Scurvy in Children

A Report of Three Cases

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Scurvy in an adult patient was first described in 1669, by Poupart, who found crepitus in the epiphyses of the bones and noted that there were separations of the epiphyses from the shafts of the long bones on post mortem examination. In 1859, Moeller described a disease in infants characterized by swelling of the extremities, pseudoparalysis and bleeding of the gums, and thought that it was an acute form of rickets. In 1883, Barlow recognized the scorbutic origin of the disease described by Moeller, and proved his statements anatomically. Some of the confusion between rickets and scurvy resulted from the fact that the two diseases may occasionally be seen together. The German literature still calls infantile scurvy "Moeller-Barlow Disease."[1]

Later on, in addition to the clinical description of scurvy, Fraenkel, Pelkan and Wimberger[2] described the various radiological features which bear their name today. Although early medical writers such as Colbatch (1699) observed that findings of scurvy subsided with treatment by fresh fruit and plant juices, James Lind was the first author to use controlled series of patients to show that this disease is preventable, and completely curable, by fresh fruits and vegetables (1753). The effective antiscorbutic agent was isolated in 1932 by Szent-Gyorgyi, and was named "Ascorbic Acid".

Since most children are breast fed during infancy, and in some cases their diets include vitamin C-rich vegatables in early childhood, scurvy in children has not been a serious public health problem in Turkey.

To our knowledge, no case of scurvy has been observed among the children seen at Hacettepe Children's Hospital in the past 11 years; we

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were therefore interested to see three children with this disease in the hospital in a three month period, and thought it might be appropriate to report them and review the disease briefly.

Case Reports

Case 1- (A. P. No. 16005) This 3 year old girl was admitted to Hacettepe Children's Hospital on April 1, 1968, with complaints of pain, and for the last eight months tenderness over the arms and legs, and swelling of the knee joints during the previous two weeks.

Physical examination: This showed the patient to be pale, prostrate, and apparently in pain. She weighted 10 kg, and her height was 86 cm. Her hair was sparse, there was marked wasting of the subcutaneous fat, and she had bilateral nystagmus. The wrists and knee joints were widened, and she displayed a prominent, sharp-edged rosary on the anterior chest wall. Her legs were swollen and very painful to the touch. A grade 1 systolic murmur was heard over the heart.

Laboratory examination: The patient had Hb 6.71 gm/100 ml, W. B. C. was 5400/mm^3 with segmented cells predominating, and thrombocytes were present. A blood smear revealed hypochromia, anişocytosis and poikilocytosis. The reticulocyte count was 0.2 per cent and the hematocrit 24 mg/100 ml. Urinalysis was normal and PPD skin test was negative. NPN in the blood was 38 mg/100 ml, Na 137 mEq/Lt., K 5 mEq/lt. Cl 100 mEq/lt., P 3.4 mg/100 ml, total serum proteins 5.5 gm /100 ml, albumin 3.4 gm/100 ml, globulin 2.1 gm/100 ml and cholesterol 122 mg/100 ml. Serum electropheresis revealed the albumin to be 49 mg/100 ml, alpha lmg/100 ml, globulin 5 mg /100 ml., alpha 2 mg/ 100 ml, globulin 15 mg/100 ml, beta globulin 12 mg/100 ml and gamma globulin 16 mg/100. Serum iron was 33 gamma, and iron binding capacity 417 gamma. Ophtalmoscopic examination of the fundi was normal. Determination of vitamin C levels in the serum following Vitamin C tolerance test revealed the following values:

Fasting, 0.125, 1 hour 0.310, 2 hours 0.725, 3 hours 0.725, 24 hours 0.250 mg/100 ml.

In this test, serum vitamin C level reached only 0.75 mg/100 ml at the end of three hours, versus the normal value of 0.8–2.2 mg.

Radiological findings of this patient are summarized below in Figures 1–5.

