

Author's reply;

Hypophosphatemic Rickets and its Dental Significance

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Although the premature tooth exfoliation (PTE) can be associated with some systemic diseases such as changes in immune system or connective tissue disorders, we didn't find any report on this disorder and hypophosphatemic rickets. In bone metabolic disorders, there are a few reports on hypophosphatasia and PTE^[1-3]. Although, some factors such as severe oral infection can predispose PTE in HR patients, we recommend a study on prevalence of PTE and its related factors in HR patients in future.

The inflammation of the gingiva was evaluated by Gingival Index of Loe and Silness index^[4]. It was missed to mention in our article. As we mentioned, dental examination was performed by the same dentist in all patients^[5].

We agree with Ramezani about the prevalence of taurodontism as a common dental problem with wide range of prevalence in normal population (up to 8%^[6]), but as we reported in our paper the prevalence of this disorder was 15% that is very more common than in normal population^[5]. It was reported in 0.3% of normal children^[7].

Finally, for diagnosis of taurodontism, as mentioned in our report, orthopantomogram was performed in all taurodontism suspicious patients^[5].

Key words: Hypophosphatemic Rickets; Taurodontism; Dental Problems

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Sandifer's Syndrome: a Misdiagnosed and Mysterious Disorder

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Sandifer's Syndrome (SS) is a rare pediatric manifestation of gastro-esophageal reflux (GER) disease characterized by abnormal and dystonic movements of the head, neck, eyes and trunk. Although Sandifer initially observed the association, Kinsbourne and Oxon first reported it based on the observations of Sandifer^[1]. The syndrome is most certainly underrecognized, and delays in diagnosis are due to atypical presentations or cases in which the diagnosis is not part of the differential^[2,3].

9 month old boy was referred with a 2 months history of vomiting. 15 days ago before admitting to our hospital, he started to make bizarre head and neck movements as told by his parents. The parents provided careful video recording of these movements. The most striking feature pronounced neck dystonia with repeated rotation of the neck and tilting of the head towards the left shoulder. These movements were observed during or just after the feeding. Sometimes upward deviation of the eyes and head nodding accompanied these movements. All of these movements stopped when he was asleep. The milestones of motor and

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mental development were normal. His physical and neurological examinations were normal.

The patient had undergone an extensive evaluation, including subspecialty consultations. Routine laboratory tests were normal. Orthopedic, otorhinolaryngologic and ophthalmologic evaluations were also normal. Cranial and cervical magnetic resonance imaging was normal. The video electroencephalograms (EEG) of the patients which were taken during the routine, sleepless and paroxysmal behaviors were normal. Cow's milk specific IgE was negative and skin prick test was normal. He had a macroscopically negative upper gastro-intestinal endoscopy; but esophageal biopsy revealed reflux esophagitis, with no hiatal hernia. Esophageal pH monitoring for 24 hour confirmed gastroesophageal acid reflux.

Management of the infant with GER disease, including domperidon, sodium alginate and lansoprasole was started. The paroxysmal dystonic behaviors were dramatically diminished with treatment, but not disappeared. Cow's milk was removed from his diet and after three days, all of the movements disappeared. Fifteen days later, cow milk was introduced to the diet of the patient and the dystonic movements started again. Cow's milk was removed once again from his diet and movements disappeared. So, SS was diagnosed due to cow's milk protein allergy.

The literature includes between 40 and 65 cases of SS^[4,5]. The exact pathophysiology of the movements in SS is not known. The head and neck posturing and the dystonic movements are thought to be a response to the pain associated with GER^[6]. Why some children with GER disease present with abnormal movements and others do not remains unresolved. Research supports the theory that the movements are learned behaviors used by children to reduce reflux. Kinsbourne and Oxon suggested that the 'child having hit upon this movement by chance may have found that it temporarily relieved the discomfort and therefore continued to practice the relevant movements'^[1]. We also suggest that dystonic movements are learned movements. Cow's milk protein allergy is a reproducible clinically abnormal reaction to cow's milk protein (CMP). Improvement or disappearance of symptoms on a CMP-free diet adds substantial evidence to the diagnosis. If the reintroduction of CMP causes relapse of symptoms, the diagnosis seems established,

because a challenge test is considered as the golden standard diagnostic test^[7]. In our patient, challenge test was positive, so we diagnosed our patient as SS due to cow's milk allergy. Corrado et al reported a Sandifer's syndrome in a breast fed infant. They proposed food allergy to dietary proteins ingested by a lactating mother may play a role^[4].

The clinical manifestations almost invariably arouse the suspicion of neurological disease. Patients with SS, especially infants, are often misdiagnosed due to their paroxysmal neurobehaviors, and they receive unnecessary medication. Moreover, these patients fail to receive primary treatment as regards GER disease^[3]. A critical point for diagnosis is that an ictal EEG iconography demonstrating a normal EEG activity during the paroxysmal event for excluding epileptic disorders. The fact that the patients respond well to treatment in spite of all these findings may make one think that GER disease, as well as SS, is a benign natured disorder. However, in cases where diagnosis is late and which have accompanying anatomic defects, response and prognosis may be worse^[8].

Key words: Sandifer; Gastroesophageal Reflux; Cow's Milk Protein Allergy; Children

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