Sacral Chordoma: The Loma Linda University Radiation Medicine Experience

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What is a chordoma?

- 1st chordoma discovered in clivus by Virchow and Luschka 1856
- Rare tumor arising from notochordal remnants or ectopic notochordal tissue

Rudolph Virchow – Father of Chordomas

Metastasis

- Chordomas are not benign, can have metastatic potential
- Lungs, liver, bone most common
- Other studies report a distant metastasis rate of 5-40% in patients with long term follow up
- Survival time after metastasis was only 2.4 months

Epidemiology

- Incidence 0.08 per 100,000 people
- Rare reports of familial chordoma
- Few extensive case series
- In past, grouped with other bone neoplasms rather than studied separately

- Largest population based study for chordomas by McMaster, et al. examining SEER data
- 400 cases from 1973-1995

- 32% cranial, 33% spine, 29% sacrum, 8% other
- Male: Female 1.8:1
- Males more likely to have sacral presentation, females with cranial presentation
- Median age at diagnosis 58.5 years (range 3-95)

- Site of presentation varies with age and gender
- Youngest more likely to have cranial presentation
- Oldest patients, sacral tumors more common
- Median age of sacral presentation is 69

- Sacral lesions (32%) were more likely than cranial lesions(11%) to be treated with radiotherapy alone
- 5 year survival for sacral presentation 73.6%
- 10 year survival 32.2%

Clinical Presentation

- Symptoms can be vague
- May grow to large size before seeking workup
- Local pain in almost all
- Neurological deficits including bladder, bowel symptoms, radiculopathies

Imaging Findings

- Often 1st detected by CT scanning
- CT: partially calcified soft tissue mass eroding adjacent bone

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Treatment Options

- Surgery
- Radiation
- Surgery + Radiation
- Chemotherapy

Surgery

- Surgery is treatment of choice when possible
- Complete excision may lead to neurogenic dysfunction, pelvic instability
- Resection approach may be posterior or combined anteroposterior

- Want margins around tumor, optimally 1-2 cm margin of normal bone
- In large lesions, complete surgical excision usually impossible.

Mayo Clinic

- Mayo Clinic series of sacrococcygeal chordomas between 1980 to 2001
- 52 new presentation patients in the study
- Of these, only 40% were able to resected with wide margin

Mayo Clinic Results

- 5 yr overall survival 74%
- 44% had local recurrence
- Most important predictor of survival was wide margin

- Other studies report local control rate of 60-80% with total excision
- 25-50% with subtotal resection

York MD Anderson retrospective series

- Retrospective study of 27 patients with sacral chordomas at MD Anderson from 1954-1994
- With radical resection disease free interval was 2.2 years vs only 8 months with subtotal resection

- Some patients received post-operative radiation after subtotal and gross total resections
- An improvement in time to recurrence was seen among those receiving post operative XRT

TABLE 2. Surgical Results for Patients with Sacral Chordomas

Treatment	Recurrence	Median Time to Recurrence (yr)
All procedures[a, b]		
Subtotal excision	30/33 (91%)	0.67
Subtotal excision + XRT[c]	2/6 (33%)	2.12
Radical resection	10/21 (48%)	2.27
Radical resection + XRT	5/7 (71%)	4.00
Total	47/67 (70%)	
First procedure[a]		
Subtotal excision	10/12 (83%)	0.67
Radical resection	8/15 (53%)	3.37

[a] Differences between radical resection and subtotal excision were statistically significant for data for all procedures and the first procedure ($P = 0.001$ and $P = 0.0001$, respectively).

[b] Difference between subtotal excision with and without radiation therapy was statistically significant for data for all procedures ($P < 0.02$).

[c] XRT, radiation therapy administered in conjunction with surgery.

Radiation Options

- Photons
- Charged Particles: Protons, helium, carbon ion

Catton et al. study Princess Margaret (photon)

- Retrospective study of 48 patients with chordoma, including 23 sacral chordoma patients
- 13 sacral patients received XRT, most receiving 50 Gy in 25 fx
- Median survival after diagnosis was 62 months
- No survival advantages for patients receiving >50 Gy vs <50 Gy

Schoenthaler et al. study Lawrence Berkeley Laboratory

- 14 patients with sacral chordoma treated with charged particles helium and neon between 1977-1989
- Median dose 75.7 CGE given at 2-2.25 GyE daily
- Median follow up 5 years
- All patients received surgery (2 bx, 4 GTR, 8 STR)

- All received helium beam, 5 also received photons, 4 also received neon
- Target: entire sacrum and any evident soft tissue abnormality with 3cm margin to 50 GyE
- Cone down of gross tumor with 1-2 cm margin received 75.7 GyE

- Anterior rectum<50 GyE

Results of Schoenthaler Study

- 5 year survival 85%
- 5 year local control 55%
- 7 recurrences, with 2 of these patients recurring outside of radiation field

- Of the 4 patients who had total resection, 3 remain free of disease
- Of the 10 patients who had partial excision or biopsy, only 3 remain free of disease

Hug study(protons)

- 1980-1992 47 patients at Massachusetts General Hospital and Harvard Cyclotron Laboratory treated for chordomas, chondrosacromas, osteogenic sarcomas, giant cell tumors, osteo or chondroblastomas
- Included 14 spinal and sacral chordoma patients (8 sacral)
- Of spinal/sacral chordoma patients, mean dose given was 74.6 CGE

- 10 patients received pre and postop XRT to mean dose of 75.2 CGE
- Mean follow up time 3.2 years

- 5/14 patients (36%) failed locally
- 4 within the radiation field, 1 outside
- 1/4 patients (25%) with gross total resection failed locally
- 3/8 patients (38%) with subtotal resection failed locally

- Trend toward local control for target doses>77 CGE but not statistically sig
- 5 year local recurrence free survival rate 53%
- 5 year overall survival rate 50%

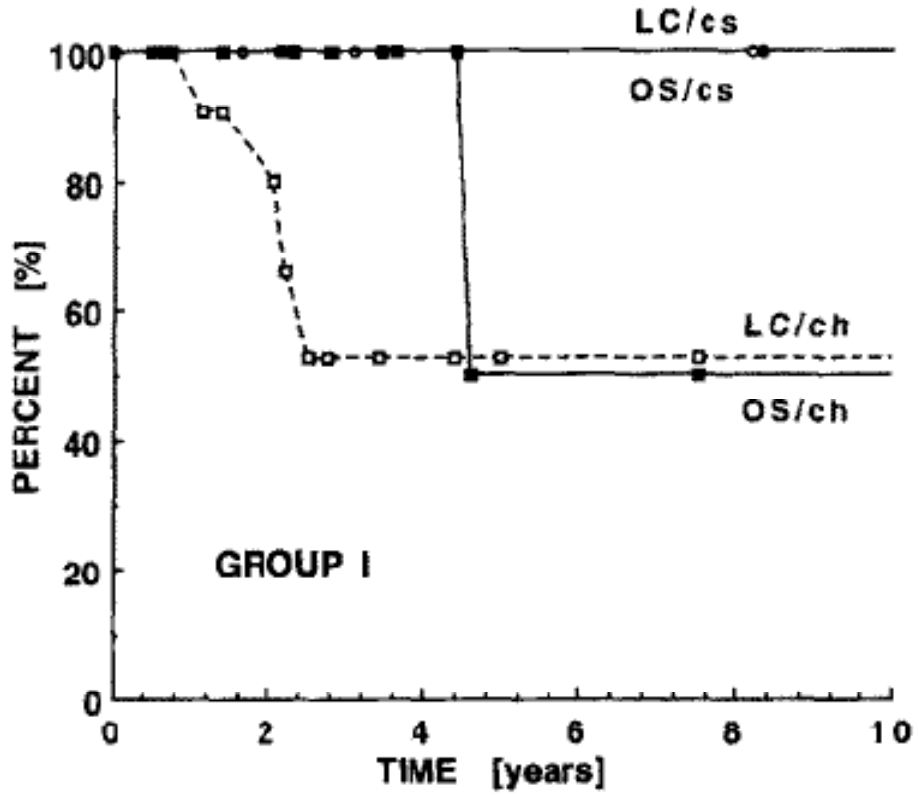


Fig. 3. Actuarial local control (LC) and overall survival (OS) of 14 patients with chordoma (ch) and 6 patients with chondrosarcoma (cs) of the thoracic and lumbar spine and sacrococcygeal region following combined photon and proton radiation therapy (group 1).

