

XLH and Dental Care:

Expert input on 26 essential questions

This document is intended to be general advice for those with X-linked Hypophosphataemia (XLH) and the dentists looking after their dental care.

Contributors:

Professor Catherine Chaussin and team (France)

Elvire Le Norcy (Orthodontist)

Martin Biosse Duplan (Perio and implantologist)

Violaine Smail Faugeron (Pediatric dentist)

Ole Rasmus Theisen and Gisela Vasconcelos (Norway)

Helena Anjou (Sweden)

Suzanne Sykes (UK)

Hanneke van Verseveld (NL)

Primary Teeth – Daily Care.

1. What is the advice for children regarding brushing the teeth?

Brushing for at least two minutes, twice a day, is recommended to remove bacterial plaque and ensure sufficient fluoride delivery to strengthen the tooth enamel.

Adult supervision is key until the child develops sufficient dexterity and learns not to swallow toothpaste, and an adult should personally brush the child's teeth before or after the child brushes until at least 8 years of age. Emphasis is placed on correct posture and brushing support (e.g., supporting the chin and ensuring visibility), with the adult positioned behind the child in front of a mirror.

Electric toothbrushes can be helpful. However, some children dislike the sensation. A regular toothbrush is also fine if used properly. Use a paediatric toothbrush with soft bristles and a small handle and small head, suited to the child's age.

Interdental cleaning (flossing) is not advised for children. Focus should remain on effective brushing.

2. Is a regular visit to a dental professional advisable?

Yes. Children with XLH should see a dental professional at least twice a year.

3. Is fluoride treatment advisable?

Yes. Fluoride toothpaste should be used from a young age:

- **Before age 6:** 1000 ppm fluoride, the size of a grain of rice up to 3 years, pea-sized age 3-6. Brush as soon as the first tooth appears.
- **After age 6:** 1450 ppm fluoride. Apply the toothpaste to cover one third to one half of the length of the toothbrush's bristled surface. Fluoride varnish should only be applied by a dental professional based on clinical need. Systemic fluoride treatment (pills/supplements) are not advised.

4. Should sealants be placed on primary teeth as a precaution?

Yes. Sealants help protect the pits and fissures of molars and should be applied to primary and permanent molars. Due to enamel microcracks, abnormal dentin mineralization, prominent pulp horns and enlarged pulp chambers in XLH, sealants can help prevent pulp infections, which can occur even without a carious lesion or history of trauma.

Primary Teeth – Treatment

5. Is it safe to extract primary teeth if needed?

Yes, extraction is safe when clinically indicated. However, extractions should be carefully planned to avoid unnecessary early loss and to consider future occlusion and space maintenance.

Antibiotics are indicated in cases where a dental infection is systemically involved. Systemic involvement includes fever, malaise, regional lymphadenopathy, or a diffuse or fascial space swelling beyond the local site of the tooth. Local abscesses are effectively managed with incision and drainage of the abscess followed by appropriate dental management.

6. What is the advice regarding pulp therapy?

A pulpotomy can be considered for primary teeth where the pulp is inflamed, but vital.

A pulpectomy followed by placing a resorbable material in the root canals can be an alternative to extracting a necrotic primary tooth. The criteria to consider in this decision include the presence of physiological or pathological root resorption and the child's level of cooperation.

7. What is the advice regarding crowns ?

Some authors recommend preformed metal crowns (such as stainless steel) as a preventive measure. However, this treatment is invasive and was mainly used before the onset of medical treatments (before 1980) and the dental resin bonding "revolution". That is why a randomized clinical trial is currently underway to evaluate whether sealants could be an effective preventive alternative.

8. What else might we consider for children's teeth?

For a child with temporomandibular disorder (TMD) a soft interocclusal appliance in the lower jaw is necessary to stop any wear to the teeth that could lead to an infection. A soft "mouthguard/night guard/occlusal splint" may be suitable for protection for children without TMD.

Permanent Teeth – Daily Care

9. What is the advice for adolescents and adults regarding brushing the teeth?

Electric toothbrushes can improve plaque removal and are often easier for maintaining good hygiene. Brushing technique is important, as is to clean for at least two minutes twice daily.

Daily brushing with fluoride toothpaste (1450 ppm) is recommended. Professionally applied fluoride varnish up to four times annually can be beneficial, especially in those with a high caries risk.

Fluoride-containing mouthwash with 0.2% sodium fluoride (NaF) after brushing may benefit those with increased risk of caries. Fluoride varnish should only be applied by a dental professional based on clinical need (post eruptive maturation of enamel in adolescents, remineralization of enamel carious lesions).

10. Is flossing advisable?

Yes. Daily interdental cleaning is recommended, as permanent teeth are more tightly spaced. This helps prevent interproximal decay and gum inflammation. Method (floss, toothpicks, interdental brushes) should be based on space between teeth and should focus on cleaning the entire tooth and root surface. A dentist or a dental hygienist can help select the right tool.

11. Are certain precautions to prevent periodontitis advisable (e.g. special toothpaste or mouthwash)?

Yes. XLH patients may be more prone to gum inflammation. Good oral hygiene is essential. Antibacterial mouthwashes (e.g. chlorhexidine) can be considered during inflammation, under professional guidance and for a limited time. Regular check-ups are important for monitoring.

12. Is a regular visit to the dental professional for monitoring and cleaning advisable?

Yes. At least twice a year for a general check-up is recommended. A dental professional can provide preventive care and monitor for early signs of periodontal disease. The frequency of follow-up and the number of visits per year are adjusted to each person's clinical needs, as is the type of care provided. Regular follow-up also helps detect dental infections early

Regarding treatment by dental hygienist. The teeth are more sensitive than normally so the removal of tarts and the cleaning has to be done very carefully.

