Dental Treatment of Patient with Hypophosphatasia

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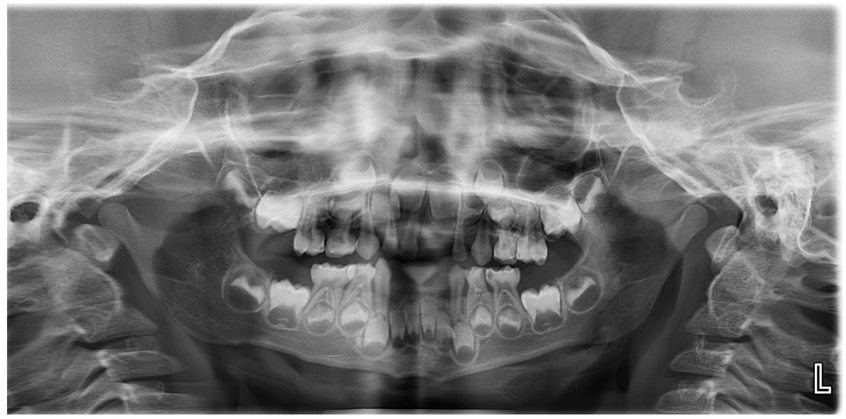
BACKGROUND:

Hypophosphatasia (HPP) is a progressive hereditary metabolic disorder characterized by defective mineralization of bones and teeth. HPP is caused by mutations in the ALPL (alkaline phosphatase) gene which encodes tissue non-specific alkaline phosphatase (TNSALP). With loss of TNSALP activity, inorganic pyrophosphate is not degraded, and phosphate is not produced; as a result, calcium and phosphate cannot bind. Ultimately, hydroxyapatite formation and outgrowth are disrupted resulting in impaired bone mineralization. Manifestations include stillbirth, impaired bone mineralization, and early exfoliation of primary teeth. Disturbances in the formation of cementum and dentin affects tooth attachment to the alveolar bone. Early diagnosis is important for patients to provide early intervention from both a medical and dental standpoint. Some existing literature has shown that enzyme replacement therapy can improve dental mineralization and aid in stabilization of periodontal tissues and improved root formation.

DIAGNOSIS:

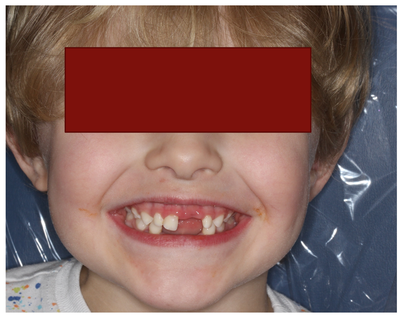
Hypophosphatasia is diagnosed according to clinical symptoms, radiographic findings, and biochemical test results. Low serum alkaline phosphatase (ALP) levels must be compared with reference values according to age and sex. Dental signs include early exfoliation of primary dentition, alveolar bone loss, enlarged pulp chambers and root canals, and thin dentinal walls. Other signs include skeletal fractures, chronic joint pain, short stature/unusual gait, and muscle weakness/fatigue.

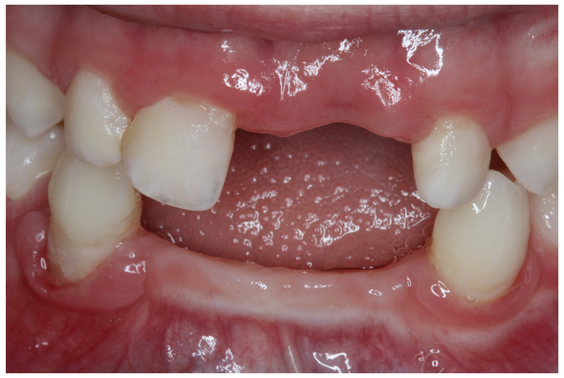
PANORAMIC RADIOGRAPH:



Reveals: significant mandibular anterior alveolar bone loss and evidence of enamel hypoplasia.

CLINICAL PHOTOS:





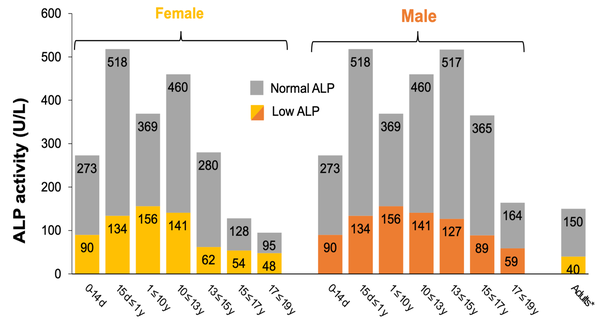
MANAGEMENT/TREATMENT:

Dentist role is to identify patients with hypophosphatasia and refer for proper management.
Treatment must include guidance on adequate oral hygiene to maintain good periodontal condition.
Partial dentures can be fabricated for primary teeth that have exfoliated.
A treatment plan for this patient was composed to address dental caries and stabilize remaining dentition which will include full mouth rehabilitation.

CLINICAL PRESENTATION:

A 4-year, 5-month-old male presents to the outpatient dental clinic at Riley Hospital for Children for an emergency dental visit. His parents were concerned of recent early exfoliation of #F. The patient's medical history is significant for hypophosphatasia. The patient is taking asfotase alfa (enzyme replacement therapy) which is administered subcutaneously 3 times per week; he has no known drug allergies. Early exfoliation of primary teeth began when patient was 2 years old. Intraoral exam reveals primary dentition and premature loss of #F, #N, #O, #P, #Q, as well as significant mobility of #L, #M, and #R. Localized gingival inflammation and periodontitis associated with #R. Clinical caries detected on #E, #I, #J, #K and #L.

AGE AND SEX ADJUSTED ALP REFERENCE RANGES:



REFERENCES:

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