



Lip biting in a patient with Chiari type II malformation: case report

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Abstract

Self mutilation of lips and tongue is considered a common type of Self-Injurious Behavior (SIB). Treatment of SIB in the form of Lip-Biting in developmentally disabled individuals has been the focus of several related reports using different oral appliances preventing or inhibiting the SIB. In this paper we report a case of SIB in the form of Lip-Biting on an infant with Chiari Type II Malformation which was treated with a Lip-bumper. The Lip-bumper demonstrated to be a viable option in treating transient and acute episodes of SIB involving the lower lip and buccal mucosa. (Pediatr Dent 21:209-212, 1999)

Self-injurious behavior (SIB) is defined as deliberate harm to one's own body without suicidal intent.¹ The three most common types of SIB are head banging against objects; self biting of hands, arms, lips and tongue; and head hitting with a fist or palm.² The prevalence of SIB in the general population is not well established, but is estimated to be about 750 in 100,000.³ SIB in the form of lip biting is a condition which appears sometimes in normal children following

a block injection with local anesthetics.⁴ Lip biting has been associated with genetic syndromes such as Lesch-Nyhan⁵⁻⁸ and congenital sensory neuropathy.⁹ Lip biting has also been reported in patients with mental retardation,¹⁰ cerebral palsy,¹¹ seizure disorders,¹² infectious disease such as encephalitis,¹³ autism⁴, and among patients in a comatose state¹⁴—which Gayton has referred to as neuropathologic chewing.¹⁵ Neurodevelopmental impairment has been associated as a predisposing factor for the development of SIB.²

The Chiari malformation (CM) described also as the Arnold-Chiari malformation, is a spectrum of hindbrain hernias from structures located in the posterior cranial fossa through the foramen magnum.¹⁶ The CM has been categorized into four types according to the abnormal position of the cerebellum in relation to the foramen magnum.¹⁶ Chiari type I describes caudally displaced cerebellar tonsils with minor degrees of elongation of the fourth ventricle and medulla oblongata. The Chiari type II malformation is described as the caudal descent of the cerebellar vermis with caudal displacement of the fourth ventricle and lower brain stem. (Fig 1) Patients with Chiari type II malformations present almost universally at birth with a myelomeningocele.¹⁷ The Chiari type III lesions are extreme displacement of the cerebellum and brain stem into a high cervical pouch. These patients generally have a guarded outlook with regard to their eventual level of functioning. Chiari type IV describes severe cerebellar hypoplasia or aplasia. All type of CM may be associated with syringomyelia (Fig 2), a condition of cystic cavities of cerebro spinal fluid within the spinal cord parenchyma, and/or syringobulbia (Fig 2), a cystic cavity which extends into or arises within the brainstem and may cause cranial nerve dysfunction.¹⁸

The CM is a common cause of progressive neurological deficit in myelodysplastic children.¹⁹ Between 20% and 33% of Chiari type II patients become symptomatic from hindbrain herniation.¹⁶ Of the symptomatic children, nearly one third do not survive beyond infancy, making the symptomatic CM the leading cause of death in treated myelodysplastics in the first two years of life.²⁰ There are two types of clinical presentations, which are age dependent reflecting different pathophysiologies.¹⁶ Symptomatic neonates present with symptoms referable to lower cranial brain and brain stem dysfunction in a dramatic fashion with swallowing and feeding difficulties, apnea, stridor, aspiration arm weakness, and opisthotonos as



Fig 1. Magnetic resonance image demonstrates Chiari type II Malformation. Structures below the arrows are hindbrain hernias located in the posterior cranial fossa through the foramen magnum.