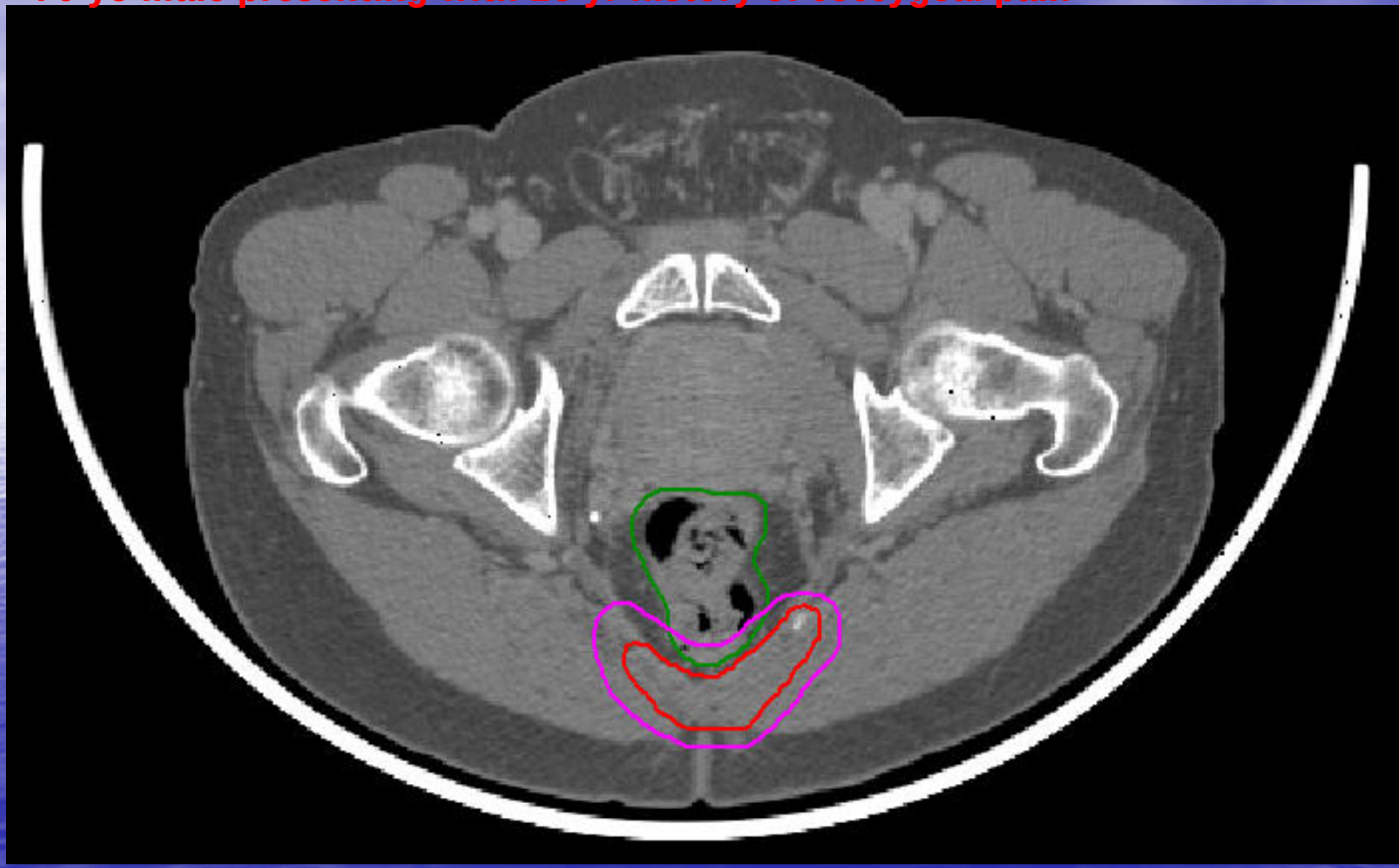
Study Design

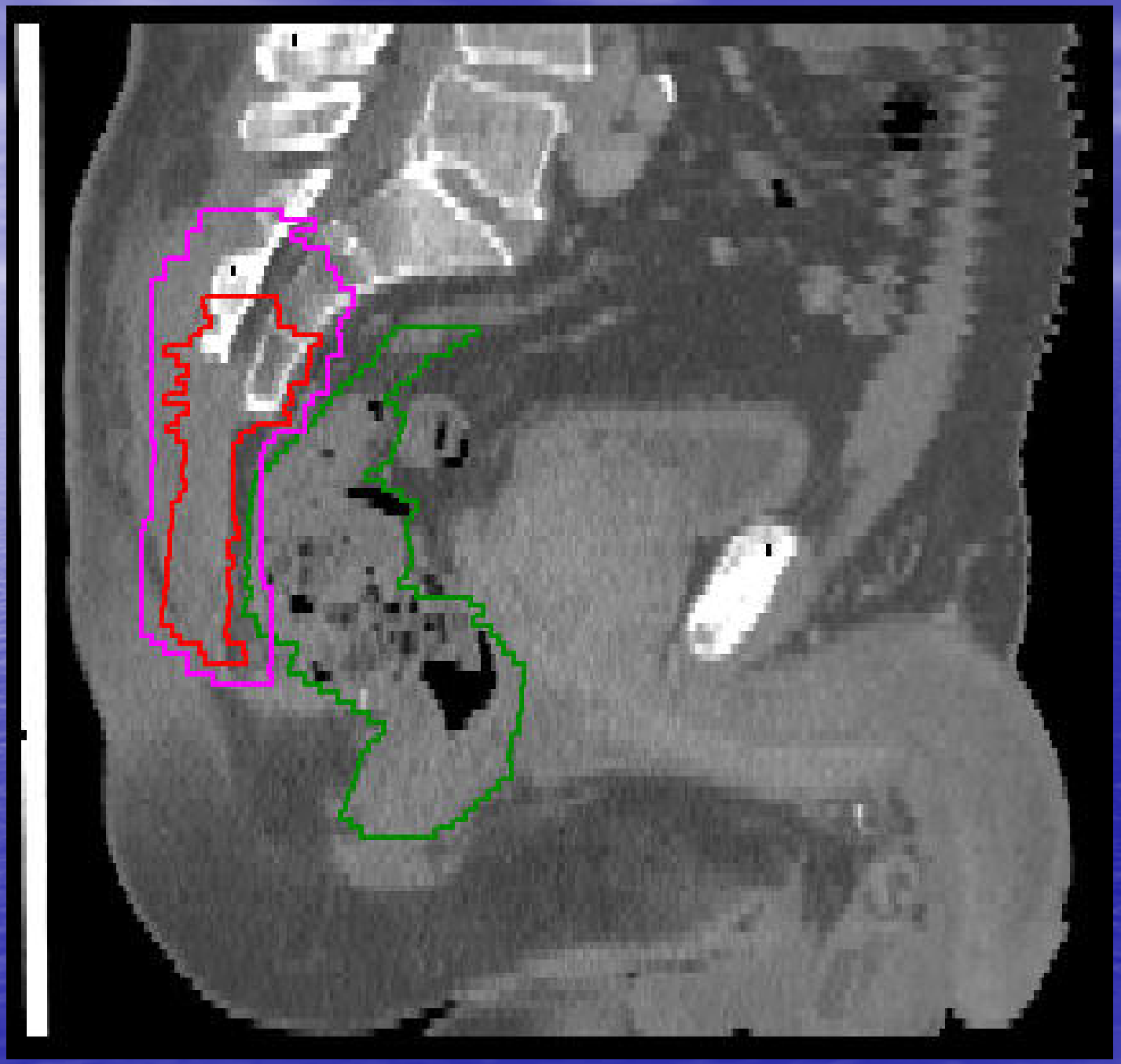
- Retrospective review of 22 patients treated at LLUMC for sacral chordomas between 1992-2004
- 3 patients excluded from analysis due to metasatic disease at presentation
- 6 records unavailable for followup

