Simultaneous maxillomandibular distraction osteogenesis in severe progressive hemifacial atrophy with two distractors

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Progressive hemifacial atrophy is a rare disorder characterized by an acquired, idiopathic, self-limited, unilateral facial atrophy involving skin, subcutaneous tissue, fat, muscle, and bone. Symmetry and contour restoration are the main treatment challenges. Among many techniques, microvascular reconstruction has been introduced as the gold standard to correct the atrophic deformity. For some patients with severe manifestations, soft tissue reconstruction alone does not obtain the desired outcome. In this series, we used an effective method to restore the severe progressive hemifacial atrophy by simultaneous maxillomandibular distraction osteogenesis with 2 distractors. The results demonstrate an improvement in both the profile and the occlusion plane of the patients with corresponding satisfactory esthetic and functional outcomes. We conclude that the simultaneous maxillomandibular distraction osteogenesis with 2 distractors is an effective method for hemifacial atrophy and bone frame reconstruction, especially ones involved in the discrepancy of the occlusal plane. (Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2011; 111:292-297)

Progressive hemifacial atrophy (PHA), also known as Parry-Romberg syndrome, is an uncommon and poorly understood degenerative condition. It is a rare pathologic process characterized by a slow and progressive atrophy affecting one side of the face and involving skin, subcutaneous tissue, fat, muscle, and bones. It often localizes and progresses according to the distribution of one or more branches of the fifth cranial nerve. The onset of the disease usually starts in the first or second decade of life, and often between the ages of 5 and 15 years. Generally, the atrophy is more commonly unilateral and more often in female individuals. The cause of the disorder is still unknown; many etiologies have been suggested, including trauma, infection, heredity, immunologic abnormality, trigeminal neuritis, and scleroderma. The treatment is systematic and directed at reconstructing the deficient tissue to improve the contour and symmetry of the face. Most patients with mild or moderate manifestations may only need soft tissue reconstruction. Microvascular reconstruction has been introduced as the gold standard to correct PHA. Many flaps have been used in clinical settings, including latissimus dorsi, groin, anterolateral thigh, scapular, and radial forearm types, among others. However, for some patients with severe manifestations, they need not only soft tissue reconstruction, but also hard tissue reconstruction to achieve the optimal esthetic and functional outcomes.

Distraction osteogenesis has become an increasingly popular procedure in recent decades for craniofacial malformations; historically it was treated with complex osteotomies together with bone grafting. In 1989, McCarthy et al.1 first reported successful mandibular lengthening by gradual distraction. Since then, mandibular distraction osteogenesis has become a reliable procedure in the management of mandible defects, and the success of this treatment has been well documented. However, the simultaneous maxillomandibular distraction osteogenesis with 2 independent distracters has not been well documented. We are reporting our clinical experiences in the reconstruction of the bony frame and occlusal relationship of PHA by 2 simultaneous maxillomandibular distraction osteogenesis procedures.

CASE SERIES
Patient data
A total of 5 patients with severe PHA received hard tissue reconstruction by maxillomandibular osteogenesis distraction and soft tissue augmentation with latissimus dorsi flaps between July 2001 and January 2009 in the Department of Oral and Maxillofacial Surgery in the Guanghua School of Stoma-
There were 4 male and 1 female patients in the series, and their ages ranged from 14 to 21 years. The average age of onset of the disorder was 10 years, and the average duration of atrophy was 6 years. All of the patients had unilateral progressive atrophy (1 patient had right-sided atrophy 4 patients left-sided). The unilateral atrophy involved the skin, subcutaneous tissue, fat, muscle, and bones and was associated with oral occlusion. A large linear fibrotic coup de sabre passed the midline of face. The soft tissue defect extended to the buccal division and labial parts of the infraorbital and submandibular regions. Most important of all, the hard tissues had atrophied, including the orbital bone, zygomatic bone, maxilla, and mandible. Consequently, the discrepancy of the occlusal plane and malocclusion were predominant (Fig. 1). There were no positive neural signs in the series.

