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OTOMANDIBULAR DYSOSTOSIS

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CLINICAL SYMPTOMS

The relevant embryology, etiology, and clinical features of the first and second branchial arch syndrome, otomandibular dysostosis, have been explicitly covered elsewhere. The syndrome, in various gradations, is thought to occur as a result of hematoma formation in the region of the embryonic stapedial artery.[32] Rarely, the condition may be bilateral, but it has been reported to present predominantly on the right side of the face.[24] The hypoplasia on the affected side is three-dimensional (vertical, horizontal or lateral, and sagittal).[26] It involves, in severe cases, the anterior and middle cranial fossae, with the orbital conus positioned inferiorly; the auditory ossicles, external auditory canal, temporal bone, zygomatic bone and arch, glenoid fossa, mandible (condyle, ramus, and body), base of the maxilla, muscles of mastication and facial expression, soft palate and pharyngeal wall and tongue, facial soft tissues, parotid gland, facial nerve, skin, and pinna (all of which may be hypoplastic or rudimentary).[6, 7, 24] Secondary nasal deformity, deviation of the chin prominence, canting of the occlusal plane, and tooth malalignment are usually present. Macrostomia occurs, with a subcutaneous scarlike band passing from the angle of the mouth toward the affected ear. Skin tags (often incorporating cartilage) and sinus tracts are frequently found anterior to the affected ear.[26]

Very rarely otomandibular dysostosis may also be associated with clefting of the lip, the palate, or both (Fig. 14–1).

EVALUATION OF THE PATIENT

Clinical Assessment

The clinical assessment of the patient is of course the cornerstone to treatment planning and cannot be dispensed with by any multitude of preoperative diagrams, measurements, and computer analyses. This evaluation is, indeed, part of the art of maxillofacial surgery. Surgical experience and intuition, an appreciation of the esthetics particularly applicable to the race and cultural background of the patient being investigated, and a knowledge of the concerns relating to his deformity are imperative.