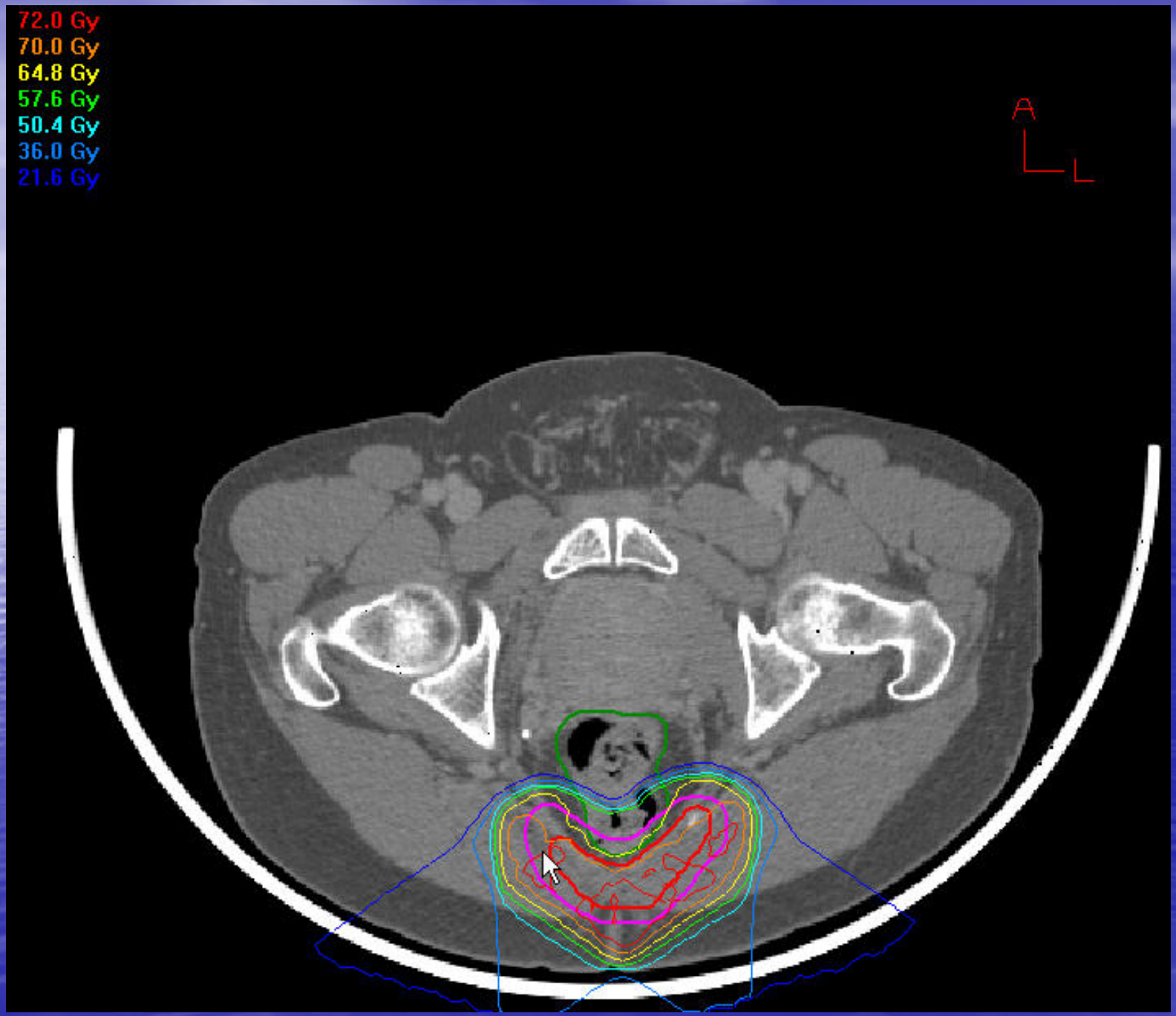
- Endpoints to be examined included overall survival, disease free survival, local control
- All patients had a surgical procedure: 1 biopsy only, 7 subtotal resection, 11 gross total resection

- Radiation dose ranged from 66.6 CGE to 81 CGE
- Average dose 73.6 CGE
- 8 patients had combination therapy of proton and photon beams
- GTV: preoperative tumor volume
- CTV: Add 0.5 cm around GTV
- Limit rectal dose to 50.4 CGE

70 yo male presenting with 15 yr history of coccygeal pain



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64.8 Gy
57.6 Gy
50.4 Gy
36.0 Gy
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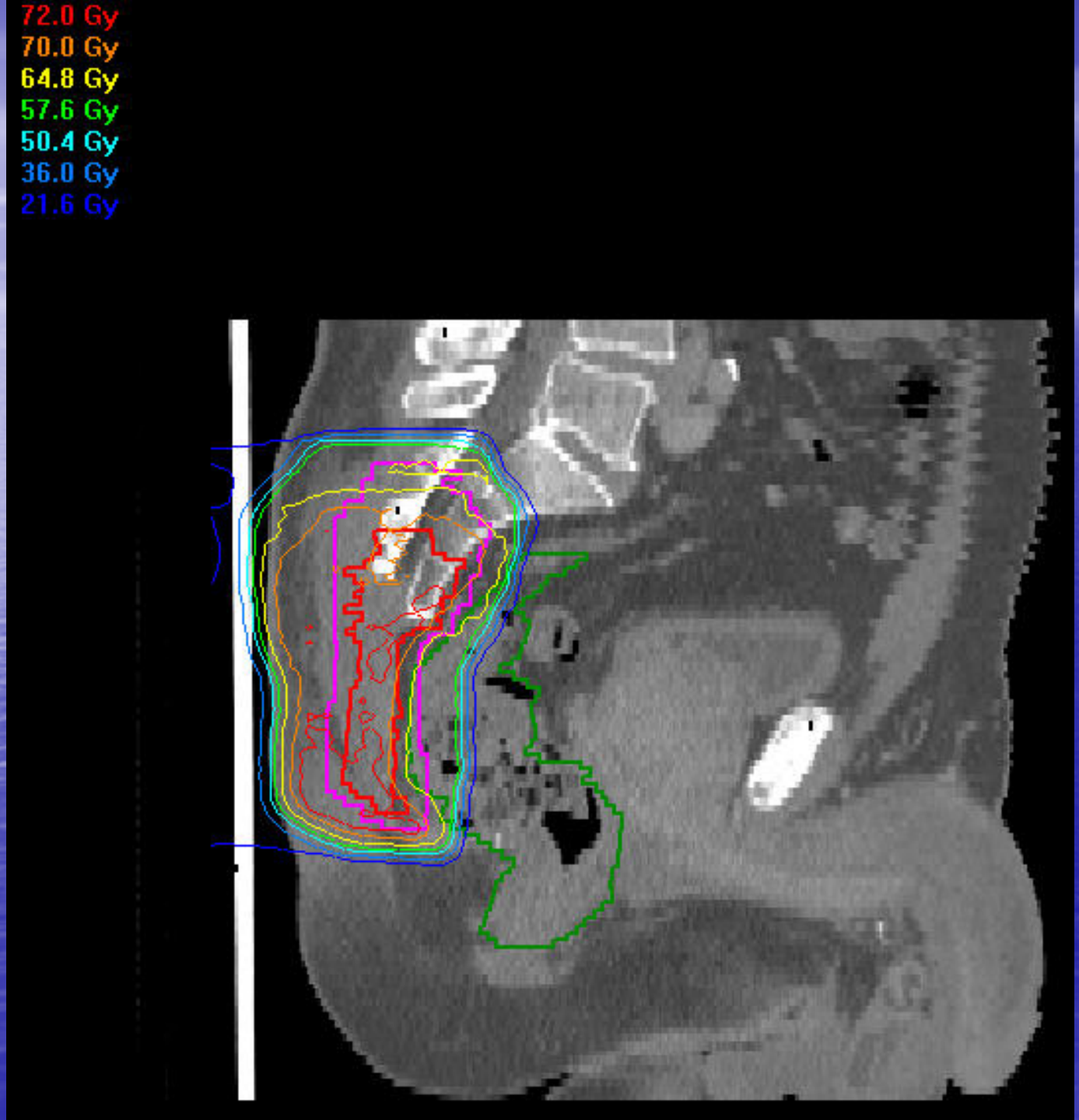
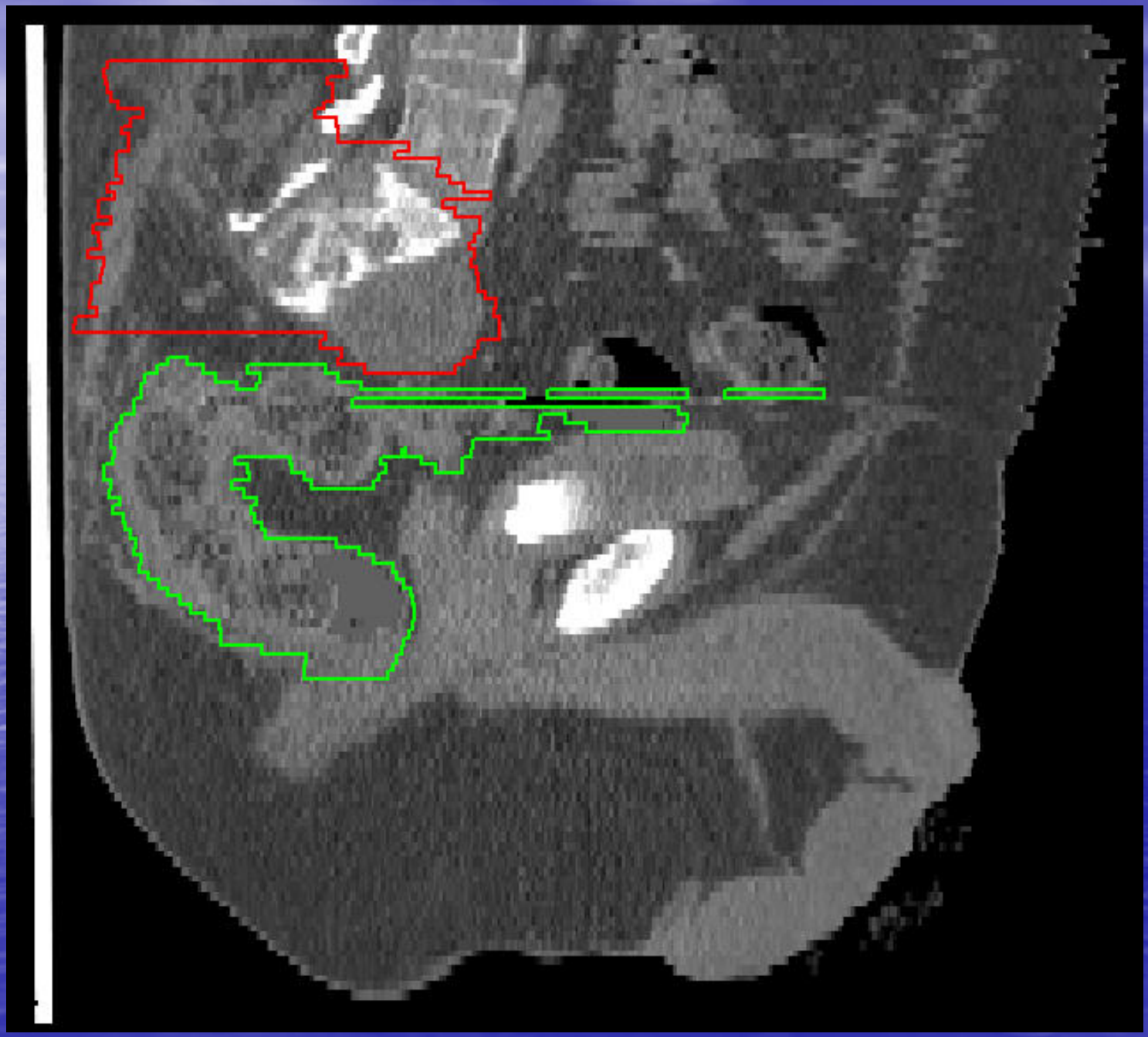

