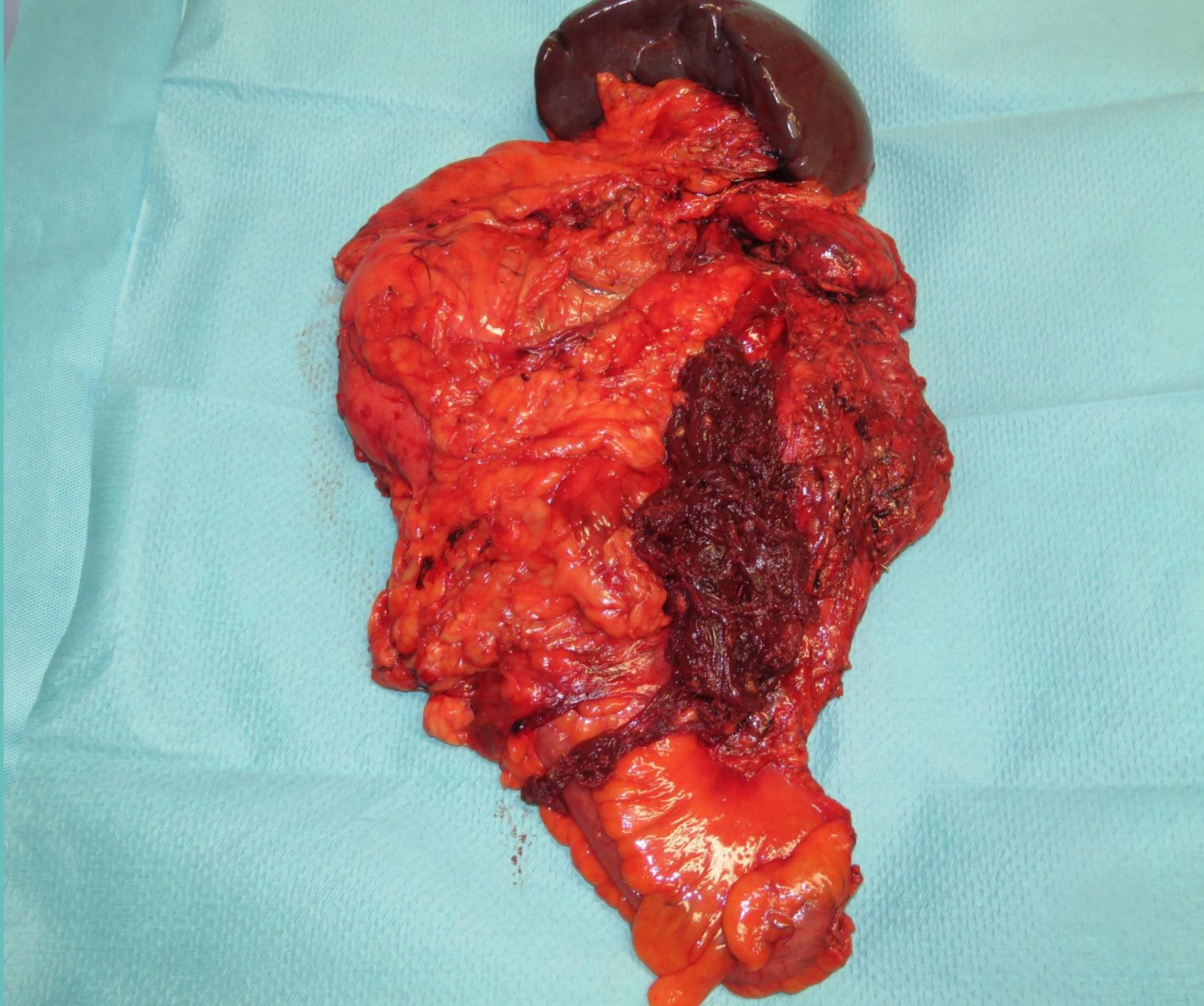












How to work up a retroperitoneal mass

Labs

- B-HCG
- AFP
- LDH
- Metanephrines/catecholamines if appropriate

Imaging

- CT abdomen preferable to MRI
- CT chest for staging
- Differential renal scan if nephrectomy anticipated

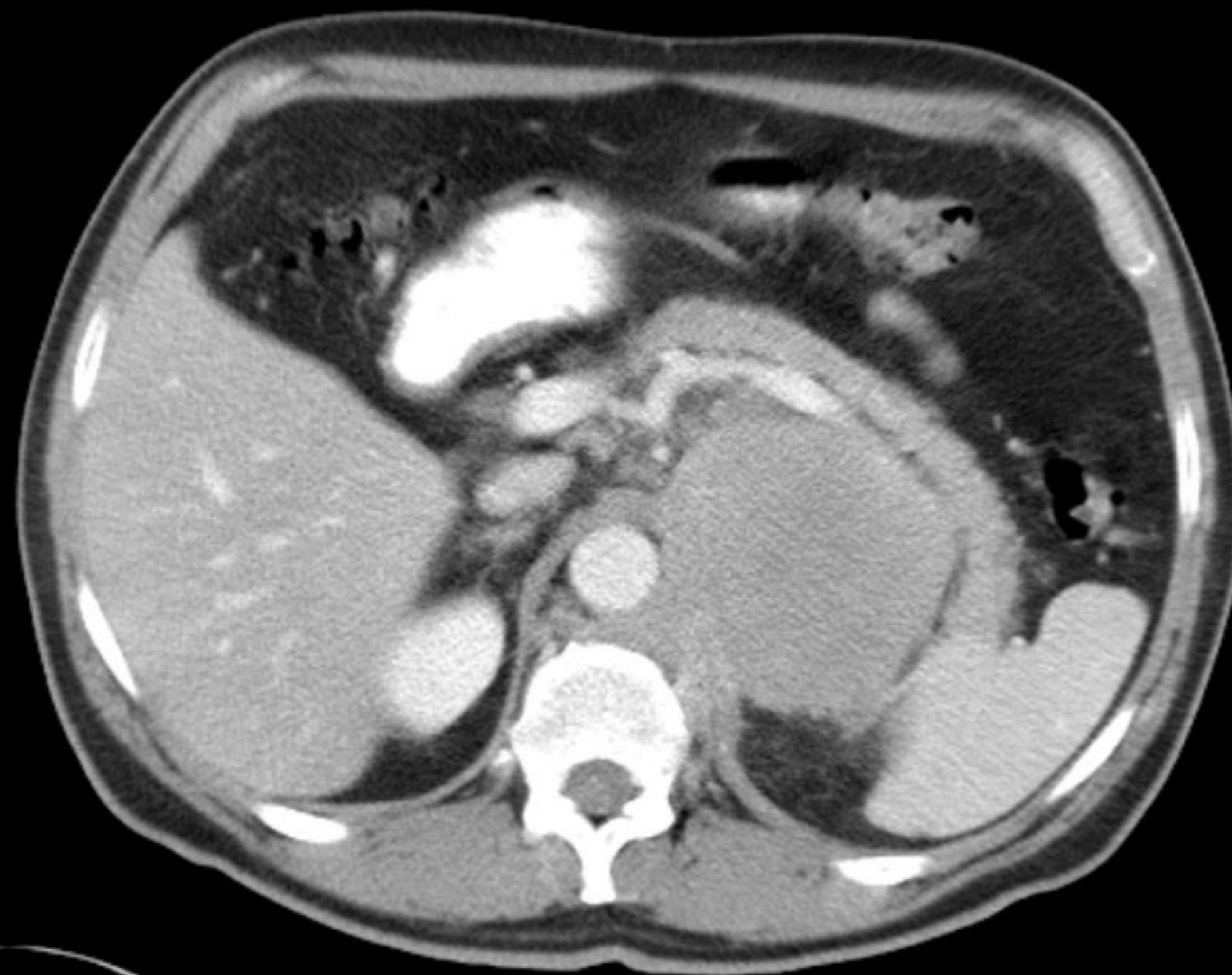
Why Biopsy?



**Establish
diagnosis:**
eliminate
nonoperative
pathology

Image size: 512 x 512
View size: 1499 x 899
WL: 20 WW: 338

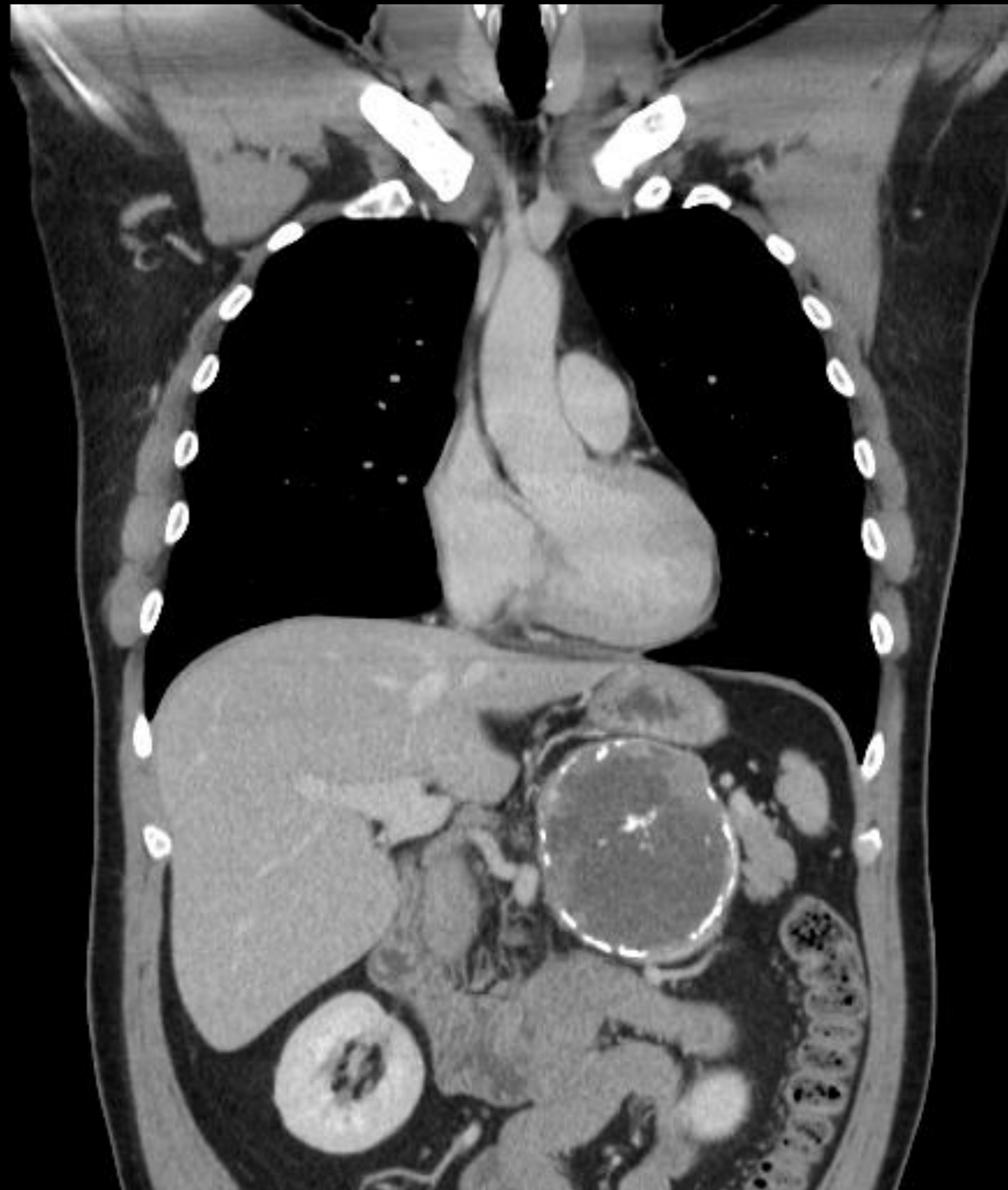
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Im: 29/61
Zoom: 176% Angle: 0
Thickness: 6.50 mm Location: -1055.50 mm



