Pediatric Osteoporosis

Approach to Diagnosis and Treatment Considerations

The Future is Bright

Egyptian Academy Bone Health & Metabolic Bone Disease

Pediatric Vs Adult Osteoporosis

Where is the difference?

Adults: BMD

T-score ≤ -2.5 SD

Peak Bone Mass

Either Hip or Spine

Children:

Have not yet reached PBM

A "fracture threshold" for BMD has not been established

The 2013 International Society of Clinical Densitometry (ISCD) guidelines

Z-score, < −2

+

A clinically significant long bone fracture history

≥2 by age 10 years OR

≥3 by age 19 years

Fracture

the presence of

≥1 vertebral compression fractures occurring without major trauma or local disease

To avoid unnecessary investigations, fracture history assessed by questionnaire should be confirmed evaluating medical documentation

Pediatric Osteoporosis
Diagnosis

Adults: BMD

T-score ≤ -2.5 SD peak bone mass

Either Hip or Spine

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Have not yet reached PBM

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Pediatric Osteoporosis
Diagnosis

DXA

- the preferred method to asses bone mass during pediatric age
- Good reproducibility
- speed,
- reduced exposure to ionizing radiation,
- large availability of reference data

pQCT

- separately analyzes trabecular and cortical bone compartments,
- allowing the analysis of appendicular bone geometry, density, and strength,
- Enable the evaluation fat and muscle composition of the limbs.
- However, pQCT use is still limited by:
- the lack of standardized scanning protocols and
- Lack normative pediatric values

Pediatric Osteoporosis: Diagnosis
In Children with bone fragility:

DXA at lumbar spine & total body less head

Skull mineralization is not affected by nutrition or environment. Skull fractures should not suggest OP.

Diagnosis of OP in paediatrics can not be established on basis of DXA alone

Pediatric Osteoporosis: Diagnosis
Interpreting DXA measures

- DXA measures the total amount of BMC (g) contained within the skeletal region scanned and the 2-dimensional projected bone area (BA; g/cm²).
- The ration of BMC and BA expressed in units of g/cm2 is referred to areal BMD (aBMD).
- DXA provides aBMD at particular skeletal site but does not allow separate assessment of these measures within the trabecular and cortical bone compartments.

Pediatric Osteoporosis: Diagnosis
DXA reporting

The terms "Osteopenia / Osteoporosis" should not appear on the Pediatric DXA reports

"Low bone mass or BMD" is the preferred term for pediatric DXA reports when aBMD Z-score is < -2.0 SD

A child can function as its own control in longitudinal follow up. Therefore DXA at the beginning of the disease is recommended.

Pediatric Osteoporosis: Diagnosis
DXA technical considerations

DXA scans should avoid areas with metal implants, contractures or fractured vertebrae.

There is no age limit to perform DXA scan in children. Normal values for a whole body DXA are available from the age of 3-years.

If a follow up DXA is indicated, the minimum interval between scans is 6-12 months.

Pediatric Osteoporosis

Causes of Pediatric Osteoporosis

Primary Vs secondary causes

Pediatric Osteoporosis
High Risk Children

Primary bone disorders	**Chronic inflammatory diseases**
Osteogenesis imperfecta	Systemic lupus erythematosus
Idiopathic juvenile osteoporosis	Juvenile idiopathic arthritis
Osteoporosis-Pseudoglioma syndrome	Dermatomyositis
Homocystinuria	Inflammatory bowel disease
Ehlers-Danlos syndrome (type I)	Nephrotic syndrome
Marfan syndrome	**Immobility or decreased activity**
GSD type I	Post trauma
Juvenile/Early-onset Paget's disease	Cerebral palsy
Catabolic state/Inadequate nutrition/Malabsorption	Spinal muscular atrophy, Muscular dystrophy
Vitamin D deficiency	**Medications**
Malignancy - Acute lymphoblastic leukemia, Lymphoma	Anticonvulsant, Glucocorticosteroids, Heparin, Methotrexate (in oncology doses)
Cystic fibrosis	**Endocrine disorders**
Psychiatric eating disorders - Anorexia nervosa/Bulimia	Hypogonadism - Gonadal dysgenesis
Chronic malabsorption (e.g. Celiac disease)	Hyperthyroidism
Acquired immunodeficiency syndrome	Cushing syndrome
Female athlete triad disorder	Growth hormone deficiency
	Delayed puberty

Pediatric Osteoporosis
Primary Pediatric Osteoporosis

Osteogenesis Imperfecta

- OI present with:
- varying degrees of fracture,
- blue sclerae,
- dentinogenesis imperfecta,
- ligament laxity, and
- hearing impairment.