72.0 Gy
70.0 Gy
64.8 Gy
57.6 Gy
50.4 Gy
36.0 Gy
21.6 Gy

52 yo male with recuurent sacral chordoma, s/p 3 subtotal resections



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BUSINESS SYSTEMS
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DANGER KEEP OFF

Patient Description

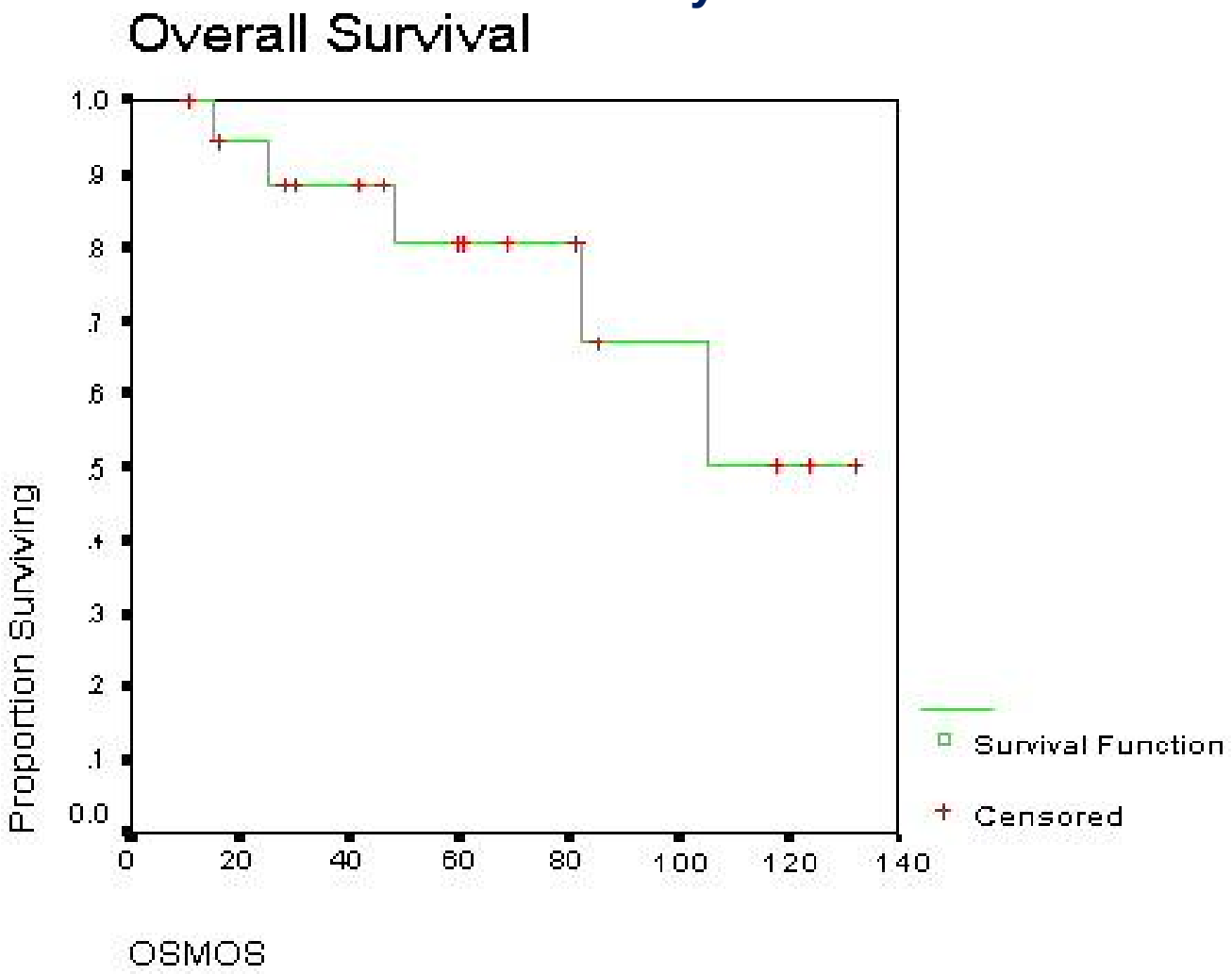
- 4 females, 15 males
- Age range: 25-82
- Average age: 60.8

Distant Mets

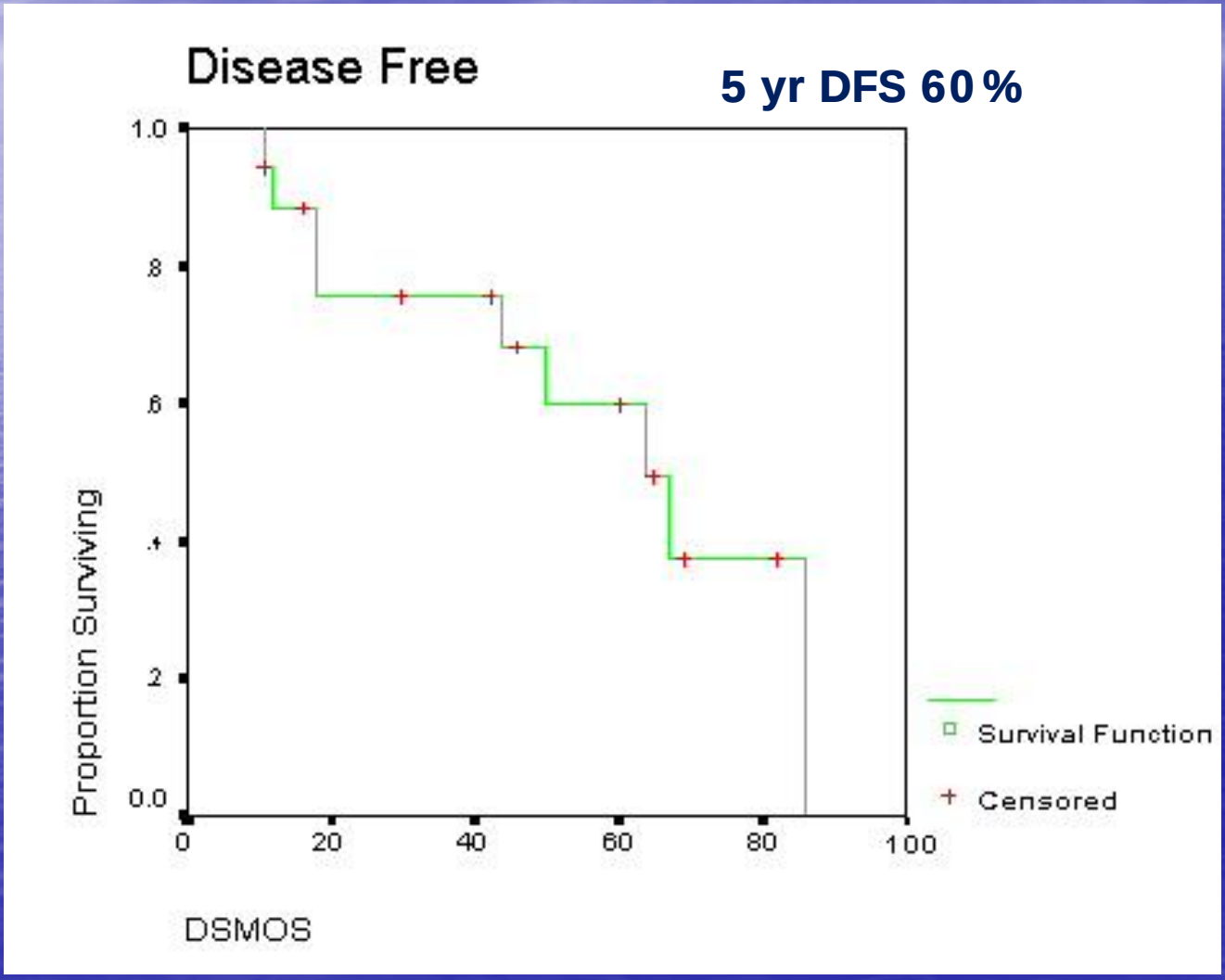
- On initial presentation 2 patients had distant metastases
- Areas include lung, liver