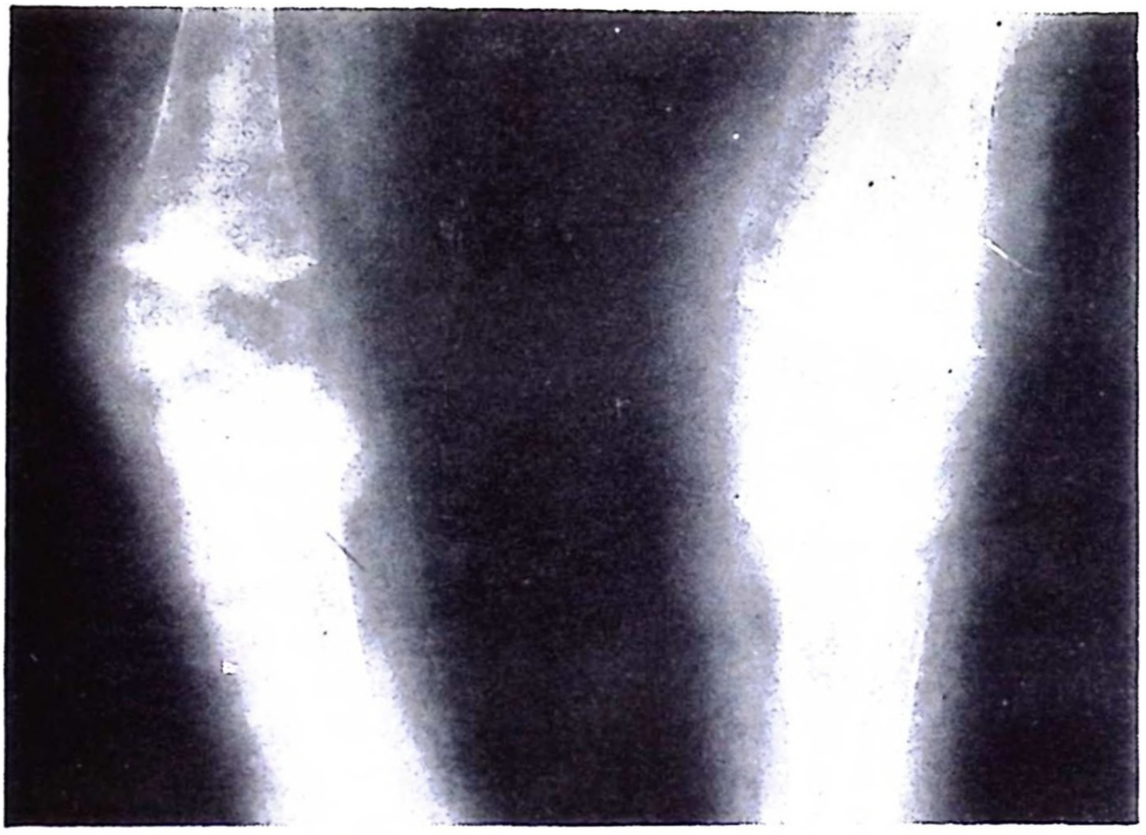

Figure. 1. Anteroposterior radiogram of knees on day of admission showing diffuse osteoporosis of the bones and marked thinning of the cortical bone. In the metaphyses bordering the epiphyseal plate (zone of provisional calcification), a line of heavy calcification is evident, and seen to end in a lateral spur (single arrow) in places. Secondary ossification centers of epiphyses display a radiolucent center surrounded by a frame-like sclerotic ring. The earliest radiological manifestation of subperiosteal hemorrhages is found in the outward displacement of the intermuscular fascia (Two arrows).

Clinical Course: The patient was initially treated with 500 mgs Vitamin C administered intravenously for three days, and was then kept on 200 mgs of oral vitamin C daily. Iron deficiency anemia was treated with ferrosanal (20 drops t.i.d.), and a concurrent urinary infection was treated with Gantrisin. Clinical improvement was rapid, with a decrease in the swelling of the legs, and the child started using her limbs freely and feeding herself. Radiological improvement was evident, with calcification of the subperiosteal hemorrhages observed on radiograms obtained 25 days later (see Figures 2 and 3).

The patient was discharged on the 35th day of hospitalization.

Case 2- (S. T. No. 24478) This patient was a one-year-old boy admitted to Hacettepe Hospital with coughing and fever on May 10, 1968. Beginning a month prior to admission, it was noticed that he was unable to stand on his feet, and this was followed by a swelling in the left leg and some weight loss.

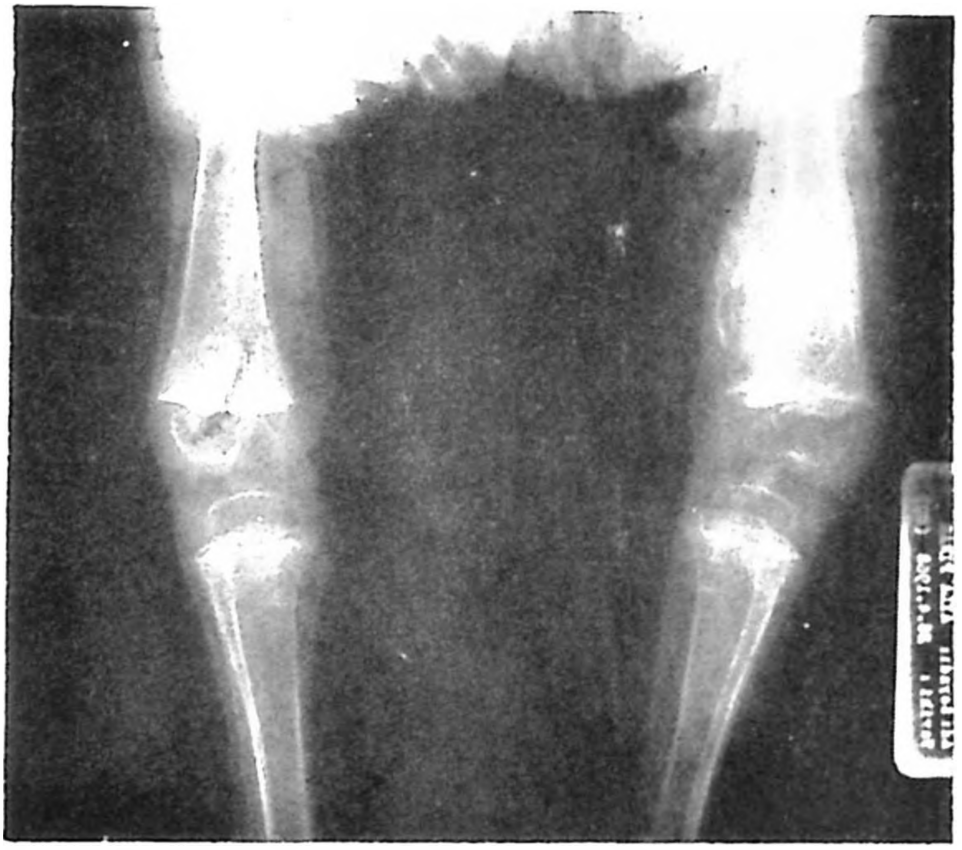

Figure. 2. Follow-up radiogram of knees of same patient, 25 days after the first, showing calcification of subperiosteal hemorrhages. Overall density of the bones has regained a more normal appearance and some bony trabeculae can now be identified following rapid deposition of calcium.

Physical examination: revealed the child to be pale, wasted and immobile. He weighed 6.2 kg, the hair appeared thin and reddish, the skin was dry, and in places showed slight desquamation. The anterior rib ends were widened, prominent and sharp edged on palpation, and the wrists and knee joints were similarly widened. The liver was palpable five cm below the costal margin.

Laboratory examination: Hb was found to be 6.73 gm/100 ml, and W. B. C. was 7600/mm^3 with segmented cells predominating. Erythrocytes revealed hypochromia, anisocytosis and poikilocytosis. PPD test was negative, and urinalysis was normal. Serum tests gave the following results: NPN 41 mg/100 ml, total serum proteins 6.3 mg/100 ml, albumin 4.5 mg/100 ml, globulin 1.8 mg/100 ml, SGOT 30 U, SGPT 10 U., CCF++, thymol turbidity 4 U., cholesterol 118 mg/100 ml, total lipids 470 mg, Ca 8.5 mg /100 ml, P 6 mg/100 ml, alkaline phosphatase 5.2 B.U., serum iron 28 gamma, and serum iron binding capacity 312 gamma. The serum vitamin C level was 0.2 mg.

