Hemifacial Microsomia (HM) is a condition of facial asymmetry due to the underdevelopment of the structures derived from the first and the second branchial arches and a unilateral mandibular hypoplasia, with a lower midline deviation on the affected side and a subsequent ipsilateral involvement of the maxilla. As described by Obwegeser [Obwegeser and Makek, 1986], facial asymmetry is more evident when the mandible is affected by a primary HM on a side and by a contralateral hemimandibular elongation, due to a reactive hemimandibular growth by stretching of the structures. HM is a complex congenital malformation involving the skeleton of the temporomandibular area and its soft tissues: some patients suffer from weakness of the facial muscles and have an incomplete facial palsy; in others, the facial nerve involvement on the affected side is more serious. Occasionally, we observe a form of HM with mandibular midline deviation to the opposite side of HM; in these cases, facial nerve involvement, and in particular palsy of the ramus marginalis, has been documented.

With a frequency of 1:5600 born, HM is the most common congenital craniofacial anomaly after clefs of the lip and palate [Grabb, 1965; Poswillo, 1973; Cousley and Wilson, 1992]. From the embryologic damage [Silvestri et al., 1996], different phenotypic grades of dysmorphism may occur, from slight to more complex malformations affecting the mandible unilaterally or bilaterally [Carvalho et al., 1999].

There are no studies to date that clearly document the incidence of ramus marginalis palsy in Hemifacial Microsomia; Takushima et al. [Takushima A et al., 2002] say that, despite the frequent facial nerve and mimetic muscle involvement, few reports described the treatment of this condition. MacQuillian reports that, although paralysis of the complete facial musculature may be present, most often a ramus marginalis mandibulae paralysis is seen [MacQuillian

Ramus marginalis mandibulae nervus facialis palsy in hemifacial microsomia

A. SILVESTRI, G. MARIANI, R.A. VERNUCCI

ABSTRACT. Aim The paralysis of the ramus marginalis mandibulae nervus facialis may occur in Hemifacial Microsomia (HM); the combination of both HM and palsy contributes to an elongation of the mandibular body. This study explores a possible correlation between neurological deficit, muscular atony, and structural deficiency.

Study Design Of 58 patients with HM who had come to the University of Rome (Sapienza) Pre-surgical Orthodontics Unit, 4 patients were afflicted with Hemifacial Microsomia and ramus marginalis mandibulae nervus palsy; these patients underwent physical, neurological, opthamologic and systemic examinations. The results were then analysed in order to determine a possible correlation between neuro-muscular and structural deficit.

Methods Electroneurographic and electromyographic examinations were performed to estimate facial nerve and muscles involvement. Results Neuroelectrographic exam showed a damage of the nervous motor fibres of the facial nerve ipsilateral to HM, with an associated damage of the muscular function, while neuro-muscular functions on the healthy side were normal. Conclusions The peripheral nervous and muscular deficits affect the function of facial soft tissues and the growth of mandibular body with an asymmetry characterised by a hypodevelopment of the ramus (due to the HM) and by an elongation of the mandibular body (due to ramus marginalis mandibulae nerve palsy), so that the chin deviation is contralateral to HM. In these forms, a neurological examination is necessary to assess the neurological damage on the HM side. Neuromuscular deficiency can also contribute to a relapse tendency after a surgical-orthodontic treatment.

KEYWORDS: Hemifacial microsomia; Ramus marginalis palsy; Facial asymmetry.
et al., 2003]. The paralysis of the ramus marginalis is often associated to systemic malformations or craniofacial disorders [Papadatos et al., 1974; Shapiro et al., 1995]. However, precise data concerning the incidence of unilateral ramus marginalis mandibulae nervus facialis involvement with alterations of the mimic expressions in the oral district in patients not affected by HM has not been collected to date; several Authors have attempted to estimate it: Harris [1983] reported 41 cases of paralysis in 18,139 consecutive births, giving an incidence of 0.23%, while Smith [1981] reported a facial paralysis of 0.05%.

In HM cases with ramus marginalis palsy «etiology of the condition remains unclear, but facial palsy associated with hemifacial microsomia is thought to be secondary to abnormal middle ear seventh nerve pathways and fallopian canal atresia. The degree of external ear deformity, rather than other bony or soft-tissue defects, is a predictor of seventh nerve involvement» [MacQuillan et al., 2003].

The ramus marginalis innervates the depressor labii inferioris, mentalis, orbicularis oris, depressor anguli oris muscles [Bernhard, 1997]; for a symmetrical development of the mandibular structure, a symmetrical function of the perioral muscular system is necessary, especially during smiling and lips protrusion. Symmetrical smiling operates like a "sling-like" muscular mechanism in the chin region that represents a brake system of the sagittal mandibular development. When it is affected with palsy, the patient shows an alteration of the mimical mechanics in the inferior facial third, the patients’ smile appears like a grimacing (Fig. 1).

