

# Chordoma Community Conference 2022

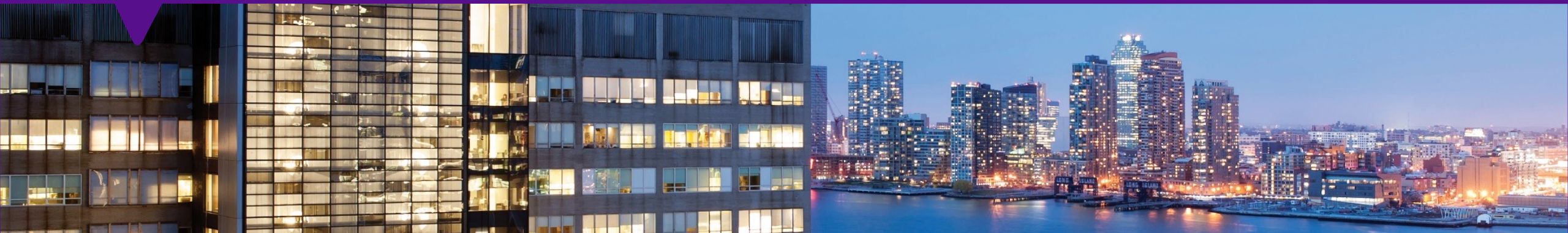
## *Sacral Surgery: Where Do We Stand?*

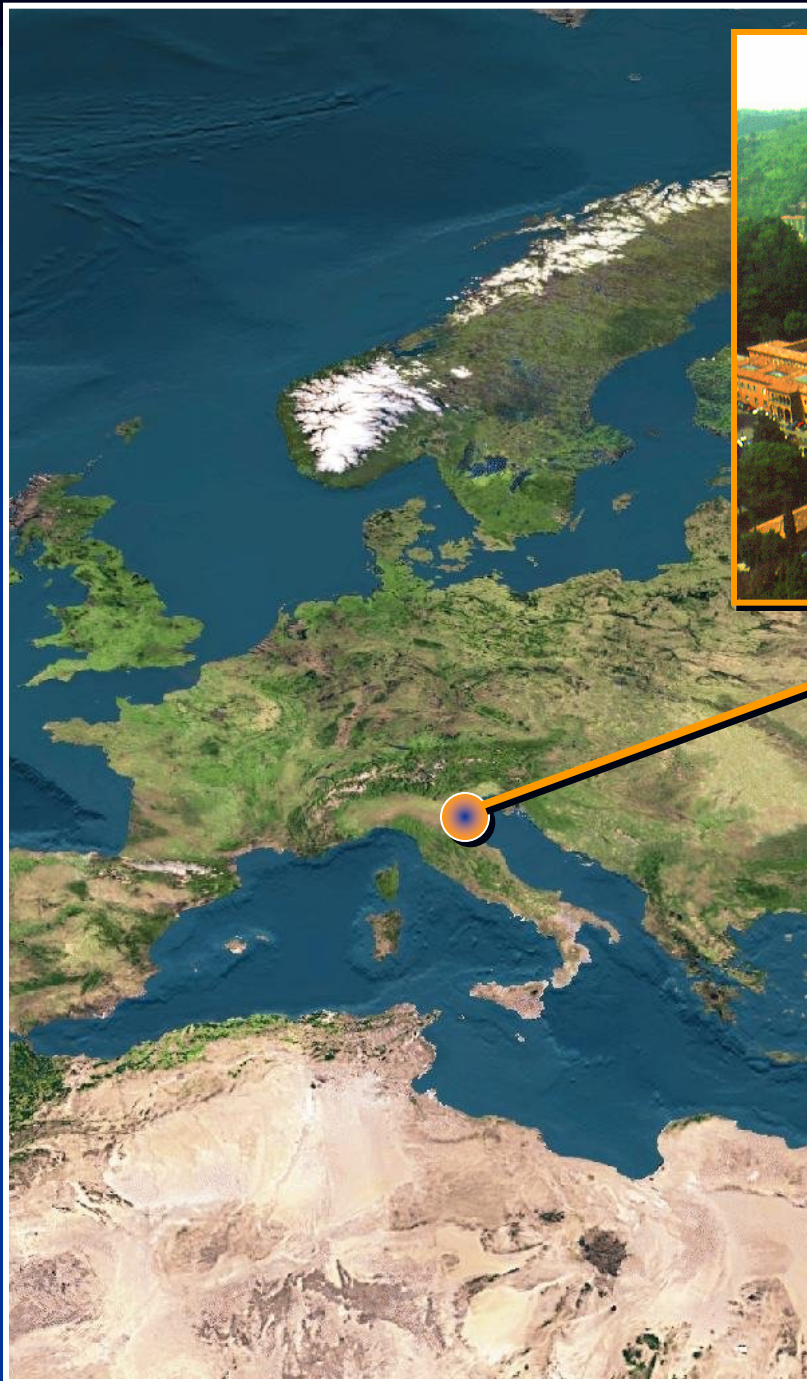
*Nicola Fabbri, MD*

*Chief Division, Orthopaedic Oncology*

*Professor, Orthopaedic Surgery*

*Department of Orthopaedic Surgery, NYU Langone*





University of Bologna Medical School

Orthopaedic Surgery Residency Program

Istituto Ortopedico Rizzoli - Bologna



# Fellowship

MS Oncology : 1995 Rizzoli



Adult Recon: 1996 Mayo Clinic

MS Oncology : 1997 Mayo Clinic



2013 - 2022



Memorial Sloan-Kettering  
Cancer Center



# Chordoma Community Conference 2022

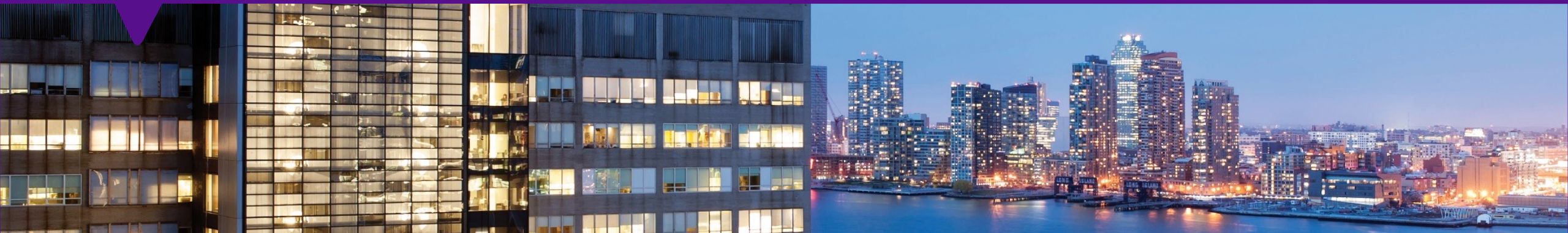
## *Sacral Surgery: Where Do We Stand?*

*Nicola Fabbri, MD*

*Chief Division, Orthopaedic Oncology*

*Professor, Orthopaedic Surgery*

*Department of Orthopaedic Surgery, NYU Langone*



# TUMORS OF THE SACRUM

<b>Primitive (on 20000 cases)</b>	<b>663</b>	
• <b>Chordoma</b>	<b>362</b>	<b>55%</b>
• <b>Giant Cell Tumor</b>	<b>80</b>	<b>12%</b>
• <b>Osteoblastoma</b>	<b>44</b>	<b>7%</b>
• <b>Chondrosarcoma</b>	<b>34</b>	<b>5%</b>
• <b>Osteosarcoma</b>	<b>27</b>	<b>4%</b>
• <b>Lymphoma</b>	<b>25</b>	<b>4%</b>
• <b>Ewing' sarcoma</b>	<b>22</b>	<b>3%</b>
• <b>Myeloma</b>	<b>21</b>	<b>3%</b>
• <b>Fibrosarcoma</b>	<b>22</b>	<b>3%</b>
<b>/M.F.Histiocytoma</b>		

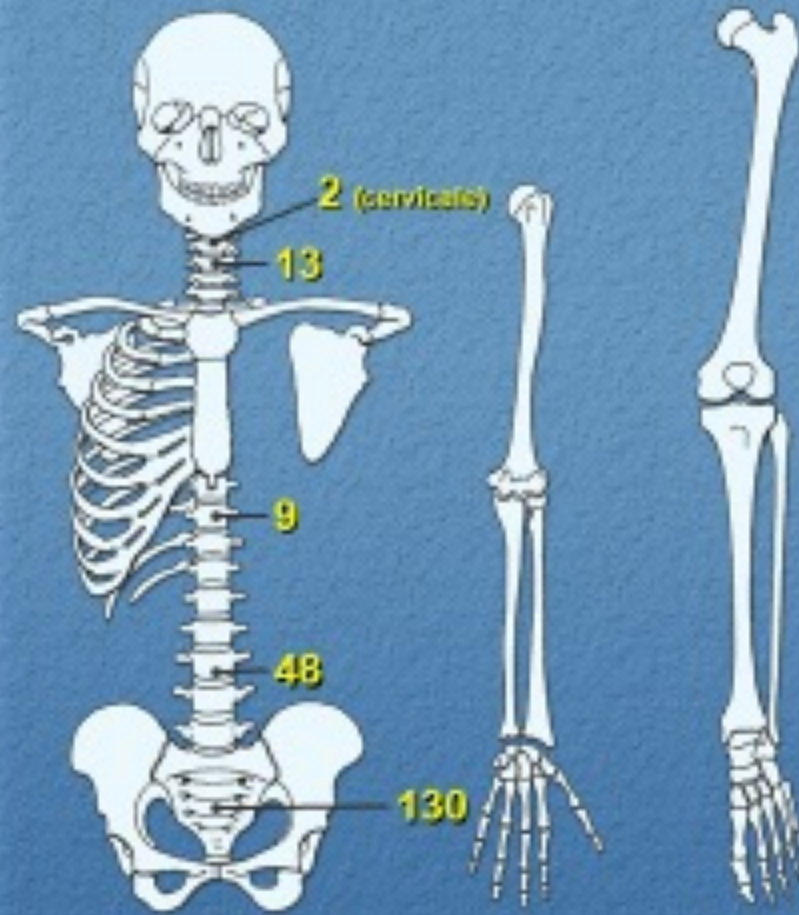
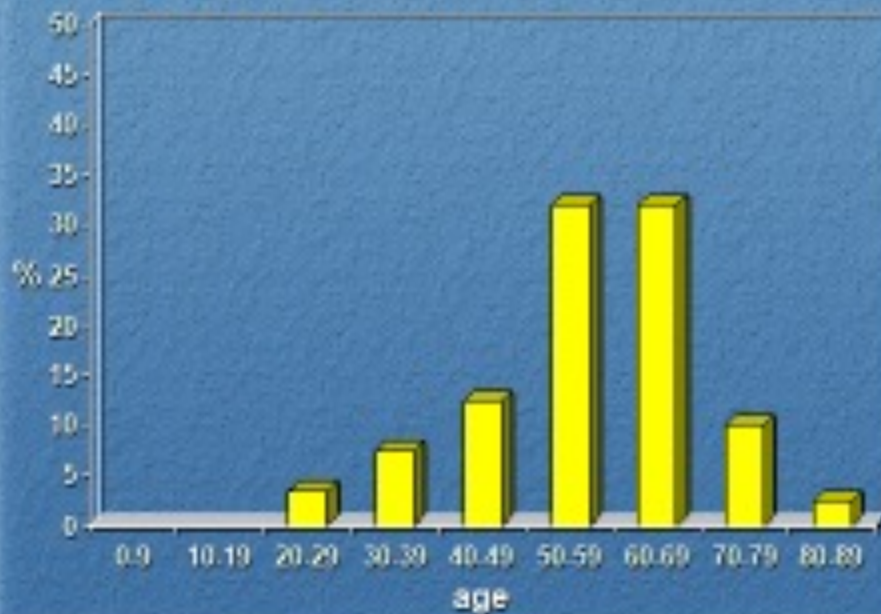


