Caring for Pediatric Patients with Malignant Bone Tumors:

The Advanced Practice Nurse's Perspective

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APHON National Conference - September 2020

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Disclosures

- No relevant conflicts of interest or disclosures to discuss!

Objectives

- To describe the presentation, diagnosis, and management of pediatric malignant bone tumors
- To explore conference identified learning gaps
- To address how to support patients through the acute and late effects of these tumors and their therapy
- To discuss the APP role and the importance of interprofessional practice in the care of these patients

Case Study #1

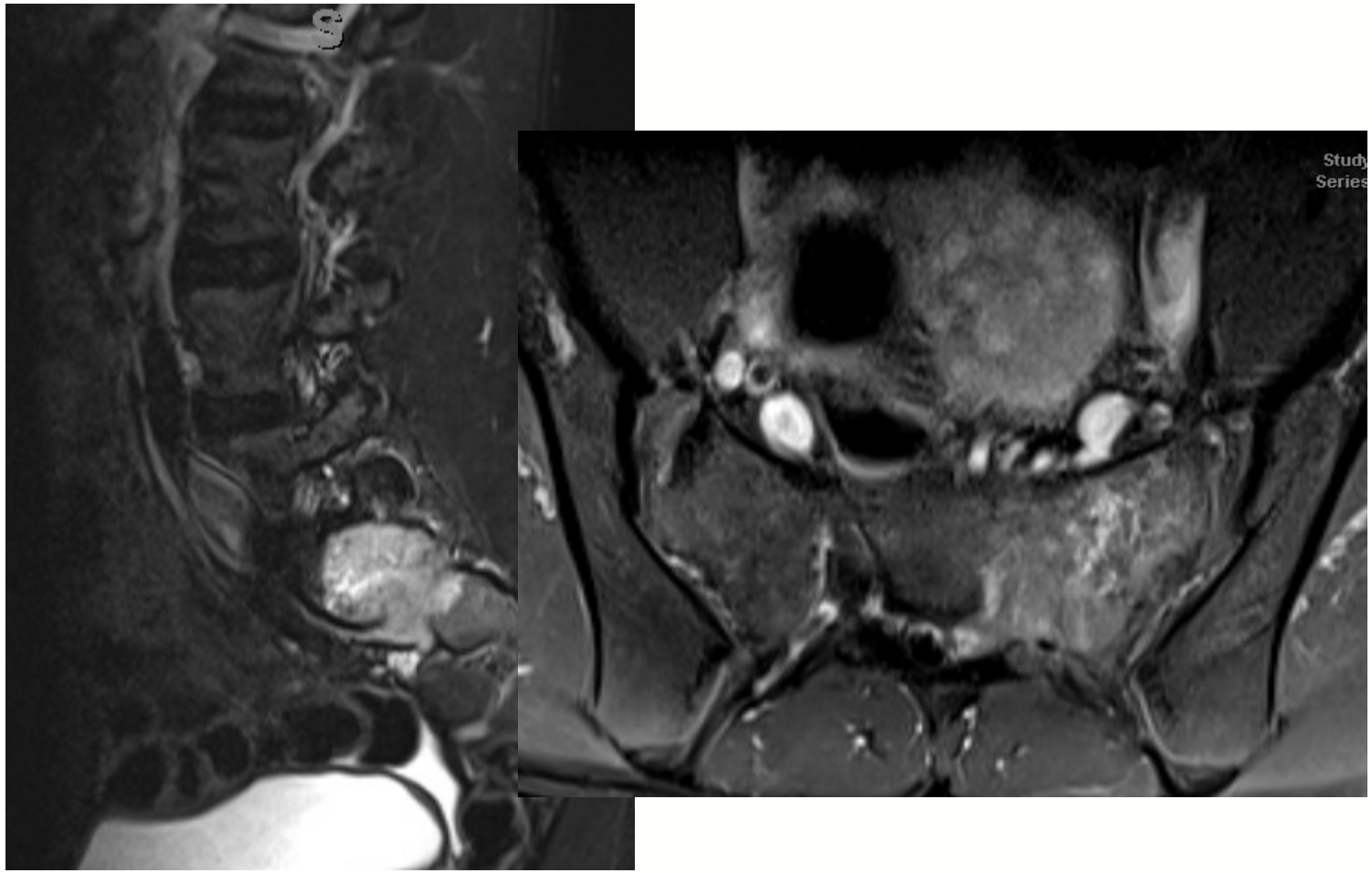
- 18 year old male
- 2 month hx of left calf stiffness and weakness
- Symptoms progressed to sharp shooting pain in the back of his left thigh, buttocks, and radiating to the calf
- No urinary or bowel incontinence, saddle anesthesia, fever, decrease in appetite
- 5 pound weight loss over the past 1-2 months

Case Study #1

- Seen in outpatient ortho clinic
- Physical examination
  - 4/5 strength in left leg
  - Hyporeflexia on left leg
  - Decreased sensation over Left thigh
  - Normal rectal tone
  - Limp
  - Unable to plantar flex left foot or do left leg calf raises
- MRI lumbar spine revealed lesion in S2
- Referral to pediatric oncology

Diagnostic Work Up

- MRI lumbar spine
- Biopsy
- CT chest
- PET scan



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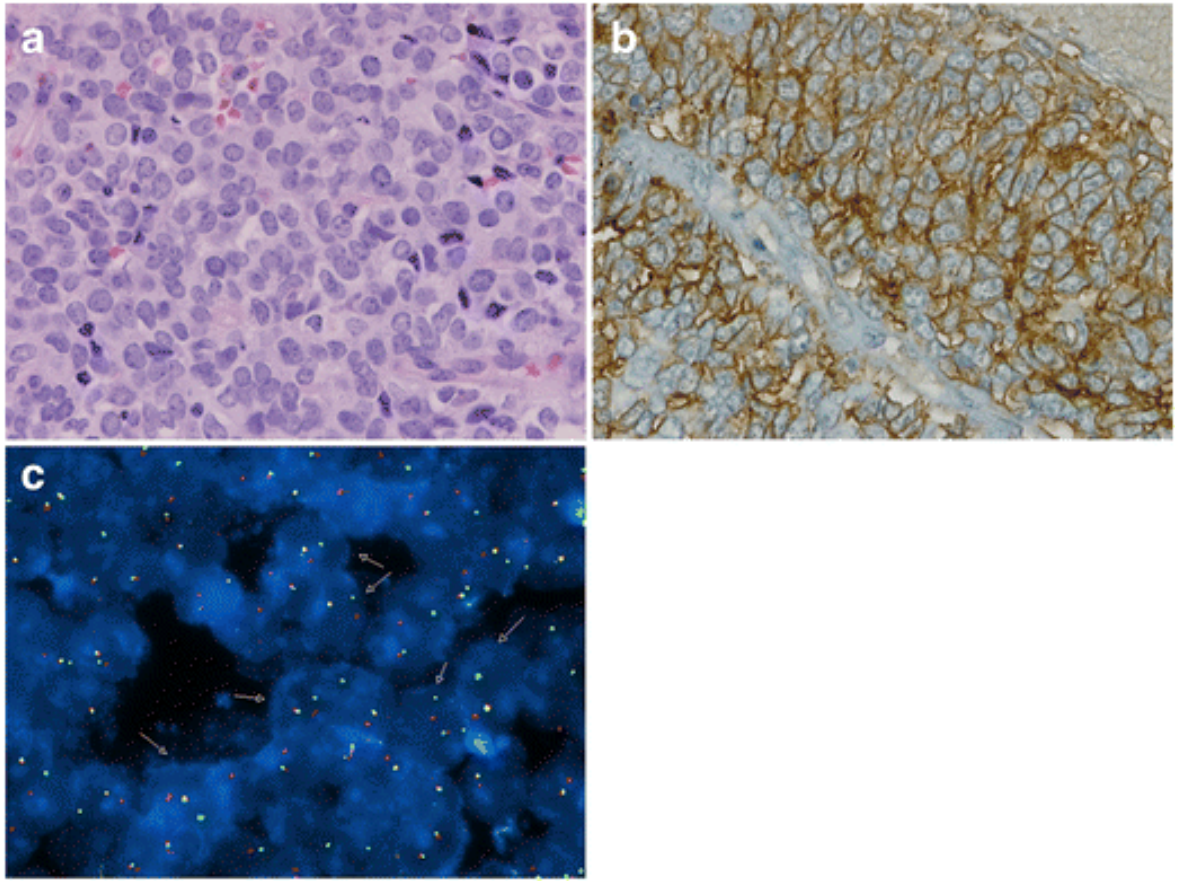
Diagnostic Work Up: Biopsy

- Needle
- Open incisional
- Lab testing

https://www.chop.edu/pediatric-fellowships/interventional-radiology/fellowship

Ewing Sarcoma

- Epidemiology
- Biology
- Genetics
- Risk factors
- Clinical presentation
- Staging
- Prognostic factors



http://europepmc.org/article/PMC/5808087

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Supportive Care at Diagnosis

- Pain control
- Physical therapy
- Education
- Interprofessional support

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Plan

- Diagnostic and staging workup
- Baseline evaluations
- Central line placement
- Chemotherapy
- Re-staging scans
- Local control
- Monitoring after therapy

