When the fracture is more: Pathologic Fractures in the Pediatric Population

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Objectives

- How to recognize a pathologic fracture
- How to evaluate a pathologic fracture
- Treatment principles in the management of pathologic fractures
- Common pathologies encountered in the pediatric population

Recognizing a pathologic fracture

- Be suspicious when
  - The mechanism of injury seems disproportionately minor
    - Example: a fall from standing results in a humerus fracture
  - The fracture pattern is atypical for location
    - Example: transverse fracture in a femur after a fall
  - The bone doesn't look normal at the fracture site
    - Example: any lesion, or appearance that is cystic, lucent, sclerotic, or with surrounding reaction such as periosteal elevation

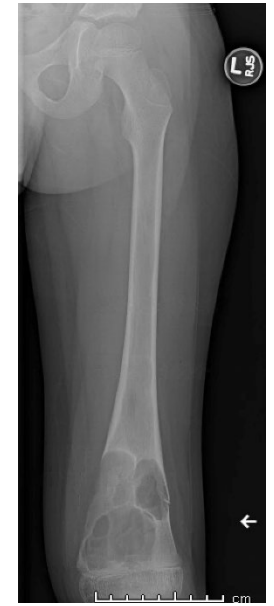
Evaluating a suspected pathologic fracture

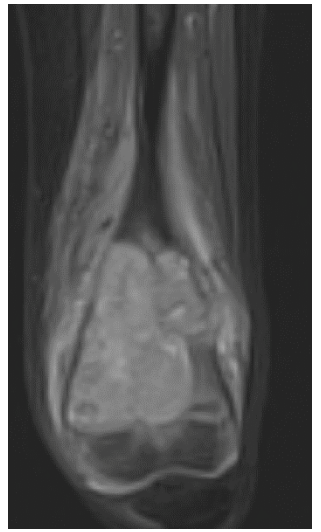
- Obtain a detailed history
  - Local and constitutional symptoms: confirm if any pain, particularly night pain, fever, weight loss, swelling, or lack of these
  - Current or past medical conditions
  - Family history
- Physical exam
  - A thorough exam includes evaluation for skin changes, swelling, soft tissue mass, lymphadenopathy

Evaluating a suspected pathologic fracture

- Diagnostic imaging
  - Plain x-rays are typically done to identify the fracture
  - Ensure complete imaging of the whole bone

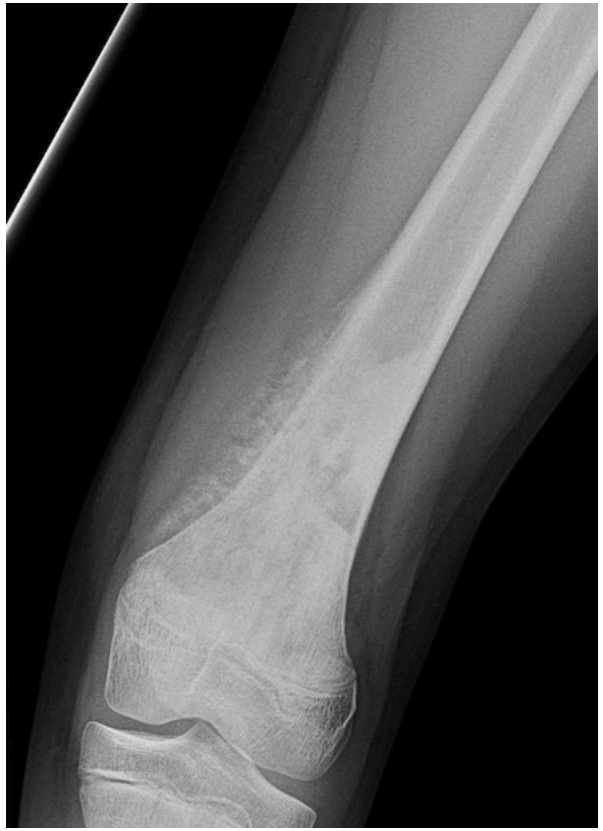
- Advanced imaging can be useful
    - MRI, CT, bone scan





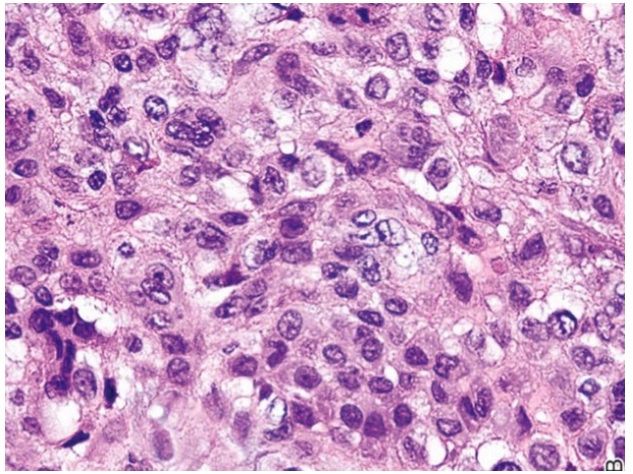
Evaluating a suspected pathologic fracture

- Diagnostic imaging
  - Evaluate the lesion
    - Location
      - Diagnosis can be specific to epiphysis/metaphysis/diaphysis
    - Character
      - Consider the presence of cyst, calcification, or matrix in the bone
    - Reaction of the underlying bone
      - Is there periosteal elevation, is the shape of the bone normal, is there a clear border to the lesion



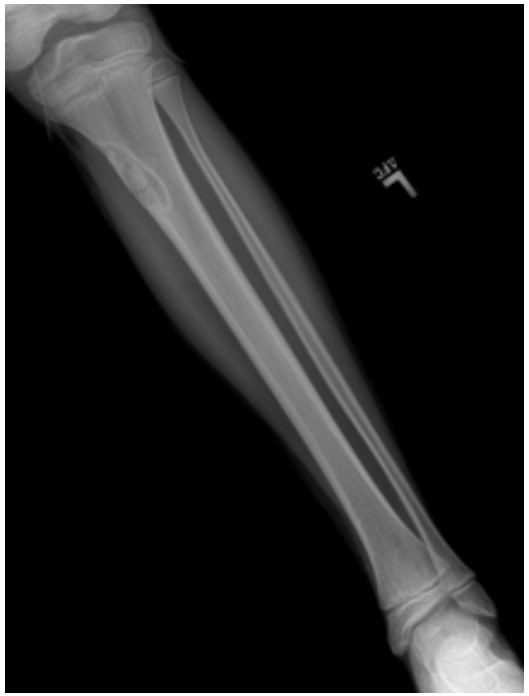
Evaluating a suspected pathologic fracture

- Biopsy
  - May be needed for diagnosis or treatment planning
  - Sometimes performed at the time of operative fracture management
  - Principles of oncology must be followed in performing a biopsy
    - Consider possibility of malignancy and need for future resection
    - Send cultures as well as tissue specimen
- Consider consultation with an orthopaedic oncologist prior to biopsy, particularly if there is concern for malignancy.



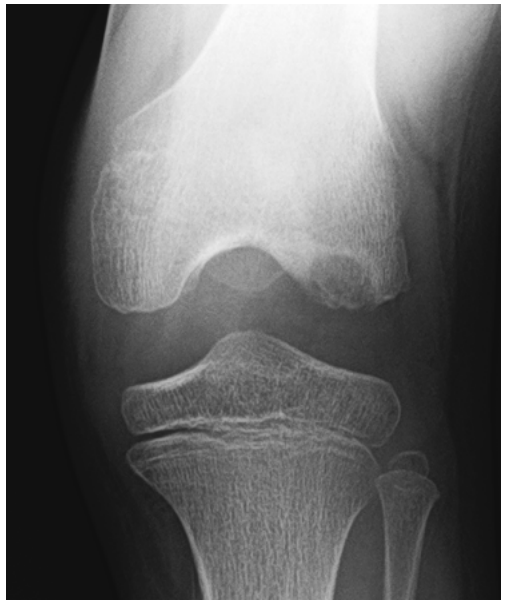
Treating a suspected pathologic fracture

- Treat the fracture
  - Not all pathologic fractures require operative management
  - Some pathologic fractures can be treated as though there were no lesion
- Treat the pathology
  - Some pathologies can be observed, or treated in a delayed fashion, i.e. after fracture healing.