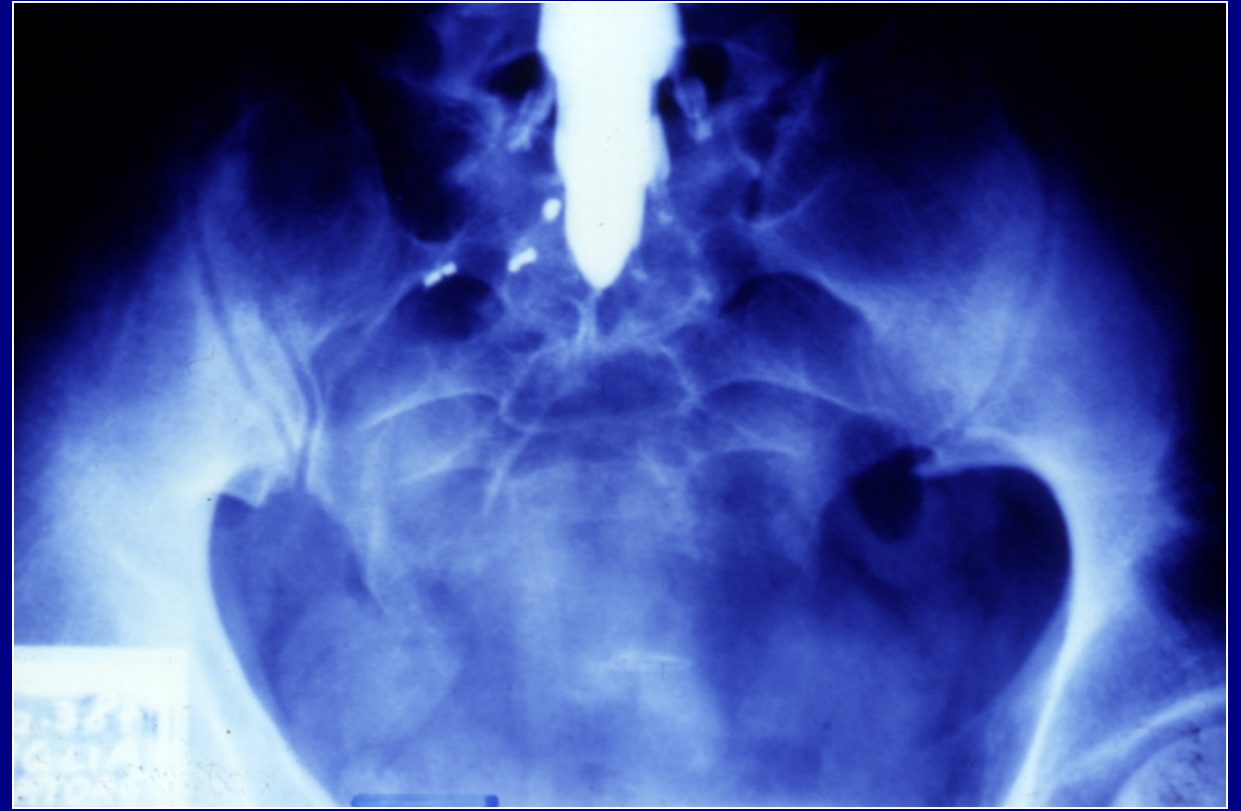
# Chordoma

## 202 cases

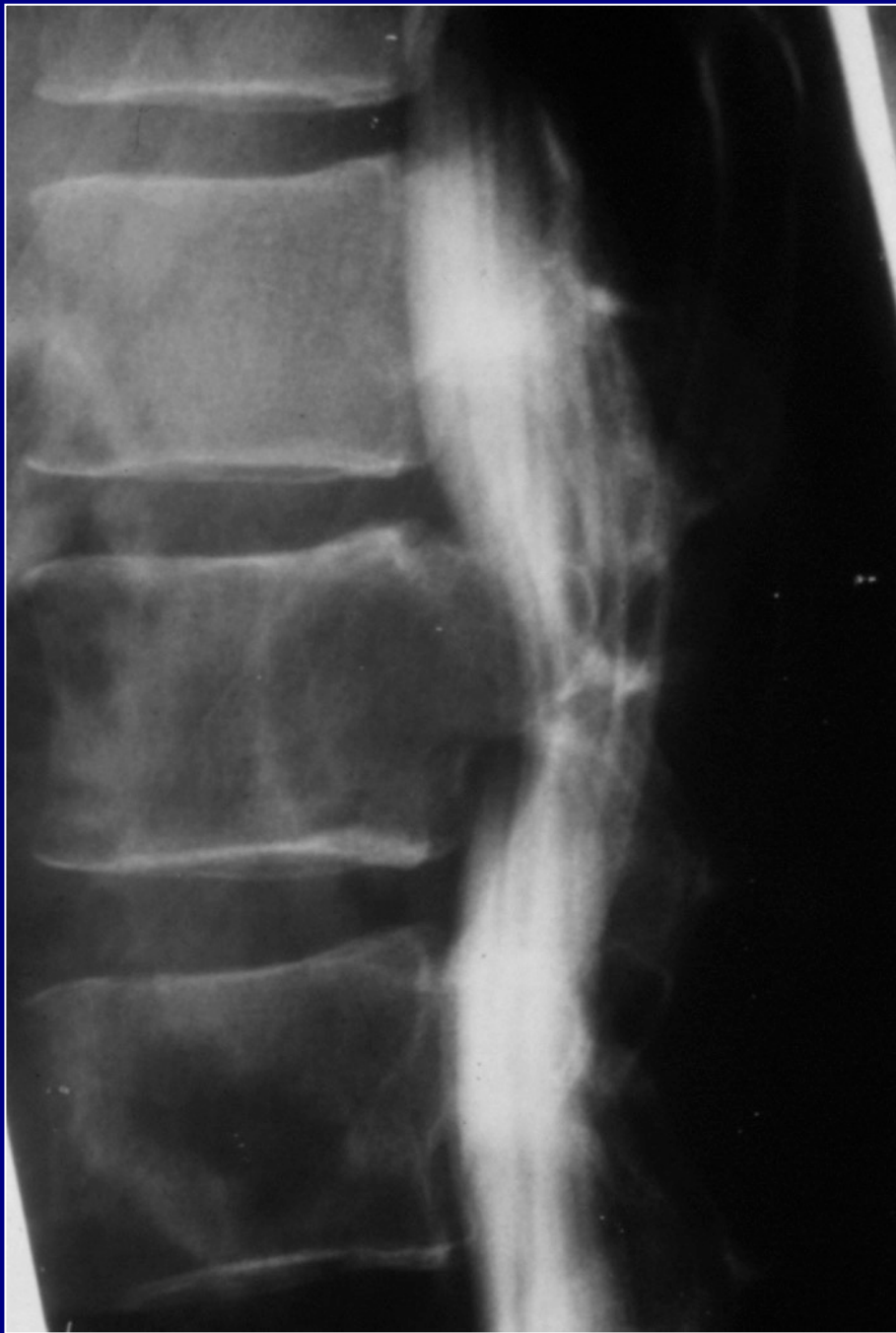


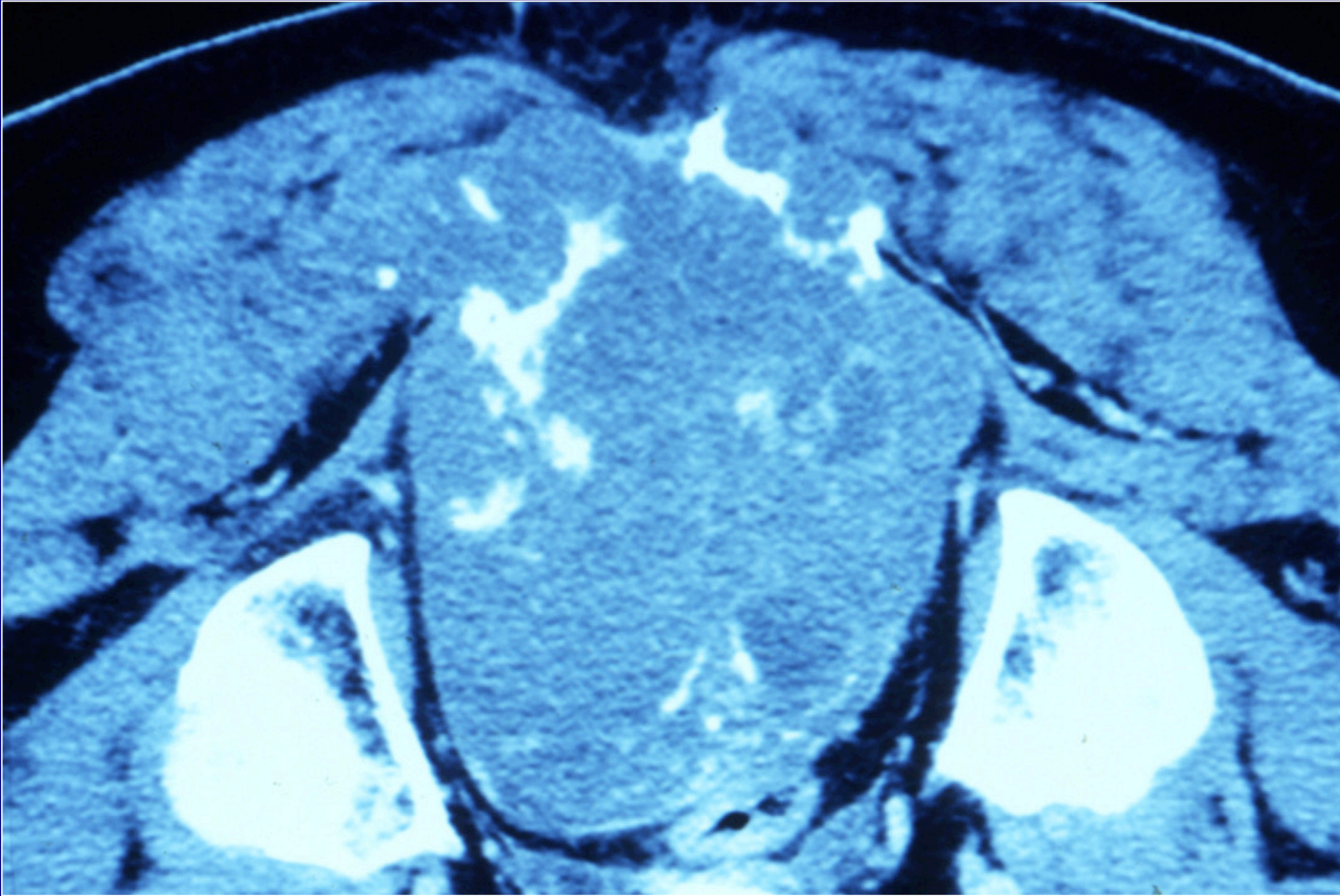
Average: 56 - Median: 58



