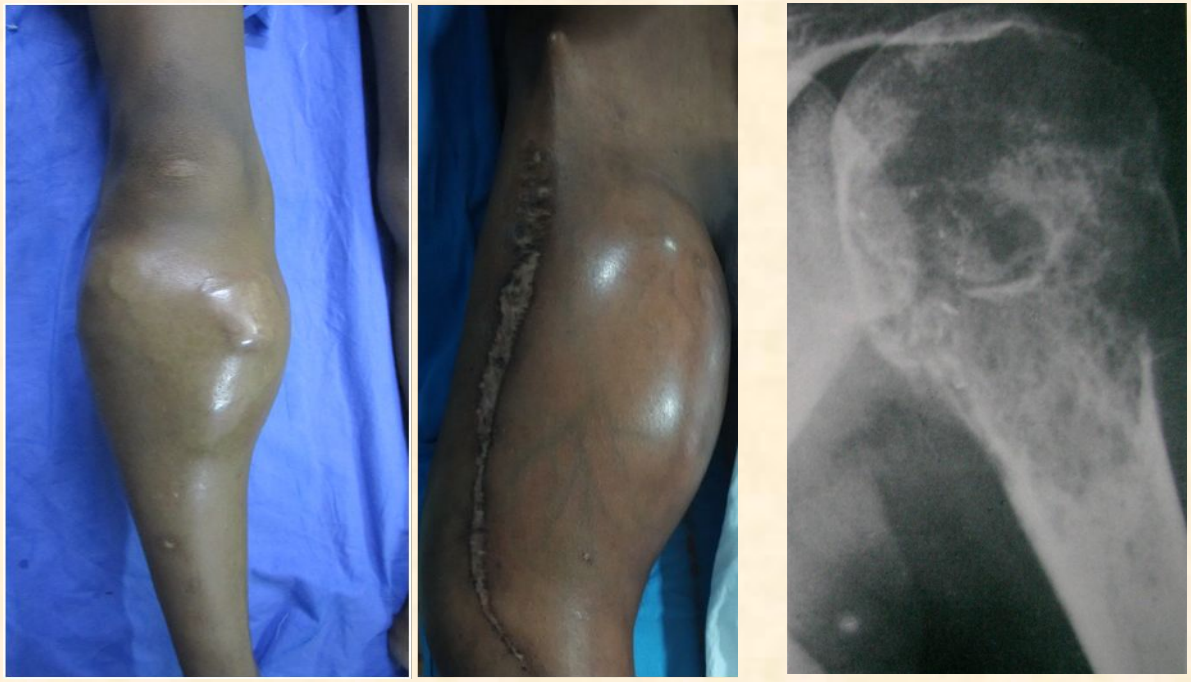
Malignant Bone Tumors



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WHO Classification

a) **Bone forming tumours**
- Benign: - Osteoid osteoma, osteoma
  - Osteoblastoma
- Indeterminate - Aggressive osteoblastoma
- Malignant - Osteosarcoma conventional, variants

b) **Cartilage forming tumours**
- Benign: - Osteochondroma (exostosis)
  - Enchondroma (chondroma)
  - Chondromyxoid fibroma
  - Chondroblastoma
- Malignant: - Chondrosarcoma

c) **Giant cell tumours (GCT)**

WHO Classification

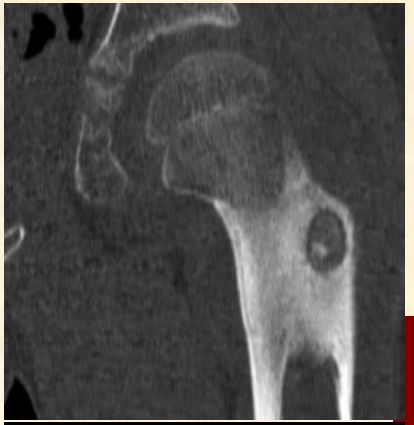
d) **Marrow tumours**
- Malignant:
  - Ewing's sarcoma
  - Plasma cell tumour
  - Multiple myeloma
  - Lymphoma

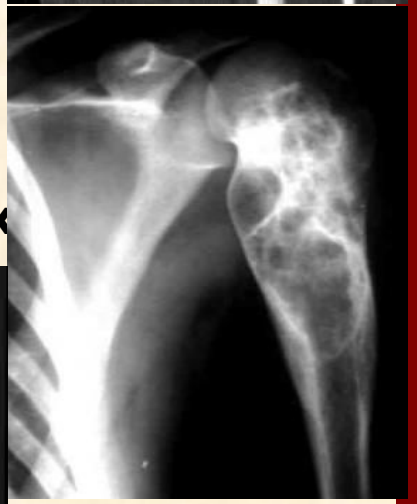
e) **Vascular tumours**
- Benign:
  - Haemangioma
  - Glomangioma
- Malignant:
  - Angiosarcoma

f) **Others**
- Benign:
  - Neurilemmoma
  - Neurofibroma
- Malignant:
  - Malignant fibrous histiocytoma
  - Liposarcoma
  - Undifferentiated sarcoma
  - Adamantinoma

g) **Tumour-like lesions**
- Bone cysts – simple or aneurysmal
- Fibrous dysplasia – mono or polyostotic
- Reparative giant cell granuloma (e.g. epulis)
- Fibrous cortical defect
- Eosinophilic granuloma

Enneking's staging for Benign Tumors





Stage 1; Latent

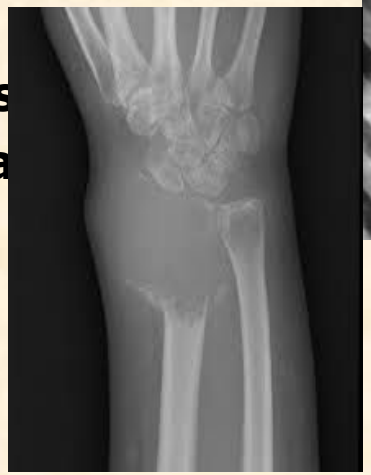
- Well defined margin
- Grows slowly and then stops
- Heals spontaneously e.g. osteoid osteoma

Stage 2; Active

- Progressive growth limited by natural barriers
- Well defined margin but may expand thinning cortex

Stage 3; aggressive

- Growth not limited by natural barriers
- Metastasis present in 5% of these pta



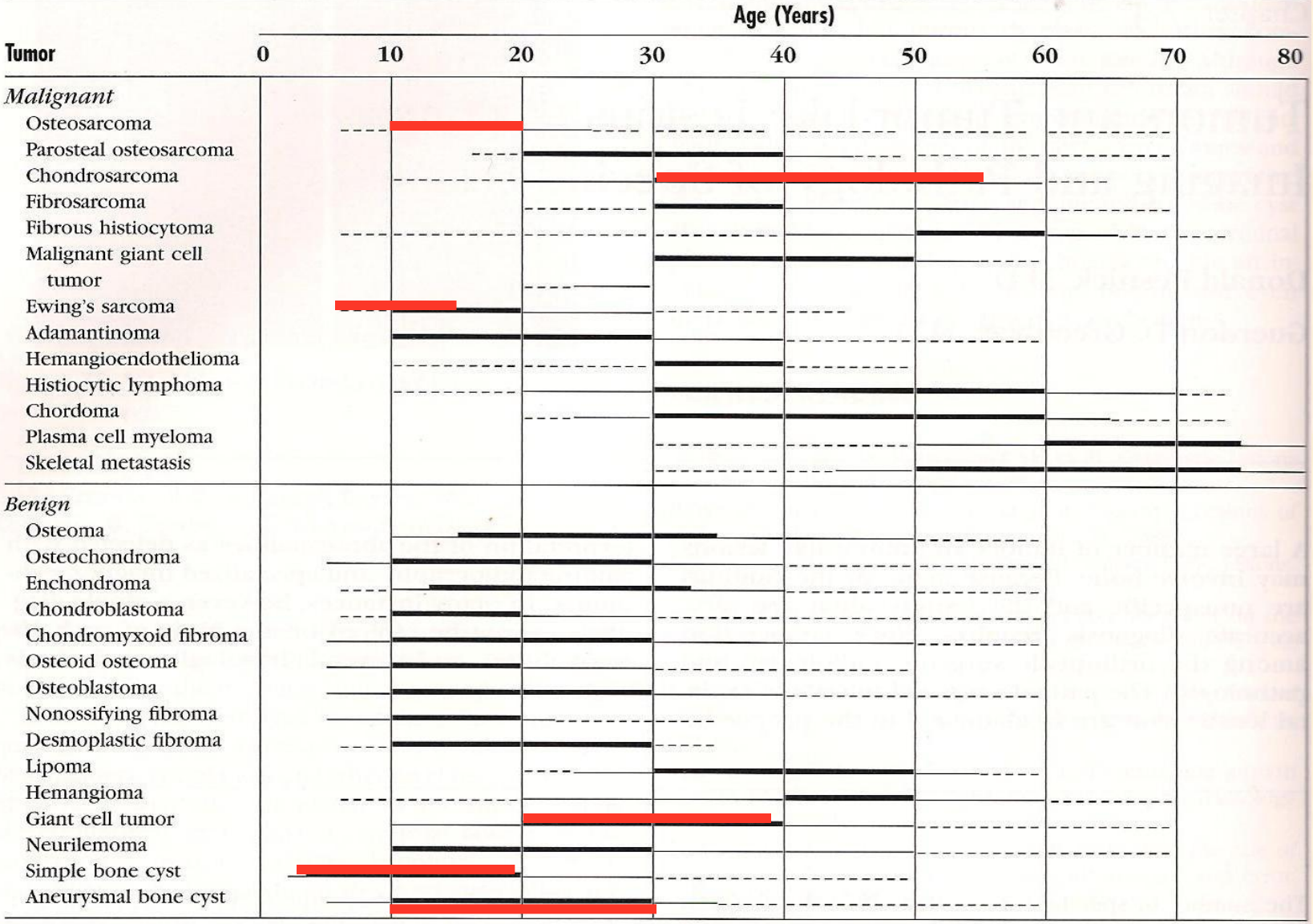
Enneking's staging for malignant bone tumors

STAGE	GRADE	SITE	METASTASES
IA	Low	Intracompartmental	None
IB	Low	Extracompartmental	None
IIA	High	Intracompartmental	None
IIB	High	Extracompartmental	None
III	Any	Any	Regional or distant metastases

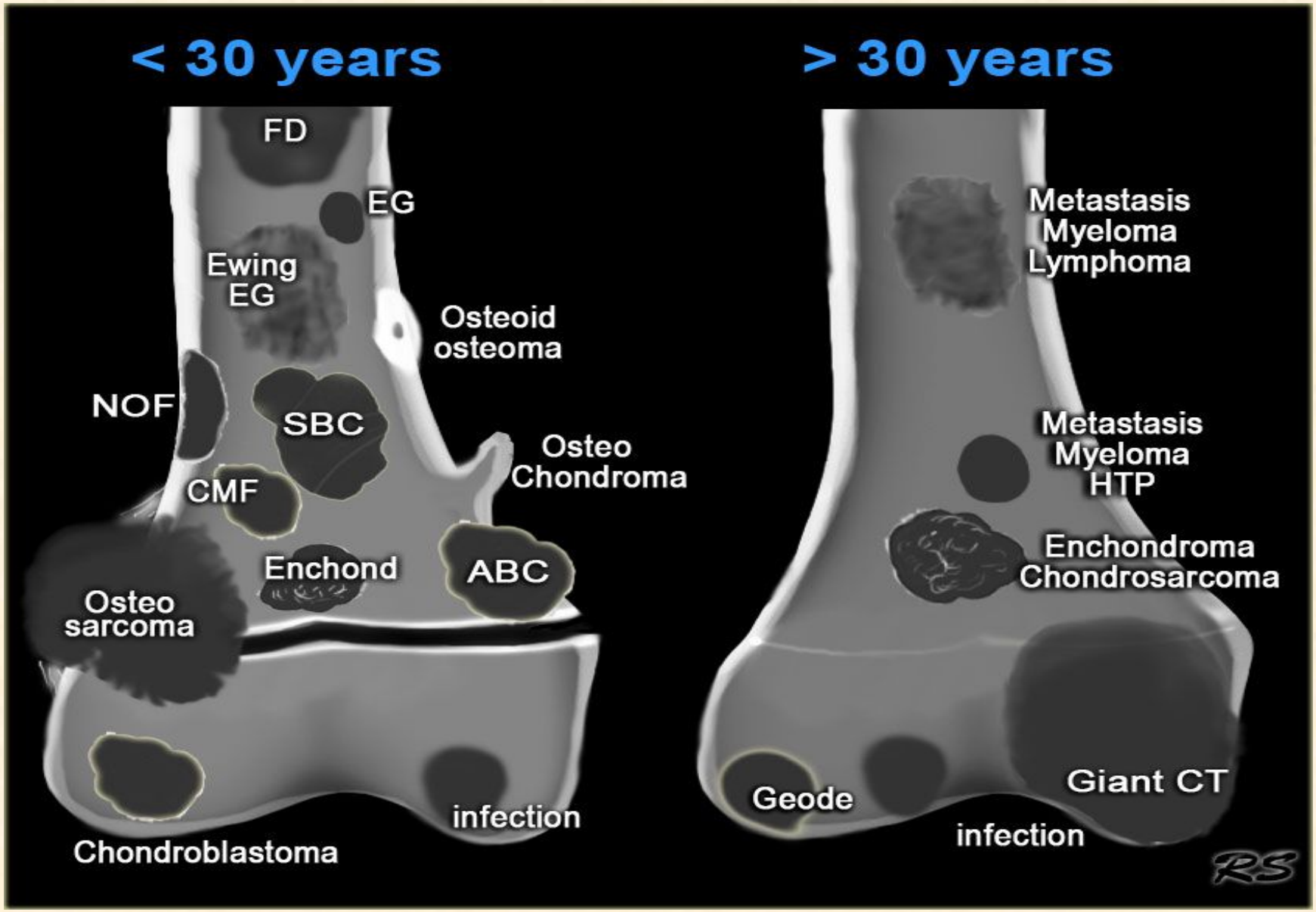
AJCC staging for Malignant Bone tumors

STAGE	GRADE	SIZE	METASTASES
I-A	Low	≤8 cm	None
I-B	Low	>8 cm	None
II-A	High	≤8 cm	None
II-B	High	>8 cm	None
III	Any	Any	Skip metastasis
IV-A	Any	Any	Pulmonary metastases
IV-B	Any	Any	Nonpulmonary metastases

TABLE 74–1. TUMORS AND TUMOR-LIKE LESIONS: TYPICAL AGES OF PATIENTS*



*Bold solid line, most typical age; regular solid line, less typical age; interrupted line, even less typical age.


< 30 years
> 30 years
FD
EG
Ewing
EG
Osteoid
osteoma
NOF
SBC
Osteo
Chondroma
CMF
Enchond
ABC
Osteo
sarcoma
infection
Chondroblastoma
Metastasis
Myeloma
Lymphoma
Metastasis
Myeloma
HTP
Enchondroma
Chondrosarcoma
Giant CT
Geode
infection
RS

J Am Acad Orthop Surg. 2011 Nov;19(11):678-89.

Osteoid osteoma and osteoblastoma.

