**Clinical Practice Guideline for Management of Patients with Fragile Bones**

*Note, this guideline EXCLUDES NICU and Oncology patients*

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**Fracture Risk Factors**

1. Immediate High Risk:
   - History of Osteogenesis Imperfecta
   - \( \geq 1 \) vertebral fractures occurring in the absence of local disease or high energy trauma
   - Abnormal DEXA (Z score \( \leq -2.0 \)) AND significant fracture history (\( \geq 2 \) long bone fractures before 10 years of age or \( \geq 3 \) long bone fractures before 19 years of age)*
   - Chronic multiple joint contractures
   - Prematurity considered a risk factor up to 2 years of age, if birth weight \( \leq 1500 \) g and/or gestational age \( \leq 28 \) weeks
   - *Combined risk factors = Diagnosis of osteoporosis

2. Present on Admission or Acquired While Hospitalized:
   - Known low bone density (by DEXA)
   - Duchenne/Muscular Dystrophies
   - Wheelchair bound or non-ambulatory for \( \geq 6 \) months
   - Intubated and/or paralyzed for \( \geq 2 \) weeks
   - Cerebral palsy (specifically, spastic quadriplegic with GMFCS level 5)
   - Myelomeningocele/Spina bifida (specifically, thoracic myelo)
   - Parenteral nutrition \( \geq 2 \) consecutive months
   - Failure to thrive, poor growth, eating disorder, malabsorption disorder
   - Taking medications that contribute to fractures, special considerations for patients with chronic kidney dysfunction and congenital heart disease

3. Medications
   - Anticonvulsants
   - Glucocorticoids
   - Methotrexate: dose dependent and/or duration of medication
   - Loop diuretics
   - L-thyroxine suppressive therapy
   - GnRH-prolonged use
   - Medroxyprogesterone acetate

4. Discharge Recommendations
   - For at-risk patients discharged within 7 days and for patients, in general, who are hospitalized for more than 7 days send the following recommendations to the patient's Primary Care Provider:
   - Consider Endocrinology consult
   - Consider Physical Therapy consult
   - Obtain 25(OH)D level
   - Referral for DXA scan for children \( \geq 4 \) years old

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Recommended Labs with normal results
These values are for Children’s Healthcare of Atlanta labs, values can be different if sending to other labs

<table>
<thead>
<tr>
<th>LAB Test</th>
<th>AGE</th>
<th>Female</th>
<th>Male</th>
</tr>
</thead>
<tbody>
<tr>
<td>ALK Phos (U/L)</td>
<td>0-15 do</td>
<td>90-273</td>
<td>153-518</td>
</tr>
<tr>
<td></td>
<td>15 do - &lt;1 yo</td>
<td>134-518</td>
<td>36-156</td>
</tr>
<tr>
<td></td>
<td>1 yo - &lt;10 yo</td>
<td>156-369</td>
<td>141-460</td>
</tr>
<tr>
<td></td>
<td>10 yo - &lt;13 yo</td>
<td>62-280</td>
<td>127-517</td>
</tr>
<tr>
<td></td>
<td>13 yo - &lt;15 yo</td>
<td>54-128</td>
<td>89-365</td>
</tr>
<tr>
<td></td>
<td>15 yo - &lt;17 yo</td>
<td>48-95</td>
<td>59-164</td>
</tr>
<tr>
<td>Calcium (mg/dL)</td>
<td>≥19 yo</td>
<td>50-136</td>
<td>8.5-11</td>
</tr>
<tr>
<td></td>
<td>1 yo - &lt;19 yo</td>
<td>8.9-10.4</td>
<td>8.5-10.1</td>
</tr>
<tr>
<td>Magnesium (mg/dL)</td>
<td>0-7 do</td>
<td>1.2-2.6 mg/dL</td>
<td>1.6-2.4</td>
</tr>
<tr>
<td></td>
<td>7 do – 1 mo</td>
<td>1.6-2.4</td>
<td>16.2-2.6</td>
</tr>
<tr>
<td></td>
<td>1 mo – 2 yo</td>
<td>1.4-2.6</td>
<td>16.2-2.6</td>
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<tr>
<td></td>
<td>2 yo – 6 yo</td>
<td>1.5-2.4</td>
<td>26.2-2.4</td>
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<tr>
<td></td>
<td>6 yo – 10 yo</td>
<td>1.6-2.3</td>
<td>36.2-2.3</td>
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<tr>
<td></td>
<td>10 yo – 14 yo</td>
<td>1.6-2.2</td>
<td>46.2-2.2</td>
</tr>
<tr>
<td>Phosphorus (mg/dL)</td>
<td>0-14 do</td>
<td>5.6-10.5</td>
<td>7.4-10.0</td>
</tr>
<tr>
<td></td>
<td>15 do - &lt;1 yo</td>
<td>4.8-8.4</td>
<td>7.4-10.0</td>
</tr>
<tr>
<td></td>
<td>1 yo - &lt;5 yo</td>
<td>4.3-6.8</td>
<td>7.4-10.0</td>
</tr>
<tr>
<td></td>
<td>5 yo - &lt;13 yo</td>
<td>4.1-5.9</td>
<td>7.4-10.0</td>
</tr>
<tr>
<td></td>
<td>13 yo - &lt;16 yo</td>
<td>3.2-5.5</td>
<td>7.4-10.0</td>
</tr>
<tr>
<td></td>
<td>15 yo - &lt;19 yo</td>
<td>2.9-5.0</td>
<td>7.4-10.0</td>
</tr>
<tr>
<td></td>
<td>≥19 yo</td>
<td>2.5-4.9</td>
<td>7.4-10.0</td>
</tr>
<tr>
<td>GGT (U/L)</td>
<td>&lt;15 do</td>
<td>23-129</td>
<td>10-21</td>
</tr>
<tr>
<td></td>
<td>15 do - &lt;1 yo</td>
<td>10-21</td>
<td>10-21</td>
</tr>
<tr>
<td></td>
<td>1 yo - &lt;11 yo</td>
<td>10-21</td>
<td>10-21</td>
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<tr>
<td></td>
<td>11 yo - &lt;19 yo</td>
<td>10-21</td>
<td>10-21</td>
</tr>
<tr>
<td></td>
<td>≥19 yo</td>
<td>12-43</td>
<td>15-73</td>
</tr>
<tr>
<td>25-Vitamin D (ng/mL)</td>
<td>0 - &lt;19 yo</td>
<td>&lt;20: Deficient 20-30: Insufficient &gt;30: Sufficient</td>
<td></td>
</tr>
<tr>
<td>1, 25 Vitamin D (pg/mL)</td>
<td>ALL</td>
<td>19.9-79.3</td>
<td></td>
</tr>
<tr>
<td>PTH (pg/mL)</td>
<td>ALL</td>
<td>8.5-77.1</td>
<td></td>
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<tr>
<td>Urine Calcium/Creatinine Ratio (recommendations per Nephrology)</td>
<td>&lt;6 mo</td>
<td>&lt;0.8</td>
<td></td>
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<tr>
<td></td>
<td>6-12 mo</td>
<td>&lt;0.6</td>
<td></td>
</tr>
<tr>
<td></td>
<td>12-24 mo</td>
<td>&lt;0.4</td>
<td></td>
</tr>
<tr>
<td></td>
<td>≥24 mo</td>
<td>&lt;0.2</td>
<td></td>
</tr>
</tbody>
</table>

