الكلية : الطب

القسم او الفرع :طب الاطفال وحديثي الولادة

المرحلة: الخامسة

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اسم المادة بالغة العربية : طب الاطفال وحديثي الولادة

اسم المادة باللغة الإنكليزية : طب الاطفال وحديثي الولادة

اسم المحاضرة الأولى باللغة العربية: الكساح

اسم المحاضرة الأولى باللغة الإنكليزية : Rickets

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Rickets

Definition ;failure of meniralization of the osteoid tissue in a growing bone due either to deficiency (nutritional rickets) or defective metabolism of vitamin D .

Rickets of three major groups ;

Hypocalcemic rickets

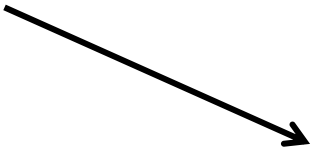
#Hypophosphatemic rickets

#End organ resistance to vit d

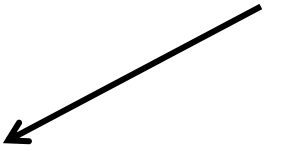
Pathophysiology of rickets ;

Vit D deficiency or

defective metabolism

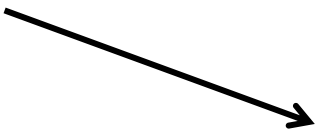


low calcium or phosphorus

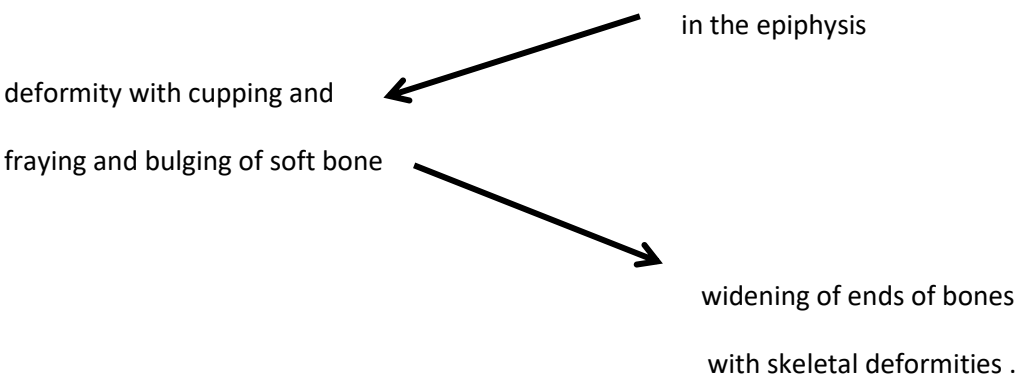


defective mineralization

of the new osteoid tissue



soft osteoid tissue



Hypocalcemic rickets ;

the body trys to elevate the serum calcium by the excessive secretion of parathormone (PTH) with parathyroid hyperplasia (hyperparathyroidism).

The PTH will mobilize the bone calcium and phosphate to increase the serum calcium and reduces the renal tubular phosphate reabsorption causing phosphate urea .so the patient will have normal or low serum calcium ,low phosphate and high serum PTH , tetany ,convulsion ,rachitic myopathy ,angular bowing of legs ,harrison's sulcus , rachitic rosary ,chest deformties and enimal defects of the teeth .

Urine exam shows aminoaciduria ,phosphaturea , bicarbonateurea and glucoseurea .

In severe cases metabolic acidosis (rta) may be associated .

Treatment by calcium and vit D .

Examples;

-neutritional rickets .

-hepatic rickets ,by effecting the metabolism and absorption of vit D.

-rickets due to drugs (anticonvulsant), by stimulation of hepatic cytochrome 450 , so converts the vit D to inactive metabolite .

- vit D dependant rickets (type 1)

-renal osteodystrophy .