The radiological findings of the patient are shown in (Figures 6 and 7) below:

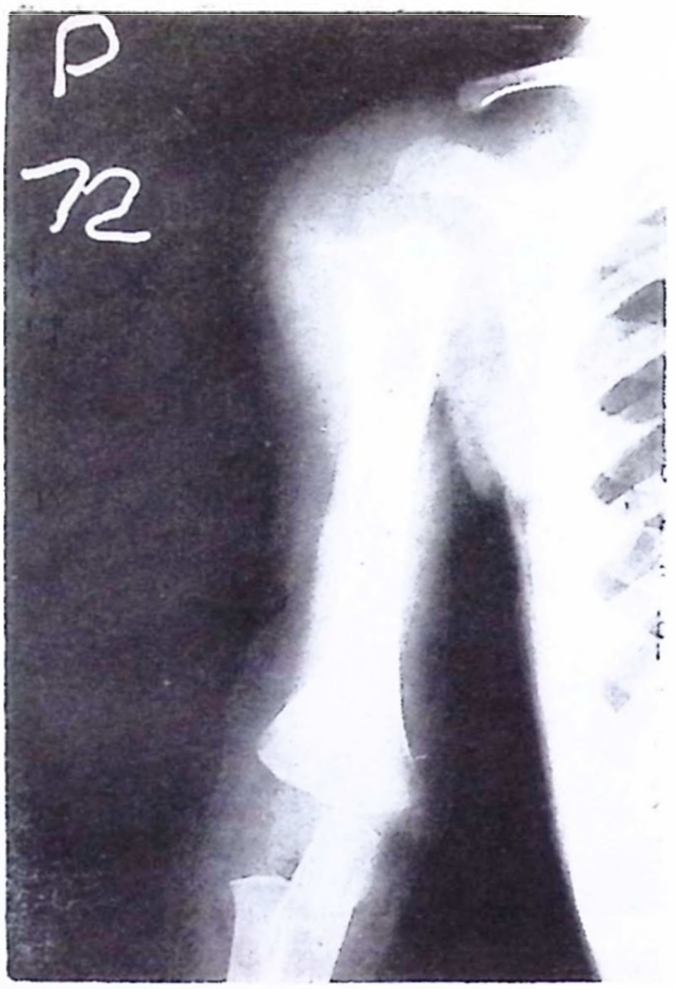

Figure. 3. Radiogram of right humerus showing a characteristic lateral spur formation at the level of the proximal humeral metaphysis. The stripped periosteium has started to lay down new bone.

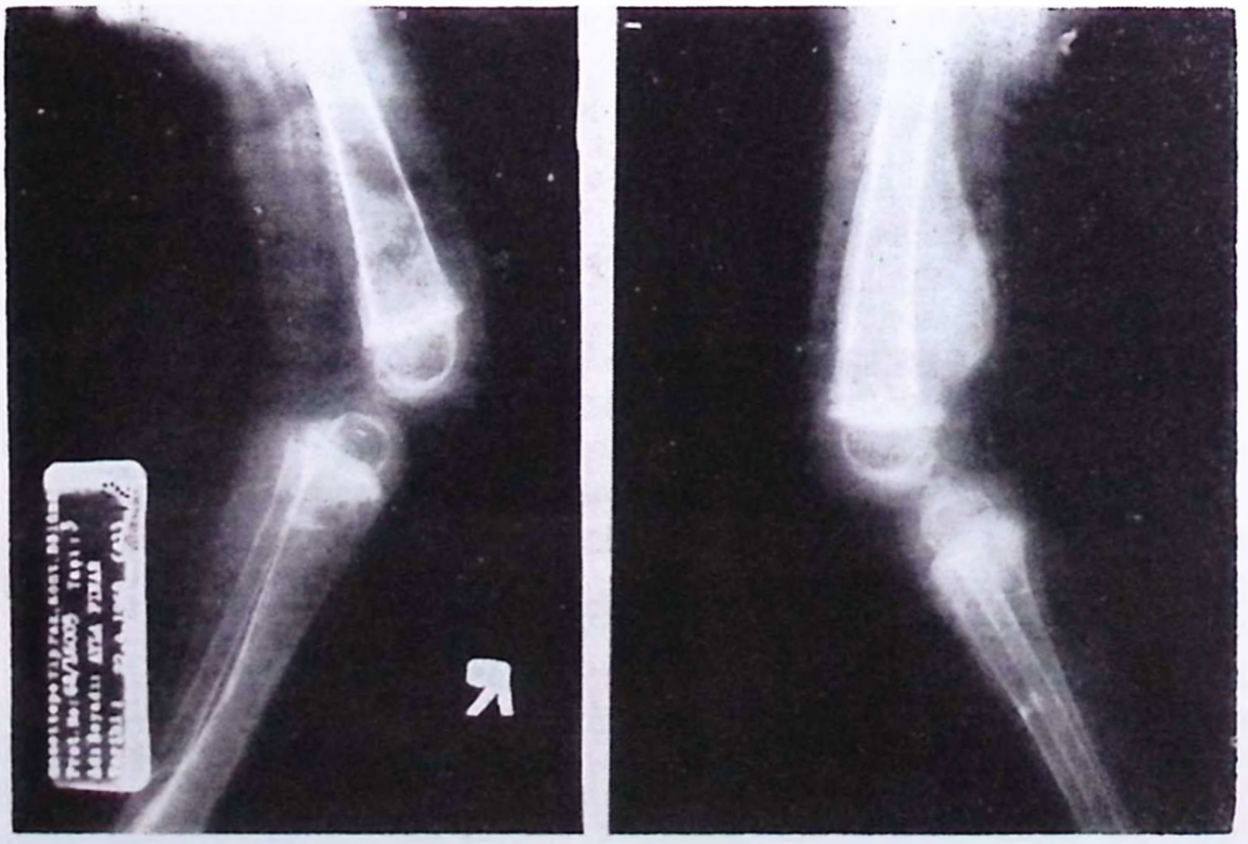

Figure. 4. Lateral radiograms of knees of same patient showing massive, calcified subperiosteal hemorrhage to better advantage.

