



**MEDICAL UNIVERSITY - PLEVAN  
FACULTY OF MEDICINE**

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**Department of Pediatrics**

**Lecture № 9**

# RICKETS

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# RICKETS



Rickets



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# DETERMINATION

- **Rickets** is a disease of the growing bone that is unique to children and adolescents
- It is caused by a failure of osteoid to calcify in a growing person
- Failure of osteoid to calcify in adults is called **Osteomalacia**.
- **Rickets** is determined as an impaired mineralization of the bone matrix or osteoid tissue of the growing bone due to mineral deficiency, which lead to a delay of the bone growth and delay in the bone age
- Bone growth is increased by : growth hormone, thyroid hormone, insulin, sex hormones and decreased by glucocorticosteroids

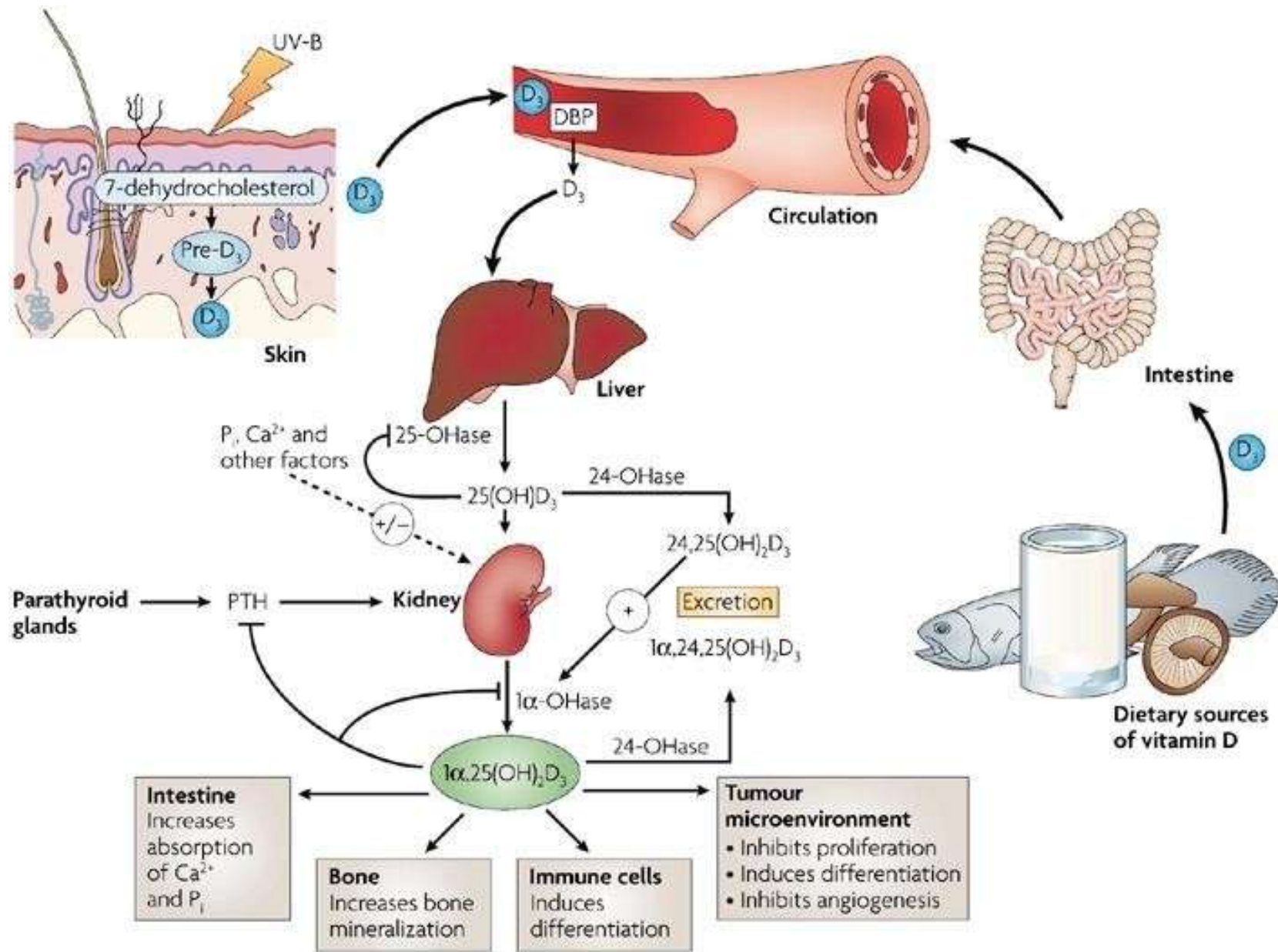


# VIT D3 FORMATION AND METABOLISM

- Vit D 3( Cholecalciferol) is formed in the skin from a derivate of ergocalciferol( vit D2) under the stimuli of Ultraviolet Blight.
- In the liver is the hydroxylation on the 25-th place, and than in kidneys - on the 1-st place
- Final product is called 1,25(OH)<sub>2</sub> D3- calcitriol
- Vit D3 function :
  - increase bone mineralization
  - Increased resorption of Ca and P in intestinal mucosa
