Scurvy
Yes, it exists

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Abstract
Scurvy is not gone from the face of the planet. In the past it was seen commonly in sailors and soldiers who had limited access to fresh fruits and vegetables over extended periods. Scurvy still exists today within certain populations, particularly in patients with neurodevelopmental disabilities, psychiatric illness or unusual dietary habits.
We describe a case of scurvy occurring appeared in a 4-year-old boy with a special diet such as milk and cookies from a young age, presented in a month earlier: asthenia, diffuse lower-extremity musculoskeletal pain, and refusal to walk.

Introduction
Scurvy stands out in our minds as something that is so basic and easy to avoid, and yet these people have ended up falling victim to an illness that simply should not exist in a developed country. Humans cannot synthesize vitamin C. It needs to come from external sources, especially fruits and vegetables, or fortified foods.
Scurvy can occur in children whose diet is restricted as a consequence of developmental or socioeconomic circumstances\(^1,2\). Almost exclusively in children with autism, developmental and behavioral issues, malabsorptive processes, or iron overload, and in disorders causing dysphagia or impaired swallowing\(^3,4\).
Until recently, in developed countries, the occurrence of scurvy in children had become a historical footnote, with most radiologists having never encountered acase\(^5\).

Case Presentation
A four year old boy with history of worsening musculoskeletal pain and refusal to walk for one month. His mother had noticed weight loss, lethargy, anorexia, fatigue and weakness. A bone story for which he landed in childhood surgery then the patient was referred to us. In his personal antecedents include the notion of six febrile convulsions brought under Depakine
Physical examination disclosed impaired general condition (weight loss, severe asthenia, anorexia), apyretric, hemodynamically stable, weighed 12kg (-2 DS according to the WHO curve) with a size of95 cm (between -1 and -2 DS according to the WHO curve)mucosal skin examination finds pallor, gingivitis perifollicular hemorrhages and extensive ecchymoses with increased pigmentation involving the lower extremities and a diffuse onpruritic perifollicular rash, edema of the lower limbs and feet (Figure 1). On physical
examination, with knees and hips flexed in antalgic posture, he had a marked limp favoring the left side and limited abduction and internal and external rotation of the left hip. Neurologic examination result was normal but the child was listless and indifferent. Further laboratory work was sent, which showed an deficient hypochromic microcytic anemia at 6g/dl of hemoglobin for which he received a blood transfusion. Bleeding disorder work up including prothrombin time, partial thromboplastin time, international normalized ratio (INR), platelet aggregation tests, and factor assay for factors VII, XIII and IX were normal. C-reactive protein, coagulation profile, and chemistry including calcium, phosphorus, and magnesium were also normal. Vitamin C blood test: less than 03µmol /l(normal 23–114 mmol/L), there were no other abnormalities, we even dosed the vitamin d which returned to 18.9 ng / ml. Given his progression of symptoms to the point of acutely not bearing weight, additional imaging was done. An ultrasound showed trace joint fluid the left hip, Radiographic findings in our case are a Wimberger ring sign, which denotes a thin sclerotic cortex surrounding a lucent epiphysis. Periosteal new bone formation secondary to subperiosteal hemorrhage, a dense provisional calcification immediately adjacent to the physis (Frankel line), and an adjacent lucent band more diaphyseal in location (Trummerfeld line) are also classically described. (Fig 2)

The patient was started on vitamin C supplementation at 100 mg every 8 hours for 1 week followed by 100 mg daily along with iron, vitamin D, and a multivitamin. On hospital day 7, there was a decrease in the number of petechia and in pain (the child could sit for a few minutes)

On day 20: the child became quiet, responsive and playful, disappearance of the pain (the child can sit down) with persistence of some petechia, weight gain (02kg), the child however remained always suspicious and apathetic. Rehabilitation was started as soon as the pain had subsided. He was discharged with follow-up appointments, including nutrition and feeding therapy. At his 3-month follow-up, his pain had resolved, and he was ambulating with persistence of a little stiffness and lameness, persistence of gingivitis and some petechiae in the trunk.

A marked improvement was noted on the psychological level since the child spoke laughed and even played with us during the consultation.