Chemotherapy

- Neoadjuvant and adjuvant therapy
- Importance of interval compression
- Standard of care
  - VDC
  - IE
- Notable toxicities r/t chemotherapy
- Supportive care during chemotherapy

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Surgery

- S2 vertebral tumor - challenging location
- Risks
  - Loss of bladder or bowel function
  - Loss of sensation to foot or ankle
  - Loss of motor function to foot or ankle
  - Tumor recurrence or positive margins

IN DEPTH: Radiation for Ewing Sarcoma

- Radiosensitive tumor
- Efficacy
  - Tumor size and location may influence outcome
  - No randomized controlled trials comparing radiation and surgery
- Generally used when surgery is not an option or if poor functional outcome is expected with surgery

Radiation: Dose

- Measured in Gy
  - Absorption of one joule of radiation per kilogram of matter
  - 1 Gy = 100 cGy
- Definitive Dose for Ewing Sarcoma: 55.8 Gy (5580 cGy)
  - Pre-chemo volume to 45 Gy (4500 cGy)
  - Boost to residual tumor (determined by re-staging scans)
- Dose is divided into daily fractions

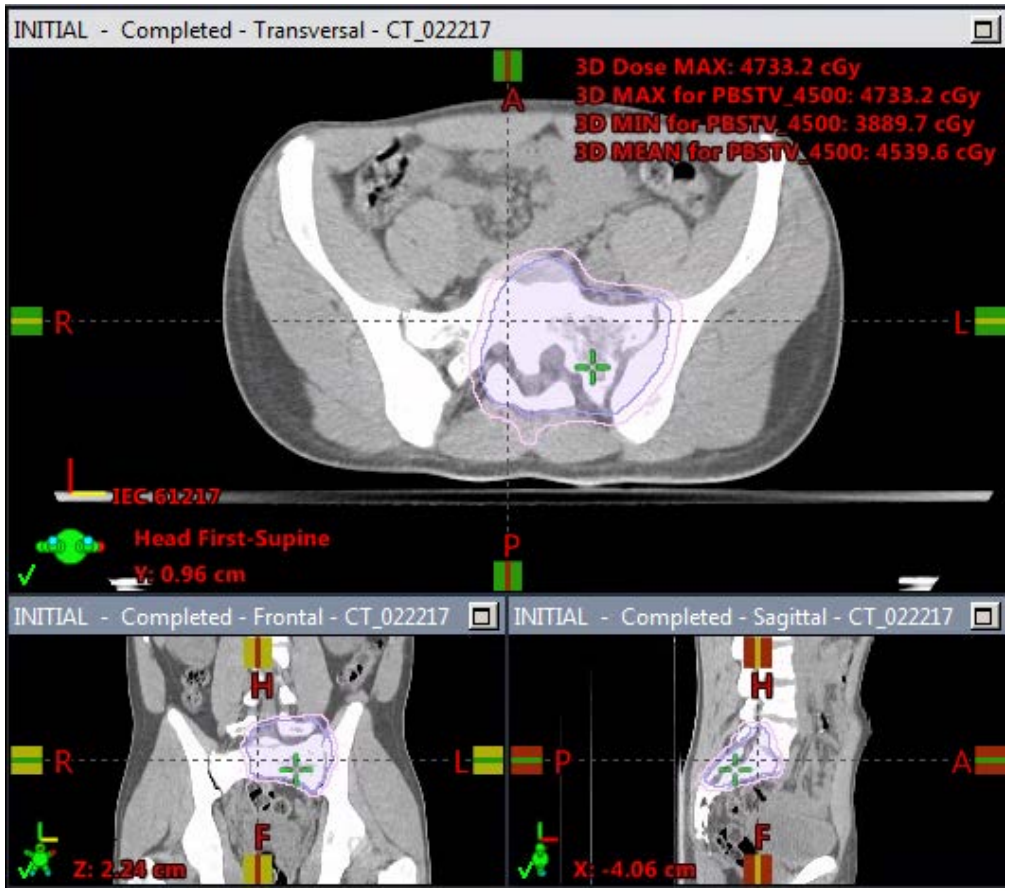
Radiation: Planning Process

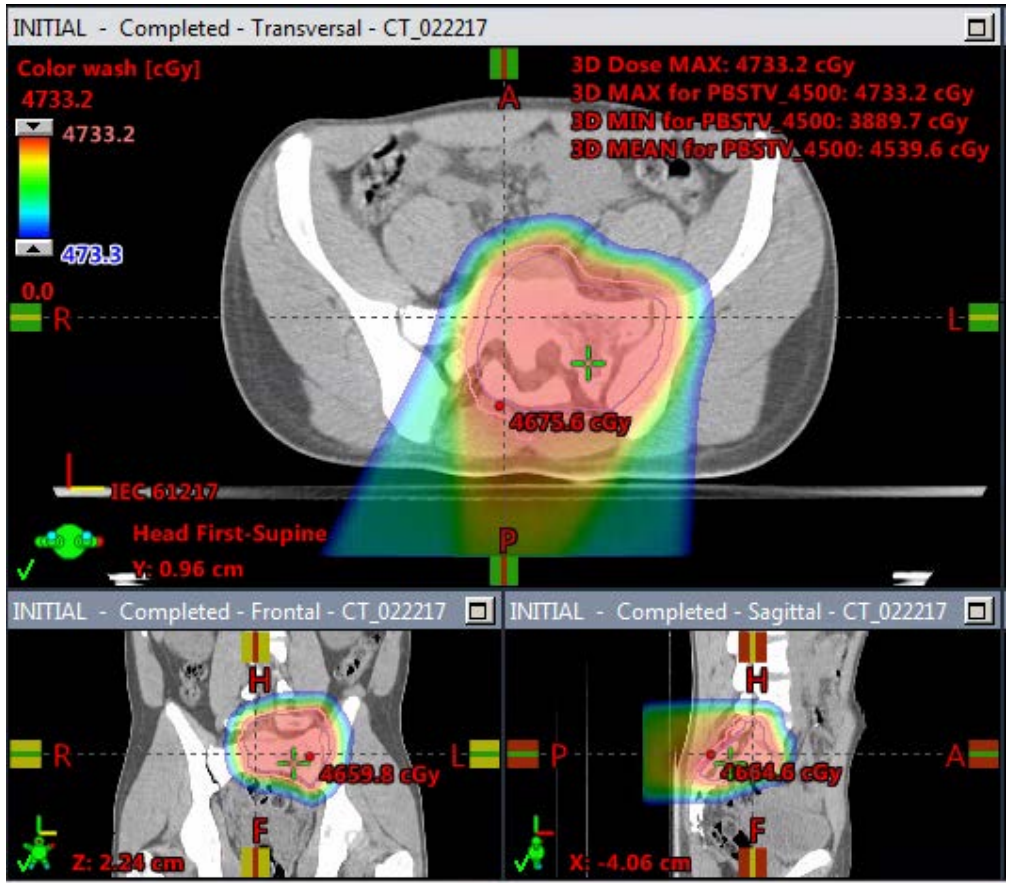
- CT-Simulation
  - Non-contrast scan
  - Create immobilization devices and place tattoos to replicate position
  - Anesthesia may be required for young children

- SIM CT is fused with diagnostic imaging
- Radiation Oncologist contours the target volumes
- Dosimetrists and Physicists design the beam angles
- Radiation Oncologist reviews and approves the plan (or sends back for revisions)
- Rigorous quality assurance process