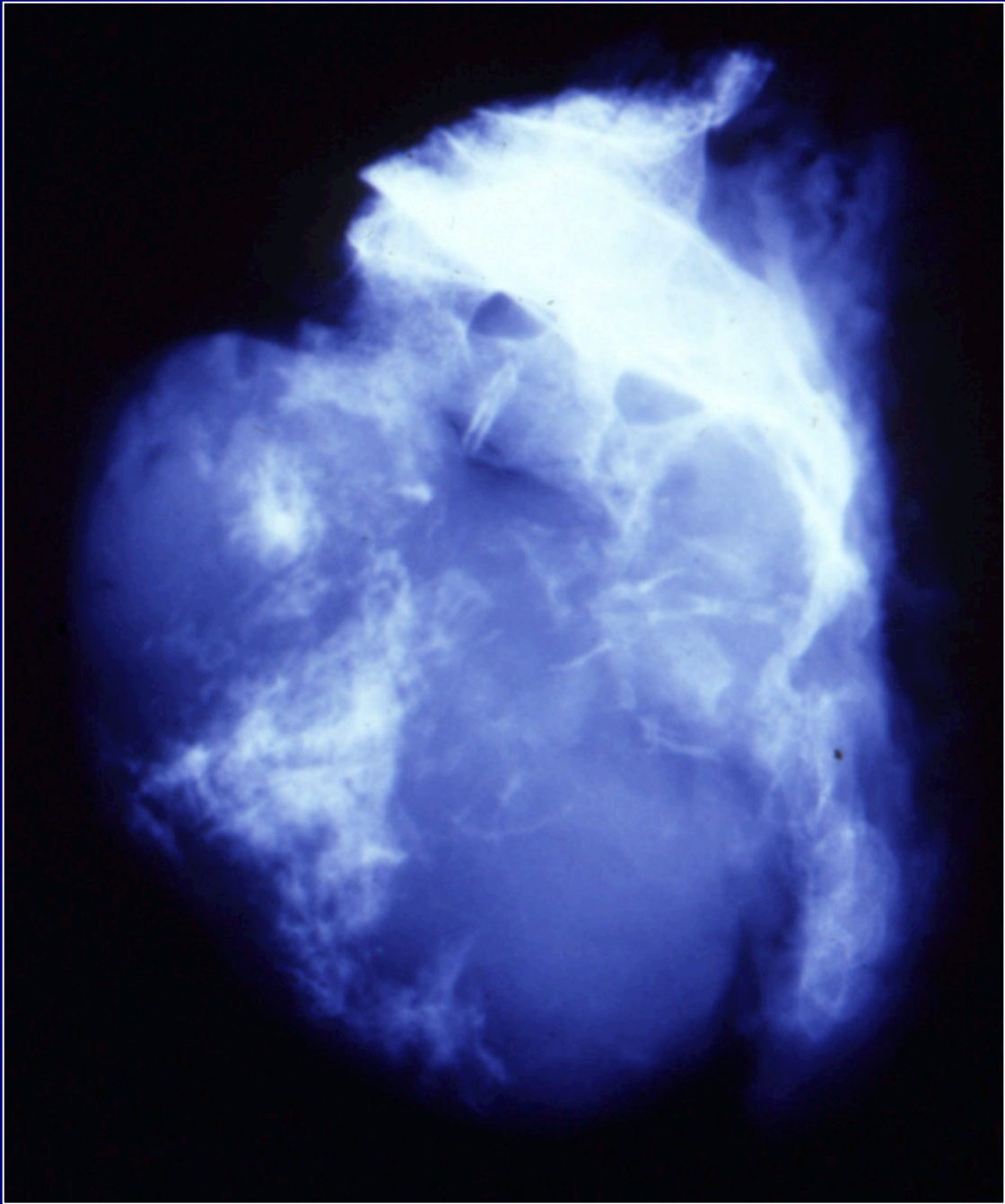


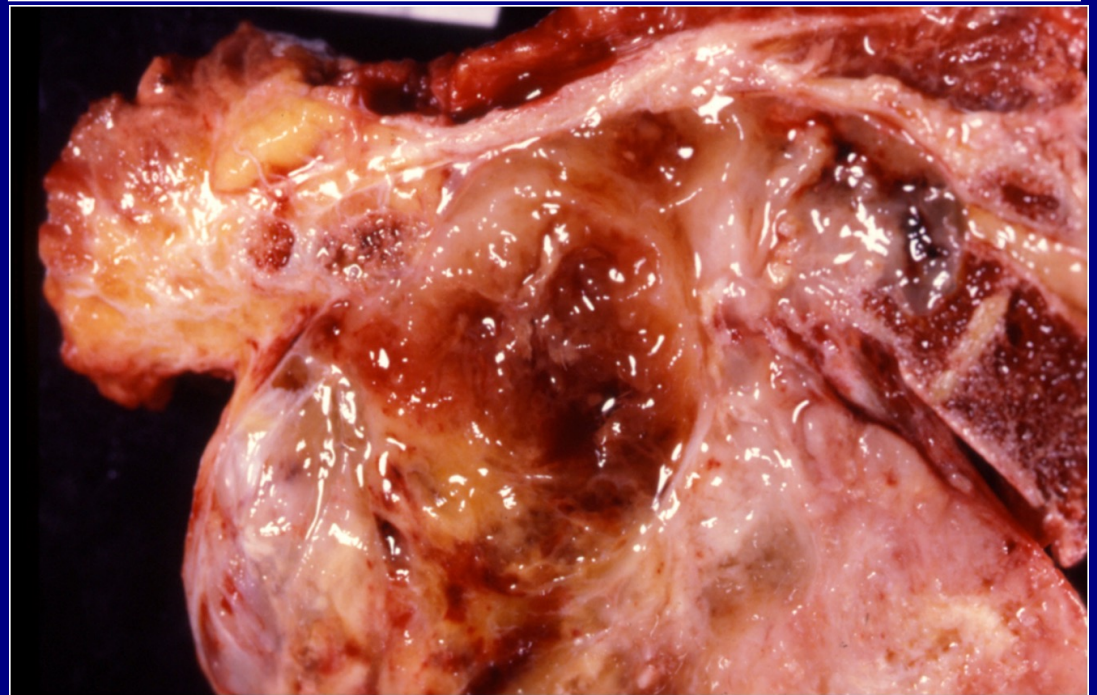
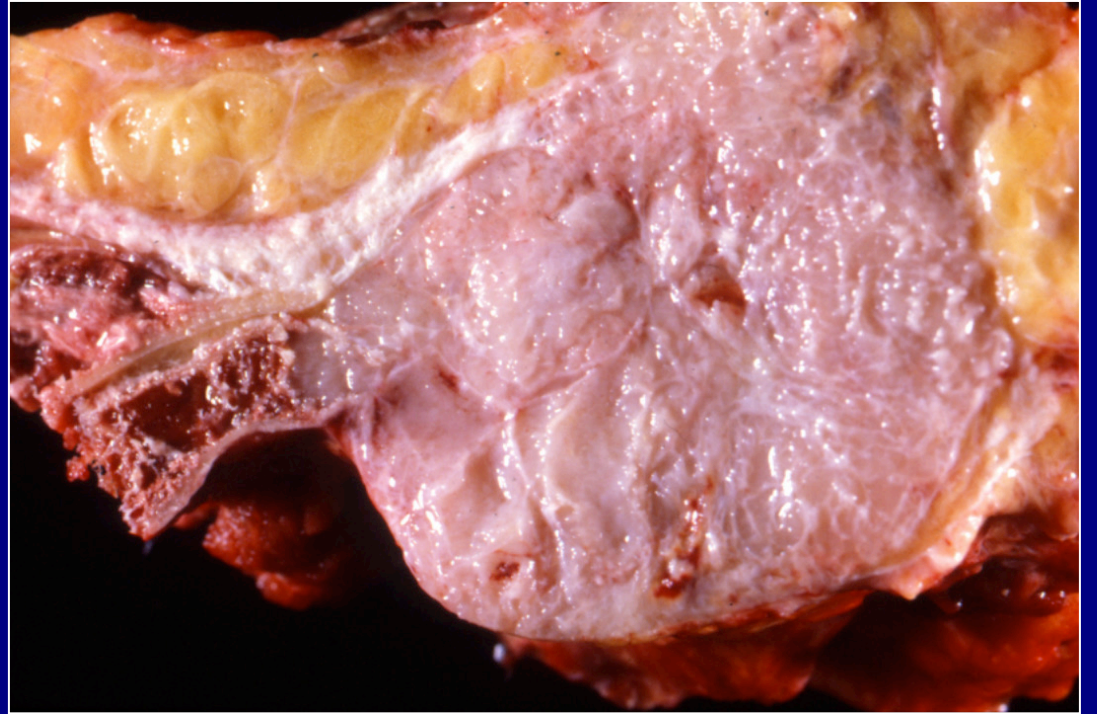
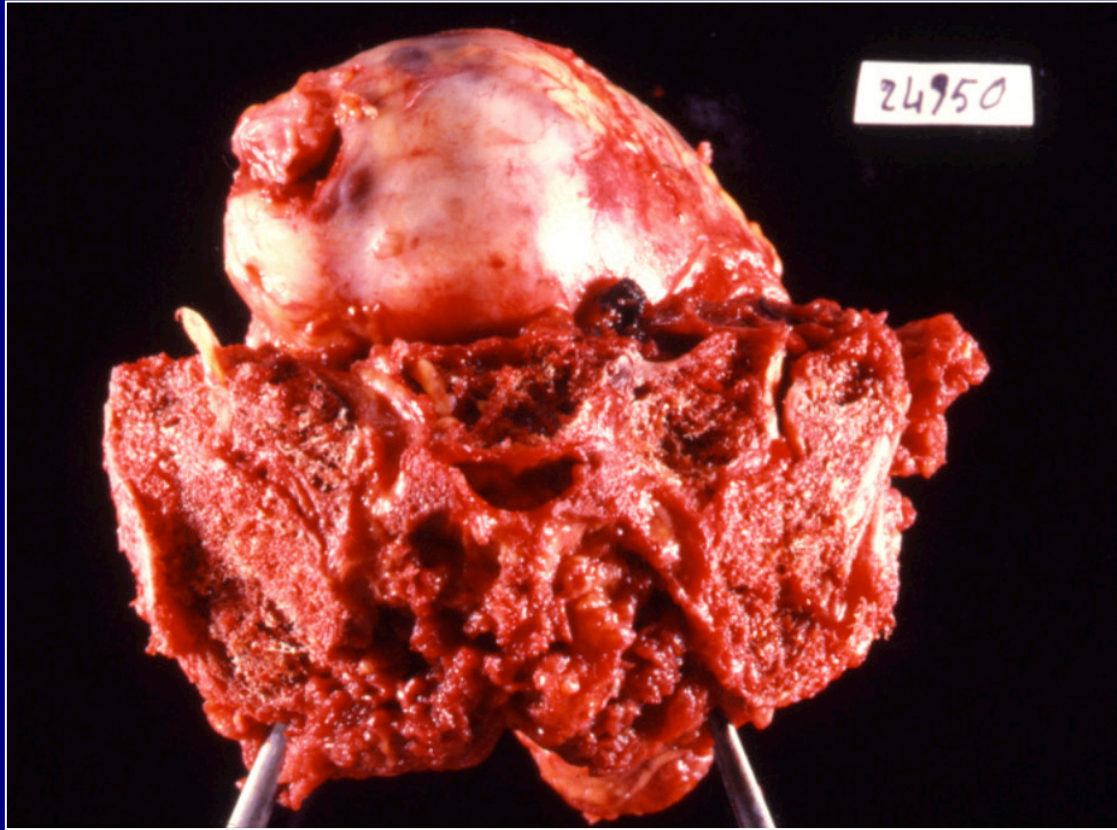