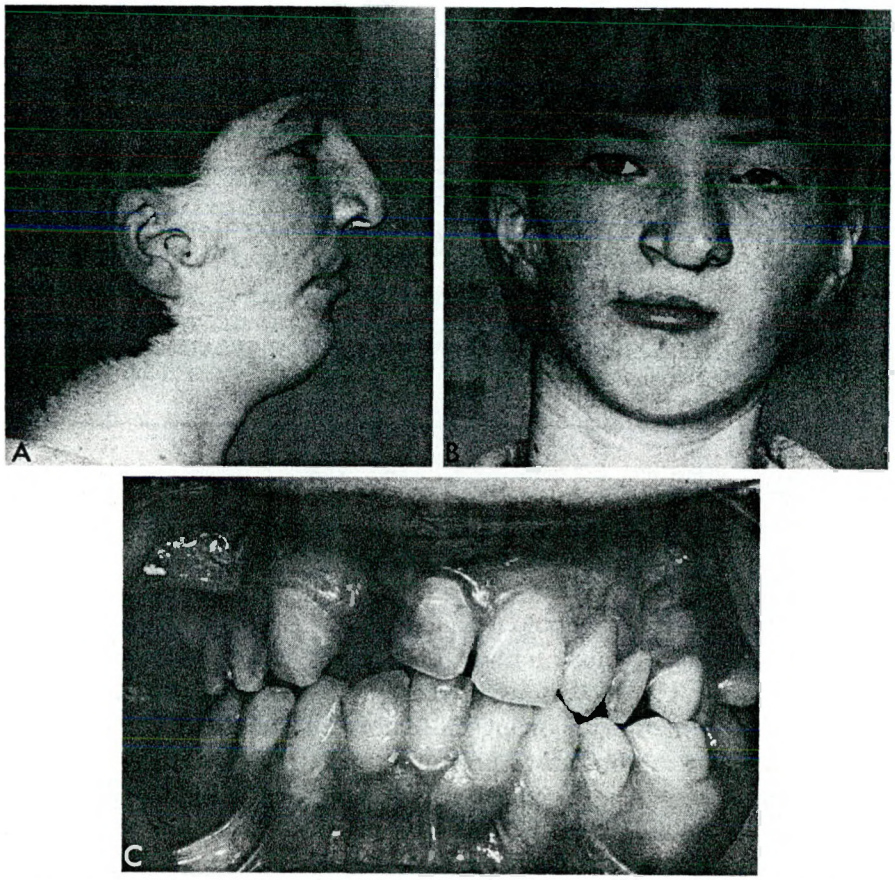


Figure 14–1. *A, B,* and *C,* Preoperative photographs of patient with a previously repaired right total cleft lip and palate as well as otomandibular dysostosis.

Illustration continued on opposite page

Planning Prerequisites

RADIOGRAPHS. Standard radiographic projections are obtained initially to assist in the clinical evaluation and treatment planning as well as to serve as records with which to assess the accomplishment of preoperative goals. Radiographic projections should include Towne's or Water's view, posteroanterior and lateral skull views, orthopantomogram, posteroanterior view of the mandible during maximum mouth opening, temporomandibular joint and ramus tomograms, computed tomograms, and frontal linear tomograms.

Extensive measurements of the lateral cephalogram are not considered necessary and can, on the contrary, often prove to be misleading.[10] It is important to determine the various angles and other measurements pertaining to the spatial relationship between the anterior teeth and the respective jaw base, as normalization of this relationship must be of prime concern in preoperative orthodontic treatment.

TRACINGS. Acetate paper tracings and cutouts of the maxillary, mandibular, and chin segments to be repositioned surgically, although they form part of the preoperative work-up, serve only as guides.

