Orthodontic Management of a Patient with Moebius Syndrome

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INTRODUCTION

Moebius syndrome (MBS) is a congenital, non-progressive, neurological disorder that is characterized by weakness or paralysis of multiple cranial nerves, with the sixth and seventh cranial nerves being the most commonly affected.[1]

Clinical presentation depends on which nerve is affected and the extent of paralysis. There could be simultaneous involvement of other cranial nerves, limb malformations, underdevelopment of the fingers, and orofacial malformations such as cleft lip and palate. Existing studies have found a higher prevalence of Class II (56%) in patients with MBS, with micrognathia and excess maxillary development. This was thought to be due to the lack of lip seal. Other dental implications include: microstomia, mandibular hypoplasia, tongue weakness, open bite, and dental malocclusion.[2] Patients with MBS are also found to have a higher caries experience than patients without the syndrome.[3]

ETIOLOGY AND EPIDEMIOLOGY

- The exact cause of MBS is unknown.[4] Genetic and environmental components are thought to be involved.
- Clinical prevalence is estimated to be 1/250,000 live births.
- There is equal incidence in both sexes.
- Most cases are sporadic, with mutations in the PLXND1 and RE3L genes.[5]
- Two percent of cases are found to be autosomal dominant.

DIAGNOSIS AND MANAGEMENT

- The diagnostic criteria determined at the 2007 biannual Moebius Syndrome Foundation defined MBS as "congenital, uni- or bilateral, non-progressive facial weakness and limited abduction of the eye(s)." [6]
- There is no specific treatment for MBS, rather management is supportive according to symptoms.
- A multidisciplinary team will often include pediatricians, neurologists, plastic surgeons, pediatric dentists, speech pathologists, audiologists, ophthalmologists and other healthcare professionals is recommended.[7]
- Surgery has been performed to correct strabismus, improve limb and jaw deformities, and to improve smile. Physical and speech therapy has been recommended to improve coordination as well as control of eating and speaking abilities.

Figure 1. Pre-treatment records A) Frontal profile B) Lateral profile

CASE REPORT

A 16 year old female patient presented to the orthodontic clinic at the Children's Dental Health Clinic of the Long Beach Memorial Hospital with a chief complaint of 'crooked teeth'. The patient's medical history is significant for Moebius syndrome with complete paralysis of her left and right sixth cranial nerves, left seventh cranial nerve, and a partial paralysis of her right seventh cranial nerve. She does not take any medication and has no known drug allergies.

