Scurvy in Pediatric Patients : A Review of 28 Cases

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Abstract

Objective : To study the dietary factors, clinical findings, plasma vitamin C level and post-treatment outcome of scurvy in pediatric patients at Queen Sirikit National Institute of Child Health.

Study design : A retrospective study.

Method : The medical and radiographic records of pediatric patients, diagnosed with scurvy at Queen Sirikit National Institute of Child Health from 1995 to 2002 were reviewed.

Results : Twenty-eight pediatric patients were diagnosed with scurvy. Their ages ranged between 10 months - 9 years 7 months (median age of 29 months). 93 per cent of the cases were between 1-4 years of age. All were fed with well-cooked foods and small amounts or no vegetables and fruits. Supplementation with ultra heat temperature (UHT) milk was found in 89 per cent, average 5.8 boxes/day and 14.3 months in duration. Eighty-six per cent of cases were misdiagnosed previously. Clinical manifestations involved limp or inability to walk (96%), tenderness of lower limbs (86%), bleeding per gum (36%), fever (18%), and petechial hemorrhage (3.6%). All cases had abnormal radiographic findings compatible with scurvy and 2 cases had epiphyseal separation. All had clinical improvement within the first week after vitamin C supplementation.

Conclusion : Vitamin C deficiency was found in the children's intake of small amounts or no vegetables and fruits together with UHT-milk. Frequent manifestations were limping and inability to walk and pain in the lower limbs. Response to vitamin C treatment was dramatic.

Key word : Vitamin C Deficiency, Scurvy, UHT Milk, Limp, Gum Bleeding, Children

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The epidemiology of scurvy or vitamin C deficiency, differs from the past to the present. During the 14 th and 16 th centuries, scurvy occurred among sea voyagers who had traveled long distances without fresh fruits and vegetables[1-3]. At the end of the 19th century, usage of heated milk and proprietary food lacking of vitamin C, led to infantile scurvy[1,2]. Now scurvy is unusual after food fortification and supplementation with fresh vegetables, fruit or fruit juice, but it occasionally exists among refugees[4]. In Thailand, although plenty of tropical fresh fruits and vegetables are available around the year, scurvy in children still occurs because of improper feeding by their parents [5-7]. It is difficult for inexperienced physicians to diagnose scurvy. From 1985 to 1994, 21 pediatric patients were treated as scurvy at Queen Sirikit National Institute of Child Health (QSNICH), the tertiary referral center for children[5]. From the year 1995 to 2002, the diagnosis of scurvy still remains. Scurvy is an easily preventable disease, and could have been eradicated. A study on the contributing factors of the disease and clinical findings may find out some differences from the past and provide some light on scurvy prevention in the population.

MATERIAL AND METHOD

A retrospective study was performed on medical and roentgenographic records of patients, diagnosed with scurvy from 1995 to 2002, at Queen Sirikit National Institute of Child Health. The data were analyzed on the demographic data, weight, dietary history, symptoms and signs of scurvy, laboratory and radiographic findings, and results after treatment. Cases with inadequate clinical data and radiographic findings were excluded from the present study.

Nominal data were analyzed into number and per cent. Numerical data were analyzed as range, median, mean, and standard deviation (SD). Nutritional status was analyzed using the Thai standard growth reference (1999) as % weight for age (%W/A). %W/A was calculated from the patient's weight in kilograms x 100, divided by the standard median weight of the patient's age. The Gomez's classification was as follows: %W/A was $\leq$ 90, > 90-110, and > 110, classified as underweight, normal and overweight respectively[8].

RESULTS

During the eight-year period (1995 to 2002), thirty-one patients were diagnosed as scurvy. Three cases were excluded from the study. Twenty-eight patients comprised of 17 males (61%) and 11 females (39%), with an average age of 31.9 months (range 10 months - 9 years 7 months, median age 29 months) in Fig 1. Nutritional status was classified as underweight 12 cases (43%), normal 10 cases (36%), and overweight 6 cases (21%). Dietary history revealed that the intake of fresh vegetables and fruit or fruit juice was small in