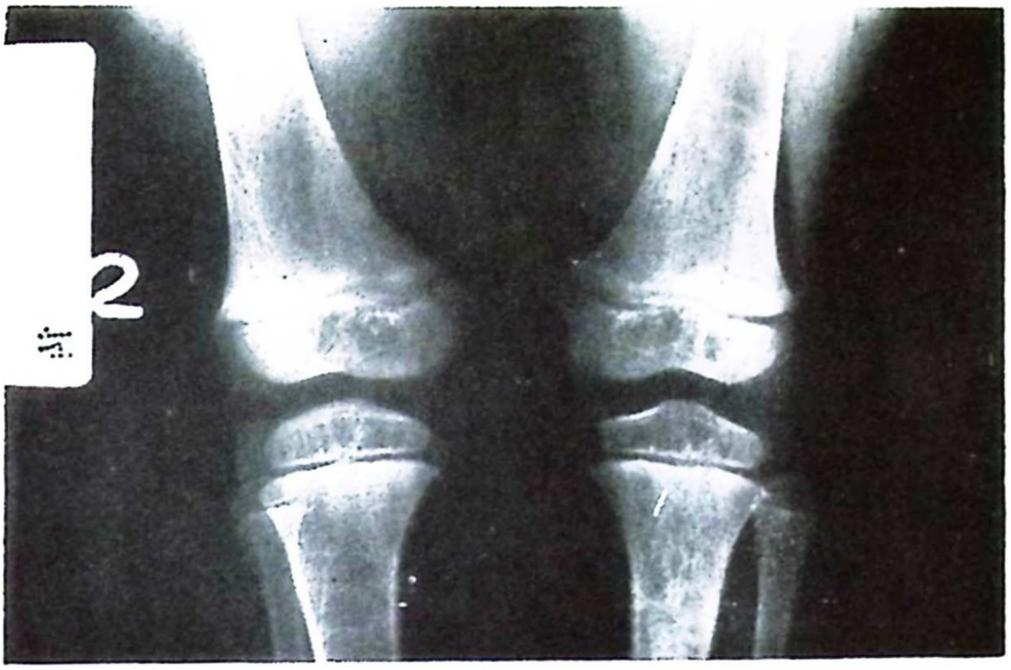

Figure. 5. Radiogram of knee joints of same patient 15 months after initial radiograms, showing transverse sclerotic line deeply buried in the shaft by this time, following normal growth of bone. The relatively radiolucent cores within the epiphyseal ossification centers are the other radiological remainders of the past scorbutic process.

Clinical Course: The patient was treated with 500 mg vitamin C intravenously for the first three days, and was then given 100 mg of oral vitamin C daily. A concurrent pulmonary infection was treated with penicillin and streptomycin, and iron deficiency anemia with ferrosanal. There was a rapid clinical and radiological improvement, and the child was discharged on the 40 th day of hospitalization.

Case 3- (Y. O. No. 19942) This four-year-old boy started complaining of pains in the legs, and showed a reluctance to stand four months prior to admission to Hacettepe Hospital on April 17, 1968. Two months later he developed urinary and fecal incontinence, and involuntary movements of the eyes.

Physical examination: The patient was seen to be pale and weak, with marked wasting of the subcutaneous fatty tissues. He weighed 11 kg, the lower extremities were atrophied and painful to the touch, there were widespread areas of purpura over the skin, and bilateral nystagmus and slight midriasis was present. Ophthalmoscopic examination showed areas of congestion.

Laboratory examination: Hb was found to be 7.21 mg, W.B.C. was 13600/mm^3 with segmented cells predominating, and normal throm-

