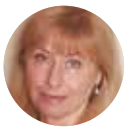
Moebius syndrome: The challenge of dental management



D. Cudzilo*,
T. Matthews-Brzozowska*,**

*Department of Maxillofacial Orthopaedics and Orthodontics, Institute of Mother and Child, Warsaw, Poland
**Department and Clinic of Maxillofacial Orthopaedics and Orthodontics of Poznan University of Medical Sciences

e-mail: dcudzilo@gmail.com

DOI 10.23804/ejpd.2019.20.02.12

Abstract

Background Moebius syndrome (MBS) is a rare disorder which involves unilateral or bilateral paralysis or lack of the VI and VII cranial nerves. This is accompanied by abnormalities affecting the head, mouth, upper and lower limbs, and chest. A distinguishing feature of this syndrome is the so-called mask-like face. A small number of publications relating to occlusal and dental abnormalities and documenting the progress of orthodontic treatment has led the authors to present the case of a patient with Moebius syndrome.

Casew report Aa patient with Moebius syndrome was initially treated by means of removable appliances and later by means of a fixed appliance after permanent tooth extractions. As a result of the treatment, there was an improvement in the ratio of the posterior facial height to the anterior facial height: Sgo/NGn increased from 55.8% to 59.1%, which is closer to the Caucasian population norm.

Conclusions The aesthetic result was satisfactory both for the patient and for her parents.

KEY WORDS Craniofacial anomalies, Dental management, Moebius syndrome, Occlusal and dental disorders.

Introduction

Moebius syndrome (MBS;OMIM 157900) is an extremely rare disease, its incidence irrespective of gender is estimated at 1:50,000 and 1:500,000 [Linsay et al., 2010; Briegiel, 2012;Morales-Chavez et al., 2013]. Originally the condition was described by von Graefe in 1880, later it was more broadly defined by Moebius in 1888 and 1892 and since then it has been known as Moebius syndrome. It involves unilateral or bilateral paralysis or lack of the VI and VII cranial nerves with concurrent abnormalities affecting the head, mouth, upper and lower limbs, and chest. A distinguishing feature of this syndrome is the so-called mask-like face, which involves abnormalities in or a complete absence of facial expression with accompanying cranio-facial abnormalities [Sensat, 2003; De Magalhaes et al., 2006; Escoda-Francoli et al., 2009]. The aetiology of Moebius syndrome has multiple factors connected with genetics or environment, and is not well defined [Cai et al., 2012; Cudzilo et al., 2012; Morales-Chavez et al., 2013]. A literature review conducted on the basis of publications obtained from PubMed indicates that there are not many publications which extensively discuss the issues of dental and occlusal abnormalities or document the progress of orthodontic treatment undertaken in these subjects.

This paper presents the case of a patient who was diagnosed with Moebius syndrome at the age of 6. The patient displayed the majority of the typical clinical symptoms that characterise this disorder [Cudzilo, 2012]. The orthodontic treatment was initially conducted using removable appliances and later, after permanent teeth had erupted, it was continued using fixed appliances.

Case report

At the age of 9 the patient was referred by her family doctor for her first appointment with an orthodontist. A clinical examination revealed a preserved symmetry between the left and right sides of the face, a flat forehead, elongated lower third part of the face, short and hypoplastic upper lip, full exposure of central mandibular incisors and no lip closure. The face was typical for Moebius syndrome in that there was

FIG.1 Laterals and frontal intraoral view before orthodontic treatment.

a complete lack of facial expression, and an inability to smile or cry. An intraoral examination revealed the presence of mixed dentition, tooth crowding in the maxilla and mandible (Fig. 1), Angle's Class II bilaterally, and hypoplastic enamel in permanent mandibular incisors. Additionally there were numerous tooth fillings as well as hyperemic periodontium and mucous membrane. The orthopantomogram revealed the existence of all permanent tooth buds (Fig. 2).

In the first stage of orthodontic treatment functional therapy was undertaken using removable appliances: upper and lower Schwartz plates. Although the patient was fully cooperative, little improvement was achieved. Because of a dento-alveolar discrepancy as well as significant damage to the crowns of first molars on the left side, a decision was made to extract teeth 26 and 36 for general dental health reasons, as well as teeth 14 and 44 on the right side for orthodontic reasons.

The patient returned for further treatment after 3 years; the break in the visits was due to the tooth extractions being performed at long intervals, but also for family reasons. A clinical examination was performed, diagnostic models were prepared, and photographic and radiological documentation was produced, including a panoramic x-ray and a lateral cephalometric radiograph (Fig. 3). The orthodontic treatment plan was revised: extraction of tooth 24 was recommended. The aim of the treatment was to reduce the inclination of the incisors in the maxilla, obtaining Angle's Class I on the right side and Class II on the left. Due to an increased risk of caries, a segmented fixed appliance was initially fitted on the left side in order to achieve tooth 23 distalisation; after 8 months the remaining elements of the upper appliance were added and then the lower appliance was fitted. After 7 months of treatment with the fixed appliance, improvement was noted in the upper and lower arches in terms of tooth alignment as well as overbite. When the fixed appliance treatment was completed a radiological examination was conducted including a panoramic x-ray and a lateral cephalometric radiograph (Fig. 4), and a cephalometric analysis was performed. The panoramic x-ray revealed a distal cavity in tooth 47 and the patient was referred for conservative treatment.