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Author information

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Abstract

Osteoid osteoma and osteoblastoma are commonly seen benign osteogenic bone neoplasms. Both tumors are typically seen in the second decade of life, with a notable predilection in males. Histologically, these tumors resemble each other, with characteristically increased osteoid tissue formation surrounded by vascular fibrous stroma and perilesional sclerosis. However, osteoblastomas are larger than osteoid osteomas, and they exhibit greater osteoid production and vascularity. Clinically, osteoid osteoma most commonly occurs in the long bones (eg, femur, tibia). The lesions cause night pain that is relieved with nonsteroidal anti-inflammatory drugs (NSAIDs). Osteoblastoma is most frequently located in the axial skeleton, and the pain is usually not worse at night and is less likely to be relieved with NSAIDs. Osteoblastoma can be locally aggressive; osteoid osteoma lacks growth potential. Osteoid osteoma may be managed nonsurgically with NSAIDs. When surgery is required, minimally invasive methods (eg, CT-guided excision, radiofrequency ablation) are preferred. Osteoblastoma has a higher rate of recurrence than does osteoid osteoma, and patients must be treated surgically with intralesional curettage or en bloc resection.

Chondroblastoma

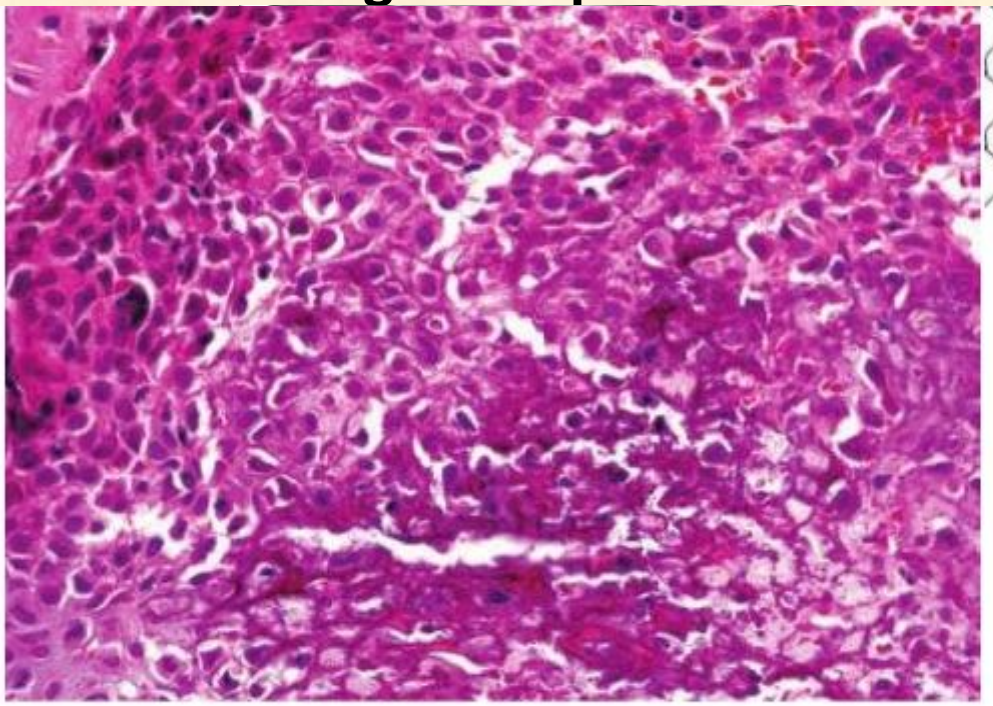
Teenagers – before obliteration of growth plates. EPIPHYSISEAL

Male : Female 2 : 1

Treatment

Curettage + BG

RFA for smaller lesions



- The calcification in chondroblastoma is seen between individual tumor cells-"chicken-wire"

Giant Cell Tumor

Incidence is 8.6 % of all bone tumors

Incidence in 3rd (commonest) & 4th decades

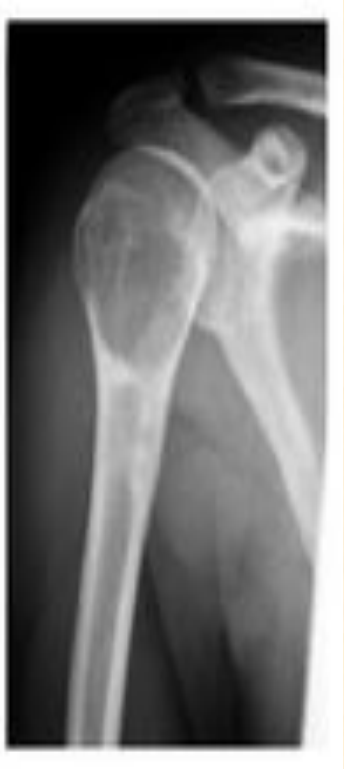
(rare in skeletally immature patients)

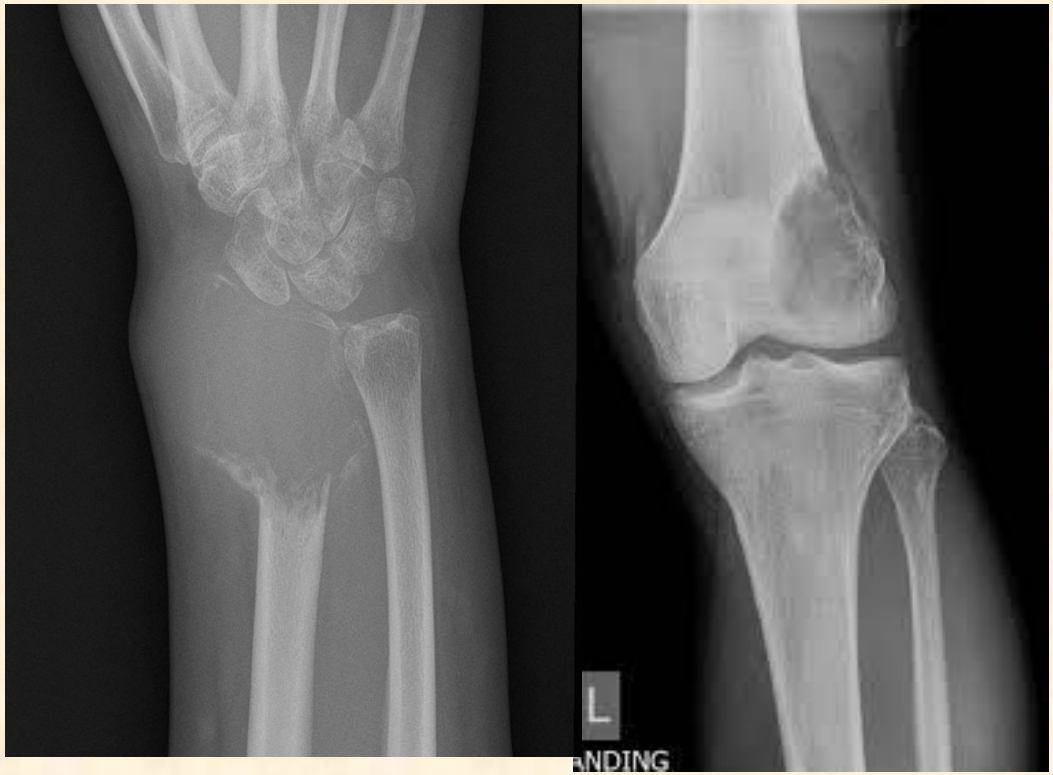
Female > male(1.5 : 1)

Commonly occurs in Distal femur, Proximal tibia, Distal radius & Proximal humerus.

Most are stage II/III at presentation.

GCT radiology



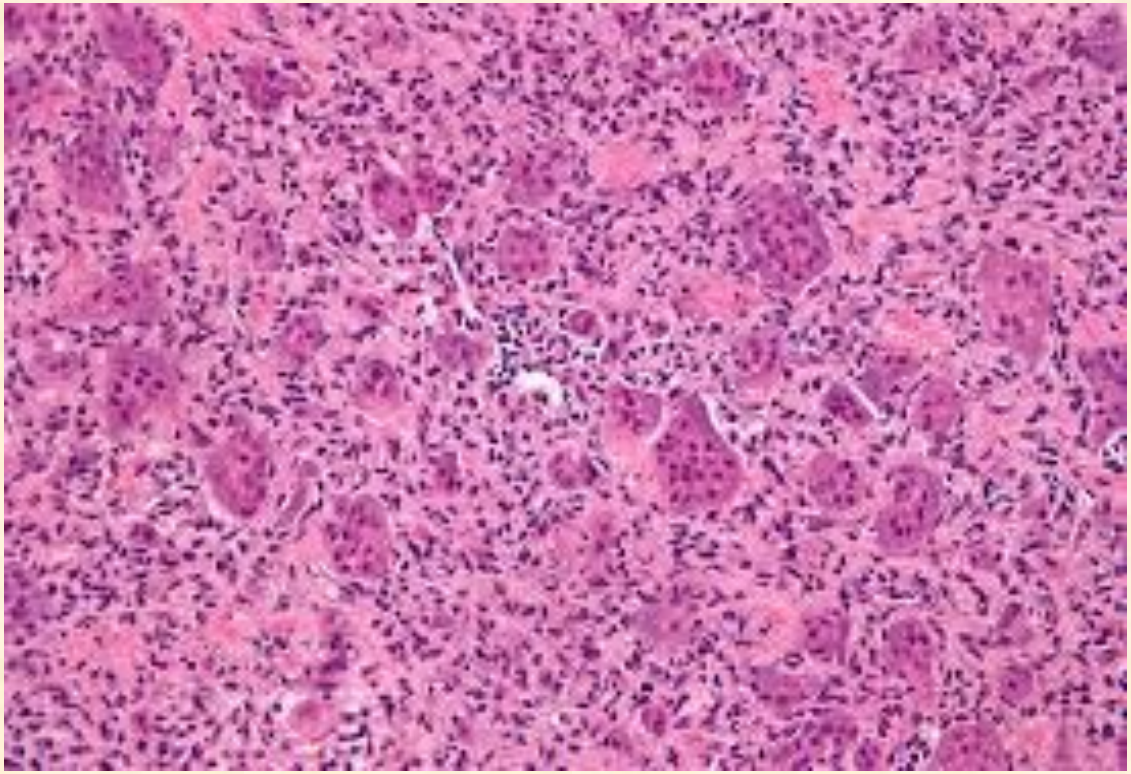


ECCENTRIC

EPIPHYSEAL

EPANSILE **L**YTIC

GCT Histopathology



Multinucleated Giant Cells , Mononuclear Stromal spindle Cells

Treatment

Stage I, II

Extended Curettage + BG

Adjuvants
High speed Burr
Phenol
H_2O_2
Liquid N2
Cement (PMMA)

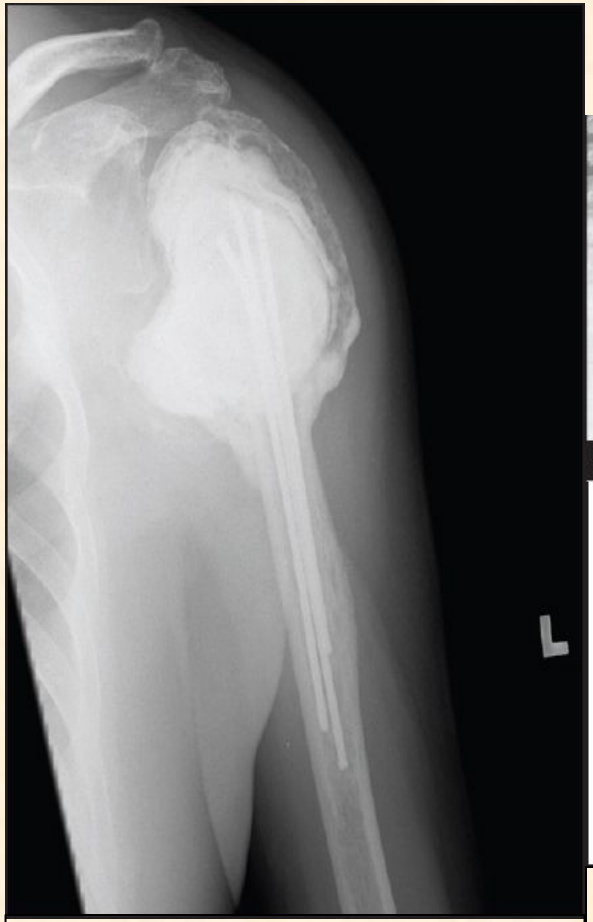
Downgrading

Bisphosphonates (Alendronate, Zolindronic Acid, Pamidronate)

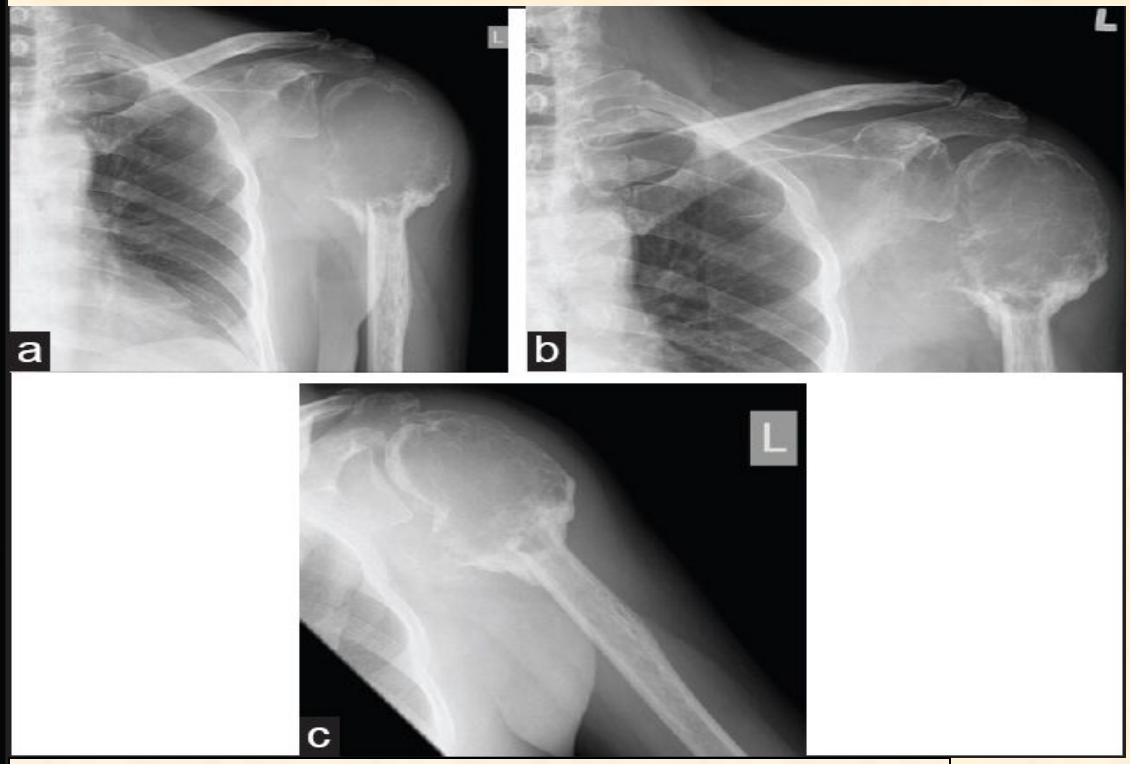
Denusumab

Stage III

Wide Excision



2 years post op
Curettage + Cementation
No recurrence



Response to Denusumab, 3 Injections at interval of 3 weeks

Osteochondroma

Osteochondroma is a bony exostosis projecting from the external surface of a bone