Hypophosphatemic rickets ;

When the hypophosphatemic rickets is not associated with a mechanism leading to hypocalcemia ,the patient will have no rachitic rosary ,harreson's sulcus , or chest deformties ,and no tetany or convusions

The teeth defect is in the dentine not in the enimal , with defective intra globular dentine reccurent tooth abcesses .

The serum phosphate is low , the calcium is normal and the parathormone (PTH) is normal .the urine shows phosphate urea with no amino acidurea ,bicarbonateurea or glucoseurea .

Note\\ When the hypophosphatemic rickets associated with hypocalcemia and hyperparathyroidism eg .fanconi syndrome , the child will have the signs of hypocalcemic rickets as convulsions , tetany ,rachitic rosary ect .with hypophosatemia ,and urinary bicarbonateurea , phosphateurea ,calciurea and aminoaciduria .

Treatment by vit D and oral phosphate

Examples;

-vit D resistant ricket

-oncogenic ricket ;neurofibromatosis and mesenchymal tumers liberates substances that suppresses the renal 1a.hydroxylase enzyme causing phosphateuric rickets.

-phosphate deficiency or malabsorption .

-fancony syndrome(cystenosis ,tyrosenosis ,galactosemia,lowe syndrome and acquired types).

-rickets of renal tubular acidosis (RTA), the proximal type 2

Nutritional rickets

Common at 6 to 24 months age . more common in prolonged breast feeding without adding solid food .

Causes;

-low intake .

-inadequate direct sunlight exposure(uv light doesnot pass the window glass)

-Prolonged breast feeding.

-dark skin (nigro).

-steatorrhea ,celiac desease ,chronic pancreatitis with fat malabsrorption .

-Drugs interfere with vit D metabolism (anticonvulsants)

Clinical manifestations;

-skull ;excessive sweating , craniotabies ,large fontanel with delayed closure ,large skull , caput qudratum , delayed teething and frequent tooth caries .

-Thorax; chest deformity , harrison's sulcus , rachitic rosary .

- spinal colomn ;scoliosis ,kyphosis ,lordosis ,compression of vertebra.

-stature ;rachitic dwarfism .

-pelvis ; deformity , contracted pelvis .

-ligaments; lax ligaments , over extensibility of joints .

-extremeties ;widening of joints ,bow leg , knock knee , anterior convexity of femur and tebia ,coxavara ,bone pain and green stick fracture .

-muscles ;poor development ,hypotonia , pot belly abdomen ,delayed standing and walking ,tetany , carpopedal spasm , laryngospasm ,convulsions .

-infections; reccurent chest infections .

Diagnosis;

Serum calcium low or normal , serum phosphorus always low .

High serum alkaline phosphatase .

Low serum $25(oh)_2D3$

-GUE shows aminoaciduria ,phosphaturea bicarbonateurea with alkaline urine and glucosurea in severe rickets .

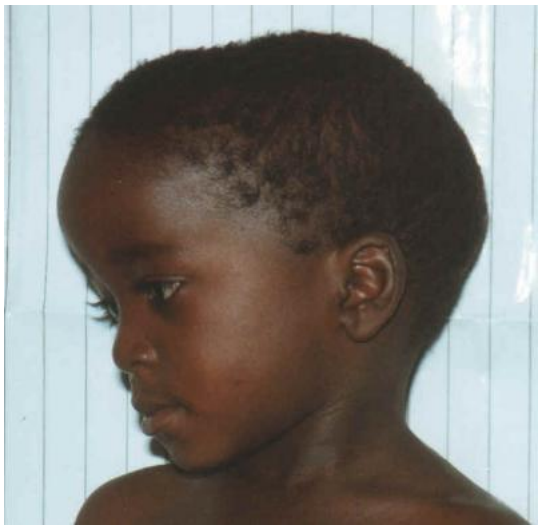
X-ray ;shows widening of the ends of the long bones ,cupping fraying ,decreased bone density ,increasing of the joint space ,delayed bone age , double contour of bones and may be pathological fracture.