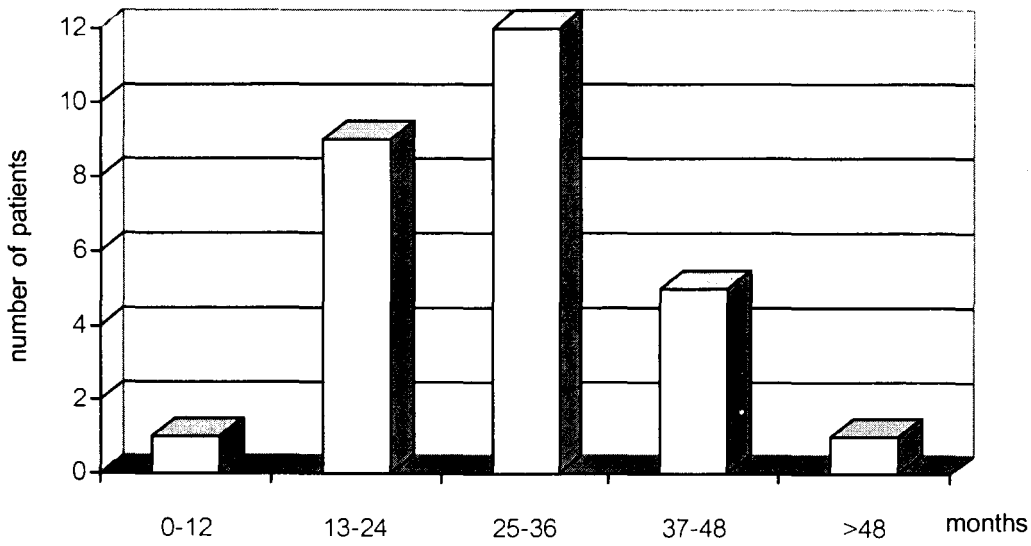

Fig. 1. Age distribution.

11 cases (39%), or none 17 cases (61%) and were fed daily with cooked food. Twenty-seven cases had milk intakes as follows: UHT milk (cow's milk and soy milk) 24 cases (89%), homemade soy-milk 2 cases (7%), and diluted powder whole milk 1 case (4%). The average intake was 5.8 boxes (250-300 ml) per day (SD = 1.5, ranged 3-10 boxes) with a mean feeding duration of 14.3 months (SD = 6, ranged 6-24 months). Two non-ambulated cerebral palsy boys with mental retardation were dependently fed with non-milk diet by their parents. These two boys were a 3-year-7-month malnourished boy with fever and marked swelling in both knees from epiphyseal separation of the distal femur and ipsilateral distal tibia, and the oldest boy, aged 9 years and 7 months, presented with ecchymosis of the right eye from minute trauma in his bed.

The average time of delayed diagnosis (after having symptoms) was 3.9 weeks (SD = 3, ranged 1-16 weeks). Twenty-four patients (86%) were previously treated by at least one physician before diagnosis of scurvy at QSNICH. The symptoms and signs were as follows: inability to walk (Pseudoparalysis) or limp 96 per cent, pain in the lower limbs 89 per cent, small trauma accounted for 46 per cent, tenderness of lower limbs 86 per cent, swollen knee and ankle joints 46 per cent, and gum hypertrophy and bleeding 36 per cent as shown in Table 1. Laboratory findings revealed anemia 76 per cent (19 cases), and 95 per cent were diagnosed with iron deficiency anemia (hypochromic-microcytic red blood cells). The cases that were checked for plasma vitamin C level, all had low levels and radiographic changes of the knee joints as shown in Table 2. Two cases of epiphyseal separation reported as epiphyseal separation or fracture of the distal femur and distal tibia in one cerebral palsy case and the other was distal femoral physeal collapse, who underwent closed reduction and percutaneous pinning. Two boys including a 1-year-9-month-old and a 2-year-10-month-old boy came with fever, limb pain and clinical right-sided heart failure who were treated as heart failure and with vitamin C and thiamin supplementation. Pulmonary hypertension was diagnosed by echocardiogram. Moderate pericardial effusion was only found in the younger boy and hypertension was recorded in the older boy. Both of them did not have any neurological signs of beriberi and diuretic effects from thiamin treatment. Clinical signs of limb pain and heart failure improved after a few days and normal echocardiograms were recorded 1 week after treatment. Hypertension in the older boy was slow improvement in two weeks.