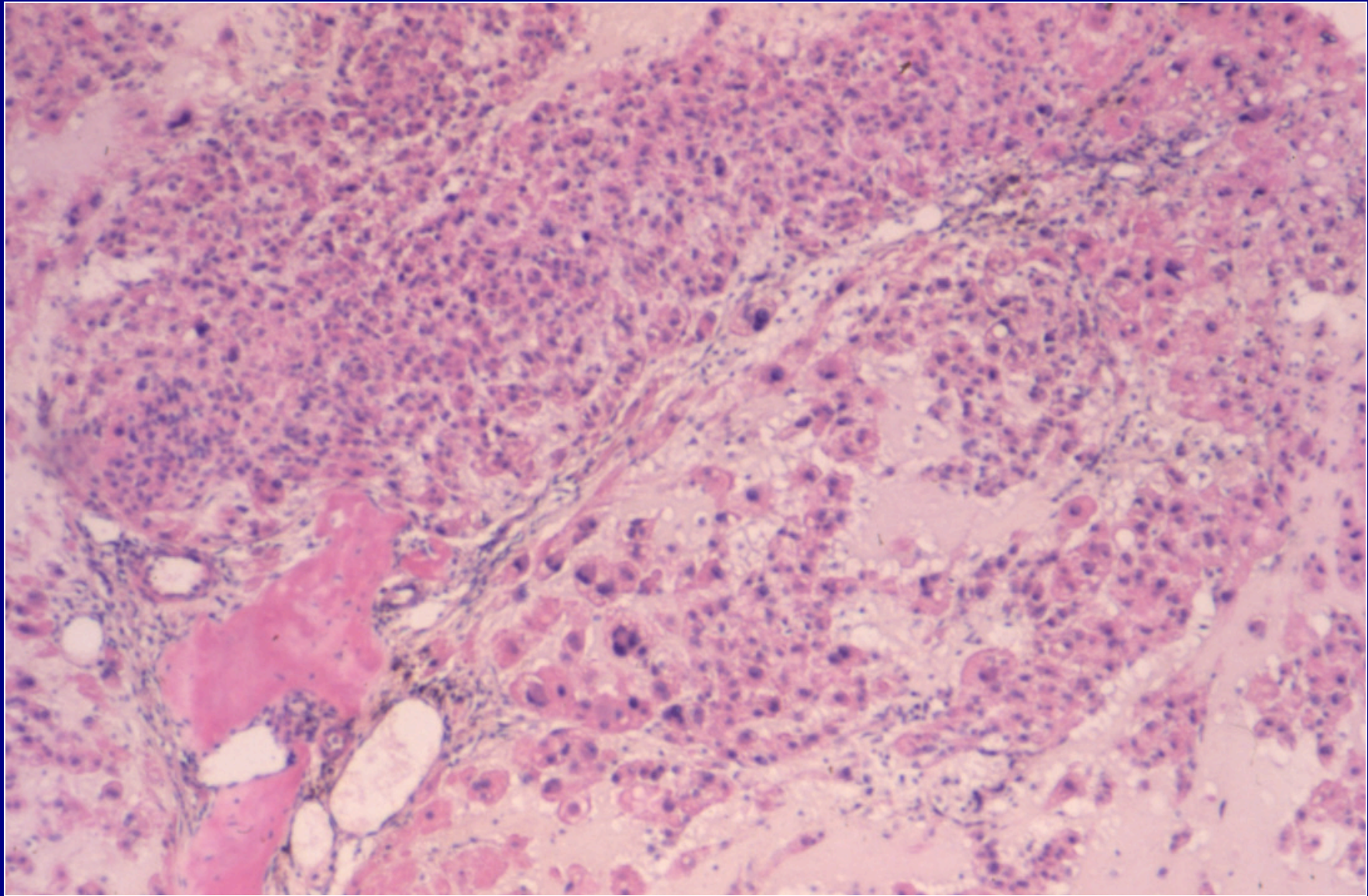




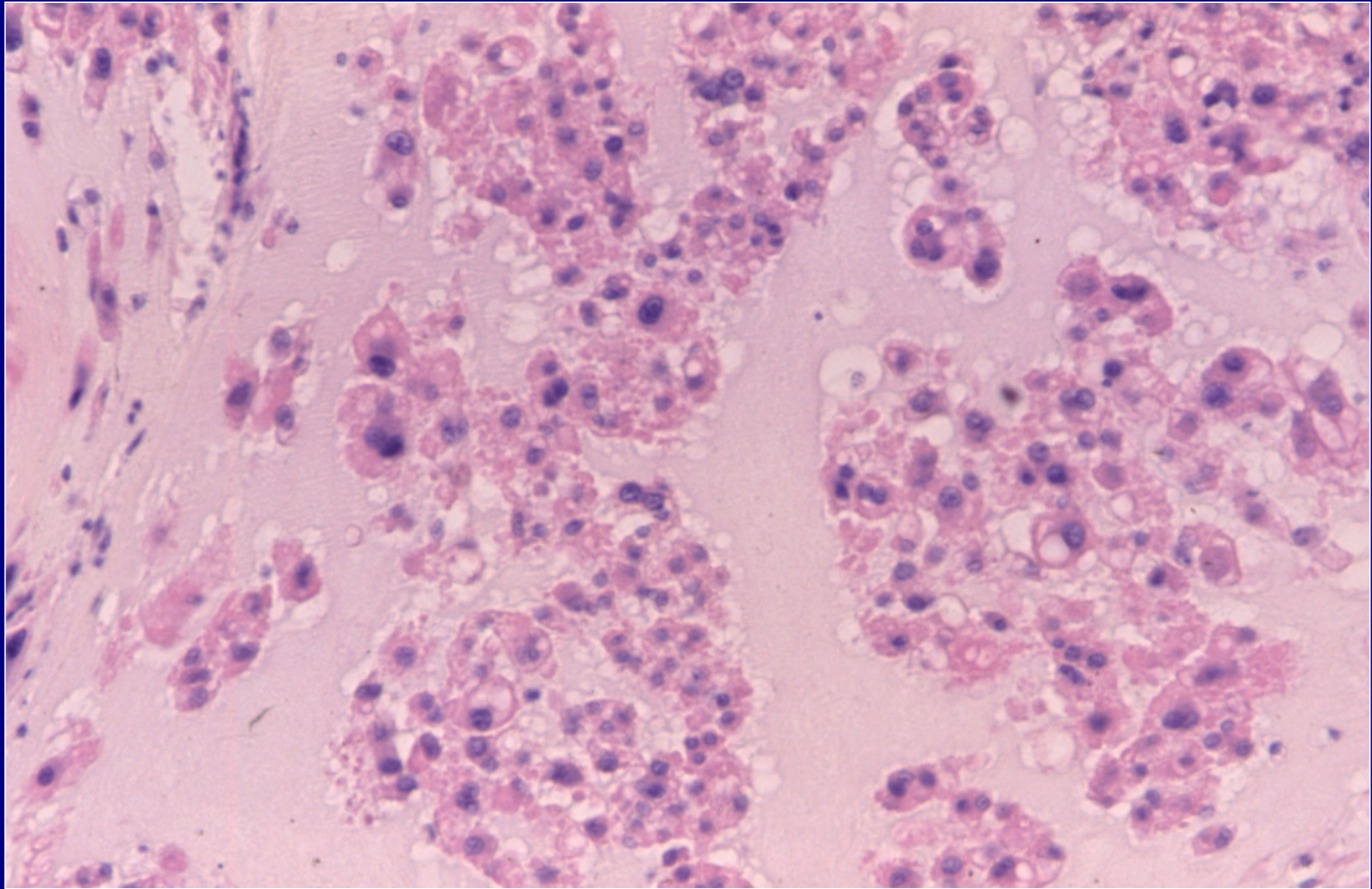


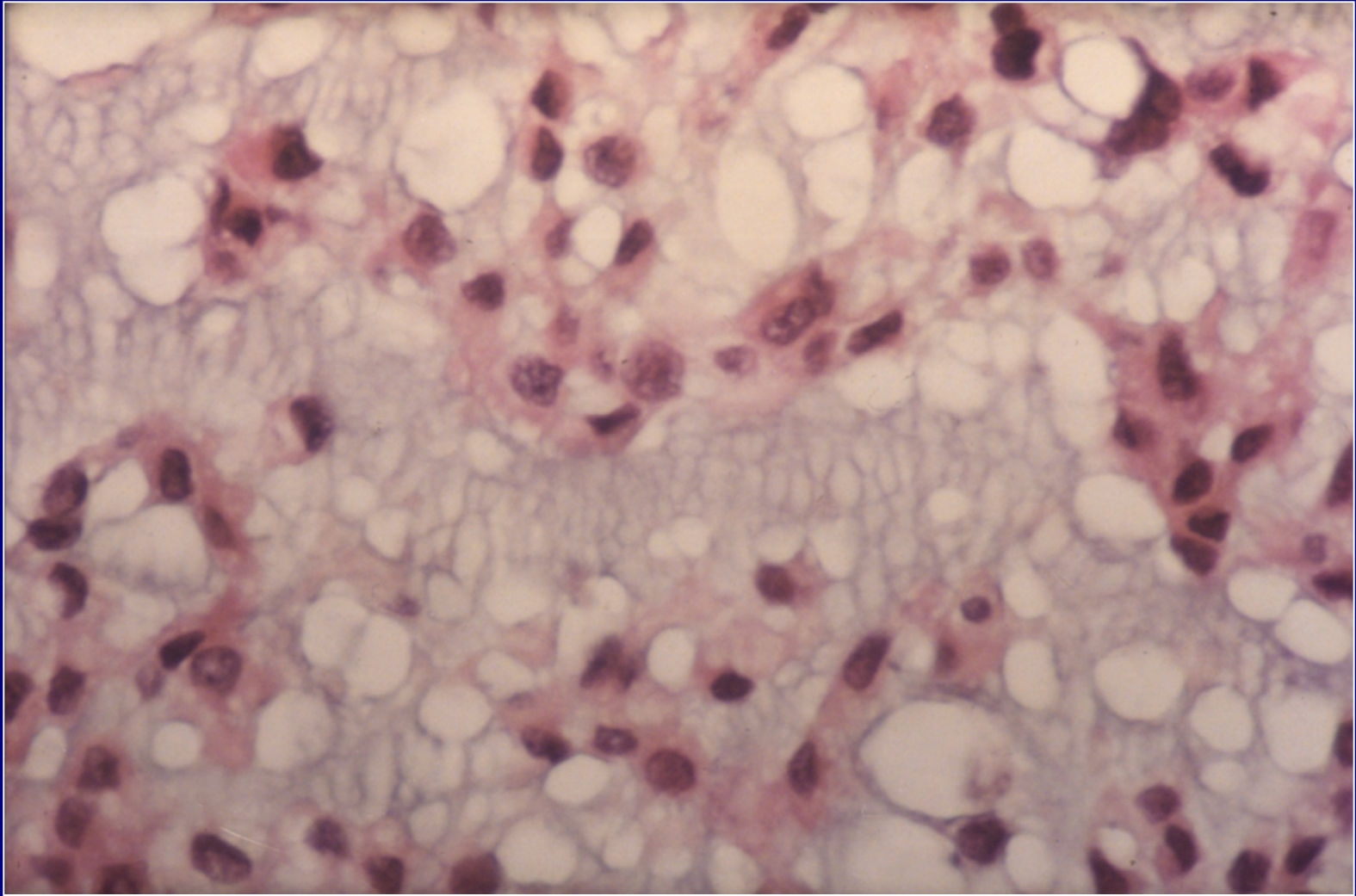
















## Sacral resections: experience of the I.O.R. bone tumor center

R. Capanna\*, A. Briccoli\*\*, R. Biagini\*, R. Casadei\*, N. Fabbri\*, N. Guernelli\*\*,  
M. Campanacci\*

\* Clinica Ortopedica dell'Università, Istituto Ortopedico Rizzoli, Bologna, Italy

\*\* Istituto di Patologia Chirurgia, Università, Modena, Italy

R. Capanna\*, A. Briccoli\*\*, R. Biagini\*, R. Casadei\*, N. Fabbri\*, N. Guernelli\*\*, M. Campanacci\*

Tab. I.  
Surgical complications and their outcome.

	Complications (21 cases)		Treatment
	Surgical Approach		
	Double	Post.	
Wound slough and/or infection	5	8	All healed (7 after surgical debridement)
Pulm. embolism	1	—	Died
Colitis with sepsis	3	—	1 died, 2 healed (1 after colostomy)
Intra-op. hemorrhage (hypogastric vein)	1	—	Clamped without consequence
Post-op. hemorrhage (gluteal vessels)	—	1	Clamped after reintervention
Rectus wall opening	—	2	1 spont-repaired 1 intra-op. sutured