PHOTOGRAPHS. Preoperative photographs in standard views are essential: frontal

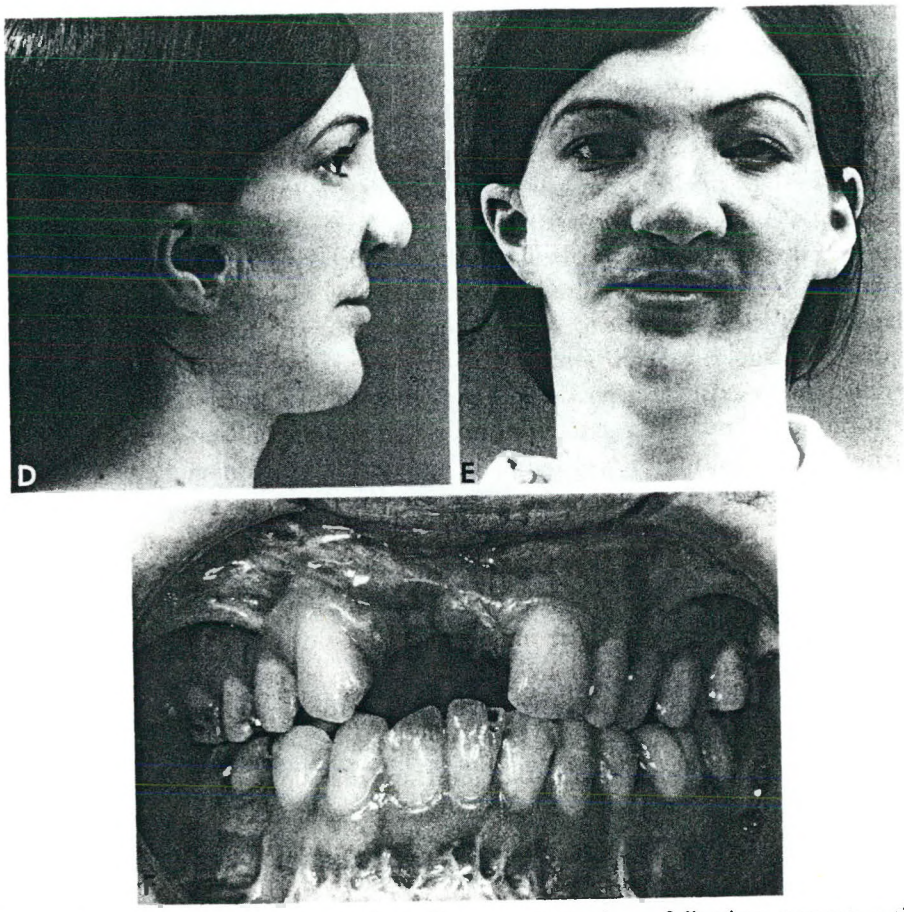


Figure 14–1 *Continued. D, E,* and *F,* Postoperative views following reconstruction and recontouring of the ascending right ramus and right zygomatic arch with autogenous rib, closure of a residual palatal cleft, chin correction, and various nose and lip correction procedures. Further refinements and correction of the left-sided coloboma will follow.

views of the face in centric occlusion, with maximum mouth opening, and when biting on a wooden spatula placed between the teeth to reveal the angular difference between the occlusal plane coronally and the interpupillary plane; left and right profile views; and an angled view of the face from below. Anterior left and right occlusal views together with intraoral mirror views of the mandibular and maxillary dental arches should also be taken.

MODELS. Stone models cast from impressions serve as a record of the preoperative intermaxillary relationship, and plaster operation models are used for sectioning to simulate movement of the respective jaws following the intended osteotomies. These models are mounted in a semi-adjustable articulator. When taking the face-bow reading, one should be cognizant of the anteromedial location of the deformed condyle, which renders the reading inaccurate in unilateral cases and virtually meaningless in cases of bilateral asymmetric otomandibular dysostosis.

The model operation also assists in assessing the amount of transplant bone required for stabilization. Usually autogenous pelvic bone or rib (or both) is used, although deep-frozen bank bone and allogeneic lyophilized cartilage may be used in addition.[36]

TIMING OF TREATMENT

The surgeon is often faced with the decision of when to initiate skeletal surgical intervention. In a mature patient the issue presents no immediate problem. When faced by distraught parents and a child obviously aware of and psychologically upset by his deformity, however, the surgeon is under great pressure to effect an all-encompassing treatment as soon as possible.

Appropriate guidelines to the timing of surgery for treating otomandibular dysostosis may be drawn from the work of Poswillo, who reasons that surgery in an area already affected in early development by necrosis may lead to insidious and irreversible damage.[32, 33] Additionally, facial skeletal reconstruction where obvious muscle deficiencies exist is, in terms of Moss's functional matrix theory, at best a short-lived restoration of facial balance, requiring repeated surgical replacement procedures, and further compounding the possibility of greater tissue damage.[14, 33]

The advocates of early skeletal surgical intervention hope to alleviate the patient's and the parents' psychologic trauma while also attempting to minimize secondary compensatory changes induced in adjacent soft tissues, bones, and dentition.[2, 8, 9, 13, 30] Converse and associates proposed a vertical osteotomy of the hypoplastic ramus, displacing the mandible downward on the affected side, creating an open bite, and inducing caudal growth of the maxillary alveolar process with the teeth.[2] Munro, Murray, and Ortiz-Monasterio await the eruption of the first permanent molars before employing Obwegeser's standard treatment approach with reconstruction of the zygoma, in which the temporomandibular joint and deficient ramus are reconstructed, the lower half of the face is rotated, and the chin prominence is relocated.[15, 16, 23, 24, 26, 29]

It has as yet not been clearly established whether early radical surgery in severe cases results in an adequate, long-term improvement of the lateral (horizontal) and sagittal hypoplasia in the mandibular, temporal, and zygomatic bone regions. Thus, following completion of growth, further major surgery may be necessary to reconstruct the temporomandibular joint prior to repositioning the lower half of the face, reconstructing the ramus, and correcting an asymmetric chin.[24, 26] Unequivocal evidence does exist that growth of congenitally hypoplastic midface skeletal areas corrected by osteotomies in childhood is further retarded.[3, 4, 28]

In most cases, and particularly in severe cases, early onlay bone grafting to camouflage the deformity may provide a modicum of esthetic improvement, sufficient to allay the psychologic trauma experienced by both patient and parents. Until growth has ceased, however, surgery on the facial skeleton and subsequent soft-tissue correction should be delayed for as long as possible.