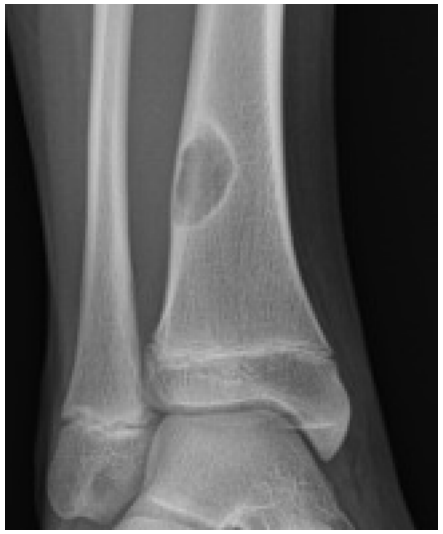


Common benign pathologies by location

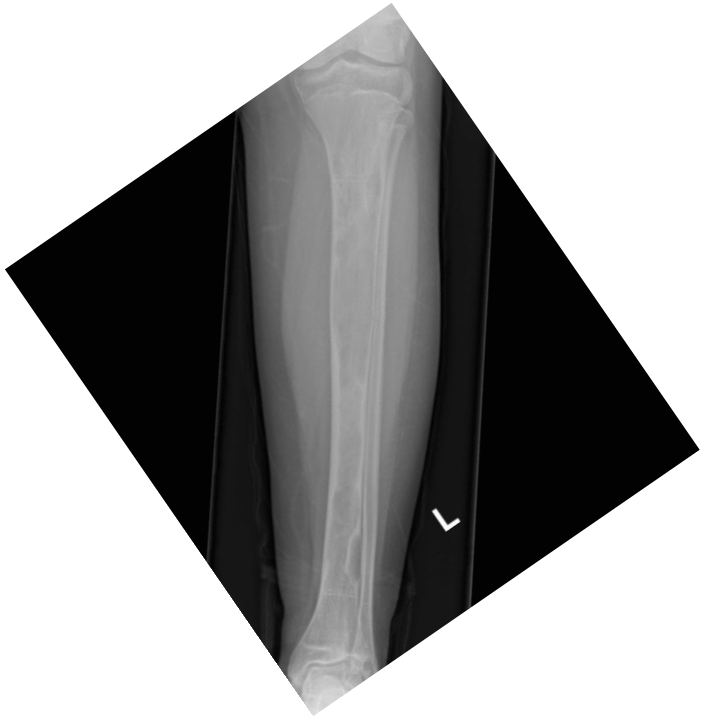
- **Epiphysis:**
  - Chondroblastoma
  - Giant cell tumor



- **Metaphysis**
  - Osteomyelitis
  - Nonossifying fibroma
  - Unicameral bone cyst
  - Aneurysmal bone cyst
  - Fibrous dysplasia

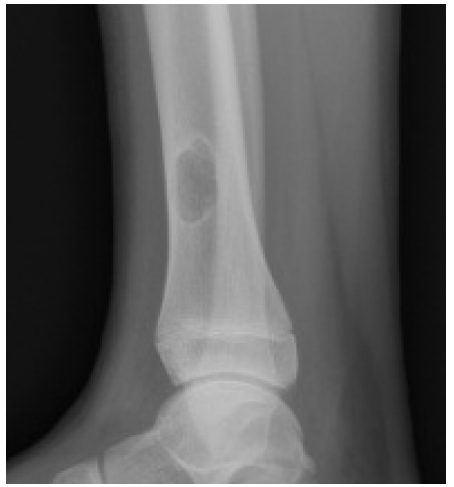


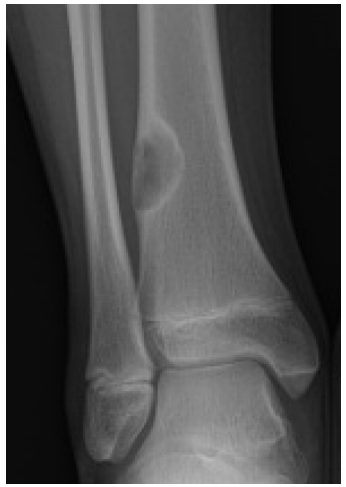
- **Diaphysis:**
  - Fibrous dysplasia
  - Osteofibrous dysplasia

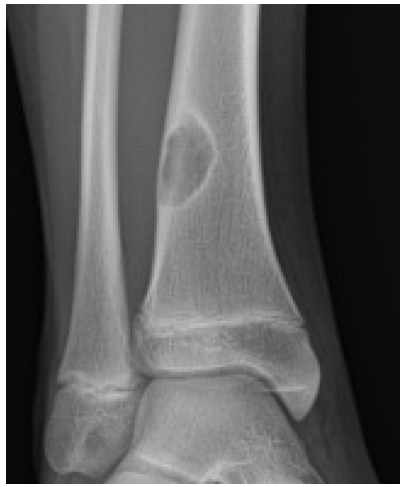


Nonossifying Fibroma

- Most common benign lesion
  - Common incidental finding on xrays
- Eccentric, metaphyseal, multilocular lucency with a sclerotic border
- *Often classic enough in appearance that no further imaging is necessary*
  - Differential diagnosis: UBC, ABC
  - Other names for this include:
    - Fibrous cortical defect
    - Fibroxanthoma