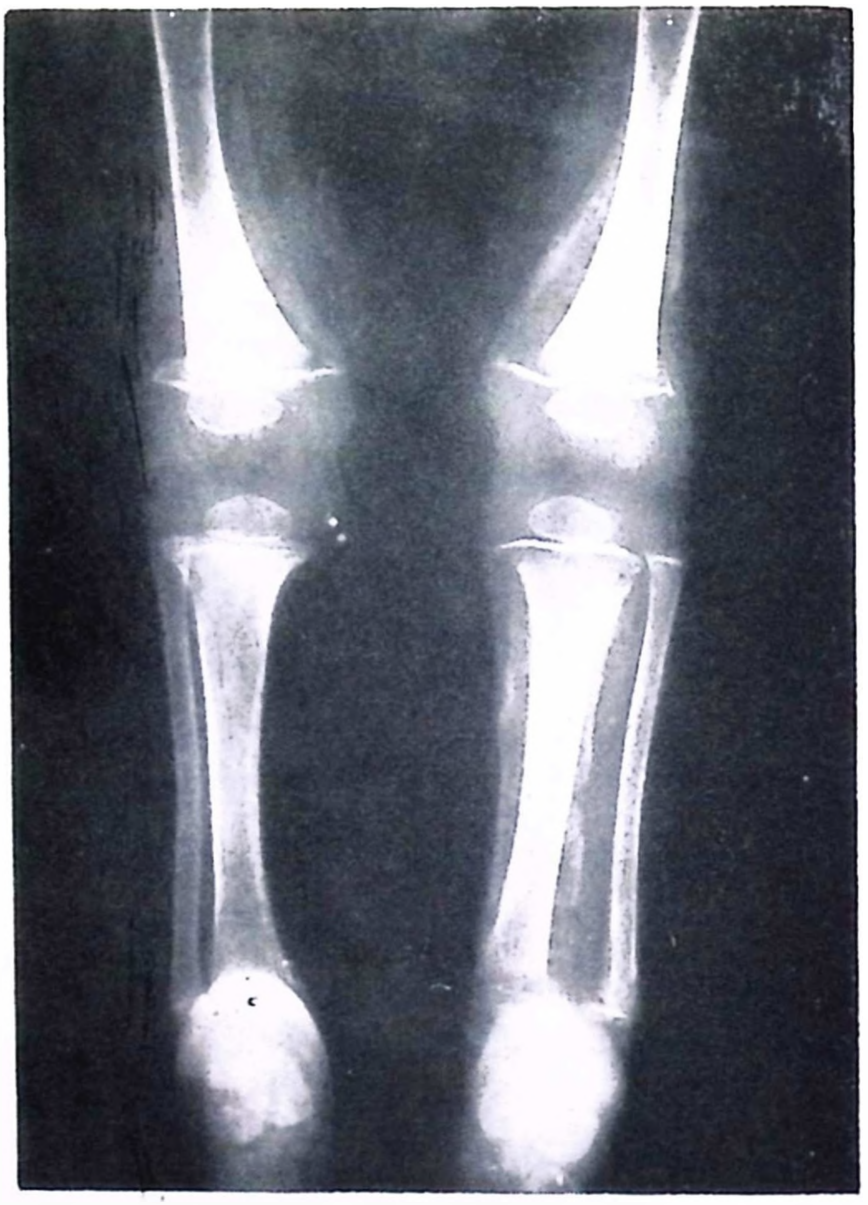

Figure. 6. – A radiogram of the lower extremities of (Case No. 2 two days prior to admission), revealing characteristic selerotic lines in the metaphyses of he long bones, proximal to which is a band of diminished density (the "scurvy line"). There is a fracture through the distal epiphyseal plate of the right femur with lateral displacement of the ossification center, and wide-spread subperiosteal hemorrhages, already partially calcified, are seen.

bocytes were present. Erythrocytes showed hypochromia, anisocytosis and poikilocytosis, and the reticulocyte count was 1.4 mg/100 ml, Urinalysis was normal. NPN was 43 mg/100 ml, total proteins 6.8 gm/100 ml, albumin 3.7 gm/100 ml, globulin 3.1 gm/100 ml, Ca 9.9 gm/100 ml, p 4.4 gm/100 ml, and alkaline phosphatase was 16 B. U. In the serum electropheresis, albumin was 33 gm/100 ml, alpha 1/100 ml, globulin 11/100 ml, alpha 2/100 ml, globulin 17/100 ml, beta globulin 14/100 ml and gamma globulin 26/100 ml. PPD skin test was negative. Vitamin C levels in the serum following the loading dose were: fasting 0.166 per cent,

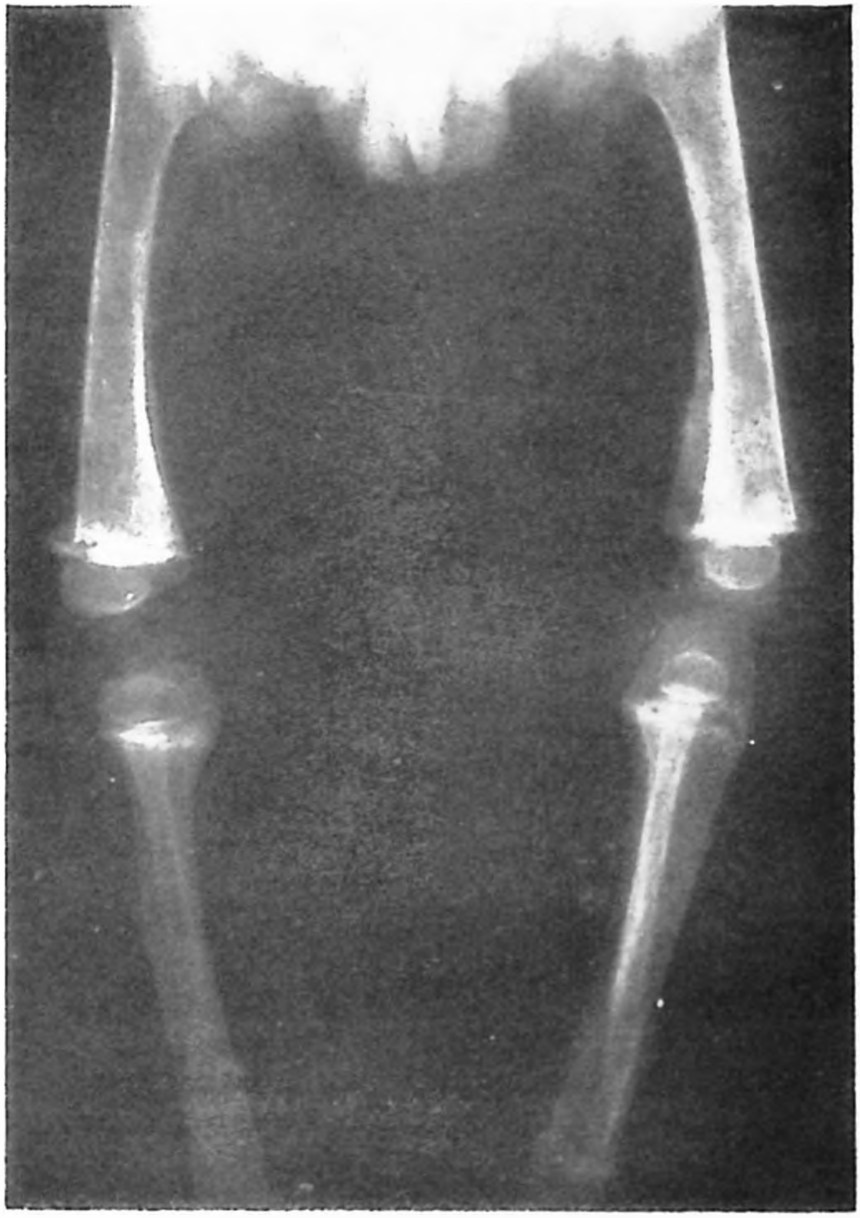

Figure. 7. Radiogram of the same region three weeks later; the radiolucent "scurvy lines" have started to fill in with normal bone.