Treatments of scurvy included vitamin C 150-300 mg per day in all cases. The underweight cases were supplemented with multivitamins and iron was supplemented in cases suspected of iron deficiency anemia. After treatment, all symptoms and signs was gradually reduced and appetite increased after a few days. In 23 follow-up cases, pain and tenderness of the lower limbs disappeared, on average 2.2 weeks (SD = 1.2, ranged 1-4 weeks), normal gait was seen on average 3.5 weeks (SD = 1.7, ranged 1-8 weeks). The swollen and bleeding gums appeared normal on average 1.5 weeks (ranged 1-3 weeks). All cases with fever became afebrile within one week. Radiographic bone changes of all 20 follow-up cases revealed an increase in bone density and resolution of the radiolucent line after an average of 4 weeks (SD = 1.2, ranged 2-6 weeks). Subperiosteal bleeding with periosteal new bone formation along the shaft of the long bone in a follow-up roentgenogram was seen in 7 cases (33%). The parents of all cases were educated and advised about complete nutrient intake and the appropriate food for their children.

Table 1. Symptoms and signs.

Symptoms and signs	Number	%
Inability to walk or limp (previously ambulated cases = 25)	24	96
Pain in the limbs	25	96
Small trauma accounted	13	46
Gum bleeding	12	43
Tenderness of lower limbs	24	86
Swelling of the lower limb joints	13	46
Gum hypertrophy or bleeding	10	36
Fever	5	18
Petechial hemorrhage	1	3.6

Table 2. Laboratory and radiographic findings of distal femur.

Findings	Number	%
Anemia (hemoglobin < 11g/dl) (n = 25)	19	75
Microcytic and hypochromic anemia (n = 16)	15	95
Low plasma vitamin C level (normal 0.31–2.1 mg/dl) (n = 13)	13	100
Dense white line of Freankel (accumulation of calcified cartilage)	28	100
Radiolucent line (Scurvy line as diagnostic of scurvy)	28	100
Bone atrophy (decreased radiodensity)	22	79
Metaphyseal spur (Pelken's spur)	9	32
Ring epiphysis (Wimberger's sign)	8	29
Subepiphyseal infraction (Corner sign)	4	14
Epiphyseal separation	2	7

Table 3. Number of pediatric scurvy cases : Age distribution comparison between three studies.

Age (mo)	The past 50 years study, Canada[12]	Previous study,[5] 1985-1994	Present study, 1995-2002
0-12	55	0	1
13-48	11	20	26
>48	-	-	1

DISCUSSION

Vitamin C is an essential micronutrient required for normal metabolic functioning of the body [3,9-11]. Humans cannot synthesize vitamin C so exogenous sources must be obtained through the diet. From Table 3, scurvy in the present and previous studies at QSNICH, were mostly preschool-age, but the average age of the present study was 31.9 months, higher than 27.1 months in the previous study[5]. Because a boy, aged 9 years and 7 months, was included in the present study. No infants were included in the previous study [5]. From 1953 to 1962, 66 cases of infantile scurvy were recognized, due to feeding with pasteurized or boiled in which vitamin C was destroyed and 83 per cent were aged between 5-12 months[12]. The cases at QSNICH were fed with breast milk or fortified infant formula, and changed to UHT milk after infancy. In the present study, only one 10-month old infant was underweight and fed with diluted formula. Scurvy occurs more frequently in children older than 6 months of age. Because of adequate stores of vitamin C due to higher fetal and infant plasma vitamin C levels and sufficient amounts of vitamin C in breast milk, that is 3-10 times greater than maternal plasma level[3,9,13]. In the past most affected cases were fed with boiled or pasteurized milk which is different from nowadays [1,12]. Males had a higher prevalence of scurvy than females, male to female ratio was 1.5 : 1, which was similar to the previous report with ratios of 2 : 1[6]. and 2.5 : 1[5]. In a US population, males of all ages consistently ingested ~ 20 per cent more vitamin C than females[9]. Nutritional statuses classified as underweight, normal, and overweight were 43 per cent, 36 per cent, and 21 per cent respectively in the present study, compared to the previous study which found that underweight was similar to 41 per cent of the previous study, but overweight was higher than 6 per cent of the previous study[5].