  - Increased resorption of Ca in distal renal tubuli
  - Increased secretion of calcitonin

Daily need of vit D 3 is 400-1000 U/day for term babies and 800- 2000 U/day for preterm babies





# ETIOLOGY OF THE RICKETS

- Intensive growth - SGA babies, preterm babies, puberty
- Early infancy - after 2-d month
- Sex: male>female
- Season : winterbabies
- Race: blackbabies
- Malabsorption: celiac disease, CF, acute pancreatitis, etc.
- Therapy with antiseizure medications
- Increase needs
- Intake of caw milk



# PATHOGENETIC

- Disturbance between the bone growth and mineralization
- Impaired of endochondral ossification
- Osteoblast cells produce osteoid , which does not calcified

Pathway:

Low vit D ⑦ poor Ca and P from the intestines ⑦ l o w serum Ca ⑦ increased Parathormone ⑦ C a and P from the bones , increased alkalic phosphatase ⑦ osteoclast stimulation ⑦ Phosphaturia, hypophosphatemia, acidosis, ~~aminoaciduria~~



# RICKETS CLASSIFICATION HARRISON 1994

## 1. Deficiency of 1,25(OH)D<sub>3</sub> and calcipenia

- A. Deficiency of vit .D<sub>3</sub>
- B. Resistance of the peripheral organ : vit D dependent rickets ( receptor disease)

## 2. Deficiency of HPO<sub>4</sub> and phospopenia

- A. Increased loose of P ( hypophosphatemic rickets, De Toni - Debre- Fanconi, cystonosis, Lowe s-me, M. Wilson, RTA)
- B. Low P intake : preterm babies, total parental feeding





