Moebius syndrome

Oral implant rehabilitation in a patient with Moebius syndrome

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Abstract
Introduction: Moebius syndrome is a rare congenital disorder characterized by unilateral or bilateral involvement of the sixth and seventh cranial nerves, resulting in a lack of facial expression and eye movements. These patients suffer a series of oral manifestations that may complicate their dental treatment, such as facial and tongue muscle weakness, uncontrolled salivation secondary to deficient lip sealing, micrognathia, microstomia, bifid uvula, gothic and fissured palate, fissured tongue, and glossoptosis. The underlying etiology remains unclear, though vascular problems during embryogenesis appear to be involved.

Clinical case: We report the case of a woman with Moebius syndrome and total edentulism. Eight years ago she underwent complete oral rehabilitation with the placement of two implants in each dental arch.

Discussion: Moebius syndrome has still an unknown etiology, although it is related to disorders during pregnancy. This kind of patient can be rehabilitated using oral implants.

Key words: Moebius syndrome, congenital disorder, complete oral rehabilitation, implant treatment.

Introduction
Moebius or Möbius syndrome (MS) was described by Moebius in 1888, and was posteriorly published in 1892, after the observation by Von Graefe of the first case of congenital facial diplegia in 1880 (1-6).

MS is an infrequent and non-progressive congenital disorder characterized by uni- or bilateral involvement of cranial nerves VI (external oculomotor nerve) and VII (facial nerve), and resulting in total or partial paralysis with a lack of patient facial expression and eye movements (1-4,7). The condition is often associated with the alteration of other cranial nerves (II, III, V, IX, X and XII) (1,2,6), the limitation of joint movements (congenital multiple arthrogryposis) (5,8), mental retardation (8), Poland syndrome in 15% of the cases (aplasia of the greater temporal muscle) (1,6,8,9) and multiple malformations of the upper and lower limbs in 25-50% of all affected patients - including congenital amputations, syndactyly, polydactyly, symbrachydactyly, clinodactyly, ectrodactyly, partial absence of hands or feet.
and/or varus, equinovarus or club foot (1-6,8-10). More rarely, MS can be associated with infrequent Goldenhar syndrome with or without cardiac vitium, Klippel-Feil anomalies, hypoglossia - hypodactyly anomalies, and/or different unilateral craniofacial defects (9). Patients with MS moreover suffer a number of oral manifestations that greatly complicate dental treatment, such as a gothic and fissure tongue, tongue and facial muscle weakness, a hypoplastic upper lip, uncontrollable salivation secondary to deficient lip sealing, oligodontia, hypoplastic teeth, periodontal disease, carries, micrognathia, microstomia, open bite, bifid uvula, the incapacity to perform excursive mandibular movements (lateral and protrusive), and glossoptosis (2-4). As a result, these patients have problems maintaining adequate oral hygiene, speech and pronunciation. This and the lack of facial expression cause these subjects to be introverted, shy and with learning problems (1,4,5). MS is a rare congenital disorder with epidemiological characteristics that remain to be determined (3,5). Nevertheless, it is estimated that the risk of developing this syndrome may be in the range of 2% in the general population without family antecedents (9). No gender predilection has been observed (5). Although the underlying etiology is not clear, MS appears to be related to vascular problems during embryogenesis, when in the sixth week of pregnancy the primitive subclavian artery fails to provide the required blood flow to the developing structures it irrigates (3,5,7,9,11). Other possible explanations are the different expression of one same autosomal gene (6), parental consanguinity, gestational diabetes, hyperthermia, hypoxia (5,9), and certain toxic agents (drugs) or infections (rubella) (5,8). Misoprostol is a synthetic E1 prostaglandin with activity when administered via the oral route. It was initially used for the treatment of peptic ulcer, and is wrongly used in different developing countries as an ineffective aborting agent - causing generalized vascular disruption in the first trimester of pregnancy, and resulting in multiple congenital malformations (4,8-10). Other pharmacological and toxic agents related to the development of MS are benzodiazepines, misoprostol, thalidomide, methaqualone, oral contraceptives, alcohol and cocaine (1,4,5). This latter substance is closely related to multiple congenital malformations of this type (1,8).

Patients with MS require control by a multidiscipline health care team, and in this context the dental professional obviously must carry out periodic controls to ensure correct prevention and treatment of the frequent buccodental disorders found in these patients - including functional, orthopedic, periodontal, caryogenic and even temporomandibular joint (TMJ) problems (1,3,5,12).