Initial Radiation Plan

- Pre-chemo volume to 4500 cGy





Proton Plan

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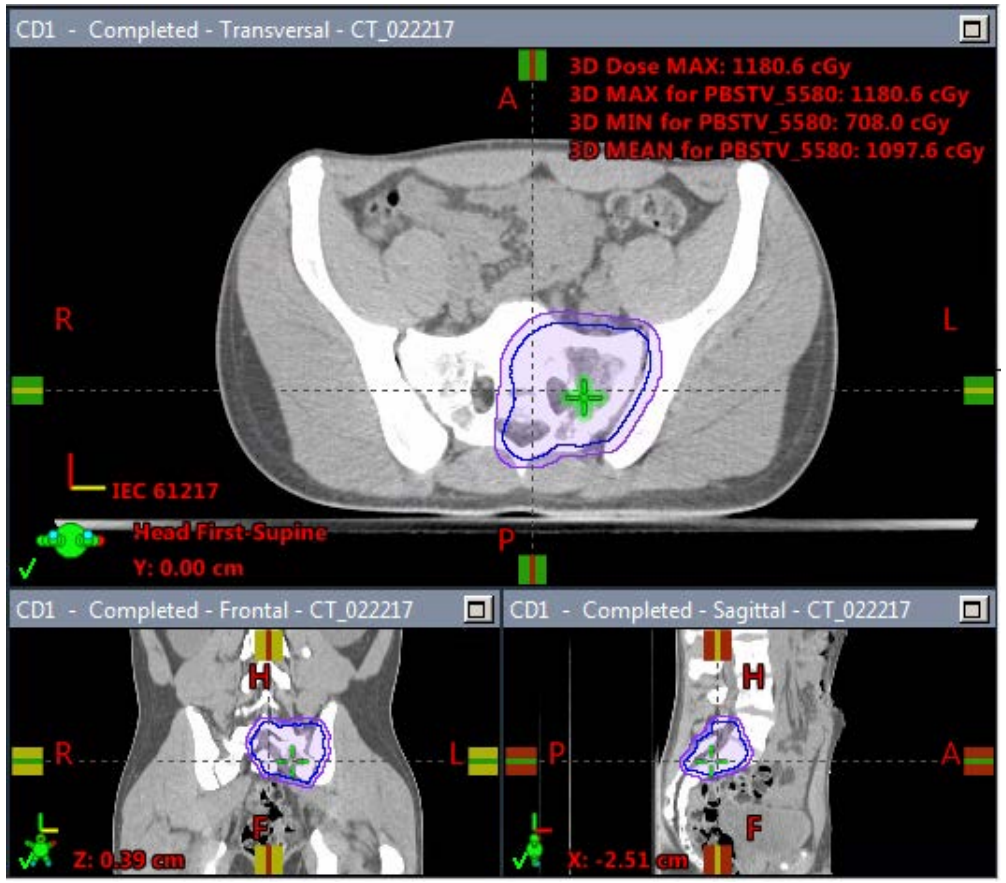
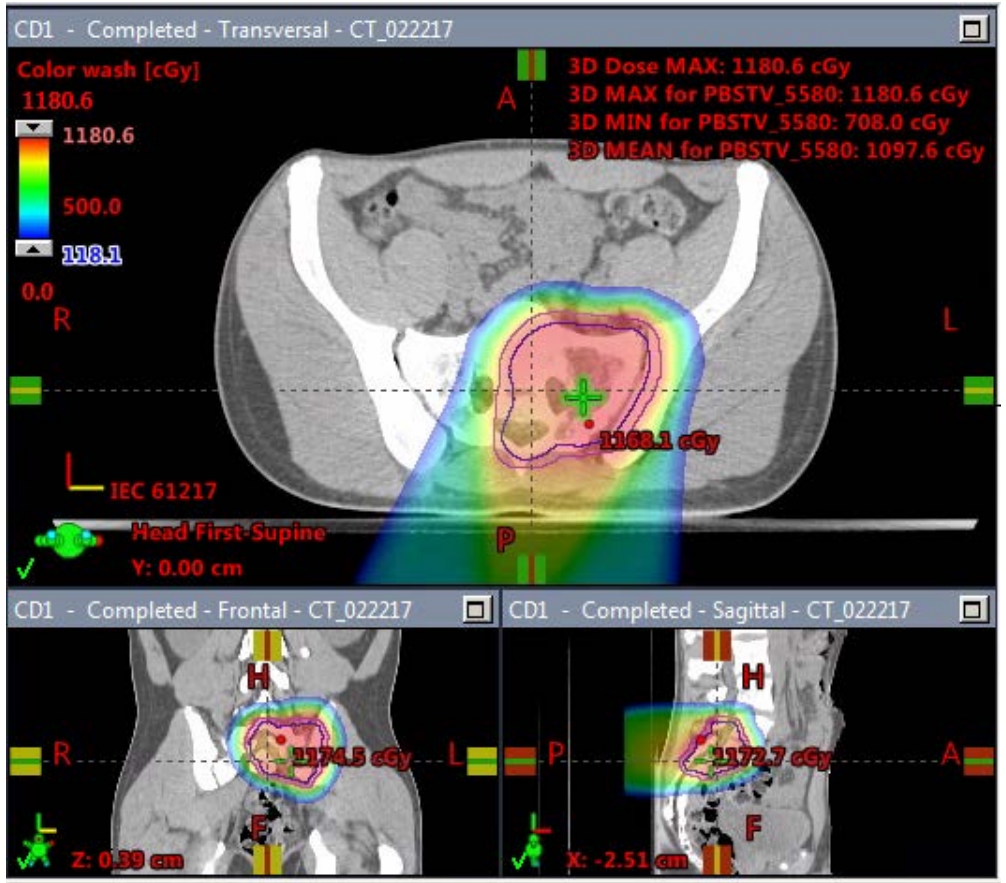
Boost Volume

- Residual tumor volume to 5580 cGy





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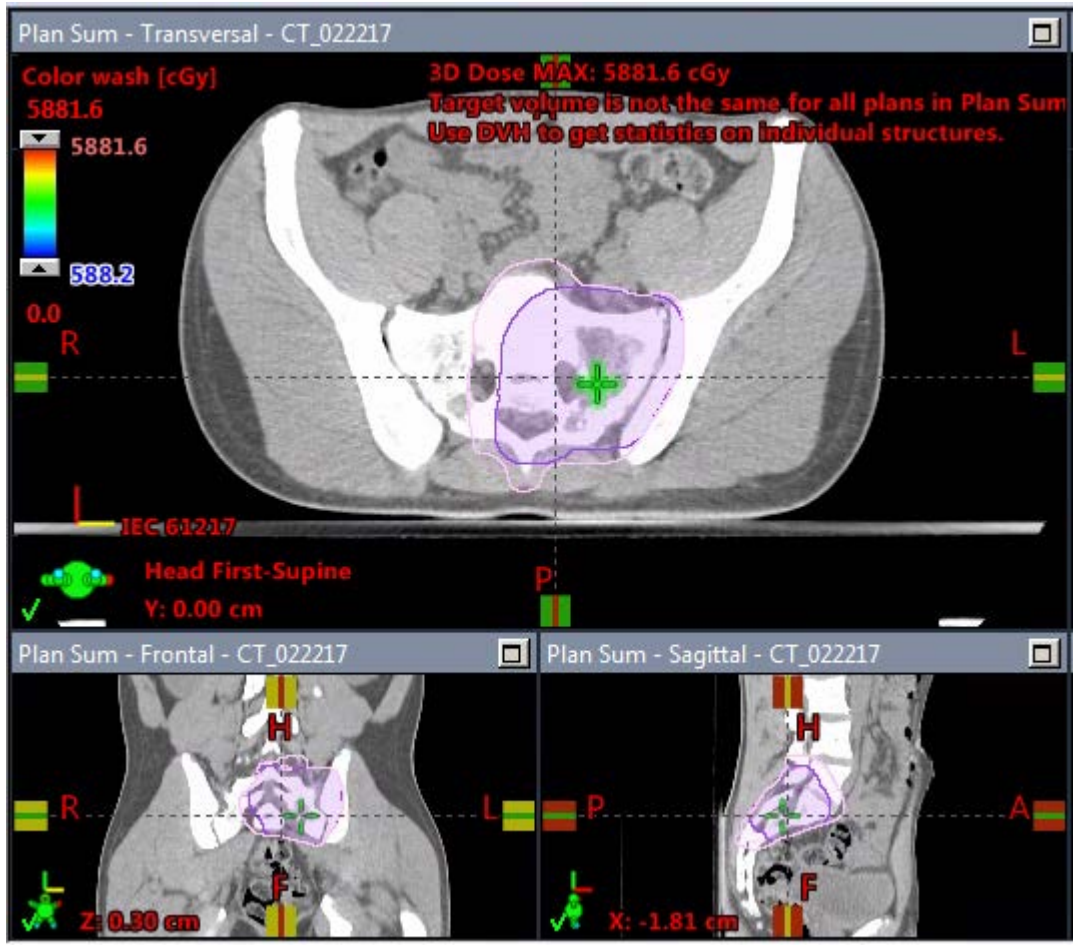
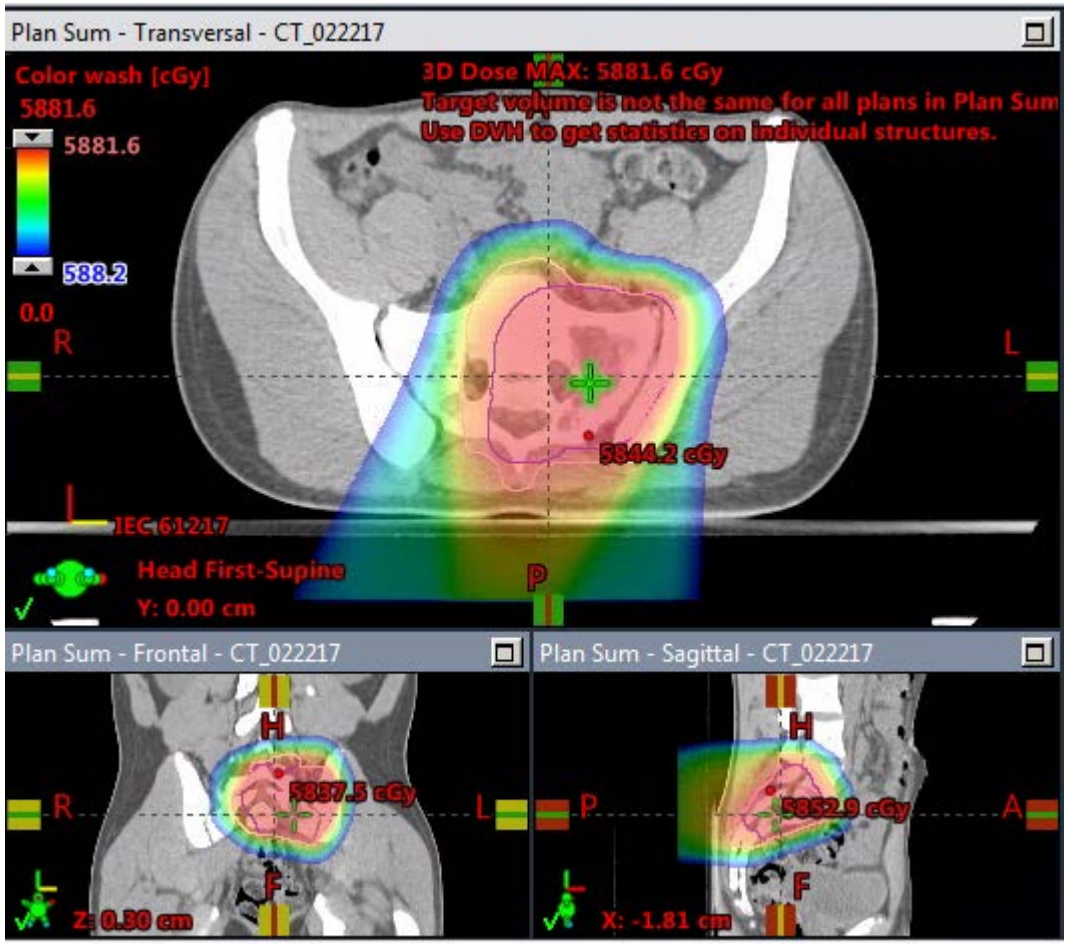
Complete Radiation Plan