Overall Survival

5 yr OS 80%

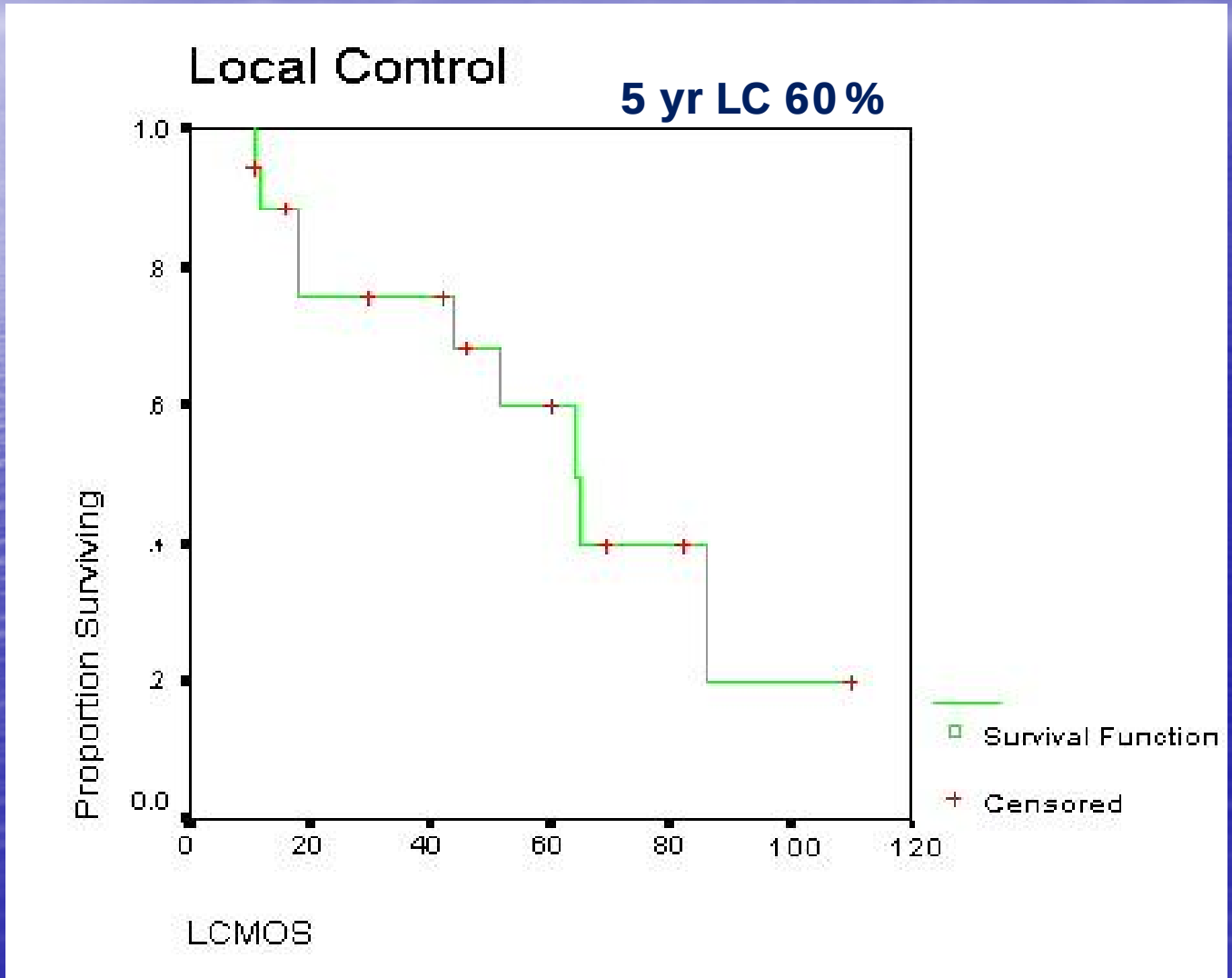


Disease Free Survival

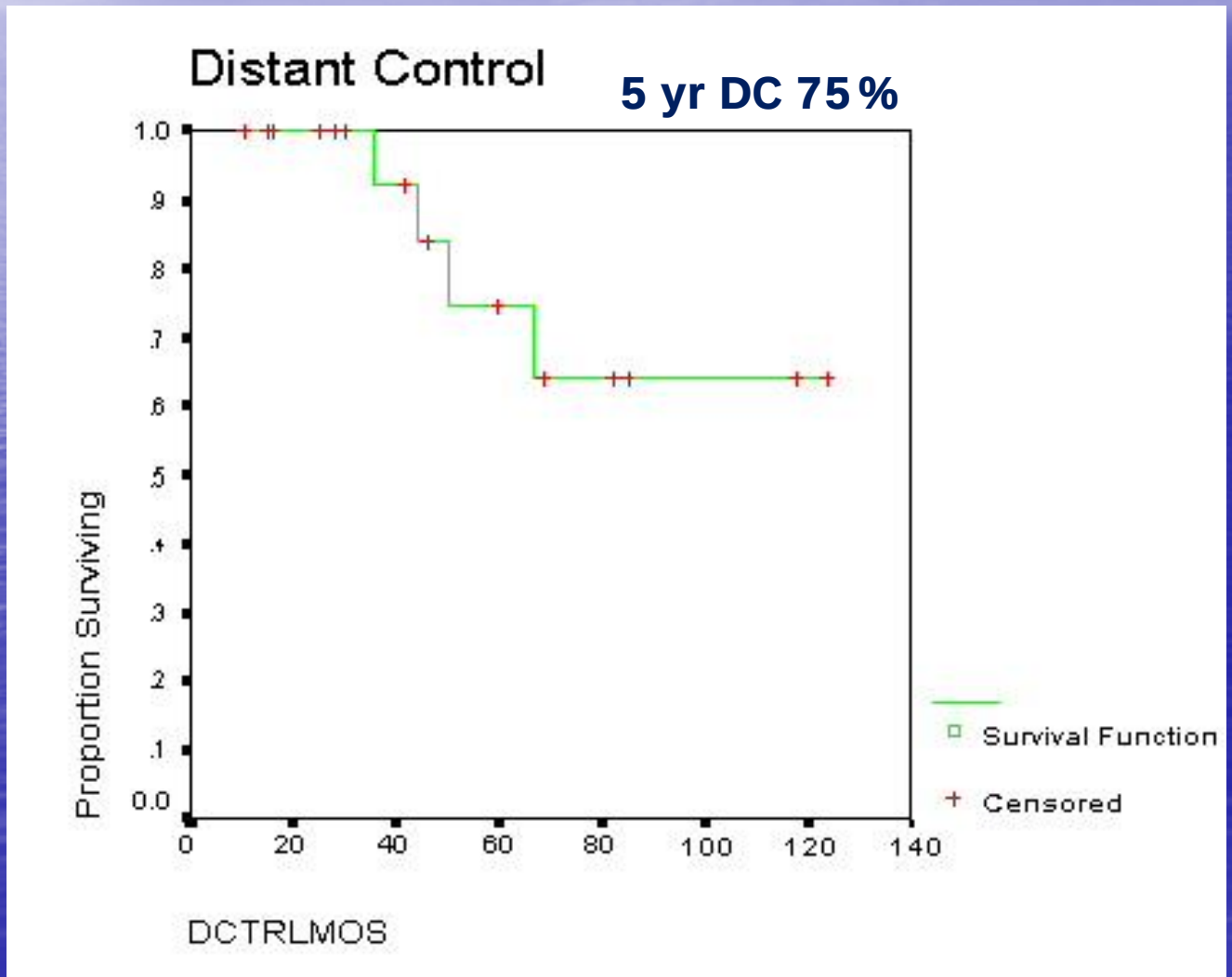


- Median time before recurrence 64 months (95% CI of 43 to 85 months)