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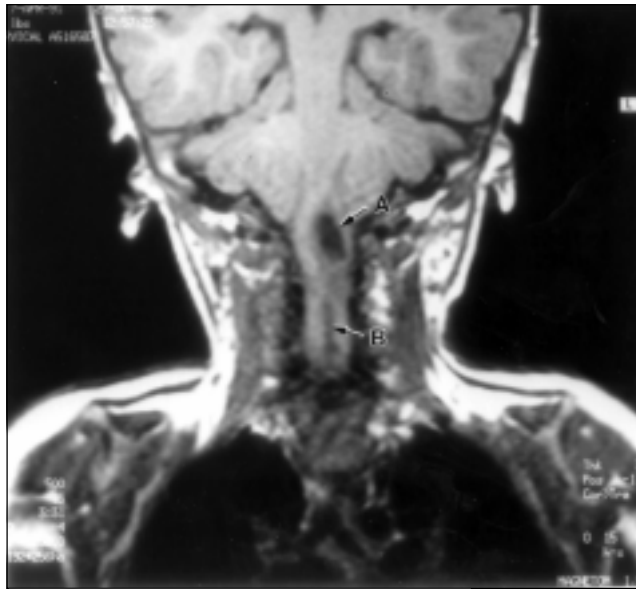


Fig 2. Magnetic resonance image
A=Syringobulbia, and B=Syringomyelia.



Fig 3. Extensive trauma on lower lip as a result of uncontrolled lip biting.



Fig 4. Lip-bumper fabricated with .036 labial arch and acrylic.

the most common symptoms. The presentation of symptoms in older children are slower, less life threatening, and may present as upper extremity weakness and spasticity.

Hydrocephalus develops in about 90% of affected children with CM.²¹ When signs and symptoms of overt hydrocephalus are observed, a ventriculo-peritoneal shunt is required. Over one half of children with shunts will require at least one revision within the first six years of life and 20% will require multiple revisions.²² Most of the patients with hydrocephalus can be expected to have normal or borderline normal intelligence particularly if they are maintained with normal intracranial pressure and intracranial infection is avoided.¹⁹ Respiratory difficulties are common and lethal manifestations of Chiari type II infants, occurring in up to 76% of patients.¹⁶ Feeding and swallowing difficulties are the next most common presenting symptoms and are found in 59% to 71% of the patients.²¹

Magnetic resonance imaging (MRI) is the preferred method to evaluate patients with suspected CM as this imaging provides good detail of hindbrain herniation.¹⁷ (Fig 1 and 2). Early recognition of the CM is essential in the successful management of this condition. Following diagnosis, once symptoms are confirmed, treatment typically consists of shunt revision. If symptoms are persistent, surgical decompression of the CM is performed.¹⁶ Preoperative neurologic impairment, such as central hypoventilation and bilateral vocal cord paralysis, is commonly associated with poor surgical outcome.²³ Older children, adolescents, and young adults respond consistently to surgical decompression of the CM, infants frequently have persistent postoperative symptoms.²³

Treatment of SIB in the form of lip biting in developmentally disabled individuals has been focused in an array of different oral appliances preventing or inhibiting the SIB.³ Appliances range from intraoral removable like a soft mouthguard¹² to fixed lip bumpers soldered onto orthodontic bands.^{4,24} Other appliances include combination of intraoral and extraoral appliances.¹ If these methods are unsuccessful, extractions of the offending teeth and orthognathic surgery to create an open bite have been described.²⁵ In this paper we report a case of SIB in the form of lip biting as a postoperative symptom following decompressive surgery on an infant with Chiari type II malformation with Syringobulbia and Syringomyelia (Fig 1 and 2).

Case Report

A thirty three month nonambulatory infant male was seen as an emergency with the chief complain of lip biting. His medical history revealed a diagnosis of spina bifida with myelomeningocele approximately at L4 level and hydrocephalus at birth. The myelomeningocele was closed and a ventriculoperitoneal shunt (VP) was placed at five days postnatally. The VP shunt was revised six months afterward. Surgical decompression of the Chiari type II malformation was performed at 11, 16, 21, and 28 month of age as well as syringo subarchnoid shunt at 21 and 28 months. At 16 months a tracheostomy was performed soon after paralysis of the left vocal cord was diagnosed. The patient required oxygen during sleep and naps as well as 24 hours nursing supervision. A gastric tube was implanted at 17 months, as a result of feeding difficulties. From the history it was noted that following the last suboccipital craniotomy and cervical laminectomy for decompression