- Plan Sum





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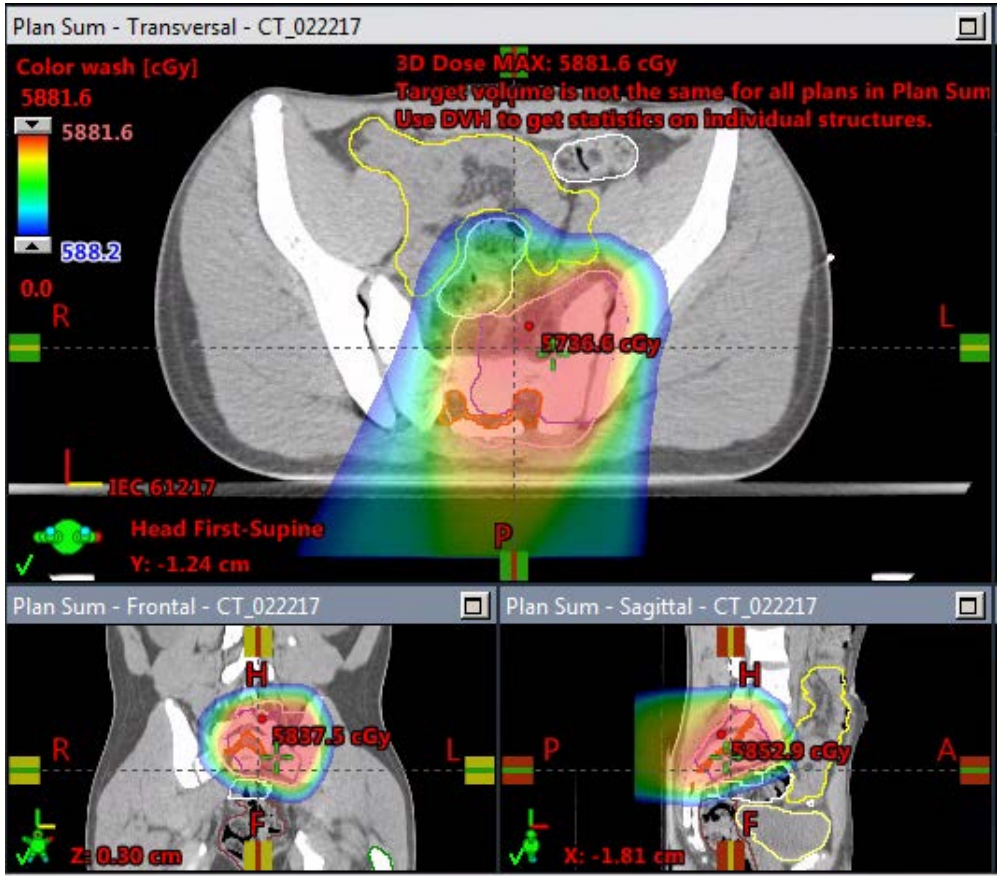
Radiation Treatment

- Goals
  - #1: Treat the tumor
  - #2: Minimize toxicity to normal cells
- Consider the real estate
  - Bladder
  - Bowel (large and small)
  - Cauda
  - Rectum
  - Femoral heads
- Determine the optimal modality



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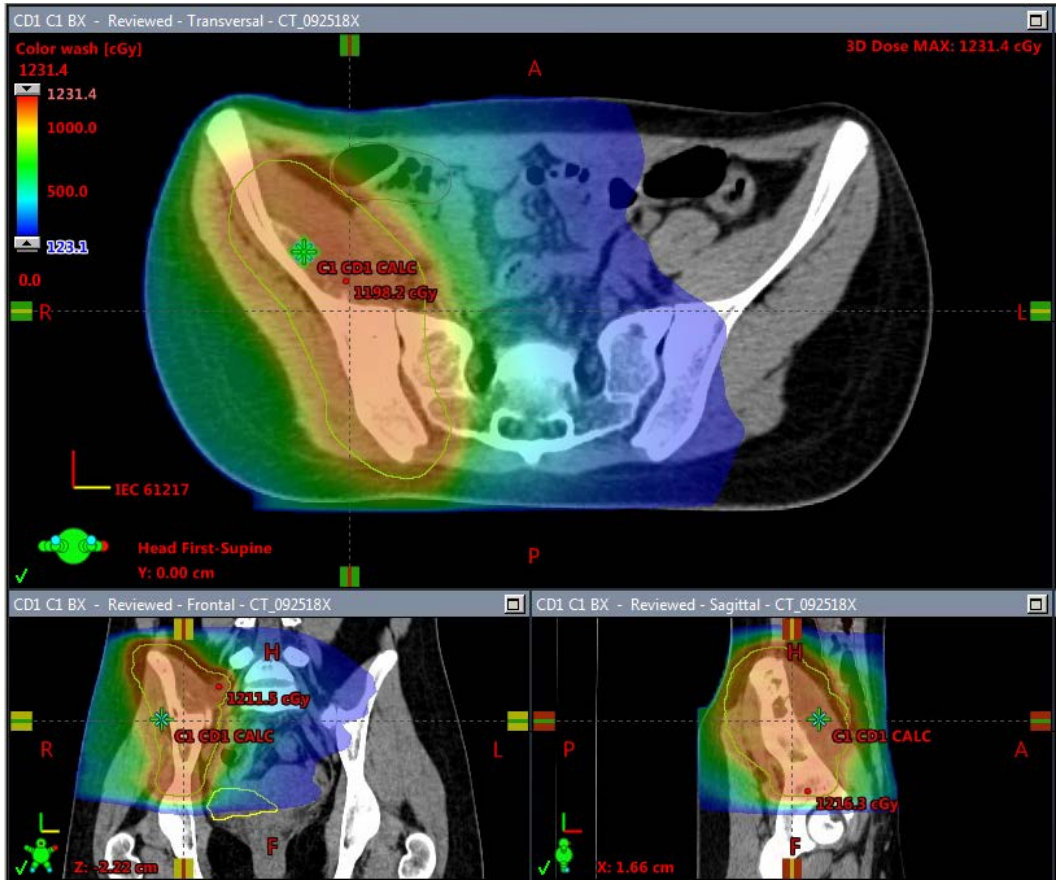
Radiation Modalities