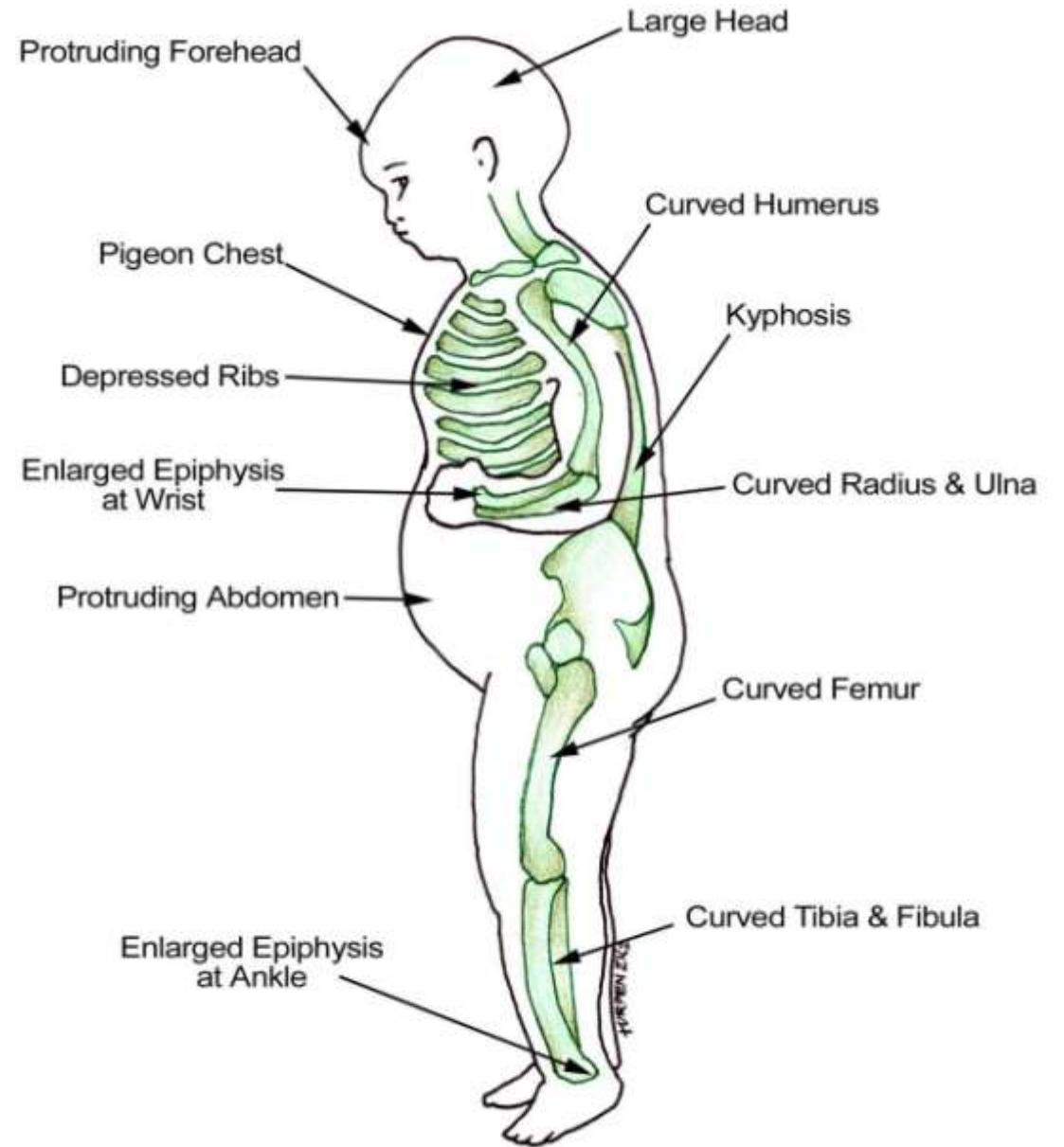
# CLINICAL SYMPTOMS

- **A. Central nervous symptoms** : early : impaired sleep, anxiety, sweating, hair loss .
- **B. Muscle hypotonia**
  - Decreased movement development
  - Froggy belly
  - Intestinal hypotonia and constipation
  - Respiratory muscle hypotonia ➔ pneumonia
- **C. Bone changes** : proximal - > distal direction
  - Osteomalacia : craniotabes
  - Osteoid hyperplasia: caput quadratum, rickets bracelet, etc.
  - Bone deformations: Harrison groove, bell chest, pectus carinatum/excavatum, scoliosis, genua valga, vara

