Restitution of Facial Form in a Patient with Hemifacial Microsomia

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a case report

Hemifacial microsomia involves unilateral underdevelopment of the ear, mandible and associated structures. This report describes an approach to early surgical restoration of function to improve growth, illustrated with a case report.

KEY WORDS: • Congenital malformation • Function • Growth • • Hemifacial microsomia •

Hemifacial microsomia is the term used by Gorlin and Pindborg (1964) to describe a syndrome characterized by varying degrees of underdevelopment of the ear, mandible and contiguous bony structures of the cranium and face. They suggest that oculoauriculovertebral dysplasia (Goldenhar Syndrome) is a variant of hemifacial microsomia which is sometimes also known as *first arch syndrome* or *first and second branchial arch syndrome.*

A branchial arch dysplasia should be regarded as a nonspecific symptom complex, the pathogenesis of which may have several different etiologies (Poswillo 1974).

While the etiology of hemifacial microsomia is still unresolved and complicated, Poswillo (1973) produced remarkably similar deformities in animals by causing fetal hemorrhages centered on the stapedial artery system. He postulates that hemifacial microsomia may be caused by a vascular occlusion in the second month of fetal development. The site of such a vascular occlusion would thus determine the extent of the facial hypoplasia.

In patients with severe forms of hemifacial microsomia the ramus is reduced to a thin lamina of bone, or there may be complete agenesis of the ramus. The

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zygomatic arch is absent and the zygoma hypoplastic to some degree. Absence of the lateral pterygoid muscle limits mandibular movement toward the normal side. In these patients the facial asymmetry becomes progressively worse during the formative years as the growth disparity causes the mandible to deviate laterally and remain relatively high on the affected side.

While these growth characteristics are probably genetically determined, with each individual following a specific pattern, external form and internal structure are also related to functional stresses (Gans and Bock 1965). As the mandible grows, it adjusts within the craniofacial complex in an organized manner around the associated neurologic distribution (Ricketts 1975).

To explain growth variations from an established genetic type, an analysis of the force couples surrounding the mandible supports the theory that severe deviations from the genetic pattern may occur under the influence of abnormal muscle conditions (Ricketts 1975).

— Treatment —

Cognizance of the determinants of mandibular form and growth is crucial in the treatment of congenital defects such as hemifacial microsomia (Harvold 1975). Treatment requires that an extension to the mandibular ramus be constructed and maintained. Maintenance of this additional bone depends in turn on the creation of appropriate functional loading of the bone as a whole.

Treatment planning and results obtained in a specific patient illustrate the principles involved. A six-year-old girl presented with severe left hemifacial microsomia (Fig. 1). The left zygoma was hypoplastic, the zygomatic arch was absent, and there was no glenoid fossa. The mandible on the left side was also hypoplastic, with only a small remnant of ascending ramus present. In frontal view, the occlusal plane tilted up to the left and the midline of the mandible was grossly deviated to the same side. In the profile, marked retrognathia was the most striking feature.

Full-thickness bone grafts were taken from the right parietal bone in strips 3.8cm × 2.0cm. A zygomatic arch constructed from this excised parietal bone was extended from the zygoma to the region of the missing glenoid fossa. A depression with the approximate position and shape of a glenoid fossa was created on the under surface of this new zygomatic arch. Strips of bone were also used to construct a left lateral orbital wall and to build out the left temporal fossa.

Two layers of skull-bone graft were used to reconstruct the left mandibular body and ascending ramus. The mandible was over-corrected to the right side, and that portion of the bone graft corresponding to the condyle was positioned into the newly-constructed glenoid fossa. The graft was wired to the mandible but not to the glenoid fossa, where it was held in position only by the pull of the soft tissues.

The postoperative course was uneventful. Facial nerve function was the same as before the operation. Interdental fixation was maintained for four weeks.

Although this report is essentially based on clinical observations, cephalometric records were also made. Tracings of the lateral cephalometric radiographs taken immediately before the surgical procedures and two years postoperatively show a marked reduction in the A–N–B angle over this period (14° to 6°). The angle S–N–A remained relatively constant at 77°. Permanent upper incisors had not erupted at the time of the follow-up lateral cephalometric radiographs, and eruption of these teeth could result in a change in the angle S–N–A.

Fig. 1 The six-year-old patient with hemifacial microsomia.

Lateral and postero-anterior cephalometric radiographs (Figs. 2 to 5) show the marked improvement in facial form and symmetry.