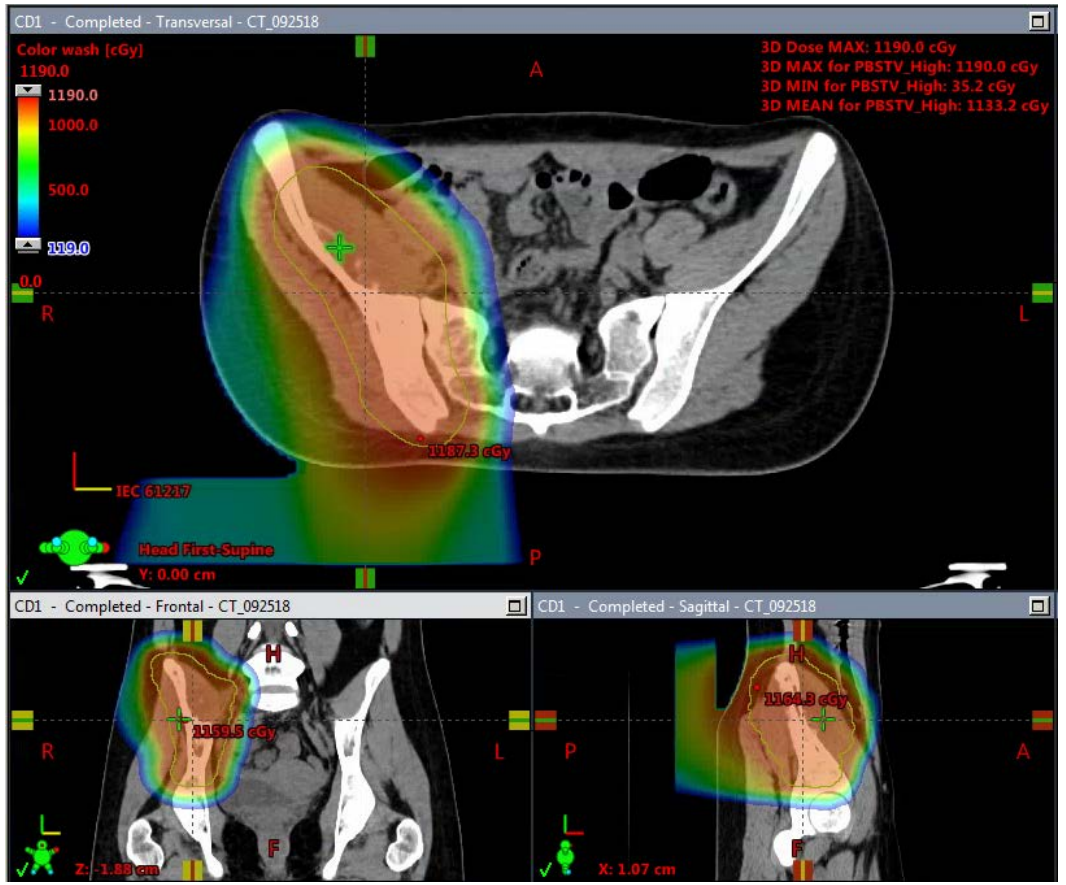
- **Photons** (x-rays)
  - AP/PA (2D)
    - Most basic, used in the emergent setting
  - 3D
    - Basic - extremities, whole abdomen, whole brain
  - Intensity Modulated Radiation Therapy (IMRT)
    - Superconformal
- **Protons**
  - Double scatter
    - Less conformal, increased dose to skin
  - Pencil Beam Scanning
    - Dose paint, conformal, less dose to normal tissue

Plan Comparison of Radiation Modalities

- Photons vs. Protons



IMRT Plan



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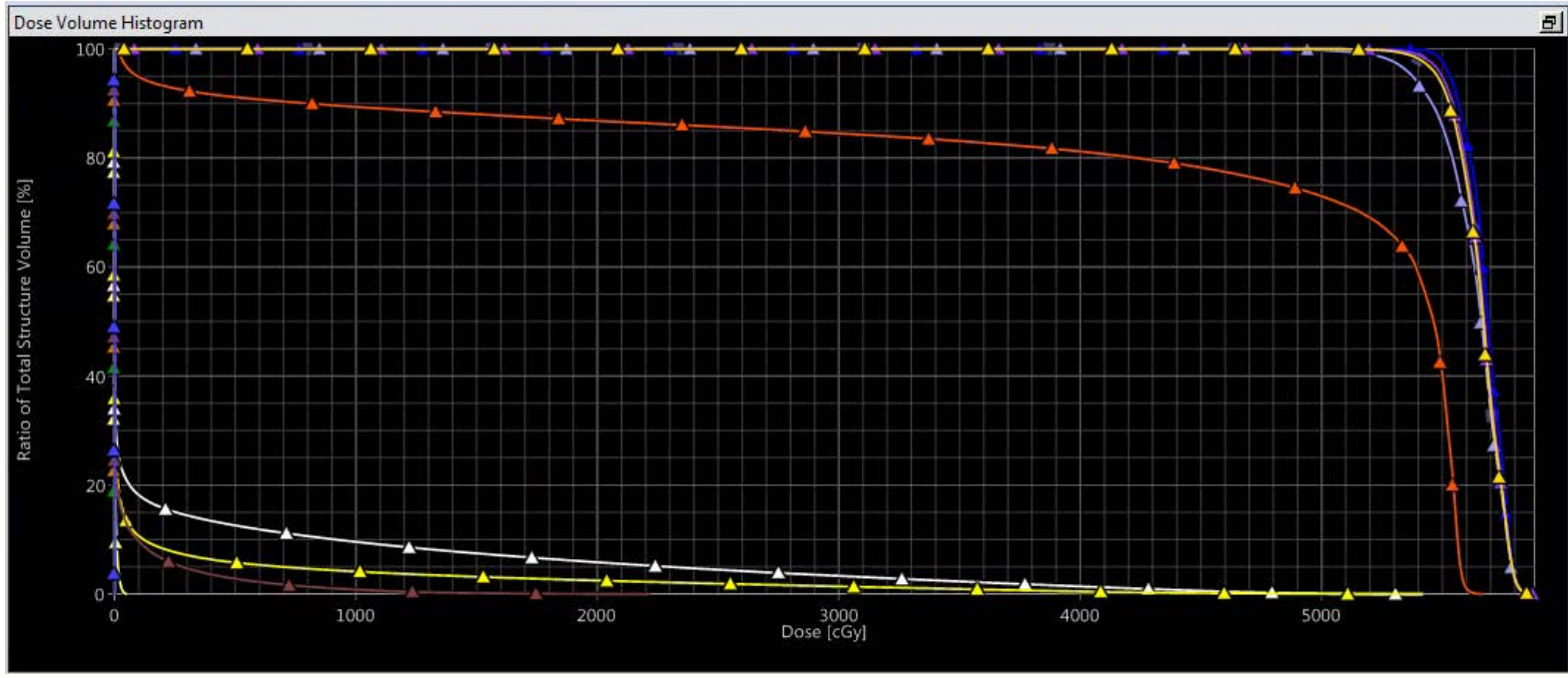
Dose Volume Histogram

- Dose to bladder, bowel, and rectum very low
- Cauda- mean dose is 46 Gy
- Highest dose to tumor



Show DVH	Structure	Approval Status	Plan	Course	Volume [cm³]	Dose Cover.[%]	Sampling Cover.[%]	Min Dose [cGy]	Max Dose [cGy]	Mean Dose [cGy]
☑	RECTUM_RTOG	Approved	Plan Sum	C1 PELVIS	143.6	100.0	100.0	0.0	2222.1	45.7
☑	PTV_5580	Approved	Plan Sum	C1 PELVIS	274.4	100.0	100.0	4830.0	5881.6	5663.9
☑	PBSTV_5580	Approved	Plan Sum	C1 PELVIS	288.4	100.0	100.0	4829.6	5881.6	5656.8
☑	Genitalia	Approved	Plan Sum	C1 PELVIS	264.0	100.0	100.0	0.0	0.0	0.0
☑	FemoralgHead_R	Approved	Plan Sum	C1 PELVIS	171.1	100.0	100.0	0.0	0.0	0.0
☑	FemoralHead_L	Approved	Plan Sum	C1 PELVIS	165.3	100.0	100.0	0.0	4.5	0.3
☑	CTV_5580	Approved	Plan Sum	C1 PELVIS	175.0	100.0	100.0	5364.3	5878.2	5681.2
☑	CTV_4500	Approved	Plan Sum	C1 PELVIS	250.8	100.0	100.0	4778.5	5881.6	5631.6
☑	CAUDA	Approved	Plan Sum	C1 PELVIS	29.0	100.0	100.1	10.6	5669.6	4607.4
☑	Bowel_Small	Approved	Plan Sum	C1 PELVIS	545.9	100.0	100.0	0.0	5422.5	132.8
☑	Bowel_Large	Approved	Plan Sum	C1 PELVIS	394.3	100.0	100.0	0.0	5428.6	286.9
☑	Bladder	Approved	Plan Sum	C1 PELVIS	208.5	100.0	100.0	0.0	56.1	2.5
☐	PTV_4500	Approved	Plan Sum	C1 PELVIS	384.9	100.0	100.0	4311.8	5881.6	5564.0

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Acute Side Effects of Pelvic Radiation

- Fatigue
- Skin erythema (radiation dermatitis)
- Nausea and vomiting
- Diarrhea
- Anemia, thrombocytopenia
- Delayed count recovery

Late Side Effects of Pelvic Radiation

- Decreased velocity of bone growth
- Arthritis in hip joint (if in the field, usually avoided)
- Slightly increased risk of bowel obstruction, altered bowel habits
- Increased risk of fracture
- Risk of second malignancy

Case Study #2

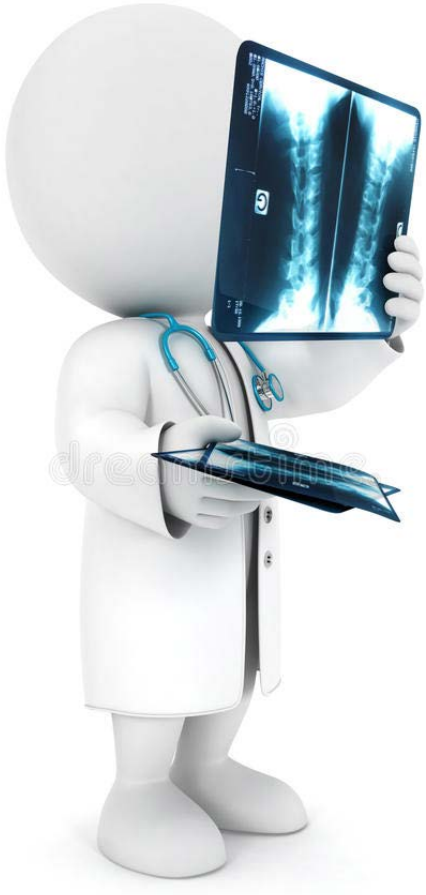
- 15 year old female
- Presented to orthopedic clinic with 4 weeks hx of right shoulder pain
- Pain when doing push ups during color guard practice
- Pain worsened over several months
- PCP ordered x-rays → referred to local orthopedist
- MRI right shoulder → concern for tumor

