



Case Report

Scurvy in pediatrics, revisiting a historical disease: A report of two children

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Received: 29 July 2025
Accepted: 13 October 2025
Epub ahead of print: 25 November 2025
Published:DOI
10.25259/JMSR_359_2025

Quick Response Code:



ABSTRACT

Scurvy is a forgotten disease of childhood that occurs secondary to prolonged severe dietary deficiency of vitamin C. It can manifest as a multisystemic disease resulting from prolonged severe deficiency of vitamin C. Musculoskeletal symptoms can be the presenting signs in children with physical or mental disabilities. We are presenting two children with scurvy. Patient A is a 4-year-old irritable boy maintained on boiled food since birth and presenting with wrist and hip pain. Radiographs revealed a subperiosteal hematoma, Wimberger ring sign, and white line of Fraenkel. Patient B is a 30-month-old girl with bowed legs, maintained on milk only since birth, and presented with bilateral leg/knee arthralgias and decreasing ambulation; she also had pithed frog leg posture. Pelvic radiographs revealed osteopenia, narrow physis, pencil-thin cortex, and Pelkan spurs. Vitamin C levels were low (<0.2 mg/dL), and symptoms resolved with vitamin C supplementation. Despite the rarity of vitamin C deficiency among pediatric patients, clinicians need to consider scurvy in the context of proper clinical findings to avoid misdiagnosis and delayed treatment.

Keywords: Arthralgia, Child, Scurvy, Subperiosteal hematoma, Vitamin C deficiency

INTRODUCTION

Scurvy is an often-overlooked childhood disease that arises from prolonged and severe dietary deficiency of vitamin C.^[1] Although considered a rare disease, it has been increasingly reported in children with restrictive nutritional habits, physical or mental disabilities.^[2] Vitamin C deficiency is estimated to affect 7% of the United States population, and as per US pediatric data (2016–2020), the incidence of scurvy increased from 8.2 to 26.7/100,000.^[3,4]

Vitamin C, also known as ascorbic acid, is an essential component found in fresh fruits and vegetables but is vulnerable to heat, ultraviolet radiation, and oxygen.^[3] It is biologically involved in the absorption of iron, wound healing, collagen formation, neurotransmitter synthesis, and the functioning of the immune system.^[3] Vitamin C also acts as a cofactor to prolyl hydroxylase and lysyl hydroxylase that stabilizes collagen and provides structural strength; hence, deficiencies result in poor osteoid bone formation and disruption of enchondral bone formation, connective and soft tissue disturbance exhibiting musculoskeletal manifestations reaching 80% and fragile capillaries, leading to bleeding complications from mucosa, intramuscular, intraarticular, and subperiosteal.^[3]

How to cite this article: Hammad AM, Barakat A, Abdelnoor J. Scurvy in pediatrics, revisiting a historical disease: A report of two children. J Musculoskelet Surg Res. doi: 10.25259/JMSR_359_2025

Scurvy can mimic several conditions, including autoimmune diseases, infections, and neoplasia.^[2] It should be suspected in patients with musculoskeletal complaints in the proper clinical settings. The latter is usually symmetrical and manifests as limb or joint pain, limping, and even refusal to walk in pediatric patients, often presenting as the initial symptom.^[2,5] The disease spectrum varies widely and additionally involves dermatologic, dental, and systemic manifestations.^[1] Common radiographic findings include subperiosteal hematoma, ring epiphysis, metaphyseal white line of Fraenkel, Trümmerfeld refraction zone, and epiphyseal slips.^[1]

The report at hand presents 2 cases of scurvy in pediatric patients. It aims to highlight the clinical and radiographic manifestations of pediatric scurvy, discuss the importance of dietary habits and vitamin C deficiency in the development of systemic disease and associated musculoskeletal symptoms that can be mislabeled as psychoneurological diseases. It also questions whether scurvy is still present in modern pediatric populations, and how careful clinical assessment and radiographic evaluation can help differentiate it from more common conditions. Both oral and written informed consents were obtained regarding the case study and future publications.

CASE REPORT

Patient A is a 4-year-old boy born healthy and full-term, who was referred to our care because the patient has been irritable and psychologically disturbed most of the day and nighttime, which was attributed to psychological problems that needed further investigation. The patient was seen by a psychologist but was irritable with an unclear diagnosis and was not started on treatment. On physical examination, the patient appeared to be neurologically intact; however, he experienced joint pain, primarily in the wrist and hip joints, which did not interfere with ambulation. Hence, the patient was referred to the orthopedics clinic. In the context of irritability and arthralgias, the pelvic radiograph was normal, but the wrist radiograph revealed subtle signs at the distal radius corresponding to subperiosteal hematoma, Wimberger ring sign, and white line of Fraenkel [Figure 1]. With the suspicion of vitamin C deficiency, the mother reported an unusual dietary habit of boiling all food, even vegetables, before feeding her child. Serology test showed normal laboratory values, including inflammatory markers, sedimentation rates, hemoglobin, coagulation profile, serum calcium, phosphorus, alkaline phosphatase, and vitamin D levels. However, vitamin C levels were low ($<11 \mu\text{mol/L}$, $<0.2 \text{ mg/dL}$).

