

Self-inflicted Oral Trauma in a Baby with Moebius Syndrome

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ABSTRACT

Moebius syndrome (MS) is a rare disorder mainly characterized by bi- or unilateral palsy of the facial and abducens nerves. Among the various orofacial characteristics related to this syndrome, the following are pointed out, difficulty with sucking, tongue malformation, mandibular hypoplasia, saliva drooling, arched palate and micrognathia. In spite of the diversity of oral features described for these patients, self-inflicted oral trauma has not yet been reported. Therefore, the purposes of the present report were to describe an unusual case of a baby with Moebius syndrome who had a parafunctional habit that resulted in a persistent traumatic lesion of the lower lip, and discuss the treatment performed. (J Dent Child 2007;74:224-7)

KEYWORDS: MOEBIUS SYNDROME, SELF MUTILATION, INFANT, MOUTH GUARD

Moebius syndrome (MS) consists of congenital nonprogressive paralysis of the facial (VII) and abducens (VI) cranial nerves which may be complete or partial, unilateral, or bilateral. Other cranial nerves may also be involved.¹⁻³ Because of this paralysis, lack of facial expression is observed in the affected individuals.⁴⁻⁶

As the syndrome is uncommon, the incidence in the general population has not been determined³ and both boys and girls are affected with equal frequency.¹ According to the Moebius Syndrome Foundation, Brazil has the second-largest number of MS patients, with a total of 300 individuals, whereas the United States has 800 MS patients.

Because most of the reports address cases occurring sporadically, the etiology and pathogenesis of MS have been intensively discussed, but not completely defined.^{1-3,7,8} The most commonly accepted hypothesis, however, would be a failure in vascular system development, resulting in transient

fetal ischemia and, consequently, reduced placental blood flow to the fetus.⁵ For some authors, the lack of a clear hereditary pattern in the majority of cases does not rule out genetic causes,⁹ since more than one case of MS in the same family has been reported.¹⁰ Moreover, environmental factors^{3,8,11} influencing pregnancy are thought to be associated with MS, such as infections, hyperthermia, gestational diabetes, use of cocaine or alcohol and some medicines, namely, misoprostol, benzodiazepines, and thalidomide.

MS can usually be clinically diagnosed soon after birth and present with incomplete closure of the eyelids during sleep, saliva drooling and difficulty with sucking.^{1,4,8} In addition, in cases of bilateral palsy, it is observed that affected children are unable to smile or move their facial muscles when crying—a condition known as “mask-like facies”.¹ MS is frequently associated with malformation of the limbs and the orofacial structures and results in significant motor deficiencies.^{9,12} Moderate mental retardation has been reported in 10% to 50% of all cases.^{13,14}

Among the craniofacial manifestations present in these patients, the following conditions may be found: (1) strabismus; (2) ear defects; (3) mandibular hypoplasia; (4) arched palate; (5) cleft palate; (6) tongue malformation; (7) lack of protrusive and lateral mandibular movements; and (8) inability to establish a good perioral seal.^{2,3,7-9} As most patients do not have satisfactory oral hygiene, early

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carious lesions and gingivitis can be observed.³ Absence of teeth in both deciduous and permanent dentition has also been reported.^{8,15}

In spite of the great diversity of oral manifestations, there have, as yet, been no reports of lesions of the soft tissue resulting from parafunctional habits. Therefore, the aims of this report were to describe a pediatric patient with Moebius syndrome who presented with persistent traumatic lesion as a result of a parafunctional habit and describe the treatment performed.

CASE REPORT

The patient, a 14-month-old boy, was brought to the Department of Pediatric Dentistry of the Federal University of Rio de Janeiro, Brazil, because his parents complained of an increasing injury to the baby's lower lip, which appeared after the mandibular teeth erupted. During anamnesis, it was found that the child had been under medical treatment because of convulsive crises since the first week of life. In addition, the child presented with facial palsy, nondetermined inborn error of metabolism and difficulty with sucking, which prevented the child from feeding properly.

Also reported were frequent episodes of pneumonia, deafness, delayed motor development, cerebral palsy, and epilepsy. Medicines taken daily by the patient included, L-carnitina, sodium benzoate, carbamazepine, and Gardenal.

The patient is an only child and was born prematurely by Caesarean section, because the mother had developed hypertension and gestational diabetes during the last 3 months of pregnancy. During the first 4 months of life, the child was kept in the intensive care unit due to convulsions and feeding difficulties.

Upon extraoral examination, convergent strabismus and facial palsy on the right side were observed (Figure 1). Based on the patient's medical history and facial aspect, the dentist suspected the existence of MS, which was later confirmed by a neurologist who diagnosed unilateral palsy of the VI and VII cranial nerves.



Figure 1. Asymmetric facial expression of the patient and convergent strabismus.

Intraorally, an irregularly shaped fibrous lesion was found, located in the inner edge of the bottom lip next to the mandibular central incisors (Figure 2), which were the only existing teeth. These teeth were buccally positioned and had great mobility due to trauma. Because of muscle hypotonicity, both saliva drooling and presence of angular cheilitis were observed. On radiographic exam, the central and lateral mandibular incisors were normally developed and had open apices, with no associated osseous and periapical alterations (Figure 3).



Figure 2. Initial intraoral examination. Note the buccal position of mandibular central incisors.



Figure 3. Radiographic examination showing normal development of mandibular incisors.

Based on the lesion's history, the traumatic ulcer caused by the anterior teeth was thought to be the result of a parafunctional habit. As the teeth had great mobility, the central incisors were extracted under local anesthesia. The patient was co-operative throughout the entire procedure. Two weeks later, the lesion was slightly better.

