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Increase in the frequency of scurvy in children with food selectivity: A case series

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ABSTRACT

Scurvy is a disease caused by vitamin C deficiency. Although rare, in recent years, the number of scurvy cases in children with eating disorders has increased.

Its manifestations are varied because vitamin C is a cofactor in numerous processes, such as collagen synthesis. The typical skin manifestations include petechiae, bruising, and hyperkeratosis. Mucosal involvement manifests as gingivitis with hypertrophy, bleeding, and loss of teeth. The diagnosis is based on clinical findings and may be confirmed by measuring plasma vitamin C levels.

The objective of this study was to describe a cohort of patients diagnosed with scurvy in recent years, its clinical manifestations, and findings in relation to their eating behavior and neurodevelopmental disorders.

Keywords: *vitamin C; scurvy; pediatrics; restrictive diets.*

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INTRODUCTION

Scurvy is one of the oldest diseases.[1,2] In 1753, English physician Sir James Lind associated scurvy with vitamin C deficiency.[3,4]

Ascorbic acid plays a key role in the hydroxylation of collagen in the skin, mucous membranes, adnexa, blood vessels, bones, and teeth. Ascorbic acid is also involved in other biological processes, such as the synthesis of corticosteroids, aldosterone, and carnitine, iron absorption, hair keratin binding, and the regulation of tyrosine metabolism. It functions as an antioxidant by reducing free radicals and the damage they cause to lipids, proteins, DNA, and blood vessel walls. It is also involved in the hydroxylation of dopamine to noradrenaline.[5] Within the immune system, the most prominent functions of ascorbic acid include the stimulation of leukocytes (neutrophil functionality and monocyte movement) and the improvement of macrophage, neutrophil, and NK cell activity.[5]

Since it is not synthesized by humans, it depends exclusively on intake. Fresh and raw fruits and vegetables are the greatest source of vitamin C. The fruits containing the highest amount of vitamin C are citrus fruits, kiwi, mango, pineapple, strawberry, watermelon, and melon. The vegetables with the highest concentration are broccoli, cauliflower, chard, spinach, potato, red pepper, and tomato.[6,7]

Vitamin C deficiency causes defects in collagen formation and alterations in the production of chondroitin sulfate and the development of skin symptoms and subperiosteal bleeding. The clinical manifestations of vitamin C deficiency include asthenia, arthralgia, myalgia, bone pain, functional impotence, and hemorrhagic lesions, such as bruising, petechiae, gingival enlargement, and gum bleeding and necrosis.[4,8]

During breastfeeding, a vitamin C intake between 7 and 10 mg/day help to prevent deficiency. An intake of 30 mg/day is recommended during the first 6 months to provide an amount that promotes iron absorption and immune system functioning. Well-nourished, non-smoking pregnant women who eat fresh fruits and vegetables have adequate vitamin C levels in their breast milk.[9]

Although scurvy is currently a rare entity, we have observed an increase in the number of cases in our hospital in recent years.

A total of 38 cases were diagnosed between 2009 and 2017 (3.8 cases/year). Between 2018 and July 2023, 54 cases were reported (10.8 cases/year); this means that, in our hospital, cases trebled.

The objective of this study was to describe a case series of patients diagnosed from 2018 to date in a tertiary care children's hospital and the importance of an adequate history-taking on eating habits —especially in those patients with neurodevelopmental disorders— and of supplementing patients with restrictive diets.

CASE REPORTS

In the past 4 years, 54 patients were diagnosed with scurvy in our hospital.

Among all patients diagnosed between 2018 and 2023, 81.48% were male. The reason for consultation varied depending on the disease stage at the time of diagnosis. The prevailing symptom was pain in the lower limbs, followed by asthenia, adynamia, and fatigue. Petechiae and bruising, gingivitis, and bleeding were also common (*Figure 1*). Anemia was confirmed in most patients, with a mean of 9.1 g/dL in boys and 8.5 g/dL in girls (*Table 1*).

Supplementary tests showed that 25 patients (46.29%) had X-ray alterations (*Table 1*), including metaphysis thickening (white line of Frankel), subperiosteal bleeding, and radiolucent y transverse bands (Trummerfeld zone) *(Figure 2)*.

Neurodevelopmental disorders were noted in 45 patients (83.33%), with autism spectrum disorder (ASD) as the most common one. Four children had extreme food selectivity; an infant consumed boiled cow's milk and another received a plant-based beverage.

Among all analyzed patients, 49 had a normal weight (90.74%).

The mean plasma vitamin C levels in patients in whom it was measured was 0.16 mg/dL (*Table 1*). The reference values used by the laboratory established that a level between 0.6 and 2 mg/dL was normal.