Local Control

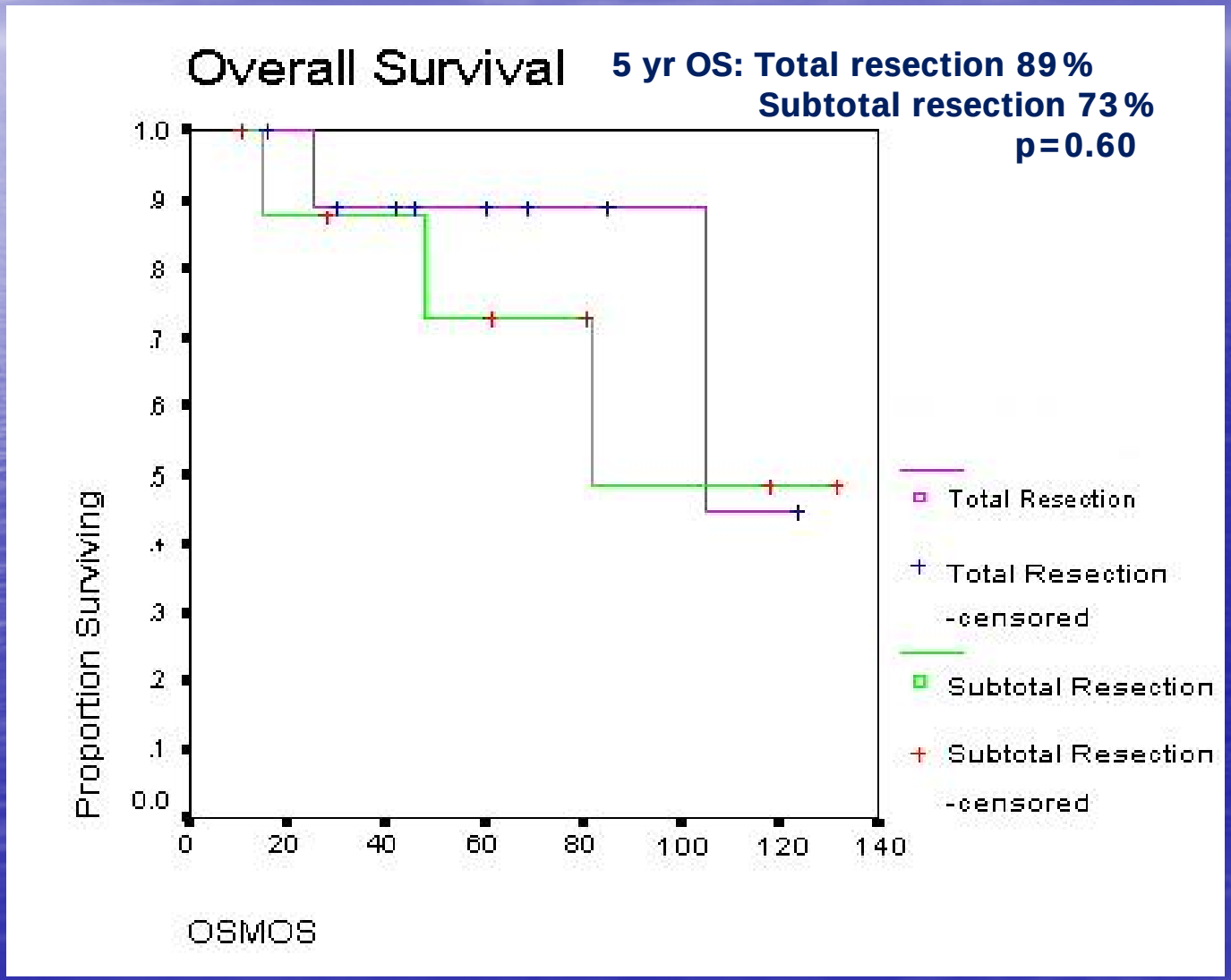


Distant Control



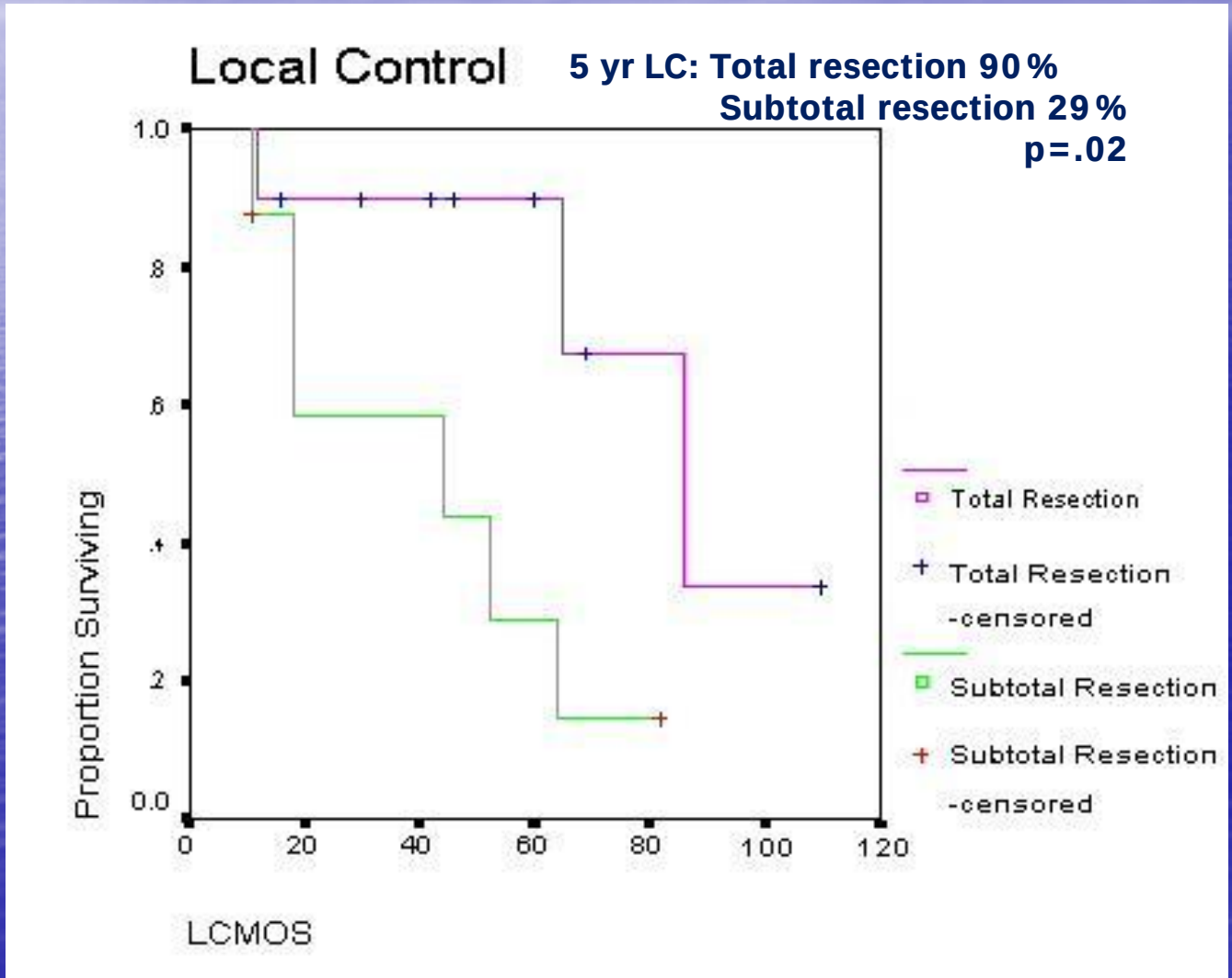
- Analyzed by extent of surgery:
- total resection vs subtotal resection