Producing an intraoral photo which would show the occlusal relationships in the lateral sections was impossible in the early diagnostic stages due to microstomia (Fig. 1). A complete photographic documentation of the patient, including lateral intraoral projections with teeth in central relation, was only possible at the age of 13, after the fixed appliance was removed and the treatment was completed (Fig. 5).

FIG. 2 Panoramic radiograph showing the existence of all permanent teeth.

FIG. 3 Lateral cephalometric radiograph before orthodontic treatment.

FIG. 4 Lateral cephalometric radiograph after orthodontic treatment with fixed appliances.

Results

A comparison of the results of cephalometric analysis before treatment with a fixed appliance and after the appliance has been removed shows that the values of the SNA, SNB and SNPg angles increased, which may indicate an anterior growth of the maxilla and mandible in relation to the cranial base. The values of the +1/SN and -1/ML angles, which describe the position of the incisors in relation to the cranial base and the mandibular base, decreased, which produced a more vertical position of the incisors in relation to each other. This is also confirmed by an increase in the interincisal (+1/-1) angle from 110° to 122°. The ratio between the posterior facial height and the anterior facial height increased and at the end of the treatment and reached a value close to the norm. The value of the angle describing the position of the mandible in relation to the cranial base did not change (Table 1).

FIG. 5 Patient`s lateral and frontal photos with the intraoral occlusal view after treatment with fixed appliance.

Discussion

It is believed that Moebius syndrome is not gender-specific, occurring in both males and females [Briegiel, 2012; Linsay et al., 2010; Sensat, 2003]. Nevertheless, an analysis of some documented cases indicates that it is more frequent in boys. Stromlad described 25 patients with MS, two-thirds of whom were male; three-quarters of the cases described by Pinto were males; similarly, over three-quarters of Sjogren's cases were male patients; in Cai's study 2 out of the 3 cases described were boys; and in Bate's research again two-thirds of the cases were males [Sjogreen et al., 2001; Stromland et al., 2002; De Serpa Pinto et al., 2002; Cai et al., 2012; Bate et al., 2013]. This observation suggests that this issue ought to be more closely examined.

Craniofacial anomalies are prominent symptoms of Moebius syndrome. They include different types of malocclusion, flat forehead, flat cheeks, hypotonic mimetic and lip muscles, dental enamel hypoplasia, tongue atrophy or hypertrophy, and many others. As regards dental and occlusal disorders, they may include open bite or deep overbite, maxillary hypoplasia, maxillary narrowing, high arched palate, mandibular hyperplasia or features indicating mandibular hypoplasia. The main masticatory defects in patients with Moebius syndrome are associated with micrognathia. In the case reported the patient had deep overbite, but in the literature there are also reports of skeletal open bite, where combined orthodontic and surgical treatment was implemented to improve occlusion [Cai et al., 2011; Guijarro-Martínez and Hernández-Alfaro 2012]. There is a scarcity of publications discussing the difficulty

Parametr	Before treatment - T0	After treatment – T1	Difference T1-T0	Caucasian population norms	SD
SNA	84.7°	86.2°	1.5°	82°	3.5°
SNB	77.8°	78.5°	0.7°	80°	3.5°
SNPg	75.5°	76.9°	1.4°	81°	3.5°
ANB	6.85°	8.1°	1.25°	2°	3.0°
SN/ML	45.6°	45.8°	0.3°	33°	6.0°
+1/SN	110.8°	106.9°	-3.9°	104°	6.5°
-1/ML	93.3°	85.9°	-7.4°	94°	7.0°
+1/-1	110.4°	122°	11.6°	127°	8.5°
SGo/NGn	55.8%	59.1%	3.3%	60.5%	2.5°

TABLE 1 Cephalometric parameters according to Steiner`s analysis before orthodontic treatment and after treatment with the use of fixed appliance.

in dealing with craniofacial anomalies and presenting the results of treatment for Moebius syndrome patients. Despite some promising titles, an orthodontist is unlikely to find in the reports a great deal of information or practical guidance regarding treatment plans. There are hardly any descriptions of initial examinations, observations recorded during the course of treatment or presentations of final treatment outcomes [Rizos et al., 1998]. Few of the publications available in the PubMed database discuss the orthodontic treatment of MBS patients. Optimal orthodontic treatment is connected with the type of disorder; as standard solutions are insufficient in Moebius syndrome, they ought to be modified according to the clinical situation.

The treatment of open bite using appliances with a tongue crib was discussed by Lima et al. [2009]. However, there is no discussion of long-term outcomes. There is a possibility that functional treatment consisting of muscle tension change may not be very effective in the treatment of MBS patients due to the causes of the condition.

Based on our own experience, it must be noted that the treatment of the patient was technically very difficult for the orthodontist. Microstomia made it extremely difficult and time-consuming to take diagnostic impressions and to fit and monitor the fixed appliance in the lateral sections. Excellent interpersonal relations between the patient and the orthodontist were very helpful in this case.