1 hour 0.332 per cent, 2 hours 0.498 per cent, 4 hours 0.498 per cent, and 24 hours 0.332 mg per cent. Bleeding time was 2', clotting time 10', and prothrombin time was 12'. Serum iron was 22 gm and the iron binding capacity was 358 gm. The radiological findings in this patient are shown below in (Figures 8, 9, and 10).

Clinical Course: After the diagnosis of scurvy had been made, the patient was treated with 200 mg of oral vitamin C daily. He also had a bronchopneumonic infection which responded well to antibiotic treatment. He developed rubeola on the thirty-first day of hospitalization, but the findings of scurvy showed continous improvement and he was discharged on the 66 th day.

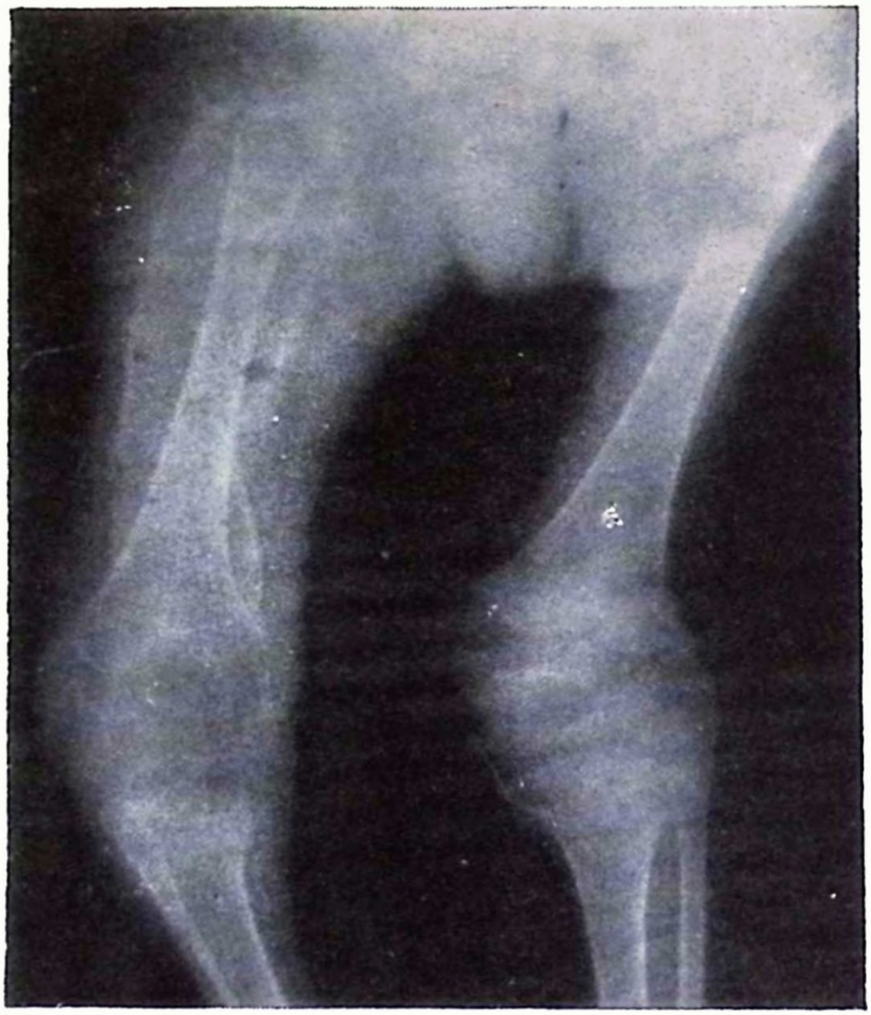

Figure. 8. Radiogram of femurs and knee joints of Case 3 on admission showing partially calcified subperiosteal hematcma surrcunding the entire shaft of the right femur. The osteoporotic bones display thinning of the cortices. The patient was unable to extend the lower extremities fully because of pain.

Discussion

Survy in children has only been recognized in the last 100 years, whereas the adult type of the disease was relatively well known much earlier. This is closely related to the introduction of artificial feeding for infants[3]. Since human milk contains about four to eight mg vitamin C per 100 ml, breast-fed infants usually have an adequate supply of this vitamin. But if it is not added to the food of an artificially fed baby, clinical or subclinical findings of scurvy will develop. If the pregnant women has a diet adequate in vitamin C, her infant obtains stores of it in utero, which can last him for almost the first five months of his post-natal life, providing the demands from his vitamin C depots, such as infection, are not excessive. Vitamin C is stored in the tissues, especially in the adrenal and other ductless glands, and is in transit in the plasma and white blood cells. Be-