Hyaline lined cartilaginous cap

The cortex and spongiosa of the lesion merge with that of the host bone

Most common skeletal growth/tumor

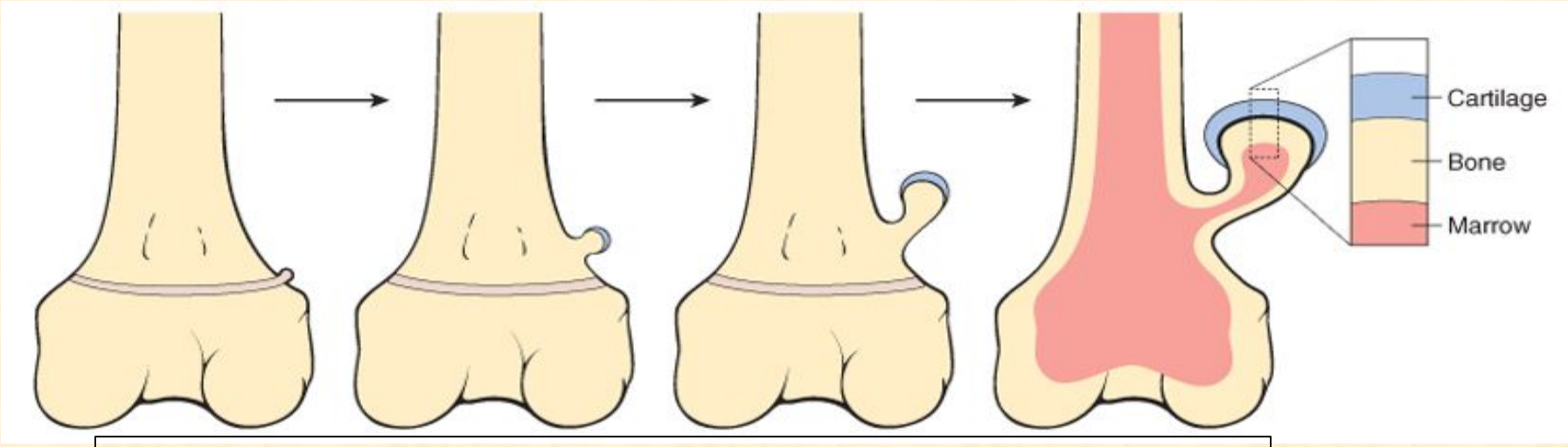
Approximate incidence is 50% of all benign bone tumors

Male : Female ratio 2:1

Most are encountered in childhood and adolescence

Widespread ostechondromas are associated with a positive familial history, and the condition is known as Heriditary Multiple Exostosis

Osteochondroma

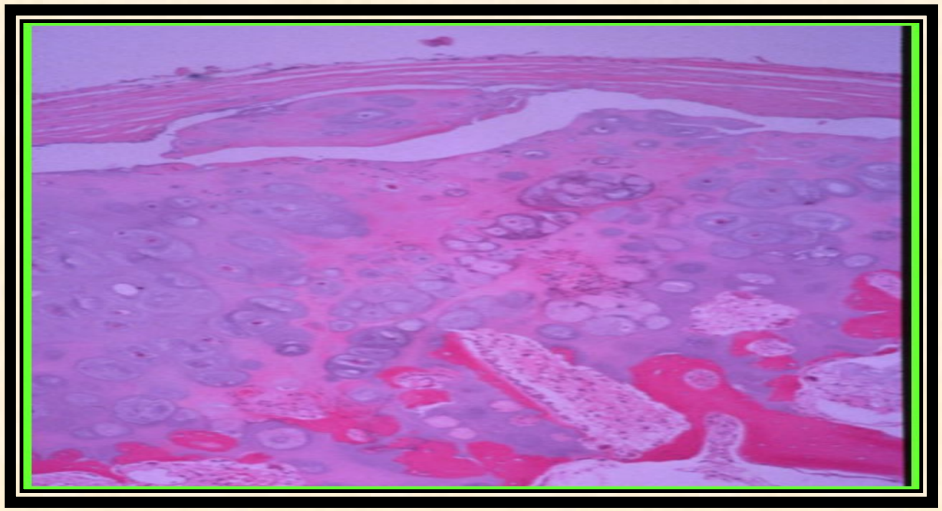


Osteochondroma, beginning with an outgrowth from the epiphyseal cartilage

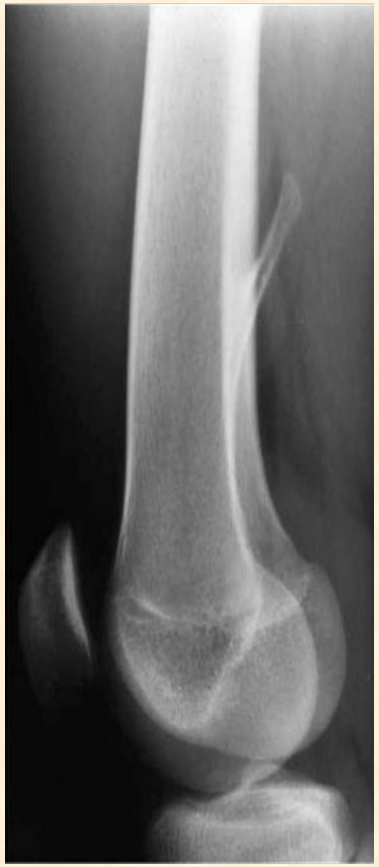
Covered by thin layer of periosteum

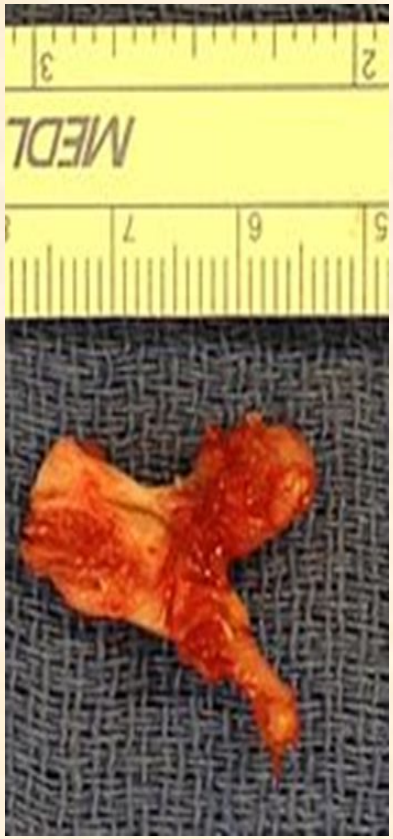
Binucleate chondrocytes in lacunae

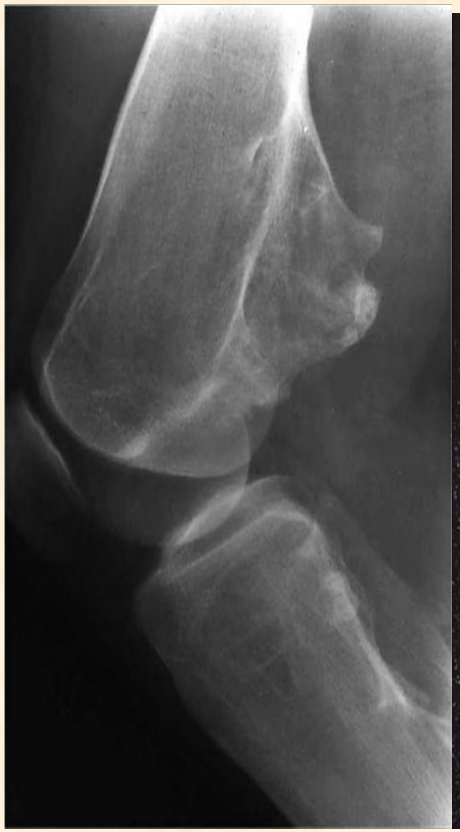
Contains hyaline cartilage, bony tissue and normal bone marrow particle

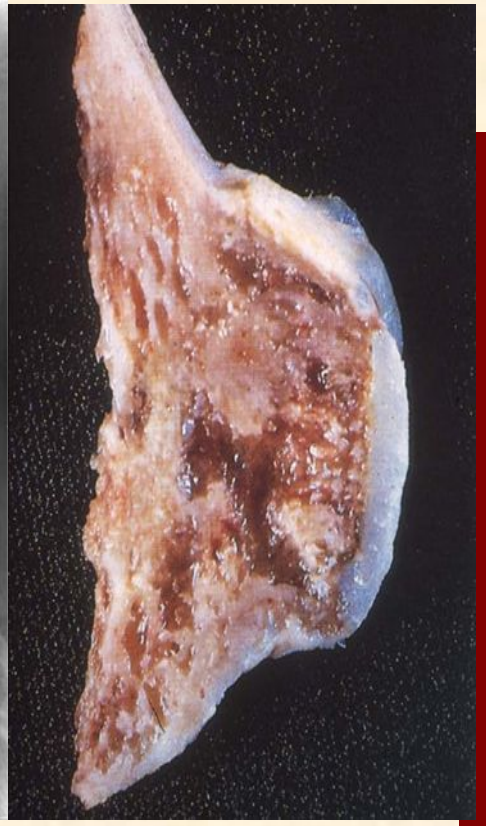


Pedunculated vs Sessile









Causes of Pain in Osteochondroma

1. Mechanical irritation of muscles, ligaments or tendons during growth
2. Nerve compression and irritation of nerve
3. Fracture through stalk
4. Bursitis Exostica
5. Malignant transformation into chondrosarcoma
6. Associated Pseudoaneurysm

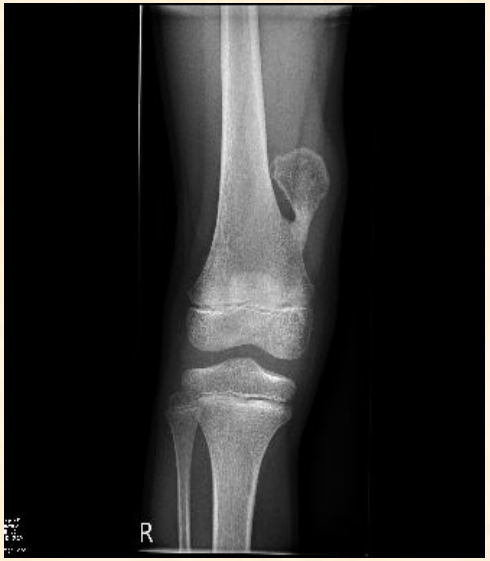
X ray findings

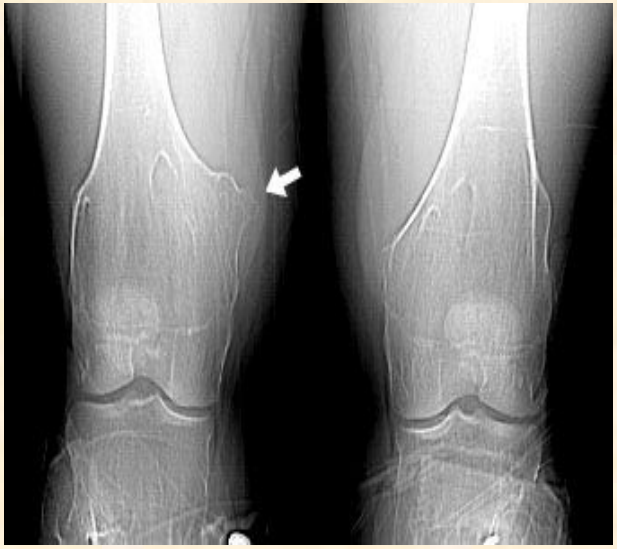
Bony protuberance on a pedunculated stalk or a sessile base forming ‘trumpet shaped deformity’, pointing away from joint

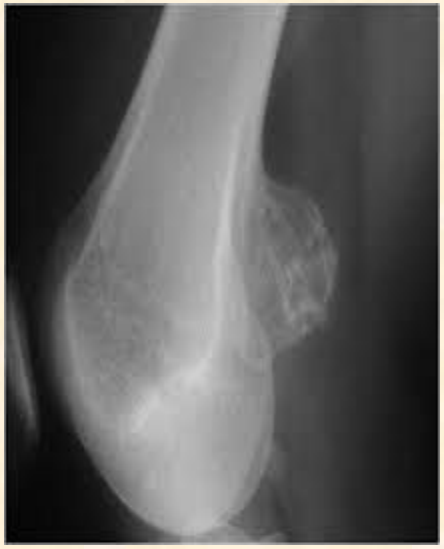
Metaphyseal or diaphyseal location , widening of metaphysis from where it arises

The trabecular pattern of lesion blends with that of intramedullary host bone

Cortex of lesion flares into cortex of host bone







When to suspect malignant Transformation?

Osteochondroma usually ceases to grow after skeletal maturity.
Continued growth & sudden onset pain after skeletal maturity, point towards malignant transformation.

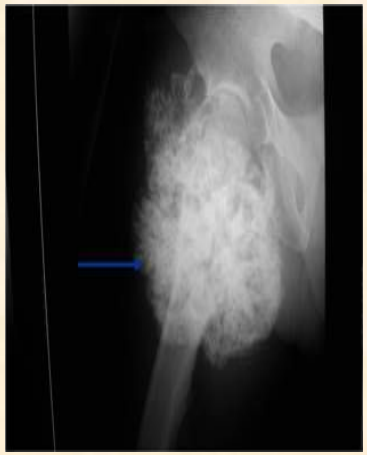
Radiological features of malignant transformation

1. Stippled calcification and variable mineralization of cartilage cap

2. Soft tissue mass

3. Bony destruction

4. Cartilage cap thickness > 1.5 cm on USG or MRI (controversy exists , using 2 cm cartilage cap as a cutoff on MRI is used for differentiating osteochondroma from secondary chondrosarcoma)



Treatment

Asymptomatic lesions should be observed periodically

Indications for excision

1. Pressure symptoms on adjacent nerve /vessels
2. Mechanical block in joint ROM
3. Fracture of pedunculated stalk
4. Bursitis
5. Malignant transformation

Cosmesis: not an absolute indication but most common reason for excision

Enblock Extraperiosteal resection (without elevating periosteum and perichondrium) should be attempted.

Care should be taken for not damaging the physis and not leaving cartilaginous cap or perichondrium in resection bed to avoid recurrence.