Orthodontic treatment involving jaw expansion may carry the risk of opening the bite. In the case described there was a dental-skeletal discrepancy but the method of tooth extraction made it possible to keep the angle denoting the inclination of the maxilla in relation to the mandible (SN/ML) unchanged. Since none of the publications available from the PubMed database present a detailed account of orthodontic treatment it is impossible to discuss the values of these angles. Moreover, in the case presented here it was observed that, despite the fact that there was no stimulating muscle activity, there was a slight development and growth of the maxilla and mandible, which did not follow the functional matrix hypothesis formulated by Moss [Graber et al. 2012]. However, this issue also requires further observation and analysis.

In the literature it is noted that patients with Moebius syndrome are highly susceptible to dental caries [De Serpa Pinto et al., 2002; Scarpelli et al., 2008]. During the orthodontic treatment of the patient described in this paper, despite very good oral hygiene, a caries appeared in tooth 47. It was invisible during regular check-ups and it was only diagnosed on the basis of an x-ray taken at the end of the orthodontic treatment.

Conclusions

Patients with Moebius syndrome require extensive treatment by a team of specialists in highly specialised centres. Prompt restoration of the functional and aesthetic balance is of the utmost importance. In the case analysed, it was not possible to restore normal function; however, a satisfactory aesthetic result was achieved, which proved highly gratifying for both the patient and her family. A scarcity of publications discussing the difficulties involved in planning and conducting orthodontic treatment in patients with Moebius syndrome has prompted the authors of this paper to explore and present these difficult and complex issues.

References

- Bate S, Cook SJ, Mole J, Cole J: First report of generalized face processing difficulties in Möbius sequence. PLoS One 2013; 8: e62656. doi:10.1371/journal.pone.0062656
- Briegel W. Self-perception of children and adolescents with Möbius sequence. Res Dev Disabil 2012; 33: 54-59.
- Cai M, Shen G, Fang B, Zhu M, Mao LX. Treatment of severe skeletal open bite deformity in patients with Möbius syndrome: a report of 3 cases. J Oral Maxillofac Surg 2012; 70: e389-399.
- Cudzilo D, Obloj B, Obersztyn E, Bocian E, Matthews-Brzozowska T. Moebius syndrome with facial-dental impairments- a rare or rather seldom diagnosed syndrome? Dev Period Med 2012; 4: 273-279
- De Magalhaes MHCG, Araujo LCA, Chiaradia CC, Zamunaro MT, Mantesso A. Early dental management of patients with Mobius syndrome. Oral Diseases 2006; 12: 533-536.
- De Serpa Pinto MVX, De Magalhaes MHCG, Nunes FD. Moebius syndrome with oral involvement. Int J Paediatric Dent 2002; 12: 446-449.
- Escoda-Francoli J, Sanchez-Garcez MA, Gay-Escoda C. Oral implant rehabilitation in a patient with Moebius syndrome. Med Oral Patol Oral Cir Bucal 2009; 14 : e 295-298.
- Vanarsdall RL, Musich DR. Adult interdisciplinary therapy: diagnosis and treatment. In Graber LW, Vanarsdall RL, Vig KWL, editors. Orthodontics, current principles and techniques. Elservier; Mosby 2012; pp 874
- Guijarro-Martínez R, Hernández-Alfaro F. Management of maxillofacial hard and soft tissue discrepancy in Möbius sequence: clinical report and review of the literature. J Craniomaxillofac Surg 2012; 4: 11-16.
- Lima LM, Diniz MB, Santos-Pinto L. Moebius syndrome: Clinical manifestation in a paediatric patient. Pediatr Dent 2009; 31: 289-293.
- Lindsay RW, Hadlock TA, Cheney ML. Upper lip elongation in Mobius syndrome. Otolaryngology Head Neck Surg 2010; 142: 286-287.
- Morales-Chávez M, Ortiz-Rincones MA, Fabiola Suárez-Gorrin F. Surgical techniques for smile restoration in patients with Möbius syndrome. J Clin Exp Den. 2013; 5: e203–e207.
- Rizos M, Negrón RJ, Serman N. Möbius syndrome with dental involvement: a case report and literature review. Cleft Palate Craniofac J 1998; 35: 262-268.
- Scarpelli AC, Vertchenko TB, Resende VL, Castilho LS, Paiva SM, Pordeus IA. Möbius syndrome: a case with oral involvement. Cleft Palate Craniofac J 2008; 45: 319-324.
- Sensat ML. Mobius syndrome: a dental hygiene case study and review of the literature. Int J Dent Hygiene 2003; 1: 62-67.
- Sjogreen L, Andersson-Norinder J, Jacobson K. Development of speech, feeding, eating, and facial expression in Mobius sequence. Int J Pediatr Otorhinolaryngol 2001; 60: 197-204.
- Stromland K, Sjogreen L, Miller M, Gillberg C, Wentz E, Johanson M et al. Mobius sequence – a Swedish multidiscipline study. Eur J Paediatric Neurol 2002; 6: 35-45.