Idiopathic Juvenile OP

- . IJO typically presents before puberty and spontaneously remits after puberty.
- Characteristic features are:
- bone pain,
- walking difficulties, and
- metaphyseal and vertebral fractures.

Pediatric Osteoporosis
Secondary Pediatric Osteoporosis

Conditions with reduced bone formation:

- Immobilization or prolonged bed rest
- Medications: especially corticosteroids, diuretics and cyclosporine
- Burn injury
- Hepatic osteodystrophy with chronic cholestasis
- Aluminum toxicity in association with total parental nutrition (TPN) or renal osteodystrophy
-Prolonged total parental nutrition (TPN) use

Conditions associated with high bone resorption

- Corticosteroid-induced bone loss
- Immobilization or bed rest
- Juvenile Paget's
- Primary and secondary hyperparathyroidism
- Rickets due to vitamin D, calcium, or phosphorus deficiency
- Idiopathic juvenile osteoporosis
- IBD

Pediatric Osteoporosis

Conditions with with low BMD for age and gender without known etiology

- Sickle cell anemia
- - Thalassemia
- - Celiac disease
- - Type I diabetes
- - Myelomeningocele
- - Long-term oral anticoagulant therapy
- - Epilepsy
- - Acute lymphoblastic leukemia
- - Cystic fibrosis

Pediatric Osteoporosis

Management Algorithm

Who

- ≥1 vertebral compression Fracture. (>20% loss of height)
- Low Force – Long bone fracture
  ≥2 long bone fr. <10-years
  ≥3 long bones Fr. Any age up to 19-years
- Children at high risk of Osteoporosis (table 1) +fracture ◊

Exclude

- Other causes of fracture (e.g. rickets): Bone profile, vitamin D, X-ray wrists
- Systemic illness e.g. malignancy, inflammatory condition, neuromuscular disorders etc.

Assess aBMD Z-score

DXA Scan

- DXA: aBMD > -2 No Fracture → **No Osteoporosis◊**
- DXA: aBMD > -2 + Long bone fracture → **Osteoporosis**
- DXA: aBMD < -2 + Fracture → **Osteoporosis**

Consider

Factors to consider before treatment: age at diagnosis of osteoporosis, pubertal status, potential for spontaneous recovery, stature, growth, pubertal development, neurological, back examination/tenderness

Assess aBMD

DXA: aBMD > -2
No Fracture

DXA: BMD > -2 + Long bone fracture

DXA: aBMD < -2 + Fracture

No Osteoporosis

Osteoporosis

Is there underlying systemic condition / risk factor?

Yes
Secondary Osteoporosis

No
Possible Genetic bone fragility

Management

- Monitor bone health
- Spine radiograph
- Rectify/ manage risk factor
- Ensure proper nutrition
- Monitor new incidence of low force-long bone fracture

Osteoporosis

Management

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Possible spontaneous recover from Osteoporosis

Type I collagen mutation analyses

Sec. OP + Fr.

Yes
Min. risk factors
Nutrition
Manage secondary causes*
Monitor bone health to document spontaneous recovery, including increases in BMD Z-scores appropriate for height, reshaping of vertebral fractures, absence of new non-vertebral fractures

Positive: Osteogenesis Imperfecta

Negative: Genetic analyses for a mutation in one of the other bone fragility genes

No

Positive: Genetic Bone Fragility

Negative: Consider bone biopsy for signs of OI (hyperosteocytosis) or JO (thin osteoid seams, low bone turnover), if available

Start Treatment

Assess aBMD

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Children Osteoporosis Treatment

1.Stabilization Phase (usually last for 2-years)

Start intravenous bisphosphonate therapy with standard, published regimens (table 2) until the patient is clinically stable** (typically for a minimum of 2 years