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Made In OsiriX





Why Biopsy?

**Establish
diagnosis:**
eliminate
nonoperative
pathology

**Identify
histologic
subtype:**
tailor treatment
strategy



How do histologic subtype and management strategy AFFECT RECURRENCE?

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 Rutkowski, 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†††*

Background: Retroperitoneal sarcomas (RPS) are rare tumors composed of several well defined histologic subtypes. The aim of this study was to analyze patterns of recurrence and treatment variations in a large population of patients, treated at reference centers.

Methods: All consecutive patients with primary RPS treated at 6 European and 2 North American institutions between January 2002 and December 2011 were included. Five, 8, and 10-year overall survival (OS) and crude cumulative incidence (CCI) of local recurrence (LR) and distant metastasis (DM) were calculated. Multivariate analyses for OS, CCI of LR, and DM were performed.

Results: In all, 1007 patients were included. Median follow-up was 58 months (first and third quartile range 36–90). The 5, 8, and 10-year OS were 67% [95% confidence interval (CI), 63, 70], 56% (95% CI, 52, 61), and

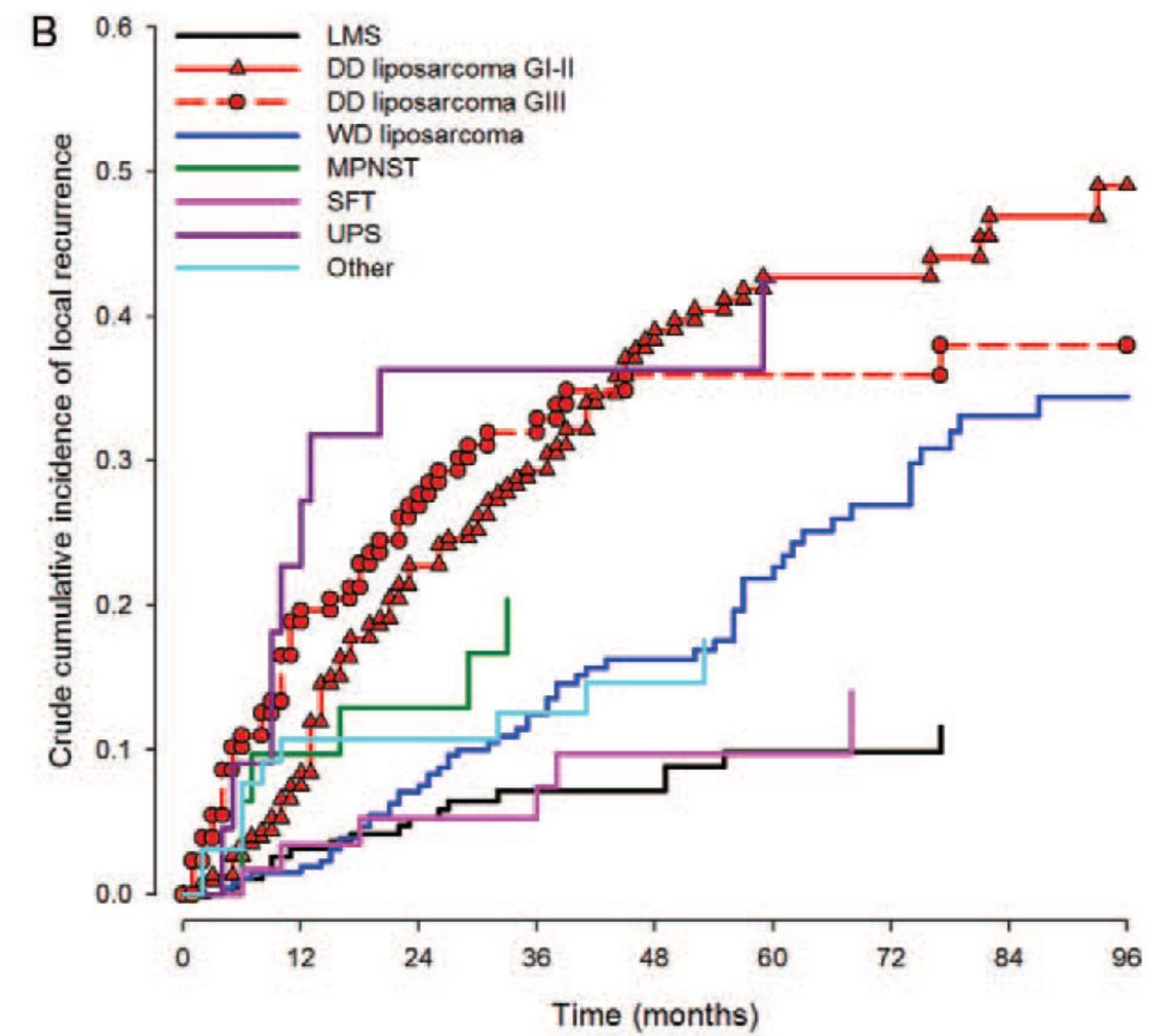
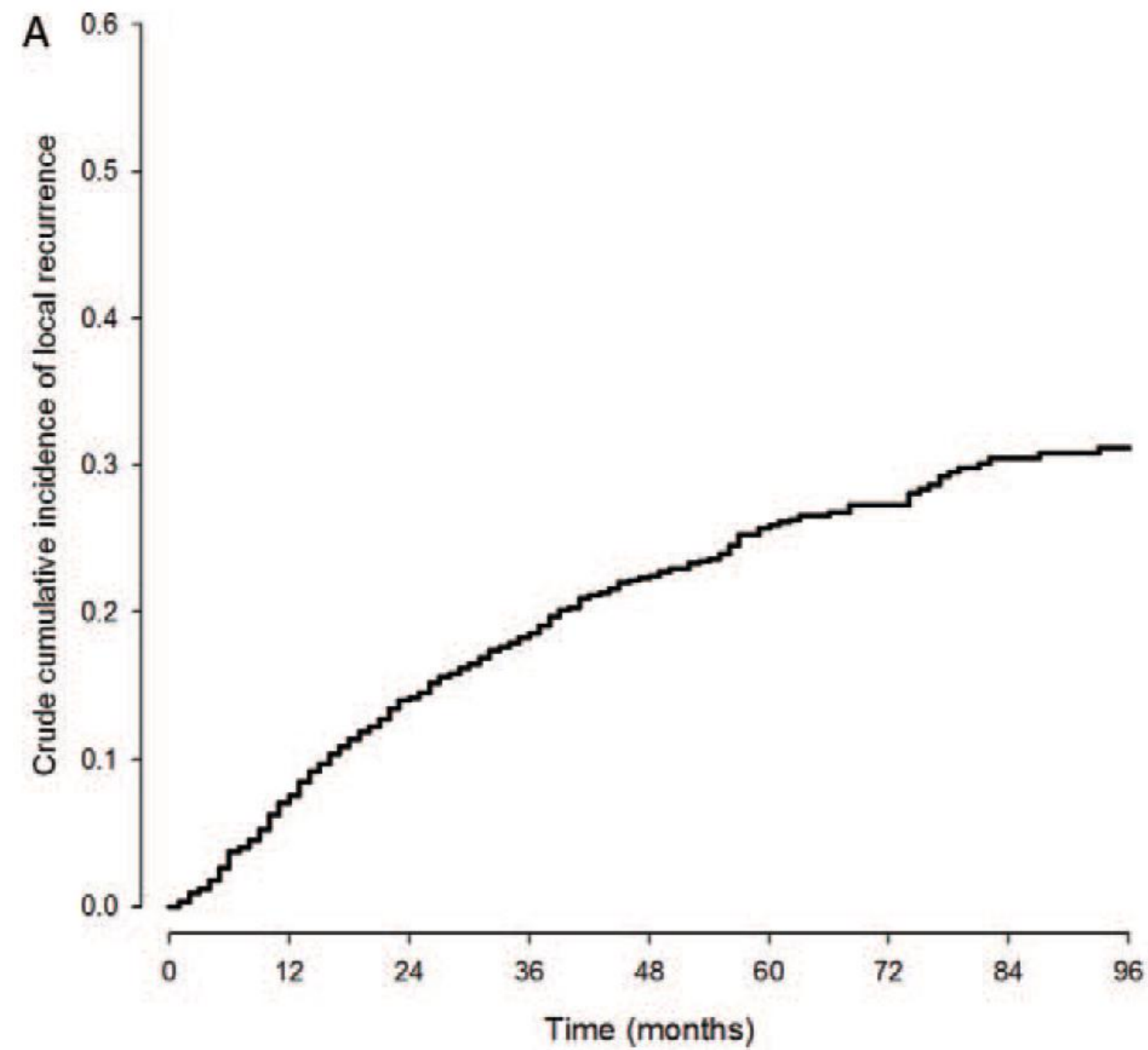
Different treatment policies at participating institutions influenced LR of well differentiated liposarcoma without impacting OS, whereas discrepancies in adjuvant systemic therapies did not impact LR, DM, or OS of leiomyosarcoma.

