INFANTILE SCURVY IN MALAYSIA

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INFANTILE SCURVY in the tropics has been reported to be a rare disease. (12) (6) In Malaysia and Singapore, only three cases of scurvy have been described, all in Singapore, and all in older persons, aged 4, 17 and 50. (17) (18) (7) Only one was an infantile scurvy. At the time that these reports were made, the authors and others (14 (9) felt the disease to be a rarity.

There have been no comprehensive surveys of Vitamin C nutrition in Malaysia or Singapore. However, a survey of 2,000 consecutive admissions (13) in the Pediatric Department of the General Hospital, Singapore, in 1939 showed 118 clinically diagnosed cases of rickets, 8 1 with X-ray diagnoses as well. Congenital syphilis was ruled out but scurvy was not. Many were diagnosed on the enlargement of the costochondral junction alone, which may have been rachetic or scorbutic. The author has subsequently stated in retrospect that scurvy may have been present in a percentage of these cases. (15)

Four cases of scurvy were diagnosed at Gombak Aborigine Hospital and the University Hospital, Kuala Lumpur in a 5-month period, April-August, 1969. This paper includes the case reports of these four cases and a brief review of some of the radiological signs and diagnostic laboratory tests in scurvy.

CASE 1

This patient was a 9-month old female Aborigine Senoi child with a history of drinking unsupplemented sweetened condensed milk since birth because the mother was not able to breast-feed her. Two other siblings, who had received the same diet, were well. Three months prior to admission, she developed an intermittent cough, fever and diarrhoea and failed to thrive.

Three weeks prior to admission, she became irritable, crying when moved or picked up and was unable to sit up or move her left arm or legs well. She developed a continuous fever one week prior to admission, became anorexic and began to show bilateral knee swelling.

On examination, she was irritable, emaciated, dry and pale, lying on her back in a "frog leg" position, an expression of pain on her face and appearing to cry but producing no sound. Weight was 7 lbs. 12 ozs. There were no petechiae or purpuric spots. Her skin appeared rough and red with a light fine maculopapular rash over her scalp and forehead. There was

Fig 1
Case 1, showing enlarged costochondral junctions, the "scorbutic rosary".

no neck stiffness and the fontanelle was moderately depressed. Her eyes rolled vertically and to the left and there was frequent blinking. Gums were normal around 4 erupted teeth. Respiratory, cardiovascular and lymphatic systems were normal. The abdomen was protuberant and the colon appeared to be filled with faeces. Liver and spleen were not palpable.

There was marked enlargement of the costochondral junctions ("rosary"), which were round to the touch and painful when pressure was applied. (Fig. 1)

There was no voluntary movement of her left shoulder or either leg and passive movement of these joints gave pain. Both knees and dorsums of the feet were oedematous. On admission, laboratory data

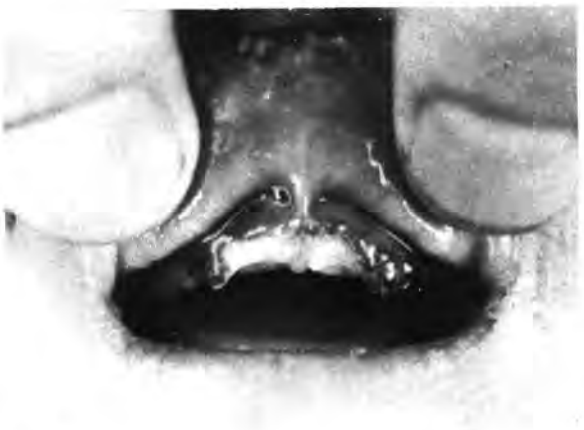

Fig. 3
Case 2, hypertrophic, bleeding, bluish coloured gums, characteristic of scurvy.

