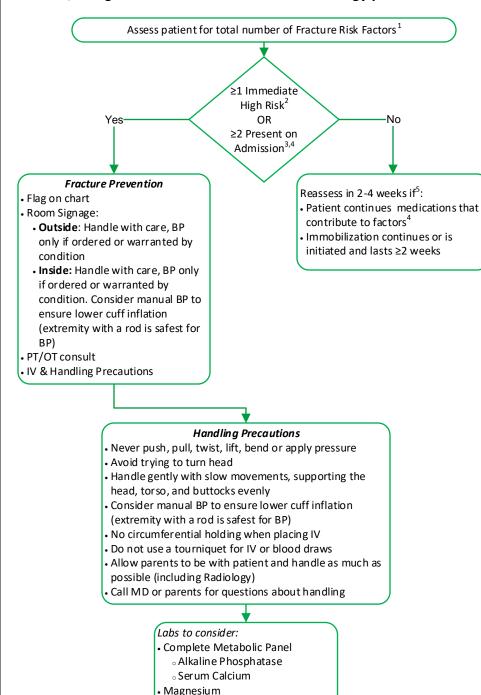
CLINICAL PRACTICE GUIDELINE FOR MANAGEMENT OF PATIENTS WITH FRAGILE BONES

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

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*Note, this guideline EXCLUDES NICU and Oncology patients



• Serum Phosphorus

• 25-Vitamin D

1,25 Vitamin D

GGT

¹Fracture Risk Factors

²Immediate 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

³Present 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

⁴Medications

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

Discharge Recommendations

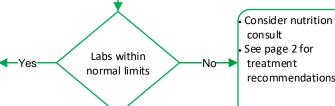
For at-risk patients discharged within 7 days and for patients, in general, who are hospitalized for more than 7 days send the following recommendations to the patients Primary Care Provider:

- Consider Endocrinology consult
- Consider Physical Therapy consult
- Obtain 25(OH) D level
- Referral for DXA scan for children ≥4 years old

Urine Calcium/Creatinine Ratio

Repeat labs as indicated by both diagnosis and patient age

Intervention per primary/consult team



(check prior to starting rocaltrol)

consult See page 2 for treatment recommendations⁵

Recommended Labs with normal results

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				
	<u>></u> 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 10-21					
	11 yo - <19 yo						
	≥19 yo	12-43	15-73				
25-Vitamin D (ng/mL)	0 - <u>≥</u> 19 yo	<20: Deficient 20-30: Insufficient >30: Sufficient					
1, 25 Vitamin D	ALL	19.9-79.3					
(pg/mL)							
PTH (pg/mL)	ALL	8.5-77.1					
Urine	<6 mo	<0.8					
• • • • • • • • • • • • • • • • • • • •	.5 1110	\U.U					
Calcium/Creatinine	6.40						
Ratio	6-12 mo	<0.6					
(recommendations per	12-24 mo	<0.4					
Nephrology)	>24 mo	<0	0.2				

MANAGEMENT OF PATIENTS WITH FRAGILE BONES Page 2 of 2								
Recommended Labs with normal results			sults	LAB/Diagnostic Test	Screening Evaluation and Treatment			
These values are for Children's Healthcare of			-	Alkaline Phosphatase	If high OR low compared to age/gender appropriate norms, then Consult Endocrine			
Atlanta labs, values can be different if sending			ending		If HIGH , Consult Endocrine If LOW :			
to other labs								
LAB Test	AGE	Female	Male		•Correct Magnesium, if indicated			
ALK Phos (U/L)	0-15 do	90-2	273		Obtain ionized Calcium and/or serum Albumin to determine			
	15 do - <1 yo	134-518		Serum Calcium	available calcium			
	1 yo - <10 yo	156-	-369		 For emergent treatment, administer IV calcium Central access preferred; PIV only in emergency If Calcium persistently low or additional tests become 			
	10 yo - <13 yo	141-	-460					
	13 yo - <15 yo	62-280	127-517					
	15 yo - <17 yo	54-128	89-365		abnormal, Consult Endocrine			
	17 yo - <19 yo	48-95	59-164	Serum Phosphorus	If high OR low compared to age/gender appropriate norms, then			
	≥19 yo	50-:	136	Serum Phosphorus	Consult Endocrine and/or Renal, as indicated			
Calcium (mg/dL)	0 - <1 yo	8.5	-11	Vitamin D	Subspecialty team to manage and supplement per condition;			
	1 yo - <19 yo	8.9-	10.4	Vitalilli	target level >30ng/mL			
	≥19 yo	8.5-10.1		Urine Calcium/Creatinine	If abnormal, Consult Nephrology			
Magnesium (mg/dL)	0-7 do	1.2-2.6	mg/dL	·	Use to determine extent of multiple fractures, in particular in suspected NAT setting and for cases of genetic bone disease •Ordered by Endocrine, Genetics, or Provider •Consider if patient on steroids for >2 years and/or DEXA Z score <-2.0			
	7 do – 1 mo	1.6-	-2.4	Skeletal Survey				
	1 mo – 2 yo	1.3-	-2.6					
	2 yo – 6 yo	1.5-	-2.4	Thoracic Lumbar AP lateral				
	6 yo – 10 yo	1.6-	-2.3	spine films				
	10 yo – 14 yo	1.6-	-2.2		•If on medications listed on page 1 and/or pathological fracture			
	≥14 yo	1.5-2.3			•Spine bone mineral density (BMD) is low if DXA Spine Z-score			
Phosphorus (mg/dL)	0-14 do	5.6-	10.5		<-2.0 or history of high dose steroids-scan recommended yearly			
	15 do - <1 yo	4.8-	-8.4	DEXA Scan*	•Adjust for height for any patient with significant short stature			
	1 yo - <5 yo	4.3-	-6.8	DEAA SCAIL	Consider Endocrine consult if any questions on how to order Consider Consult with Child Life and/or Physical Therapy to be			
	5 yo - <13 yo	4.1-	-5.9					
	13 yo - <16 yo	3.2-	-5.5					
	16 yo - <19 yo	2.9-	-5.0		present for scan			

*Additional Recommendations for DEXA scan

- 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
- 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