ABSTRACT

Aim Moebius syndrome is a rare condition characterised by bilateral facial and abducens nerve paralysis. In the present study, it was investigated the effect of the long term facial muscles hypotactivity on temporomandibular joint movements development.

Methods Accordingly with Terzis classification [Terzis, 2003], a wide sample of A type and B type Moebius patients was investigated for mandibular range of movements. Moebius patients were compared with a sample of healthy subjects.

Results Both type A and type B Moebius patients develop a severe articular movement reduction, especially during mouth opening. A highly significant difference was found between Moebius patients and the healthy group. No difference was found between A and B Moebius sub-samples.

Conclusion The authors stress the importance of an early adequate rehabilitation of Moebius patients, in order to avoid the limitation of TMJ movement range.

Keywords Mandibular movement reduction; Moebius syndrome; TMJ dysfunction.

Introduction

Moebius syndrome is a rare, congenital anomaly characterised by bilateral facial and abducens nerve paralysis, often accompanied by limb, orofacial, muscular and ocular malformations. The association of VI and VII cranial nerve pairs paralysis was first described in 1880 and 1882 [Harlan and Von Graefe, 1880; Chrisholm 1882], but only in 1888 the syndrome was clearly identified by the neurologist Paul Julius Möbius [Möbius 1888, 1892].

In Moebius patients, several other cranial nerves as III, V, VII, IX, X, XI and XII pair might be affected by a reduced function. For this reason, if the mimic muscles paralysis and convergent strabismus are the typical symptoms, a wide range of other dysfunctions may be observed in the orofacial area [Gillberg, 1984; Cohen, 1987; Miller, 1989; Kumar, 1990; Carr, 1997; Abramson, 1998]. Sometimes it is possible to identify other eye movements disturbances, as well as problems of face and tongue sensitivity, chewing strength, pharynx motility and sensitivity, swallowing movements.

The pathogenesis of the syndrome is still unclear, most likely vascular accidents occurring between weeks 13 and 17 of pregnancy [Terzis, 2003; Stabile, 1984; Cortez, 1996], but also the genetic assessment of the patient are involved. The facial nerve paralysis makes patients unable to show any emotion as happiness, sadness or anger. In young developing patients, the lack of facial animation and emotional expression may lead to severe psychological introversion and reclusive personality. Sometimes the paralysis is partial and some orofacial functions may be recovered with a timely and adequate therapy. In any case, treatment of this complex condition requires the expert team efforts of paediatrician, neurologist, maxillofacial surgeon, orthodontist, orthopaedic, ophthalmologist, psychologist and speech therapist. In selected cases the so-called “smile surgery”, or gracilis muscle autotransplantation [Hamilton, 1984; Harii, 1976; Terzis, 1989; Zuker, 1989; Terzis, 1995; Zuker, 2000], may dramatically improve the expressive ability of the patient.

The decreased orofacial muscular functions limit also the normal temporomandibular joint function development during growth, with subsequent limitation in the TMJ movement range. In a previous study authors showed the preliminary results of a TMJ functional analysis in Moebius patients [Di Blasio, 2010]. However, due to the rarity of the syndrome, it was necessary to widen the sample in order to confirm the data. The aim of this sample-control study is to investigate the TMJ function in a sample of 50 Moebius patients compared to 100 healthy subjects.

The study confirmed the severe loss of TMJ motility of Moebius patients, especially with regard to mouth opening. In the lateral and protrusive movements the observed limitation was less severe. It also confirmed that no difference in TMJ function was found between complete bilateral and incomplete paralysis cases.

Materials and methods

In 2002, a dedicated centre for Moebius syndrome was created as part of the Head and Neck Department of Parma University, in cooperation with the A.I.S.M.O. (Italian Association for Moebius Syndrome). In a single day, Moebius patients are visited by several specialists:
maxillofacial surgeon, orthodontist, ophthalmologist and speech therapist. The first goal is to confirm the diagnosis and correctly assess the patient based on the Terzis’ classification [Terzis, 2003], which has replaced the previous classification of Abramson [Abramson, 1998], which was less precise. It analyses five districts: cranial nerve (C), lower limb (L), upper limb (U), face (F) and thorax (T) (C.L.U.F.T.). The first district, “C” as cranial nerve, is divided into three groups based on severity of paralysis.

- Group A: bilateral complete paralysis of VI and VII nerve pair (Moebius syndrome).
- Group B: paralysis of VI and VII nerve pair with residual function of some nerve-muscle units in one side of the face (incomplete Moebius syndrome).
- Group C: unilateral facial paralysis (Moebius-like syndrome).

The neurological assessment of the patients about complete or incomplete facial paralysis was carried out by a neurologist. The study included Terzis group A and B Moebius patients. Exclusion criteria were: 1) Terzis group C; 2) acquired VII pair paralysis; C) any other muscular or articular TMJ disorder. In ten years of clinical activity the authors identified 71 cases of Moebius syndrome, 38 males and 33 females, that met the criteria for inclusion in the study. The mean age of the sample group was 15 years and 9 months. Thirty-two patients were classified as Terzis group A, 17 male and 15 females (mean age 15 years and 7 months); 39 patients were classified in Terzis group B, 21 male and 18 females (mean age 16 years and 4 months).