The correlation between functional neuromuscular damage and skeletal alterations has long been recognised. This study tests the hypothesis that, regarding the pathogenetic mechanism of these forms of HM with ramus marginalis palsy, a possible relationship exists among neurological deficit, muscular atony, and structural deficit.

**Materials and methods**

From 1980 to 2007, 58 patients affected by HM were treated at the “Sapienza” University of Rome (Maxillofacial Surgery Dept. and Orthodontics Dept.); in 28 patients, only orthodontics was applied (48.2%); 30 patients were treated with a combined surgical-orthodontic therapy (51.2%).

Among the 58 patients, 4 patients (6.9%) suffered from ipsilateral ramus marginalis mandibulae nervus facialis palsy: three showed a mandibular deviation on the side opposite the one affected with HM as well as palsy, one showed a mandibular deviation on the same side affected with HM (Table 1).

Our study concerned the 4 patients who suffered from ipsilateral ramus marginalis palsy. All patients were subjected to ophthalmologic, neurological and systemic examination. Each patient was submitted to routine evaluations (photos, skull x-ray, cephalometric tracing, dental casts), classified with O.M.E.N.S. method by Vento et al. [1991] and subsequently submitted to a complete electroneurographic (ENOG) and electromyographic (EMG) study to point out facial nerve and muscles involvement. EMG/ENOG evaluations were performed at the Department of Clinical Neurology and Otorhinolaryngology of Rome “La Sapienza” University with a Medelec-Synergy Electromyograph. Electromyographic signals were recorded bilaterally from the orbicularis oculi, orbicularis oris and frontalis muscles, by means of concentric bipolar needle electrodes, at rest and during different levels of isometric contraction (patients seated on a comfortably chair). Motor Unit Potentials (MUP) analysis was performed. The motor nerve conduction in the facial nerve was analysed by means of concentric

<table>
<thead>
<tr>
<th>Case</th>
<th>Age</th>
<th>Sex</th>
<th>HFM Side</th>
<th>Omens Classific.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 M.T.</td>
<td>5 yrs</td>
<td>M</td>
<td>Right</td>
<td>O0 M2a E1 N72 S2</td>
</tr>
<tr>
<td>2 G.P.</td>
<td>10 yrs</td>
<td>M</td>
<td>Left</td>
<td>O21 M2a E2 N72 N31 S1</td>
</tr>
<tr>
<td>3 D.G.</td>
<td>7 yrs</td>
<td>F</td>
<td>Right</td>
<td>O21 M2b E2 N2 S2</td>
</tr>
<tr>
<td>4 A.Z.</td>
<td>9 yrs</td>
<td>M</td>
<td>Left</td>
<td>O0 M1 E0 N2 S3</td>
</tr>
</tbody>
</table>

**Table 1 - Clinical features of the patients involved in the study.**
bipolar needle electrode at the recording side and by bipolar surface electrodes, with interelectrode distance of 2 cm, at the stimulating site. The Compound Muscular Action Potentials (CMAPs) were recorded from orbicularis oculi, orbicularis oris and frontalis muscles while percutaneous electrical stimulation of the extracranial facial nerve was executed just distal to its exit from the stylo-mastoid foramen, where the nerve trunk is still undivided near the earlobe; rectangular electric shocks with a duration of 0.1 ms were used to perform the excitability test and to obtain the CMAPs; latency was measured at the first deflection of the CMAPs; amplitude was calculated peak-to-peak. The analysis time was 20 ms, the intensity was set at 500 mV and the filter band-pass was 3Hz-10kHz. During the study, room temperature was maintained between 22°C and 24°C, and skin temperature between 34°C and 35°C [Kumura, 1989].

Case 1
Patient was seen in 1987 at the age of 7. He showed right HM, classified O0 M2a E1 N72 S2, and characterised by a right ramus marginalis palsy, ipsilateral elongation of the mandibular body with a left mandibular and interincisor line deviation (opposite side of HM). He underwent functional therapy with Asymmetrical Functional Activator (AFA Silvestri), and, three years after first visit, we began fixed orthodontics with an Edgewise appliance; this therapy continued until the patient was 17 [Stricker et al., 1990; Silvestri et al., 1996; Silvestri and Pozzi, 1997]. Surgical repositioning of the skeletal bases was not necessary with this orthodontic treatment. Physical examination revealed facial asymmetry at rest and during voluntary activity, with facial deviation on the left side and no voluntary or spontaneous movement of the lower division of the right facial nerve, while the upper division of the right facial nerve and the left side were normal. No additional acatastasia was identified on ophthalmologic, neurological and systemic examination (Fig. 2).