Panoramic radiographs (Fig. 6) graphically illustrate the effects of the bone graft on the mandibular morphology. The angle of the mandible on the affected side is better developed and the unerupted second permanent molar is situated in newly-formed bone of the reconstructed mandibular body. The constructed condyle is seated in the new glenoid fossa.

There appears to be a functional relationship between the mandible and the reconstructed fossa. The patient is able to open her jaws normally, and the deviation toward the affected side is very slight. There is still a dental crossbite on the right side.

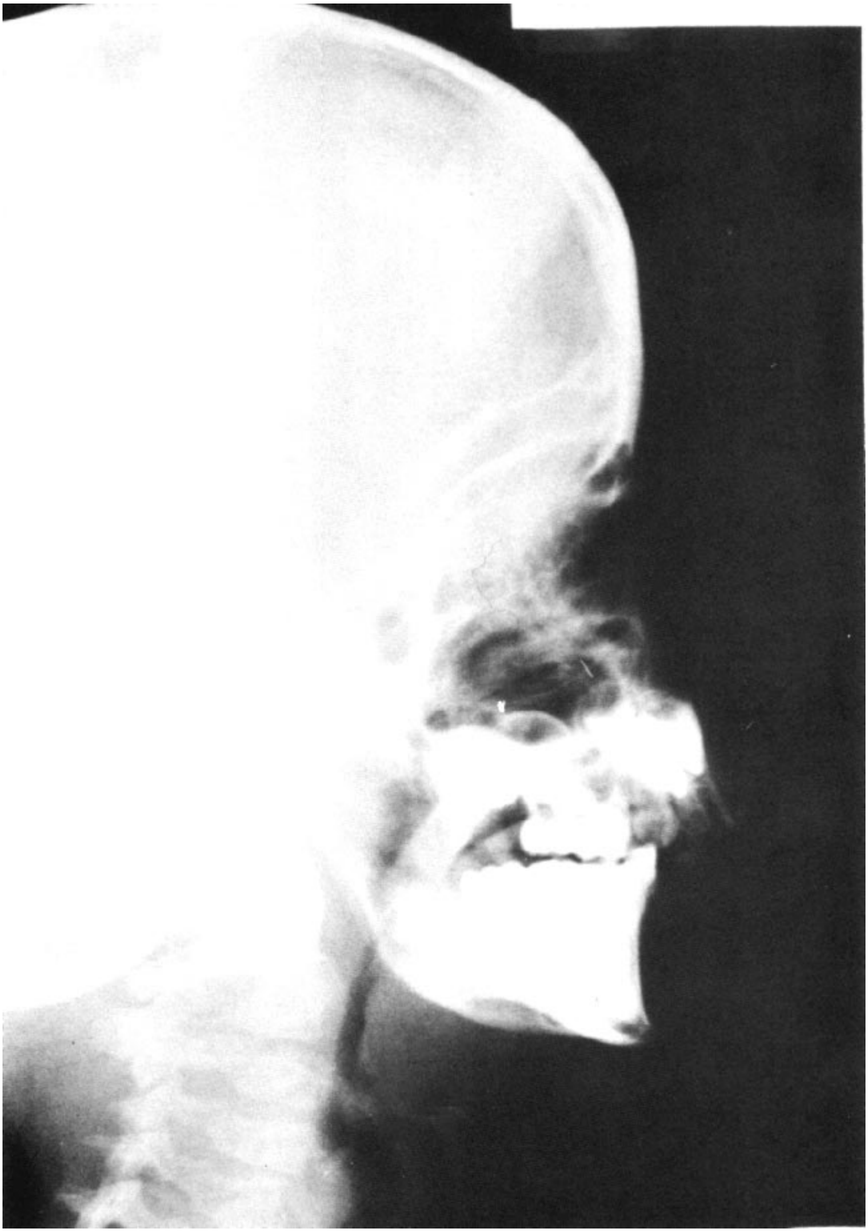