Conclusions: Reference centers are critical to outcomes of RPS patients, as the management strategy requires specific expertise. Histologic subtype predicts patterns of recurrence and should inform management decision. A prospective international registry is under preparation, to further define our understanding of this disease.

Keywords: leiomyosarcoma, liposarcoma, prognostic factors, retroperitoneal sarcoma, sarcoma, solitary fibrous tumor, surgery, survival

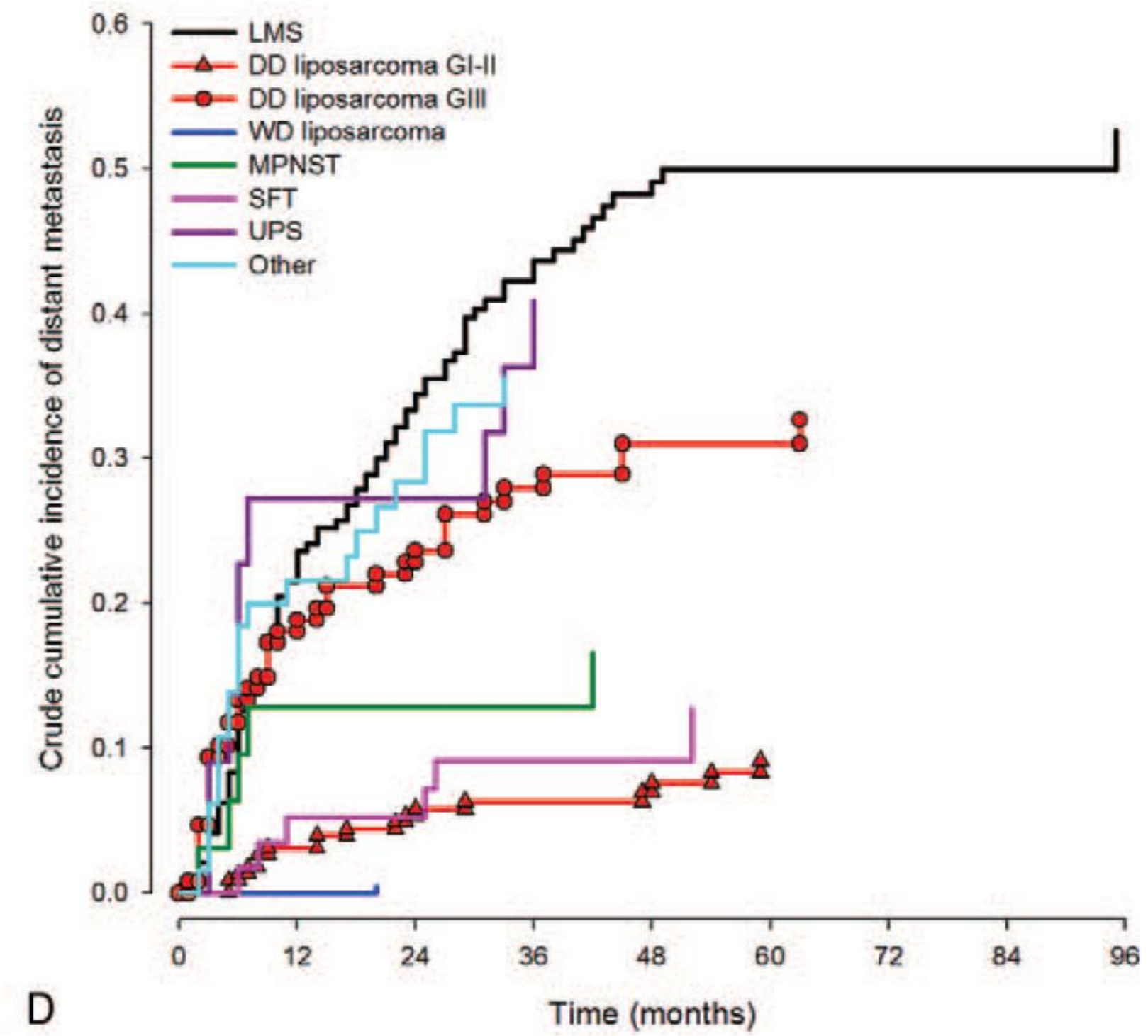
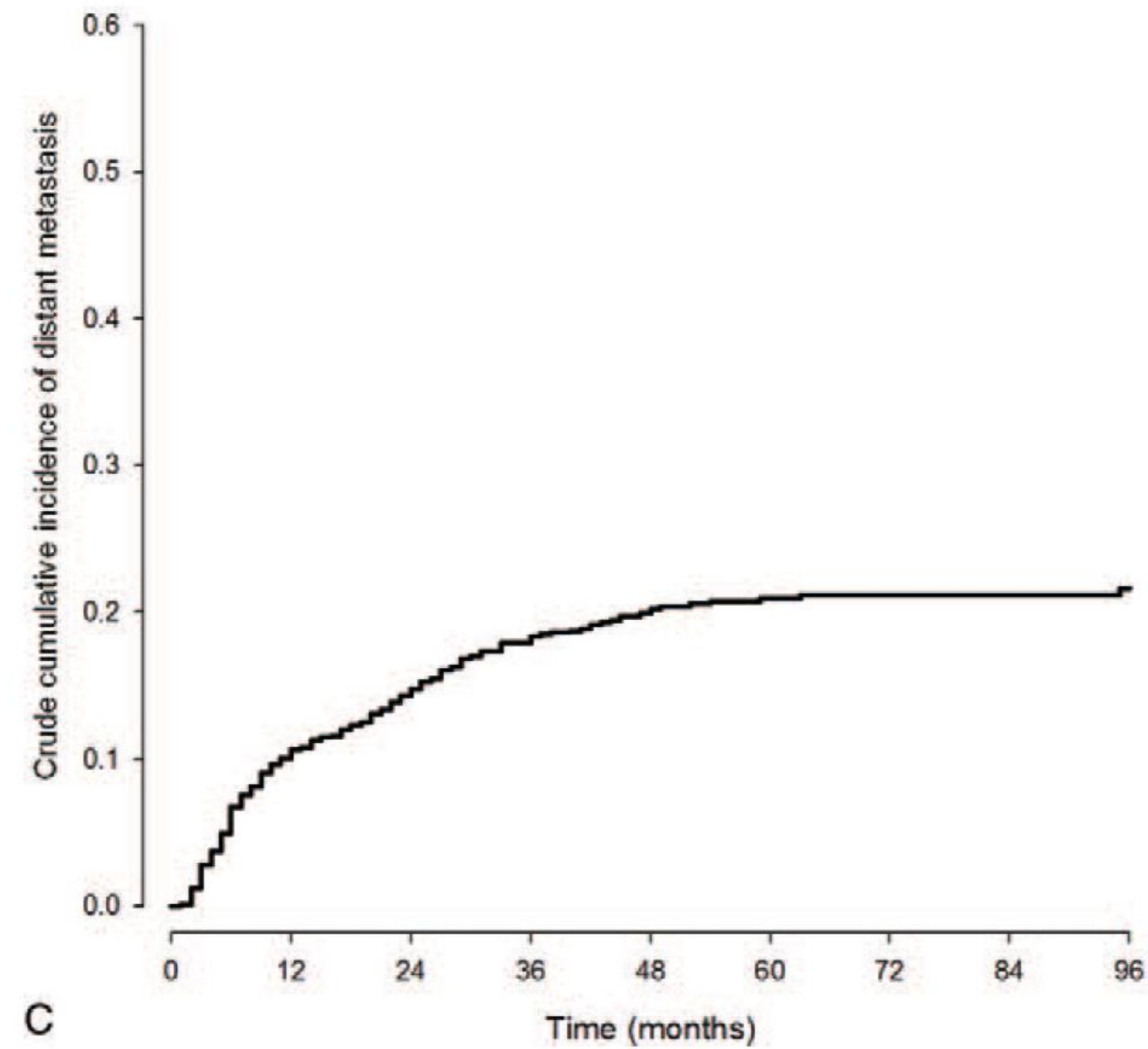
(*Ann Surg* 2015;xx:xxx–xxx)