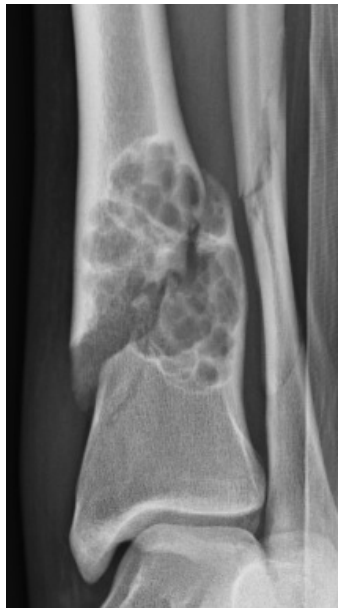






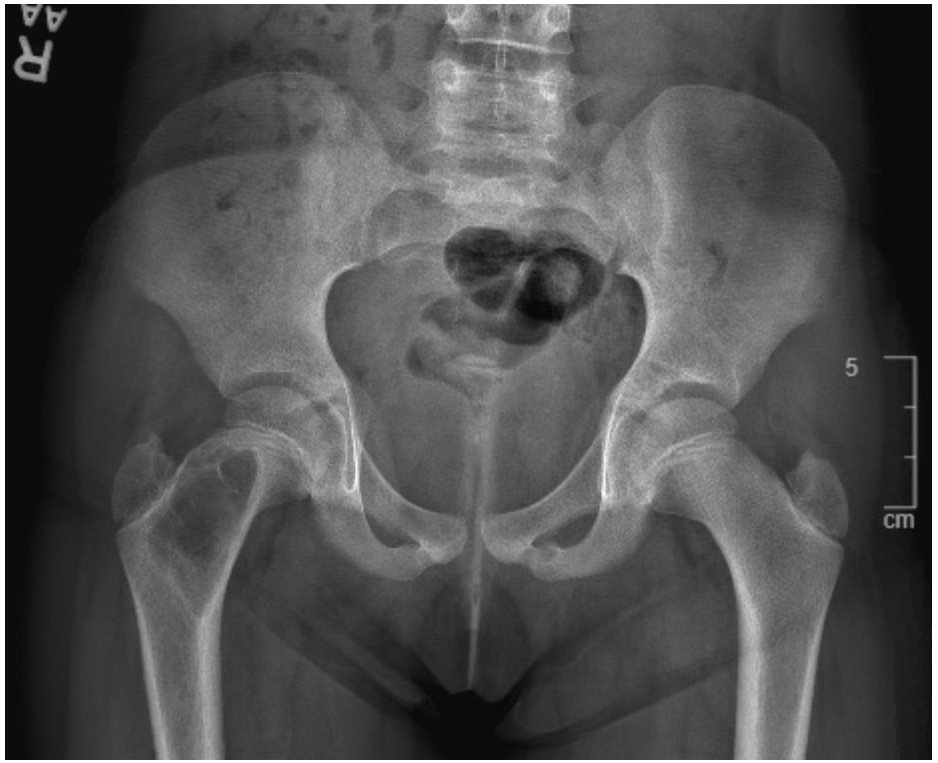
Nonossifying Fibroma

- Observation is mainstay of treatment
- Pathologic fracture may be treated nonoperatively with predictable healing
- Large enough lesions causing fracture or bone pain may indicate surgical management
  - Curettage and grafting
  - Local recurrence rare



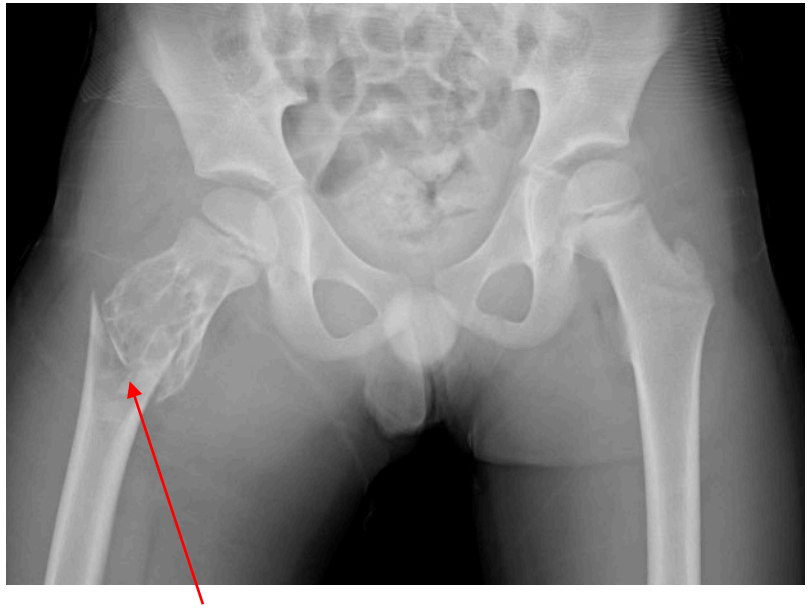
Unicameral Bone Cyst (UBC)

- Benign cystic lesion
- 2:1 male:female ratio
- Common between ages 4-10
- Proximal humerus (59%) and proximal femur (26%) most common
- Uncommon to grow or recur after skeletal maturity



Unicameral Bone Cyst (UBC)

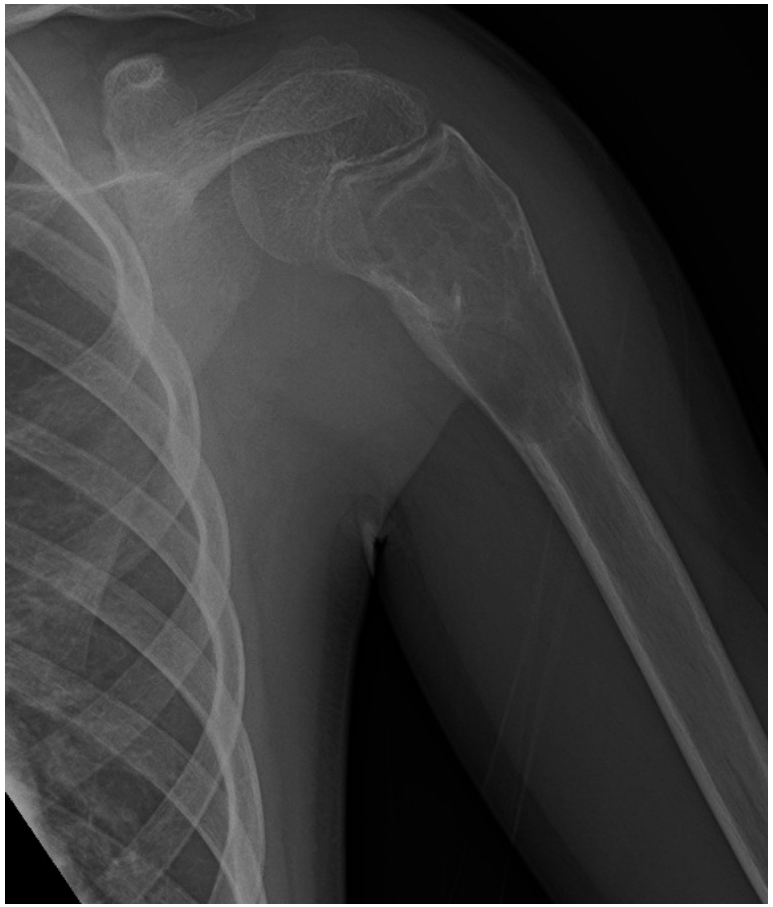
- Well-defined, radiolucent lesion centrally located within the metaphysis
  - Cortical thinning
    - No periosteal reaction
  - Mild cortical expansion
    - NOT wider than the epiphysis
- Fallen leaf sign
  - Fragment of bone floating inside the fluid-filled cavity
  - Typical after a fracture



Fallen leaf sign: cortical fragment within the lesion

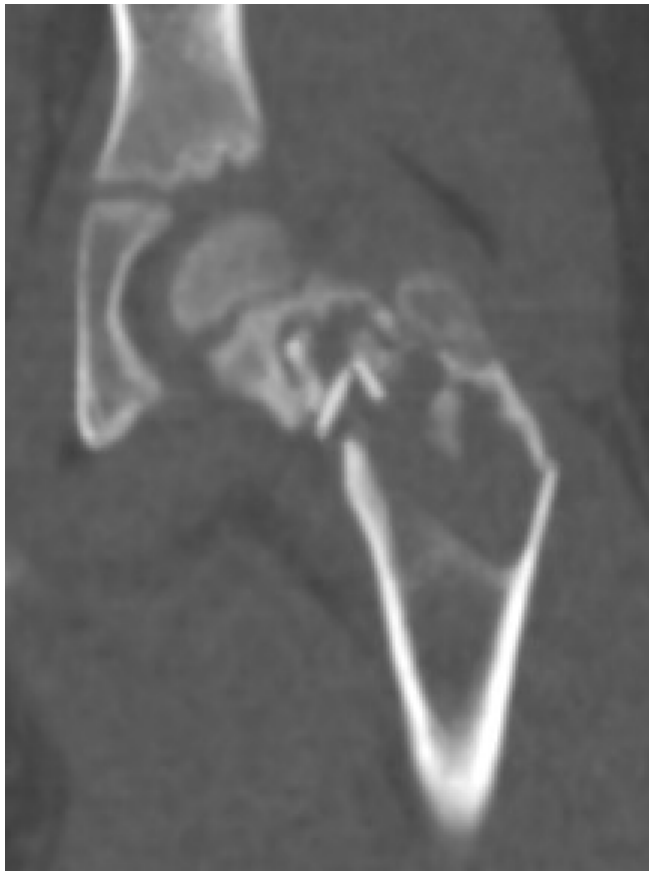
Unicameral Bone Cyst (UBC)