Case Study #2

- On exam
  - Firm and fixed mass palpable at right proximal humerus
  - Pain with shoulder abduction + internal/external rotation
  - Tenderness to palpation
  - Neurovascular exam intact
  - No deficits in strength

Diagnostic Work Up

- MRI right shoulder
- IR guided biopsy
- CT chest
- PET scan



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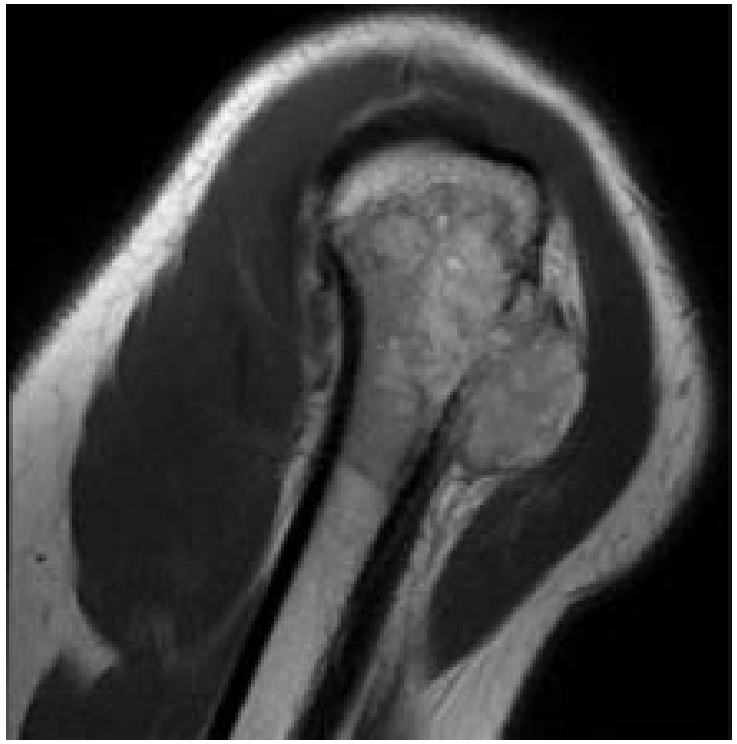
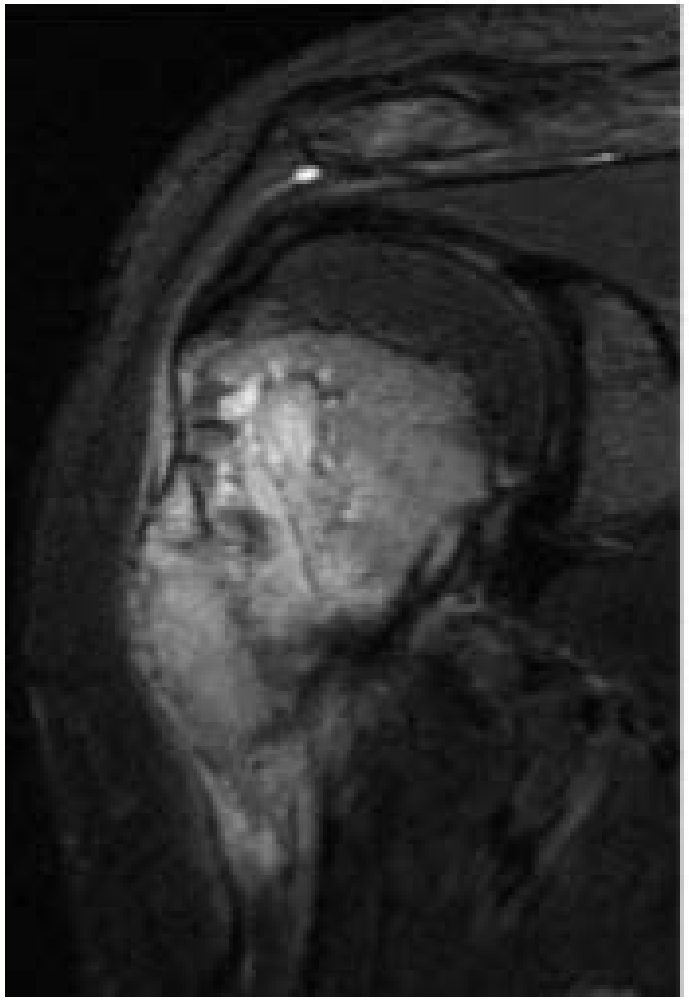
Diagnostic Work Up: Imaging





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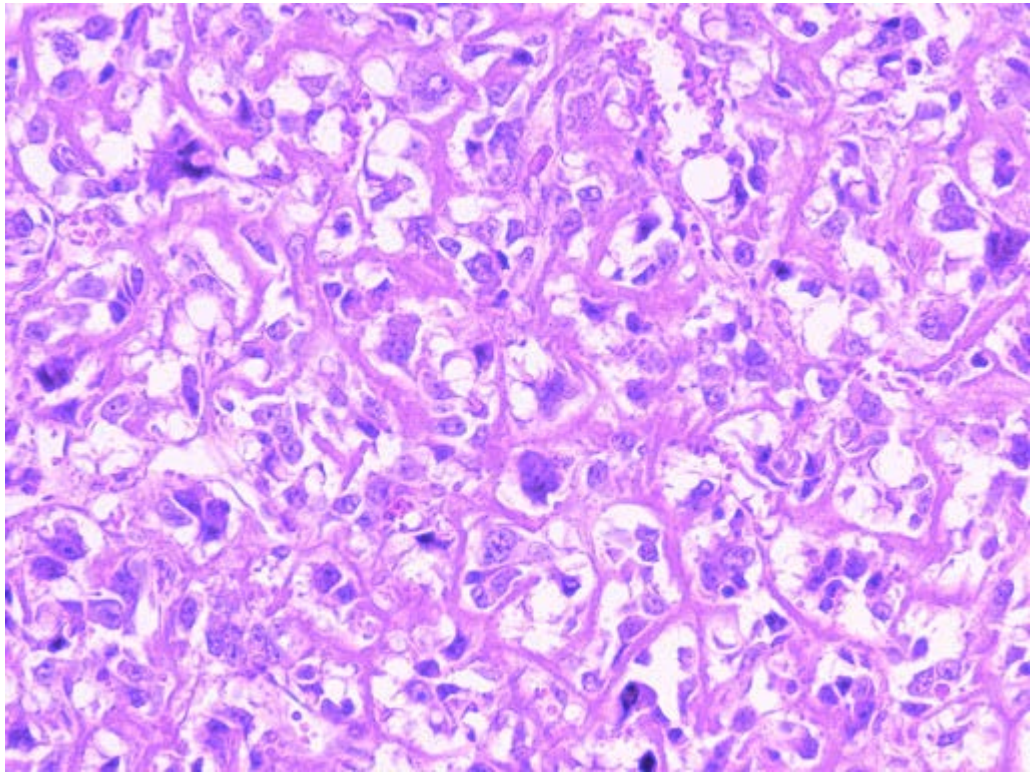
Osteosarcoma

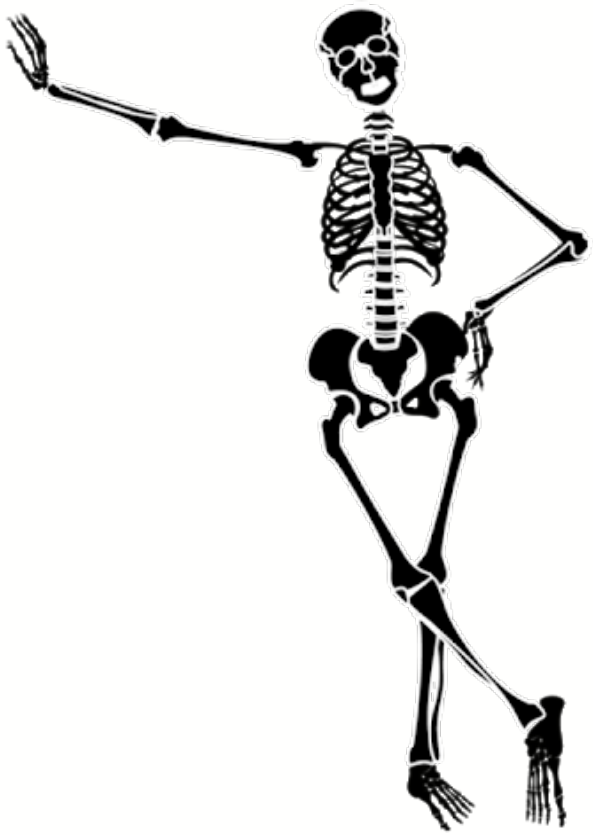
- Epidemiology
- Biology and genetics
- Risk factors
- Predisposition syndromes
- Clinical presentation
- Staging
- Prognostic factors



http://medcell.med.yale.edu/histology/bone_lab/osteosarcoma.php

Supportive Care at Diagnosis

- Pain control
- Occupational vs physical therapy
- Weight bearing precautions
- Education
- Interprofessional support