Case 2
Patient was seen in 1998 at the age of 10. He showed a left HM classified O2↓ M2a E2 N72 N31 S1; a left elongation of the mandibular body with a right chin and interincisor line deviation, on the opposite side of the lesion. Physical examination revealed a complete left facial nerve palsy with no voluntary or spontaneous movement of either the upper and lower divisions of the left facial nerve, with facial asymmetry at rest and facial midline deviation on the right. No additional abnormalities were identified on ophthalmologic, neurological and systemic examination. He underwent fixed orthodontic treatment before surgical repositioning of the upper and lower jaws at the age of 17 (Fig. 3).
Case 3
Patient was seen in 1992 at the age of 7. She showed a right HM classified O2, M2b E2 N2 S2, and a left mandibular and interincisor line deviation caused by an elongation of the right mandibular body (the same side of HM and facial nerve palsy). She started functional therapy using an asymmetrical functional activator (AFA Silvestri) until 11 years old, then fixed orthodontics was performed; at 13 years old she underwent surgical repositioning of the upper and lower jaws. After two years from surgery she showed the tendency to a relapse of the mandibular deviation and facial asymmetry, due to neuromuscular alterations of the facial nerve palsy; for this reason, the patient was submitted to a second operation for chin and right mandibular angle repositioning (Fig. 4).

Case 4
Patient was seen in 1982 at the age of 9. He showed a left HM classified O0 M1 E0 N2 S3, with a left ramus marginalis palsy and left chin deviation. Patient came again to us when he was 15, past the age when functional treatment can be performed. We performed pre-surgical fixed orthodontics, and at the age of 17 he underwent to surgical treatment for mandibular angle remodelling and chin repositioning. Soft tissues graft on the left facial side was done for aesthetical purposes (Fig. 5).

Results
Electroneurographic exam revealed damage to the nervous motor fibres of the ipsilateral facial nerve, with an associated reduction of muscular function, both for trophism and voluntary activity (Table 2). Analysis of the contralateral healthy side demonstrated a functional normal range, except in case 2, where reduced function on both sides was noticed.

In case 1, Electroneurography (ENOG) demonstrated Compound Muscular Action potentials (CMAPs) from the right perioral muscles (orbicularis oris and mentalis) to be about 15% of the amplitude obtained from the clinically normal opposite side, with polyphasic morphology and increased duration. Nerve excitability was reduced on the affected side.

Electromyographic (EMG) examination of the orbicularis oris, depressor anguli oris on the affected side showed a reduced motor unit activity with polyphasic, high amplitude and long duration Motor Unit Potentials (MUPs). At rest there were no denervation potentials.

ENOG and EMG on the left side facial muscles were normal (Fig. 6).

In case 2, ENOG showed low amplitude, high duration and polyphasic morphology of the CMAPs from orbicularis oris, depressor anguli oris and orbicularis oculi on both right and left facial nerves. The EMG revealed a reduced motor unit activity with polyphasic, high amplitude and long duration
Motor Unit Potentials (MUPs) on both sides (Fig. 7).

In case 3, ENOG showed a moderate involvement of both mandibular and zygomatic branch of the right facial nerve, with low amplitude (about 50% compared to the opposite side), normal duration and moderate morphologic abnormalities of the CMAPs. Nerve excitability was reduced on the affected side.

The EMG revealed reduced motor unit activity with polyphasic, high amplitude and long duration Motor Unit Potentials (MUPs) on the left side. At rest there were no denervation potentials.

### Table 2 - Comparison of facial nerve CMAPs and amplitude of the affected and unaffected side.

<table>
<thead>
<tr>
<th>Case</th>
<th>Latency Side</th>
<th>Amplitude Side</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Affected</td>
<td>Normal</td>
</tr>
<tr>
<td>1 M.T.</td>
<td>3.7 ms</td>
<td>3.45 ms</td>
</tr>
<tr>
<td>2 G.P.</td>
<td>3.7 ms</td>
<td>3.45 ms</td>
</tr>
<tr>
<td>3 D.G.</td>
<td>3.05 ms</td>
<td>3.8 ms</td>
</tr>
<tr>
<td>4 A.Z.</td>
<td>2.3 ms</td>
<td>2.75 ms</td>
</tr>
</tbody>
</table>

**Fig. 6** - Case 1. Orbicularis oris CMAPs latency, amplitude and image from affected side compared to unaffected side. Diagrams of the amplitude and CMAP image show unilateral axonal right facial nerve injury on the HM side.