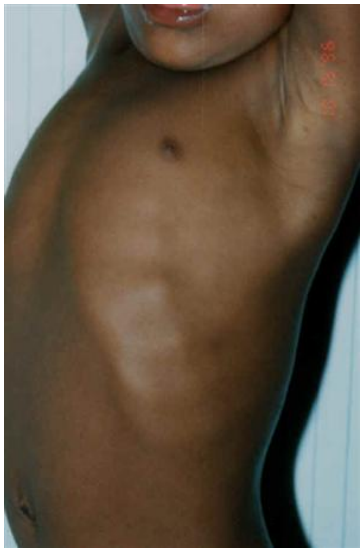
Treatment; oral cholecalciferol (D3)2000-6000 iu \day (50-150 ug) or 1.25(oh)2d3 (calcitriol) 0.5-2 ug\day orally . or by giving single dose of vit d 600,000 iu orally or im .

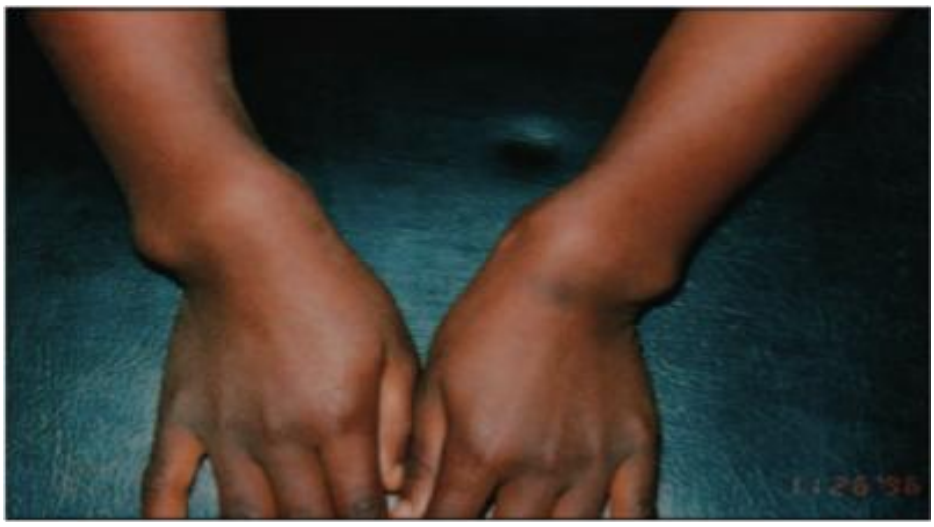
(Healing seen 2-4 weeks on x ray .)

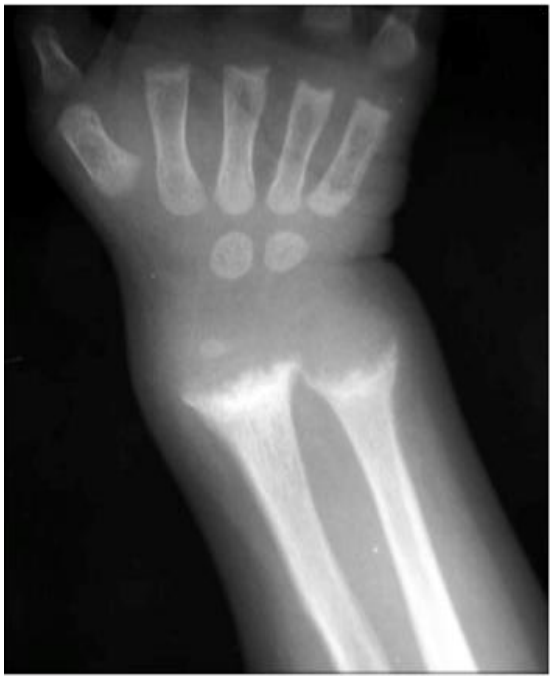
After complete healing ,continue with vit D rich foods and promote exposure to sunlight .