Exostosis-host bone junction should be removed

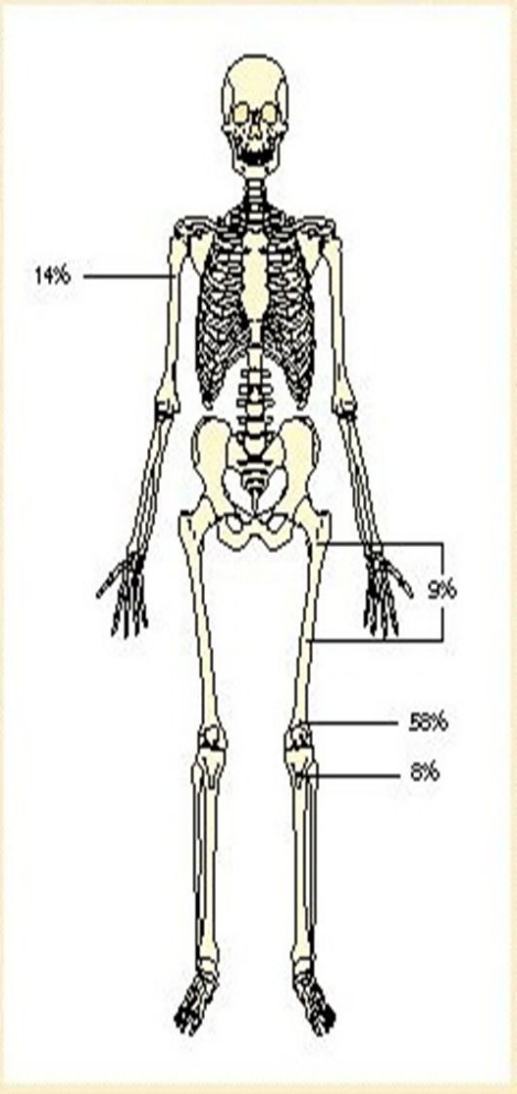
Osteosarcoma

Highly malignant tumor of mesenchymal

Spindle shaped cells that produce osteoid

Most common primary malignant bone t

Age at presentation: 10-20



PRIMARY OSTEOSARCOMA: Variants

- Conventional /classic osteosarcoma (high grade, intra medullar y)
- Low-grade intramedullary osteosarcoma
- Parosteal osteosarcoma
- Periosteal osteosarcoma
- High-grade surface osteosarcoma
- Telangiectatic osteosarcoma
- Small cell osteosarcoma

SECONDARY OSTEOSARCOMAS

More common in >50 years of age

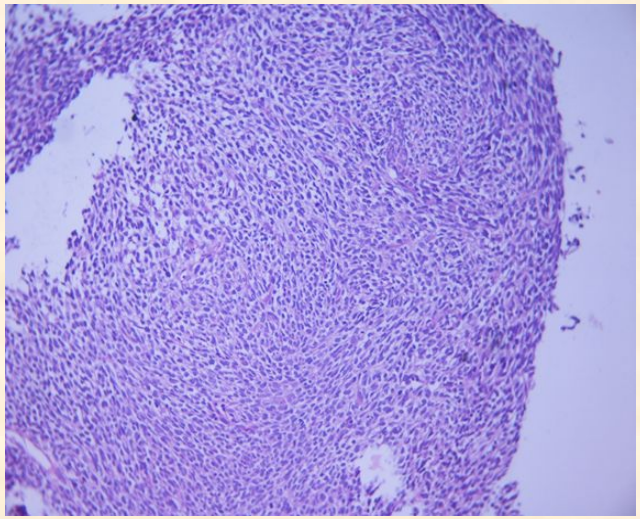
- Paget disease
- Previous radiation treatment
- Fibrous Dysplasia

Ewing's sarcoma

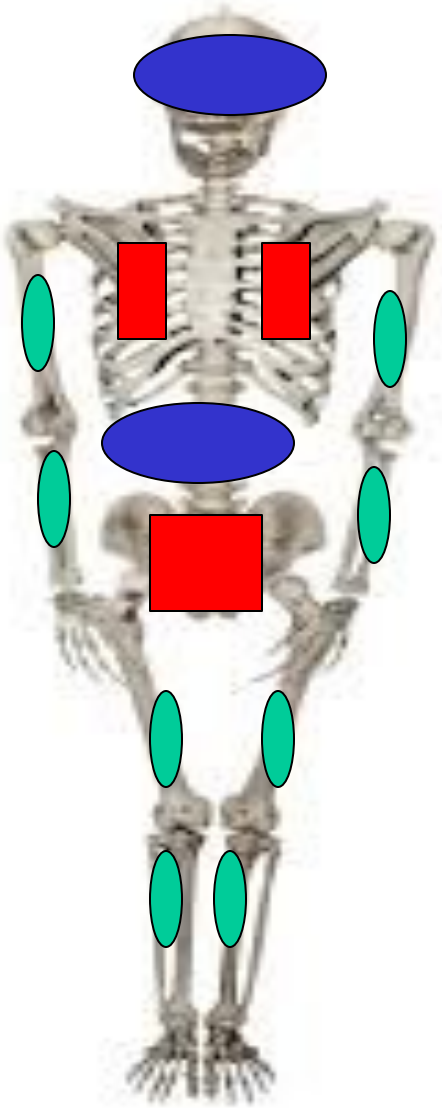
Small round cell tumor with neuroectodermal differentiation

5–10% of primary bone tumors

More common than osteosarcoma in children less than 10 years of age



Site of Occurence



Diaphysis/Metadiaphysis of long bones

Pelvic bones/ Ribs

Vertebrae/skull/small bones

Soft tissue Ewing's Sarcoma

Chondrosarcoma

Chondrosarcoma is composed of transformed cells that produce cartilage

It is resistant to chemotherapy and radiotherapy

Age :30-60

Axial skeleton>Appendicluar skeleton

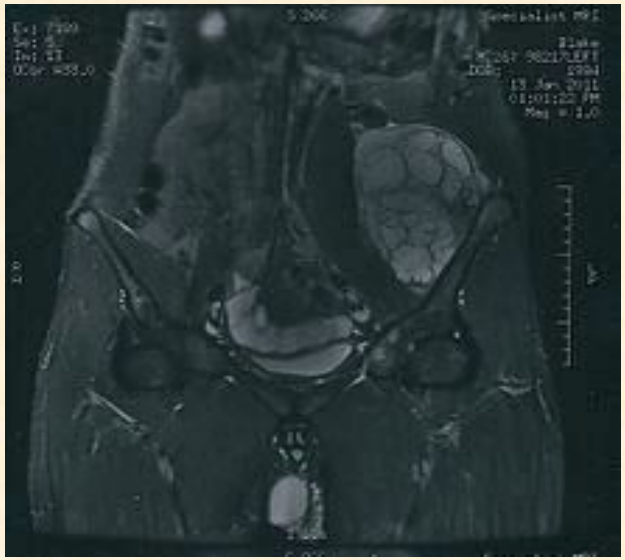
<u>Secondary chondrosarcoma</u>

Enchondroma

Osteochondroma

Maffuci syndrome

Ollier disease



Clinical Presentation

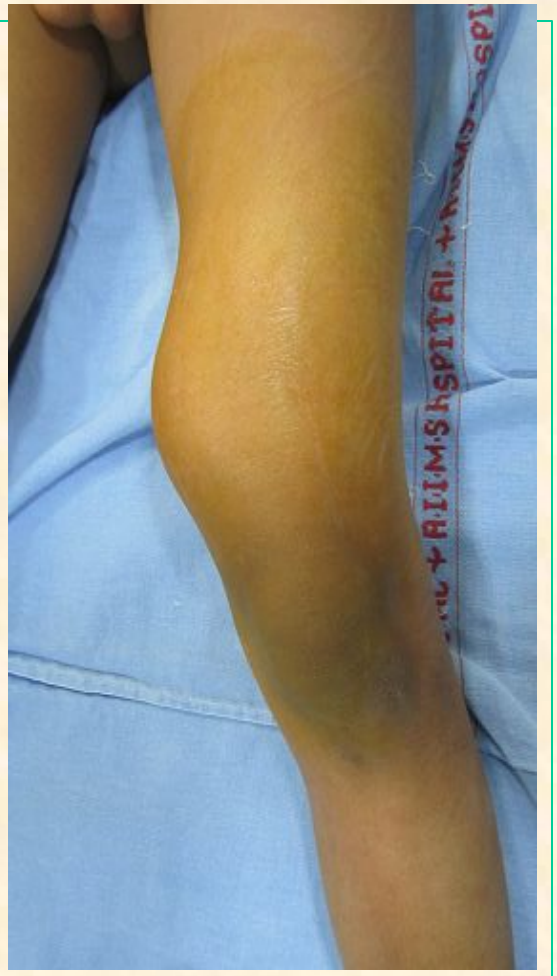
Malignant swelling

History (5 points)

1. **Pain → Swelling**
2. **Duration: weeks-Months**

(Rapid Progession)

3. **Activity: Loss of function**
4. **Symptoms of metastasis**
5. **History of chemotherapy**



Clinical Presentation

General Examination (2 points)

1. **Cachexia (**ill built, poorly nourished,anemia, hair loss, nail changes)

2. **Findings suggesting metastasis**
   (PLEURAL EFFUSION)

Clinical Presentation

INSPECTION (5 points)

1. **Stretched shiny skin**
2. **Dilated veins**
3. **Wasting of limb**
4. **Irregular Surface**
5. **Ulceration/Fungation**

Clinical Presentation

Palpation (5 points)

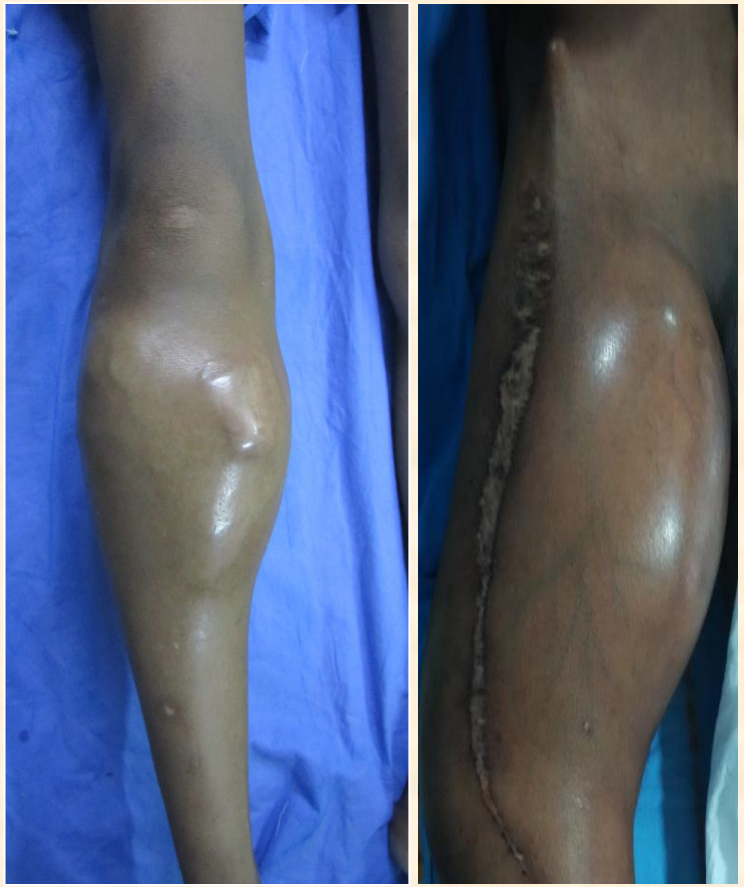
1 Raised temperature

2. Significant Tenderness

3. Ill defined Margins

4. Variable consistency

5. Non-pinchable skin

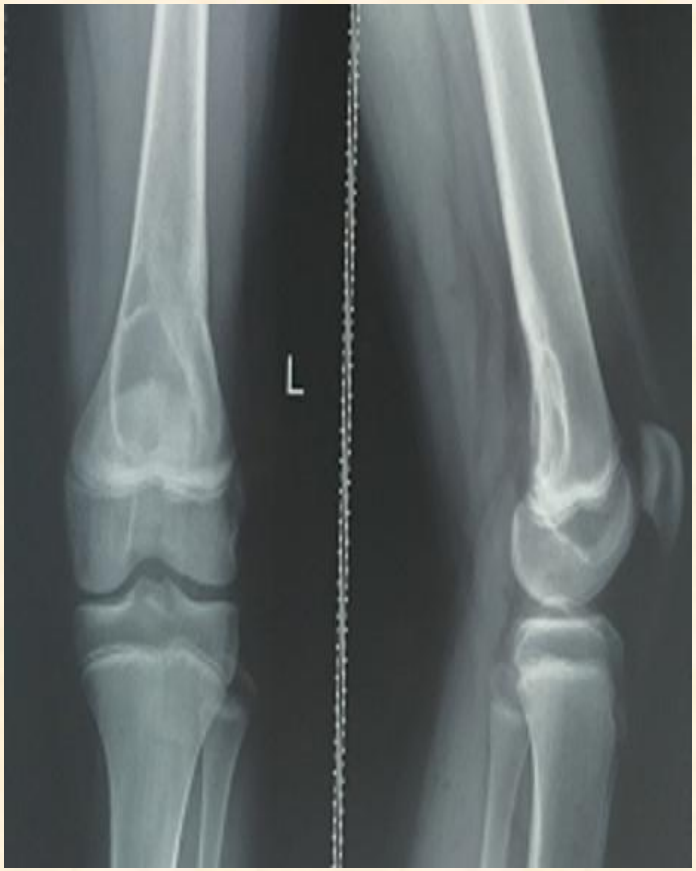


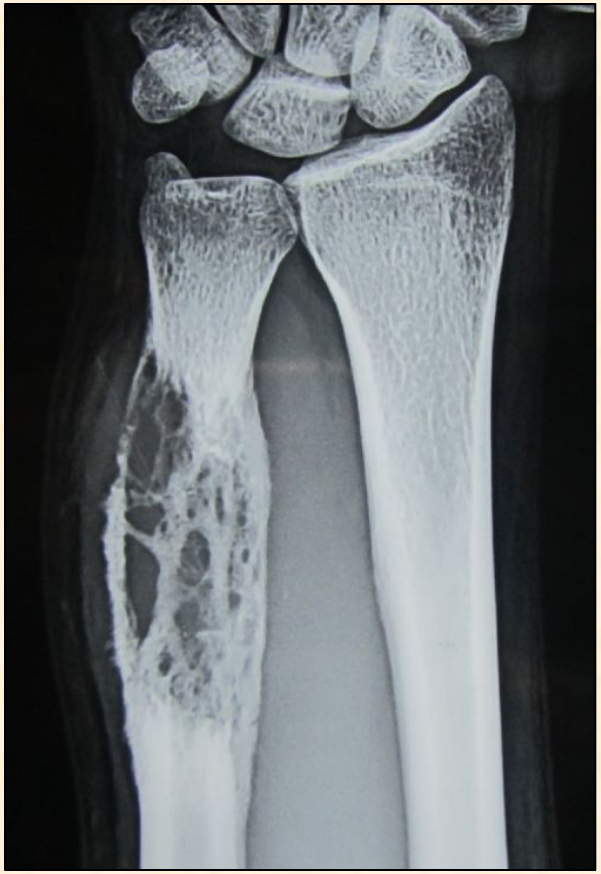
2 others:

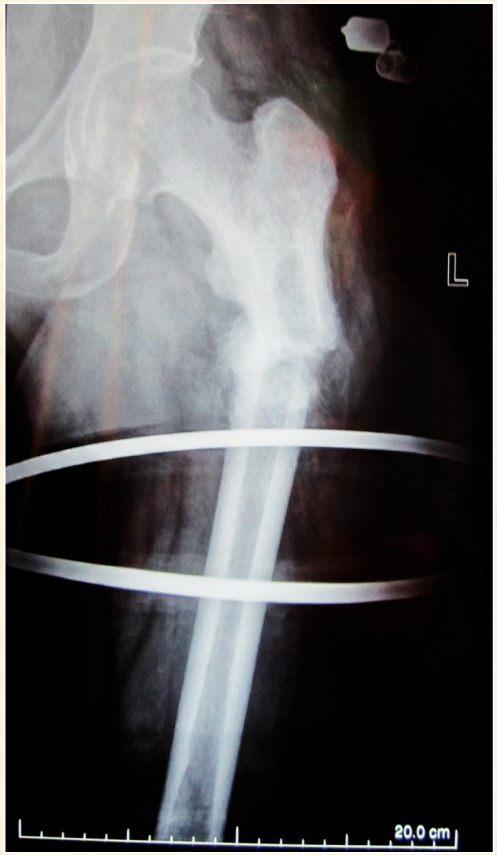
Restricted ROM

DNVD

How to Investigate???