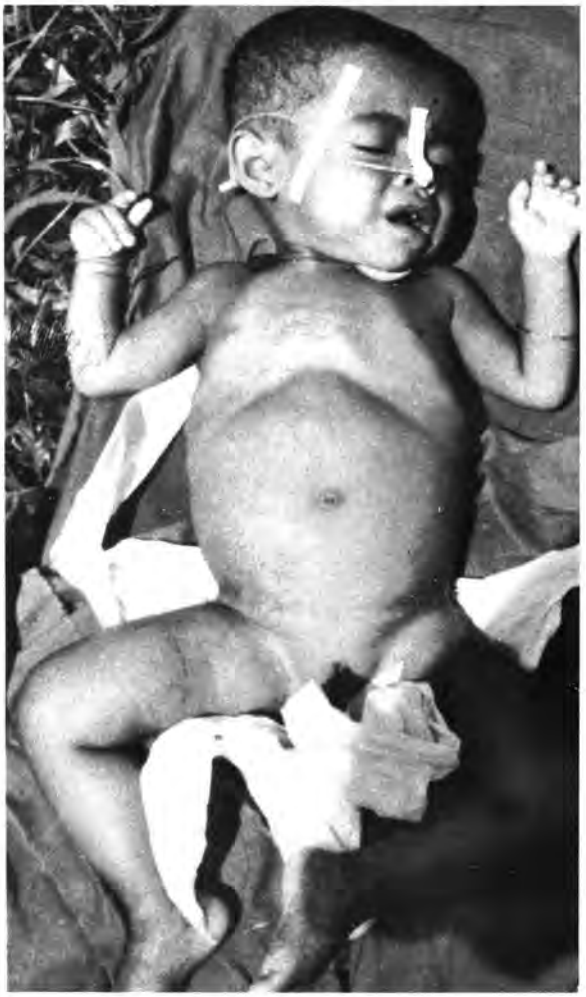

Fig. 2
Case 2, child in "pithed frog" position. Also seen are scorbutic rosary and leg swelling bilaterally.

were an hematocrit of 30; WBC of 9,800 with normal differential; CSF colorless, clear and non-cellular under low pressure. A tournique test was negative. CSF culture was negative and stool culture grew salmonella $^{c}2$. Radiological examination of her chest revealed normal lung and heart shadows but prominent large costochondral rib junctions. (Fig. 4) X-rays of the extremities showed decreased density and loss of trabecular pattern in the diaphyses. There was cupping, spreading and spurring of the metaphyses and ring epiphyses in the ankle joints, knees, hips and wrists. (Fig. 5) There was a fracture dislocation through the left proximal humeral ephiphyseal plate. These findings were consistent with scurvy.

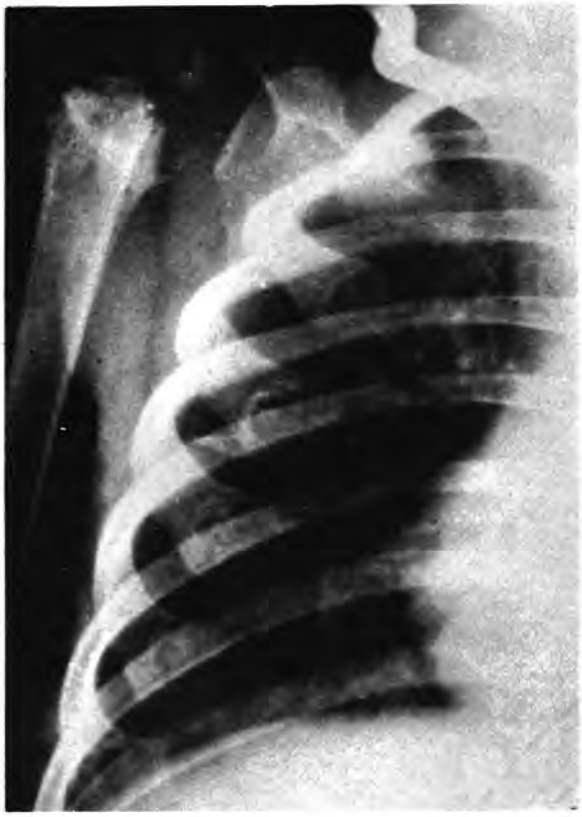

Fig 4
Case 1, chest X-ray showing enlarged costochondral junctions and a dislocation-fracture of epiphysis of humerus.