Local Recurrence



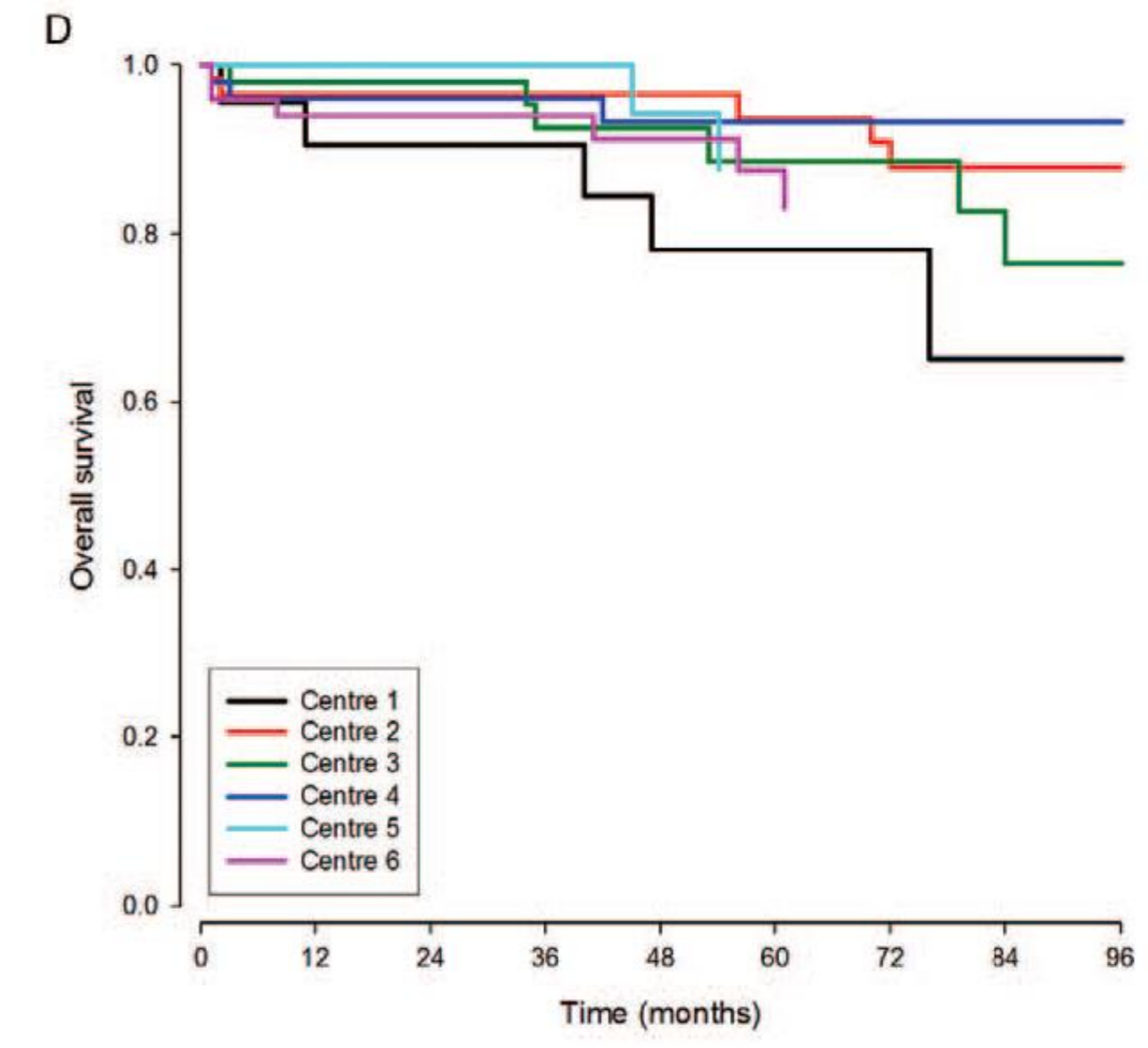
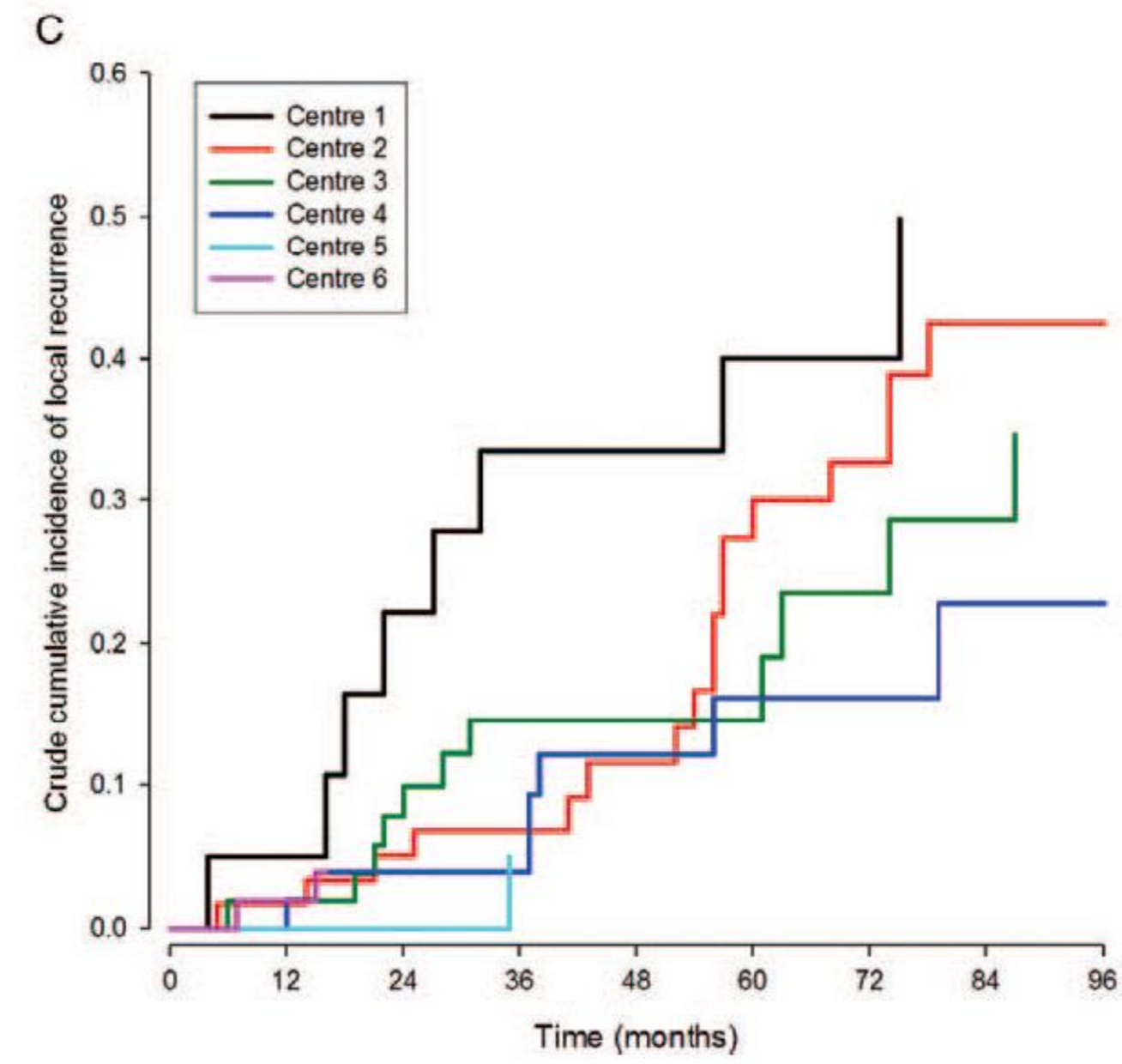
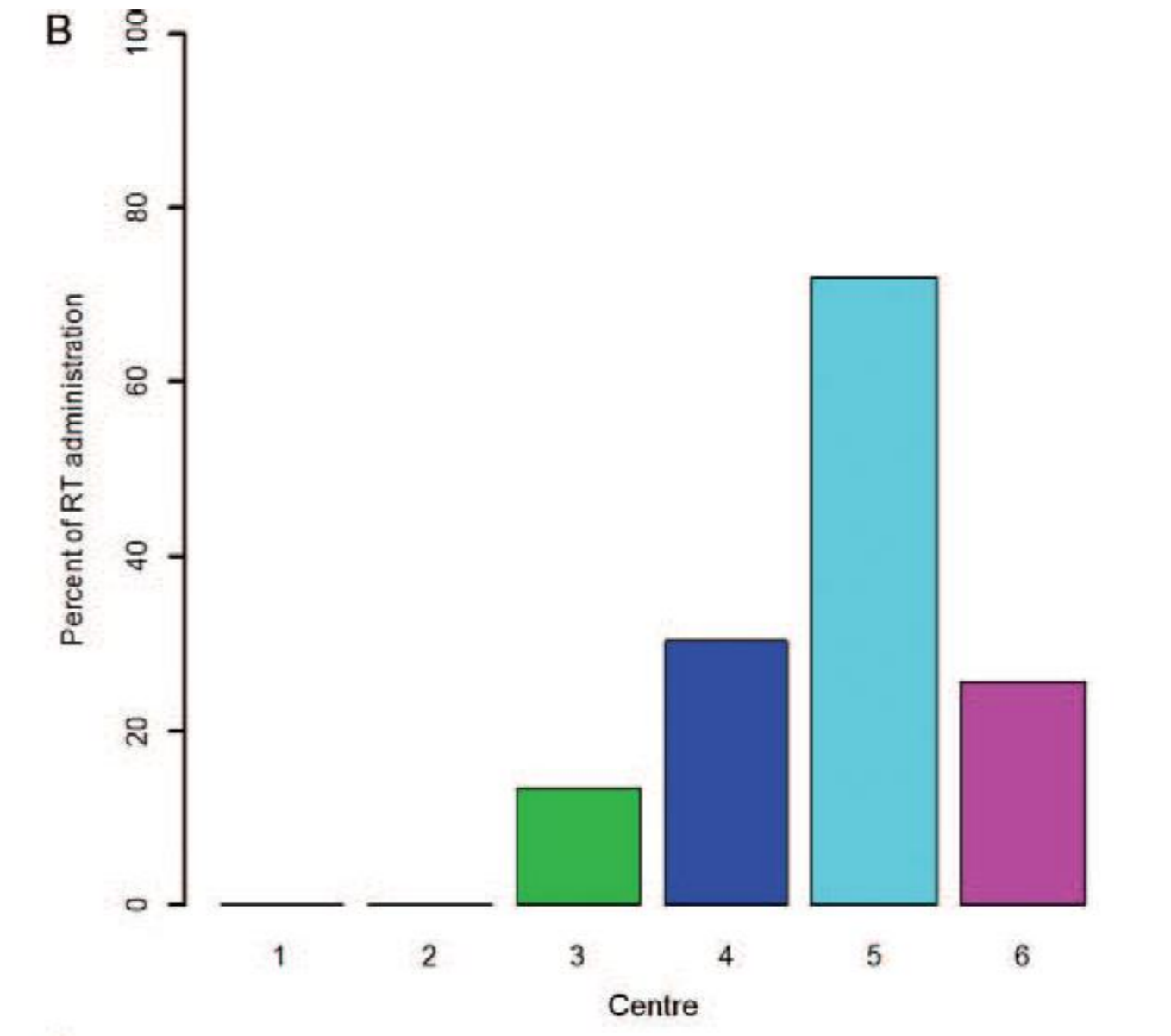
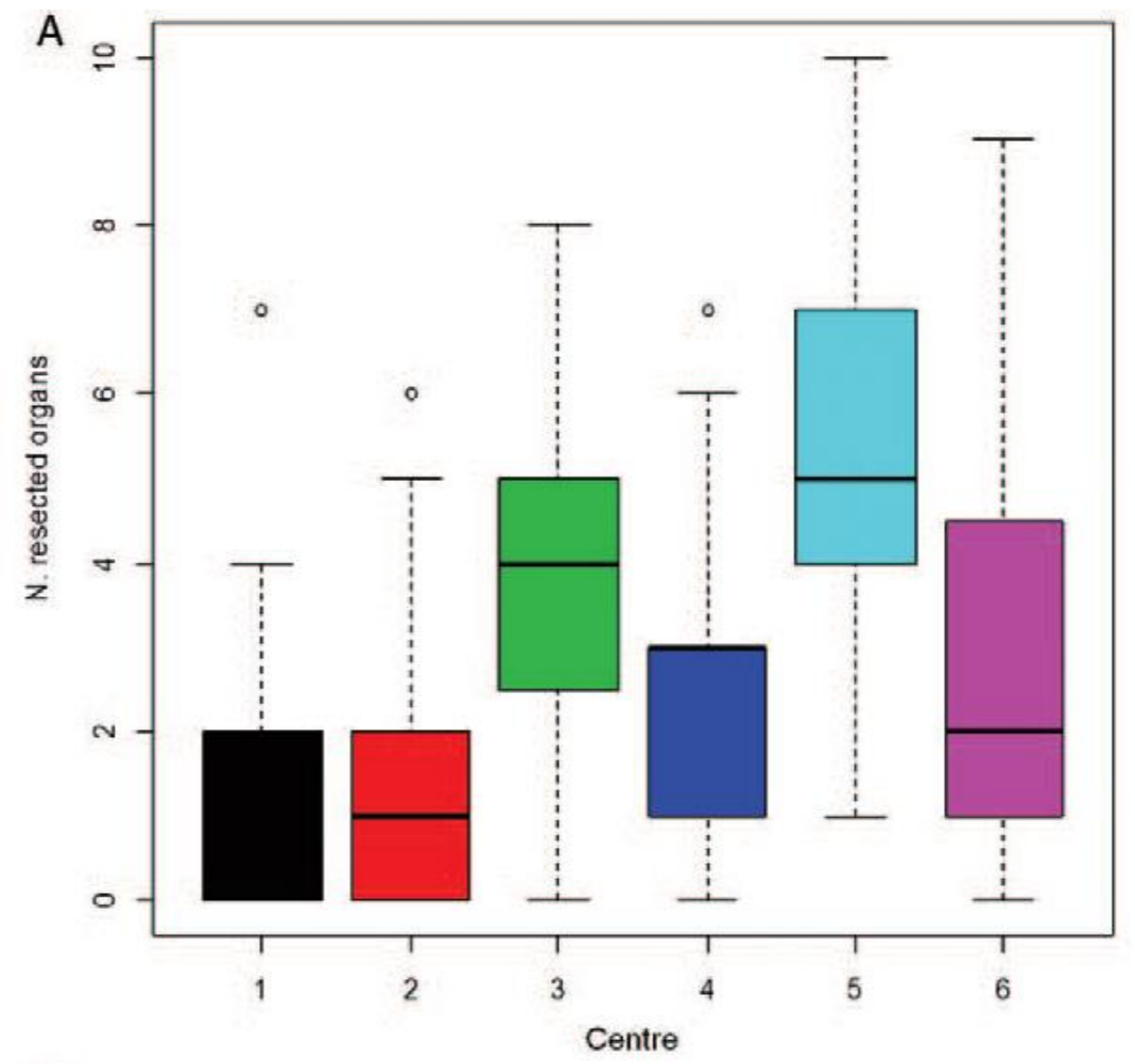
5y LR 24%
10y LR 33%