Fig. 2 Preoperative lateral radiograph

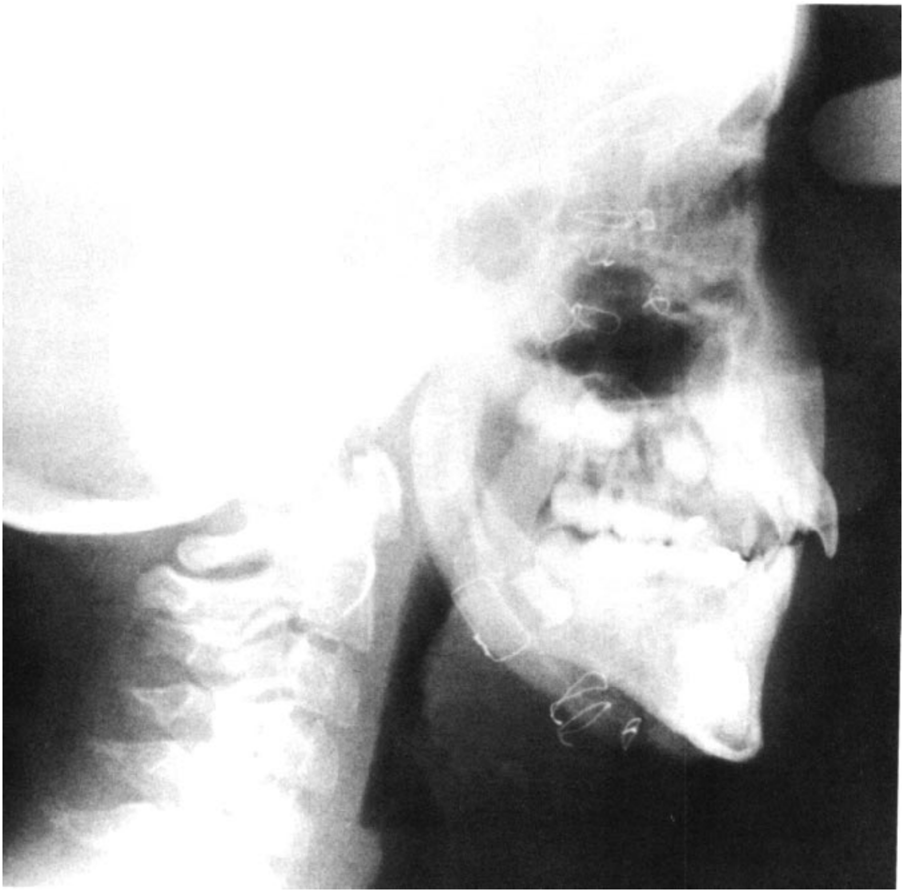

Fig. 3 **Postoperative lateral radiograph**

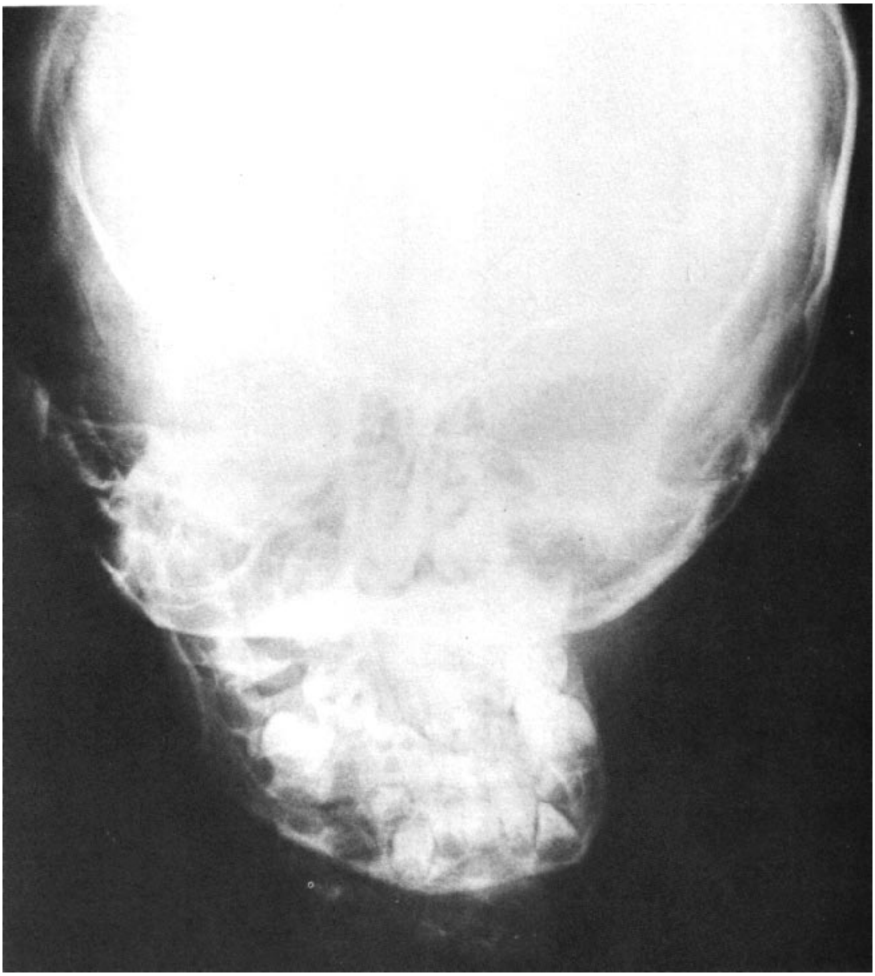

Fig. 4 **Preoperative postero-anterior radiograph**

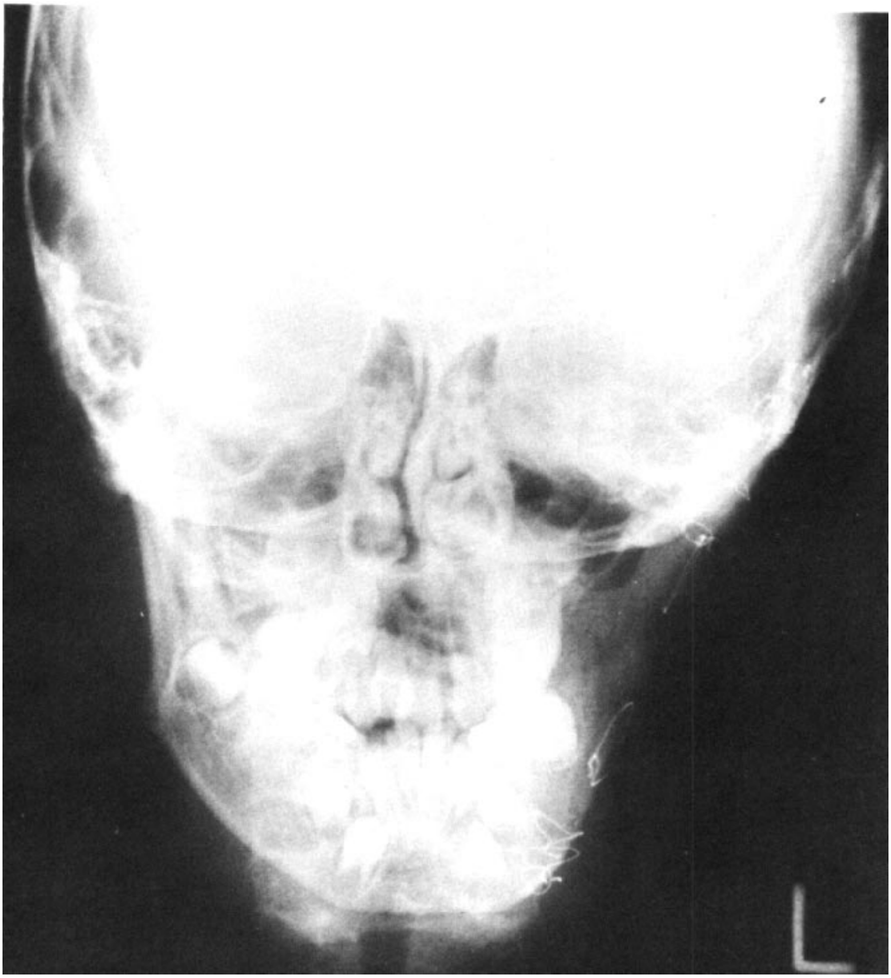

Fig. 5 **Postoperative postero-anterior radiograph**

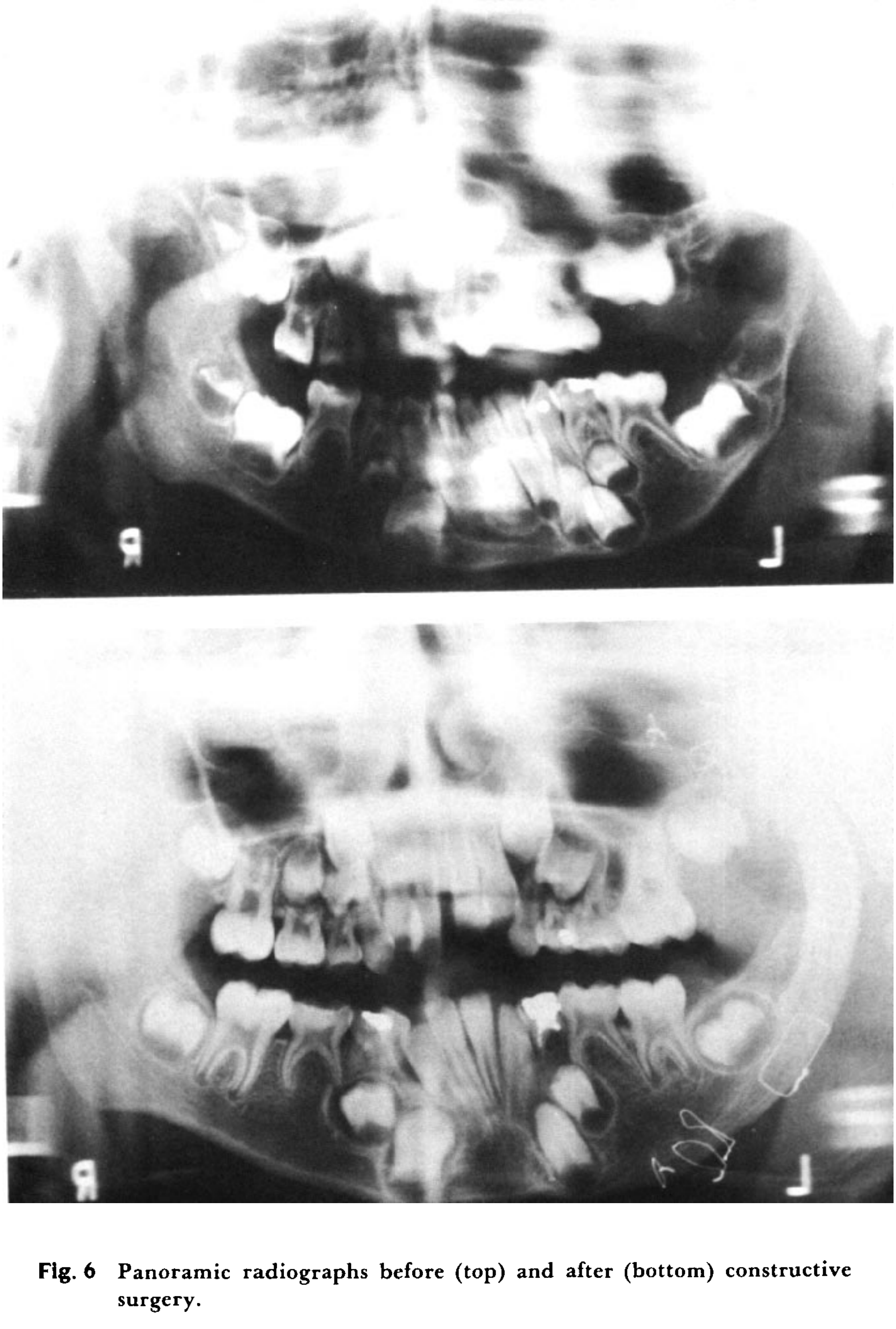

Fig. 6 Panoramic radiographs before (top) and after (bottom) constructive surgery.

— Conclusions —

The damaging effects of hemifacial microsomia on facial form become increasingly apparent with age, and there is evidence that the restoration of function has a beneficial effect in cases with dysplastic condyles (COCCARO 1969). O'RYAN AND EPKER (1982) suggest that surgical alteration of mandibular growth can be accomplished in growing individuals, noting Moss's belief that mandibular growth is a compensating process with the mandible a part of a "functional matrix".

These principles prompted the early surgical intervention in this six-year-old patient with a hemifacial microsomia. The rationale behind the mode of therapy reported here is to use the jaw on the affected side more and more, rather than less and less, particularly during the critical years when mandibular growth is active. At operation the skeletal deficiencies in the left temporal fossa were restored by appropriate bone grafts, and the mandibular ramus was augmented with grafted bone.

The significant improvement in function appears to have been associated with a demonstrably favorable growth response of both maxilla and mandible. Restoration of more normal functional stimuli at a relatively young age is assumed to have secured considerable advantages in terms of more balance in both skeletal growth and muscle development. With this improved function and growth, the patient has also experienced a marked positive psychological change. Further surgical soft tissue correction is planned at about 16 years of age. A/O

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