Authors included in the control group healthy subjects with normal motility of mimics muscles, as assessed by neurological examination. Exclusion criteria were the same as in the Moebius group. The sample group was selected from consecutive young patients referred for different reasons to the Odontostomatology Unit of Parma University. One hundred subjects, 58 male and 42 females, met the inclusion criteria for the control group. The mean age of the control group was 15 years and 1 month. The range of TMJ movements in Moebius and control group was measured twice by two different expert operators and the mean measurement was employed for the study. The TMJ mobility was assessed by means of an orthodontic calliper measuring spontaneous, non-forced and pain-free:
- maximum opening,
- maximum protrusive movement (along the occlusal plane),
- maximum right lateral movement (along the occlusal plane),
- maximum left lateral movement (along the occlusal plane).

**Results**

In the Moebius group the mean maximum opening was 38 mm (range 14 to 56 mm) (Table 1). The mean maximum protrusive movement was 7.5 mm (range from 0 to 11.5 mm). The mean maximum lateral movement on the right side was 8.5 mm (range 0 to 12.5 mm) and on the left side was 7.5 mm (range 0 to 12.5 mm).

In the Moebius group A (Table 2) the mean maximum opening was 37 mm (range 15 to 50 mm). The mean maximum protrusive movement was 7 mm (range 5 to 10 mm). The mean maximum lateral movement on the right side was 8.5 mm (range 0 to 11 mm) and on the left side was 7.5 mm (range 0 to 10 mm).

In the Moebius group B (Table 3) the mean maximum opening was 38 mm (range 14 to 56 mm). The mean maximum protrusive movement was 7.5 mm (range 0 to 11.5 mm). The mean maximum lateral movement on the right side was 8.5 mm (range 5 to 12.5 mm) and on the left side was 8 mm (range 4 to 12.5 mm).

In the control group (Table 4) the mean maximum opening was 51 mm (range 37 to 62 mm). The mean maximum protrusive movement was 8.5 mm (range 3 to 14.5 mm). The mean maximum lateral movement on
the right side was 10 mm (range from 6 to 12.5 mm) and on the left side was 9.5 mm (range 4 to 12 mm).

Discussion

A 50 mm. opening and 8-10 mm lateral and protrusive movement are generally considered a normal TMJ functional range. However, the real lower cut-off to recognize a problem in mandibular movement range is not clearly described in the scientific literature [Farrar WB, 1982; Kaplan AS, 1991; Dworkin SF, 1992; Laskin DM, 2006]. For this reason the authors preferred to compare Moebius patients to an healthy group, instead than to a theoretical value of normal limit of movement.

The healthy group showed a wide range of movements, as expected. The total (A and B) Moebius group on the contrary showed a clearly reduced range of movement, especially in mouth opening. This movement drops from 51 mm in the healthy group, to 37 mm in the Moebius patients, with a reduction of over 25% (Fig. 1).

This value, as assessed by the Mann-Whitney statistical test, showed an highly statistically significant difference at p<0.01.

The mandibular lateral movement ability of Moebius patients seemed to be less severely compromised. However also these movements, compared to the healthy group showed a statistically significant difference at the Mann-Whitney test, p<0.01. Mandibular protrusion did not show any statistically significant difference.

In the Moebius patients, the constant limitation of TMJ movement during growth seems to lead to the permanent limitation in the articular movement range. A few Moebius patients showed a severe limitation in mandibular opening due to a coronoid process hypertrophy [Turk A.E. 1999], in all other cases the limitation is probably related to the described general muscular hypoactivity. It is well known that the Moebius syndrome affects mimic muscles activity when muscles related to mandibular opening, protrusive and lateral movements are not directly damaged. However several other cranial nerves are often involved in the syndrome and the complex oral disfunction generates an overall facial hypoactivity, thus reducing TMJ mobility.

**FIG. 1** Healthy vs. total Moebius (A+B) group.

**FIG. 2** Moebius group A vs. group B.
The difference in general dysfunction between complete (A) and incomplete (B) Moebius syndrome is very mild (Fig. 2). In B patients only a few muscle-nerve units in one side of the face are active. The mandibular extreme movements did not show any differences between the two sub-samples. Only the maximum opening is slightly better in the B group. However both parametric (Anova) and non-parametric (post-hoc) tests did not show a statistical significant difference. The authors hypothesise that the mild functional advantage of B patients may be not sufficient to create a more favourable condition for a normal TMJ development.

Conclusion

In Moebius patients non only mimic muscles are compromised but the syndrome creates a general and complex muscular weakness in the whole face. In this unfavourable environment, the TMJ seems unable to develop a wide range of normal movements. In the affected patients the more evident limitation seems to be mouth opening. However also lateral movements, even if less severely compromised, show a statistically significant reduction. The presence of a unilateral, limited preserved function (Moebius “B”) does not warrant a proper TMJ development. These observations suggest the importance of an adequate functional rehabilitation programme, starting from an early age, to improve TMJ movements. Even if in some cases a residual function may be observed, the patients still need to undergo the same rehabilitation programme because also Moebius group B patients may eventually develop the same disorders as group A.

References

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