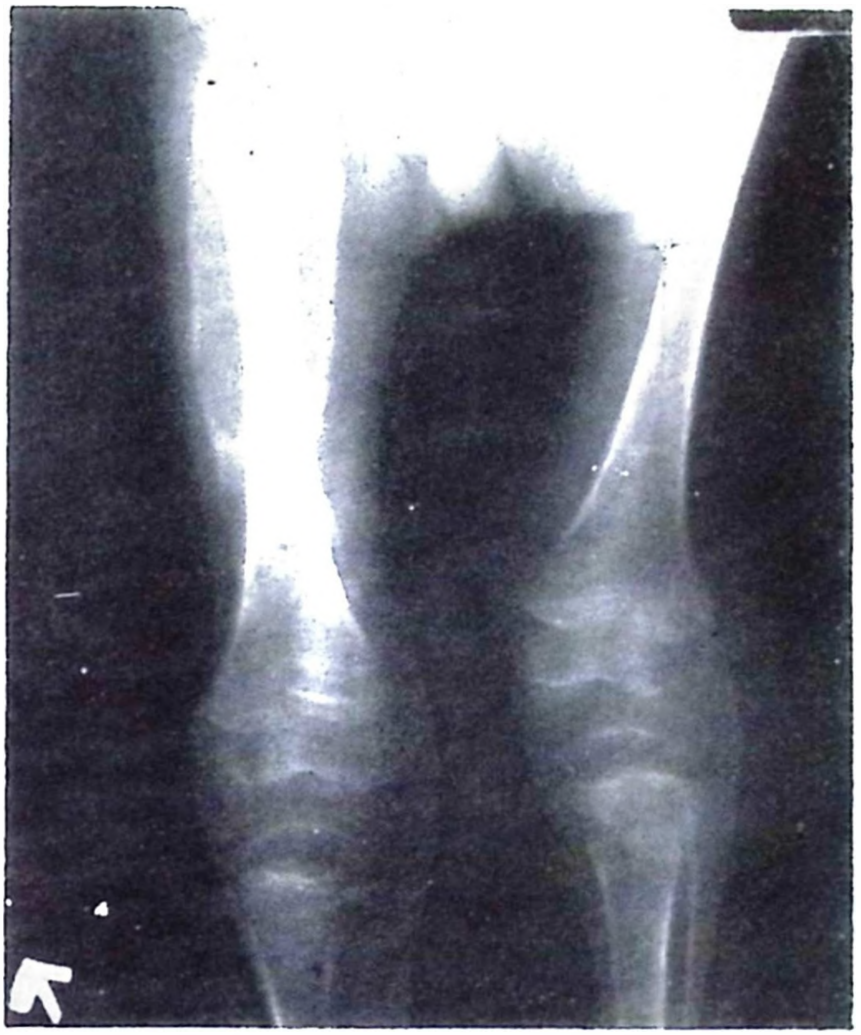

Figure 9. Radiogram of the same region a month later showing an increase in over-all bone density and beginning of "remodelling" of new subperiosteal bone along the right femur by resorbtion.

cause of pre-natal storage of this vitamin, infantile scurvy very rarely becomes manifest before the fifth or sixth month after birth.[4]

After a variable period of vitamin C depletion, vague symptoms of irritability, digestive disturbances and loss of appetite are usually the first clinical evidence of the disease.[5] As the disease advances, irritability increases and the patient develops general tenderness, which is most marked in the legs. The infant refuses to be picked up and cries when his diaper is changed. Because of pain the child shows pseudoparalysis and holds the legs in a typical "frog" position, as this causes him the least pain (Figure 11). Edema of the legs may develop, and at times a subperiosteal hemorrhage along the shaft of a long bone may be palpable. A "rosary" may be present at the costochondral junctions, which is different from the rachitic rosary, in that the flaring rib ends display a sharp margin, and the cartilage of the junction shows a depression. The gums may have bluish-purple swelling, and hemorrhages may occur in the skin and mucous membranes. In-

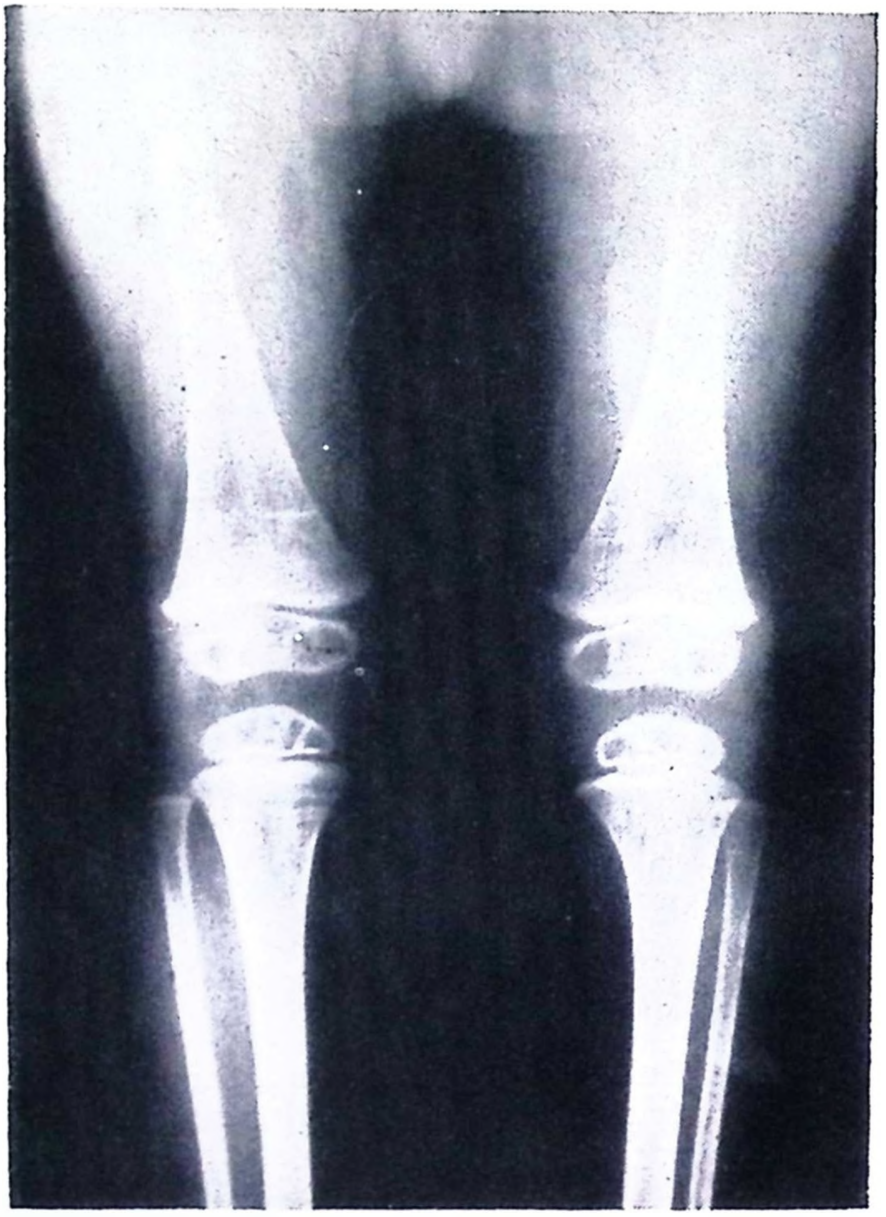

Figure 10. A. 6-month follow-up radiogram showing complete resorbtion of the subperiosteal hematoma around the right femur. The only remainders of scurvy are the transverse sclerotic lines in the shaft ends, and the insets of central rarefaction remaining within the epiphyseal ossification centers.