Fig1a: Gingivitis

Fig1b: perifollicular rash, edema
Discussion

Humans cannot synthesize vitamin C, so we depend strictly on exogenous contribution. Fruits and vegetables are the primary dietary source of this essential vitamin. Vitamin C is an essential cofactor for collagen synthesis, and defective collagen synthesis in scurvy leads to characteristic dermatologic and skeletal findings. Malfunction in collagen synthesis is the primary cause of clinical manifestations of vitamin C deficiency. Common findings include perifollicular hemorrhage, “corkscrew” hairs, ecchymoses, gingivitis, arthralgias, impaired wound-healing, and anemia.\(^6\)

Additional manifestations can include lower extremity edema, splinter hemorrhages, alopecia, poor wound healing, cardiac hypertrophy, psychological changes, and conjunctival hemorrhages, among many others.\(^7\)

There have been reports of scurvy re-emerging in certain special groups. These groups includes children with oral aversions, cerebral palsy, developmental delay, autism, food faddism and predominant dairy consumption\(^8-9\). Although scurvy can occur at any age, the incidence of scurvy peaks in children aged 6-12 months who are fed a diet deficient in citrus fruits or vegetables, as well as in elderly populations, who sometimes have "tea-and-toast" diets deficient in

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**Fig 2a:** Wimberger’s sign - circular, opaqueradiologic shadow surrounding epiphyseal centres of ossification

**Fig 2b:** Fraenkel’s line- densezone of provisional calcification; c, f: pencilthin cortices: d: ground glass appearance; e:Pelkan spur
vitamin C. Scurvy is uncommon in the neonatal period.\[10\]
Our case was a developmentally normal child albeit malnourished. His malnutrition was predominantly due to scarcity of food in general as well as excessive consumption of exclusive dairy products.

Pediatric patients commonly present with musculoskeletal manifestations. With damage to synovial blood vessels and microfractures, there is pain and swelling secondary to subperiosteal hemorrhages and hemarthroses.\[11\]

A case series from Thailand reported that 96% of children diagnosed with scurvy presented with inability to walk.\[12\]

Anemia is another hallmark feature of scurvy and can be multifaceted. Iron deficiency anemia often accompanies scurvy in part due to decreased absorption and also due to bleeding manifestations. It is a well-known fact that vitamin C helps in enhancing iron absorption by reducing iron to ferrous form. Thus in our child features suggestive of Iron deficiency anemia could have likely resulted from lack of vitamin C facilitated iron absorption.\[13\]

Our patient had microcytic anemia that required one blood transfusions.

Classic radiographic findings may include white line of Fraenkel, which is an irregular, thickened white line that appears at the metaphysis and represents an increased calcification of the cartilage matrix; the Trummerfeld zone, which is a zone of rarefaction beneath the Fraenkel line, which denotes subperiosteal hemorrhage; Wimberger ring sign, which is a white line that surrounds ossification nucleus in the epiphysis; and the Pelkan sign, which is the presence of metaphyseal spurs that appear later due to the repair of microfractures.\[14\]

Osteopenia, and cortical thinning, many signs have been reported in the literature.\[16-17\]

All of these signature radiological signs associated with scorbatic bone changes were present in our child and helped in clinching the diagnosis in this case.

The diagnosis of scurvy is primarily based on clinical presentation, x-ray findings of long bones, and a history of poor intake of vitamin C. The absence of characteristic radiologic changes in the context of significant clinical signs of scurvy is not uncommon.\[18\]

The dosage of serum vitamin C levels is considered specific but laboratory tests are insensitive. It is known that serum concentrations do not always correspond with tissue storage of ascorbic acid. A reliable indicator of body storage is the measure of urinary excretion after intravenous ascorbic acid administration\[19\]. The prompt resolution of symptoms after substitutive treatment represents the main evidence to confirm the diagnosis of scurvy\[20\].

Scurvy is preventable disease. Treatment of scurvy is based on replenishing the level of vitamin C to counteract the symptoms. There is no established regimen for vitamin C supplementation in scurvy. Children require at least 15 mg/day of VC to meet daily metabolic requirements and prevent clinical/subclinical development of ascorbic acid deficiency symptoms. The daily allowance recommended for infants is 40 to 50 mg, and for children 1 to 3 years old, it is 15 mg\[21\]. It is demonstrated, that ascorbic acid administration at an initial dose of 300 mg per day leads to complete resolution of symptoms within 4 weeks\[22\]. We chose the former regime with good results.

Constitutional symptoms generally significantly improve within days while bone abnormalities and ecchymoses gradually improve over several weeks\[23\]. Improvement usually begins in 24 hours, with pain diminishing in 2 to 4 days and gingival lesions recovering in 2 to 3 weeks. Once treated, the only damage from scurvy is lost teeth\[24\]. The evolution in our patient was not as rapid.
Conclusion
Scurvy is a diagnosis that could be missed easily. High index of suspicion together with detailed history taking are important to diagnose it early which lead to prompt initiation of treatment. It should be considered in high risk groups such as those with food faddisms, poor nutrition, and malabsorption. A possibility of scurvy should be entertained in any child with new onset pain and weakness especially in the absence of any neurological cause. Children with a restricted diet can be referred to a nutritionist or feeding therapist to ensure that they receive adequate nutrition from their diet to avoid easily correctable nutritional deficiencies and diseases such as scurvy. We also recommend that tests for vitamin C levels should be made available in the hospital laboratories, since scurvy is a disease that can still be seen nowadays.

Bibliography