Successful growth center transplants (metatarsal bone, costochondral junction, head of fibula, sternoclavicular joint, or rib) serve to restore an adequate functional matrix during the growth period, and in cases in which electromyography has revealed good masticatory muscle function, these transplants may make possible earlier surgical correction of otomandibular dysostosis.

ORTHODONTIC TREATMENT

Preoperative orthodontic preparation of the dental arches is necessary in order to decompensate the dentition and facilitate intraoperative interdigitation of the teeth and intermaxillary fixation. Although functional efficiency may, in fact, initially be reduced preoperatively, correct alignment of the two arches is essential for postoperative

occlusal stability. The repositioning of the maxillary and mandibular premolars and molars, usually lingually inclined on the side of the deformity, helps to support the soft tissues of the cheek.

Particular care should be taken, when correcting the axial inclination of the teeth (tilted toward the affected side) and the crossbite frequently encountered on the normal side, to position the teeth well within the confines of the available alveolar bone. Intra-arch crowding and particularly transverse arch size discrepancies should be treated by means of planned extractions and additional osteotomies rather than by exaggerated buccolabial placement of teeth with consequent root fenestration and relapse.

Profile esthetics is a reflection of the spatial relationship between the bony bases constituting the face, i.e., the cranial base, the maxillary base, and the body of the mandible, rather than of mere dental arch support to the soft tissues.

Fine orthodontic adjustments may be undertaken postoperatively.

SURGICAL TREATMENT

Preparation for Surgery

Continuous-loop wire splints are applied under local anesthesia, topical anesthesia, or intramuscular neuroleptanalgesia.[18] Variation in splinting is obviously required in patients sufficiently edentulous for intermaxillary fixation with continuous-loop wiring to be unstable. In most cases, however, it suffices, as the patients are young and have good dentition. An acrylic occlusal-wafer splint may be constructed on the articulated plaster model to assist intraoperatively in positioning the mobilized maxilla with respect to the unmobilized mandible. A similar acrylic wafer splint may also be constructed to adapt the mandible, once mobilized, to the repositioned and immobilized maxilla. The former splint tends to reflect errors in face-bow readings and articulator adjustments as well as inaccuracies in the plaster model operation, so, although the splint may be of some use, operator experience in intraoperatively assessing the angle of the occlusal plane relative to the interpupillary line and Frankfort plane, the maxillary midline relative to the vertical facial midline, and incisal edges of the maxillary teeth relative to the length of the upper lip is essential to ensure correct placement of the mobilized maxilla. The rotation of the lower half of the face necessary to correct severe forms of otomandibular dysostosis requires the surgeon to have a good mental grasp of the intended three-dimensional movement of the jaws that is being attempted; reliance on a piece of acrylic fabricated in a laboratory is insufficient. The latter occlusal-wafer acrylic splint, to relate the mobilized mandible to the repositioned maxilla, is usually superfluous, as preoperative orthodontic treatment and an adequate dentition ensure that correct occlusion can be easily ascertained and stable intermaxillary fixation achieved.

General Considerations

Incisions

A tragus border incision placed just endaurally is curved along the upper attachment of the ear sufficiently posteriorly to avoid the posterior branch of the superficial temporal artery.[35] From there the incision passes superiorly within the hairline and is

directed so that it can, if necessary, be extended to meet a similar contralateral incision, allowing a bicoronal forehead flap to be turned if required.[1, 27] This approach is used for the reconstruction of the glenoid fossa and for temporal and zygomatic bone augmentation as well as for transplantation of gluteal fat to the cheek.