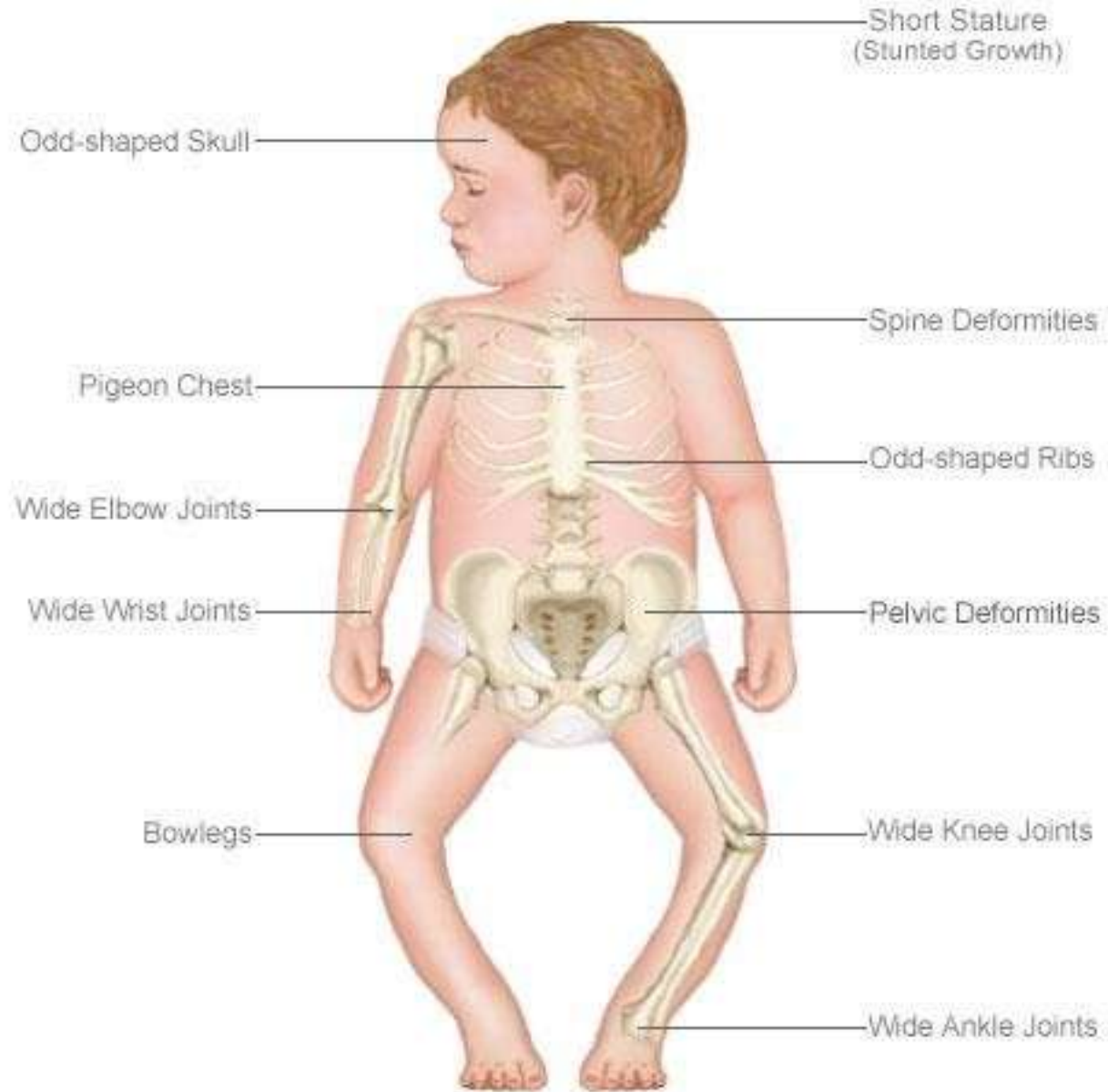


# SYMPTOMS

- Delayed growth
- Delayed motor skills
- Pain in the spine, pelvis and legs
- Muscle weakness
- Bowed legs or knock knees
- Thickened wrists and ankles
- Breastbone projection