- Most commonly identified at time of fracture (85% of cases)
- Thin cortical rim
  - Microfracture of this can lead to pain
  - Pathologic fracture possible after minor trauma, e.g. fall from standing
- More potential for growth when closer to the physis
  - Active lesions are near physis
  - Latent lesions are remote from physis



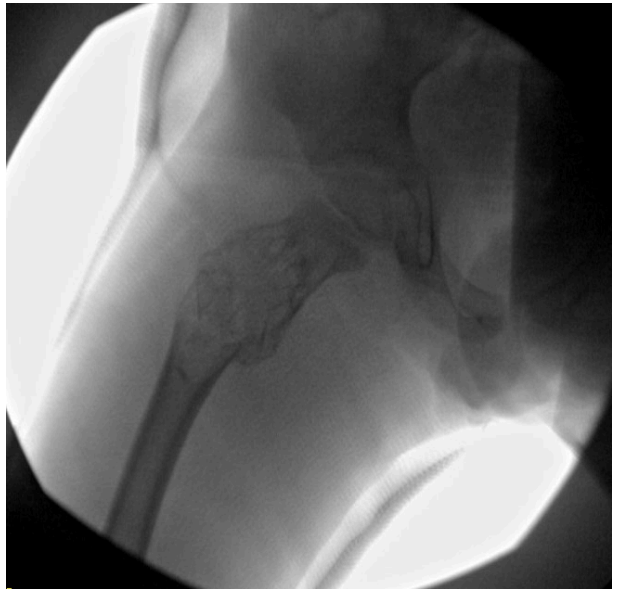
Unicameral Bone Cyst (UBC)

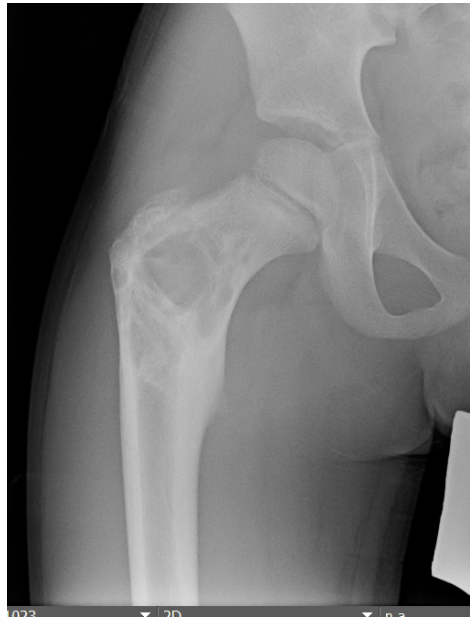
- X-rays usually sufficient for diagnosis
- CT or MR can be considered if diagnosis uncertain
  - Axial skeleton
- Aspiration of straw-colored fluid is diagnostic



Unicameral Bone Cyst (UBC)

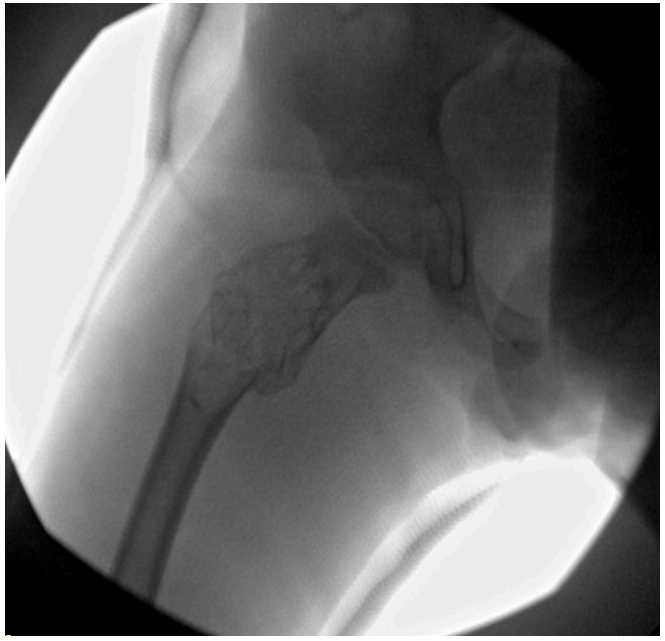
- Treatment of the fracture can be guided by typical fracture treatment principles
  - Conservative treatment is most often indicated
- Low rates of spontaneous healing of cyst with fracture healing

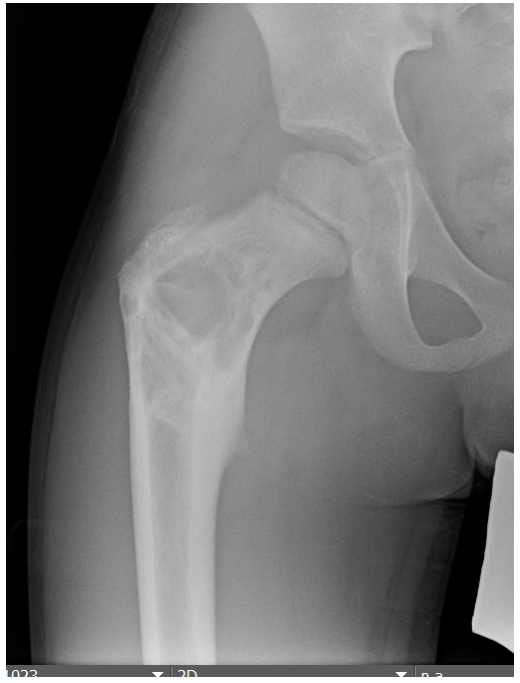


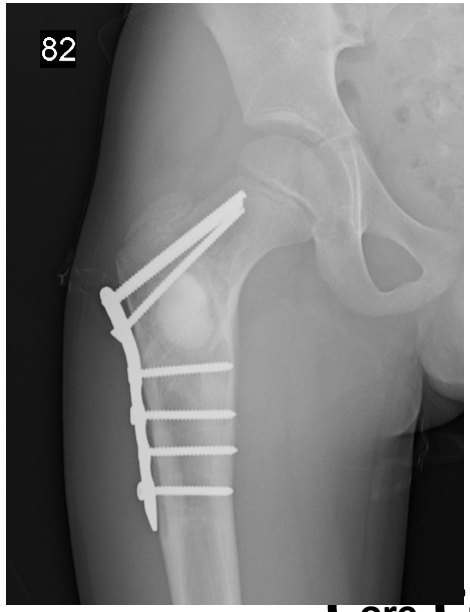


Unicameral Bone Cyst (UBC)

- Treatment of the cyst can proceed upon fracture healing
- Treatment goals include preventing recurrent fracture
  - Prevent complications of fracture→deformity or growth arrest



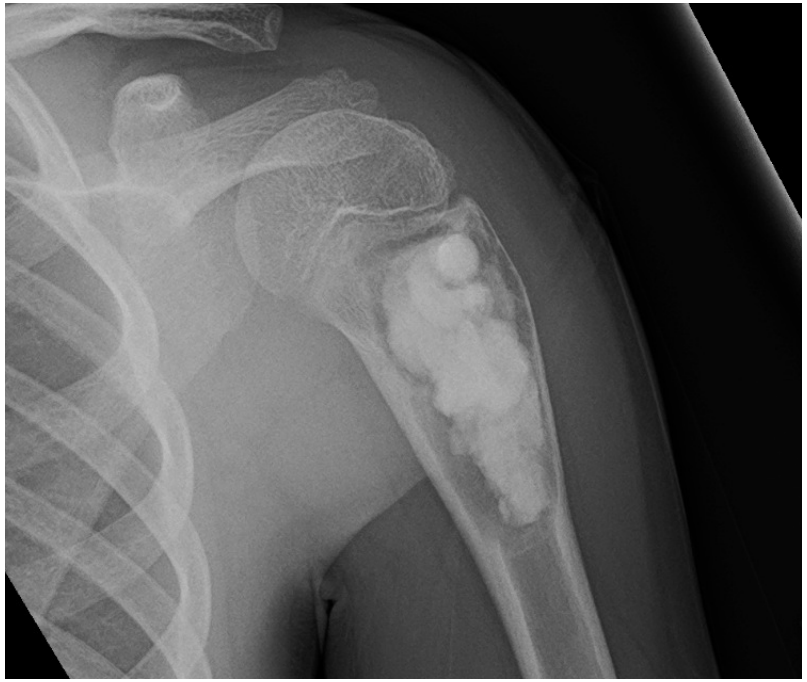




Unicameral Bone Cyst (UBC)