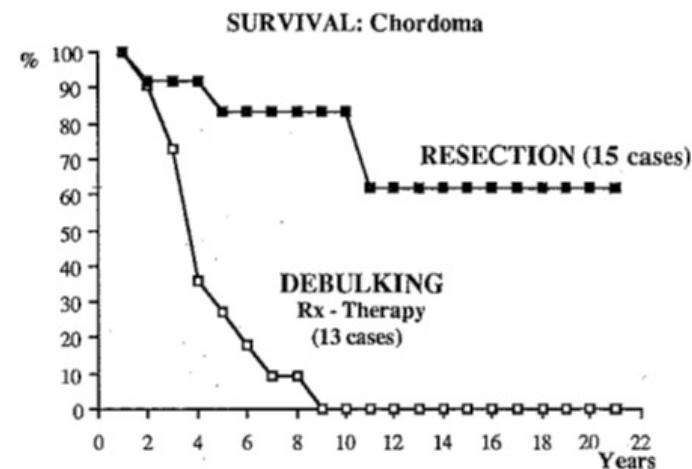
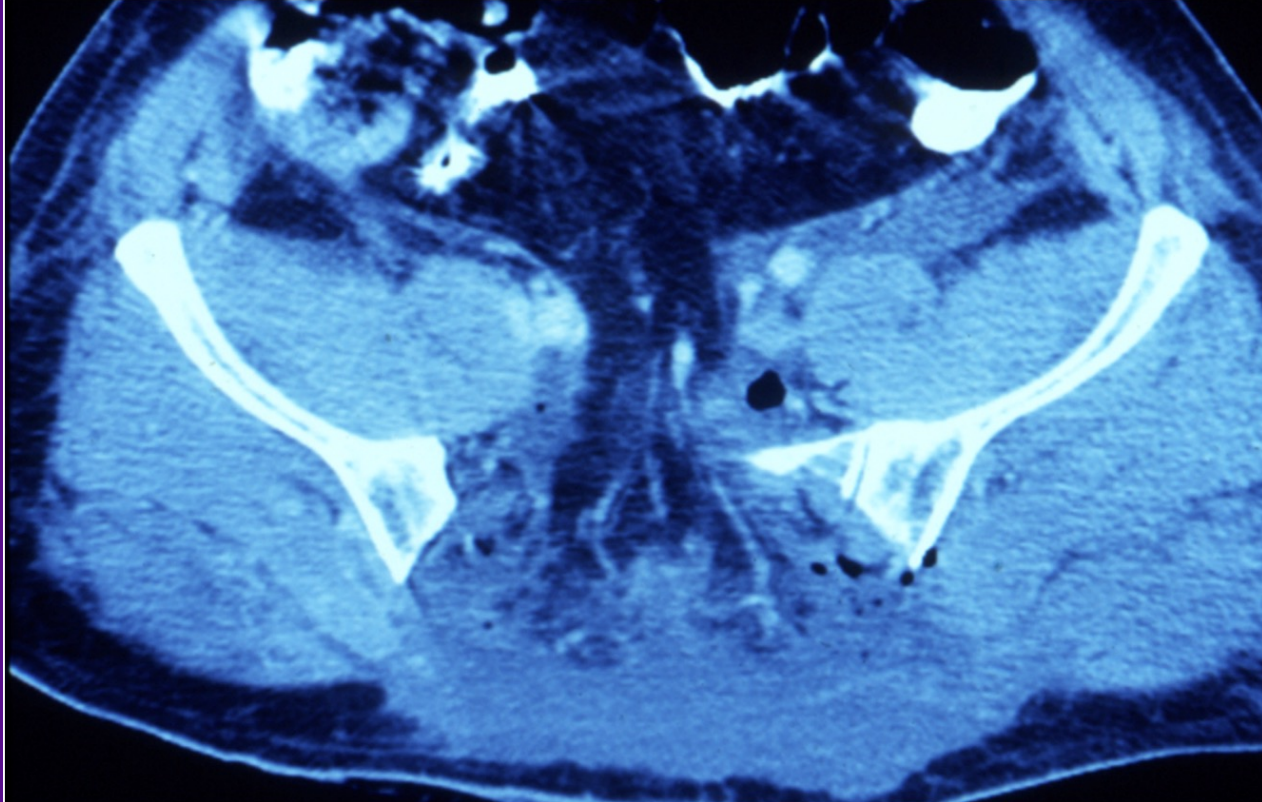
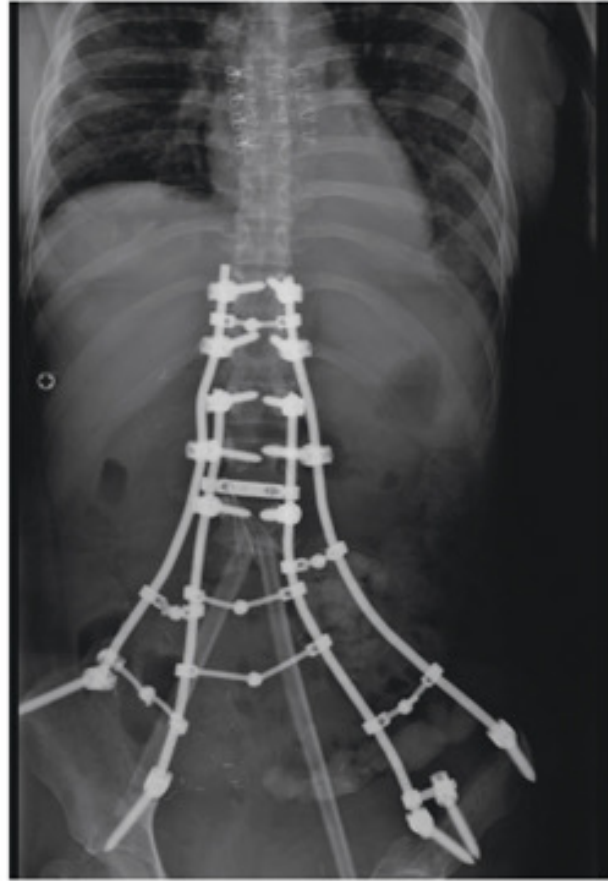
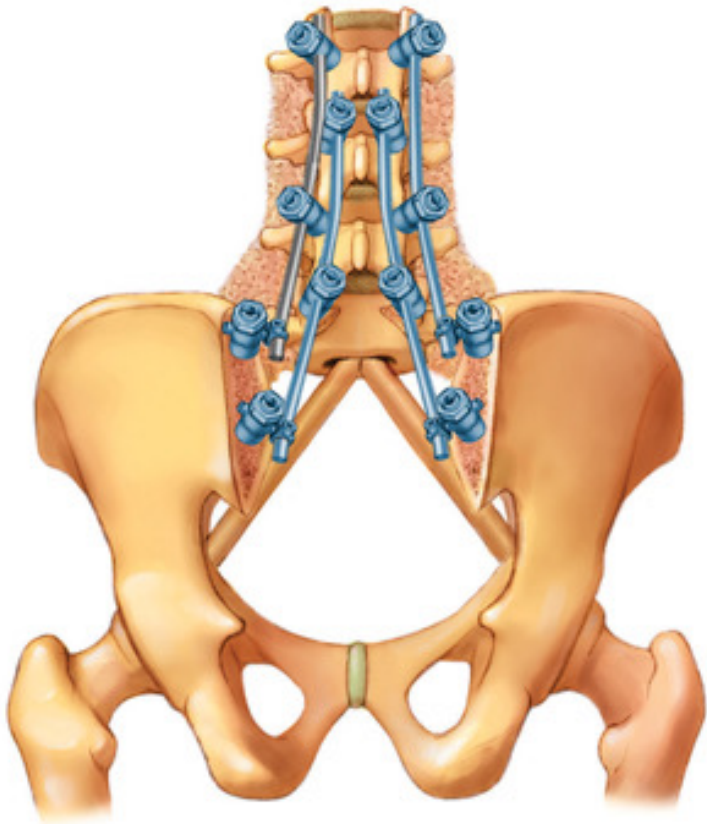


Fig. 1.  
Actuarial curve of survivors for patients affected by chordoma treated with oncologically adequate surgery (resection: 15 cases) versus curettage combined with radiotherapy (13 cases).









Contents lists available at [ScienceDirect](#)

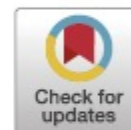
# European Journal of Surgical Oncology

journal homepage: [www.ejso.com](http://www.ejso.com)



## The sacral chordoma margin

S. Radaelli <sup>a,\*,1</sup>, P. Fossati <sup>b,1</sup>, S. Stacchiotti <sup>c</sup>, T. Akiyama <sup>d</sup>, J.M. Asencio <sup>e</sup>, S. Bandiera <sup>f</sup>, A. Boglione <sup>g</sup>, P. Boland <sup>h</sup>, S. Bolle <sup>i</sup>, Ø. Bruland <sup>j</sup>, A. Brunello <sup>k</sup>, P. Bruzzi <sup>l</sup>, D. Campanacci <sup>m</sup>, F. Cananzi <sup>n</sup>, R. Capanna <sup>o</sup>, R. Casadei <sup>p</sup>, A. Cordoba <sup>q</sup>, C. Court <sup>r</sup>, A.P. Dei Tos <sup>s,t</sup>, T.F. DeLaney <sup>u</sup>, A. De Paoli <sup>v</sup>, T.M. De Pas <sup>w</sup>, A. Desai <sup>x</sup>, L. Di Brina <sup>y</sup>, D.M. Donati <sup>p</sup>, N. Fabbri <sup>h</sup>, M.R. Fiore <sup>z</sup>, A. Frezza <sup>c</sup>, M. Gambarotti <sup>aa</sup>, A. Gasbarrini <sup>f</sup>, P. Georg <sup>b</sup>, G. Grignani <sup>ab</sup>, N. Hindi <sup>ac</sup>, E.B. Hug <sup>b</sup>, R. Jones <sup>ad</sup>, A. Kawai <sup>ae</sup>, A.D. Krol <sup>af</sup>, F. Le Grange <sup>ag</sup>, A. Luzzati <sup>ah</sup>, G. Marquina <sup>ai</sup>, J.A. Martin-Benlloch <sup>aj</sup>, K. Mazzocco <sup>ak</sup>, F. Navarria <sup>v</sup>, P. Navarria <sup>y</sup>, P.D. Parchi <sup>o</sup>, S. Patel <sup>al</sup>, E. Pennacchioli <sup>am</sup>, M.G. Petrongari <sup>an</sup>, P. Picci <sup>ao</sup>, R. Pollock <sup>ap</sup>, L. Porcu <sup>aq</sup>, V. Quagliuolo <sup>o</sup>, C. Sangalli <sup>ar</sup>, S. Scheipl <sup>as</sup>, G.M. Scotto <sup>ai</sup>, M. Spalek <sup>at</sup>, T. Steinmeier <sup>au</sup>, B. Timmermann <sup>av</sup>, A. Trama <sup>aw</sup>, M. Uhl <sup>ax</sup>, C. Valverde <sup>ay</sup>, P.P. Varga <sup>az</sup>, R. Verges <sup>ba</sup>, D.C. Weber <sup>bb</sup>, C. Zoccali <sup>bc</sup>, P.G. Casali <sup>c,bd</sup>, J. Sommer <sup>be</sup>, A. Gronchi <sup>a</sup>

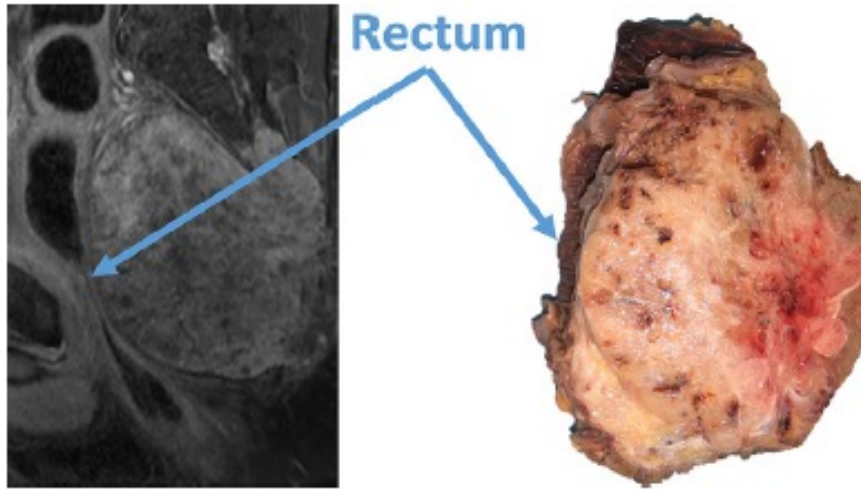




# Surgery

**Table 1**  
Series of sacral chordoma patients reporting oncologic outcome according to the adequacy of surgical margins. \*Data extrapolated from KM curves on available information.