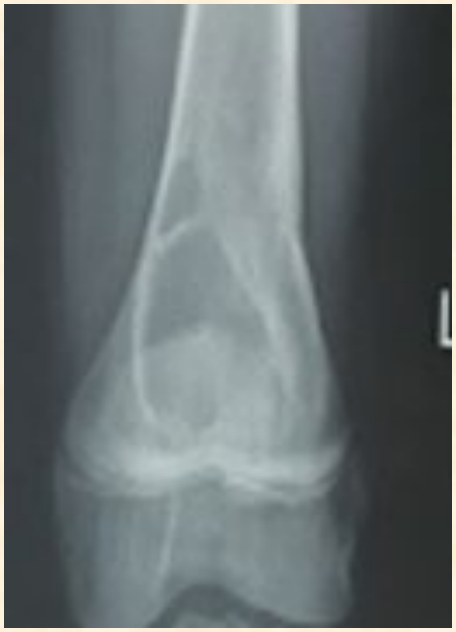




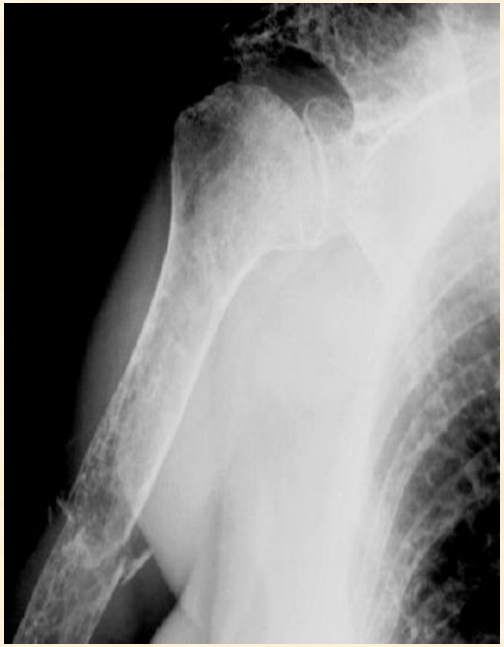
What on x-ray ??

Site of lesion

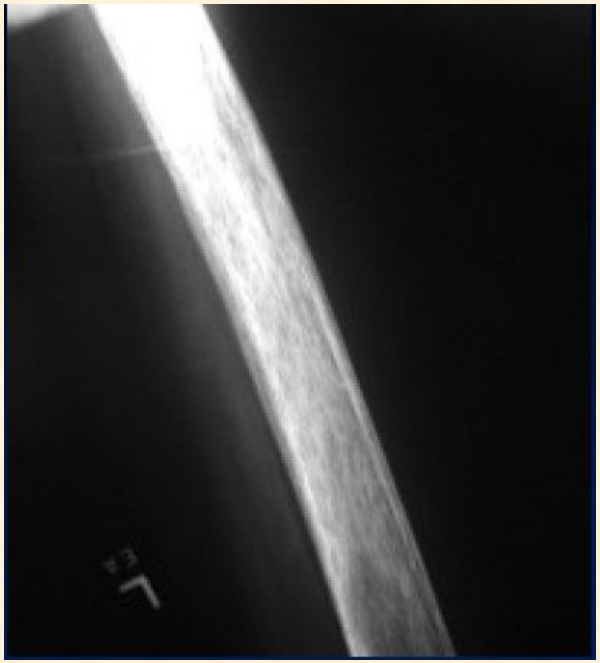
Pattern of destruction



Geographic



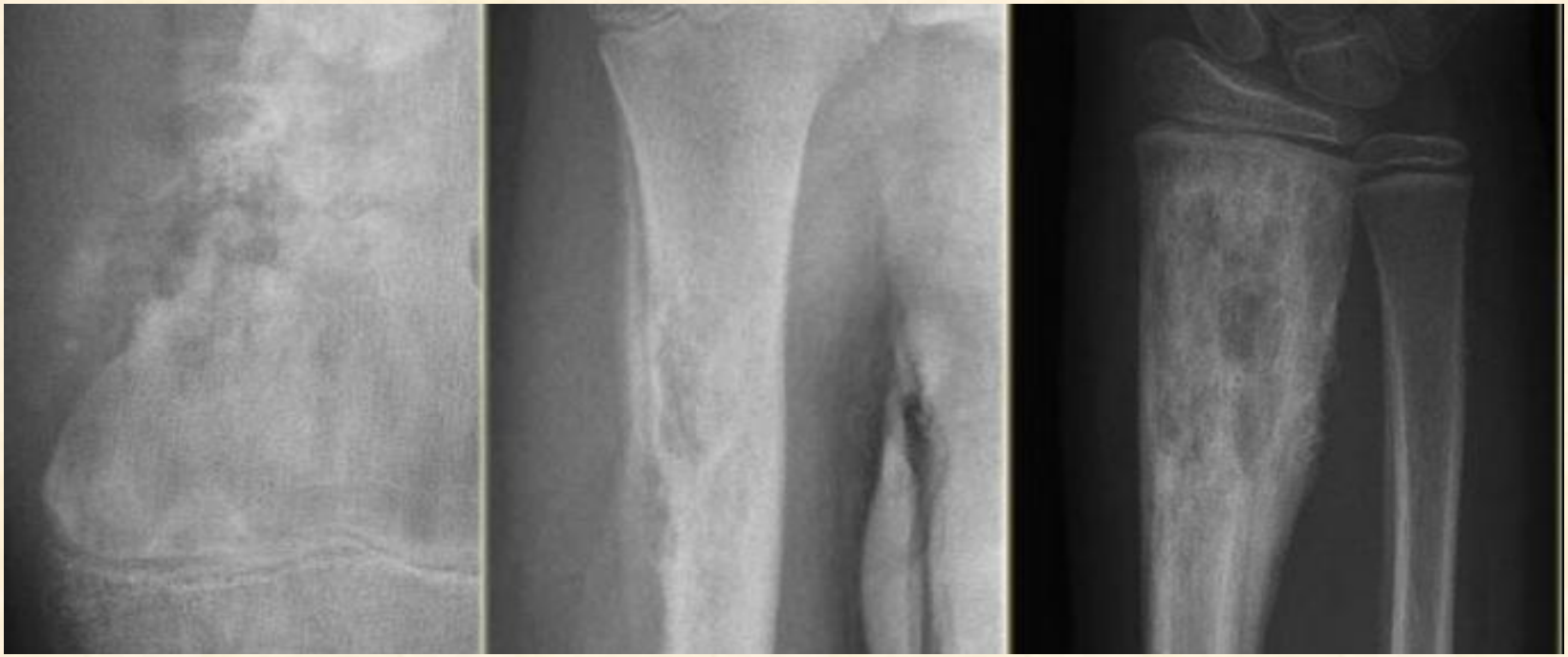
Moth Eaten



Permeative

What on x-ray ??

Zone of transition



Wide zone of transition

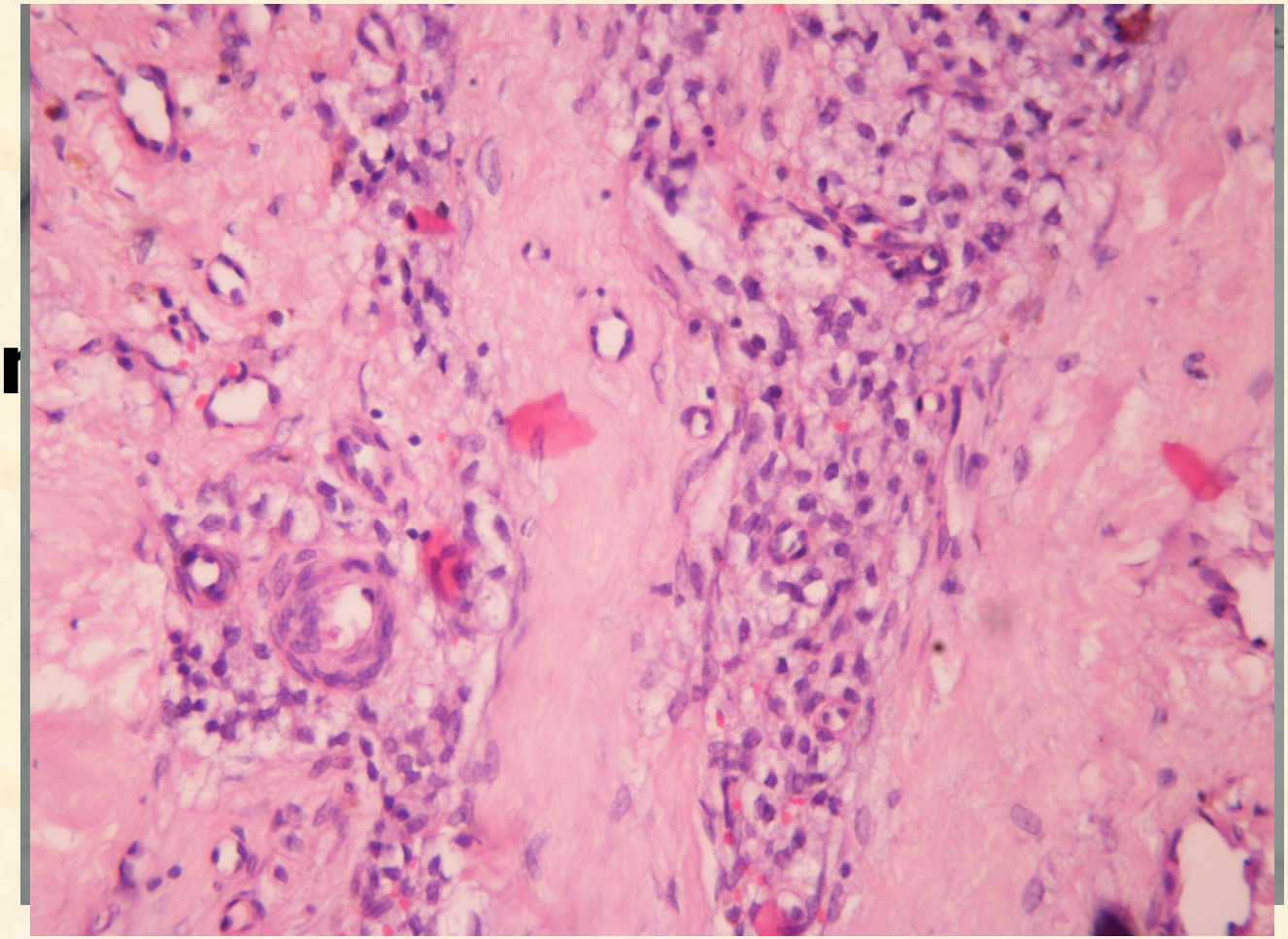
What next ???

MRI

CT chest

Bone Scan

Biopsy



Principles of Biopsy

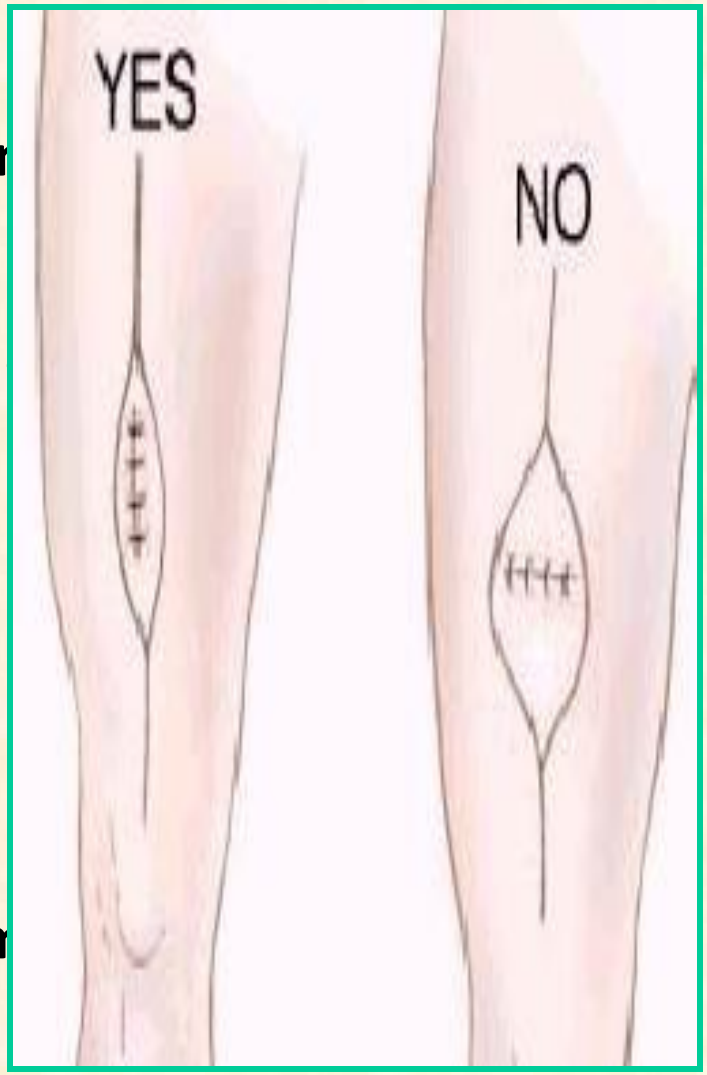
Should be planned and executed by sar

All clinical ,lab, radiological evaluation

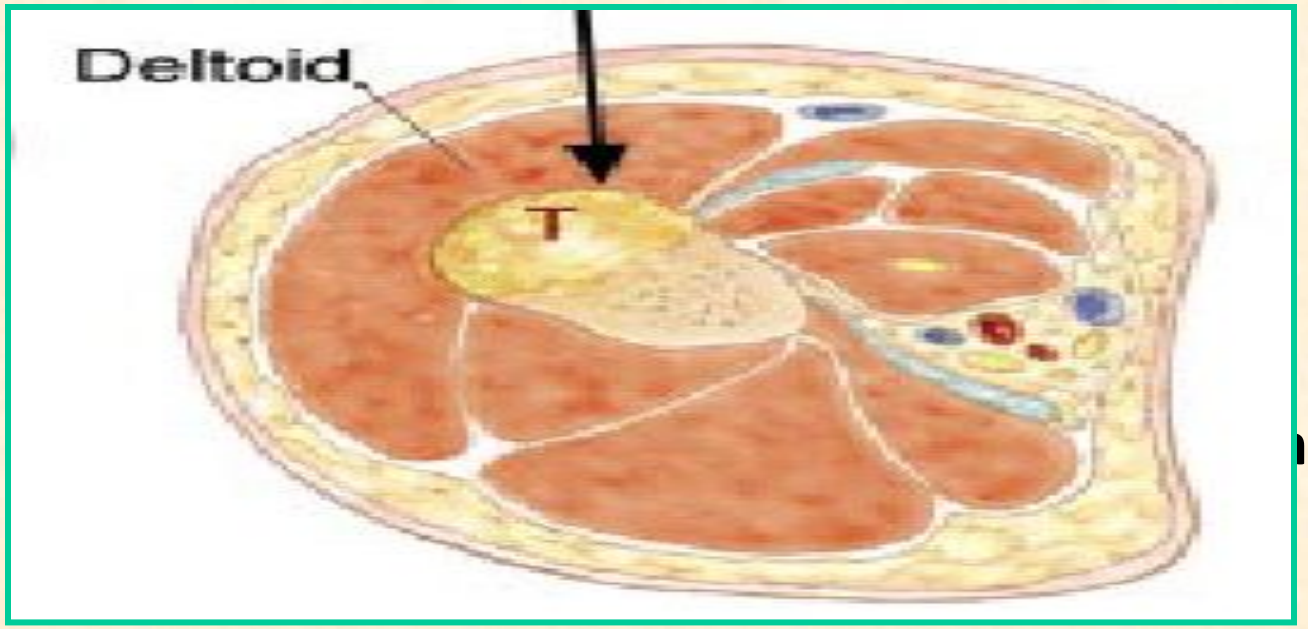
Incision – long axis of the extremity

Do not raise flaps

Deep dissection through one compartm



Principles of Biopsy…..