Distant Metastasis

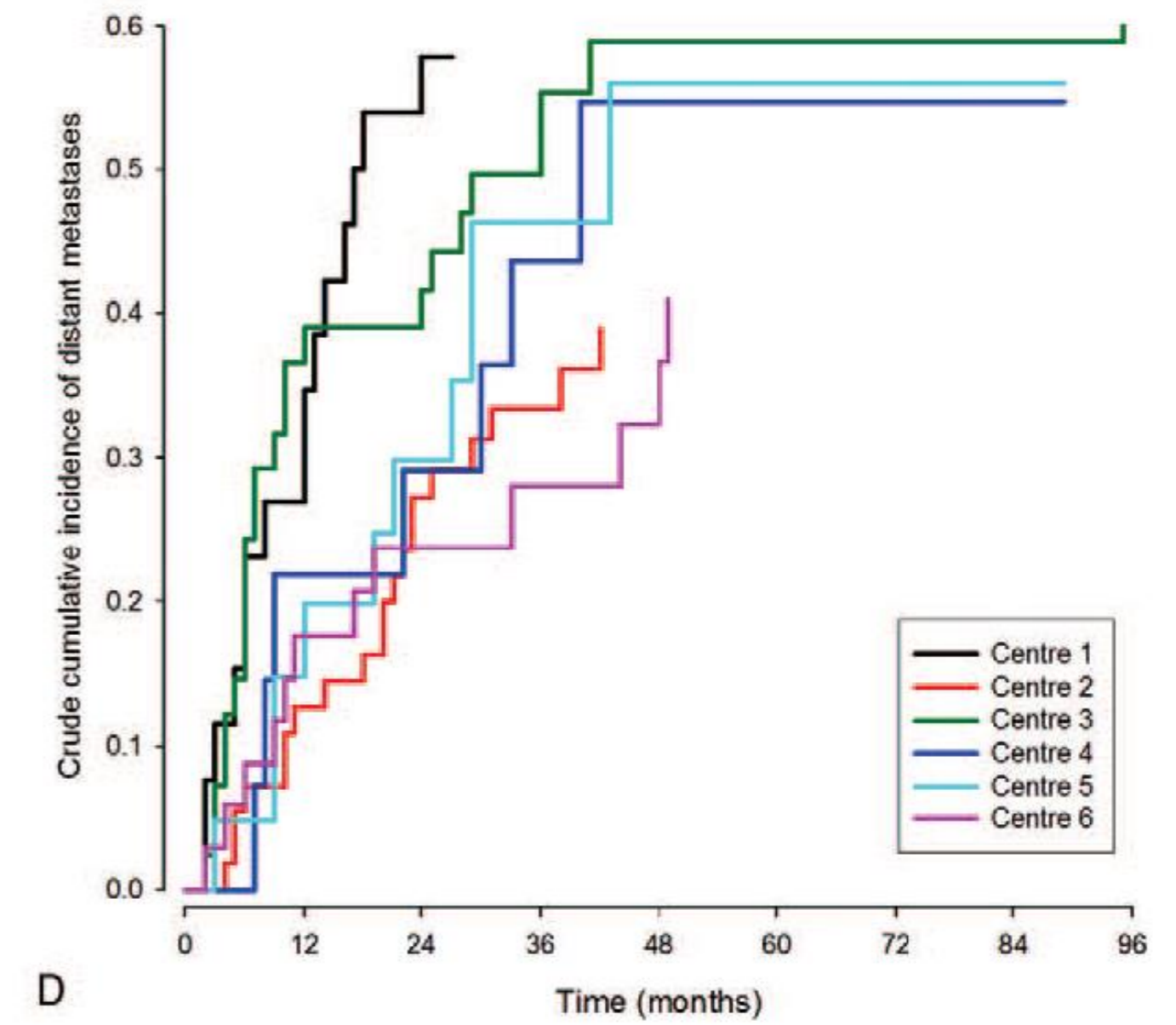
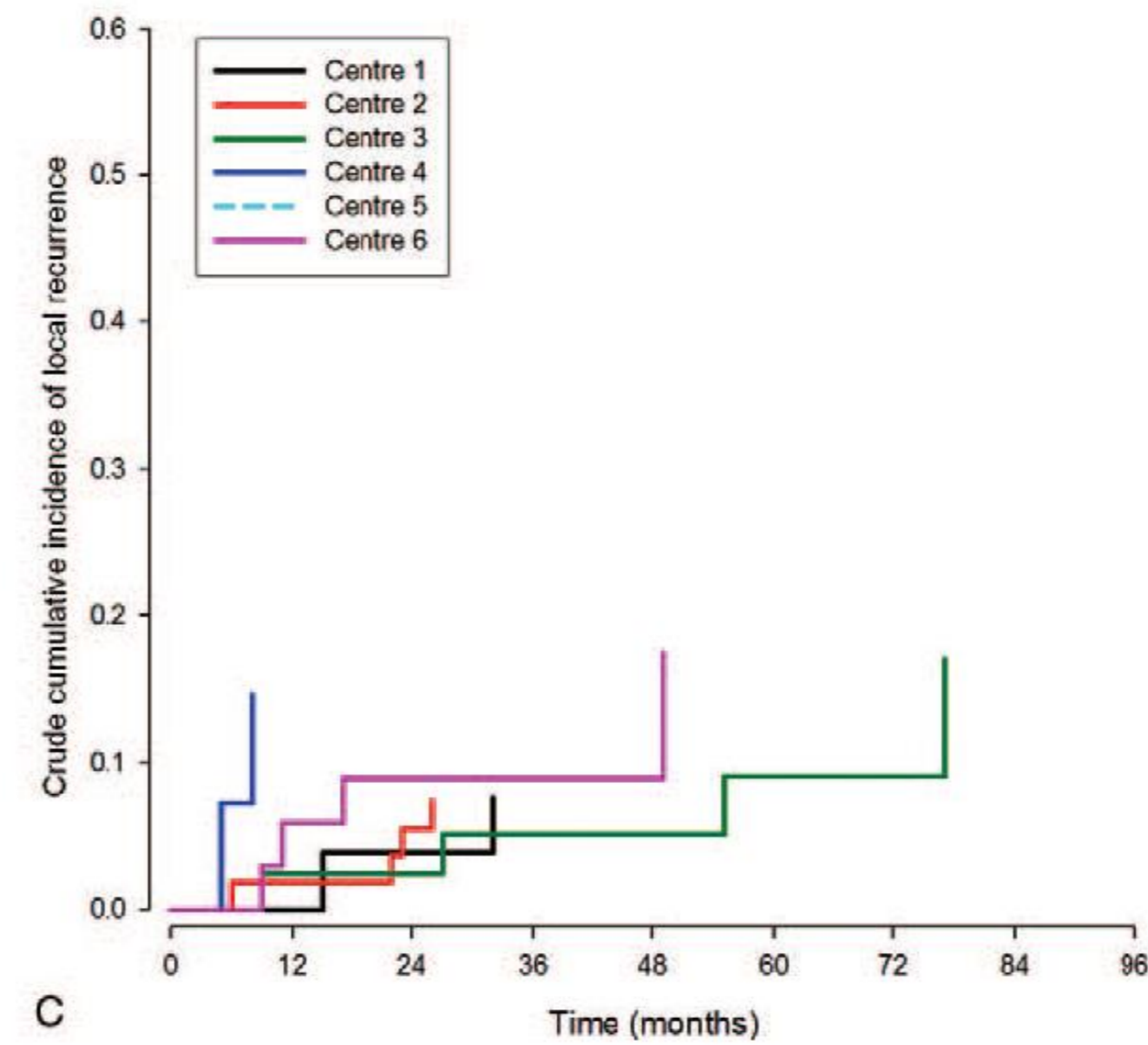
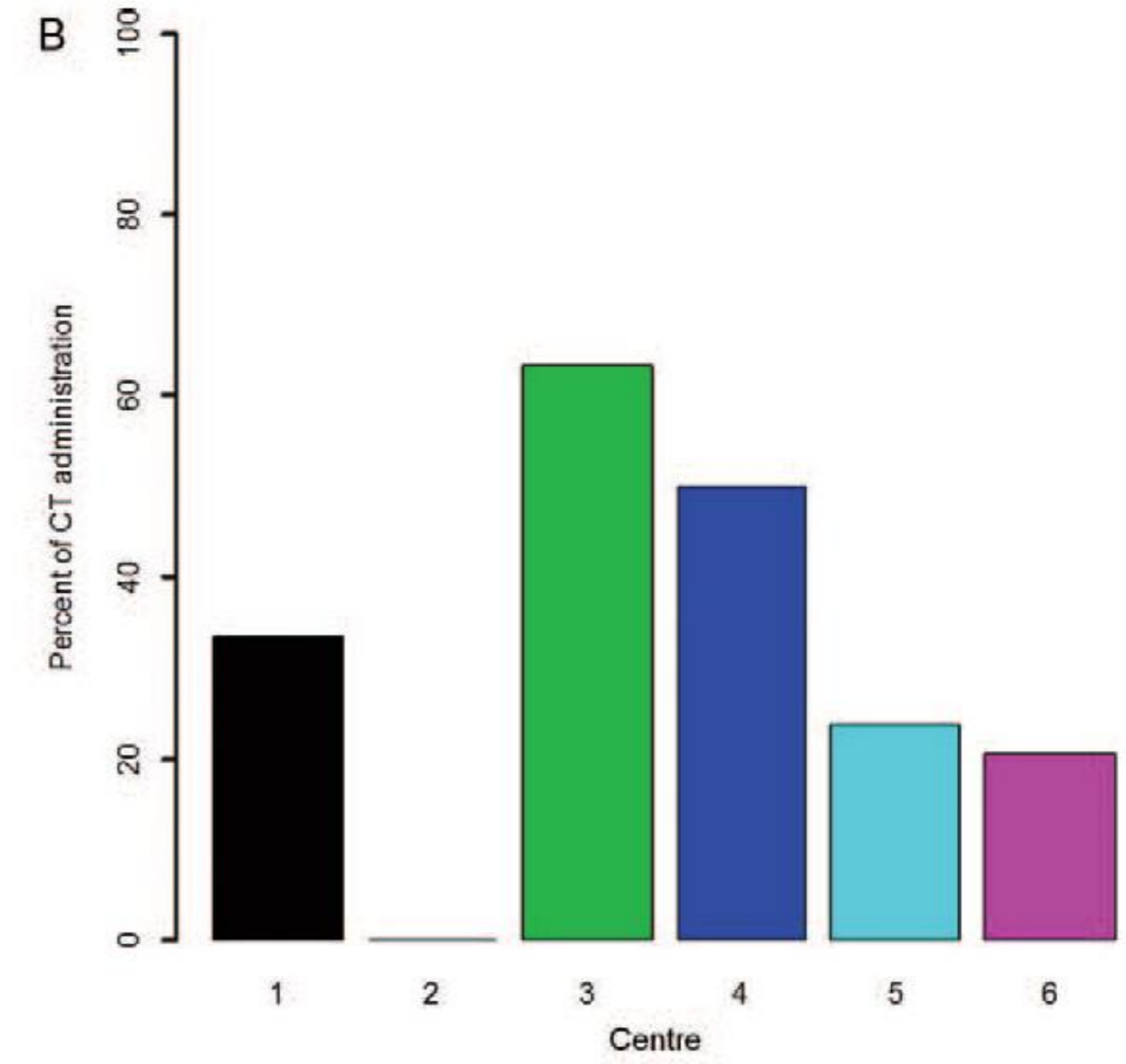
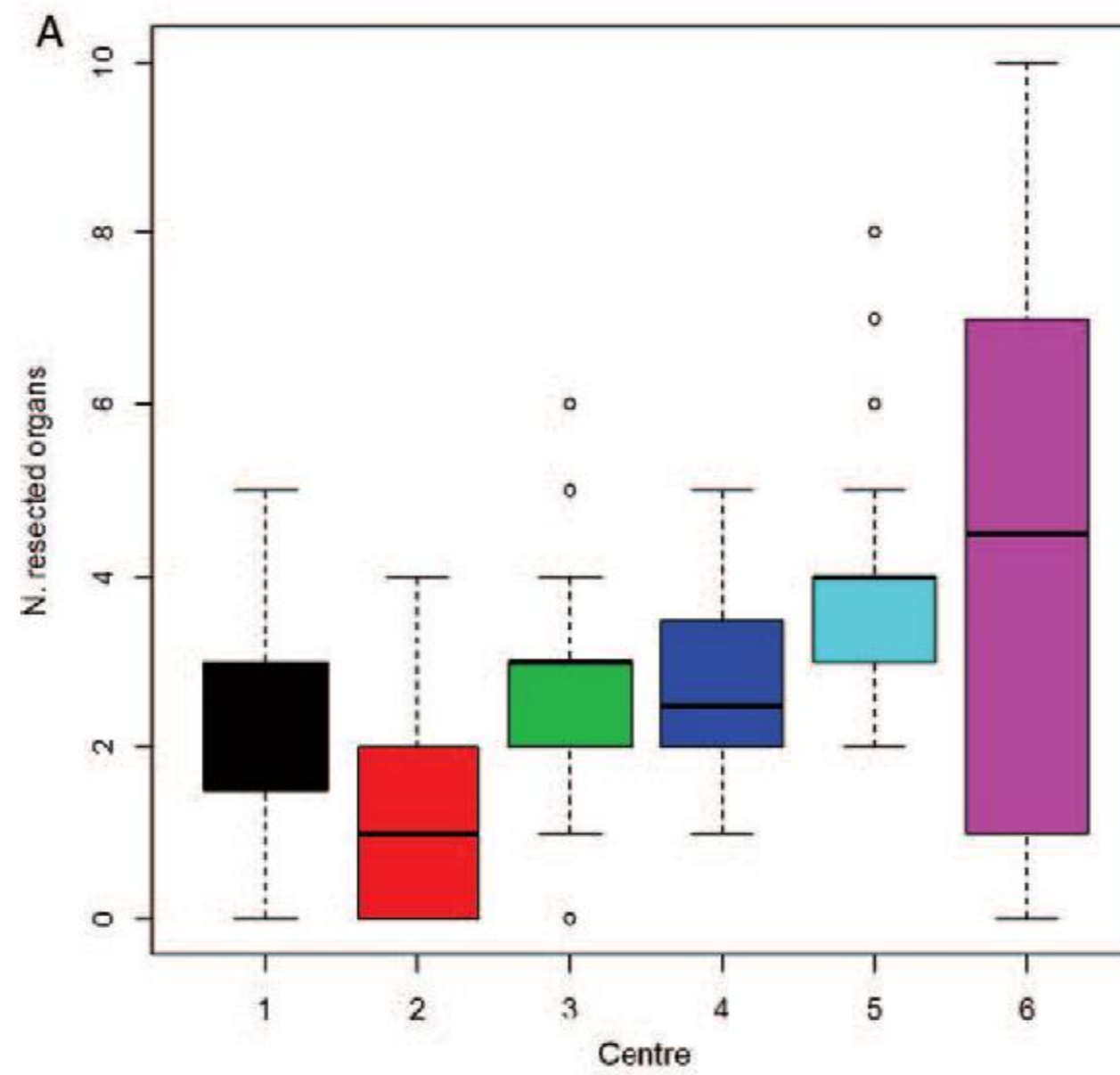


5y DM 21%
10y DM 21.6%

Institutional Management Strategies: WDLPS



Institutional Management Strategies: LMS



Why Biopsy?