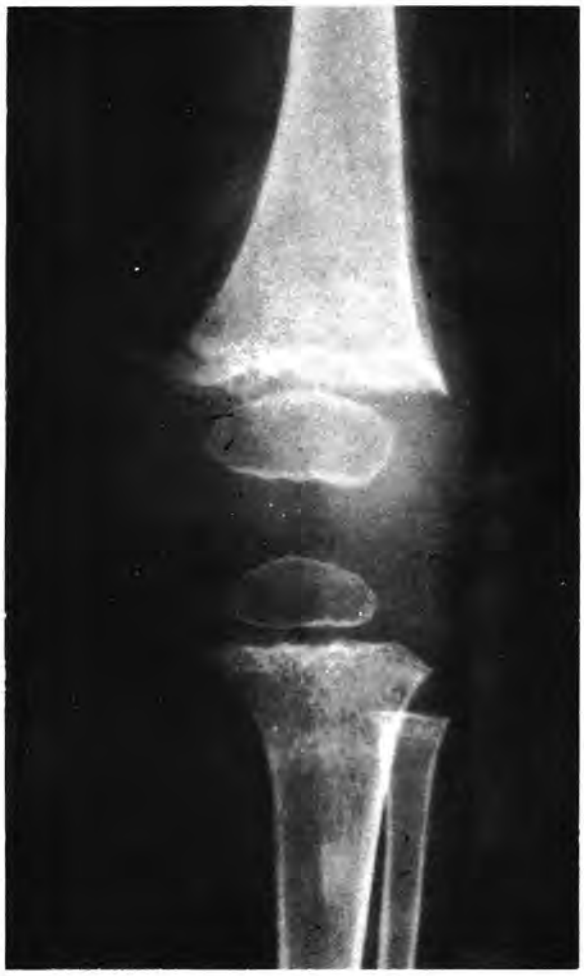

Fig. 5
Case 1, X-ray of right knee at admission, showing thickening and spurring of provisional line of calcification, a radiolucent wedge-shaped "corner sign of scurvy" and a dense outlining of the epiphysis, the "ring sign of scurvy".

The child was treated with ascorbic acid 50 mgm IM q6h., tube feeding, multivitamins and antibiotics for the Salmonella. Within one week, abnormal eye movements and blinking had ceased and the child could again cry. In 10 days, the frog leg position was lost, leg and arm tenderness gone and knee swelling down. On X-ray, calcification of subperiosteal hematomas about the distal ends of the femurs appeared. (Fig. 7) At one month, she completely desquamated an outer layer of skin but recovered well and was discharged after three months. Her weight on discharge was 10 lbs. 4 ozs. (4.66 kg.)

CASE 2

This 12-month-old Aborigine female presented with a history that she had been breast-fed until the age of six months. At this time, the supply of breast milk failed and she was fed on sweetened condensed milk supplemented occasionally with rice. She had had fever and cough for 3 weeks and swollen bleeding gums, swelling and tenderness of the legs for two weeks prior to admission.