Types of Biopsy

BIOPSY TYPE	TISSUE OBTAINED	ADVANTAGES	DISADVANTAGES
Fine-needle aspiration	Cells	Cost effective Fewer complications Good for obese patient or tumor near neurovascular structure	Small sample size Need expert pathologist
Core needle	Small tissue core	Cost effective More tissue than fine-needle aspiration	More complications* than fine-needle aspiration
Incisional biopsy	Adequate sample of mass/lesion	Adequate tissue sample (gold standard)	Increased complications* May compromise definitive resection
Excisional biopsy	Entire lesion removed	Removes entire lesion Indicated for small lesion or expendable bone	Increased complications*

*Complications include infection, bleeding/hematoma, pathological fracture, tumor contamination/seeding.

Management

Neo-adjuvant Chemotherapy

??ROLE OF RADIOTHERAPY

Reassessment and Preparation for Surgery

Is Limb Salvage feasible??

What Surgical Procedure???

??ROLE OF RADIOTHERAPY

Post Op Chemo therapy

Neo Adjuvant Chemotherapy

Prior to 1970's, bone sarcomas were routinely treated with amputation. Yet most patients died from metastatic disease.

Over last two decades, limb salvage surgery for bone sarcomas has gained popularity throughout the world and around 85% patients are now offered limb salvage.

Advances in modern multi-agent chemotherapy regimens

Neo-Adjuvant Chemotherapy

Benefits of pre-operative chemotherapy:

1. **Reduces cellular volume and viability**

2. **Eliminates micro-metastatic disease**

3. **Formation of Pseudo-capsule around the tumor: barrier against local spread, more differentiation of planes during surgery**

Pre-operative Radiotherapy..Ewings

Centrally located lesions such as spine/pelvis where attaining wide surgical margins is not possible

Inoperable tumors with palliative intent

Post-operative Radiotherapy..ewings

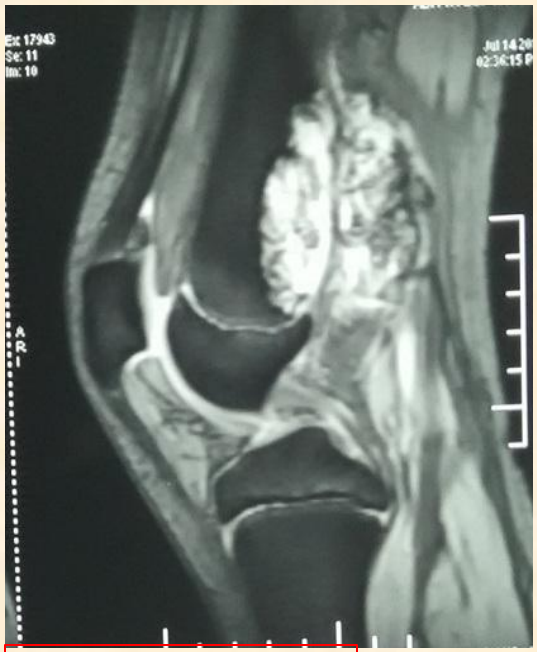
Poor response to Neo-adjuvant Chemotherapy (Necrosis <90 % in HPE of resected specimen)

Surgical Margins if Inadequate

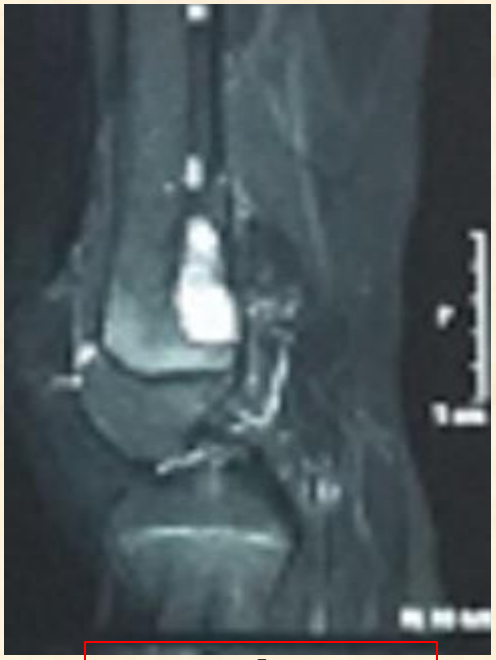
Preoperative Assessment

RE-STAGING of tumor

(Repeat MRI is MUST…!!)



Pre-chemo



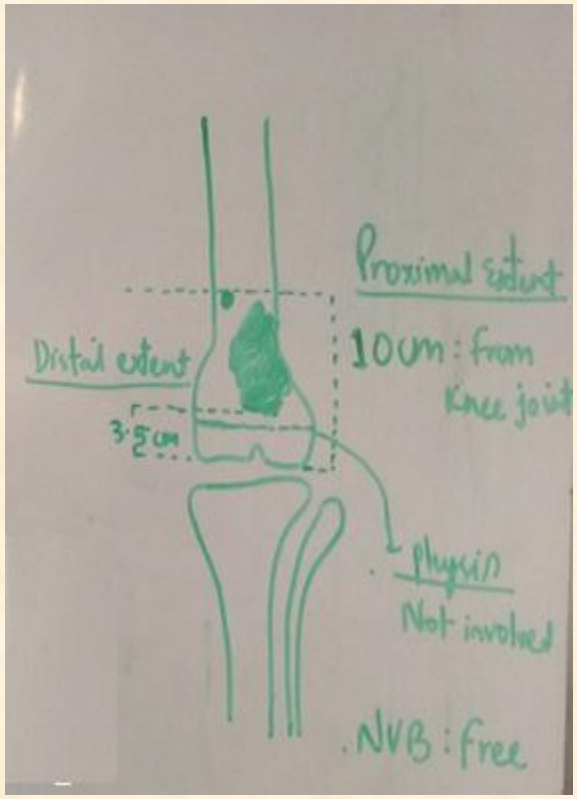
Post-chemo

Discussion with Radiologist Regarding..

Status of Neurovascular Bundle

Extent

Physis/Joint involvement



Surgery

The goal of surgery must always be complete tumor removal

Amputation :

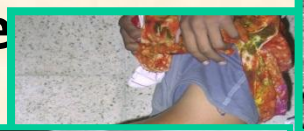
Attaining adequate surgical margins is not feasible (major NVB encasement)

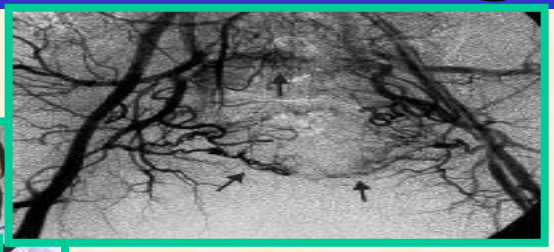
Expectant poor distal extremity function post-operatively.

Limb salvage surgery should only be performed in institutions where the margins of surgical excision can be accurately assessed.

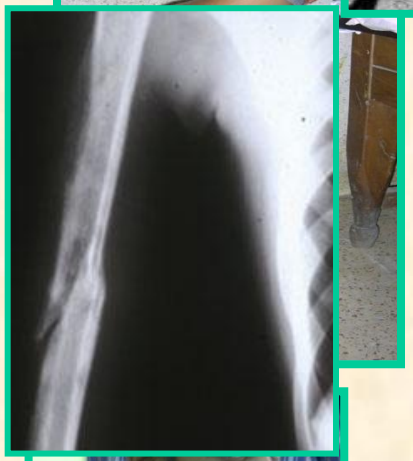
Contraindications to Limb Salvage

Neurovascular involveme



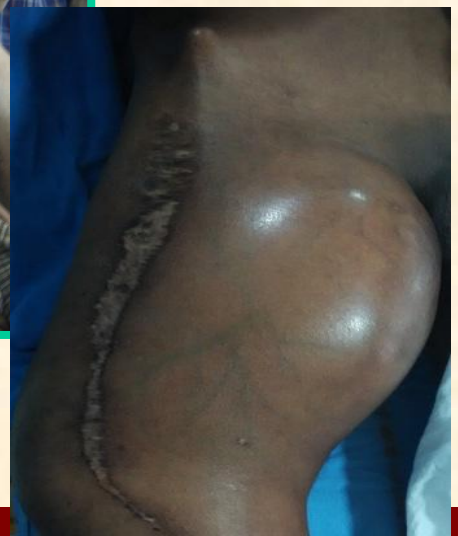


Large sized tumour



Displaced pathologic fracture (relative contraindication)

Fungating and infected tumors



Recurrence of malignant tumors

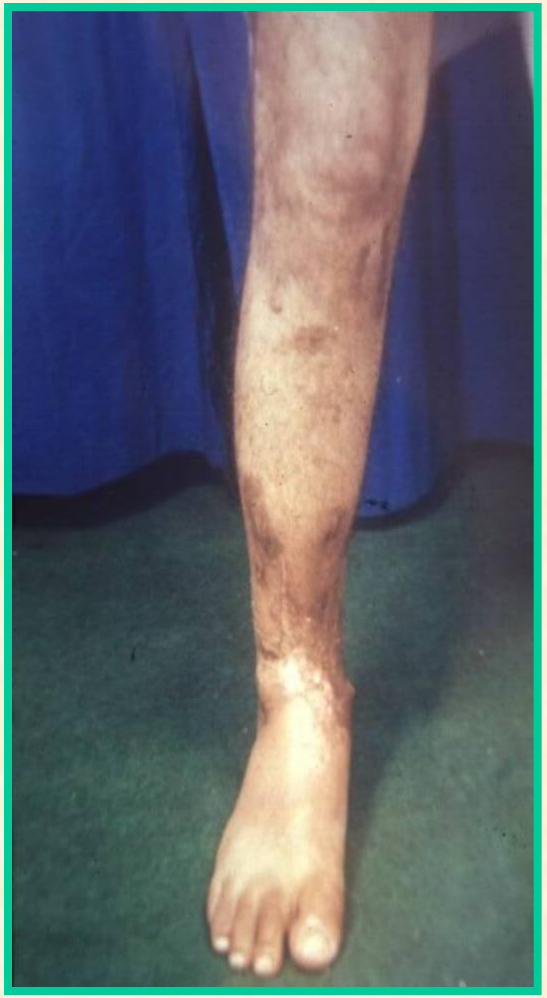
The Salvaged Limb

Acceptable degree of function

Cosmetic appearance

Minimal amount of pain

Durable enough to withstand the demands of normal daily activities



Surgical margins: Enneking

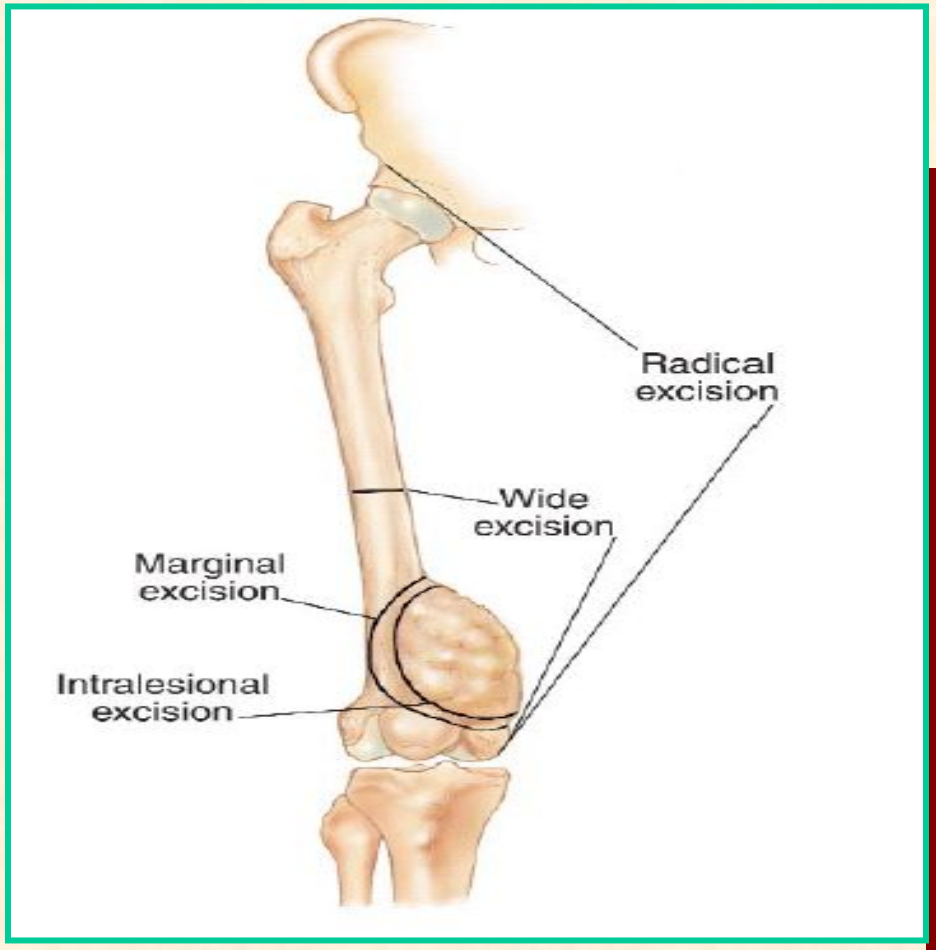
Enneking WF. A system of staging musculoskeletal neoplasm. *Instr Course Lect*. 1988;37:3–10.