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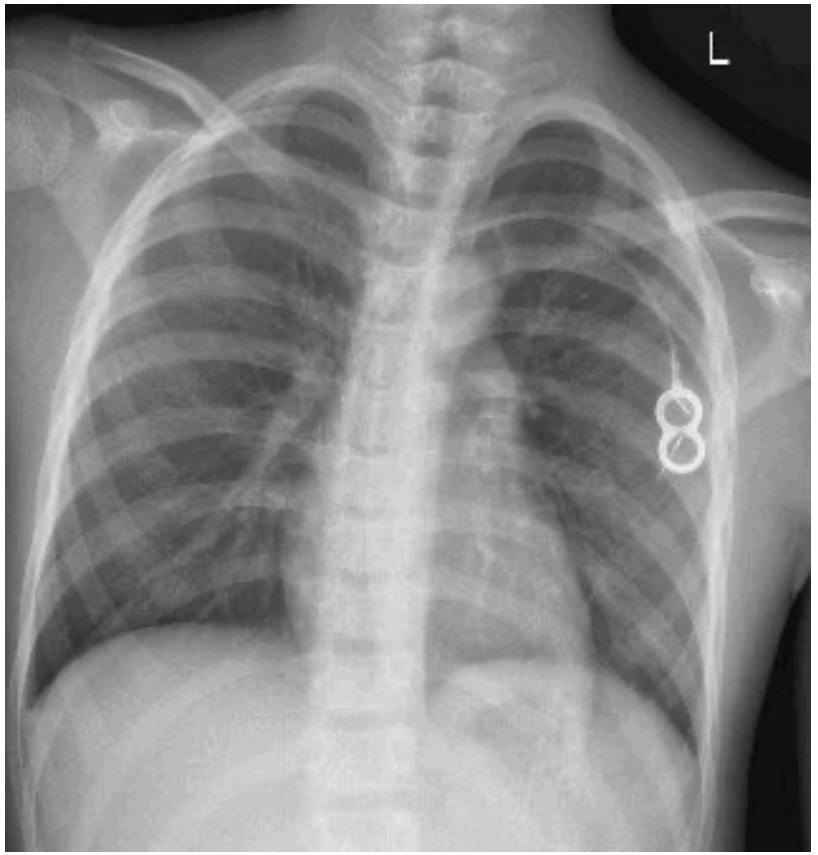
Plan

- Diagnostic + metastatic workup
- Baseline evaluations
- Central line placement
- Chemotherapy
- Surgery
- Monitoring after therapy



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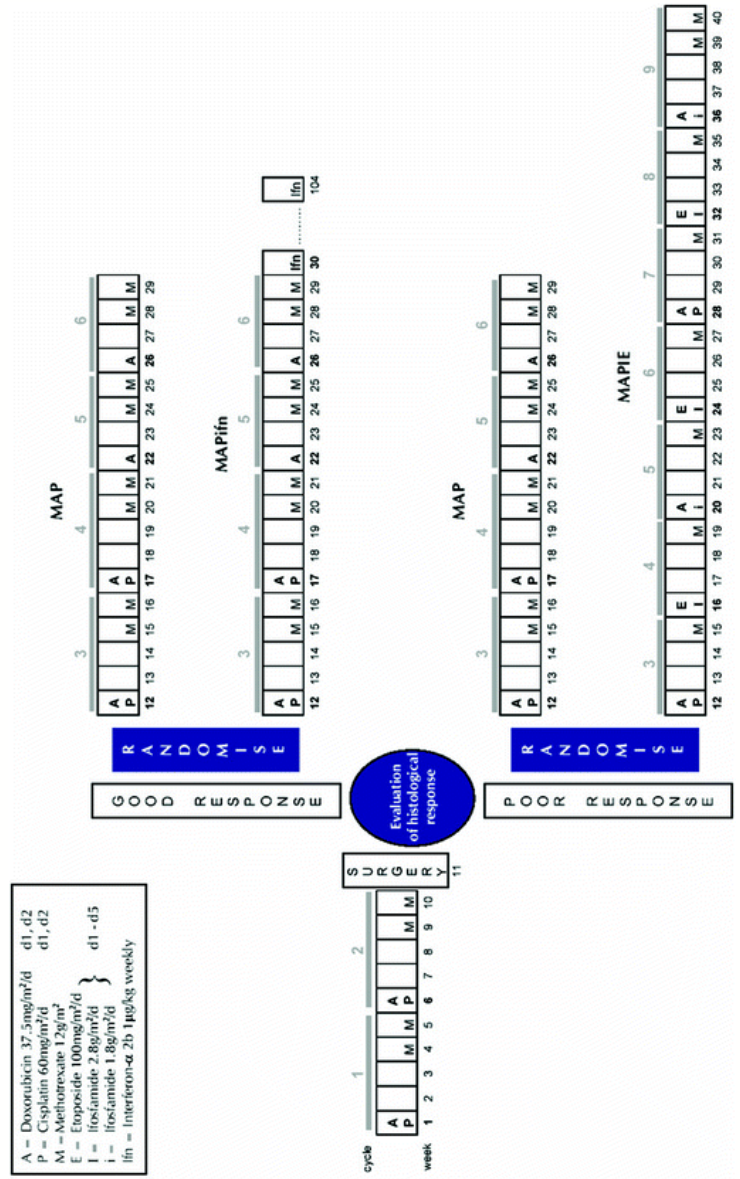
Chemotherapy

- Neoadjuvant and adjuvant therapy
- Standard of care = MAP
  - Methotrexate
  - Doxorubicin
  - Cisplatin
- Local control
- Notable toxicities
- Supportive care



https://link.springer.com/chapter/10.1007/978-1-4419-0284-9_18

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Role of Radiation in Osteosarcoma

- NOT a radiosensitive tumor
  - Requires high doses (7000-8000 cGy)
  - Limited by the normal structures in the field
- Considered if no surgical option
- Palliation

IN DEPTH: Surgery

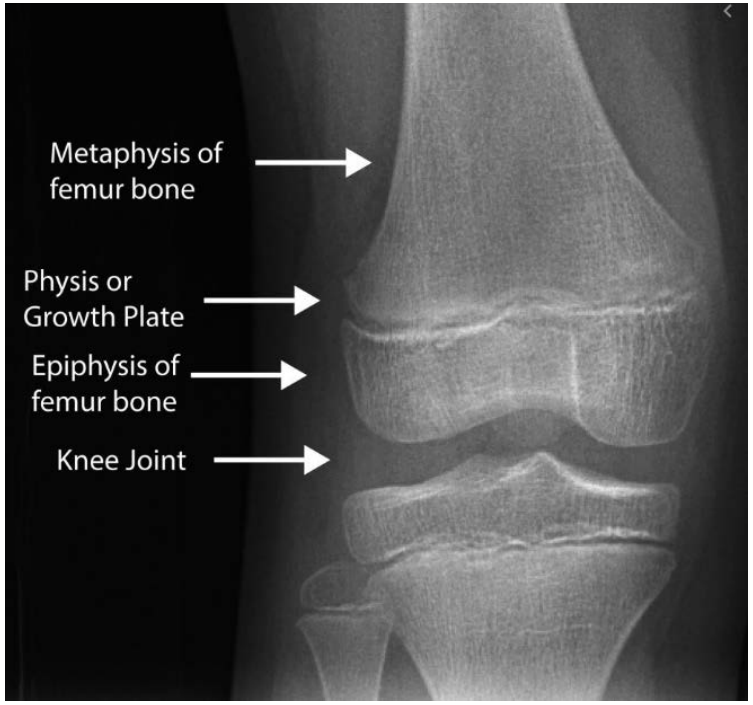
- Local control surgery = wide resection
- Goals
  - Negative margins
  - Maintain function
- Pre-op MRI

Surgery: Limb Salvage

Radical Resection

- Physeal-sparing
- Joint-sparing
- Limb-sparing

Huvos Classification	
Grade	Necrosis
I	Little or no effect, <60% tumor necrosis
II	Areas of viable tumor, 60-89% necrosis
III	Scattered foci, 90-99% tumor necrosis
IV	No histological evidence of viable tumor – 100%