**Fig. 7** - Case 2. Orbicularis oris CMAPs latency, amplitude and image from affected side compared to unaffected side. Diagrams of the amplitude and CMAP image show bilateral axonal facial nerve injury on the HM side.
The ENOG and EMG on the left side facial muscles were normal (Fig. 8).
In case 4, the ENOG revealed an involvement of both mandibular and zygomatic branch of left facial nerve, with low amplitude (about 10% compared to the opposite side). Nerve excitability was reduced on the affected side.
Electromyographic analysis showed a lower recruitment in the mandibular division of the left facial nerve than in the zygomatic division, polyphasic, high amplitude and long duration MUPs without spontaneous activity at rest. The ENOG and EMG on the right side were normal (Fig. 9).

**Discussion**

The treatment of 58 subjects with HM was analysed, of these 4 were clinically evaluated and showed a
deficit of the ramus marginalis of the facial nerve ipsilateral to HM. Upon, physical examination, each of the 4 patients showed a marginalis nerve palsy ipsilateral to HM: three of whom had a mandibular and interincisor line deviation contralateral to HM, one of whom had an ipsilateral deviation.

The neurophysiologic data obtained supported our clinical observations. The neurological deficit, associated with the muscular hypotony of the HM side, caused an asymmetric function of the perioral muscles and the resulting monolateral action of the "sling-like" mechanism in the chin region. All of the above contributed to a skeletal hemimandibular iper-growth, appearing as a hemimandibular elongation of the mandibular body, on the side affected with HM and palsy of the ramus marginalis.

The influence of the muscular system on the morphology of the bones has long been recognized [Moss, 1962]. Regarding the relationship between bone and muscle in HM, Kane [1997] supports the hypothesis that the extent of hypoplasia of the facial muscles predicts the extent of the osseous dysplasia; Vargervik and Miller [1984] also designed a study to establish the extent of neuromuscular abnormalities in patients with HM and related their findings to the skeletal malformations in these patients. Both authors support the idea that the epigenetic regulation of the bone is performed by muscles, according to the functional matrix theory. It is also reported that the asymmetry of neuromuscular functions and the masticatory apparatus can lead to an asymmetry of the mandible and the cranium and facial asymmetry inducing muscle dysfunction too [Captier et al., 2006].

Polley and coworkers [1997] explained that mandibular and facial asymmetry in HM is not progressive in nature and that in these patients the growth of the affected side goes together with that of the unaffected side. On the other hand, Kearns and coworkers [Kearns GJ et al., 2000] observed that facial asymmetry in HM patients is progressive with time. On the basis of our experience in treating HM, we agree with Polley’s theory concerning the non-progressive behaviour of facial asymmetry during growth.

Functional therapy is useful in all cases in which it is necessary to stimulate the growth of the structure affected by HM during pre-pubertal growth; it’s impossible to perform the same therapy in the case of palsy of the ramus marginalis, because the malformative picture is due to the lengthening of the hemimandibular body on the HM side rather than to hypo-development. In these forms we clinically observe a vertical hypo-development of the ramus; a stimulation of the vertical growth of the ramus is useful to restore symmetry. At present, it is possible to correct the deficit of this bony structure early, with functional therapy in the less severe cases and with distraction osteogenesis of the ramus in the more serious ones.

The neurological deficit of the facial nerve should be treated as soon after birth as possible; facial muscles are embryologically not related to the temporo-mandibular area and they are unaffected, so, if the paralysis is detected early, it is possible to perform a neuro-surgical intervention for restoring the neurological function in the region of the palsy and a symmetric perioral neuro-muscular function. Various treatment options [MacQuillan et al., 2003] are described in literature: a crossed facial nerve graft [Iñigno et al., 1993], transposing the anterior belly and the tendon of the digastric muscle to the region of the lower lip’s orbicularis oris [Conley et al., 1982], using a neurovascular free muscle [Takushima et al., 2002]. Without this, a late mandibular repositioning obtained with surgical treatment cannot assure long term stability (see case 3) because the functional deficit of the neuro-muscular system will progressively condition the morphology of the skeletal structures: for this reason, it is very important to identify the pathogenetic mechanism of this condition for an accurate therapeutic program and to avoid the relapse of the pathology after an orthodontic or maxillofacial surgical treatment.

Conclusions

Patients with HM with facial nerve palsy have a deviation of the mandibular interincisor line towards the opposite side of HM. This clinical picture is quite different in patients with HM and no facial nerve palsy. Our study reports the correlation between the peripheral nervous and muscular deficit and the abnormal growth of the mandible; the mandibular deviation in these cases is clinically due to an elongation of the mandibular body ipsilateral to HM, secondary to the alteration of the “sling-like” muscular mechanism in the chin caused by the ramus marginalis palsy.

In our opinion, restoring a functional, neurological and structural symmetry of the perioral structures is critical, though not always possible. For this reason, in patients with HM and facial nerve involvement, it is critical to perform an early neurological examination in order to assess the neurological damage on the HM side.
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