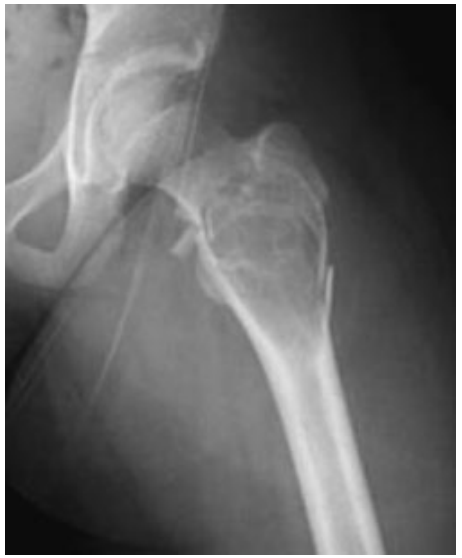
- Treatment options include injection, decompression, and curettage with grafting

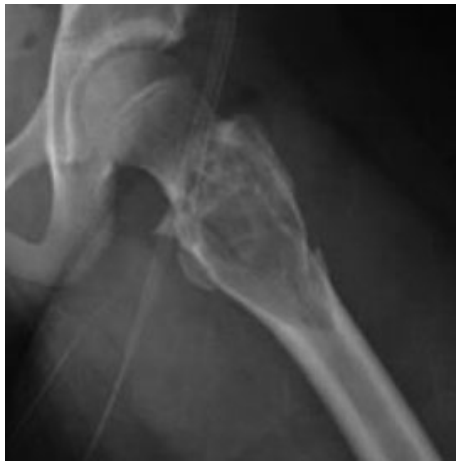
- Comparable healing rates (~80%)
    - Bone marrow or steroid injection
    - Curettage and grafting with either autograft, allograft, or bone substitutes
    - Decompression with IM nails or cannulated screw



Aneurysmal Bone Cyst (ABC)

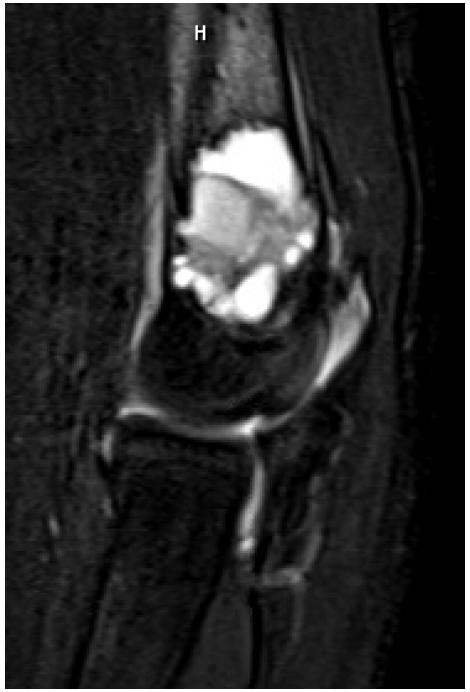
- Benign, but locally aggressive tumor
- Most commonly in teenagers (80%)
- Much less common than UBC
- Most common in long bones and spine
  - 50% long bones
  - 30% spine
- Typically metaphyseal
  - Occasionally found in the epiphysis or diaphysis

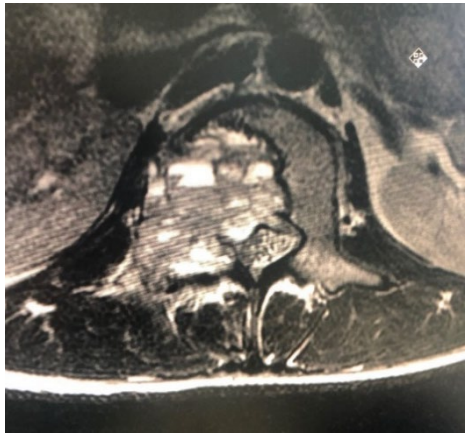




Aneurysmal Bone Cyst (ABC)

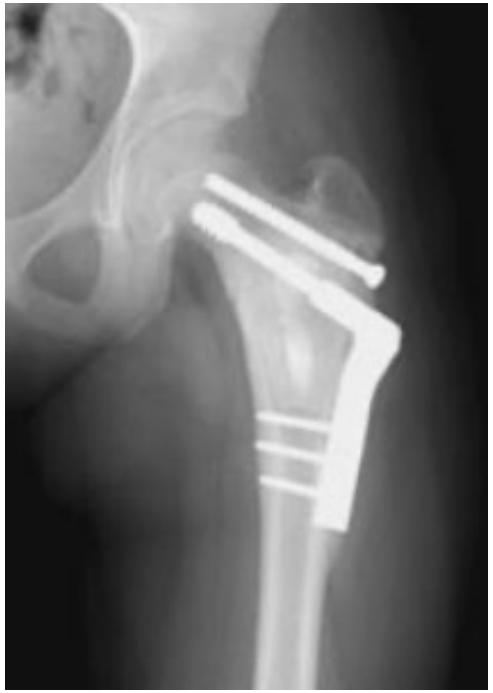
- Radiolucent, eccentric, expansile lesion, most commonly in the metaphysis
  - May see periosteal reaction due to aggressive nature of this benign tumor
- Fluid/fluid levels on MRI characteristic
- Differential diagnoses: telangiectatic osteosarcoma, giant cell tumor, UBC, secondary ABC
- Biopsy recommended to confirm diagnosis





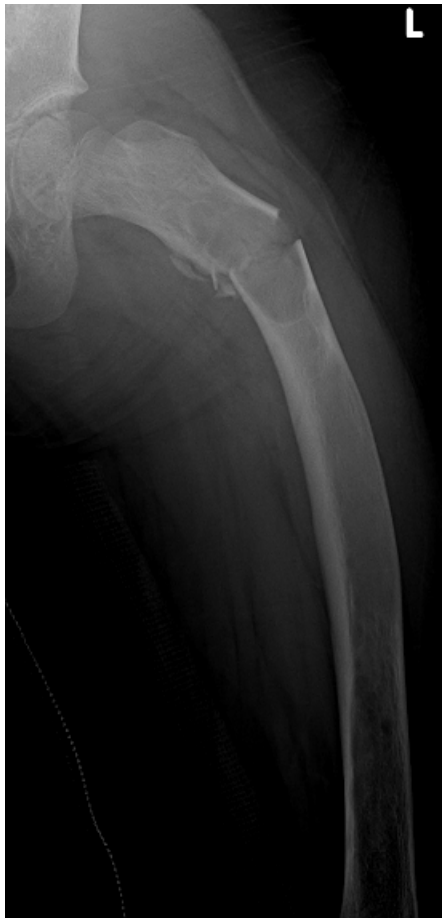
Aneurysmal Bone Cyst (ABC)

- Treatment is most commonly curettage and grafting, with or without adjuvant therapy
  - Embolization used pre-operatively to reduce bleeding
- High rate of local recurrence
- Radiation limited to inoperable lesions