**Surgical technique**

Before surgery, the extent of discrepancy was measured according to the distance from the oculopupillary plane to the occlusal plane. Model surgery was performed to predict the effect and guide the operation. During surgery, the standard submandibular incision was performed in the atrophied side to expose the mandible. An osteotomy was performed next in the mandibular ramus parallel to the mandibular occlusal plane, taking care to preserve the mandibular branch of the trigeminal nerve. Before finalizing the mandibular osteotomy, a distractor was fixed to the mandibular ramus on each side with titanium screws (Fig. 2, A). Simultaneously, a modified Le Fort I osteotomy was performed to down-fracture and mobilize the maxilla. Meanwhile, another distraction device was fixed to the maxilla on each end of bone (Fig. 2, B). The osteotomies were then completed, and the distractors were activated to assure adequate movement. The distractors were then deactivated to their original position. A 7-day latency period was observed to allow for healing of the periosteum and soft tissue. After the latency period, the maxillomandibular distraction was initiated without using simultaneous intermaxillary fixation. The mandibular distraction rate was 0.5 mm, and there was a twice-daily activation. The maxillary distraction rate and rhythm were adjusted according to the mandibular protocol (~0.2-0.5 mm per day). The distraction period lasted 3-5 weeks. The devices were activated until achievement of the following desired clinical end points: horizontalization of the mandibular occlusal plane and displacement to the midline of the chin position or slight overcorrection to the contralateral side.

At the completion of the distraction phase, a consolidation phase of 12-20 weeks was secured to allow for bone healing and remodeling. After the consolidation, the distractors were removed. The processes of distraction osteogenesis were monitored, and the new regenerates were evaluated with radiography (Fig. 3). At 6-8 months after completion of distraction osteogenesis, latissimus dorsi flaps were used to reconstruct the soft tissue defects (Fig. 2C and D). The flaps ranged from 10 × 11 cm to 15 × 18 cm and were raised by regular methods and trimmed to fit with the temporal and midline areas. On the recipient site, the standard preauricular and submandibular incisions were performed. A pocket was made through the dissection and run between the subcutaneous tissue and muscle layers, being careful to not to damage the facial nerve. Meanwhile, the facial vessels and external jugular veins were clearly exposed. The flaps were inserted into the pockets above the muscle and sutured into the temporalis fascia and periosteum. An end-to-end anastomosis of the thoracodorsal vessels to either the facial vessels or the external jugular vein was performed. Finally, some anchoring sutures were made between flaps and the pockets along the rim of atrophy in the midline and incisions, especially in the

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**Fig. 1. Clinical findings before surgery intervention.**

A, C, E, Frontal views showing facial asymmetry: The atrophy was severe and involved the soft tissue and bone. B, D, F, The discrepancy of occlusal planes could be distinguished clearly.
endocanthion, temporalis, basis nasi, labial commissure, and genion. The donor sites were closed directly.

RESULTS
After distraction osteogenesis, the mandibles were elongated to a simultaneous average of 20 mm. All of the patients experienced a marked esthetic improvement. No device failure, fracture, or infection was found other than an unerupted molar injured during the Le Fort I osteotomy. There were no dental necrosis cases resulting from screws within the dental root. Lateral cephalometric radiographs showed a mild change in the anteroposterior position of the maxilla and mandible (Fig. 4). At the same time, the occlusal plane discrepancy was corrected (Fig. 5). However, the atrophy of soft tissue was more obvious. These deformities improved predominately after latissimus dorsi flap transplantation. An ancillary surgery was performed to mold the lips in 1 patient due to the vermilion atrophy. There were no cases of postoperative facial nerve paresis.

DISCUSSION
The causes and mechanisms of PHA are still poorly understood. Many conservative treatments, such as physiotherapy, acupuncture, and immunotherapy, have been unable to achieve optimal outcomes. The most recent therapies surgically resolve symptoms. In all cases, the aim of treatment is to restore facial symmetry and contours and reconstruct the atrophic bones and soft tissues. Most surgeons suggest that surgical intervention should be performed after cessation of ongoing atrophic process, usually after a period of ≥1 year. Before the surgical interventions are performed, it is crucial to evaluate the degree of atrophy. Inigo et al.2,3 proposed the following classification of PHA based on the skin and subcutaneous tissue atrophy and bone involvement in trigeminal nerve territories: mild: atrophy of the skin and subcutaneous tissues affecting the territory of only one of the branches of the trigeminal nerve, no bone involved; moderate: atrophy of tissue affecting two trigeminal territories, no bone involvement; and severe: all three trigeminal territories affected or bone involved.