## The Effects of Rickets





Knock knee



Bow leg



Windswept



Sabre tibia

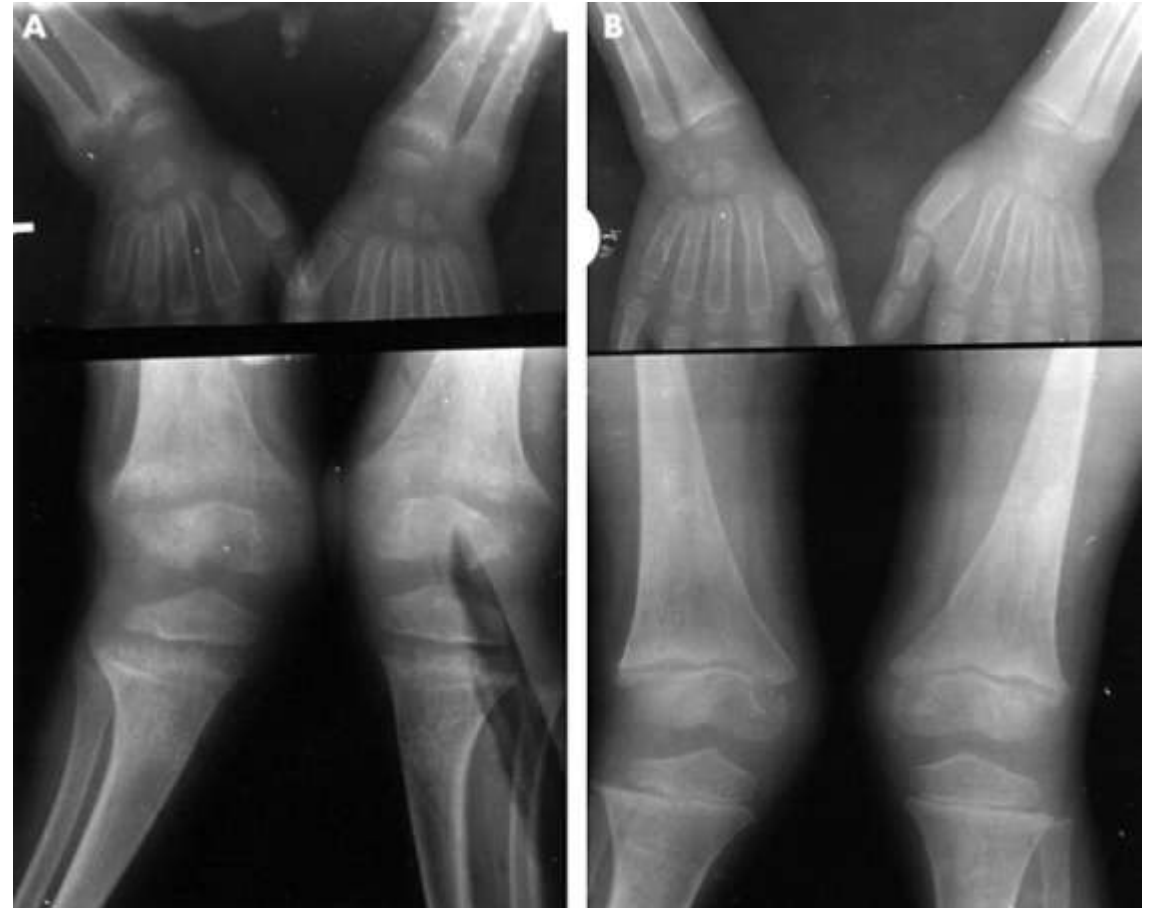
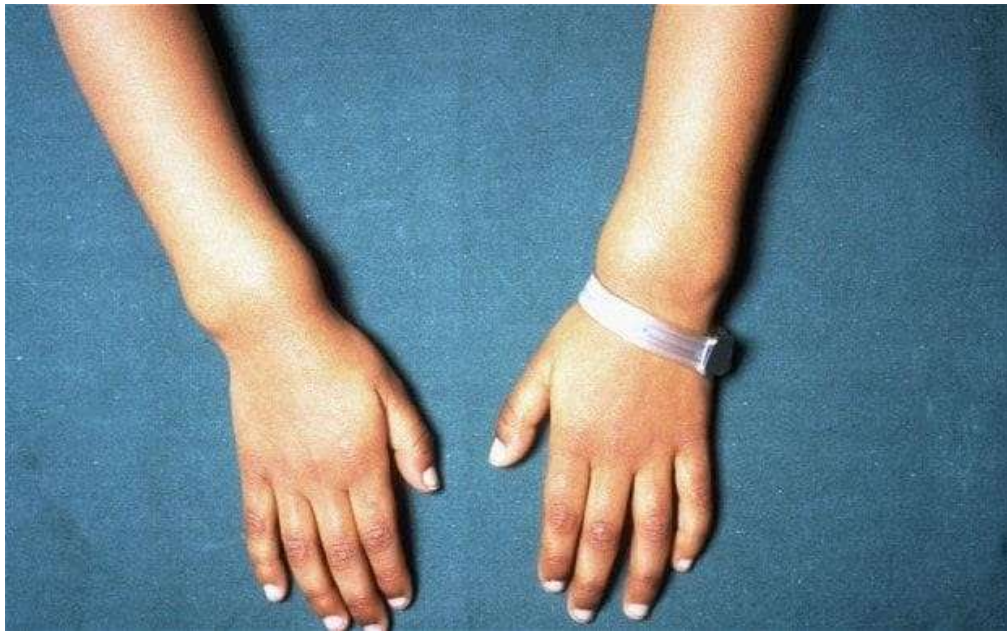