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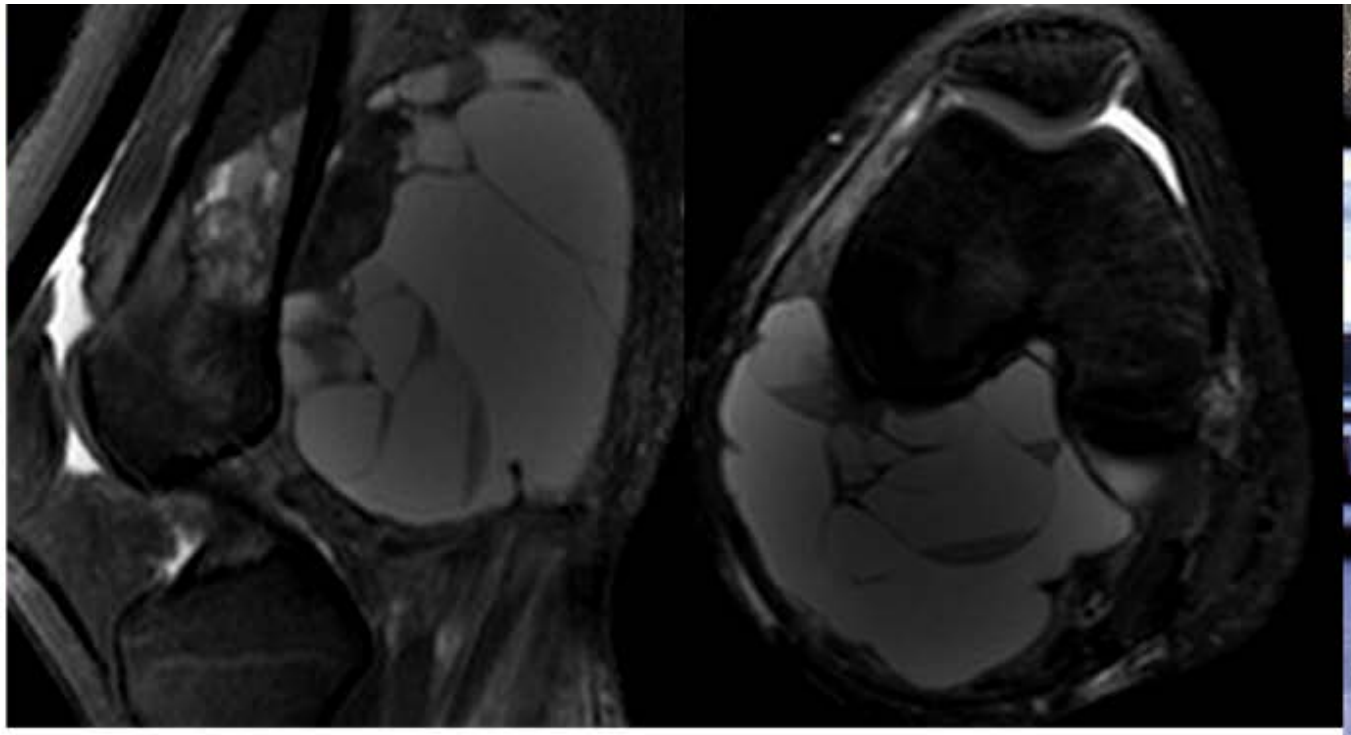
Local Control Surgery: Amputation



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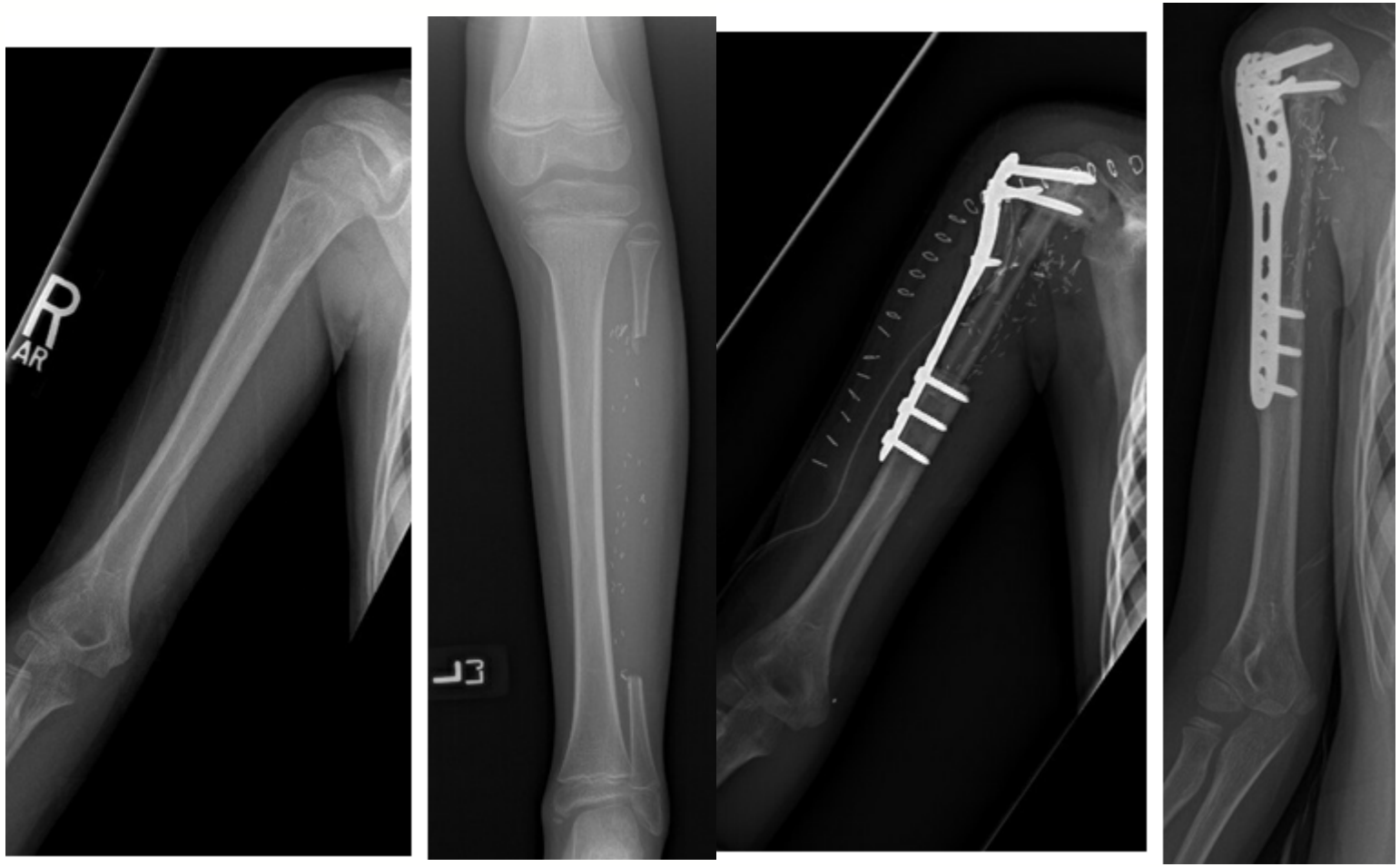
Limb Salvage: Vascularized Fibular Reconstruction

- Fibula = appendix
- Microsurgery
- Plate & screws
- Long term considerations



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Limb Salvage: Endoprosthetic Reconstruction

- Joint replacement
- Static vs Growing
- Recovery to walk
- Metal and plastic < bone
- Activity level

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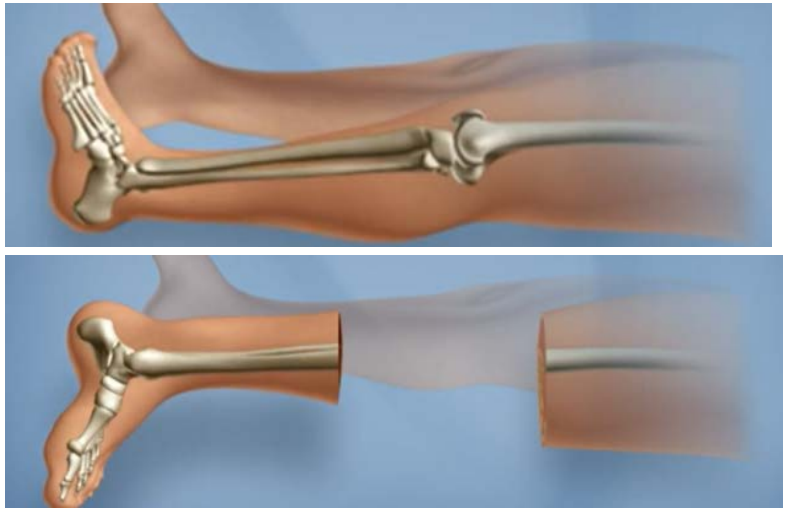
Local Control Surgery: Van Nes Rotationplasty

- "Almost" Limb Salvage, "Functional Amputation"
- Full activities
- Often will be one and done
- For distal femur/proximal tibial tumors



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Case Study #2: Surgery

- Limb salvage
  - Endoprosthetic reconstruction
  - 100% tumor necrosis, negative margins
- OT for shoulder rehabilitation
- Is now >3 years post-surgery
- Doing well!

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Local Control Surgery: Postoperative Management

- Wound healing- 2-3 weeks post-op before resuming chemo
- Monitor @ 2 weeks, 6 weeks, 3 months; then every 3-4 months x 3 years, every 6 months year 4 & 5
- Possible complications
  - Infection
  - Non-union
  - Fracture
  - Limb length discrepancy

Local Control Surgery: Take Home Points

- Not “one size fits all” approach
- Primary goal = wide resection
- Chemo is a double edged sword
- Kids are resilient!

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Role of the Nurse Practitioner

- Education
  - Patient and family
  - Nursing
  - Medical team
- Advocacy
- Clinical management, monitoring, and supportive care
- Research

Future Directions

- Standard of care
- Phase 2 clinical trials
- Future breakthroughs
  - Chemotherapy
  - Immunotherapy
  - Personalized medicine
  - Imaging
  - Radiation
  - Surgical techniques

https://nfcr.org/blog/sarcoma-awareness-month

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Questions?

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