

HPP children need special care for their teeth.

Early & regular dental care is very important.

Preventative Advice for Parents & Caregivers:

- Teeth in HPP children are very vulnerable. See the dentist by the child's first birthday!
- Most children need assistance with brushing until ~ 8 years of age
- Use fluoride toothpaste: use the size of a grain of rice if under 3 years; for children over 3 years, use a pea-size amount
- Regular flossing
- Dental sealants can be used to protect tooth enamel

Dietary Considerations:

- Dental-friendly snacks may help prevent cavities: these include fruits, vegetables, and cheese.
- Vitamin D-rich foods and supplements
- Limit drinks containing sugar and minimize snacking between meals
- Consider sugar substitutes

Good dental hygiene and regular dental exams are critical for HPP patients. Quality lifelong dental care should be tailored for each patient based on individual risk assessment.

Asfotase Alfa Treatment

Enzyme replacement therapy (ERT) with asfotase alfa (human recombinant bone-targeted alkaline phosphatase) is now available. It is approved in Canada for infants and children with HPP under 18 years of age who meet accepted criteria for ERT treatment.

Soft Bones Canada - Making a Difference

Given the rarity of HPP and the non-specific nature of many of its symptoms, physicians and dentists may not have seen patients with HPP in their practices or considered this diagnosis. This speaks to the critical role that patients and their families play in working together in partnership with their health care providers.

There is a need to bolster education and training on HPP in the medical and dental communities and to encourage health care providers to partner with patients in their treatment and care. Patients also need more resources and support. Soft Bones Canada is here to encourage those partnerships and support education about HPP!

Soft Bones Canada supports research, education, awareness, policy, and patient advocacy. Our Scientific Advisory Board is dedicated to seeing that the best care is available to patients across Canada. Our most important role is to provide resources and supports to patient and their caregivers.

How Can You Help?

Volunteer: We need volunteers to help Soft Bones Canada fulfill its mission.

Donate: Your donations help us support patients and families, distribute educational resources, and fund research. Donations can be made through our website. www.softbonescanada.ca

Awareness: Spread the word and tell people what you know about hypophosphatasia or direct them to our resources to access more information.

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**Hypophosphatasia:
A Dental Disease**

Hypophosphatasia (HPP)

Hypophosphatasia (hypo-fahs-fuh-TAY-zhah) is an inherited disorder that can vary in severity both between families and within families. People with HPP have low levels of an enzyme simply known as alkaline phosphatase, which is caused by a variant (also known as a mutation) in the ALPL gene. Low alkaline phosphatase levels cause decreased mineralization, specifically calcification, in bones and teeth. Without normal mineralization, bones become weak, soft, and deformable, resulting in skeletal deformities and increased risk of fracture.

Teeth are also affected. Children with HPP may experience early tooth loss attributable to decreased mineralization. These lost teeth often have the roots intact. In adults and in children, the crowns of teeth may also have defects that increase the risk of tooth decay and can contribute to pain.

Comprehensive dental care is recommended for all HPP patients. For the majority, ongoing dental treatments will be essential in treating and managing dental issues arising from HPP.

Towards a Diagnosis

Diagnosis of HPP is established by a medical professional who will take medical/dental history, radiographic assessments, biochemical indicators, and other systemic findings into consideration. Low serum alkaline phosphatase is a hallmark indicator of HPP, and is considered together with many other criteria. For most patients in Canada, formal diagnosis of HPP is made by a geneticist or metabolic specialist, who will oversee treatment and symptom management.

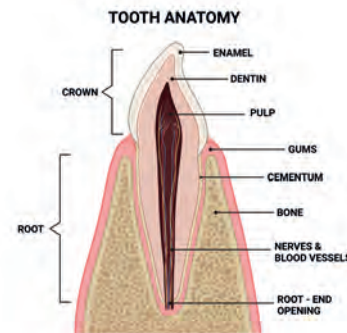
Dentists often play a critical role in the early detection of HPP. This is especially true for patients with milder forms of the condition, which can also be more challenging to diagnose.

If you think you or a family member may have symptoms of HPP, work with your dentist and family doctor to gather the needed medical information and to locate a specialist experienced with HPP.

Soft Bones Canada can assist in connecting patients with resources and specialists knowledgeable about HPP. Contact us for more information.

HPP: Common Dental Symptoms

- Premature loss of primary teeth, sometimes as early as 12-18 months of age and usually before age 5; early loss of baby teeth is nontraumatic and typically the full root is attached
- Limited cementum formation and irregular dentin formation, enlarged dental pulp areas, enamel defects including missing enamel or pits
- Increased risk of tooth decay and cavities: treating small cavities is important
- Unattractive defects may be treated cosmetically with resin fillings or other prosthodontic procedures
- Delayed eruption of permanent teeth
- Possible effects on the permanent dentition including possible loss of permanent teeth



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There is a wide range of HPP dental symptoms, which have sometimes been classified according to age of symptom onset. HPP patients may have some or all of these symptoms, and these could present at various ages. Patients with HPP dental symptoms but lacking skeletal manifestations might be classified as odontohypophosphatasia (the purely dental form of the disease). Common dental symptoms are summarized in the chart below.

Summary of HPP Dental Symptoms
Early loss of primary dentition (anterior teeth typically most affected)
Deciduous tooth loss is spontaneous and roots often fully attached
Reduced dentin thickness
Failure of cementum formation on roots
Enamel abnormalities including pits or missing enamel
Abnormal dentition
Delayed eruption of permanent teeth
Severe caries
Enlarged pulp chambers
Adult tooth loss

Dental Radiographs

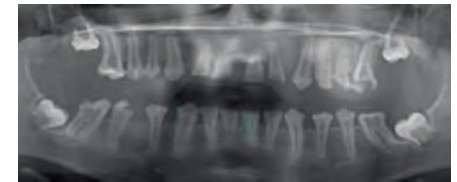
How often should dental X-rays be taken?

Regular radiographs are recommended based on a child's risk for cavities and following suggested management protocols. If low risk, approximately every 12-24 months. For moderate or high risk patients, radiographs are recommended approximately every 6-12 months.

What do teeth with HPP look like on X-rays?

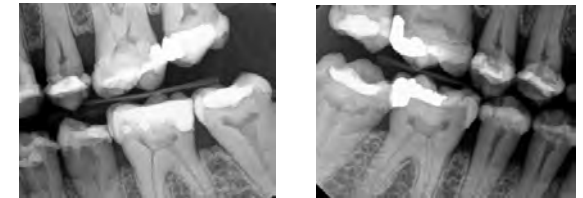
Enlarged pulp chambers (where nerves and blood vessels are located), less bone height, and bony defects may appear with mobile primary front teeth.

Dental Features in Children with HPP



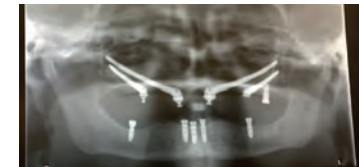
Dental radiograph of a child with HPP

Dental Features in Adults with HPP



Irregular enamel formation: "muffin top" and different pulp chamber anatomy. Bright white regions in the crowns are cavity fillings.

Dental Needs of Adults: Implants



Many adults with HPP require complex dental rehabilitative care, which may involve dental implants to replace missing permanent teeth. Current evidence suggests that the success of implants integrating into bone depends on the quality of the bone. Adults with or without skeletal manifestations may be appropriate candidates for implants. Ask your dental provider.