



Lesch-Nyhan Syndrome in a Pediatric Patient

Kylie R Bontrager, DDS • LaQuia A Vinson, DDS, MPH

Indiana University School of Dentistry, Indianapolis Indiana – Riley Hospital for Children

INTRODUCTION

Lesch-Nyhan Syndrome is an X-linked recessive genetic disorder most often found in males^[1]. It is characterized as a neurological and behavior condition with an abnormal build up of uric acid levels in the body, often causing arthritis and kidney stones^[1]. Behavioral disturbances experienced by these patients include muscle tensing, jerking, and flailing of the limbs. Self-mutilation, including biting and head banging, is often a result of these disturbances^[1, 2].

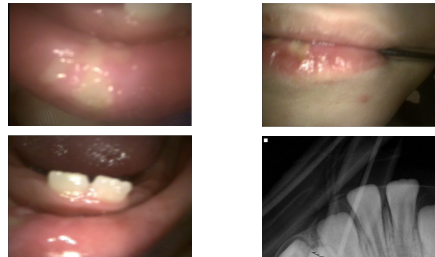
ABSTRACT

Case Report: A 10-year-old male presented to the Riley Children's Hospital dental clinic for evaluation. Parent's chief concern is lip and finger biting. His health history included Lesch-Nyhan syndrome, learning delay and having a g-tube. The patient exhibited self-mutilative behaviors and was wheelchair bound with wrist restraints. This report will include different treatment options performed prior to completely edentulating the patient, as well as the sequencing of extractions both chair side and under general anesthesia, in order to reduce self-harm.

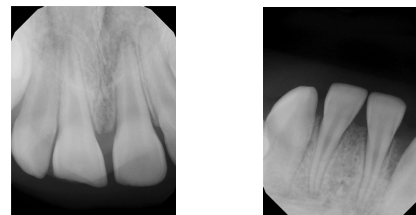
PAST DENTAL MANAGEMENT

A 5 year-old male with only primary dentition previously presented to the clinic with a chief concern of lower lip trauma due to biting and self-mutilation. A mouth guard was fabricated in an attempt to reduce self harm, but ultimately failed. The patient could not tolerate the occlusal mouth guard due to a strong gag reflex, and thus it was determined the best option would be to have multiple primary anterior teeth C, D, E, F, G, H, M, N, O, P, Q, and R extracted while under general anesthesia as this would best manage the effects of the self-mutilative behavior. The primary molars were left in the mouth as there was no history of posterior cheek biting prior to treatment. However, within two months of treatment the patient returned to the clinic with a chief concern of cheek biting with posterior teeth. The patient was then treated again in the operating room to have all remaining primary teeth, including A, B, I, J, K, L, S, and T, extracted under general anesthesia. After eruption of all 6 year molars, the patient returned to clinic nine months later with another chief concern of cheek biting. In an attempt to help reduce self harm, the patient was treated in the OR under general anesthesia a third time to complete the extraction of first permanent molars #3, 14, 19, and 30. Two months after treatment the patient returned to clinic with fully erupted lower anterior central incisors and a chief concern of lower lip biting. In another attempt to help reduce self harm, the patient was treated in an ambulatory setting with nitrous oxide and protective stabilization to have #24 and #25 extracted in the dental clinic.

LOWER LIP BITING



PERI-OPERATIVE



CHEEK BITING



CURRENT MANAGEMENT

A 10-year-old male presented to Riley Children's Hospital dental clinic for evaluation. Parent's chief concern is lip and finger biting. Patient currently has multiple fully erupted permanent anterior teeth. Tooth #8 presented with non-complicated coronal fracture that the parent reported was a result of head banging from self-mutilation. In an attempt to help reduce self harm, the patient was treated in the OR under general anesthesia to have anterior permanent teeth #7, 8, 9, 10, 23, 26, and 27 extracted. Radiographic evaluation completed at the time of OR revealed several permanent anterior and posterior teeth that were close to eruption, but could not be extracted at that time. At the 6 month evaluation, the patient presented to clinic with a chief concern of posterior cheek biting on the left side. The patient had partially erupted #5, 6, 11, 12, 21, and 22. In an attempt to reduce self harm, to allow for further eruption of the current dentition prior to extraction, and to reduce the frequency of having treatment completed in the OR under general anesthesia, enameloplasty of all cusp tips was completed with nitrous oxide in the dental clinic.

DISCUSSION

While there are currently no established methods of practice in regards to the treatment of self-mutilation in patients with congenital insensitivity to pain like those with Lesch-Nyhan Syndrome, it is recommended to attempt nonsurgical options like occlusal splints and mouth guards prior to extracting teeth^[2,3]. These patients often present with facial bruising, abrasions, dental fractures, and intraoral and extraoral traumatic ulcerations that can be concerning to care givers and physicians alike^[2]. Therefore, each patient should be assessed based on their individual clinical findings and when conservative treatment options fail, the benefits of surgical procedures should be considered^[1]. Unfortunately, the hypoxanthine-guanine phosphoribosyltransferase (HPRT) enzyme deficiency that results in the subsequent buildup of uric acid, also results in a spectrum of clinical presentations of developmental delay^[1]. Some patient's may gain an appreciation for avoiding self-mutilating behaviors as they mature while others may not^[2].

REFERENCES

1. Nanagiri, A., & Shabbir, N. (2021). Lesch Nyhan Syndrome. In StatPearls. StatPearls Publishing.
2. Dean, J. A., McDonald, R. E., Jones, J. E., Sanders, B. J., Walker, V. L. Q. A., Yepes, J. F., & Scully, A. C. (2022). Gingivitis and Periodontal Disease. In McDonald and Avery's dentistry for The child and adolescent (pp. 286–323). essay, Elsevier
3. Cusumano, F. J., Penna, K. J., & Panossian, G. (2001). Prevention of self-mutilation in patients with Lesch-Nyhan syndrome: review of literature. ASDC journal of dentistry for children, 68(3), 175–178