Intraoral vestibular incisions are used for completing the sagittal splitting of the mandibular rami, the genioplasty, and the Le Fort I osteotomy.[19–26]

Conventional pelvic crest and thoracic wall incisions are used to acquire the requisite additional bone for augmentation and skeletal support.

Case Assessment

Thorough preoperative assessment of the case to determine both the degree and the exact nature of the deformity as well as the correction required allows one to plan the staging of the surgical procedures according to Pichler's axiom "first the bone, then the soft tissue."[31]

The syndrome of otomandibular dysostosis may find minimal expression as a minor condylar movement discrepancy or take the form of severe three-dimensional craniofacial hypoplasia (Figs. 14–2 and 14–3).

Surgical intervention may be called for in cases with minimal clinical signs and symptoms in which only bone or lyophilized bank cartilage camouflage onlays are needed (Fig. 14–4). Even these mild cases may, however, require a Le Fort I osteotomy combined with bilateral sagittal splitting of the rami as well as genioplasty in order to correct the occlusal plane, facial midline, and esthetics (Fig. 14–5).

Moderately severe cases invariably require initial temporomandibular joint reconstruction followed by rotation of the lower half of the face, ramus reconstruction, and genioplasty.

Severe cases likewise require temporomandibular joint reconstruction, rotation of the lower half of the face, ramus reconstruction, and genioplasty before improvement of extensive soft-tissue malformation can be accomplished. The soft-tissue procedures include correction of ear malformations and macrostomia, and gluteal free-fat grafts to the hypoplastic cheek region (Figs. 14–6 and 14–7).

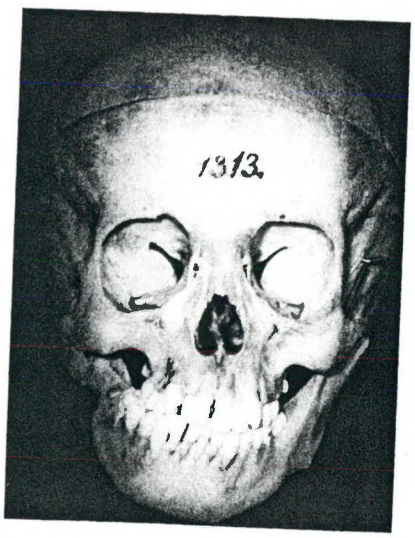


Figure 14–2. Skull demonstrating the three-dimensional asymmetry of unilateral dysostosis otomandibularis. On the affected side there is vertical, sagittal, and horizontal hypoplasia of the mandible, maxilla, zygoma, and temporal bones.

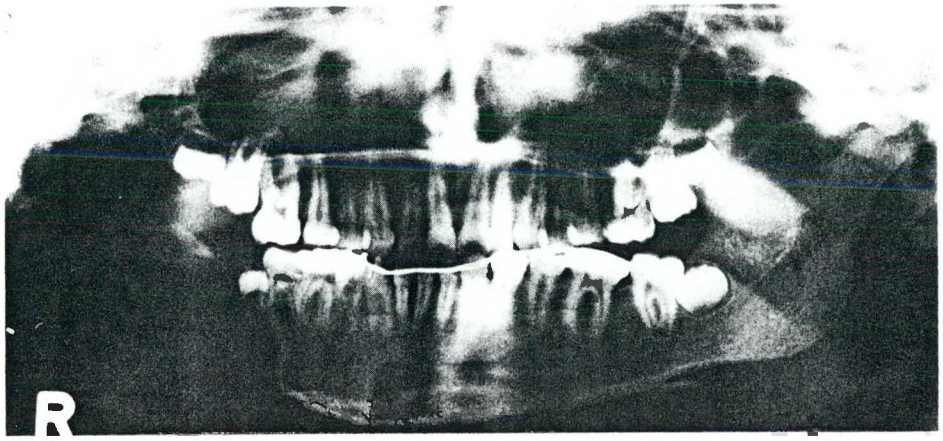


Figure 14–3. Orthopantomogram of a patient with severe unilateral otomandibular dysostosis. The right glenoid fossa and mandibular ramus are completely absent, as is half the right side of the body of the mandible.

