

## DUAL THICK OCCLUSAL SPLINT: SHIELDER FOR SELF-INJURIOUS ORAL TRAUMA IN LESCH-NYHAN SYNDROME, A CASE REPORT

<sup>1</sup>Shijee M, <sup>2</sup>Ponnudurai Arangannal, <sup>3</sup>Jeevarathan, <sup>4</sup>Janani Vinodhini, <sup>5</sup>Madhumitha, <sup>6</sup>Ramyachellamal, <sup>7</sup>Ishwarya Dhevi G R

<sup>1</sup> Postgraduate, <sup>2,3</sup> MDS, Professor and Head of the Department, <sup>4</sup> Reader, <sup>5,6,7</sup> Senior Lecturer

<sup>1,2,3,4,5,6,7</sup> Department of Pedodontics and Preventive Dentistry, Sree Balaji Dental College and Hospital, BIHER, Pallikaranai, Chennai

DOI: <https://doi.org/10.63001/tbs.2024.v19.i02.S2.pp213-215>

Received on:

16-07-2024

Accepted on:

26-10-2024

### ABSTRACT

Lesch- Nyhan Syndrome (LNS) is an X-linked disorder caused due to mutation in Hypoxanthine guanine Phospho Ribosyl Transferase (HPRT) gene. It is characterized by hyperuricemia, neurobehavioral issues, motor impairment and self-injurious behaviour (SIB). SIB such as lip biting and tongue biting causes deformity of the oral and perioral tissues. A lot of approaches have been tried so far to control this behaviour with variable success rates. This case report presents a 15 year old boy demonstrating the typical LNS with self-biting of lower lip and tongue. An occlusal splint using soft thermoplastic splint material which avoided the forceful contact of teeth with his lower lip and tongue was fabricated and inserted which helped in the healing of the traumatic lesions.

### INTRODUCTION

Lesch Nyhan Syndrome (LNS) got this name since it was first described by Lesch and Nyhan in 1965 who found certain symptoms such as neurobehavioral issues, hyperuricemia, motor impairment and self-injurious behavior in 2 brothers (1,2). The etiology of LNS is mutation of a gene located in the long arm of X chromosome which leads to congenital deficiency of Hypoxanthine guanine Phospho Ribosyl Transferase (HPRT), which in turn interrupts with purine metabolism (2). As a result of this, there occurs increased uric acid production, ultimately leading to the deposition of urate crystals in tissues under skin (subcutaneous tophi, sometimes manifested as ear bumps), joints (gouty arthritis) and kidneys (nephrolithiasis). Other clinical manifestations associated with uric acid accumulation include recurrent urinary tract infections, joint discomfort and oedema. Neurological symptoms include dystonia / hypotonia, dysarthria and failure to meet developmental milestones (1). The most unique symptom is the compulsive self-injurious behaviour (SIB) such as biting fingers, tongue and lips, scratching and hitting one's head against objects (2, 3). The postulated reason for SIB is substantial decline in brain dopamine concentration in basal ganglia. Serotonin, also a neurotransmitter, has been shown to be regulated by dopamine release in presynaptic channels, and depletion of dopamine leading to subsequent depletion of serotonin will result in high-frequency stimulation in Lesch-Nyhan patients (4). Pain sensitivity is evident in these children, but they can't control themselves from this behaviour. Therefore, they are relieved from injury if this habit is controlled.

Treatment approaches for this syndrome are administering drugs such as allopurinol, benzodiazepine, botulinum toxin and dopamine replacement therapy (2). But these medications are not effective in controlling SIB. While learning through the treatment options suggested from the past, we came across

various management strategies like extraction of front or all teeth and intraoral protective appliances such as lip shields, lip bumpers, tongue protectors, occlusal bite plane, occlusal splints, Hawley appliance with lateral acrylic shields (5,6,7,8,9,10,11,12) as well as extraoral protective appliances such as acrylic splints retained by headgear and appliance attached to bubble helmet to interrupt head banging habit as well (13,14)

This report presents the most feasible management of a patient with Lesch Nyhan Syndrome aimed at prevention of soft tissue damage and promoting the healing of already damaged tissues.