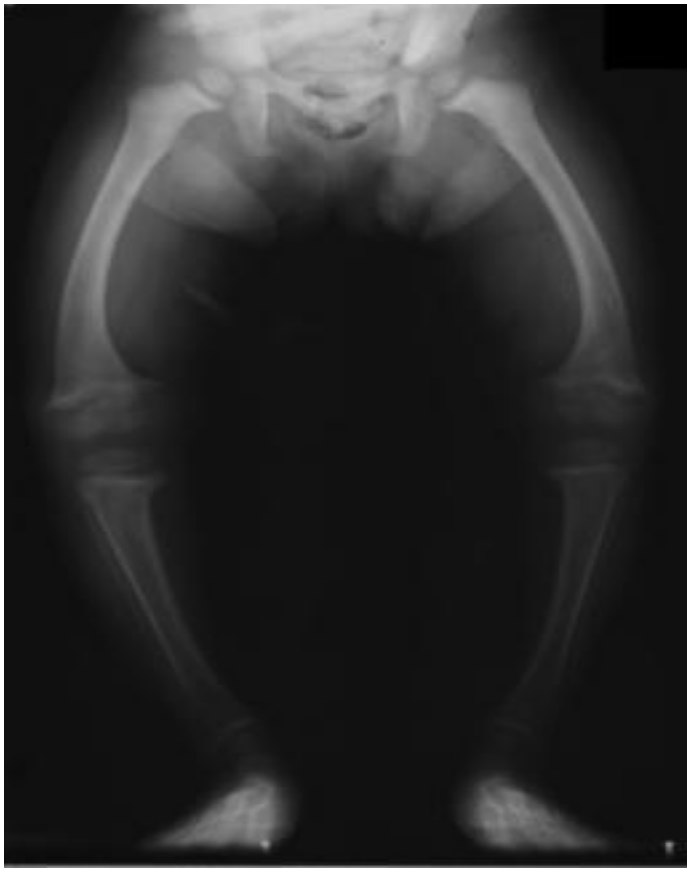
If no healing by x ray occure on this treatment ,indicates resistant type of rickets ..











Vit D dependent rickets 1 ;

Autosomal recessive AR inheritance ,hypocalcemic rickets ,due to deficiency of kidney 1.a hydroxylase.

Presented early during the first 3-6 months ,with severe rickets ,with signs of hyperparathyroidism and hypocalcemia as rachitic rosary ,harrison's sulcus ,chest deformity ,tetany ,convulsions ,rachitic myopathy ,enimal changes of teeth.

Diagnosis; low serum ca and ph and 1.25(oh)2D3 ,high alk phosphatase and parathormone .

GUE shows aminoacidurea ,phosphateurea ,glucoseurea and bicarbonateurea .

X ray shows signs of severe rickets .

Treatment ;massive doses ; 200,000 iu vit D .treat the metabolic acidosis ,give calcium containing food like milk .

Vit D –resistant rickets;

x-linked dominant ,hypophosphatemic rickets due to deficiency of 1.ahydroxylase+ defective phosphate reabsorption in the renal tubules ,leads to phosphate urea ,hypophosphatemia and rickets.

Patient presented late in the second year after starting walking ,with progressive rounded bowing of legs short stature ,waddling gait ,genu vulgus or varus ,tooth dentine defect ,((**absence of signs of hypocalcemia and hyper parathyroidism as tetany,convulsions ,rachitic rosary …ect)) .**

Diagnosis;

Normal s. ca , <u>low ph ,high alk phosphatase</u> ,normal parathormone(parathyroid hormone)

GUE; shows phosphaturea .

X-ray ;shows cupping and fraying in the knee ,ankle ,and wrist joints .

treatment ;0.5 -1 gm phosphate \day orally . and vit D 2000 iu \kg \day orally ..