Overall Survival by Surgery Type

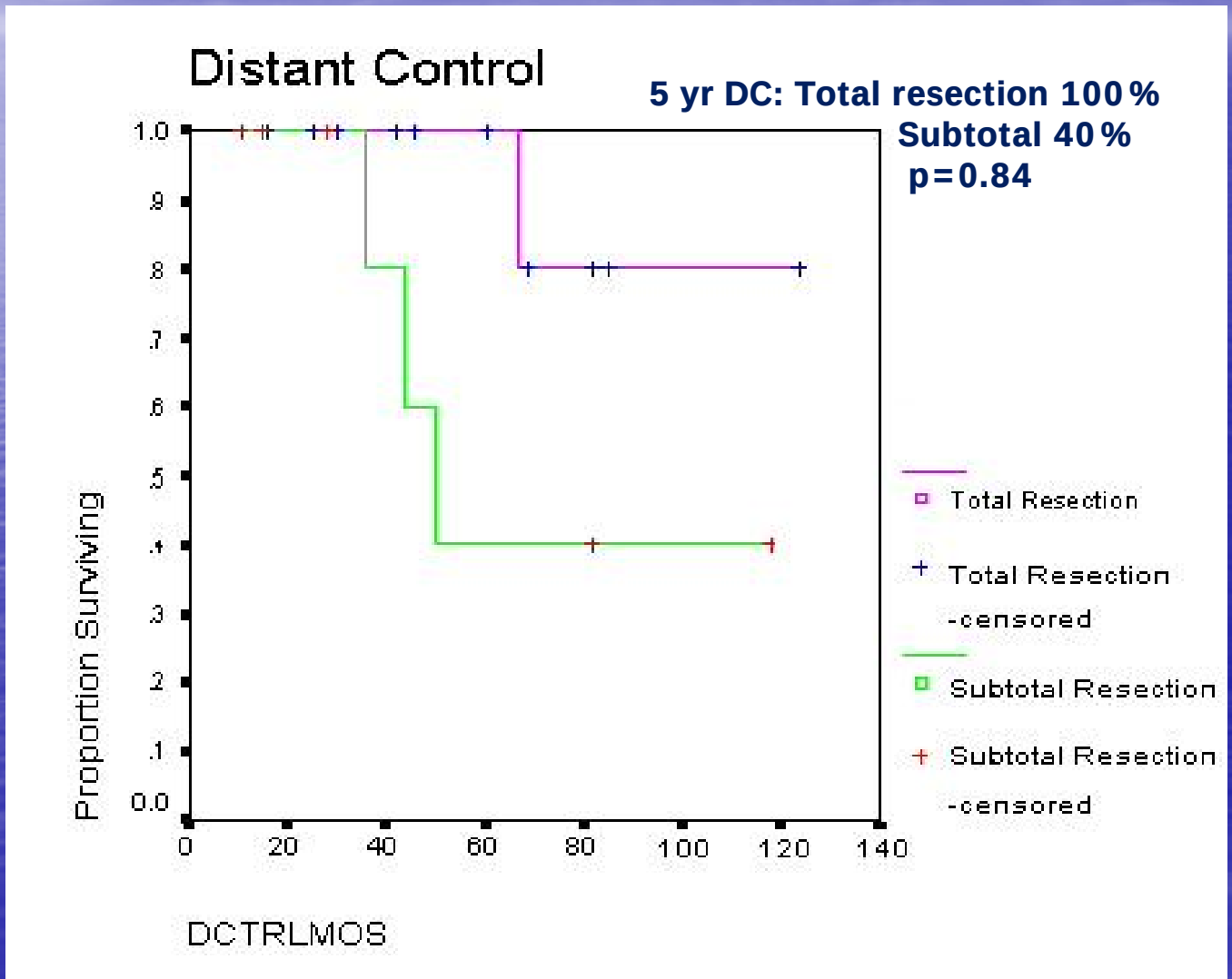


- Median time to recurrence 86 months for patients with total resection
- Median time to recurrence 44 months for patients with subtotal resection