testinal bleeding and hematuria may be present,[6] and low grade fever and moderate secondary anemia are also usually seen.[7] The radiological findings in scurvy are quite characteristic, and the disease is often diagnosed by means of x-rays. The earliest radiological change is generalized bone atrophy,[7] which is seen with marked osteoporosis, giving the bones a "ground glass" appearance, and thinning of the bony cortex. Since such a change can be produced by a variety of other pathological processes, it is not diagnostic of the disease at this stage. The next more advanced x-ray change is the development of a sclerotic line at the metaphysis of the long bones bordering the epiphyseal cartilage plate (the zone of

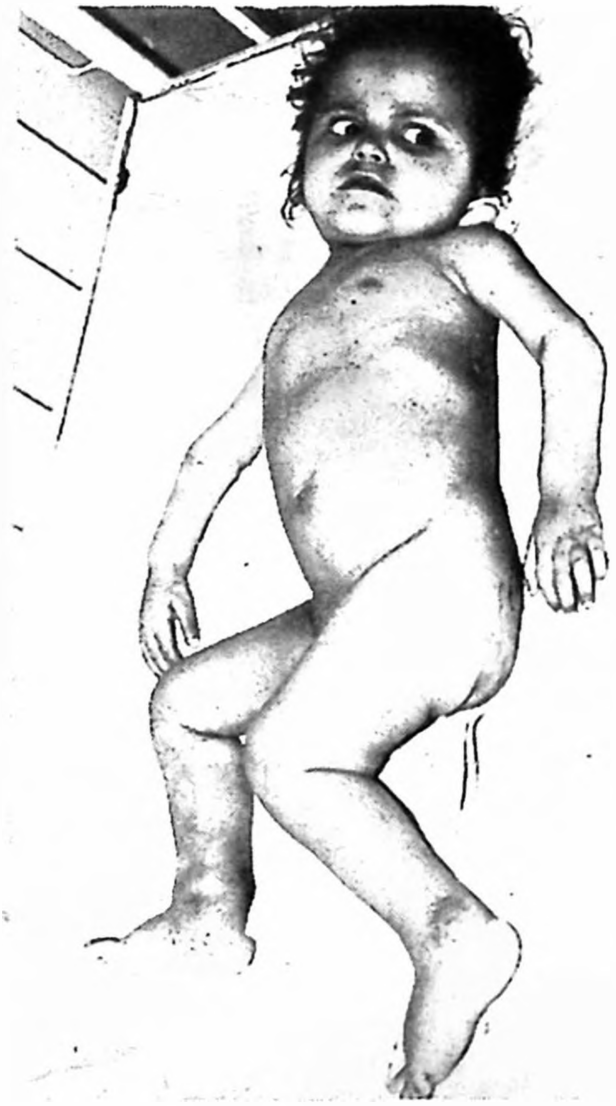

Figure 11. Lying position of Case No. 1

provisional calcification).[9] This white line, which is called "Frankel's line," is caused by a heavy deposit of calcium salts in the endochondral region, with disturbed growth. Since bone growth is most rapid in the knee area, these changes first become recognizable in this region; at the same time and through the same pathological process, a sclerotic white frame-like ring develops around the osteoporotic ossification centers in the epiphyses (Wimberger's sign), and at this stage the radiological findings are diagnostic of scurvy. As the disease advances, a radiolucent transverse band forms just proximal and parallel to the white line, which may itself show characteristic spur-like extensions on its lateral margins,[10] due to impaction fractures of this weak, poorly ossified zone. Similarly, fractures through the epiphyseal plate may easily develop, and the epiphyseal ossification centers may be displaced. Subperiosteal hemorrhages can be detected early if there is outward displacement of the muscle shadows. These become readily discernible when the elevated periosteum

starts to show a shell-like calcification, which is usually a sign that healing has begun.

Without the aid of x-rays, clinical differential diagnosis may be difficult in the early stages, and arthritis, or suppurative osteomyelitis may be considered. Among the laboratory aids, a fasting vitamin C level of the blood plasma over 0.6 mg/100 ml excludes probability of scurvy; however, a lower level of vitamin C is not always diagnostic of the disease. Diagnosis of scurvy is not usually based on chemical data, and x-rays of the long bones are of the greatest help in establishing the presence of the disease.

When adequate treatment is administered, which consists of 100 to 200 mg vitamin C daily, orally or parenterally, the prognosis is very good. Pain ceases in a few days, but roentgen changes in the long bones take a much longer time to return to normal, usually months. But even in very severe cases with epiphyseal fractures and massive subperiosteal hematomas, the long bones eventually regain normal tubulation and structure.

The pathologic signs of vitamin C deficiency outlined above are almost entirely confined to supporting tissues of mesenchymal origin (bone, dentine, cartilage, and connective tissue). Thus scurvy is characterized by failure of the formation and maintenance of intercellular materials, which in turn causes typical symptoms, such as hemorrhages, loosening of the teeth, poor wound healing, and easy fracturability of the bones.[11]

The interesting observation of horizontal nystagmus in two of our three patients could very well be incidental. We were unable to establish any relation between this finding and the physiopathological scorbutic processes, though we are not in a position to state that no such relationship exists.

One common factor in the histories of our three patients was a striking lack of a balanced diet. All three, coming from rural areas, were given almost exclusively farinaceous foods, rich in carbohydrates, and occasionally cow's milk, low in vitamin C, but their diet included no vegetables or fresh fruits.

Finally, we wish to point out that the observation of these three cases in such a short period clearly indicates that infantile scurvy, which should not exist today, definitely does in Turkey. This must be recognized in order to diagnose the disease at an early stage.

Summary

Three scorbutic children diagnosed within a short period are presented and the clinical and radiological aspects of scurvy are briefly reviewed. Attention is drawn to the presence of this completely preventable disease in Turkey today.

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