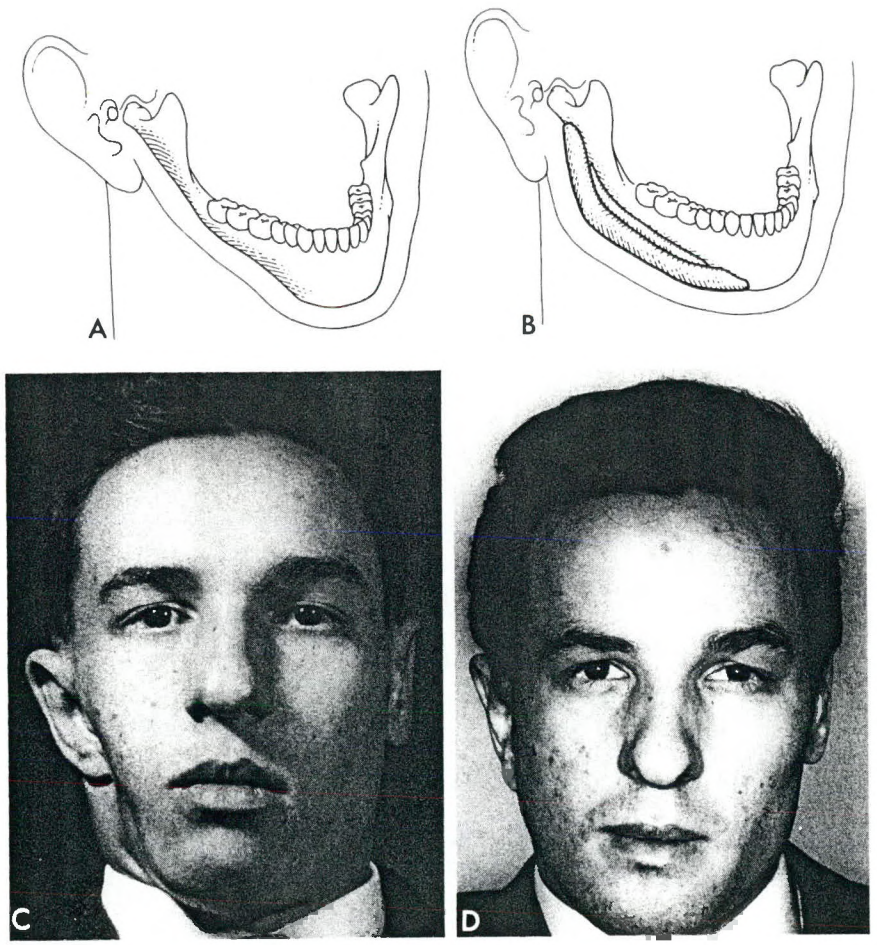


Figure 14–4. *A* and *B*, Mild degree of unilateral otomandibular dysostosis corrected by onlays of lyophilized rehydrated bank cartilage via an intraoral approach. *C*, Preoperative facial appearance of the patient. *D*, Postoperative view following correction of the deformity as depicted in *B*.