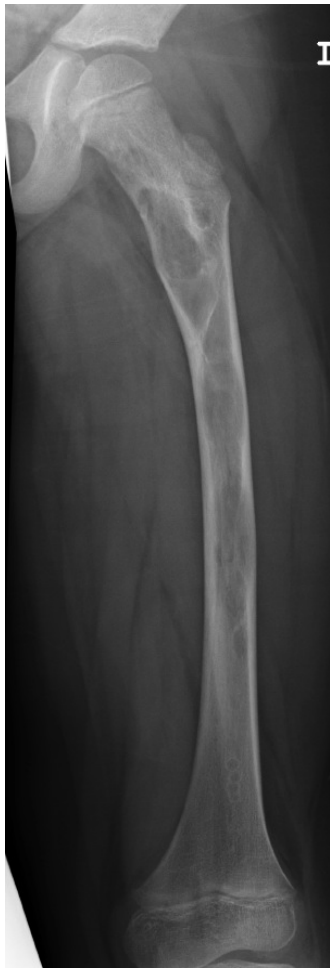
Fibrous Dysplasia

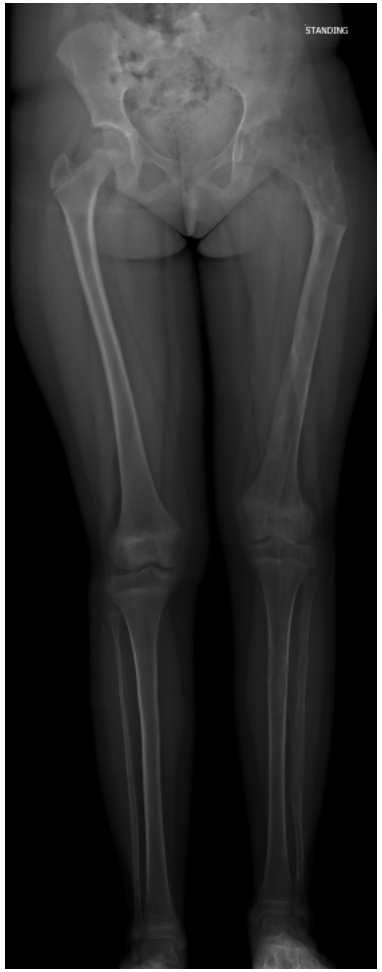
- Benign lesion
- Most common in long bones, pelvis
  - *Long lesion in a long bone*
- Mutation in $G_s\alpha$ gene
- Failure of maturation of bone
  - Immature matrix leads to decreased mechanical strength of bone
  - This can lead to pain, pathologic fracture, deformity
- Clinical presentation with bone pain or fracture



Fibrous Dysplasia

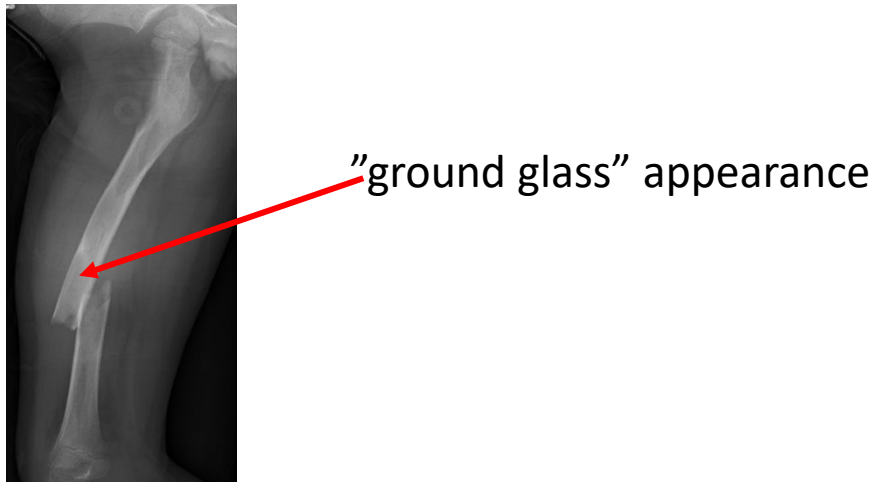
- Monostotic more common
  - Femur most common site
- Polyostotic form can be more severe
  - Larger lesions and secondary deformity
    - Deformity can be caused by microfracture and progressive structural deformation
    - Shepherd's crook deformity of the proximal femur
  - Can be associated with endocrine abnormalities
    - McCune-Albright syndrome→precocious puberty

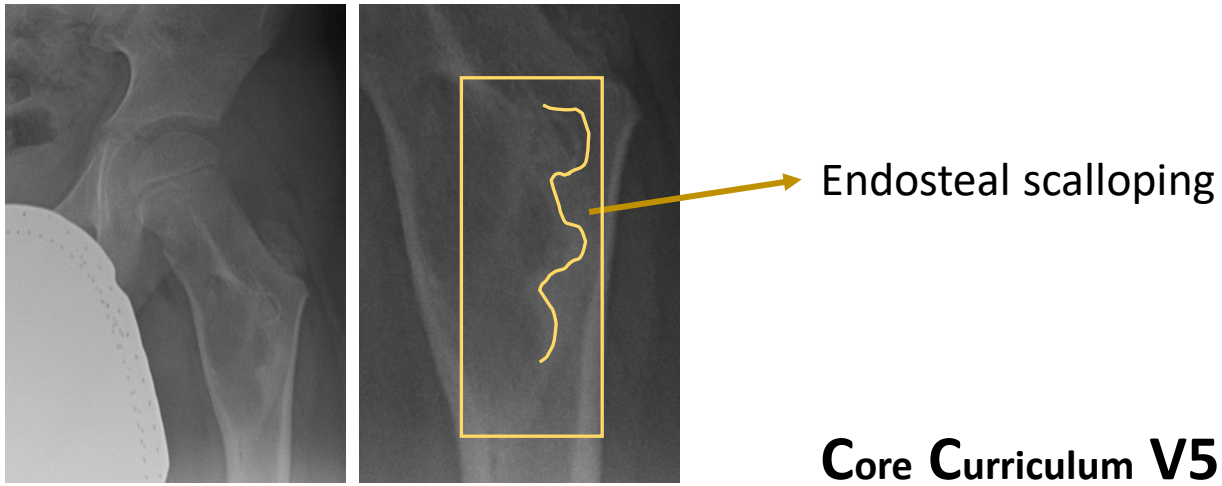




Fibrous Dysplasia –Imaging

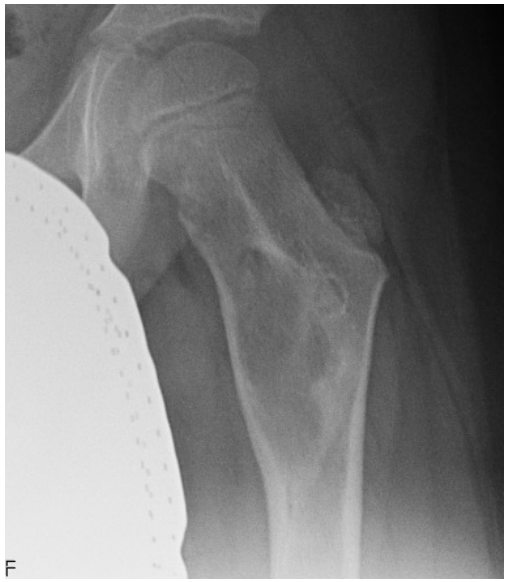
- Xrays will demonstrate a “ground glass”
  - Irregular, metaplastic woven bone replacing trabecular bone
- Lesion may appear expansile, with endosteal scalloping
- Periosteal reaction not typically seen
- *Differential diagnoses: UBC, NOF*