She presented as a very irritable, pale, crying child with a grimacing expression, unable to close her painful mouth. She lay in a "frog leg" position, showing little voluntary leg movement and would not sit up. (Fig. 2) Her gums were blue, bruised, hypertrophic and bleeding around the few erupted teeth. (Fig. 3) There was a fine rash of small hyperkeratotic nodules over the forehead and back through the hairline. Other than a few rales in the base of the left lung, respiratory and cardiovascular systems were normal. Lymph nodes were palpable

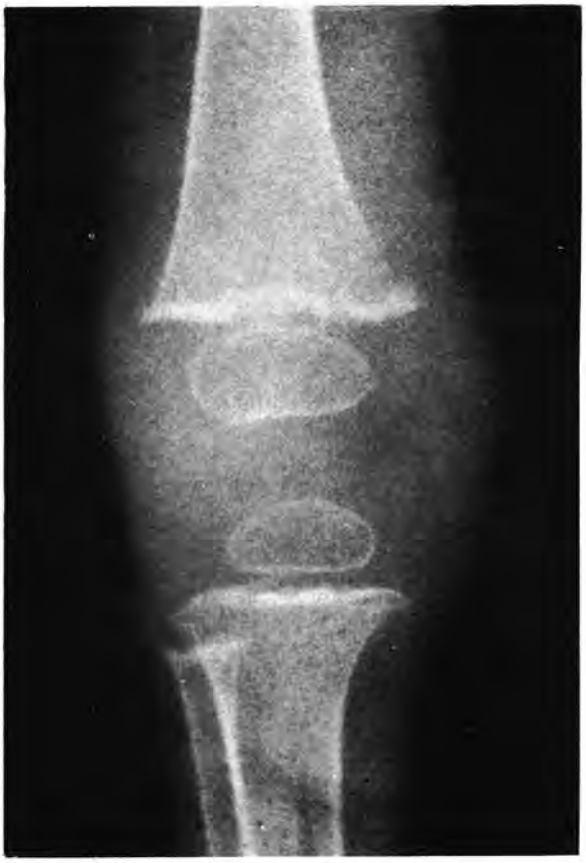

Fig. 6
Case 2, X-ray of right knee at admission, showing a thickening and spurred provisional line of calcification and an adjacent radiolucent "scurvy line".

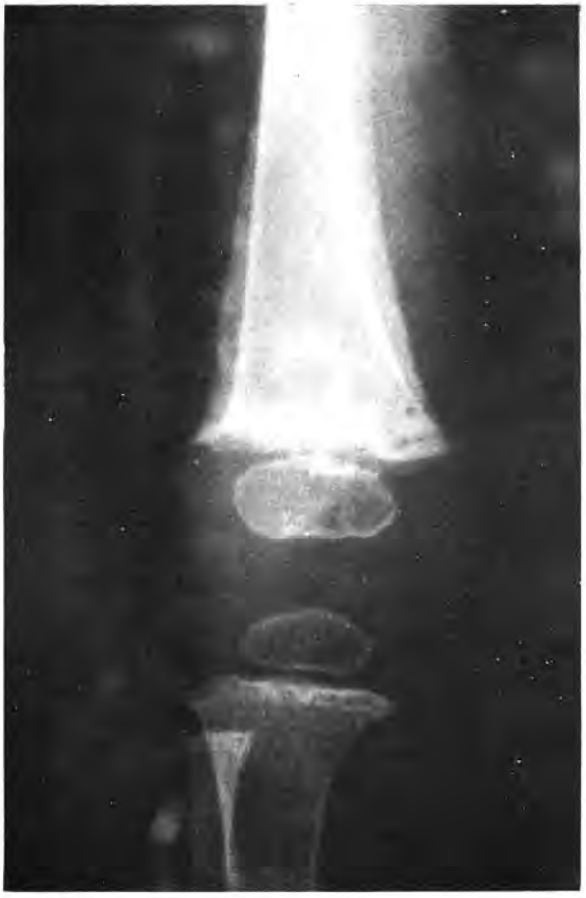

Fig. 7
Case 1, follow-up X-ray of right knee two weeks after admission showing periosteal calcification.

bilaterally in the neck and in the left axilla. The spleen was palpable 2 cm. below the costal margin. A well demarcated rosary was present, each costochondral junction feeling quite round to the touch. There were no petechiae or purpuric spots and the tournique test was negative.