In the present study, the dietary history revealed that intakes of fresh vegetables and fruit or fruit juice were inadequate. Vitamin C is plentiful in fresh fruits, and vegetables[3,9,10]. UHT milk supplementation was high, 89 per cent in a previous study, 100 per cent[5] and 50 per cent in Siriraj Hospital[6]. The possibility of a high intake of UHT milk may result from both availability and accessibility in feeding practices. A few products of vitamin C fortified UHT milk are also available in the market but are more expensive. Dairy products, meat, fish and eggs contain smaller amounts and grains contain none[3, 9]. A few cases had 3 meals with rice and meat as in previous reports[5,6]. Also approximately 90 per cent of vitamin C were significantly reduced because vitamin C may be lost in cooking and storage as follows: destruction by heat and oxygen and its loss in cook-

ing water[3,9,13]. In the present study, the intake of UHT milk ranged from 3 -10 boxes per day which was similar to a previous study, which ranged from 5-10 boxes per day[5]. A high intake of UHT milk provides enough caloric need for preschool children but it fills them up. Mean feeding duration of the present study was 14.3 months which is longer than 10.2 months in the previous study[5]. When vitamin C intake is low, body stores of vitamin C fall. Signs of scurvy are noted when the body store is below 300 mg, plasma vitamin C level may be near zero for as long as 3 months[3,11,13,14]. Feeding for young or handicapped children depends on their parents so proper supplementary food depends on the proper knowledge, attitude and practice of their parents. Older children are less dependent on their parents. So certain children with peculiar dietary habits result from parental misguidance or neglect[13,16]. As in the present study, most children with peculiar dietary habits of drinking many boxes of UHT milk and incomplete nutrients. Child neglect may be one of the predisposing causes of scurvy and malnourishment.

Twenty-four cases had delayed diagnosis because diagnosis for scurvy is not easy in an inexperienced physician, so it is possible to delay the diagnosis or even misdiagnosis it. Scurvy in children present a different picture from that of an adult, children are motionless from fear of pain, swelling of knee and gum, but there are no bruises or phrynodermia[2]. Clinical findings in the present study included inability to walk or limp in 96 per cent, pain in the knee 96 per cent, and gum bleeding 43 per cent, which is similar to other studies[5-7,15,17]. Skin manifestations which are common in older child and adult scurvy[7,18], was reported only in the oldest boy in the present study, who presented with ecchymosis of the right eye, swollen-bleeding gum, and petechial hemorrhage. With insufficient vitamin C, connective tissue disorders result in a weakening of collagenous structures, causing tooth loss, joint pains, bone and connective tissue disorders and poor wound healing are the associated manifestations of scurvy[3,13], Five cases had fever and four cases had subperiosteal hemorrhage. Anemia in the present study was reported to be 76 per cent and 95 per cent was iron deficiency anemia with hypochromic-microcytic red blood cells, that was lower than the other study which found iron deficiency anemia of 100 per cent[6], and was higher than a previous study 25 per cent[5]. Anemia in scurvy is believed to be multi-factorial, including blood loss, concurrent vitamin deficiencies (especially folate), and decreased iron absorption[3,9]. Vitamin C plays a role in the regulation of iron metabolism by enhancement of iron absorption[3,9]. All samples sent for plasma vitamin C levels had low levels and all cases had radiographic changes of the knee, as in another study[6]. Clinical signs of scurvy are associated with low plasma vitamin C values[9]. The radiographic findings in the present study were typical for scurvy [19] and quite similar to a previous study at QSNICH. [5] Radiographic changes are seen at the cartilage shaft junction and appear earliest at sites of most active growth, such as distal femur and the end of the tibia and fibula[11]. The clinical findings in the present study had some differences from the previous study [5], as follows. Two cases of epiphyseal separation reported as a rare complication of scurvy in another studies[20], and two cases of pulmonary hypertension and right-sided heart failure diagnosed by echocardiogram without available investigation for vitamin B1 level. Both cases of pulmonary hypertension did not have any neurological signs of beriberi and diuretic effects from thiamin treatment. So the pulmonary hypertension might not relate to thiamin deficiency. There was no previous report on pulmonary hypertension in scurvy but two cases in the present study responded to vitamin C treatment. A 2-year-10-month-old boy had hypertension and slow improvement after one week as a case of scurvy in 9-year-old developmentally delayed girl without an apparent secondary cause of severe hypertension[21].

