Fibrous Dysplasia Evaluation
H&P to identify: limp, bone pain, fractures, limb length discrepancy.¹

Age <5 years²:
- High clinical suspicion for significant FD
  - Skeletal survey
  - Vision and hearing evaluation
  - Serum phos., TRP³
  - ⁹⁹Tc-MDP bone scan at age 5 years⁴

- Low clinical suspicion for significant FD
  - Monitor clinically
    - ⁹⁹Tc-MDP bone scan

Age ≥5 years:
- Abnormal ⁹⁹Tc-MDP bone scan
  - Significant FD⁵
    - Baseline skeletal survey
    - Baseline head CT for craniofacial FD
    - Serum phos., TRP

- Normal ⁹⁹Tc-MDP bone scan
  - Trivial FD
    - Consider baseline XR of affected area(s)
  - Low likelihood for significant FD
    - Monitor clinically.⁶

¹Performed at initial presentation in all patients suspected of having MAS. ²Areas of clinically significant FD will be apparent on bone scan by age 5 years. Prior to age 5, a normal Tc99 does not rule out the possibility of significant FD. ³TRP = tubular reabsorption of phosphate = 1 – [(Uphos/Sphos)*(Scr/(Ucr*1000))]. Calculated from a spot urine collection. FGF23-mediated phosphate wasting is associated with high FD burden; may worsen during rapid skeletal growth, and improve or resolve in adulthood as FD becomes less active. ⁴Consider performing ⁹⁹Tc-MDP bone scan in children < 5 years regardless of clinical suspicion for bone disease in instances where establishing the diagnosis of MAS may alter management – i.e. patients for whom diagnostic surgery is being considered, such as oophorectomy or skeletal biopsy. ⁵Significance of FD is determined by both the amount and location of affected bone. Clinically significant disease is frequently associated with the craniofacial area, proximal femurs and spine. ⁶A normal ⁹⁹Tc-MDP bone scan at age 5 years or older effectively rules out clinically significant FD, and no further radiologic monitoring is required.

References

Legend
⁹⁹Tc-MDP = technetium-99 conjugated with methylene diphosphonate; CT = computed tomography; FD = fibrous dysplasia; H&P = history & physical exam; TRP = tubular reabsorption of phosphate; XR = x-ray