Clinical Case
The present case involved a 49-year-old Caucasian woman with a height of 1.30 meters and a weight of 41 kg, who worked as a monitor for mentally retarded people in a specialized center. In early 1999 she reported to our Orofacial Implantology Service (Dental Clinic of the University of Barcelona, Spain) with complete edentulism of both maxillas. The patient was dissatisfied with her two complete removable dentures, which caused ulcerations and occlusal instability when chewing. She had no known allergies, and as sole toxic habit reported the consumption of a single glass of red wine a day. She had a history of tuberculosis a number of years before (PPD+). The surgical history comprised uterine leiomyoma detected in 1993, with annual control by her gynecologist with a view to future surgical removal. As regards disease antecedents, the patient suffered MS with congenital achondroplasia, a gastroduodenal ulcer diagnosed in 1987, metrorrhagia episodes in the past (1998), frequent ecchymosis, transient vision loss lasting up to 30 minutes prior to migraine-type headaches (one every 2-3 months), and sporadic lower back pain. Her dental history comprised multiple severe bleeding episodes following tooth extraction. The patient explained that at 20 years of age all her permanent teeth were removed due to severe “double-row” crowding that caused intense oral pain. Since then, the patient had always eaten with her two complete removable dentures. At physical examination she was seen to suffer dwarfism, a symmetrical face without facial expression (Fig.1), and malformations of the upper and lower limbs. As to the oral conditions, the patient showed microstomia, total edentulism of both maxillas, a palatal fissure without premaxillary involvement, a narrow palate, acceptable oral hygiene, and severe resorption of both maxillas probably accentuated as a result of 29 years wearing the two complete removable dentures. Following evaluation and planning of the case, we prescribed 2 grams of amoxicillin (Clamoxyl®, Glaxo-SmithKline, Madrid, Spain) one hour before the surgical implantation procedure, which was carried out under local anesthesia (4 carpules of 4% articaine + epinephrine 1:100,000). The usual implant technique was used to place four fixations by means of a an exclusively crestal incision in the upper maxilla and a crestal incision together with a releasing incision on the mandibular midline. After preparing the four implant beds, two Mk II implants (Bränemark System, Nobel Biocare, Gothenburg, Sweden) measuring 11.5 mm in length and 3.75 mm in diameter were placed in the upper arch, together with two implants of the same brand and type measuring 10 mm in length and 3.75 mm in diameter in the lower arch. Following a postoperative course without complications
and a series of control visits, the second implant phase of the four fixations was completed after 6 months. The prosthetic phase began one month later, with final preparation of two overdentures (upper and lower) retained by the two osteointegrated implants in each maxilla. Annual controls were made during the next 8 years (Fig.2 and 3). No incidents were recorded, except in the seventh year when the two retainers of the lower removable dentures failed as a result of the cumulative wear. Once the lower overdenture retention problem was solved, the patient reported the same level of comfort and satisfaction as before.

Discussion
Although the etiology underlying MS is not clear, we feel that the vascular theory, with necrosis and/or aplasia of the nuclei of the affected cranial nerves (1,3,5,7,9,11), is the most plausible explanation in those cases where no toxic agent appears to be involved. However, a number of interesting studies point to a possible familial predisposition characterized by a dominant autosomal hereditary trait in relation to band q12.2 of chromosome 13 (1,3,5,9,11). It would be advisable to explore the possible genetic etiology of this disease more in depth, with a view to adopting preventive measures.

The incidence of MS has not been clearly established (3,11), though it is known that the risk of having offspring with the disease increases with parental consanguinity, and is between 25-30% - with no racial or gender predominance (1,5).

The principal characteristic of MS is facial paralysis and uni- or bilateral involvement of the sixth cranial nerve (1,3,5,9). In our patient, sixth cranial nerve damage caused convergent strabismus and limitation of lateral eye movement. On average, the life expectancy of these patients is the same as that of any healthy individual, provided there is no nerve damage affecting the function of the upper airways (1). Our patient is presently 49 years old, and her health condition is very good, with no breathing problems.

The oral manifestations are multiple and varied, depending on the case (2-4). In our patient the findings were limited to microstomia, micrognathia, weakened facial muscles, macroglossia and fissured tongue.

The correct treatment of MS always requires a multidiscipline approach (1,3,5,12). In this context, the dental professional may be of great help in young patients, in whom life-threatening situations such as asphyxia due to glossoptosis must be avoided (3). In these young developing individuals with respiratory problems, the early application of functional orthopedic devices has been shown to reduce morbidity and mortality in later periods of life (3).

After consulting the scant literature on MS in dentistry, and specifically in reference to oral implantology, the present study appears to be the second report of a case of MS subjected to implant rehabilitation. The first case was published by our group in the year 2001 (1). The interest of the present case is that complete oral rehabilitation was achieved with 4 implants (the first case involved two unit implants in positions 1.4 and 1.5) that afforded important comfort for this adult patient (19 years older than the first patient). Moreover, in this case follow-up was prolonged (versus only 6 months in the first case) - thereby confirming the good results obtained by our group and referred by our patient. The oral implant rehabilitation was planned using only two fixtures for each maxilla, due to the economic limitations of the
patient; we used balls as prosthetic retainers in order to simplify this case, to our opinion, enough special.

In conclusion, dental implant treatment in patients of this kind is clearly indicated provided there are no absolute contraindications due to relevant medical problems, and assuming that the patient demands better chewing function. Patients with MS or their care givers (in cases of mental retardation) must be instructed on the importance of correct oral hygiene, which should be assessed on each regular visit, during the full lifetime of the patient.

References