Intralesional → **Inadequate**

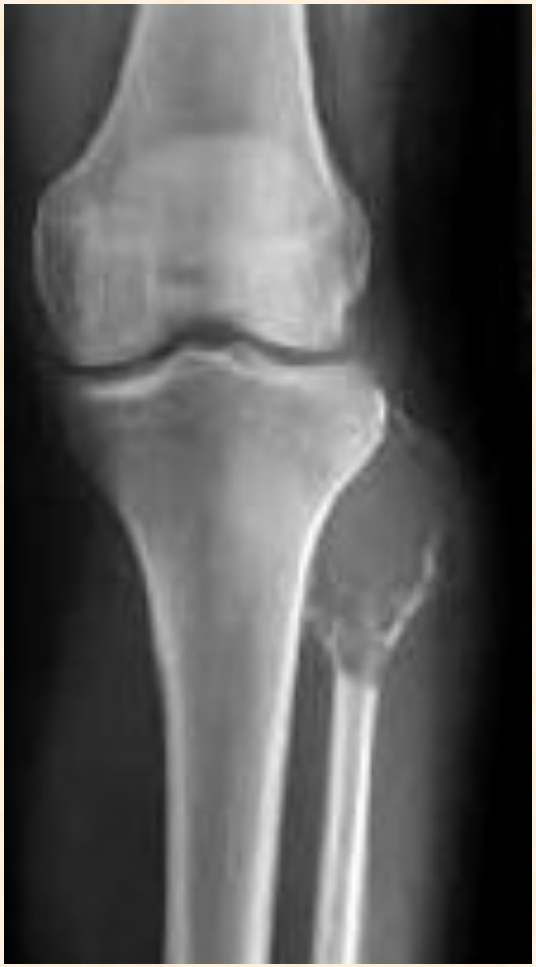
Marginal → **Inadequate**

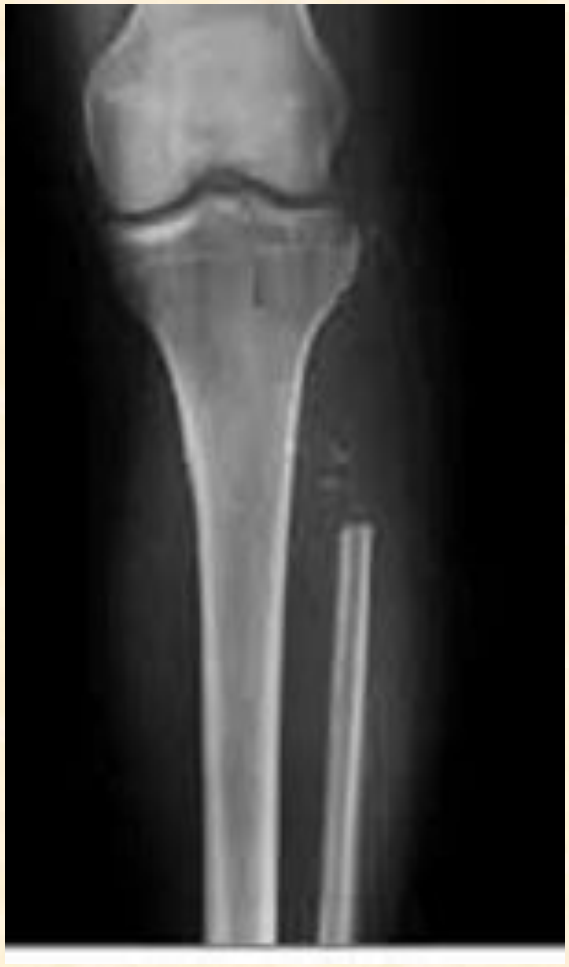
Wide resection → **Adequate**

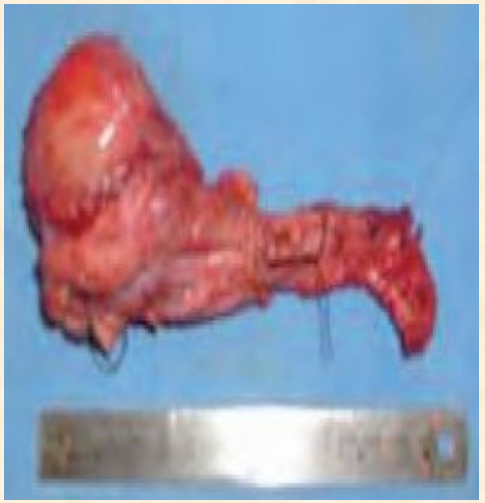
Radical resection → **Adequate**



Excision Alone







Arthrodesis

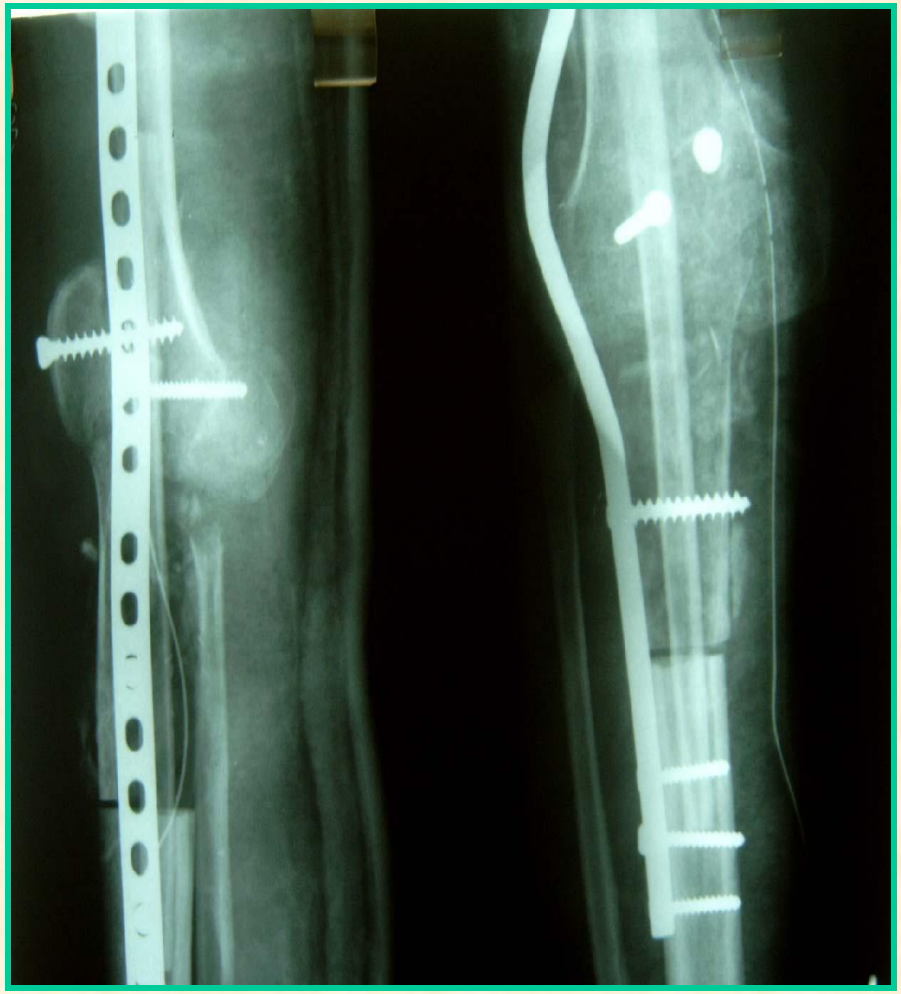
Indicated in young adults with high functional demands

Extensive muscle resection

Need for an extra-articular resection

Cost-effective

Relevant to the Indian Scenario

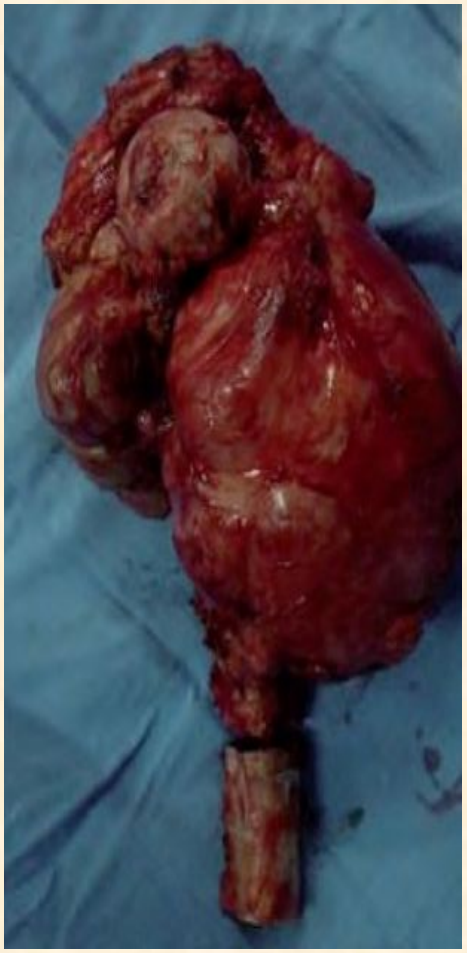


Endoprosthetic Reconstruction

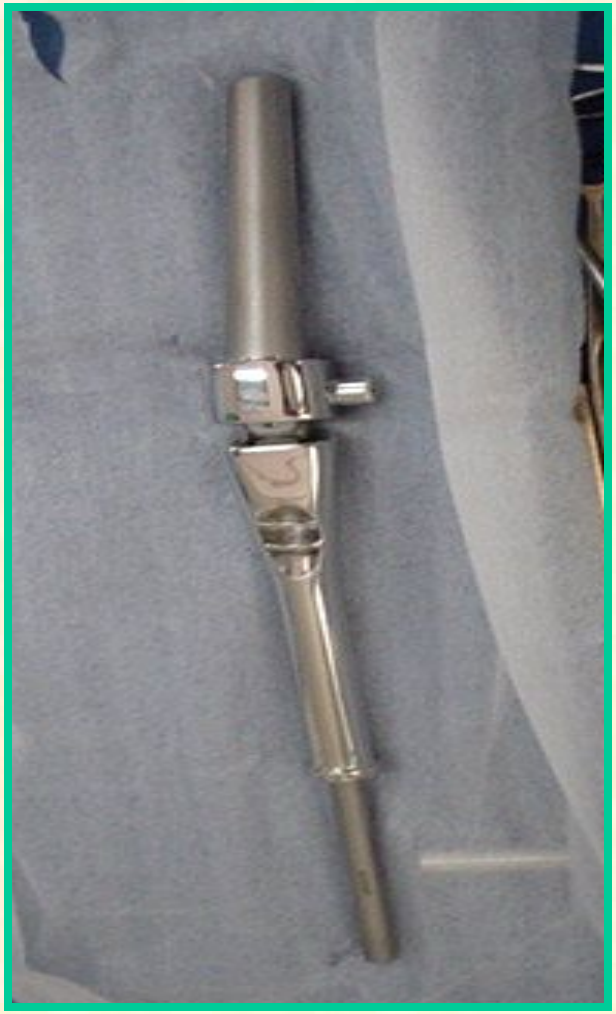
Large metallic device designed to replace the excised length of bone and the adjacent joint

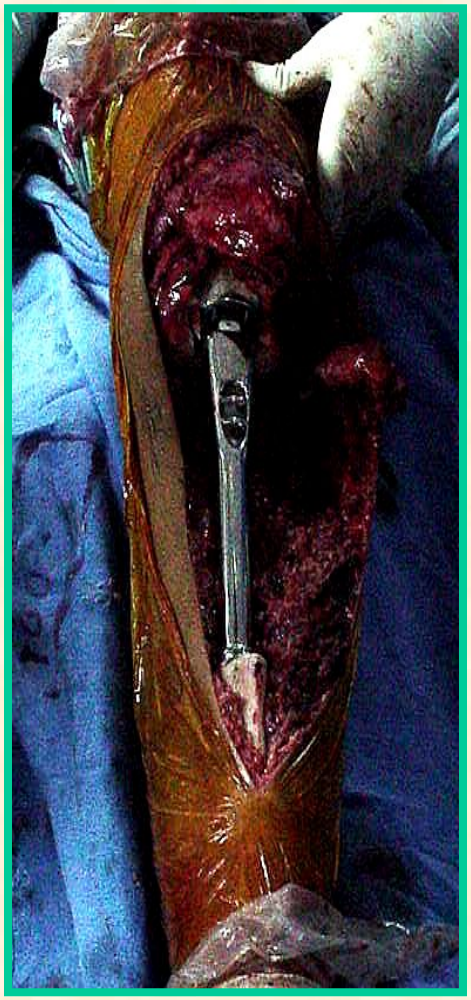
Available for all the major anatomical regions of the body

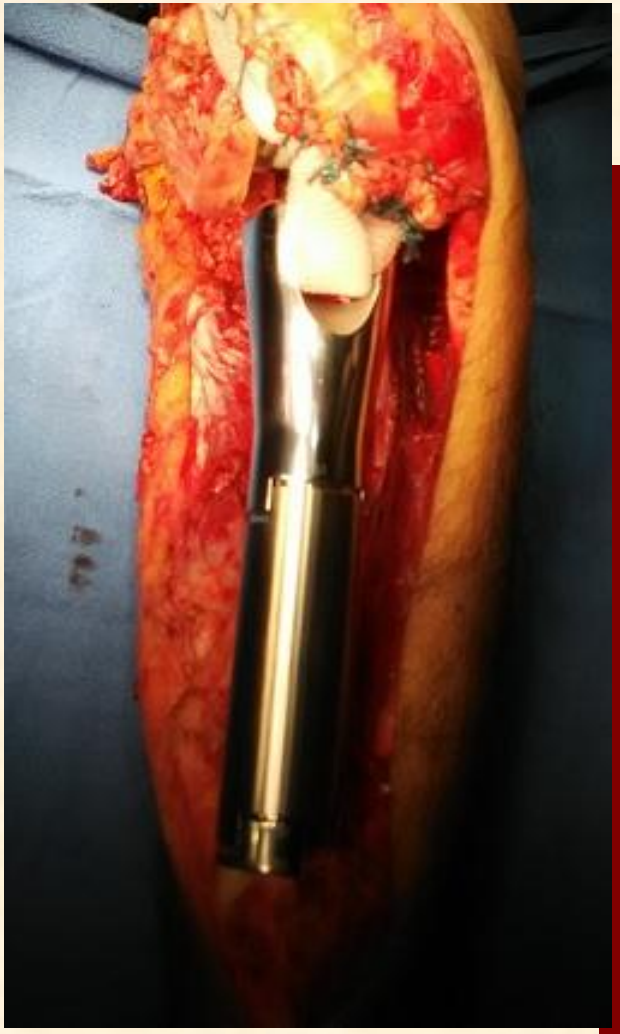
Available as custom made prosthesis and as modular system