As the maxillary incisors began erupting, however, the bottom lip lesion's clinical aspect became worse, apparently due to lip-biting (Figure 4). Consequently, after the maxillary teeth were fully erupted, a protective oral appliance was designed for covering all the maxillary teeth, in order to prevent injuries to the bottom lip region. Impressions were made with silicone-based material to obtain a working model and an acrylic resin appliance was made to cover all the maxillary incisors (Figure 5).



Figure 4. Clinical aspect of the lesion after eruption of maxillary incisors.



Figure 5. Oral appliance made of acrylic resin covering maxillary incisors.



Figure 6. Clinical aspect after 2 weeks of cementing.



Figure 7. Clinical aspect after 6 months.

The protective appliance was fixed to the teeth by means of ionomer cement, and after a period of 2 weeks the lesion's clinical aspect improved (Figure 6). The mother reported that the child had accepted the appliance well and that he no longer bit the lower lip. The child has been periodically followed-up in order to remove and clean the appliance and to instruct the mother on oral hygiene and topical fluoride application. After 6 months, evident improvement in the clinical condition could be noted (Figure 7). After this period, the appliance became dislodged once. As the habit persisted, it was recemented in place, and the child's condition improved.

Parallel to the dental treatment, the child was also submitted to specific physiotherapy to develop the facial muscles. A multidisciplinary team consisting of a neurologist, nutritionist, pediatrician, physiotherapist, speech therapist, and psychologist have followed-up with the patient, and this approach will be maintained until the patient reaches adulthood.

DISCUSSION

MS can cause several craniofacial alterations, mainly those involving facial nerve palsy. As facial nerve paralysis is crucial for diagnosing the MS,^{8,12} the facial characteristic observed in the patient were helpful findings for confirming the MS in the present case. Other findings were also useful for diagnosing MS, such as difficulty with sucking, angular cheilitis, deafness, delayed motor development, and strabismus.

As MS patients have both eating and sucking difficulties, their parents generally prepare pasty foods that are often rich in sugar and carbohydrates. Moreover, these feeding difficulties may result in affected children being bottle-fed to a later-than-normal age, and lead to severe carious lesions.^{6,7,15} Furthermore, even the older MS patients have difficulties

with performing oral hygiene adequately because of tongue movement and mastication muscle limitations.⁶ In 2003, Serpa Pinto *et al* observed a high incidence of caries and gingivitis, which had been correlated with both poor oral hygiene and inadequate diet, in all 12 cases described by them.

No carious lesion was observed, however, in the patient described in the present case report. This may be because:

1. The child was brought to the pediatric dentistry clinic at a very early age.
2. Since then, the mother has been instructed about the baby's oral hygiene.
3. The child's diet was restricted to:
 - a. ingesting low protein milk, due to the inborn error of metabolism; and
 - b. the use of a spoon instead of bottle-feeding, because of the sucking difficulty.

Probably the higher propensity for caries was controlled by all the above-mentioned factors. As the fluoride status of the family's community water supply could not be assessed, the choice was to provide topical fluoride varnish applications to all teeth.

Self-inflicted oral trauma is a characteristic that has not been related to MS patients, although such behavior was described in patients with Lesch-Nyhan syndrome, Tourette's syndrome, and infectious diseases such as encephalitis.¹⁶ In the present report, the patient had the parafunctional habit of biting his lower lip, thus traumatizing the region and causing the lesion previously described. The pain resulting from this type of traumatic lesion can impede proper feeding and cause nutritional problems in the child, something not observed in the present case. This lack of painful sensitivity in the lower lip region can be explained, because MS patients may occasionally have a partial defect of the sensory root of the trigeminal nerve, thus leading to loss of sensitivity in the lips, lower jaw, forehead, and corneas.⁹

Self-inflicted oral injuries can be treated in different ways,¹⁶ including wearing down the teeth¹⁷, restoring the teeth, extracting the teeth¹⁸, or wearing protective appliances.^{16,17} The first choice was to extract the mandibular central incisors, because of their great mobility due to trauma. This procedure was not sufficiently effective, however, because the lesion increased as other teeth erupted. Therefore, it was decided to make a fixed acrylic appliance for protecting the lower lip, and this was facilitated because the child was co-operative enough to allow an adequate impression to be made and the appliance to be cemented. The family also cooperated, by attending all scheduled appointments and keeping themselves motivated during the child's treatment. Before choosing this treatment option, although the appliance would be cemented, the authors discussed the risk of aspiration with both parents and the child's physicians. Instructions were given to the parents regarding cleaning and verifying the appliance stability. The authors chose a

yellow-colored appliance so that it would be easier for the family to see it.

The lesion showed significant improvement after the acrylic appliance was placed on the teeth, indicating the treatment's success. Biopsy was not necessary, because the traumatic lesion improved as the etiologic factor was eliminated. The patient has been periodically followed-up to evaluate the other teeth as they erupt. In the future, the acrylic appliance can either be extended to other teeth or tooth extraction can be considered. If necessary, another treatment option to be considered is adhesive restoration of the incisal surface of the tooth involved. As the patient gets older, it is expected that the habit will disappear, although, as stated by his neurologist, his future neurological condition cannot be predicted at present because he is too young.

Therefore, it is crucial to diagnose this type of lesion and the parafunctional habit as early as possible so that the treatment can be less invasive and oral protective devices can be successfully used. Similarly, given the diversity of oral findings, it is clear that MS patients should be monitored by the dental team. Furthermore, by knowing the common characteristics of these patients, the dentist will be able to recognize the limitations involved and establish more adequate treatment procedures.

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