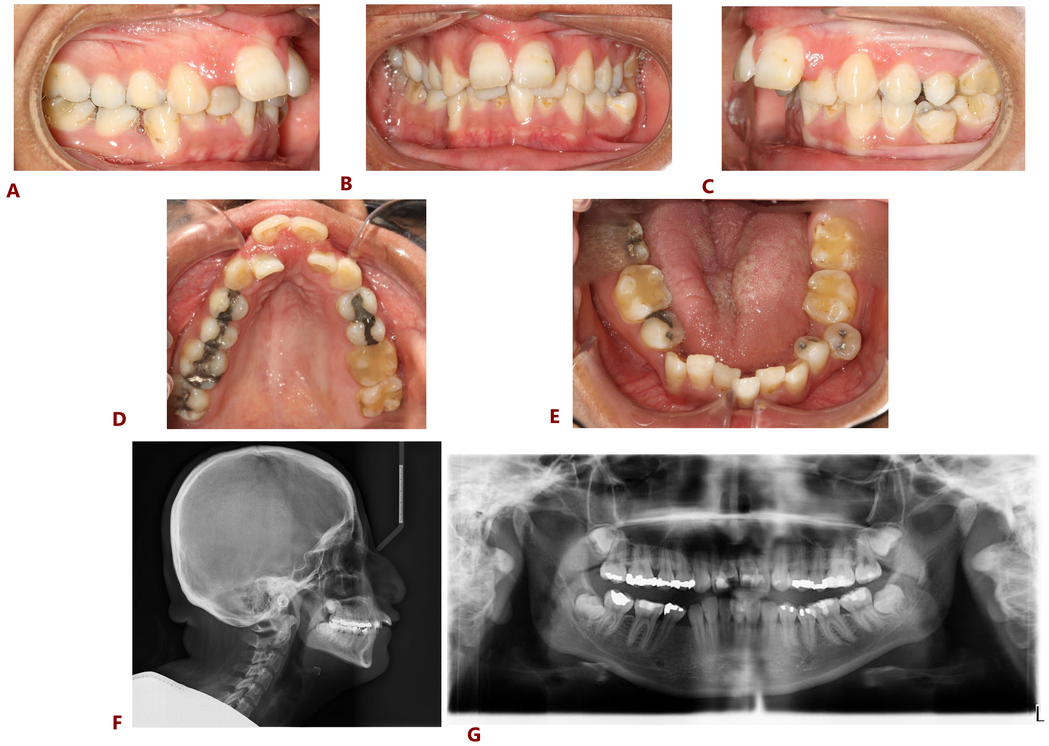

Figure 2. Pre-treatment records A) Right Buccal B)) Intraoral Frontal C) Left Buccal D) Maxillary Occlusal E) Mandibular Occlusal F) Lateral Cephalometric Radiograph G) Panoramic Radiograph

PROBLEM LIST

Skeletal:
- Convex profile and acute nasolabial angle
- Prognathic maxilla
- Retrognathic mandible
- Maxillary Transverse Deficiency

Dental:
- Microstomia
- Lip asymmetry and incompetence
- Class II division 1 malocclusion
- Severe overjet
- Severe Dental Crowding with buccally displaced #20
- History of tooth #28 extraction related to unrestorable caries
- Generalized severe enamel hypoplasia, high caries risk
- Localized mild gingival inflammation around teeth #8 and #9
- Gingival recession #27
- Impacted third molars

ORTHODONTIC TREATMENT

Due to the patient's maxillary transverse deficiency and severe dental crowding commonly seen in Moebius Syndrome, the orthodontic treatment plan consisted of maxillary expansion followed by extractions of the maxillary first bicuspids. The options for maxillary expansion included full removable or fixed appliances.

The first option presented to the patient was to use removable appliances with clear aligners. With the patient's high caries risk from generalized severe enamel hypoplasia, this option would be less damaging to the patient's hypoplastic enamel as it would facilitate maintenance of good oral hygiene.

A second option with a fixed appliance was proposed as an alternative. The fixed brackets would make it more difficult for the patient to maintain good oral hygiene, thereby increasing the caries risk. Our long-term goal for this patient is to improve her dental health, function, and esthetics.

Option #2 was chosen by the family due to financial constraints. More frequent recalls and cleanings will be recommended. Will follow-up with possible periodontal referral if recession continues.

TREATMENT MODIFICATIONS

A patient with Moebius syndrome can present multiple challenges for the dental providers. It is thought that decreased neuromuscular function leads to disturbed oral and maxillofacial development and function. Decreased activity of the tongue and masticatory muscles can lead to increased risk of caries. The lack of lip seal typically contributes to the often observed dry and inflamed gingiva, proclined maxillary incisors and severe overjet.

Dental management of patients with Moebius syndrome should involve education and preventative care, including fluoride treatment and saliva substitutes. Orthodontic treatment will be aimed at reducing the overjet and improving dental esthetics and function. Early orthodontic intervention during the mixed dentition stage can be beneficial. Furthermore, orthognathic jaw surgery may be indicated in severe open bite or severe overjet cases that do not respond to the traditional orthodontic treatment.

CONCLUSION

This case report presents the orthodontic approach to treating a child with Moebius Syndrome and the modifications and considerations that must be made when treating a child with this condition.

REFERENCES

1. Picciolini, O., Porro, M., Cattaneo, E., Castelletti, S., Masera, G., Mosca, F., & Bedeschi, M. F. (2016). Moebius syndrome: clinical features, diagnosis, management and early intervention. Italian journal of pediatrics, 42(1), 56.
2. Chen, B., Li, L. X., & Zhou, L. L. (2021). Dental management of a patient with Moebius syndrome: A case report. World journal of clinical cases, 9(24), 7269–7278.
3. Castro T, Ortega AO, Mussi MC, Braga MM, Gallottini M. Caries Experience in Individuals with Moebius Syndrome. Pediatr Dent. 2016 Jan-Feb;38(1):68-71.
4. Mulliken JB. Mobius syndrome. NORD Guide to Rare Disorders. Lippincott Williams & Wilkins. Philadelphia, PA. 2003:223-224
5. Tomas-Roca, L., Tsaalbi-Shtylik, A., Jansen, J. et al. De novo mutations in PLXND1 and REV3L cause Möbius syndrome. Nat Commun 6, 7199 (2015).
6. Miller G. The mystery of the missing smile. Science. 2007;316:826–7.
7. Magnifico M, Cassi D, Gandolfini M, Toffoli A, Zecca PA, Di Blasio A. Orthodontics and Moebius syndrome: an observational study. Minerva Stomatol. 2018 Aug;67(4):165-171.