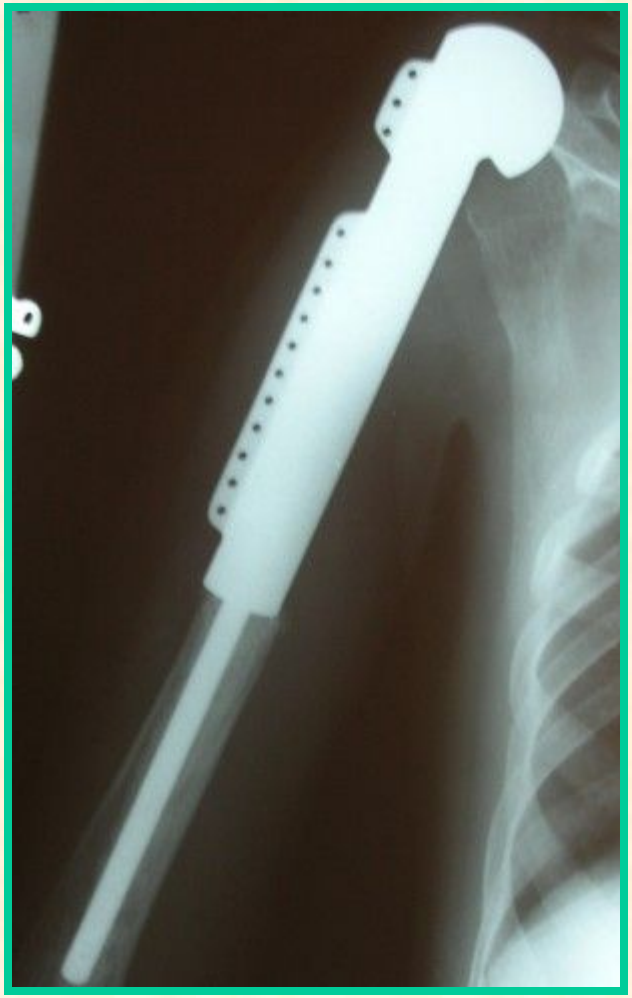
Endoprosthetic reconstruction

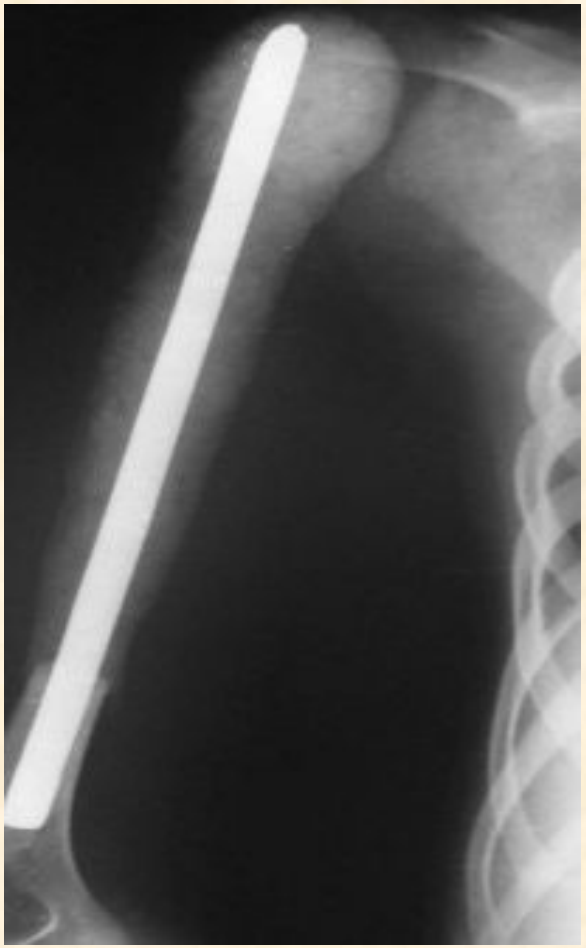






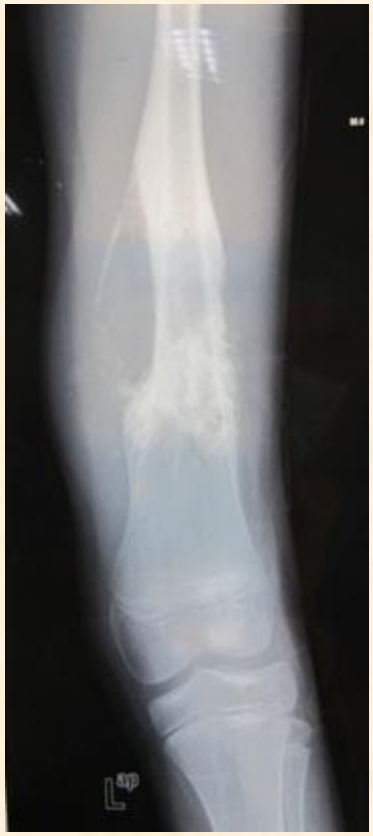
Endoprosthetic Reconstruction

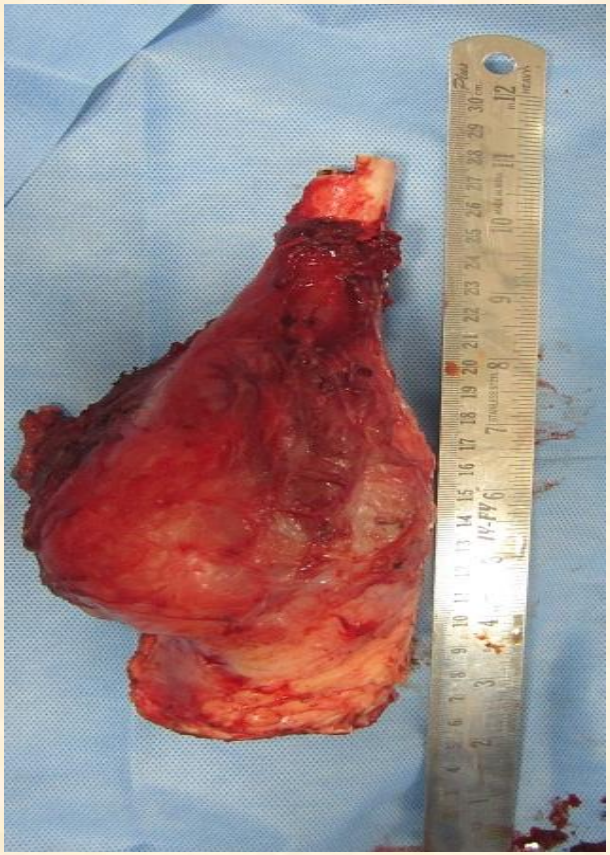


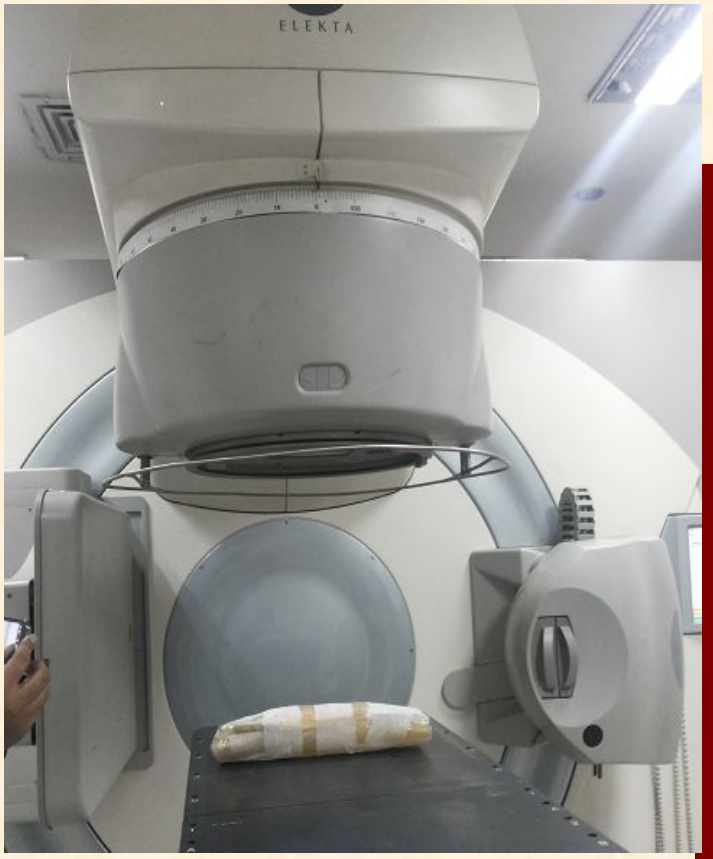


Recent trend: nail Cement spacer if large portion of deltoid involved by tumor

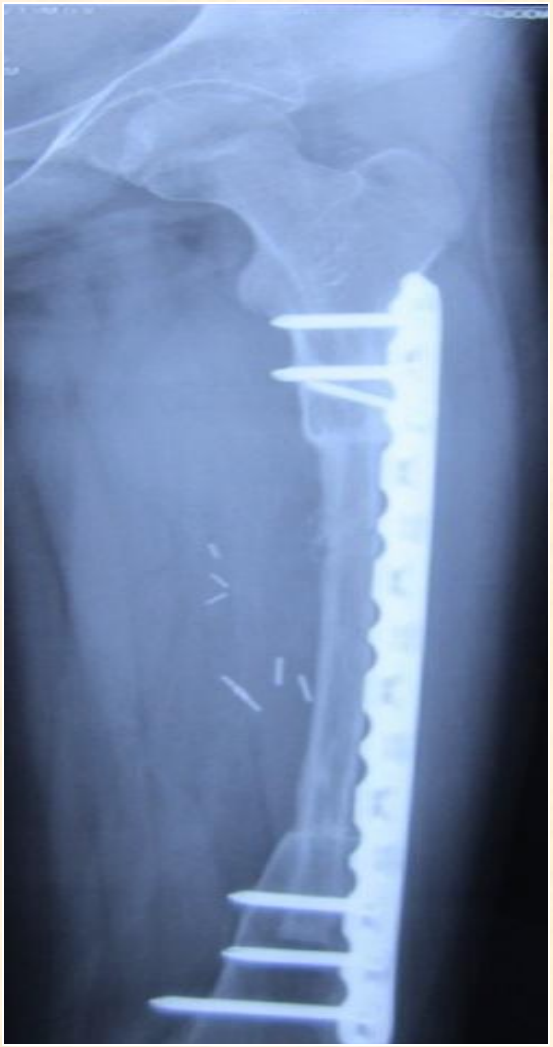
ECRT

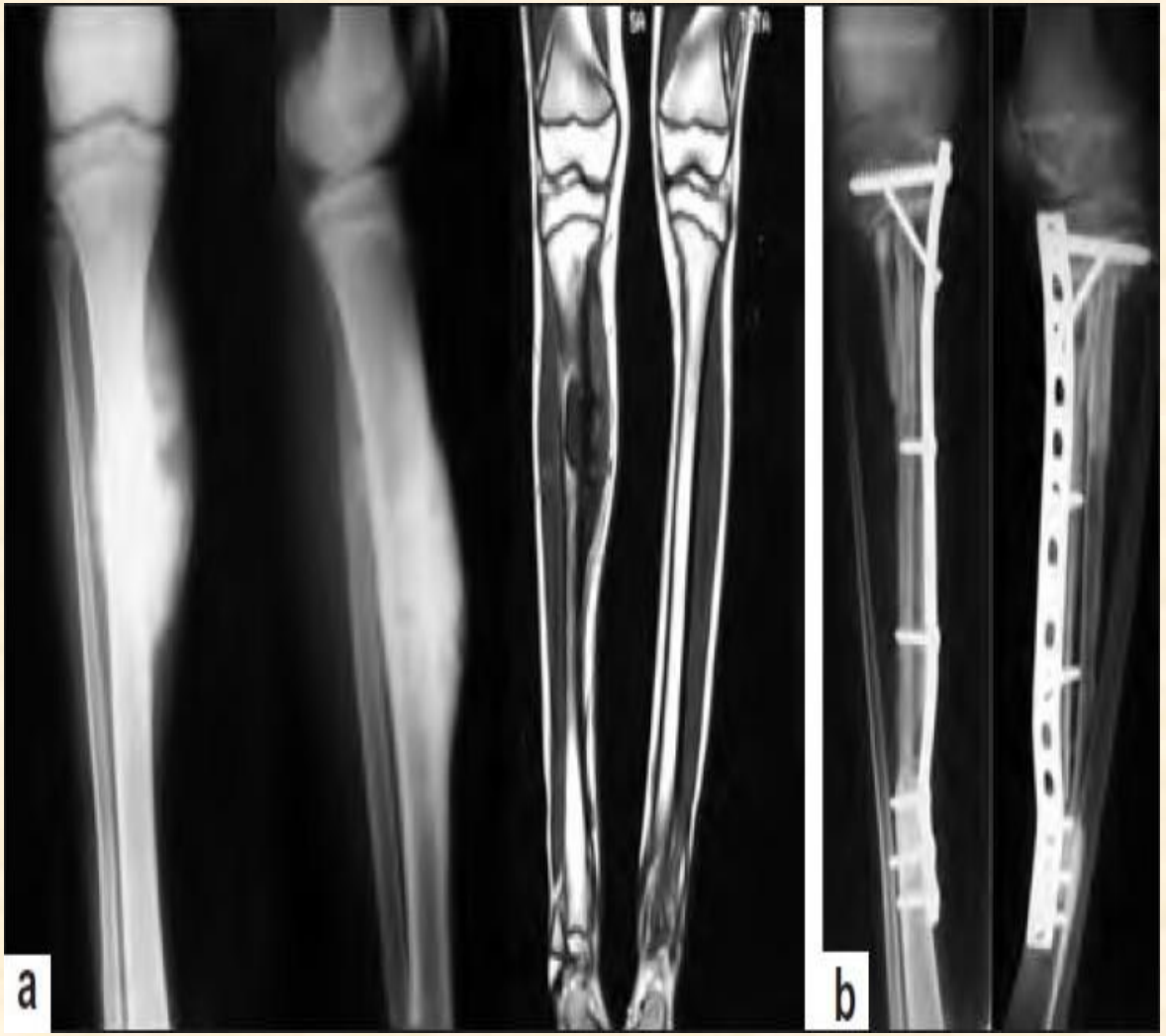




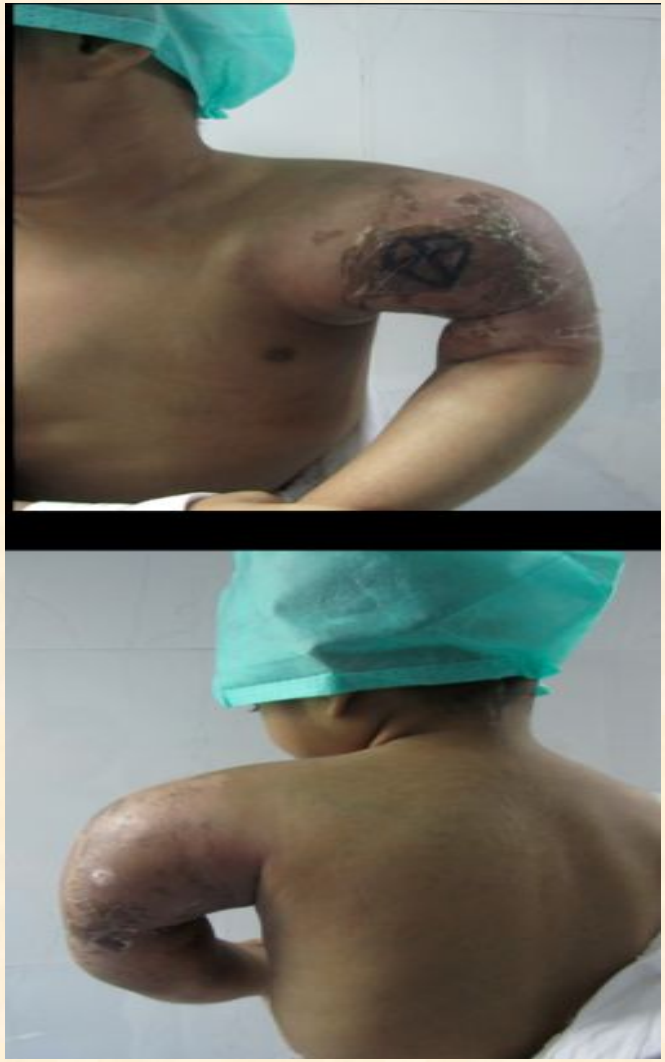


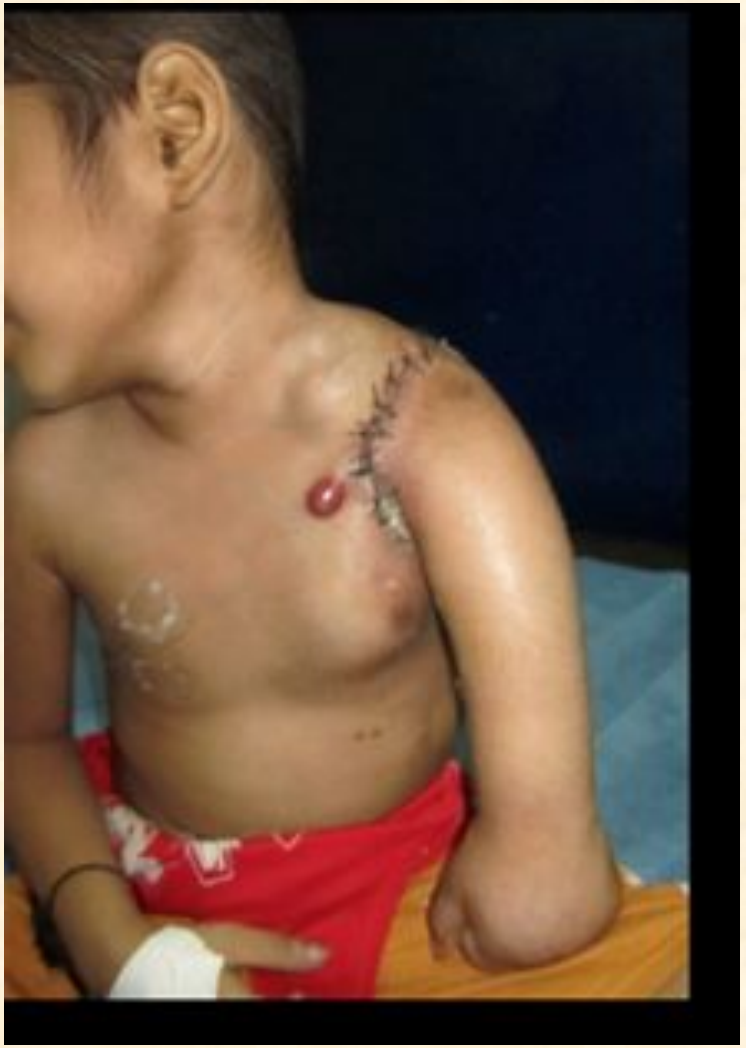
Intercalary Reconstruction





Forearm Reimplantation





THANK U