*Additional Recommendations for DEXA scan

General:
• Please include Height Age if patient is <5% as the Z-score will be falsely 'low' due to bone size-particularly pertinent to Osteogenesis Imperfecta and Cystic Fibrosis patients
• Height age can be designated in the order section as DEXA does not automatically calculate it
• If in doubt, DEXA can be done for both Chronological Age and Height Age

Site-specifics:
• Total body less head (TLBH) -Not in patients with severe contractures and/or hardware that precludes accurate positioning and/or analysis
• Spine ->3 years of age
• If needed and child <3 years of age, BMD can be hand plotted for estimated Z-score
• DO NOT order if spine fusion or spine rodding

Location for Scan:
If patient is on a stretcher or wheelchair bound, scan needs to be completed at Scottish Rite as the room at Egleston cannot accommodate

**Screening Evaluation and Treatment**

- **ALK Phosphatase**: If high or low compared to age/gender appropriate norms, then consult Endocrine
- **Serum Calcium**: If high, consult Endocrine; if low:
  - Correct Magnesium, if indicated
  - Obtain ionized Calcium and/or serum Albumin to determine available calcium
  - For emergent treatment, administer IV calcium
  - Central access preferred; PIV only in emergency
  - If Calcium persistently low or additional tests become abnormal, consult Endocrine
- **Serum Phosphorus**: If high or low compared to age/gender appropriate norms, then consult Endocrine and/or Renal, as indicated
- **Vitamin D**: Subspecialty team to manage and supplement per condition; target level >30ng/mL
- **Urine Calcium/Creatinine**: If abnormal, consult Nephrology
- **Skeletal Survey**: Use to determine extent of multiple fractures, in particular in suspected NAT setting and for cases of genetic bone disease
- **Thoracic Lumbar AP lateral spine films**: • Ordered by Endocrine, Genetics, or Provider
  - Consider if patient on steroids for >2 years and/or DEXA Z-score < -2.0
  - If on medications listed on page 1 and/or pathological fracture
  - Spine bone mineral density (BMD) is low if DXA Spine Z-score < -2.0 or history of high dose steroids; scan recommended yearly
  - Adjust for height for any patient with significant short stature
  - Consider Endocrine consult if any questions on how to order
  - Consider consult with Child Life and/or Physical Therapy to be present for scan
- **DEXA Scan***

- *Additional Recommendations for DEXA scan*
  - General:
    - Please include Height Age if patient is <5% as the Z-score will be falsely 'low' due to bone size-particularly pertinent to Osteogenesis Imperfecta and Cystic Fibrosis patients
    - Height age can be designated in the order section as DEXA does not automatically calculate it
    - If in doubt, DEXA can be done for both Chronological Age and Height Age
  - Site-specifics:
    - Total body less head (TLBH)
    - Not in patients with severe contractures and/or hardware that precludes accurate positioning and/or analysis
    - Spine
      - >3 years of age
      - If needed and child <3 years of age, BMD can be hand plotted for estimated Z-score
      - DO NOT order if spine fusion or spine rodding
    - Distal Radius
      - >12 years of age
      - Good site in children with hardware and/or severe contractures
      - If hardware or contractures in left arm, then you can scan OTHER arm and designate in the order

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