Admission laboratory data included an hematocrit of 22.5%, a WBC of 9,300, polys 36%, lymphs 62%. The blood film showed microcytic hypochromic changes. Serum Vit. C was 0.2 mgm%. Urine vitamin C values were, for the 2nd 24 hours after admission, 0.4 mgm/150 cc, for the 3rd 24 hours 2.4 mgm/150 cc, and the 4th 24 hours 149 mgm/150 cc. Radiological examinations of the chest showed prominent rib castochondral junctions bilaterally. X-rays of the extremities showed changes in the elbows, knees and ankles. There was thickening and spurring of the provisional line of calcification with adjacent radiolucent bands. The epiphyses were outlined with a radio-dense line. The X-rays were considered diagnostic of scurvy. (Fig. 5) The child was given 50 mgm Vitamin C P.O. with 500 mgm IM on the 2nd day. In three days after beginning treatment, the gum hypertrophy had diminished markedly, there was no further bleeding and the forehead rash had almost disappeared. One week after admission, the fever and leg tenderness were gone and the child was sitting by the 12th day. Follow up X-rays at 10 days showed periosteal calcification around the lower ends of the femur. The child continued to improve uneventfully.

CASE 3

This 15-month-old Chinese female child was well until 4 days before admission when she stopped walking and protested when her right leg was moved passively. She was the fourth child of a shop assistant

TABLE 1 (19)

Frequency of symptoms in scurvy patients

Tenderness of legs	77%
Irritability	70%
Pseudoparesis of legs	64%
Anorexia	31%
Fever	19%
Swelling over long bones	14%

TABLE 2 (19)

Frequency of signs in scurvy patients

Rosary	80%
Leg tenderness	67%
Pithed frog position	52%
Haemorrhagic gums	44%
Fever	44%
Palpable subperiosteal haemorrhages	48%

Frequency of symptoms and signs in infantile scurvy in a North American population.

who earned M$160 per month. She had been fed entirely on sweetened condensed milk until the age of 9 months. Subsequently, she was offered rice and fish in addition, but she seldom ate these.

Physical examination revealed an irritable child who cried whenever her lower extremeties were handled. She held her hips abducted and externally rotated and her knees flexed. There were pupuric spots on her face, neck, hands, and feet. Her gums were spongy and bled easily. A tournique test was positive. She weighed 6.65 kg. and measured 73 cm. The rest of the physical examination was unremarkable.

Laboratory data included: Hgb. 9 gm%; PCV 31%; MCHC 29%; platelets 194,000/cmm; WBC 4100/cmm; neutrophils 51%; lymphocytes 40%; eosinophils 1% and monocytes 3%. A chest X-ray showed expansion and concavity of the anterior ends of the ribs bilaterally with normal lung shadows. X-rays of the long bones revealed generalised loss of bone density and a line of increased density across the distal ends of the femur and tibia bilaterally. The X-rays were considered compatible with but not diagnostic of scurvy.

The child was offered a full cream full strength milk and multiple vitamins. In addition, she was given Vitamin C 125 mgs. q6h by mouth. Four days after admission, there was no more bleeding from the gums, the purpuric spots were fading and movements of the legs were painless. When followed up in the clinic one month later, she weighed 7.7 kgm. and was walking again.

CASE 4

A 10-month-old female Indian child presented with a history that she had fallen down from a height of about 3 feet a week prior to admission. On the day of her fall, she developed a low grade fever and productive cough. Two days later, she stopped walking and began protesting when she was handled. At the same time, a lump was noticed on her forehead. The lump increased in size until the day of her admission.

She came from a poor family and was the last of five children. Her father earned M$3 per day as a gardener. She was the product of a full-term normal delivery, weighing 5 lbs. at birth and was considered normal until the present illness. She had been fed entirely on sweetened condensed milk since birth.