13. Should sealants be placed on permanent molars as a precaution?

Yes. Fissure sealants are strongly advised for permanent molars to prevent decay and also pulp necrosis due to enamel micro-cracks, altered dentin mineralization and enlarged pulp chambers. Apply as soon as the molars erupt.

We have positive clinical experiences from using ICON/resin infiltration for the entire crown of teeth to reduce hypersensitivity in teeth. We do not yet have data on if this reduces the risk of abscesses or seals infractions.

Permanent Teeth – Treatment

14. Is it safe to extract permanent teeth if needed?

Yes, but only when absolutely necessary. Extraction should be carefully evaluated due to potential bone healing concerns in XLH patients and limited replacement options available. Avoid unless there is no restorative option.

15. Is root canal treatment safe and effective in XLH patients?

Yes, but with caution. Root canal treatment can be successful but may be more complex due to abnormal pulp chambers and dentine. Pain relief and caries/fracture assessment are the priority.

Antibiotics are indicated in cases where a dental infection is systemically involved. Systemic involvement includes fever, malaise, regional lymphadenopathy, or a diffuse or fascial space swelling beyond the local site of the tooth. Local abscesses are effectively managed with incision and drainage of the abscess followed by appropriate dental management.

16. Is it advisable to crown permanent molars and incisors?

Most teeth will not require or benefit from a crown. Crown placement in vital XLH teeth should be avoided because of larger pulp chambers and poorly mineralized dentin.

Endodontically treated permanent molars may benefit from crowns, but this is evaluated on a case-by-case basis.

17. Is pulp therapy advisable in case of exposure of the pulp during tooth preparation?

Yes, but outcomes are variable. Treatment depends on the tooth's maturity in adolescents and, for all the patients, to the pulp status. Specialist advice is recommended due to altered pulp and dentin anatomy.

18. Are white fillings advisable, or do they fail quickly in XLH?

Yes, according to the clinical situation. Resin fillings may fail more often due to adhesion issues with defective dentine. Attention to bonding procedure and potentially using a "micro-etcher"/sand-blaster may be of benefit to enhance dentine bonding in XLH (based on limited studies and clinical experience).

Orthodontic Care

19. Is orthodontic treatment (e.g. braces) safe and advisable for XLH patients?

Yes, but with caution. Orthodontic treatment can be safely performed if the patient is under systemic medical treatment, though XLH-related dental anomalies (like delayed eruption, root abnormalities, or missing teeth) may complicate planning.

Aligners may be preferable due to easier cleaning and reduced caries, however interproximal enamel reduction is prohibited due to the enamel microcracks and dentin hypo-mineralisation. Close collaboration between the orthodontist and the general dentist is essential and the systemic treatment for XLH must be optimized in paediatric and in adult patients.

20. Are there specific precautions or timing considerations for orthodontic care in XLH?

Yes. Delayed eruption and abnormal tooth development may require delaying treatment or adjusting standard approaches: early assessment is useful, but treatment may need to be postponed until dental and skeletal development is more advanced.

In order to allow proper bone remodelling during orthodontic tooth movement, patients should be under systemic treatment for XLH throughout orthodontic treatment and retention. The choice of appliance should also consider minimizing stress on bone and enamel.

21. Should orthodontists consult a specialist familiar with XLH before planning treatment?

Yes. It is strongly advised that orthodontists coordinate with a specialist familiar with XLH. This ensures treatment is tailored to the patient's skeletal and dental profile, and accounts for bone quality and dental abnormalities.

Prosthetic Care

22. Are dentures or other prosthetics safe and effective in XLH patients?

Yes, prosthetic treatments (like dentures, crowns, or bridges) are possible when necessary due to tooth loss or severe damage. Care must be taken with materials due to potentially reduced bonding strength. Careful planning is essential due to the altered bone quality and tooth structure in XLH.

23. Are there challenges with dental implants in XLH patients?

Yes. Dental implants can be more challenging because of low bone density or abnormal bone healing. Implant success is variable, and some patients may not be suitable candidates. If adjacent teeth are heavily restored, a bridge may be appropriate instead of an implant. If considered, thorough assessment and specialist involvement are required. In any case, implant placement should be performed under systemic treatment starting at least 3 months before the surgery and for 6 months after the surgery. Healing time after implant placement and before the placement of the prosthesis should be increased to at least 6 months.

24. Is it better to delay prosthetic treatment until adulthood?

Often, yes. In growing children, prosthetic options like implants are usually delayed until skeletal growth is complete. Temporary solutions (e.g. removable prostheses) can be used in the meantime.

Other / General Recommendations

25. Are regular dental check-ups more important for XLH patients than the general population?

Yes. Due to the higher risk of dental complications, patients with XLH should have regular dental reviews—ideally every 6 months, or more frequently if needed. Ongoing check-ups help monitor for gum inflammation, spontaneous pulp necrosis or infections and carious lesions.

In case of bruxism, a stabilisation splint may benefit patients to protect the teeth at night, as bruxism may lead to larger or more infractions, leading to fractures of the teeth or bacterial infiltration.

26. Are there any special dietary recommendations to protect teeth in XLH?

Yes. A low-sugar, tooth-friendly diet is important, as in the general population. Because of enamel cracks, avoiding acidic and sugary foods is especially crucial. Regular hydration and use of fluoride toothpaste support enamel protection.