Fibrous Dysplasia Management

Craniofacial FD
- Vision and hearing assessment annually\(^1\)
- Baseline and periodic head CTs\(^2\)
- Avoid surgery in absence of vision and hearing deficits\(^3\)
- Encourage low impact cardiovascular activity
- Physical therapy to optimize function and attenuate loss of mobility.
- Shoe inserts for correction of leg length discrepancy
- Monitor for scoliosis; surgical fixation if Cobb angle > 50 degrees\(^4\)
- Consult with orthopedic surgeon experienced in FD as needed for fracture, severe deformity

Axial and Appendicular FD
- Focal and/or acute pain
  - Evaluate for acute or impending fracture
- Diffuse and/or chronic pain
  - Ensure appropriate treatment for hypophosphatemia\(^5\)
    - NSAIDs and supportive care
    - Bisphosphonates for persistent, moderate to severe pain\(^6\)
      - Pamidronate: children 0.5 mg/kg, adults 60-90 mg
      - Zoledronic acid: children 0.075 mg/kg, adults 4-5 mg
      - Repeat dosing intervals determined by symptoms\(^7\)

Bone pain
- Phosphorus: 15-60 mg/kg/day, divided in 4-5 doses
- Calcitriol: 15-60 ng/kg/day, divided bid
- Monitor for hypercalcheitia, hypercalcemia, GI upset

FGF23-mediated hypophosphatemia
- Patients should be evaluated yearly by a neuro-ophthalmologist; less frequently once stability is demonstrated. Patients with evidence of optic neuropathy referred to an experienced craniofacial surgical team. \(^2\)
- Repeat head CT approximately every 5 years, or sooner if vision or hearing deficits develop. \(^3\)
- Optic nerve encasement is common and usually asymptomatic. Prophylactic optic nerve decompression in the absence of optic neuropathy is contraindicated.\(^4\)
- Scoliosis may be progressive and potentially fatal in severe cases. All patients with scoliosis should be followed regularly by an orthopedic surgeon.\(^5\)
- Inadequately treated hypophosphatemia may significantly worsen bone pain, and must be addressed before considering bisphosphonates.\(^6\)
- Bisphosphonates have not been shown to affect disease progression, and use should be limited to treatment of FD-related bone pain. \(^7\)

References

Legend
CT = computed tomography; FD = fibrous dysplasia; PP = precocious puberty