Patient B is a 30-month-old girl born healthy and full-term, who presented to the orthopedics clinic due to new onset findings of bowing of both legs. On initial investigation, the patient was misdiagnosed with vitamin D deficiency due to

an insufficient vitamin D level (23 ng/mL) and was started on supplementation. The mother reported, however, that despite the vitamin D supplements and maintaining her daughter since birth on milk only without additional sources of nutrition, the pain had improved only slightly, and that her child was walking less and had pain in her legs. On further examination, the child had bilateral mild knee swelling and tenderness at the distal femur, painful knee range of motion, and inability to bear weight. There was also a notable pithed frog posture when the legs were free [Figure 2].



Figure 1: A 4-year-old boy presenting with irritability and psychological disturbance. Radiographs of the wrist of patient A showing (A) subperiosteal hematoma, (B) white line of Fraenkel, and (C) Wimberger ring sign.



Figure 2: A 30-month-old girl presenting with bowed legs in a pithed frog leg posture.

The pelvic radiographs revealed femoral osteopenia, subperiosteal hematoma, narrow physis, pencil-thin cortex, and Pelkan spurs [Figure 3]. To further assess the possibility of scurvy disease, all laboratory values were normal except for a deficient vitamin C level of <0.1 mg/dL.

Given a dietary history deficient in vitamin C, including a lack of fresh fruits and vegetables, with pathognomonic radiographic findings and low serum vitamin C levels, a diagnosis of scurvy was confirmed in patients A and B. In addition, neither patient exhibited any additional symptoms, such as petechiae, corkscrew hair, gingival bleeding, or gastrointestinal manifestations. Hence, the children were treated with a daily supplementation of 200 mg of vitamin C for 1 week, followed by 100 mg daily until symptoms resolved. At the 4-month follow-up for patient A and the 6-month follow-up for patient B, patients were symptom-free. They had returned to normal activity with complete resolution of bone-related changes clinically and radiographically [Figure 4]. It is noteworthy that the irritability and psychological distress of patient A had gradually improved until they completely resolved at final follow-up, 9 months after supplementation.

DISCUSSION

Vitamin C deficiency refers to inadequate intake or low levels of ascorbic acid that may be asymptomatic or cause mild, non-specific symptoms such as fatigue or malaise. Scurvy is the clinical disease state that develops from prolonged, severe vitamin C deficiency and is characterized by impaired collagen synthesis.^[6] Scurvy is a rare disease, especially among pediatric patients nowadays. It is often related to a known comorbidity or selective dietary habits. It should be suspected in children with musculoskeletal complaints, not only those with risk factors, but also healthy ones.^[2] Scurvy is primarily observed in children and youth with autism spectrum disorder and feeding problems.^[7] Being less common than other nutritional deficiencies, it is rarely suspected and more frequently missed.^[1] Our cases are unique in that extreme nutritional habits can alter vitamin C levels, leading to a range of multisystemic manifestations of scurvy disease that can go unnoticed or misdiagnosed as vitamin D deficiency or even psychological disease.

A thorough history and examination, followed by radiographs, are necessary to arrive at a diagnosis. Symptoms appear to develop within 1–3 months of vitamin C deprivation when the body stores have decreased to below 300 mg or the plasma level is <0.2 mg/dL.^[8] Initial symptoms are nonspecific and include loss of appetite, irritability, low-grade fever, and later ecchymosis, petechiae, hyperkeratosis, and corkscrew hair, secondary to fragile capillaries and an increased tendency for hemorrhage due to reduced thrombocyte aggregation and platelet dysfunction.^[9] Early



Figure 3: A 30-month-old girl presenting with bowing of both legs. Radiographs of the pelvis of patient B showing (A) subperiosteal hematoma, (B) pencil-thin cortex, (C) Pelkan spurs at the distal femur, and (D) narrow physis.



Figure 4: (a) A 4-year-old boy and (b) a 30-month-old girl (right) showing evidence of healing on radiographs of the wrist and knee, respectively, after supplementation with vitamin C.

symptoms are then followed by leg swelling at the thighs, knees, and ankles, then arthralgias and limping due to subperiosteal, intramuscular, and intraarticular hemorrhage and physal fractures. The latter musculoskeletal features result in the typical pithed frog posture with hips and knees semiflexed. Gingival manifestations are due to poor dentine formation.^[1]

There are several radiological findings of scurvy; osteopenia remains the most common finding, albeit nonspecific. An irregular thickened cartilage appears as a white line of

Fraenkel at the metaphysis. The Trümmerfeld zone is a late-specific feature that appears as a lucent metaphyseal band underlying the Fraenkel line, secondary to poorly formed trabeculae. At the periphery of metaphysis are the Pelkan spurs that are associated with periosteal elevation and healing fractures.^[1] A circular shadow surrounding the epiphyseal ossification center is known as the Wimberger ring sign. Plain radiography is usually enough for identifying scurvy features; nonetheless, an inexperienced radiologist should be aware of the features for proper diagnosis. Ultrasound may also reveal bony irregularity and bulky subcutaneous planes, whereas magnetic resonance imaging is performed in cases simulating malignant features, especially leukemia.