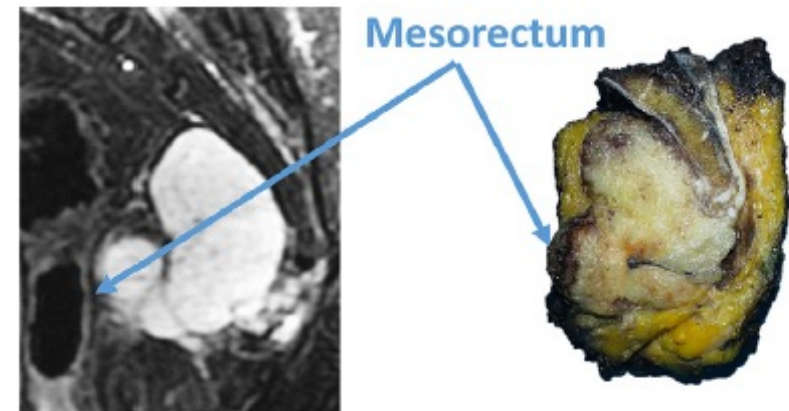
Series	Year	No. Pts	Median FU (years)	Margin status %	R0				R1			
					5-year OS	5-year LR	10-year OS	10-year LR	5-year OS	5-year LR	10-year OS	10-year LR
Bergh P.	2000	30	8.1	R0 = 70 R1 = 14 R2 = 16	90%*	10%*	95%*	76%*	50%*	50%*	100%*	100%*
Fuchs B.	2005	52	7.8	R0 = 21 R1/R2 = 31	100%*	5%	100%*	—	25%*	71%	15%*	—
Kayani B.	2015	58	3.8	R0 = 48 R1 = 42 R2 = 10	85%*	36%	38%*	—	50%*	79%	13%*	—
Angelini A.	2015	71	9.5	R0 = 77% R1 = 23%	—	28%*	—	40%*	—	55%*	—	55%*
Ji T.	2017	115	4.9	R0 = 67 R1/R2 = 33	86%	32%	—	—	67%	74%	—	—
Radaelli S.	2016	99	8.7	R0 = 47 R1 = 43 R2 = 10	95%	18%	71%	31%	95%	38%	62%	58%
Yang Y.	2017	157	4.6	R0 = 21 R1 = 39 R2 = 40	—	17%	—	—	—	43%	—	—
Colangeli S.	2018	33	4.4	R0 = 52 R1 = 42 R2 = 6	—	10%*	—	10%*	—	100%*	—	100%*



**Fig. 1.** Neoplastic invasion of the posterior rectal wall. The tumor is therefore resected en-bloc with the sacrum and the rectum.

1418

*S. Radaelli et al. / European Journal of S*



**Fig. 2.** The rectum is displaced anteriorly, without being infiltrated, by the sacral chordoma. The surgical dissection may be carried out leaving the whole mesorectum on the specimen with the posterior rectal wall exposed. Thus, the tumor is kept entirely covered achieving an appropriate anterior resection margin.



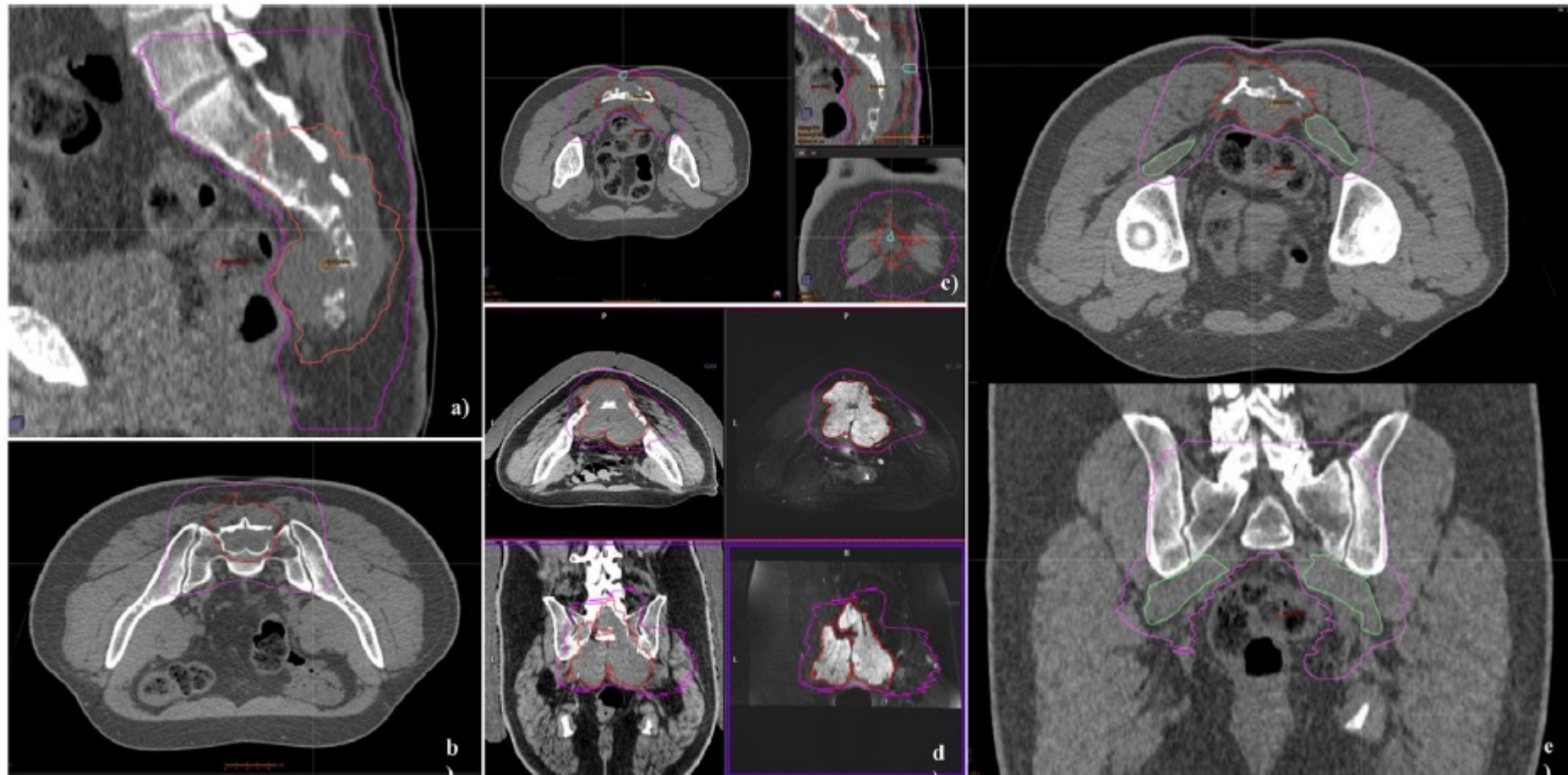
# Radiation Therapy

**Table 2**

Comparison of oncologic outcomes for sacral chordoma treated with definitive heavy-particles RT. C = carbon ion; P = proton; N = neutron; IMRT = intensity modulated radiation therapy.

Series	Year	No. Pts	Median FU (years)	Therapy	5-year OS	10-year OS	5-year LR	10-year LR
Breteau N.	1998	13	4	N	61% (4yr)	—	44% (4yr)	—
Nishida Y.	2011	7	4.1	C	53%	—	0%	—
McDonald MW.	2013	16	1.9	P	80% (2 yr)	—	15% (2 yr)	—
Mima M.	2014	23	3.2	C or P	83% (3yr)	—	6% (3 yr)	—
Uhl M.	2015	56	2.1	C ± IMRT	52%	—	21%	—
Imai R.	2016	188	5	C	81%	67%	19%	50%
Kabolizadeh P.	2017	40	4.2	P ± IMRT	82%	—	15%	—
Youn SH.	2018	58	3.5	P	88%	—	12%	—
Aibe N.	2018	23	3.1	P	10% (3yr)	—	7% (3yr)	—

# Radiation Therapy - GTV, CTV & PTV



**Fig. 3.** GTV and CTV contouring of sacral chordoma by CT/MRI scan **a)** one or two vertebral bodies rostral to the GTV and the whole sacrum caudally should be included **b)** sacroiliac joint contoured within the CTV **c)** biopsy tract or surgical scars should be included in the low-dose CTV **d)** the degree of lateral extension into the gluteal muscles is debatable albeit at least 1.5 cm margin of radiologically normal muscle should be included in the low dose CTV **e)** both piriform muscles should be entirely included in the CTV.