Once treatment was started, 48 patients (88.88%) showed a significant improvement in symptoms in the first 24 hours.

DISCUSSION

Scurvy is characterized by the involvement of various body organs and systems. Initially, the symptoms are nonspecific, beginning with asthenia, adynamia, and fatigue. Subsequently, mucocutaneous symptoms appear, which are varied, not pathognomonic, but characteristic of scurvy. Follicular hyperkeratosis, petechiae, bruising, perifollicular purpura, and dryness are

Figure 1. Before and after treatment. Petechiae, bleeding, and gingivitis are observed

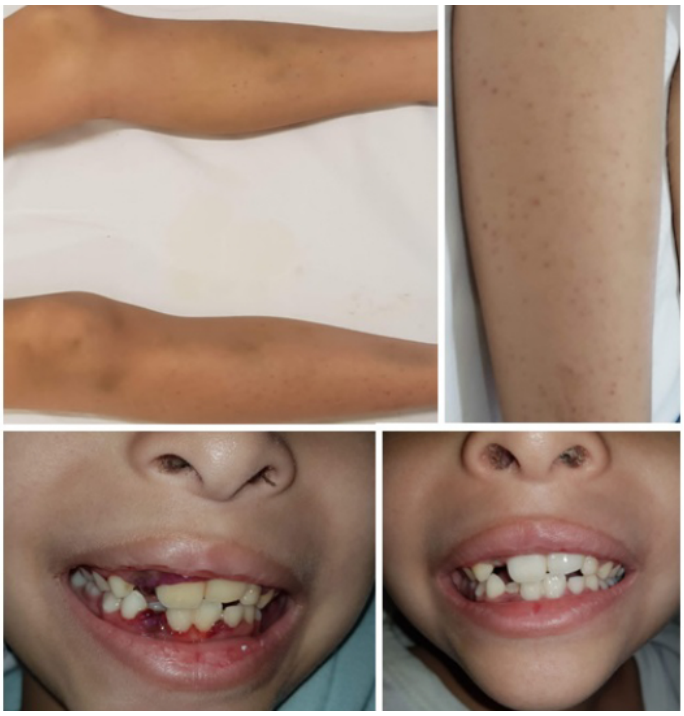


Table 1. Comparison of clinical findings and supplementary tests (N = 54 cases)

Variable	N	%
Age in years (mean)	10.68	
Sex	44 boys	81.48
Pain in lower limbs	43	79.62
Asthenia, adynamia, and fatigue	38	73.37
Petechiae and bruising	33	61.11
Gingivitis and gingival bleeding	24	44.44
Anemia	46	85.18
Pathological X-ray	25	46.29
Vitamin C measurement*	41	75.92
Plasma vitamin C level (mean and range)	0.16 mg/dL	0.08–0.59 mg/dL

N: number.

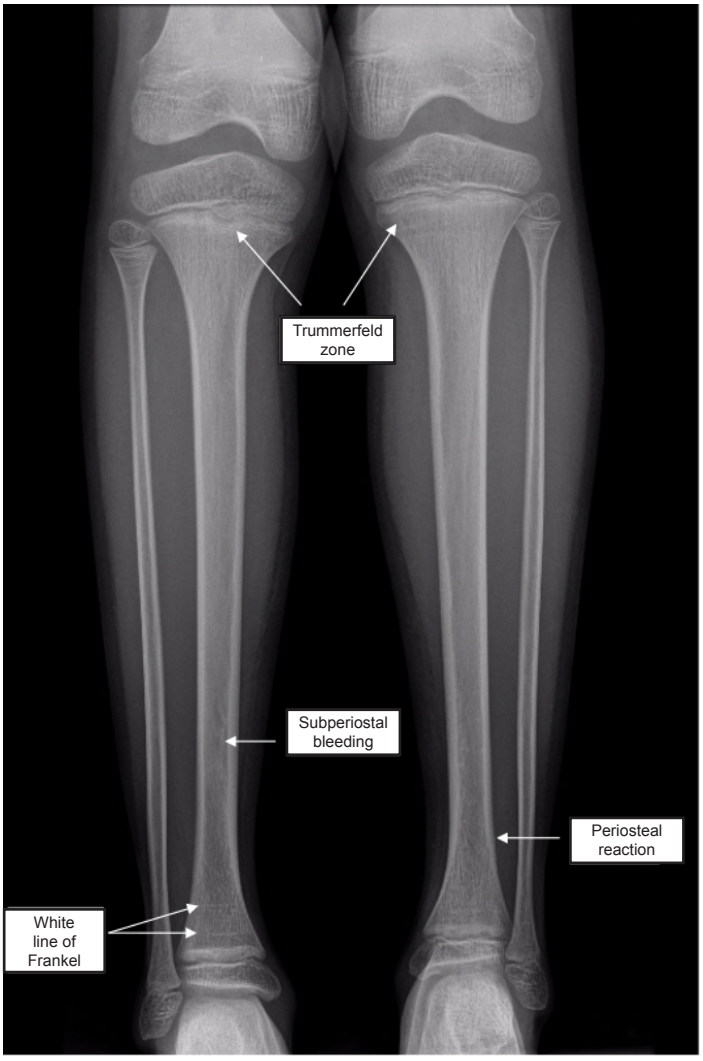
** Due to the fact that our hospital does not measure plasma vitamin C levels, in some cases, sample shipment was delayed and patients were given vitamin C.*