#### Case Report:

A 15 year old male patient reported to the Department of Paediatric and Preventive Dentistry with the chief complaint of asymptomatic decayed lower front tooth for the past 3 years. History revealed delayed developmental milestones and was diagnosed with high levels of uric acid at 10 months of age. He underwent physiotherapy which was ineffective. He had been under medication - Tab. Zyloric (powdered form) 100 mg / day from 2 years of age. He developed finger biting and head banging habit at 3 years and was diagnosed with Lesch Nyhan Syndrome. He developed lip biting and tongue biting at 5 years after his sister was born (associated with lack of maternal attention and care). The intensity and frequency of the habit increase when the patient becomes angry and aggressive. No other systemic conditions were diagnosed. The patient had undergone wound debridement in right thigh under GA before 6 years, incision and drainage of right index finger abscess due to finger biting under GA before 3 years.

Clinically, the patient was confined to a wheelchair fitted with seatbelt to keep him upright, hands restrained with wraps. The patient had motor incoordination; abnormal movements were present. He couldn't stand / walk. There was a wound in right index and middle finger due to self-inflicted biting. Extra oral examination revealed traumatized soft tissues from lower lip -

severely deformed. Intraoral examination revealed injury and disfigurement of tongue due to chronic biting [Fig 1].

As the self-inflicted trauma induced deformity of the lower lip seemed to be a very serious problem for this patient, we decided to address that first.

Alginate impression was taken for maxillary arch under physical restraint and cast was poured in dental stone [Fig 2]. Two mouldable sheets of soft thermoplastic splint material, each of 2 mm thickness, were carefully adapted over the cast using pressure moulding device such that the 2 sheets were completely fused together. The excess material was cut off using scalpel blade so that it extends buccally and palatally about 5 mm above the gingival margin. [Fig 3, 4]. The patient was recalled and the appliance was inserted. The fit and stability were checked [Fig 5, 6]. It was advised to wear the splint full time even during the night, except during eating. Initially, the patient was uncomfortable in wearing the splint. After one month, the parent reported that the patient got used it and was wearing it regularly. The parent was very satisfied with the use of splint since the patient was no longer able to bite the lips and tongue. Healing of the traumatic lesions was evident [Fig 7]. Three months later, the parents reported that they missed the splint and the lower lip had been traumatized again. As the cast was available, the splint was re-fabricated and inserted. After 2 months, the parent reported that the patient's habit got completely controlled at daytime and he needs the splint only during night. Two months later, the patient stopped his lip biting habit even without wearing the splint and the lesions got healed [Fig 8]

#### DISCUSSION

Masochistic habits or self-mutilating habits are repetitive acts that result in physical damage to individual which is much difficult to treat (15). Patient's acceptability is of prime importance in the management of such habits. Extraction of some or all teeth, which serve as the sole reason for the traumatic lesions, can of course, cease the habit; but it is an invasive approach and can lead to functional oral disability (3). Literature presents various intraoral appliances which can

resist self-injurious biting and protect oral and perioral tissues. Our idea was to control the habit by means of conservative approach. We decided to fabricate a simple, handy and easily acceptable appliance that would eliminate the upper teeth from coming in contact with the lower lip. Occlusal splint (resembling clear aligners) made up of soft thermoplastic resin are commonly used for treatment of sleep bruxism (16). It has also been effectively used in lip biting habit (17). So, we fabricated an occlusal splint of increased thickness for the maxillary arch, thereby preventing the upper teeth from coming in contact with the lower lip and it also prevents the forceful impact of upper teeth in biting the tongue.

Hanson et al in 1975 (18) stated certain criteria for an ideal intraoral appliance aimed for breaking self-injurious habits: i) it should reflect the tissues that are likely to be damaged from the occlusal table ii) it should not cause any further injury iii) it should allow movement of mandible iv) it should allow oral care v) it should withstand breakage over long period of time vi) it should allow healing of traumatized tissues vii) it should be easily fabricated and installed without any discomfort or risk to the patient. In this present case, the splint satisfied almost all these criteria. Also the patient was not able to easily remove the splint himself. The thickness of normal soft splint is very less which would eventually get worn out fastly when subjected to heavy loads. Therefore, to increase the thickness, 2 sheets were used to make this splint. Complete palatal coverage was avoided in order to avoid discomfort during speech articulation. Since, this is a thermoplastic resin, the chance of breakage and risk of swallowing or aspiration is almost nil. Another major advantage of this appliance is it protects the lips, tongue and cheeks as a whole.