Physical examination revealed an irritable pale child weighing 5.48 kgm. A soft, almost cystic swelling 7.5 by 7.5 cm., was seen on the right side of the forehead. The lower extremities were held flexed and showed very little spontaneous movement. However, she cried and moved them when touched. The costochondral junctions were prominent and the spleen and liver were enlarged. There were no haemorrhages in the skin, gums or optic fundi.

Laboratory data included a Hgb. of 7.0 gms.%; PCV of 25%; MCHC of 28.8%; a WBC of 4,600 with 66% neutrophils and 34% lymphocytes. A chest X-ray showed expansion of the anterior ends of the ribs. X-rays of the long bones showed diffuse osteoporosis. In the femurs and tibia, there were dense lines through the metaphyses with adjacent wedge shaped areas of reabsorption. The epiphyses were outlined by a ring-like dense line. X-rays of the skull were normal. The X-rays were considered to be diagnostic of scurvy. The child was treated with Vitamin C 100 mg. three times daily and was also given iron, folic acid and penicillin. She gradually lost her irritability and five days after her admission, she was moving her legs well and was free from pain and tenderness. However, she developed a bronchopneumonia one week after

admission and responded well to penicillin. Two weeks after admission, her heamoglobin was 10.1 gms.% and when discharged home a week later, she weighed 6.6 kgms.

CLINICAL PICTURE

Scurvy, as the full blown deficiency state, is easily recognized clinically in infants. It presents most frequently with the symptom triad of leg tenderness, irritability and pseudoparalysis. (19) If severe, there may be fever and anorexia. The physical signs are the scorbutic rosary, leg tenderness and swelling, the "pithed frog" position of hip abduction, lateral rotation and knee flexion and haemorrhagic hypertrophic gums.

Hypertrophic, perifollicular changes, especially over the forehead, may occur and petechiae and purpura may appear. The tourniquet test is at this stage usually positive.

Scurvy in its earlier stages presents a less clear picture. The child may become irritable and move about less, eventually regressing from an ability to stand or sit to that of lying. There may be a failure to thrive and a tendency to increased chest infections. The earliest physical signs include gum hypertrophy if teeth are present, characterized by a bluish purple discoloration of spongy swellings around the teeth which may at times conceal the teeth. As well the costochondral junctions become increasingly palpable although not yet visible and tenderness about the legs, especially the distal femur or any other epiphysis with fast growth, occurs.

The frequency of major signs and symptoms in scurvy patients will depend on the health facilities and medical standards in a given area as well as on the stage that the disease is allowed to reach before help is sought. The frequency of some of the presenting signs and symptoms in cases of scurvy in North America is shown in Tables 1 and 2.

Scurvy must be differentiated from ostiomyelitis, traumatic fractures, poliomyelitis, bone tumours, cellulitis, syphilitic periostitis, and from rickets. The gums and haemorrhogic changes must be differentiated from leukaemia, teething problems and bleeding disorders.

RADIOLOGICAL DIAGNOSIS

The radiological skeletal changes in scurvy are in general due to a decrease in normal bone cellular activity, both productive and destructive, while non-cellular activities, such as deposition of lime in the provisional zone of calcification and internal resorption of bone salts, are not disturbed. (1) This generalized bone atrophy results in a "ground glass" appearance of the shaft. With decreased cellular resorption, the zone of provisional calcification becomes thickened and casts a heavy transverse line on the radiograph. These findings are found in many non-scorbutic types of bone atrophy. But the combination of diffuse bone atrophy and multiple spurs at the cartilage shaft junction occur only in scurvy. This spurring occurs with the extension of the provisional zone of calcification laterad beyond the usual limits of the shaft. Fractures and fissures appear in this brittle zone, particularly in its lateral aspects. Atrophy and demineralisation on the metaphyseal side of the provisional zone of calcification produce a transverse radiolucent band termed the "scurvy line". Radiolucent laterad metaphyseal clefts through this weakened scurvy line result in a cortical and spongiosal defect called the "corner sign of scurvy". Both the "scurvy line" and the "corner sign of scurvy", when found with generalised bone atrophy, are valuable diagnostic features of scurvy.