observed. Delayed wound healing has also been described. Mucosal involvement is the result of gingival enlargement, gingival bleeding, and gingivitis. If symptoms progress, bone resorption results in teeth loss.[7,10]

Once scurvy progresses even more, irritability, depression, arthralgia, myalgia, and hemarthrosis may develop. Pain in the lower limbs due to subperiosteal and intramuscular bleeding is typical, causing functional impotence and even pathological fractures.

Clinical manifestations develop after 1 to 3 months of inadequate vitamin C intake. Multifactorial anemia is very common due to bleeding, iron deficiency caused by impaired absorption, and folic acid deficiency. A high erythrocyte sedimentation rate and C-reactive protein level may also be observed as a consequence of the bone and gingival inflammatory process.[3,7,11]

Figure 2. X-ray of both lower limbs



If scurvy progresses, vasomotor instability and even death caused by brain hemorrhage may occur.

The diagnosis is made based on clinical findings; however, supplementary tests will help to confirm it. An X-ray may show a ground-glass pattern in the diaphyses of bones due to trabecular atrophy, periosteal detachments, signs of bleeding, widening of the zone of provisional calcification, thickening of the metaphyseal band —called white line of Frankel—, and a transverse radiolucent band, scurvy line, or Trummerfeld zone. A magnetic resonance imaging may show acute and healing subperiosteal hematomas, periostitis, metaphyseal changes, and heterogeneous bone marrow signal intensity. The lab tests usually describe anemia and increased acute phase reactants. The determination of plasma vitamin C levels is confirmatory, although it may be difficult because it is done in few centers, requires refrigeration, and a fast processing. Sometimes, vitamin C levels do not reflect the severity of the disease, since a minimal intake of vitamin C (for example, an orange) alters the determination. Levels < 0.6 mg/dL (< 34 mcmol/L) are considered borderline; levels < 0.2 mg/dL (< 11 mcmol/L) suggest vitamin C deficiency.[4,11,12]

The best evidence that it is scurvy, beyond supplementary tests and clinical symptoms, is the resolution of manifestations once treatment is started. In children, the therapeutical dose of vitamin C is 100 to 300 mg/day for 1 month or until complete recovery. Subsequently, supplementation should be indicated until normal plasma levels are reached. In general, signs and

symptoms develop when ascorbic acid levels are below 0.2 mg/dL. The patient's general condition, fatigue, lethargy, and irritability subside within 24 hours of supplementation. Muscle pain improves in 2 to 3 days, while bruising, bleeding, gingival bleeding, and weakness usually improve within 1 to 2 weeks.[4,13]

As part of the differential diagnosis, rickets must be considered. Rickets shows low calcium and high alkaline phosphatase levels, which are normal in scurvy. Hematooncological diseases should also be taken into account, such as leukemia, thrombocytopenic purpura, rheumatic diseases, like dermatomyositis, Guillain-Barré syndrome, and some types of vasculitis. In infants and toddlers, congenital syphilis should be ruled out.

An assessment of dietary characteristics, habits, and variety should be part of pediatric health checkup in all patients, especially in the population with neurodevelopmental disorders.

It is important to be aware of scurvy and recognize its clinical manifestations, since it seems to have reemerged in association with eating disorders of diverse etiology (ASD, anorexia nervosa, extreme diets, food selectivity, replacement of breastfeeding with inappropriate foods).[14,15]

Pediatricians should consider the diagnosis of scurvy when patients present with myalgia, difficulty walking, petechiae, bruising, or gingival enlargement.

Suspecting and diagnosing scurvy in a timely manner makes it possible to start treatment immediately and thus prevent its most serious manifestations.

CONCLUSION

In recent years, we have observed an increase in the number of scurvy cases in our hospital as a consequence of extreme food selectivity, especially in patients with neurodevelopmental disorders. It is critical to address their nutrition and supplement those with a severely restrictive diet. ■

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