**Establish
diagnosis:**
eliminate
nonoperative
pathology

**Identify
histologic
subtype:**
tailor treatment
strategy

**Consider
neoadjuvant
therapy:**
radiate first

Radiation Therapy in RPS

Goal:

- Increase R0 resection rate
- Decrease local recurrence rate

Level 1 evidence of improved local control in extremity STS, no survival benefit

EORTC study 62092-22092

STRASS - A phase III randomized study of preoperative radiotherapy plus surgery versus surgery alone for patients with Retroperitoneal sarcoma (RPS)

Pre-op RT

- More accurate targeting of tumour volume
- Minimal toxicity
- Improved delivery to well-oxygenated tissues
- Increased likelihood of negative margin
- Reduced chance of intraoperative contamination/tumour rupture

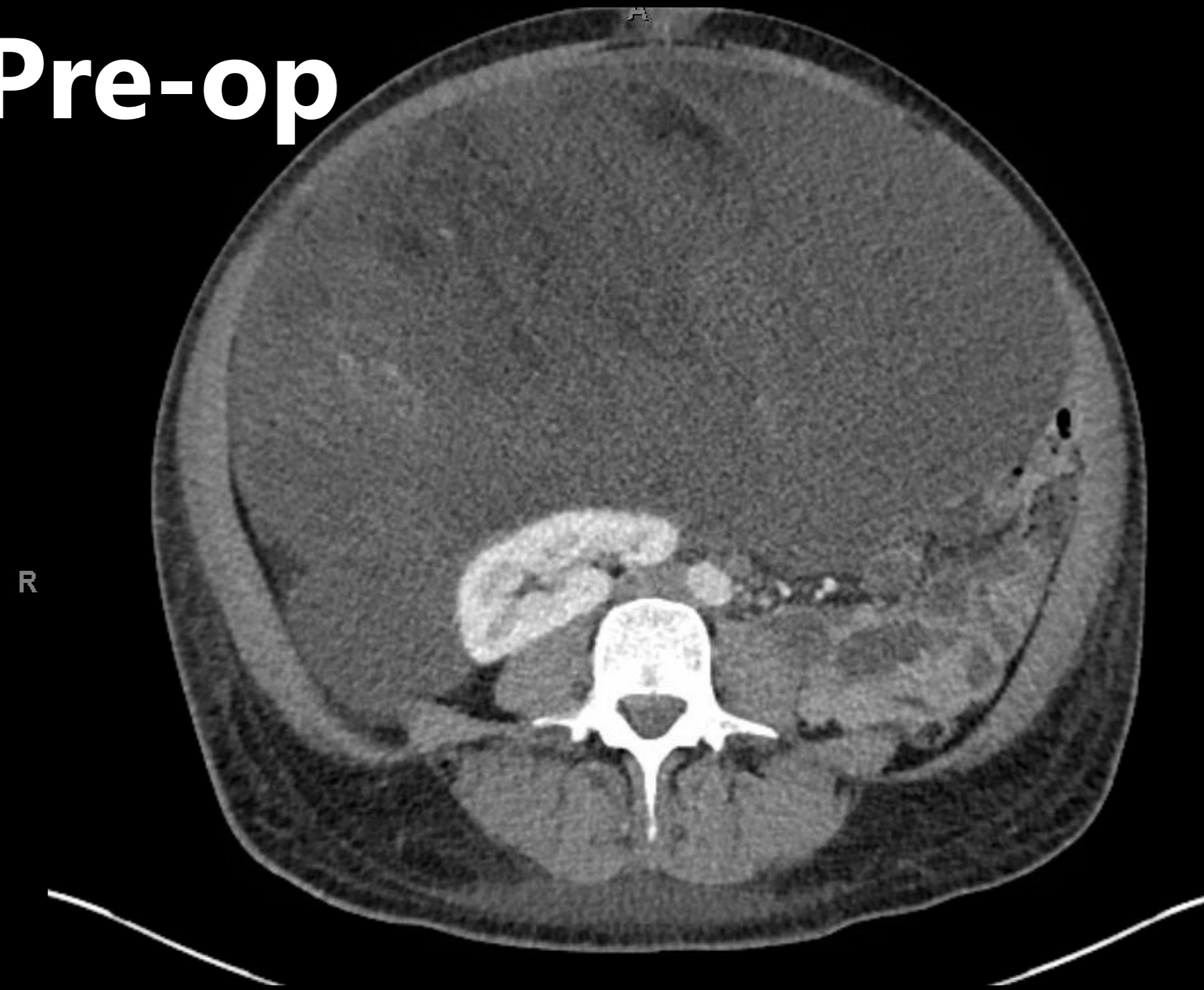
Pre-op RT

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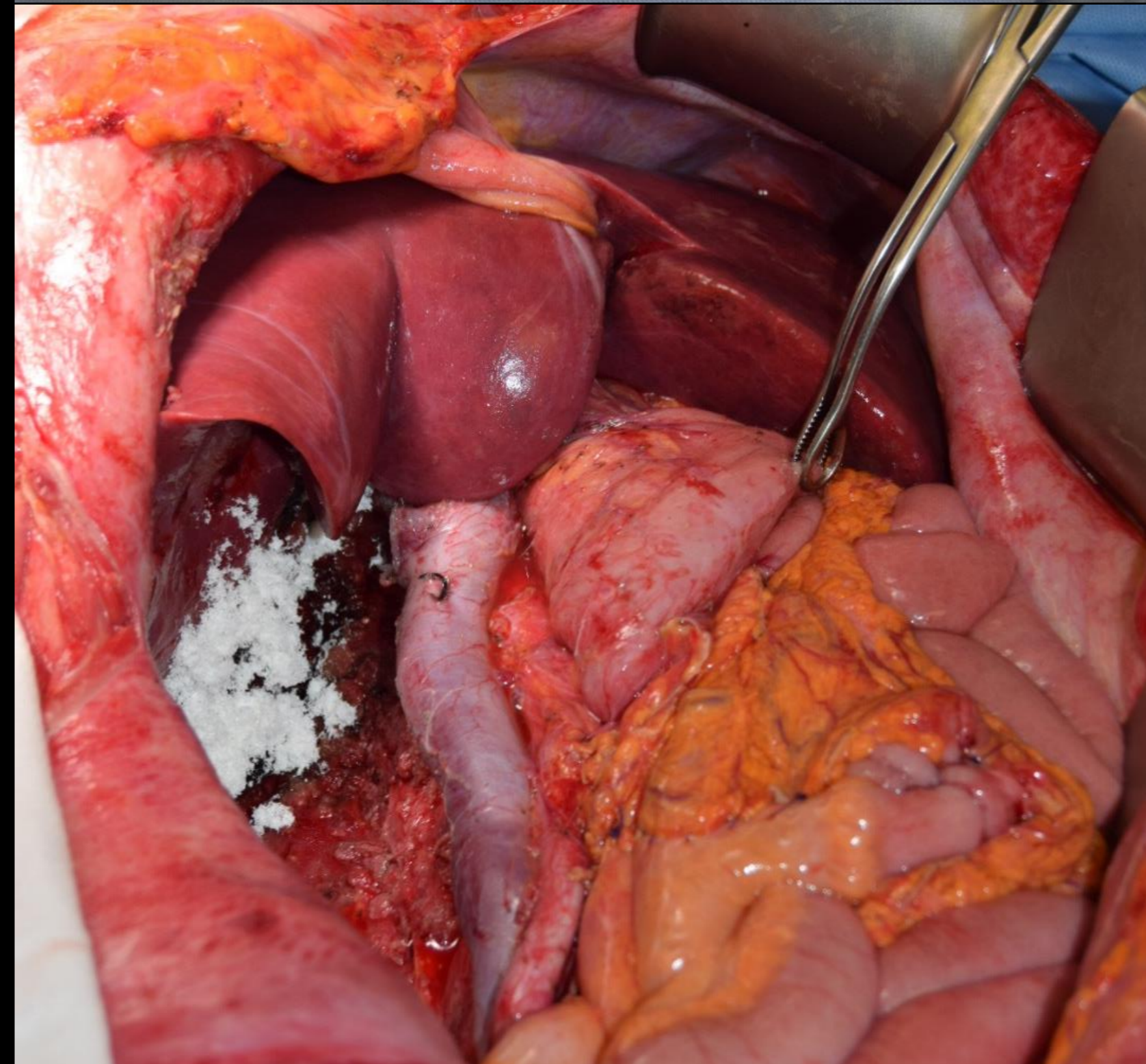
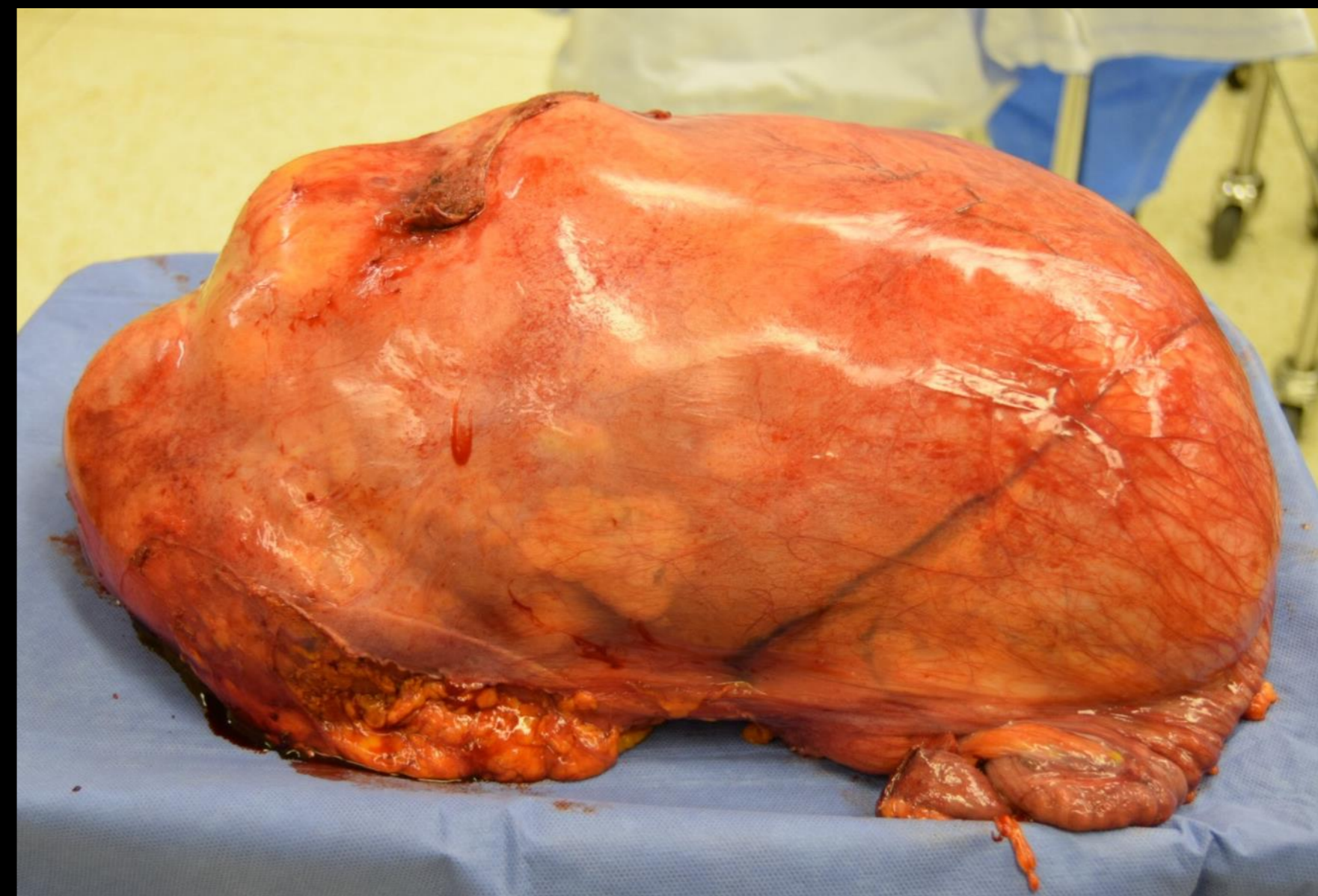
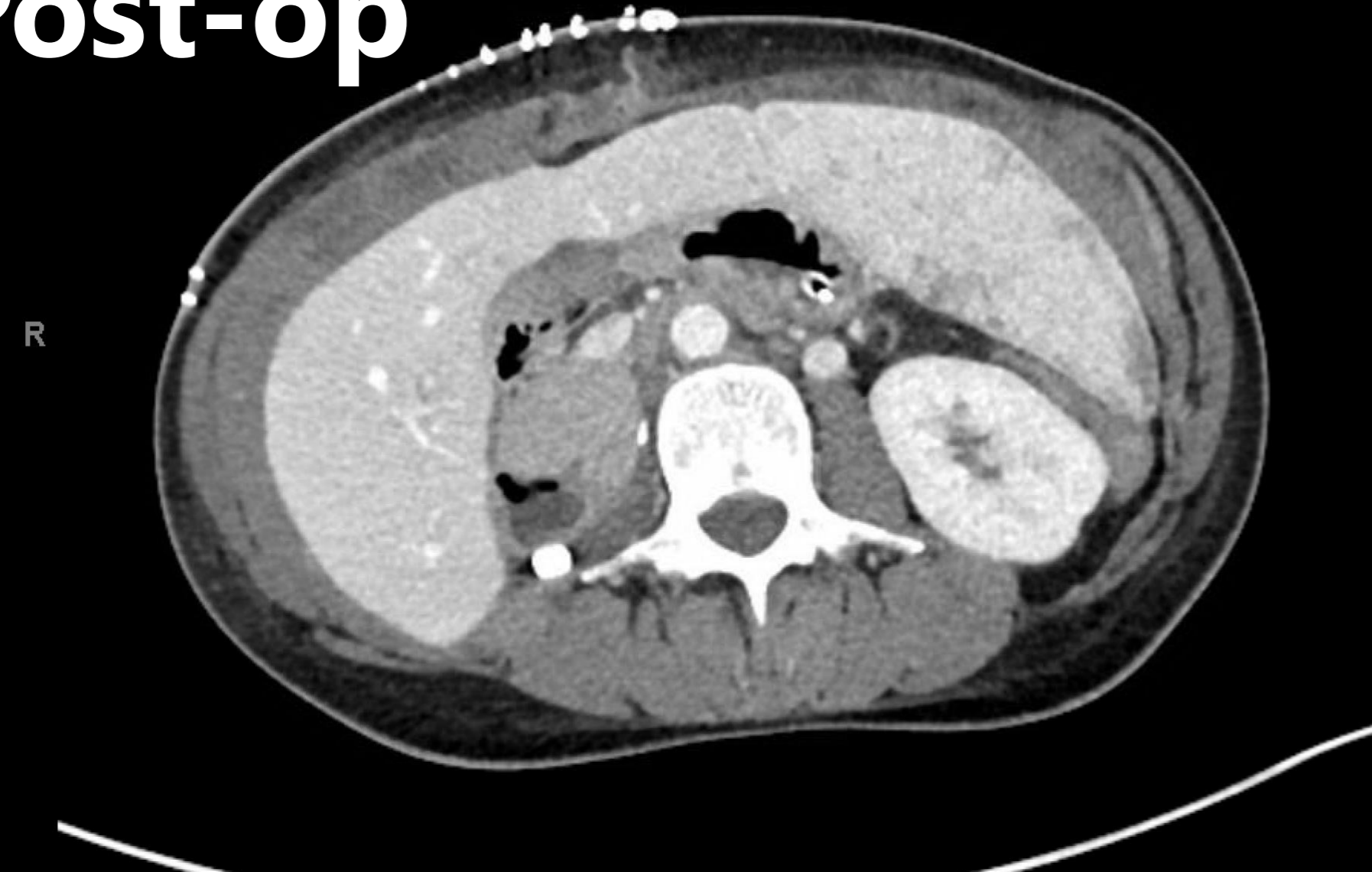
Post-op RT