To treat mild and moderate Romberg disease, soft tissue reconstruction can achieve optimal esthetic outcomes. A number of procedures have been developed to reconstruct the soft tissue defects. Among these methods, the more popular include fat injections and transplantation of dermis-fat, fascia-fat, tissue substitutes, and vascularized free flaps.4,5 Generally speak-
ing, lipoaspirated fat injection is an easy method for treating mild facial atrophy, but repeated procedures are needed because the absorption of fat transplantation is unpredictable and has a problematic survival rate. Nonvascularized grafts are restricted in both size and thickness owing to blood supply; they also have a high

Fig. 3. Panoramic radiographs showing the process of maxillomandibular distraction osteogenesis. A, Discrepancy of the occlusion plane was severe before distraction osteogenesis. B, C, D, E, The distractors were activated gradually and the bone (red arrow) was elongated. F, The new bone (red arrow) was regenerated and calcified well.

Fig. 4. Lateral cephalometric radiographs showing a mild change in the anteroposterior position of the maxilla and mandible.
degree of unpredictable absorption. The long-term effects are unsatisfactory even if the facial contour is acceptable. Biomaterials or substitutes could resist resorption but have the risk of extrusion. With the advent and rapid development of microvascular techniques, vascularized free flaps have become the gold standard for progressive hemifacial atrophy reconstruction.2,6,7

However, to treat severe hemifacial atrophy, both soft tissue restoration and hard tissue reconstruction are necessary. Before soft tissue augmentation, skeletal reconstruction is necessary to provide soft tissue support. Otherwise, excess soft tissue could lead to facial sagging and absorption, making the facial contour unsmooth. Recently, many techniques, such as bone grafts, biomaterials, and tissue bone engineering, have been developed to reconstruct bone defects along with distraction osteogenesis.8-11 All of these methods have their respective disadvantages and advantages; the choice should be flexible, based on patient status. Moreover, for the cases involving the gnathostomatic system, the orthognathic and orthodontic surgery should correct both the oral and maxillofacial deformities and reconstruct the normal occlusal relationship to provide the most optimal functional and esthetic outcome.

In the present series, all patients had severe deformities affecting not only the facial bones but also the gnathostomatic system. Clinical examination revealed a severe occlusal plane deviation and malocclusion. If only the soft tissues are reconstructed, the facial profile symmetry cannot be effectively restored. Moreover, the oral and maxillofacial deformities would not have been corrected. Therefore, the skeleton frame must be reconstructed as the first step. The autologous bone graft transplantation needs another surgical procedure, because it does not correct the occlusion deformity. The orthognathic surgery is constrained by soft tissue, because the maxilla and mandible moved 20 mm downward after osteotomy. This causes a dilemma in the treatment of severe hemifacial atrophy, which can be solved by distraction osteogenesis. The hard and soft tissues can regenerate and elongate by distraction. The skeleton frame can be reconstructed without soft tissue constraint. Most important of all, the occlusal plane discrepancy can be corrected and primary normal occlusal relationship may be established. Thus, in the present cases, at the first step maxillomandibular distraction osteogenesis was performed to reconstruct the skeleton frame and correct oral-maxillary deformity. At the second step, vascularized latissimus dorsi flaps were raised and transplanted to restore the facial contour symmetry. Finally, orthodontics adjusted the exact occlusal relationship and established a stable functional occlusion. Moreover, some ancillary surgical procedures may be necessary to achieve optimal esthetic effect.

The reconstruction of hard and soft tissues was contradictory, because of severe atrophy in the facial bones and gnathostomatic system. Distraction osteogenesis solved the dilemma successfully. The maxillomandibular distraction osteogenesis not only reconstructed the facial skeleton frame to support soft tissue, but also corrected the oral-maxillary deformity to establish a functional occlusal relationship. Finally, after ortho-
dontics and ancillary surgery, the esthetic appearance and functional occlusion were achieved.

Thus, simultaneous maxillomandibular distraction osteogenesis with vascularized flaps can provide a good option for treating severe Romberg disease, especially in cases involving the gnathostomatic system. It can not only restore the facial contour symmetry, but also reconstruct the occlusion to improve masticatory function.

REFERENCES


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