# DIAGNOSIS

- Season
- Age : early infant age
- Type of feeding
- Physical examination, typical signs
- High AP, normal or low calcium, low P
- X-ray of the wrist : osteoporosis, curly metaphyseal line, tea cup meta-epiphyseal zone



# BONE CHANGES



# TREATMENT

- ❖ Mild forms: 2000- 4000 U/day 6 weeks
- ❖ Moderate forms : 4000- 6000 U/day 6 weeks
- ❖ Severe forms : 6000 - 10 000 U/day for 6 weeks + sun exposure, massage, calcium rich food

Prophylactic therapy from 20-th day for term babies : 400-1000U, from 10 - th day for preterm babies : 1000 - 1600U/day





# DIFFERENTIAL DIAGNOSIS

- 1. **Vit D - dependent rickets** ( pseudo vitD deficiency) - Prader rickets
  - Type 1 - deficiency of 1-alfa hydroxylase in kidneys
  - Type 2- impaired tissue answer to vit D3

Clinical presentation is like a rickets

Lab. findigs : low Ca, slightly elevated P, high AP, X - ray changes

**Diagnosis: low Ca although vit D 3 treatment**





# DIFFERENTIAL DIAGNOSIS

- 2. Family hypophosphatemia (Vit D -resistant rickets)
  - X - linked, ARD
  - Defect of reabsorption of PO<sub>4</sub> in proximal tubuli
  - Impaired transformation of 25(OH) D into 25(OH)<sub>2</sub> in kidneys
  - Low P in serum ⑦ s h o r t stature
  - Bone deformations :X- , O- legs, “kitten walking”
  - Normal or low Ca, low serum P, high AP
  - **Diagnosis : Rickets appear after the active walking of the child, no muscle hypotony, short stature, low P, hyper - PO<sub>4</sub> uria**
  - Treatment : vit D high doses: 2000 U/kg/day + oral phosphate medication



# DIFFERENTIAL DIAGNOSIS

- **3. Hypophosphatasia - ARD**
  - X - ray for rickets, low AP
- **4. Rickets in hepatic disorders ( poor resorption of fatty vitamins)**
- **5. Rickets associated with tubular dysfunction**
  - Syndrome De Toni- Debre- Fanconi ( complex proximal tubulopathy) : low PO<sub>4</sub>, low K, acidosis
  - RTA proximal type
  - RTA distal type