Transverse fractures at the cartilage shaft junction may occur either through the weakened "scurvy line" or through the brittle provisional zone of calcification. These fractures may give rise to epiphyseal displacement or separation.

Although the diagnostic signs of scurvy are seen in the cartilage-shaft junctions of long bones, changes occur in other bones also. In the secondary centers of ossification, the changes parallel those described above, the center becoming atrophic and "ground glass" like and being surrounded by a dense zone of provisional calcification, called the "ring sign of scurvy". In the ribs, the costochondral junctions become enlarged and subluxed to give the "scorbutic rosary". This is a useful radiological sign as these enlarged joints can be easily seen on routine chest films. Subperiosteal haemorrhages may appear most commonly in the larger bones such as the femur, tibia and humerus and are noted as areas of increased radiolucency in the soft tissue shadows, or most easily after treatment begins, when a shell of radioopaque subperiosteal bone is laid down around the hematoma.

LABORATORY DIAGNOSIS

Although it is considered that positive clinical and radiological features are sufficient to make a diagnosis of scurvy, laboratory analyses of Vitamin C nutrition are available as additional diagnostic aids.

After beginning a completely Vitamin C deficient

diet, the plasma Vitamin C level reaches zero in approximately 40 days;(2),in the whole blood the level reaches zero in 80-90 days, (16) and in the white cell-platelet layer it reaches zero in approximately 120 days. (11) the signs of scurvy appear in approximately 130 days. (2)

Whereas the white cell-platelet layer Vitamin C level gives the best index of scurvy and a reading of zero equates well with clinical scurvy, this is a difficult study and not readily available. Serum Vitamin C levels are of little value other than for large nutritional surveys, except that a fasting Vitamin C level of over 0.6 mgm/100ml aids in the exclusion of scurvy as a diagnosis.

Many Vitamin C loading tests are available, a common one being the intramuscular administration of 200 mgm. of Vitamin C to a fasting infant, with collection of urine and blood after at 4 hours. (12) A low urine excretion of 15% of load or less and a low serum Vit. C level, is compatible with but not diagnostic of scurvy.

A more quantitative measure of Vitamin C level is a saturation test (11) based on the number of days required to produce a urinary excretion of 50 mgm. of ascorbic acid during the 4th–7th hour after a daily dose of 5 mgm/1b.

This test correlates well with the actual white blood cell-platelet layer Vitamin C level but as with most loading tests, is cumbersome and time consuming.

DISCUSSION

Four cases of scurvy have been found in two Malaysian hospitals over a five-month period, a large number for a disease which is considered rare. Several reasons are postulated for this number of cases. Both lack of recognition on the part of doctors unfamiliar with infantile scurvy and lack of medical services to the rural and poor urban populations who appear to be the ones most likely to suffer from the disease, partially explain the rarity in the past of this disease. All four children reported in this paper developed scurvy on diets of sweetened condensed milk. In the past, when a mother could not breast feed an infant, a wet nurse was found. Now bottle feeding is more common. Sweetened condensed milk, which contains no Vitamin C, is the cheapest milk product available and many manufacturers advertise it as a baby food. Unless the mothers' standards of health education and knowledge of infant nutrition are greatly improved, or manufacturers are required by law, or themselves show enough public spirit, to indicate on the cans, the dangers of sweetened condensed milk if Vitamin C is not taken as a supplement, it would appear that infantile scurvy may be on the rise in Malaysia.

SUMMARY

Infantile scurvy has been reported to be a rare tropical disease. We report four cases diagnosed during a 6-month period in two hospitals in Kuala Lumpur. The diagnostic value of some of the radiological signs and laboratory tests have been received. The reasons for the rarity of reports in the past and for possible increasing incidence in the future have been discussed.

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