Local Control by Surgery Type

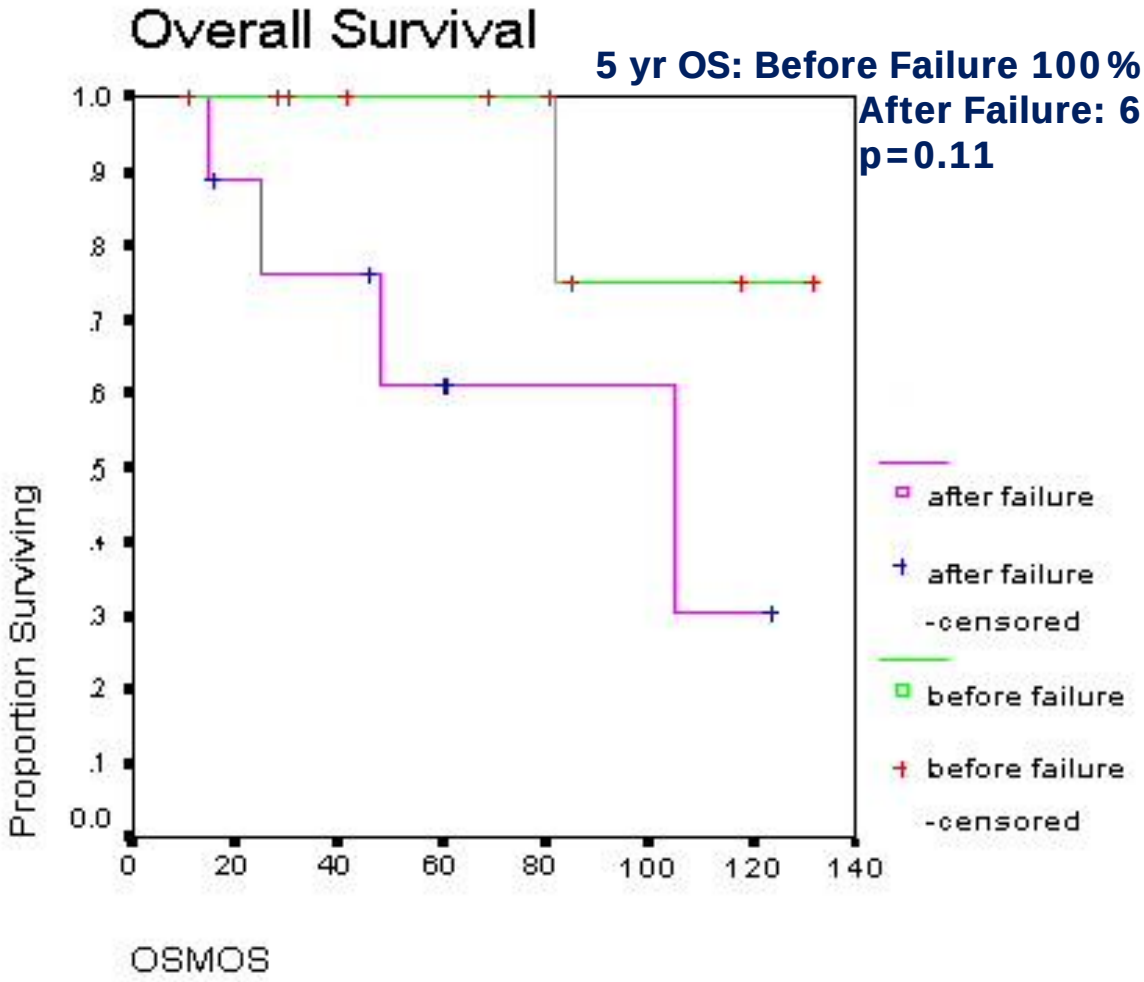


Distant Control by Surgery Type

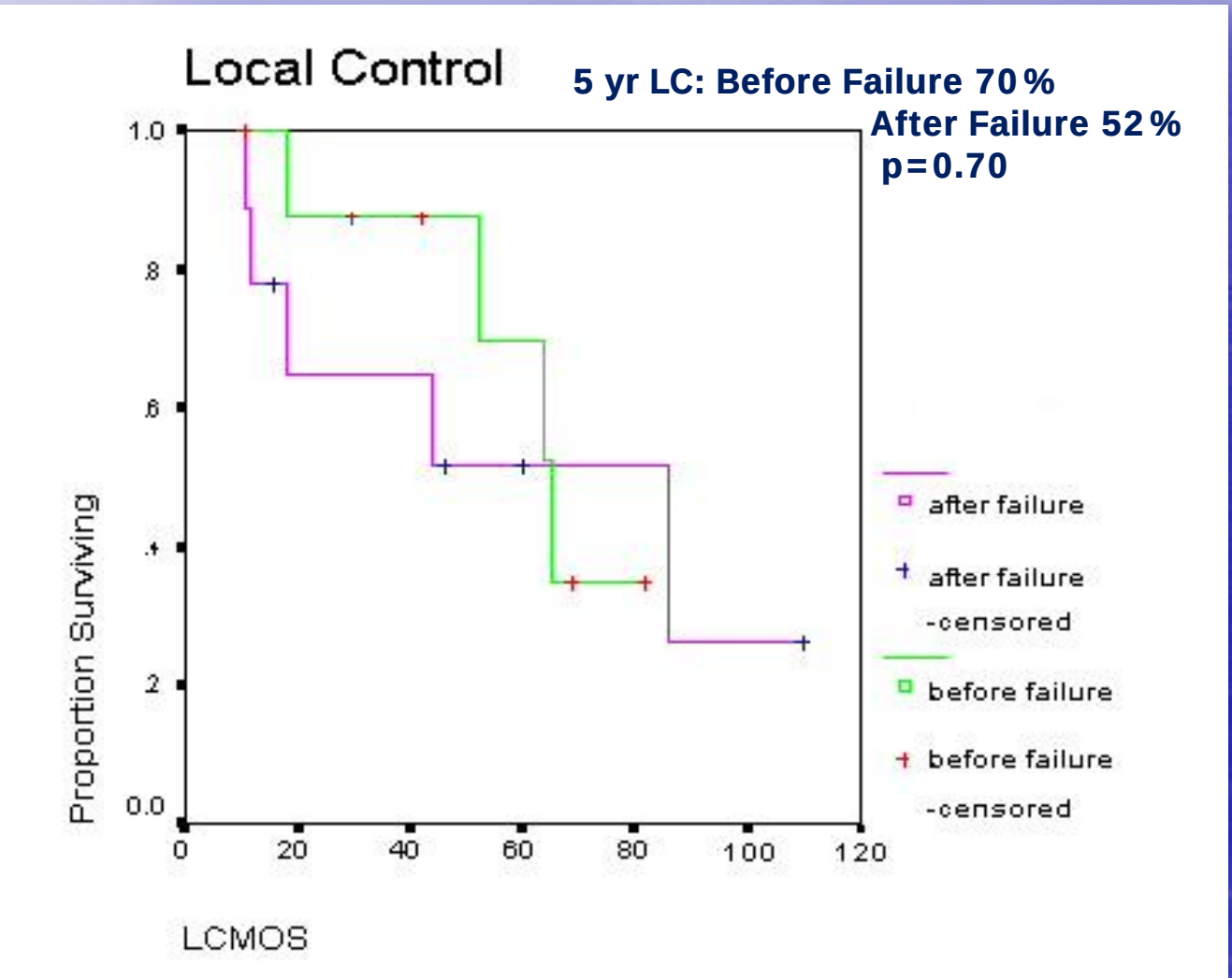


- Patients analyzed by status at study entry
- “Before Failure” group: Patients treated after initial diagnosis without recurrence
- “After Failure” group: Patients treated after recurrence

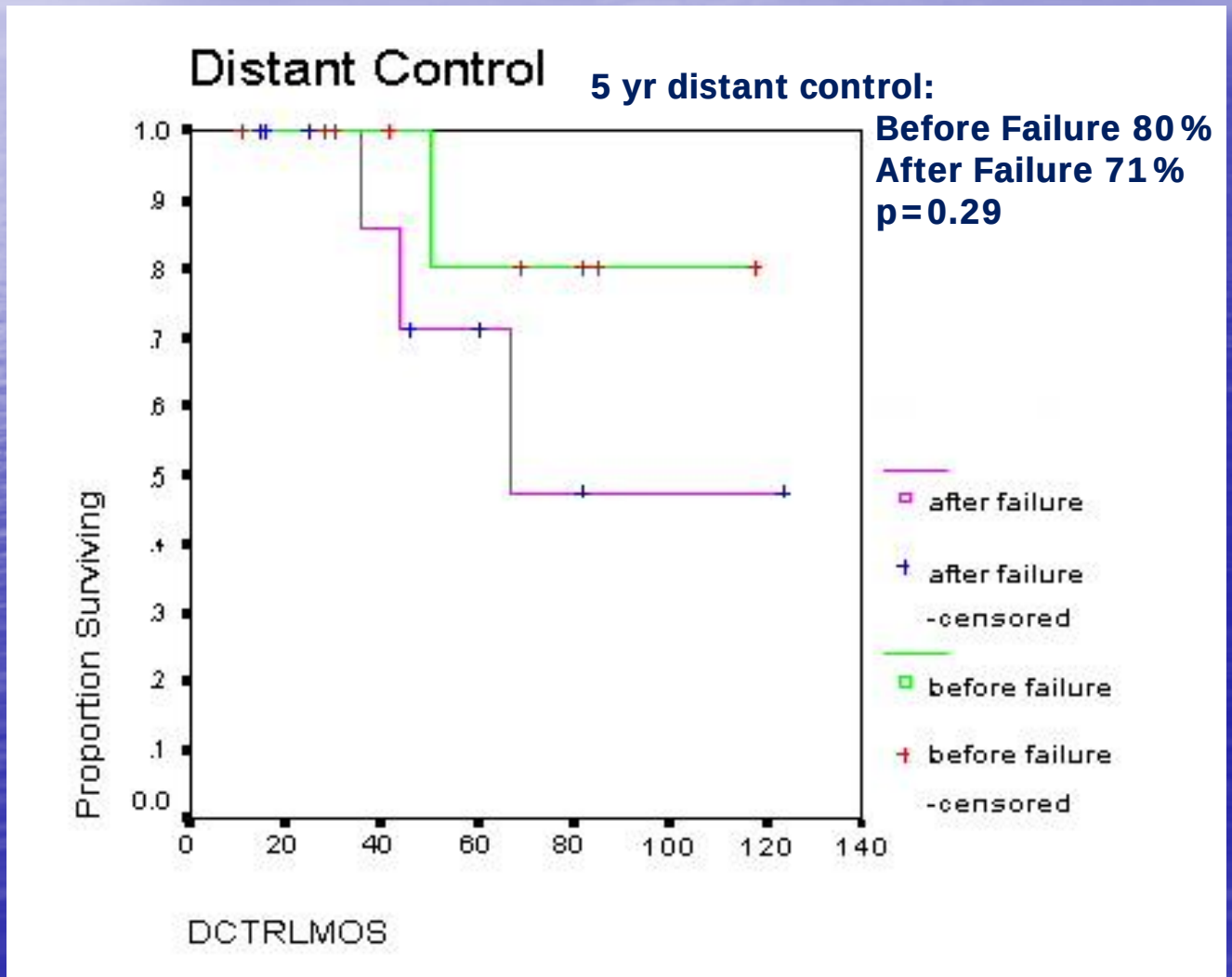
Overall Survival by Study Entry



Local Control by Study Entry



Distant Control by Study Entry



Symptomatic Relief

- 15 patients had pain on presentation
- 11 had improvement in pain
- 4 did not have pain relief and/or worsened

Motor Symptoms

- 7 had motor difficulty at presentation
- Of these 5 had improvement

Late Morbidity

ORGAN TISSUE	0	Grade 1	Grade 2	Grade 3	Grade 4	5
SKIN	None	Slight atrophy Pigmentation change Some hair loss	Patch atrophy; Moderate telangiectasia; Total hair loss	Marked atrophy; Gross telangiectasia	Ulceration	
SUBCUTANEOUS TISSUE	None	Slight induration (fibrosia) and loss of subcutaneous fat	Moderate fibrosis but asymptomatic Slight field contracture <10% linear reduction	Severe induration and loss of subcutaneous tissue Field contracture >10% linear measurement	Necrosis	
BONE	None	Asymptomatic No growth retardation Reduced bone density	Moderate pain or tenderness Growth retardation Irregular bone sclerosis	Severe pain or tenderness Complete arrest of bone growth Dense bone sclerosis	Necrosis/ Spontaneous fracture	

Late Morbidity

- Skin morbidity: No patients developed Grade 3 or higher late morbidity
- 5 patients had Grade 1, 2 scores
- Subcutaneous morbidity: 1 patient had grade 3
- 3 patients had Grade 1, 2 scores
- Only other late morbidity was one patient who developed Grade 3 bone complication

Summary

- 5 yr overall survival 80%
- 5 yr disease free survival 60%
- 5 yr local control 60%
- 5 yr distant control 75%

Conclusions

- Surgery is important factor for local control (significant difference)
- In our study patients with total resection had a 5 year local control rate of 90%, subtotal resection 29%
- However, total resection often not possible

Future Followup

- Dose escalation possibilities
- Appears most failures are within radiation field in past studies
- Investigate rectal toxicity
- Increase dose to rectum, bladder to determine if dose escalation possible for increased tumor control

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