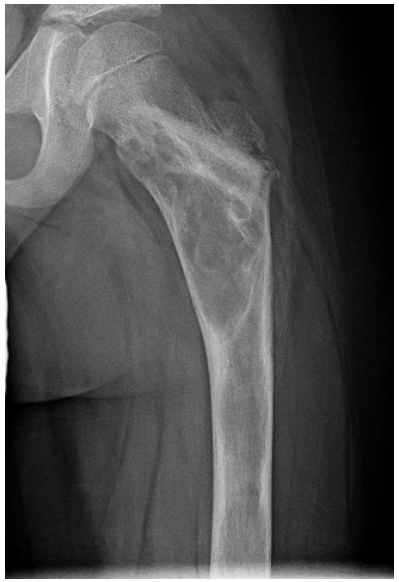


Fibrous Dysplasia - Treatment

- Observation of asymptomatic or incidental lesions is recommended
  - Follow XR needed to ensure no progression or development of deformity
    - Progressive deformity is common in polyostotic disease
      - Rare in monostotic
- Bisphosphonates can be used in polyostotic form to decrease bone pain

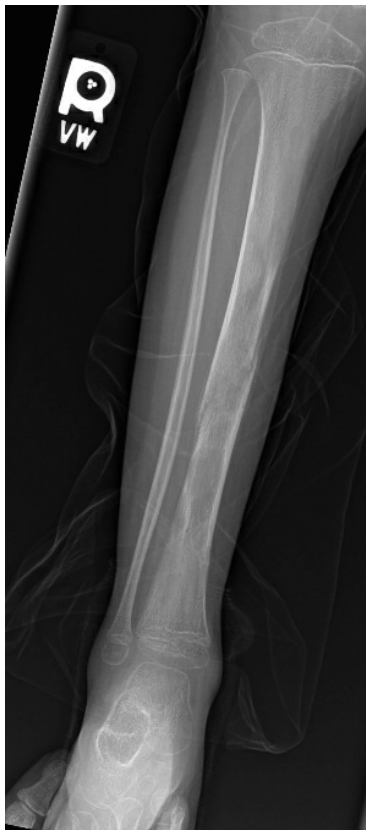


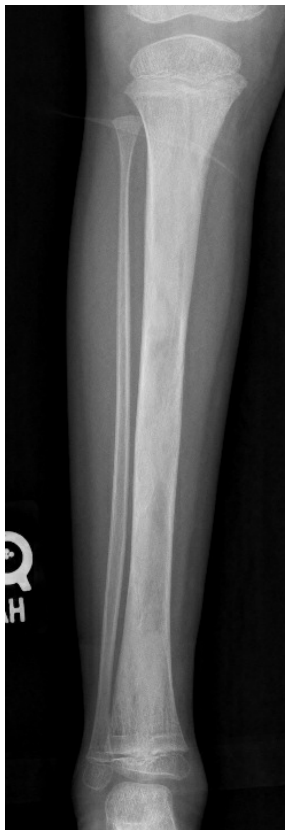
Proximal femoral varus developing



Fibrous Dysplasia – Conservative Treatment

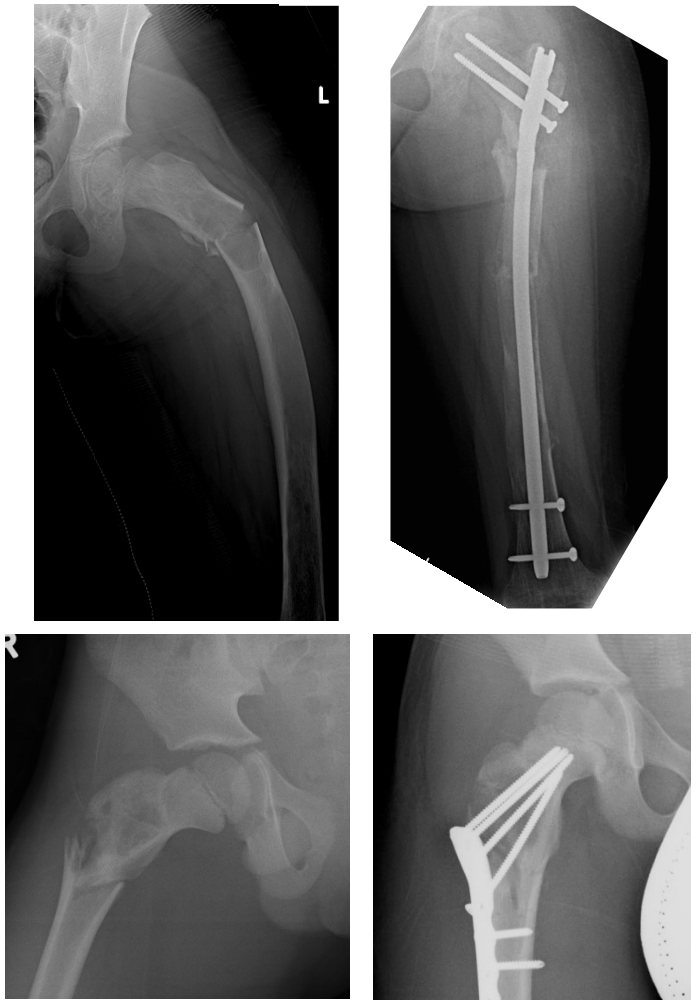
- Treatment of fracture
  - Conservative treatment most commonly indicated
  - Fractures heal rapidly
    - Good periosteal bone formation
    - Poor endosteal bone
  - Underlying bone will remain dysplastic
  - Curettage and grafting is NOT indicated
    - Graft is converted to FD bone





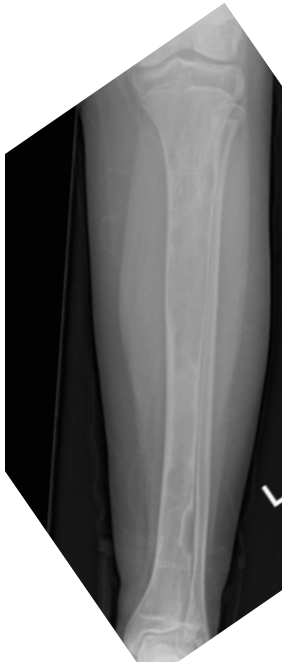
Fibrous Dysplasia – Surgical Treatment

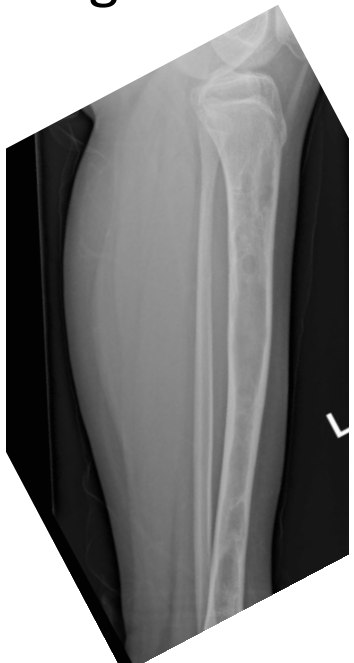
- Most often indicated for
  - Lower extremity/weightbearing bones
  - Polyostotic cases with deformity
- Intramedullary fixation is ideal
  - Load-sharing implant
  - Protect the entire bone
    - Especially in polyostotic cases
  - Additional osteotomies often needed for associated deformities
- Fixed angle constructs required in periarticular fractures when IM fixation is not sufficient