Ascorbic acid deficiency can alter hemoglobin and leukocyte concentrations, as well as the coagulation profile. In cases of scurvy, laboratory tests can show elevated erythrocyte sedimentation rate, iron deficiency, and microcytic anemia but normal serum calcium, phosphorus, and alkaline phosphatase. Leukocyte concentrations $\leq 10 \text{ mg}/10^8$ white blood cells are considered deficient and indicate latent scurvy.

Scurvy symptoms of bleeding, oral and constitutional symptoms recede with ascorbic acid replacement in a few days, whereas dermatologic and bony abnormalities and radiographic signs take a few weeks to improve.^[8] Infants and children are typically treated with 100–300 mg of vitamin C daily for at least 1 month or until full resolution of symptoms. However, children with scurvy due to restrictive feeding require ongoing intervention from a nutrition or feeding specialist.^[1] Treatment typically involves vitamin C therapy, analgesia, and rest, with no role for antibiotics.^[5]

It is of paramount importance to acknowledge that scurvy tends to occur sporadically and more often in low- and middle-income countries (LMICs). The cases presented in this study are from LMICs but resemble closely previously published reports of modern series of scurvy in children with restrictive diets. Masci *et al.*, reported on 8 children presenting with musculoskeletal symptoms, ranging from lower limb pain to overt limping (87%), and diagnosed with scurvy.^[10] Other pediatric presentations include atypical musculoskeletal symptoms such as lower extremity weakness and abnormal gait. Our patients exhibited the classic scurvy triad noted in the literature, including bleeding tendencies, corkscrew hairs, and gingival hypertrophy in addition to low vitamin C levels ($<0.2 \text{ mg/dL}$) that respond rapidly to supplementation. It is noteworthy that modern-day scurvy is seen in pediatric patients with autism, neurological, and psychiatric disorders.

Future efforts should focus on understanding the disease etiology, symptoms, and epidemiology, as well as the need for early nutritional intervention and anticipatory guidance, with the aim of preventing disease in the population at risk.^[6] When evaluating musculoskeletal and systemic

findings suggestive of scurvy, it is important to consider more common conditions with overlapping presentations. Rickets may present with bone pain, deformities, and radiographic changes such as metaphyseal cupping and fraying; however, it is secondary to vitamin D deficiency. Leukemia can mimic scurvy with bone pain, anemia, and subperiosteal hemorrhage but is usually accompanied by systemic features such as hepatosplenomegaly, lymphadenopathy, and abnormal blood counts. Septic arthritis may also present with joint swelling, pain, and functional limitation but typically demonstrates localized infection signs, elevated inflammatory markers, and positive cultures. Health professionals should be vigilant about diagnosing scurvy and the severe consequences if left untreated, especially as part of the differential diagnosis for atypical musculoskeletal pain.

CONCLUSION

Despite the rarity of vitamin C deficiency among pediatric patients, clinicians need to consider scurvy in the proper clinical findings and disease course to avoid misdiagnosis, unnecessary investigations, and delayed treatment. Scurvy can present in various forms and can mimic common diseases; therefore, a careful physical examination and a focused dietary and behavioral history, with confirmatory serology, can facilitate early diagnosis and prevent potentially fatal complications. Hence, pediatricians and orthopedic surgeons must consider scurvy in the differential diagnosis of atypical musculoskeletal pain.

Recommendations

Clinicians should maintain a high index of suspicion for scurvy in children with unexplained musculoskeletal pain and restricted diets. Early recognition through focused dietary history, imaging, and vitamin C testing allows prompt treatment and avoids unnecessary investigations. Nutritional counseling and vitamin C supplementation remain a key to both management and prevention.

Authors' contributions: JA conceived and designed the study. AB conducted research, provided research materials, and collected data. AMH analyzed and interpreted data. AMH wrote the initial and final draft of the article and provided logistical support. All authors have critically reviewed and approved the final draft and are responsible for the manuscript's content and similarity index.

Ethical approval: The research/study was approved by the Institutional Review Board at American University of Beirut Medical Center, number KA/56/2025, dated March 27, 2025.

Declaration of patient consent: The author certifies that he has obtained all appropriate patient consent forms. In the form, the patient's parents have given their consent for the patient's images and other clinical information to be reported in the journal. The parents understand that the patient's name and initials will not be

published, and due efforts will be made to conceal his/her identity, but anonymity cannot be guaranteed.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation: The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

Conflicts of interest: There are no conflicting relationships or activities.

Financial support and sponsorship: This study did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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