RICKETS

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Vitamin D : TWO FORMS D2 & D3

Both presents in dietary supplement. D3 is naturally present in human skin in provitamin stage 7-dehydrocholesterol & by action of ultra violet changed to choleclaciferol & hydroxyleted by the liver , then both D2&D3 activated in the renal cortex to (1,25 dihydroxycholecalciferol) which function as hormone . Anti-rachitic functions include facilitation of intestinal absorption of Ca^{++} & Ph^{++} & of reabsorption of phosphorus in the kidney& direct effect on mineral metabolism of bone (deposition & reabsorption) in conjunction with parathormone &calcification play major role in homestasis of Ca &Ph in the body fluids &tissues .

Rickets

Signify : failure in mineralization of growing bone or osteoid tissue &characteristic early changes seen on X-ray at the end of long bones & evidence of demineralization also exists in the shafts .

Etiology :

1- inadequate direct exposure to ultraviolet rays in sun light , these rays do not pass through ordinary window glass .
2- inadequate intake of vitamin D .
3- deficiency may occur in unsupplemented dark skinned infants or in breast fed infants of mothers unexposed to sun light .
4- conditions that interfere with the metabolic conversion & activation of vit D such as hepatic & renal lesions, or conditions that disrupt Ca & Ph homeostasis .
5- children with disorder of absorption e.g. coelic disease & steatorrhea .
6- drugs ; anticovlusant therapy e.g. phenytoin &phenobarbitol ,glucocorticoids appear to be antagonistic to vit D in Ca++ transport .

Chemical pathology :

Serum Ca ++ usually normal but may be decreased .
Serum Ph++ normaly (4.5-6.5 mg /dl) but in rickets it decreased to (1.5-3.5mg/dl)
Serum alkaline phosphatase ; in normal children less than (200 i.u./dl) is elevated in mild rickets to mor than (500 i.u./dl)
Vit D deficiency is also accompanied by generalized aminoaciduria , decrease of citrate in bone & its increased urinary excreation & decreased ability of the kidney to make an acidic urine , phosphaturia &mellituria .
The parathyroid gland hypertrophy occur in rickets &urinary cyclic AMP increased .

Clinical manifestations:

Osseous changes of rickets can be recognized after several months of vit D deficiency .In breast fed infants whose mothers have osteomalasia rickets developed within 2months , florid rickets appear toward the end of first year & during the second year of life. Rickets developed rapid growth .

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Early signs of rickets :

1- craniotabes ; is due to thinning of outer table of the skull &detected by pressing firmly over the occiput &post .parietal bones, aping pong ball sensation will be felt . Craniotabes near suture line is a normal variant . low birth weight infants are prone to early rickets &craniotabes .
2- palpable enlargement of costochondral junctions rachitic rosary&thickening of the wrists &ankles are also early evidence of osseous changes .
3- increased sweating particularly around the head .

Advanced rickets ;

Skull ; craniotabes may disappear before the end of the first year though the rackitic process contine .The softness of the skull may result in flattening , may be permanent asymmetry of the head , anterior fontanel larger than normal & its closure may be delayed until after 2nd year of life , the central parts of parietal &frontal bones often thickened forming prominences or bosses ,which give box like appearance (caput quadratum) , the head may be larger than normal & may remain so for life . eruption of temporary teeth may be delayed & permanent teeth may be affected .

Thorax ;

Enlargement of costochondral junction making beading of the ribs, palpable and even visible .The sides of the thorax become flattened and longitudinal grooves developed posterior to the rosary , the sternum with its adjacent cartilges appears to be projected forwards producing (pigeon chest deformity). Along the lower borders of the chest develops a horizontal depression (Harrison groove , which corresponds to the costal insertion of the diaphragm .

Spinal column ;

Scolosis are common and kyphosis may appear ,lordosis of the lumber region may be in erect position .

Pelvis ;

Retarded growth and deformity ,narrow pelvis enterance by a forward displacement of caudal part of sacrum and coccyx , in female hazard with childbirth so may need c/s .

Extremities :

Epiphyseal enlargement of wrist &ankle .
Bending of the shaft of the femur , tibia&fibula results in bow legs deformity , knock knee & coax vara .Green stick fracture may occur in long bone.
Deformity of the spine ,pelvis &legs result in short stature &rachitic dwarfism .
Ligment relaxation &musle are poorly developed as result with mod –sever rickets
Delay in standing & walking ,weakness of abdominal muscle lead to (pot belly).

Diagnosis:

1-wrist x-ray is best for early diagnosis , the distal ends of ulna &radius appear widened concave (cupping) &frayed (normaly slightly convex ends) ,the distance from distal ends to metacarpal bones increased since large rachitic metaphysis which not calcified not appear on x-ray , the density of the shaft is decreased .
2- serum level of Ca++ normal or low
3-serum phosphorus level below 4mg/dl
4-serum alkaline phosphotase is elevated .
5-urinary C AM P is elevated.
6-serum 25-hydroxycholecalciferol is decreased .

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 .

Prognosis

With sufficient amount of vit. D administration healing begins within few days and progress slowly till normal bone structure occur .

Enlargement of epiphysis of long bone , ribs and deformity of skull disappear only after months or years of treatment .

In advanced cases ; there may be permanent deformity in form of bow legs, knock knee, spine , pelvis deformity coax vara and dwarfism .

Rickets is not a fatal disease ,but complications ,intercurrent infections e.g. pneumonia tuberculosis and enteritis mor risky for rachitic child than normal one.

Prevetion ;

1- exposure to ultraviolet light .
2- oral administration of vit. D , the dialy requirement of vit. D(10 µg or 400 i.u.) , vit. D should also administered to pregnant &lactating mothers .

Treatment ;

1-Correction of dietary habit and exposure to sun light .

Both natural & artificial light appropriate wave length are effective .

3- specific therapy

vitamin D (50-150) µg per day of vitamin D3 or (0.5-2) µg per day of 1.25 DH choleclaciferal .

Radiological improvement will be seen within (2-4) weeks but actual treatment should be continued for several months . If improvement delayed expect vitamim D refractory rickets .

Vitamin D3 is usually adequate unless deficiency is secondary to hepatic or renal diseases .

A single dose of 15000 µg vitamin D without further therapy for several months may be advantageous .

If healing is rapid allowing earlier diagnosis from (genetic x-linked vitamin D resistant rickets) .

After healing is complete the dose of vit. D should be lowered to 10 µg /day .