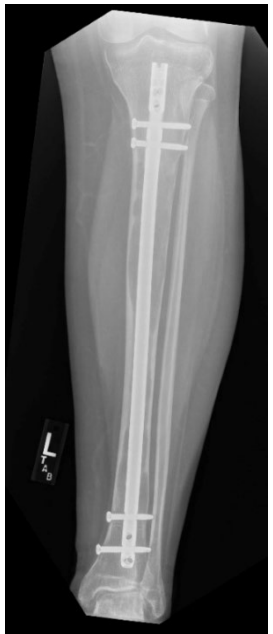


Fibrous Dysplasia – Prophylactic Treatment

- Prevention of fracture
  - Prophylactic fixation in weightbearing bones with large lesions and/or progressing deformity
  - May also present with bone pain in the absence of visible fracture
  - Minor trauma can lead to pathologic fracture

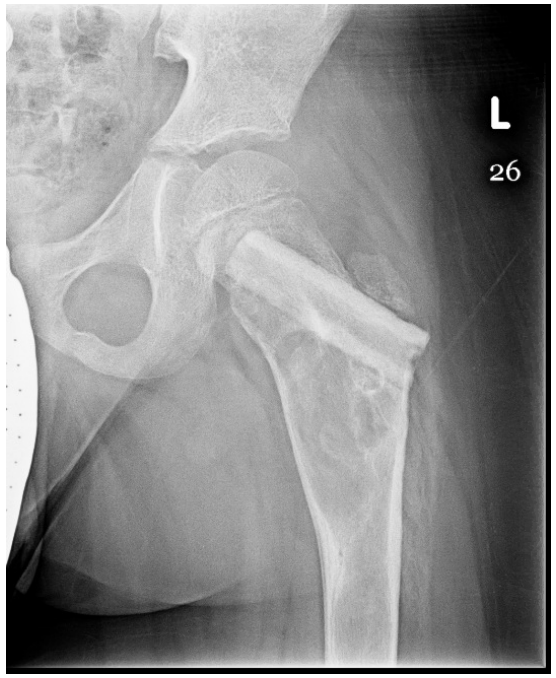




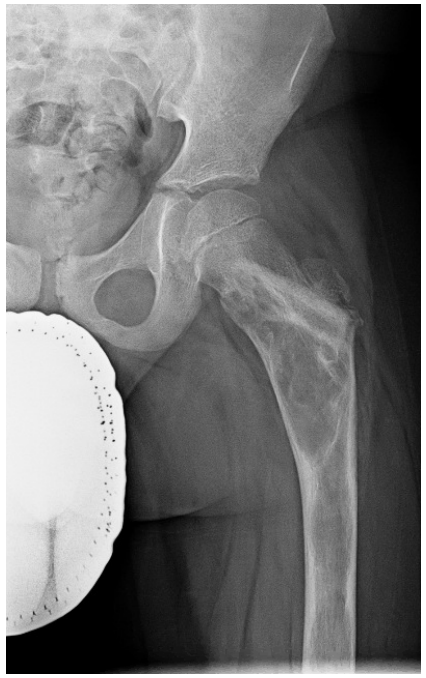


Fibrous Dysplasia - Augmentation

- Augmentation with cortical strut allograft may also improve mechanical strength
    - Bone graft will be resorbed and replaced by fibrous dysplasia over time
        - Nonstructural allograft does not have a role in treating this condition

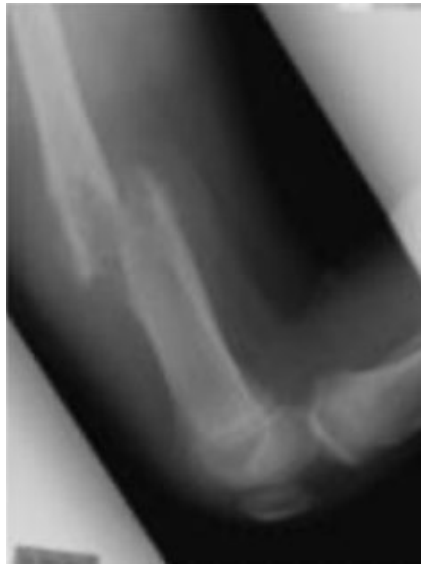


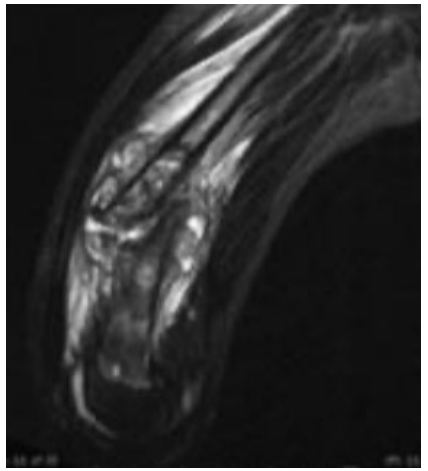
Resorption of cortical strut graft



Malignant Tumors & Metastases

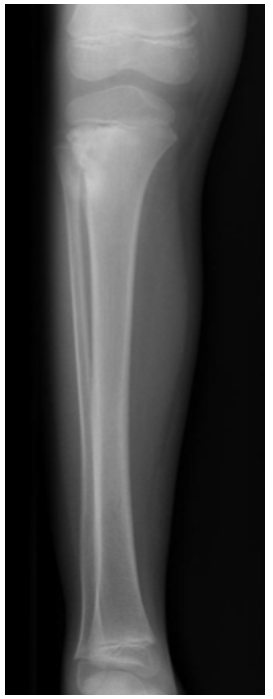
- Malignant bone tumors may cause pathologic fractures
  - Osteosarcoma and Ewing's sarcoma most common
  - Metastases
- Signs of malignancy include
  - Aggressive appearance
  - Periosteal reaction
  - Bone forming and/or destructive lesions with poorly delineated borders
    - Not well circumscribed
    - Permeative appearance
  - Associated soft tissue mass

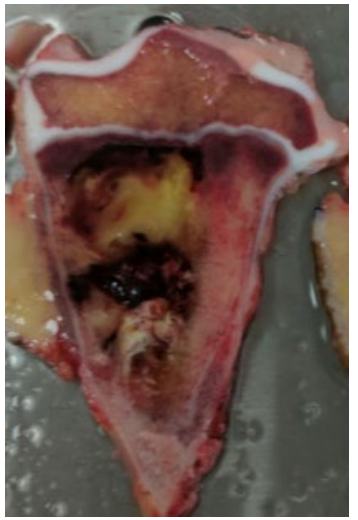




Malignant Tumors & Metastases

- Identification and diagnosis is crucial for appropriate treatment
- Work-up should be done by the treating orthopedic oncologist, especially biopsy
- Do not perform fixation before diagnosis is made
- Treatment of malignancy can include chemotherapy, radiation, and/or surgery
  - Limb salvage is often possible





Pediatric pathologic fractures

- Pathologic fractures can happen from many different etiologies, both benign and malignant
- Treatment of the fracture and treatment of the pathology are dependent on the etiology
- Many benign etiologies do not require treatment distinct from the fracture treatment
- An orthopaedic oncologist should be consulted when dealing with a suspected malignancy

References

- Kadhim, M., Thacker, M., Kadhim, A. *et al.* Treatment of unicameral bone cyst: systematic review and meta analysis. *J Child Orthop* **8,** 171–191 (2014). https://doi.org/10.1007/s11832-014-0566-3
- Rapp TB, Ward JP, Alaia MJ.J Am Acad Orthop Surg. 2012 Apr;20(4):233-41. doi: 10.5435/JAAOS-20-04-233.
- DiCaprio MR, Enneking WF. Fibrous dysplasia. Pathophysiology, evaluation, and treatment. J Bone Joint Surg Am. 2005 Aug;87(8):1848-64. doi: 10.2106/JBJS.D.02942. PMID: 16085630.
- John A Herring. Tachdjian's Pediatric Orthopaedics: from the Texas Scottish Rite Hospital for Children. 5th edition. 2014. Elsevier Saunders: Philadelphia ISBN: 978-1-4377-1549-1.