# Dental Considerations in a Patient with X Linked Hypophosphatemia

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## BACKGROUND

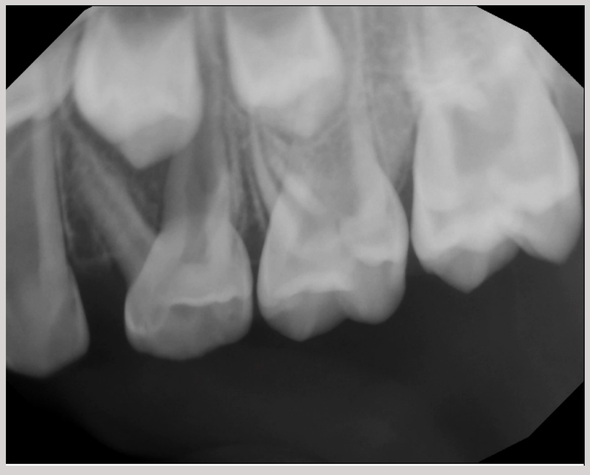
A 6 year, 8-month-old male presented to the Riley Hospital for Children outpatient dental clinic. His medical history was significant for X-Linked Hypophosphatemia for which he was taking Crysvita, a fibroblast growth factor 23 blocker. He had no reported drug allergies. This patient presented with mixed dentition and no clinical caries, but a clinical abscess associated with #I. X-Linked Hypophosphatemia (XLH) is known as the most common form of hereditary rickets. XLH is a rare, lifelong phosphate wasting disorder and is prevalent in 1 in every 20,000. XLH is due to a PHEX (Phosphate Regulating Endopeptidase X-Linked) gene mutation, which is thought to be involved in renal phosphate reabsorption and bone and dentin mineralization. Mutations in this gene ultimately lead to decreased serum calcium and potassium phosphate. Clinical presentation of XLH in children can consist of: Lower limb bone deformities, rickets, bone pain and failure to thrive. Dental manifestations of this condition include spontaneous abscesses that may occur without previous history of trauma or dental caries. The pediatric dentist plays a crucial role due to patients with XLH needing multidisciplinary care and helping to achieve optimal oral care.

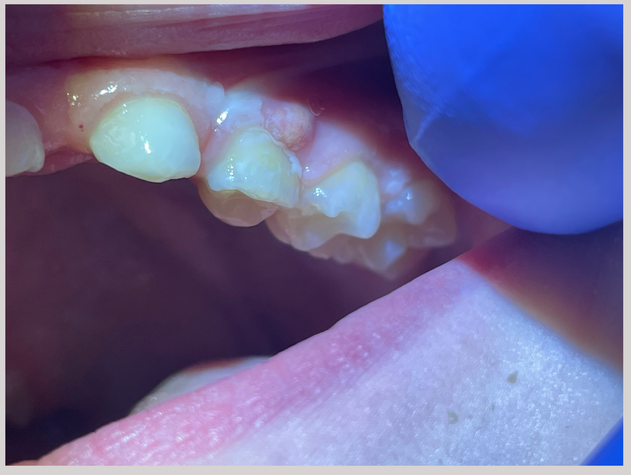
## CLINICAL PRESENTATION

An intraoral exam was performed and revealed mixed dentition. There were no clinical caries detected. Teeth #E and #F presented with a green/grey discoloration. Teeth #N and #Q were missing. A clinical abscess was detected on the buccal gingiva just apical to the crown of #I. Overall, the patient had good oral hygiene and no other significant findings.

## RADIOGRAPHIC INTERPRETATION

A periapical radiograph was taken of #I. The radiograph reveals a furcal radiolucency with internal resorption of the distal root. Widening of the PDL is also noted. No other pathology or significant findings were observed. In addition to the periapical radiograph, an intraoral photo was taken. The intraoral photo reveals a clinical abscess above the crown of #I, about 4mm x 4mm in size.





## Diagnosis and Treatment

Extraction of #I was recommended due to presence of an active infection. At the next appointment, extraction of #I was performed with no complications. The buccal periosteal space was curettaged by the resident to remove any residual infection. The patient handled procedures well and was scheduled back in 4-6 weeks for fabrication of a band and loop to preserve space for the permanent successor. The patient then returned to clinic and parents reported no pain or discomfort following the extraction. A band and loop was fabricated for #I. Post operative instructions and oral hygiene instructions were provided. The patient was then scheduled back to Riley Dental Clinic in 6 months for a periodic oral examination and prophylaxis.

## Follow Up Care

Key dental features with patients with XLH are spontaneous abscesses and dyschromic enamel. (2). With defects in enamel and dentin, patients are susceptible to caries and attrition. Other dental features that can be noticed are enlarged pulp chambers, dentinal defects, and thin enamel (3). Follow up care should consist of preventive measures such as sealants, application of topical fluoride varnish and frequent dental exams. These measures can help prevent microbial invasion that can ultimately lead to pulpitis. Overall patients with XLH can have various dental abnormalities. It is imperative that dentists identify clinical features of XLH. Providing early diagnosis, communication with the patient's pediatrician and consistent follow ups are key to dental management in patients with XLH (1).

## REFERENCES

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