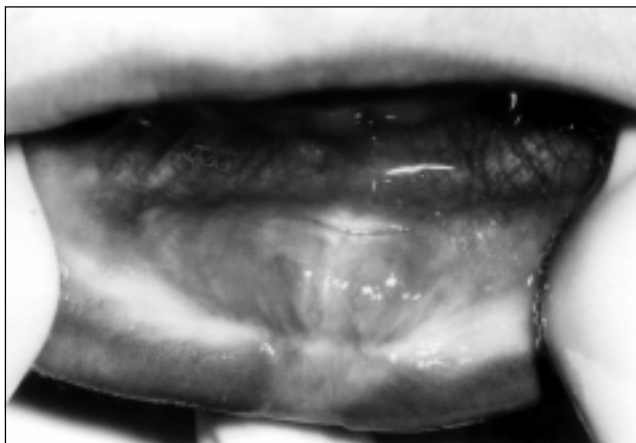


Fig 5. Habit discontinued and the lesion healed.



Fig 6. Habit discontinued and the lesion healed.

of Chiari type II malformation and syringo subarchnoid shunt, the patient woke up quadriplegic and unable to communicate. At this time he began to bite his lower lip. Damage to the trigeminal nerve was diagnosed and accounted for the insensitivity to pain. In the following months, he regained limited movement of his hands and was able to articulate words.

Intraoral examination was performed with antibiotic prophylaxis with Amoxicillin, following the recommendations by the American Heart Association, and latex allergy precautions.

The exam revealed a primary dentition with unerupted second primary molars. The lower lip displayed extensive trauma as the result of uncontrolled lip biting. The lip lesion extended equally from the midline laterally toward the right and left commissures. The lip trauma encompassed a large portion of the labial mucosa and extended to a point just short of the vermilion border (Fig 3). The lesion was firm, and appeared recent with no sign of infection. Minor hemorrhage was present.

Bands were fitted in the lower mandibular first primary molars and an impression was taken with alginate. The patient was not sedated and assisted ventilation was required. A lip bumper appliance was fabricated using an .036 labial arch wire and an acrylic shield (Fig 4). The bands were later cemented and the appliance fitted. The mother was instructed on how to place and remove the appliance. The patient wore the appliance only while awake and under visual supervision. In three months the lip biting habit was discontinued, the lesion healed, and the appliance was removed. (Fig 5 and 6).

Discussion

A lip bumper has been reported as a viable option in treating transient and acute episodes of SIB involving the lower lip and buccal mucosa.^{3,4,11,24} This appliance, when used as a semi-fixed appliance, allows for proper oral hygiene with minimum risk of breakage or swallowing. Additionally, since lip biting may be a cyclic problem, the semi-fixed appliance enables the dentist to reinsert it as needed.²⁴

The prognosis following decompression of symptomatic CM infants has been correlated to the severity of preoperative neurological impairment.²¹ Clinical manifestation of multiple deficits of the sixth, seventh, ninth, tenth, eleventh and twelve cranial nerves have been reported on CM patients,²⁰ and in the fifth cranial nerve in patients with syringomyelia.¹⁶ In this report, the patient had an outcome from surgery with sensory loss from the fifth cranial nerve, which was regained, as well as the preoperative neurological status during the following three months. It is believed that SIB has been reported as a response of an individual to unusually high level of distress.²⁷ In this case, the patient lost his ability to communicate with the external environment which, in conjunction with the sensory loss of his lower lip, is believed to have triggered the SIB. The lip bumper probably was not responsible for eliminating the aberrant behavior, but provided temporary protection for the lip, thus allowing healing to occur. As community placement of handicapped individuals continues to increase, health care providers will be consulted more often to treat and monitor patients who engage in SIB.²

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