# STAGES OF THE RICKETS

- 1. Stage: **Rachitis incipiens**: vegetative symptoms, central nervous symptoms, muscle symptoms, low Ca, normal P, high AP
- 2. Stage : **Rachitis florida** : skeletal changes ( from proximal to distal), from the skull trough chest , sternum, scoliosis, rickets wrist, legs “O”, “X”, normal Ca, low P, very high AP
- 3. Stage: **Rachitis residualis** :skeletal deformations, normal Ca, low P, high AP
- In severe forms : anemia, leukocytosis , hepatosplenomegaly, Ca:P from 2:1 to 3:1, very high AP



# RICKETS TETANIA (SPASMOPHILIA)

- Typical for spring
- Mostly observed in treated with vit. D infants with a fast recovery and Ca in the bones - latent or manifest tetany
- Latent tetany : Trusso or Hvosstec symptoms
- Manifest tetany : carpo- pedal spasmus, laryngospasmus
- Therapy : Ca gluconici i.m, i.v, + high doses vit .D



# HYPERVITAMINOSIS D

- It could be **acute** or **chronic** disease with **kidney** or **cardiac** involvement
- Pathogenesis: increased Ca reabsorption from intestines, Ca from the bones  
⑦ high serum Ca ⑦ > deposition of CaPO<sub>4</sub> into the skin, vessels, myocardium, CNS, kidneys, cornea, etc.
- Decreased secretion of ADH - polyuria, polydipsia, dehydration
- Increased blood pressure, nephrocalcinosis, renal failure, photophobia
- **Clinical symptoms** : anorexia, hypotonia, vomiting, constipation, poor appetite, seizures, high blood pressure, photophobia, pruritus, renal failure
- **Lab** : elevated Leu, high Ca, high urea, high vit. D, X - ray - osteoporosis
- **Therapy** : no Ca intake, i.v. glucosae, KCL, Furanthril, i.v. corticosteroids.



# RICKETS

Condition	Genetics	Ca	Phos.	Alk Phos	PTH	Vit D	1,25 (OH)VitD
Vitamin D Resistant Rickets (Hypophosphatemic)	X linked dominant	-	↓	↑	-	-	
Vitamin D Deficiency Rickets (Nutritional)	Nutritional	↓	↓	↑	↑	↓	
Type I Vitamin D Dependent	Auto. Recessive	↓	↓	↑	↑		↓ ↓
Type II Vitamin D Dependent	Auto. Recessive	↓	↓	↑			↑ ↑
Hypophosphatasia	Auto. Recessive	↑	↑	↓ ↓	-	-	
Renal Osteodystrophy	Renal Disease	↓	↑	↑	↑		
Hyperparathyroidism	90% adenoma	↑	↓	↑	↑		



# EVOLUTION

- 1 stage: Rachitis incipiens: low Ca, normal P
- 2 stage : Rachitis florida : low Ca, increased P H ⑦ normal Ca, but phosphaturia ⑦  
low serum P, acidosis
- 3 stage : Rachitis residualis : low Ca, low P

Phase	Early	Top	Late
AP	normal	high	Very high
PO4	normal	low	Very low
Ca	low	normal	low



# COMPLICATIONS

- Failure to grow
- An abnormal curved spine
- Bone deformities
- Dental defects
- Seizures





# PREVENTION OF THE RICKETS

- Exposure to sunlight During most seasons, 10 to 15 minutes of exposure to the sun near midday is enough.
- Because of skin cancer concerns, infants and young children are warned to avoid direct sun or to always wear sunscreen and protective clothing.
- Foods with vitamin D naturally – fatty fish ( salmon and tuna), fish oil and egg yolks
- Infant formula
- Cereal
- Bread
- Milk, but not foods made from milk, such as some yogurts and cheese
- Orange juice
- For pregnant women are recommended vitamin D supplements.



**THANK YOU FOR YOUR ATTENTION!**

