Clinical Practice Guideline for Management of Patients with Fragile Bones

Final: 12/2019
UPDATED: 9/23/21

***Note, this guideline EXCLUDES NICU and Oncology patients**

Assess patient for total number of Fracture Risk Factors[1]

≥1 Immediate High Risk[2] OR ≥2 Present on Admission[3,4]

Yes

Fracture Prevention
- Flag on chart
- Room Signage:
  - **Outside**: Handle with care, BP only if ordered or warranted by condition
  - **Inside:** Handle with care, BP only if ordered or warranted by condition. Consider manual BP to ensure lower cuff inflation (extremity with a rod is safest for BP)
- PT/OT consult
- IV & Handling Precautions

Handling Precautions
- Never push, pull, twist, lift, bend or apply pressure
- Avoid trying to turn head
- Handle gently with slow movements, supporting the head, torso, and buttocks evenly
- Consider manual BP to ensure lower cuff inflation (extremity with a rod is safest for BP)
- No circumferential holding when placing IV
- Do not use a tourniquet for IV or blood draws
- Allow parents to be with patient and handle as much as possible (including Radiology)
- Call MD or parents for questions about handling

Labs to consider:
- Complete Metabolic Panel
  - Alkaline Phosphatase
  - Serum Calcium
- Magnesium
- Serum Phosphorus
- GGT
- 25-Vitamin D
- 1,25 Vitamin D *(check prior to starting rocaltrol)*
- PTH
- Urine Calcium/Creatinine Ratio

Labs within normal limits

Yes
- Repeat labs as indicated by both diagnosis and patient age
- Intervention per primary/consult team

No
- Consider nutrition consult
- See page 2 for treatment recommendations[5]

No

Reassess in 2-4 weeks if[5]:
- Patient continues medications that contribute to factors[4]
- Immobilization continues or is initiated and lasts ≥2 weeks

[1]Fracture Risk Factors

[2]Immediate High Risk:
- History of Osteogenesis Imperfecta
- ≥1 vertebral fractures occurring in the absence of local disease or high energy trauma*
- Abnormal DEXA [Z score <-2.0] AND significant fracture history (≥2 long bone fractures before 10 years of age or ≥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 ≤1500g and/or gestational age ≤28 weeks

**Combined risk factors=Diagnosis of osteoporosis*

[3]Present on Admission or Acquired While Hospitalized:
- Known low bone density (by DEXA)
- Duchenne/Muscular Dystrophies
- Wheelchair bound or non-ambulatory for >6 months
- Intubated and/or paralyzed for ≥2 weeks
- Cerebral palsy (specifically, spastic quadriplegic with GMFCS level 5)
- Myelomeningocele/Spina bifida (specifically, thoracic myelo)
- Parenteral nutrition ≥2 consecutive months
- Failure to thrive, poor growth, eating disorder, malabsorption disorder
- Taking medications[4] that contribute to fractures, special considerations for patients with chronic kidney dysfunction and congenital heart disease

[4]Medications

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

[5]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 patients Primary Care Provider:
- Consider Endocrinology consult
- Consider Physical Therapy consult
- Obtain 25(OH)D level
- Referral for DXA scan for children ≥4 years old

Recommended Labs with normal results

These values are for Children's Healthcare of Atlanta labs, values can be different if sending to other labs

LAB Test	AGE	Female	Male
ALK Phos (U/L)	0-15 do	90-273	
	15 do - <1 yo	134-518	
	1 yo - <10 yo	156-369	
	10 yo - <13 yo	141-460	
	13 yo - <15 yo	62-280	127-517
	15 yo - <17 yo	54-128	89-365
	17 yo - <19 yo	48-95	59-164
	≥19 yo	50-136	
Calcium (mg/dL)	0 - <1 yo	8.5-11	
	1 yo - <19 yo	8.9-10.4	
	≥19 yo	8.5-10.1	
Magnesium (mg/dL)	0-7 do	1.2-2.6 mg/dL	
	7 do – 1 mo	1.6-2.4	
	1 mo – 2 yo	1.3-2.6	
	2 yo – 6 yo	1.5-2.4	
	6 yo – 10 yo	1.6-2.3	
	10 yo – 14 yo	1.6-2.2	
	≥14 yo	1.5-2.3	
Phosphorus (mg/dL)	0-14 do	5.6-10.5	
	15 do - <1 yo	4.8-8.4	
	1 yo - <5 yo	4.3-6.8	
	5 yo - <13 yo	4.1-5.9	
	13 yo - <16 yo	3.2-5.5	
	16 yo - <19 yo	2.9-5.0	
	≥19 yo	2.5-4.9	
GGT (U/L)	<15 do	23-129	
	15 do - <1 yo	10-127	
	1 yo - <11 yo	10-16	
	11 yo - <19 yo	10-21	
	≥19 yo	12-43	15-73
25-Vitamin D (ng/mL)	0 - ≥19 yo	<20: Deficient 20-30: Insufficient >30: Sufficient	
1, 25 Vitamin D (pg/mL)	ALL	19.9-79.3	
PTH (pg/mL)	ALL	8.5-77.1	
Urine Calcium/Creatinine Ratio *(recommendations per Nephrology)*	<6 mo	<0.8	
	6-12 mo	<0.6	
	12-24 mo	<0.4	
	>24 mo	<0.2	

LAB/Diagnostic Test	Screening Evaluation and Treatment
Alkaline 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
DEXA Scan*	•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

**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

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

Site-specifics:

- Total body less head (TBLH)
  - -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