

Removable Appliance for Oral Self-Mutilation in Lesch-Nyhan Syndrome: A Case Report

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Objectives Lesch-Nyhan syndrome is a rare inherited disorder associated with self-harming behaviors, delayed mental and motor development, and disturbances such as dysarthria, choreoathetosis, and spasticity. Severe inflicted behaviors in Lesch-Nyhan syndrome often include chewing and biting of the lips or fingers, which often require management. Techniques to deal with these behaviors include using appliances that restrict the behavior or ultimately extracting the teeth.

Case This case report presents a 6-year-old child with Lesch-Nyhan syndrome and self-mutilation. He had chronic wounds in his buccal mucosa due to severe biting. Two acrylic devices with anterior and posterior bite plates were designed for the maxillary and mandibular teeth. At the follow-up visits, the wounds had been healed, but due to shedding of the primary teeth, and poor patient cooperation, the appliance design was slightly changed. The design of the device used for the patient inspires similar devices for patients with self-mutilation behaviors.

Conclusion Early diagnosis and management of patients suffering from self-injurious behaviors may improve oral health-related quality of life of these patients. Acrylic appliances seem to be effective to minimize injury and to enhance fast healing of oral lesions.

Keywords Lesch-Nyhan Syndrome; Mouth Protectors; Orthodontic Appliances, Removable; Self-Injurious Behavior; Self Mutilation

Introduction

Lesch-Nyhan syndrome is a rare inherited (X-linked recessive) disorder, caused by the deficiency of hypoxanthine-guanine phosphoribosyl transferase, an enzyme involved in purine metabolism. The disorder causes hyperuricemia, hyperuricosuria, and a progressive neurological disorder which results in delayed mental and motor development (dysarthria, choreoathetosis and spasticity). Children affected by the syndrome are usually normal at birth and the disease is not detected until the neurological abnormalities appear at late infancy or early childhood.^{1, 2} However, crystalluria and hematuria may appear in the first months after birth. The prognosis is poor and death usually occurs in the first or second decades of life.¹ One of the characteristic features of the syndrome is the forced insistence on self-mutilation.²⁻⁵

Self-mutilation means inflicting harm deliberately to the body without awareness that it may cause death.^{6, 7} Three types of self-mutilations are described.^{6, 8} The first type, which is the main group, involves loss of function or structure of a body organ which is the primary target in patients with mental disorders or acute intoxication; the eyeball is usually the primary target. The second type is a stereotypic group that includes a recurring pattern of damage to one or more parts of the body. The oral cavity is usually involved, and the damage has varying degrees of severity. These damages have been mostly noticed in patients with hereditary neuropathy or encephalopathy. The third type, which is a mild superficial and chronic type, is

seen in patients with psychiatric disorders, acquired neurological disorders, and patients without comorbidities.² Severe inflicted behaviors in Lesch-Nyhan syndrome are usually chewing or biting lips or fingers, which often require management techniques such as using restrictors or extracting the teeth at an early age.^{4, 5, 9}

In this study, a case of Lesch-Nyhan syndrome treated by a type of restrictive oral appliance is presented.

Case Report

A 6-year-old boy with the chief complaint of persistent chewing of the buccal mucosa causing permanent bleeding and scar was referred to the Orthodontics Department of Shahid Beheshti University of Medical Sciences. This child was diagnosed with Lesch-Nyhan syndrome when he was 3 because of symptoms such as dystonia and delayed developmental stages. At the age of 5, the symptoms of self-mutilation behaviors were detected such as biting of his own hands. Subsequently biting of buccal mucosa appeared one year after trying to limit hand biting. Biting habits were intensified in stressful situations, such as attending school and social gatherings. Psychiatric counseling and medications were not effective.

In order to stop the biting of the buccal mucosa, two restrictive appliances including an acrylic plate with anterior and posterior bite plates, delta Adams clasps on the first and second primary molars and a labial bow were delivered to the patient for upper and lower jaws. These appliances retracted the mucosa and covered the incisal

edges and cusps of teeth (Figure 1).



Figure 1- (a) Lip and (b) buccal mucosa ulcers due to oral self-mutilation in a patient with Lesch-Nyhan Syndrome. (c) Occlusion of the patient. (d) The acrylic removable appliance designed for the patient to prevent self-harming behaviors

At the two-week follow-up, because of poor retention and the patient's attempt to remove the appliance, the lower appliance was temporarily cemented for 2 weeks in place, so the patient could adapt to the device. Following patient adaptation to the device, was used a removable appliance which could be removed before meals. After one month, the buccal mucosa ulcers had been healed, and follow-up visits were scheduled for the patient on a monthly basis to control tooth abrasion and correct any possible interference with eruption of permanent teeth.