#### CONCLUSION

Thus, in the present case, MAXILLARY DUAL THICKNESS SPLINT is simple, less cumbersome, easily fabricated, easily acceptable, non-risky and effective method of controlling the lip biting and tongue biting habit. Hence, it can be efficiently used in management of such self-injurious habit in syndromic children.



Fig 1



Fig 2



Fig 3



Fig 4

Fig 5



Fig 6



Fig 7

Fig 8

**REFERENCES**

- Nyhan WL, Oliver WJ, Lesch M. A familial disorder of uric acid metabolism and central nervous system function. II. The Journal of Pediatrics. 1965 Aug 1;67(2):257-63.
- Vani C, Andhi N. A Review on Lesch-Nyhan Syndrome: A Rare Inherited Disorder with Hypoxanthine-Guanine Phosphoribosyltransferase Deficiency
- Arhakis A, Topouzelis N, Kotsiomi E, Kotsanos N. Effective treatment of self-injurious oral trauma in Lesch-Nyhan syndrome: a case report. Dental Traumatology. 2010 Dec;26(6):496-500.
- Jathar P, Panse AM, Jathar M, Gawali PN. Lesch-Nyhan syndrome: disorder of self-mutilating behavior. International journal of clinical pediatric dentistry. 2016 Apr;9(2):139.
- Dicks JL. Lesch-Nyhan syndrome: a treatment-planning dilemma. Paediatr Dent. 1982 Jun;4(2):127-130.
- Willette JC. Lip-chewing: another treatment option. Special Care in Dentistry. 1992 Jul;12(4):174-6.
- Nurko C, Errington BD, Ben Taylor W, Henry R. Lip biting in a patient with Chiari type II malformation: case report. Pediatr Dent. 1999 May 1;21(3):209-12.
- Kozai K, Okamoto M, Nagasaka N. New tongue protector to prevent decubital lingual ulcers caused by tongue thrust with myoclonus. ASDC Journal of Dentistry for Children. 1998 Nov 1;65(6):474-7.
- FABIANO JA, THINES TJ, MARGARONE JE. Management of self-inflicted oral trauma: report of case. Special Care in Dentistry. 1984 Sep;4(5):214-5.
- Walker RS, Rogers WA. Modified maxillary occlusal splint for prevention of cheek biting: a clinical report. The Journal of Prosthetic Dentistry. 1992;67(5):581-2.
- Romero Maroto M, Ruiz Duque C, Vincent G, Garcia Recuero I, Romance A. Management of oral lesions in Lesch-Nyham syndrome. Journal of Clinical Pediatric Dentistry. 2014 Apr 1;38(3):247-9.
- Silva DR, da Fonseca MA. Self-injurious behavior as a challenge for the dental practice: a case report. Pediatric Dentistry. 2003 Jan 1;25(1):62-6.
- Chen LR, Liu JF. Successful treatment of self-inflicted oral mutilation using an acrylic splint retained by a head gear. Pediatric dentistry. 1996;18(5):408-10.
- Davila JM, Aslani MB, Wentworth E. Oral appliance attached to a bubble helmet for prevention of self-inflicted injury. ASDC J Dent Child 1996;63:131-4.
- Orian, Carmia. "Self-injurious behavior as a habit and its treatment." American Journal of Clinical Hypnosis 32, no. 2 (1989): 84-89.
- Dube C, Rompre PH, Manzini C, Guitard F, Grandmont P, Lavigne GJ. Quantitative polygraphic controlled study on efficacy and safety of oral splint devices in tooth-grinding subjects. Journal of Dental Research 2004;83(5):398-403.
- Gopalakrishnan S, Chacko T, Jacob J. Management of Lip Biting Using Clear Aligner/Clear Retainer. Journal of Indian Orthodontic Society. 2021 Jan;55(1):94-5.
- Hanson GE, Ogle RG, GIRON L. A tongue stent for prevention of oral trauma in the comatose patient. Critical Care Medicine. 1975 Sep 1;3(5):200-3.