2. Maintenance Phase

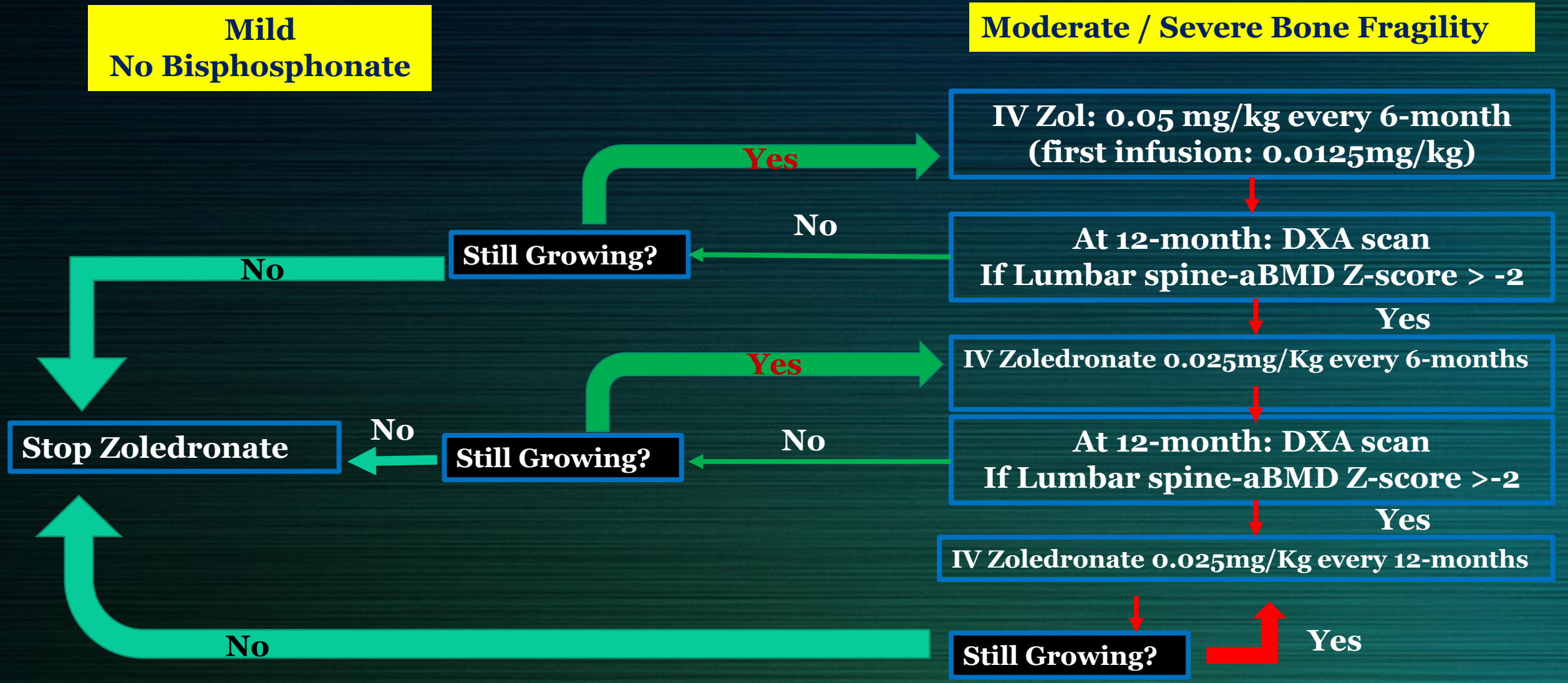
If risk factors resolve

Consider discontinuation of bisphosphonate treatment once the patient is clinically stable for at least 6 to 12 months

Ongoing risk factors (e.g. genetic bone fragility, chronic steroid therapy:

Consider continuing IV bisphosphonate treatment to the end of linear growth with titration to a lower dose with the goal to preserve the gains achieved during the stabilization phase and avoid over-treatment

Osteogenesis Imperfecta



Pediatric Osteoporosis
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Pediatric Osteoporosis
Treatment Outcomes

Subjective

-In symptomatic patients, treatment usually results in pain remittance within 2–6 weeks.

Improvement of bone and back pain

improvement in mobility

Objective

- in case of vertebral fracture, healing and subsequent bone remodeling should be visible at X-ray a few months after drug administration.
- eventual reshaping of vertebral fracture
- absence of new vertebral fracture in previously normal vertebral bodies,
- absence of additional loss of vertebral height at sites of previous fractures
- Absence of new nonvertebral fractures
- to stabilize the BMD Z-score trajectory of the patient at the follow-up DXA scan.

Pediatric Osteoporosis
Treatment Outcomes: Clinical Stability

Clinically stable includes:

Absence of new VF in previously normal vertebral bodies and absence of further loss of vertebral height at sites of previous fractures.

Reshaping of vertebral fractures.

Absence of new non-vertebral fractures, bone and back pain

Improved mobility, increases in spine BMD Z-score appropriate for height

Pediatric Osteoporosis
Treatment Outcomes: Clinical Stability

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Pediatric Osteoporosis
Stopping Osteoporosis Therapy

Clinical stability for 6-12 months

Treatment can be discontinued in patients whose underlying disease or risk factors resolve once they are clinically stable for 6–12 months

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Thank you

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