The result of vitamin C treatment in this study was dramatic with gradual improvement of all physical signs in a few days as in other studies[5-7]. Radiographic bone changes of all follow-up cases revealed improvement and some cases were found to have periosteal new bone formation along the shaft of the long bone as in previous studies[7]. The minimal requirement of vitamin C to prevent overt scurvy symptoms is 8-10 mg/day[12], which is lower than 25 mg/day of the recommended daily allowance (RDA) or the recommended dietary intake (RDI) for infant and children[3,9-11]. These recommended values are determined primarily on the basis of prevention of deficiency and based on the lower level of its content in human milk[3, 9-11]. The parents of all cases were advised to feed their children with fresh fruit or fruit juice.

SUMMARY

Scurvy can affect children of any age mostly preschool children who have an inadequate intake of

fresh fruits and vegetables and lack vitamin C. It can present with pseudoparalysis of acute onset without any other signs and respond dramatically to vitamin C supplementation. Dietary history, clinical findings and radiographic findings, as well as the rapid clinical response after vitamin C supplement diagnose scurvy. It also further emphasizes the need to accelerate public health education to prevent scurvy.

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โรคขาดวิตามินซีในเด็ก : การศึกษาทบทวนผู้ป่วยจำนวน 28 ราย

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วัตถุประสงค์ : ศึกษาปัจจัยด้านอาหาร อาการแสดงทางคลินิก ระดับวิตามินซีในน้ำเลือดและผลการรักษา ในผู้ป่วยเด็กที่เป็นโรคขาดวิตามินซี (โรคลักปิดลักเปิด) ในสถาบันสุขภาพเด็กแห่งชาติมหาราชินี

รูปแบบการศึกษา : การศึกษาข้อมูลผู้ป่วยแบบย้อนหลัง

วิธีการศึกษา : ทบทวนบันทึกเวชระเบียนและภาพรังสีกระดูกของผู้ป่วยเด็ก ที่ได้รับการวินิจฉัยว่าเป็นโรคขาดวิตามินซี ที่สถาบันสุขภาพเด็กแห่งชาติมหาราชินี ตั้งแต่ปี พ.ศ. 2538-2545

ผลการศึกษา : ผู้ป่วยเด็กเป็นโรคขาดวิตามินซีจำนวน 28 คน เป็นเพศชายร้อยละ 61 และเพศหญิงร้อยละ 39 มีอายุ 10 เดือน - 9 ปี 7 เดือน (มัธยฐาน 29 เดือน) ร้อยละ 93 มีอายุ 1-4 ปี เด็กทุกรายบริโภคอาหารที่ผ่านการหุงต้มและรับประทานผัก-ผลไม้ปริมาณน้อยมากหรือไม่ได้เลย ผู้ป่วยกินนม UHT ร้อยละ89 เฉลี่ยวันละ 5.8 กล่องและนาน 14.3 เดือน ผู้ป่วยร้อยละ 86 เคยได้รับการวินิจฉัยไม่ถูกต้องมาก่อน อาการแสดงทางคลินิกที่พบ ได้แก่ เดินกระเผลกหรือไม่ยอมเดิน ร้อยละ 96 ปวดขาร้อยละ 86 เลือดออกตามไรฟันร้อยละ 36 มีไข้ร้อยละ 18 และมีจุดเลือดออกที่ผิวหนังร้อยละ 3.6 ผู้ป่วยทุกรายมีความผิดปกติทางภาพรังสีของกระดูกขาเข้าได้กับการขาดวิตามินซี และผู้ป่วย 2 รายมีกระดูกส่วนปลายแยก ทุกรายอาการเริ่มดีขึ้นภายในสัปดาห์แรกหลังการรักษาด้วยวิตามินซี

สรุป : โรคขาดวิตามินซีในเด็ก มีสาเหตุจากการไม่บริโภคผักและผลไม้ บริโภคแต่นม UHT อาการที่พบได้บ่อย คือ การเดินกระเผลก ไม่ยอมเดิน และปวดขา การตอบสนองดีต่อการรักษาด้วยวิตามินซี

คำสำคัญ : โรคขาดวิตามินซี (โรคลักปิดลักเปิด), นมยูเอ็ชที, เดินกระเผลก, เลือดออกตามไรฟัน, เด็ก

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