After full eruption of first molars and loss of primary first molars, a new device was designed with Adams clamp on the first molars (Figure 2).



Figure 2- Second removable appliance with Adams clasps

At this time, the patient could not remove the upper appliance by hand because of the splint on his hand, but he did so with his tongue. Therefore, the bite marks appeared again and we decided to cement both appliances. In addition, the patient lost the lower appliance two times and despite that making impression from the patient was so challenging we had to make new appliances for him (Figure 3).

Eventually complete recovery was achieved, and the follow up visits still continue (Figure 4).



Figure 3- Final lower appliance delivered to the patient



Figure 4- Complete recovery of lip ulcers following treatment

Discussion

In addition to Lesch-Nyhan syndrome, self-harming behaviors may appear in patients with congenital lack of pain perception, mental retardation, autistic patients, and patients suffering from severe depression or neurological disorders.⁵⁻⁷ The self-harming behaviors in Lesch-Nyhan syndrome differ from self-harming behaviors in patients with different disorders. For example, lip biting can also be seen in Cornelia de Lange syndrome, but it seems to be accidental. In Cornelia de Lange syndrome, the damage is of low intensity and the loss of tissue is rare; also, treatment with aversive techniques that do not work with Lesch-Nyhan syndrome seems to be effective. The pattern of self-harming behaviors in patients with sensory neuropathy such as congenital insensitivity to pain is also accidental. Self-harming behaviors seen in autistic and mentally retarded children are mostly head banging and hitting and rarely biting. Therefore, at the site of trauma, hypertrophy and callus formation are seen, not loss of tissue. Extreme rapid tissue loss is the hallmark of Lesch-Nyhan syndrome. These patients certainly understand the pain and cry when they bite themselves, indicating that this is against their will.³ Therapeutic approaches for these patients depend on the severity of self-harm, including psychological and pharmacological interventions, intraoral devices and dental extractions.⁴ Although the syndrome is characterized by high levels of uric acid, lowering it has no effect on the manifestations of self-harm.³ Thus, the best choice of treatment for such cases would be intraoral devices.

Mouth guards are among the most widely used appliances

in these patients, although they show various results. Other choices are acrylic splints that can be cemented (interfering with hygiene and cause demineralization) or held in place with orthodontic loops. Another commonly used device is the lip bumper, which consists of a long element and a small acrylic shield that prevents biting by moving the lip forward and downward.⁵

Dicks et al.³ used a lip bumper device for a patient with Lesch-Nyhan syndrome that included a labial myofunctional plastic-coated arch soldered to two bands on both sides of the mandibular arch. Oral hygiene quickly became problematic and the device loosened several times and had to be re-cemented with great difficulty. Eventually, the child was able to insert the lower lip between the labial arch and the lower teeth, and the device was completely unsuccessful. However, in another study, Lee et al. concluded that a semi-fixed lip bumper may be useful as an interim appliance to block self-mutilative behaviors in children with Lesch-Nyhan syndrome.⁶

In another study, Lucavechi et al.⁷ fabricated an acrylic device with bilateral shields for an 8-year-old mentally retarded child who had severe bilateral gingival recession due to self-harm. Although the lesions recovered, the patient's self-harming behaviors remained.

Also Lerardo et al.⁸ reported successful application of a soft silicone modified bite block with the internal surface in

contact with the teeth, and the exterior part consisting of a transparent resin shell with front and rear shields to separate the lips and cheeks from the dental arches to prevent bite injuries in a Lesch-Nyhan patient.

The device in our study is somewhat similar to that designed by Yildirim et al.⁹ that was set up for a 7-year-old child with Lesch-Nyhan syndrome, except that their appliance was bulky and covered the vestibular depth; therefore, it was more likely to cause decalcification of teeth. Instead, we used a device that was less bulky and covered less area in the mouth (only teeth to their cemento-enamel junction) to avoid dental caries.

Conclusion

Early diagnosis and management of patients suffering from self-injurious behaviors may improve oral health-related quality of life of these patients. Acrylic appliances, despite some limitations, seem to be a good alternative option to invasive treatments, and a good choice of therapy to limit and prevent self-mutilation behaviors in patients with Lesch-Nyhan syndrome.

Conflict of Interest

No Conflict of Interest Declared ■

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