RICKETS

Rickets is a **disease of growing bone** that is due to unmineralized protein matrix (osteoid) at the **growth plates**, thus it occurs only in children before fusion of the epiphyses, whereas osteomalacia is present when there is inadequate mineralization of osteoid throughout the bone and occurs in children and adults. Rickets remains a persistent problem in the developing as well as developed countries.

Etiology

1/Vit D disorders: Nutritional, Congenital, Secondary, Vit D–dependent rickets (type 1 & type 2), and Chronic RF.

2/Calcium deficiency: Low intake or Malabsorption.

3/ Phosphorus deficiency: Inadequate intake, Disorders of Phosphatonin e.g. XL, AD & AR hypophosphatemic rickets, Hereditary hypophosphatemic rickets with hypercalciuria, Overproduction of phosphatonin.

4/Syndromes & diseases associated with rickets: Fanconi synd, Distal RTA & Dent disease.

Clinical manifestation

- General: FTT, listlessness, protruding abdomen, muscle weakness (especially proximal), delayed walking, waddling gait, fractures.
- Head: craniotabes, frontal bossing, delayed fontanel closure, delayed dentition with dental caries, craniosynostosis.
- Chest: rachitic rosary, Harrison groove, RTI and atelectasis.
- Back: scoliosis, kyphosis, lordosis.
- Extremities: enlargement of wrists and ankles, valgus or varus deformities, windswept deformity, anterior bowing of the tibia and femur, coxa vara, leg pain.
- Hypocalcemic symptoms: tetany, seizures, strider (due to laryngeal spasm).

Nutritional vitamin D deficiency:-

It is the **most common** cause of rickets globally. It most commonly occurs in infancy due to a combination of poor intake and inadequate cutaneous synthesis. Transplacental transport of vit D (mostly 25-D) typically provides enough vit D for the 1st 2 mo of life, unless there is severe maternal vit D deficiency.

Infants who receive formula receive adequate vit D, even without cutaneous synthesis. Because of the low vit D content of breast milk (especially if the mother was also vit D deficient), thus breast-fed infants rely on cutaneous synthesis or vit D supplements.

Diagnosis

X-ray of the wrist in AP view shows thickening of the growth plate with **fraying & cupping** of distal ends of the metaphyses. Other findings include coarse trabeculation of the diaphysis and generalized rarefaction.

Laboratory tests can be illustrated in the following table:-

Disorder	PTH	25-(OH)D	1,25-$(OH)_2D$	serum Ca	urine Ca	serum Pi	urin Pi
Vit D deficiency	↑	↓	↓, N, ↑	N, ↓	↓	↓	↑
VDDR, type 1	↑	N	↓	N, ↓	↓	↓	↑
VDDR, type 2	↑	N	↑↑	N, ↓	↓	↓	↑
Chronic renal failure	↑	N	↓	N, ↓	N, ↓	↑	↓
Dietary Ca deficiency	↑	N	↑	N, ↓	↓	↓	↑
Dietary Pi deficiency	N, ↓	N	↑	N	↑	↓	↓
XL, AD & ARHR	N	N	RD	N	↓	↓	↑
HHRH	N, ↓	N	↑	N	↑	↓	↑
Fanconi synd	N	N	RD or ↑	N	↓ or ↑	↓	↑
Tumor-induced	N	N	RD	N	↓	↓	↑

Alkaline phosphatase level is always increase in **all** types of rickets; whereas **PTH** level always increase in hypocalcemia, unless there is hypophosphatemia.

Treatment

Vit D can be given either as, **300,000-600,000 IU** orally or IM as 2-4 doses over 1 day *"stoss therapy"*, or as daily doses ranging from **2,000- 5,000 IU/day** over 4-6 wk (≈ 1 mo). Then it

should be followed by maintainance dose of daily vit D, 400 IU/day if <1 yr, 600 IU/day if >1 yr (typically given as multivitamin).

It is important to ensure that children should also receive adequate dietary **calcium and phosphorus** by milk, formula, and other dairy products.

Some children with **symptomatic hypocalcemia** can be given either IV calcium acutely, followed by oral calcium supplements, which can be tapered over 2-6 wk, or may be given IV or oral 1,25-D (calcitriol).

Prognosis

Laboratory tests should normalize rapidly, whereas radiologic

changes may heal within **few months**. Many of the bone malformations improve dramatically, but children with severe disease may have **permanent deformities** (which rarely may require surgery) and short stature.

Complication of rickets

1- respiratory infections ; bronchitis and bronchopneumonia are common , pulmonary atelectasis frequently associated with sever deformity of the chest .

2- anemia due to iron defiency or accompany infection

3- enteritis .

4- deformities of bone and dwarfism .

Prevetion

Most cases of nutritional rickets can be prevented by universal administration of a **daily multivitamin containing 400 IU of vit D to all breast-fed infants after 2 mo of age, and 600 IU/day for older children.**