- **NOT POSSIBLE!**

Pre-op



Post-op



How (not) to Biopsy?

**At sarcoma
referral centre**

**Via RP
approach**

**Targeting
dedifferentiated
areas**

**With expert
pathology
review**

How (not) to Biopsy?

At sarcoma

with DD

**No role for transperitoneal/
laparoscopic/open biopsy**

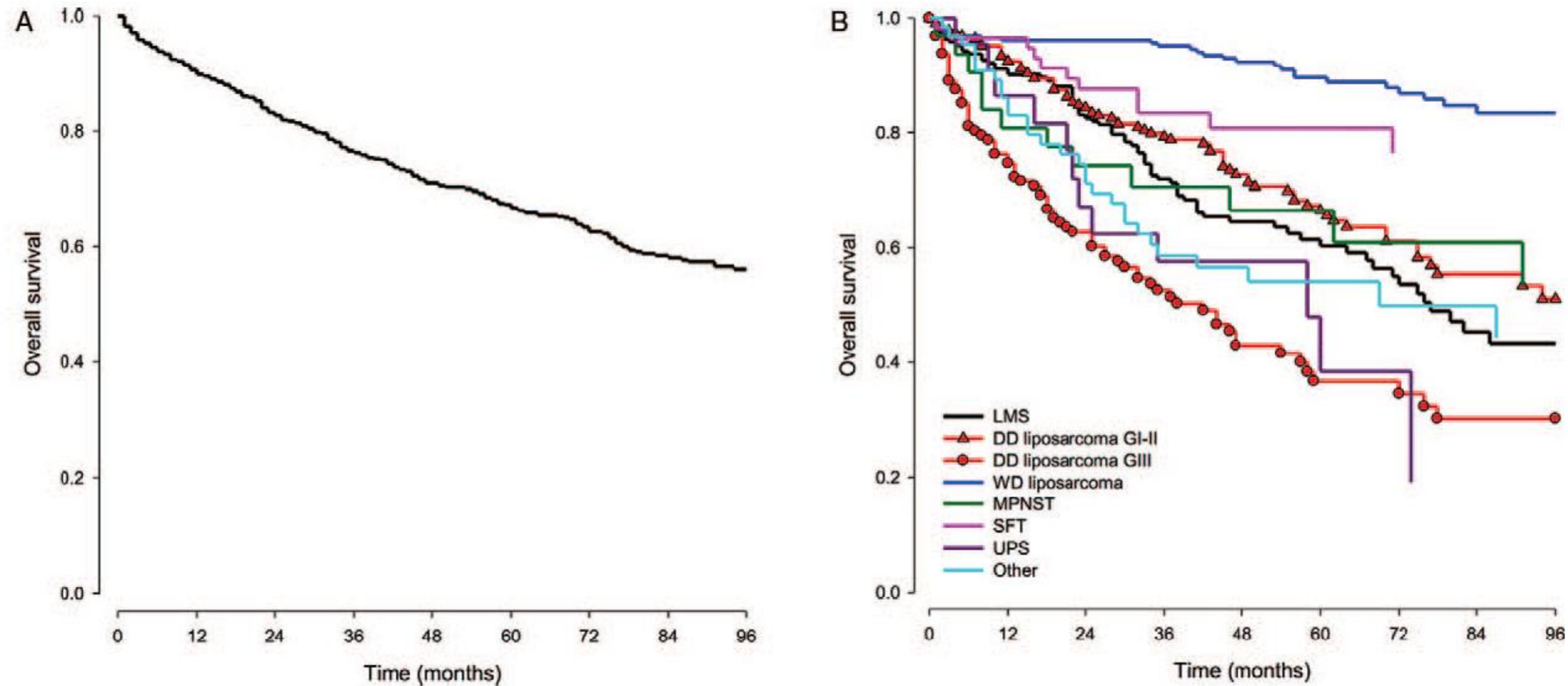
dedifferentiated
areas

pathology
review

Principles and Pitfalls

- **Best outcomes achieved at high-volume centres**

Overall Survival – TARPSWG centres



5y OS 67%
10y OS 46%

Principles and Pitfalls

- **Best outcomes achieved at high-volume centres**
- **All RPS require MDC review**

Principles and Pitfalls

- **Best outcomes achieved at high-volume centres**
- **All RPS require MDC review**

- **Large mass \neq emergency**



A

Principles and Pitfalls

- **Best outcomes achieved at high-volume centres**
- **All RPS require MDC review**

- **Large mass \neq emergency**
- **Incidental finding at laparotomy/inguinal hernia repair – do not biopsy, avoid mesh**



Take home messages

- **RPS is a family of diseases**
- **Management is multidisciplinary and must be tailored to histology**
- **Preoperative tissue diagnosis is imperative**
- **Outcomes have improved with extended resection, referral to high-volume centres**



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