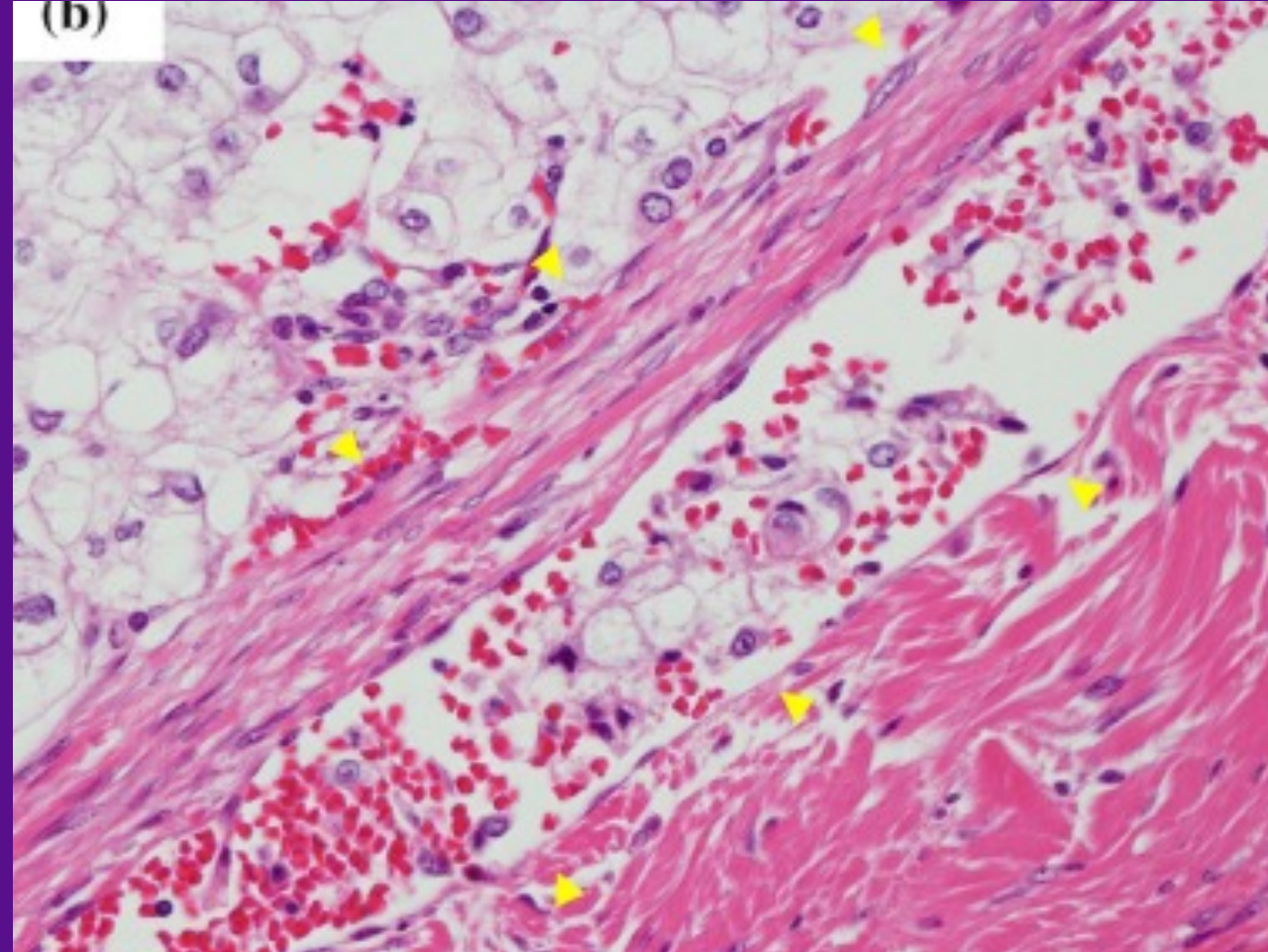
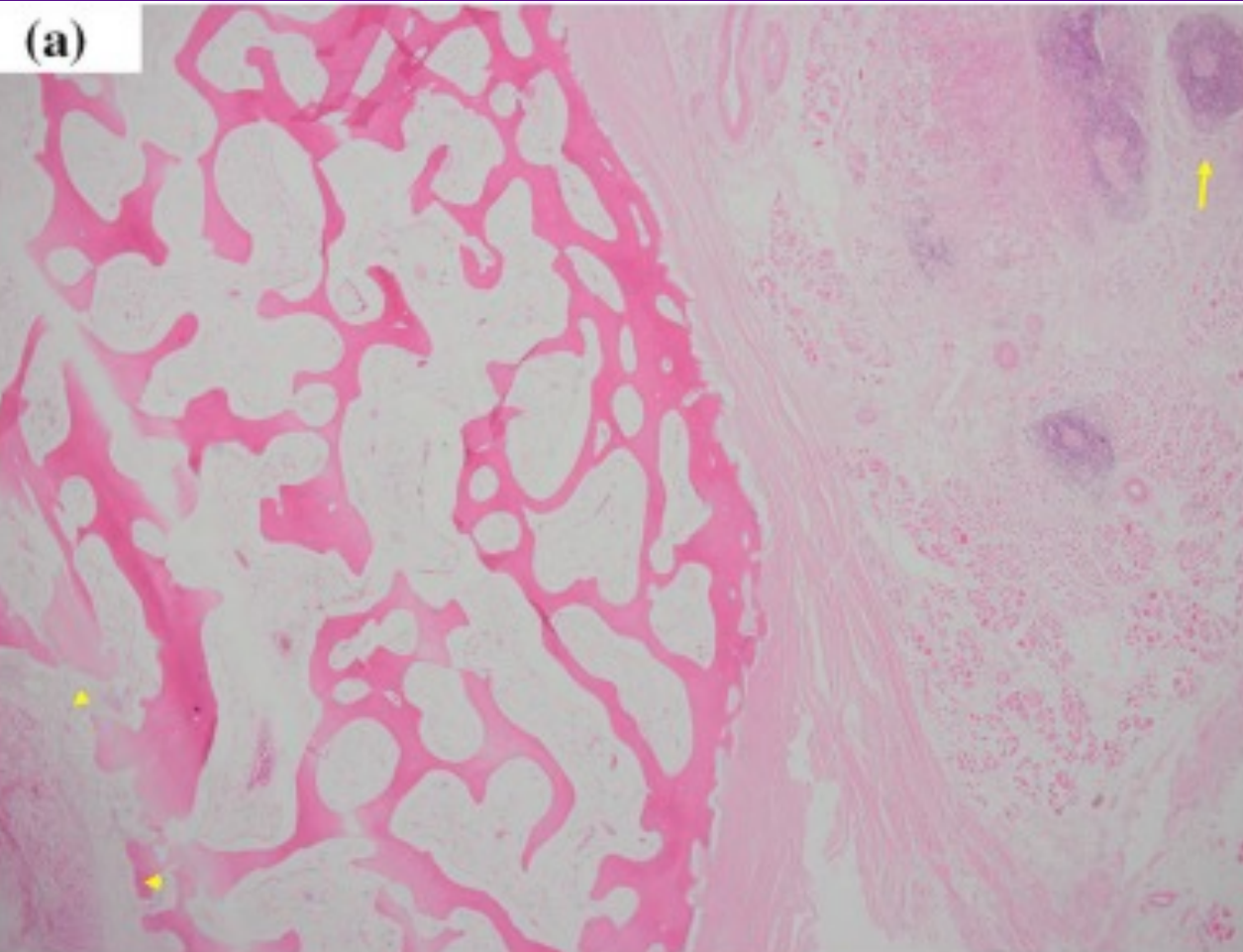
ORIGINAL ARTICLE – BONE AND SOFT TISSUE SARCOMAS

## **Analysis of the Infiltrative Features of Chordoma: The Relationship Between Micro-Skip Metastasis and Postoperative Outcomes**

**Toru Akiyama, MD<sup>1</sup>, Koichi Ogura, MD<sup>2,3</sup>, Tabu Gokita, MD, PhD<sup>4</sup>, Satoshi Tsukushi, MD, PhD<sup>5</sup>, Shintaro Iwata, MD, PhD<sup>6</sup>, Tomoki Nakamura, MD, PhD<sup>7</sup>, Akihiko Matsumine, MD, PhD<sup>7</sup>, Tsukasa Yonemoto, MD, PhD<sup>6</sup>, Yoshihiro Nishida, MD, PhD<sup>8</sup>, Kazuo Saita, MD, PhD<sup>1</sup>, Akira Kawai, MD, PhD<sup>2</sup>, Seiichi Matsumoto, MD, PhD<sup>4</sup>, and Takehiko Yamaguchi, MD, PhD<sup>9</sup>**

<sup>1</sup>Department of Orthopaedic Surgery, Saitama Medical Center, Jichi Medical University, Saitama, Japan; <sup>2</sup>Department of Musculoskeletal Oncology, National Cancer Center Hospital, Tokyo, Japan; <sup>3</sup>Department of Orthopaedic Surgery, The University of Tokyo Hospital, Tokyo, Japan; <sup>4</sup>Department of Orthopedic Surgery, Cancer Institute Hospital, Tokyo, Japan; <sup>5</sup>Department of Orthopedic Surgery, Aichi Cancer Center Hospital, Aichi, Japan; <sup>6</sup>Division of Orthopaedic Surgery, Chiba Cancer Center, Chiba, Japan; <sup>7</sup>Department of Orthopaedic Surgery, Mie University Hospital, Mie, Japan; <sup>8</sup>Department of Orthopaedic Surgery, Nagoya University Graduate School and School of Medicine, Aichi, Japan; <sup>9</sup>Department of Pathology, Saitama Medical Center, Dokkyo Medical University, Koshigaya, Japan





## Long-term outcomes of high-dose single-fraction radiosurgery for chordomas of the spine and sacrum


\*Chunzi Jenny Jin, MD, MS,<sup>1</sup> John Berry-Candelario, MD, MPH,<sup>2</sup> Anne S. Reiner, MPH,<sup>3</sup> Ilya Laufer, MD, MS,<sup>2</sup> Daniel S. Higginson, MD,<sup>1</sup> Adam M. Schmitt, MD,<sup>1</sup> Eric Lis, MD,<sup>4</sup> Ori Barzilai, MD,<sup>2</sup> Patrick Boland, MD,<sup>5</sup> Yoshiya Yamada, MD,<sup>1</sup> and Mark H. Bilsky, MD<sup>2</sup>

Departments of <sup>1</sup>Radiation Oncology, <sup>2</sup>Neurosurgery, <sup>3</sup>Biostatistics, and <sup>4</sup>Radiology, and <sup>5</sup>Division of Orthopedic Surgery, Memorial Sloan Kettering Cancer Center, New York, New York



**BRIEF REPORT**

## Poorly differentiated chordoma with whole-genome doubling evolving from a *SMARCB1*-deficient conventional chordoma: A case report

Christian Curcio<sup>1</sup>  | Robert Cimera<sup>2</sup> | Ruth Aryeequaye<sup>2</sup> | Mamta Rao<sup>2</sup> | Nicola Fabbri<sup>3</sup> | Yanming Zhang<sup>2</sup> | Meera Hameed<sup>2</sup>

<sup>1</sup>Department of Pathology and Laboratory Medicine, Hospital for Special Surgery, New York, New York

<sup>2</sup>Department of Pathology, Memorial Sloan Kettering Cancer Center, New York, New York

<sup>3</sup>Department of Surgery, Memorial Sloan Kettering Cancer Center, New York, New York

### Abstract

Evolution of poorly differentiated chordoma from conventional chordoma has not been previously reported. We encountered a case of a poorly differentiated chordoma with evidence of whole-genome doubling arising from a *SMARCB1*-deficient conventional chordoma. The tumor presented as a destructive sacral mass in a 43-year-old

- NCCN 2021-22 meeting: considerations for different guidelines than conventional chordoma



**NYU Langone**  
Orthopedics

**Thank You**