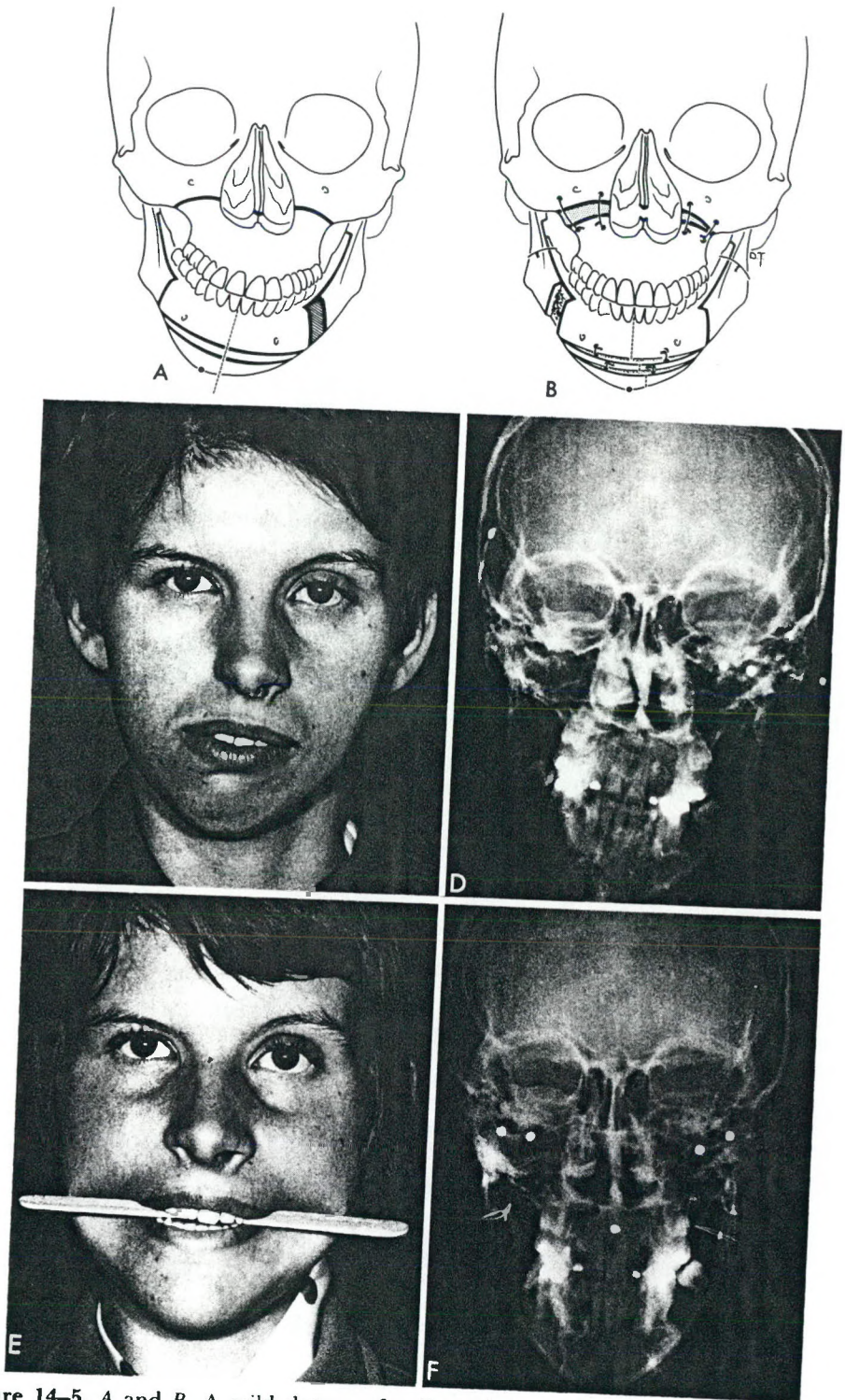


Figure 14–5. *A* and *B*, A mild degree of unilateral otomandibular dysostosis with mainly skeletal defects is corrected by rotating the lower half of the facial skeleton downward and forward by means of a Le Fort I osteotomy and bilateral sagittal splitting of the ramus together with an advancement double-step genioplasty. *C*, Appearance of twisted lower half of face prior to correction. *D*, Radiograph showing deformity prior to correction. *E*, Postoperative facial appearance. *F*, Postoperative radiograph showing symmetry following procedure depicted in *A* and *B*.

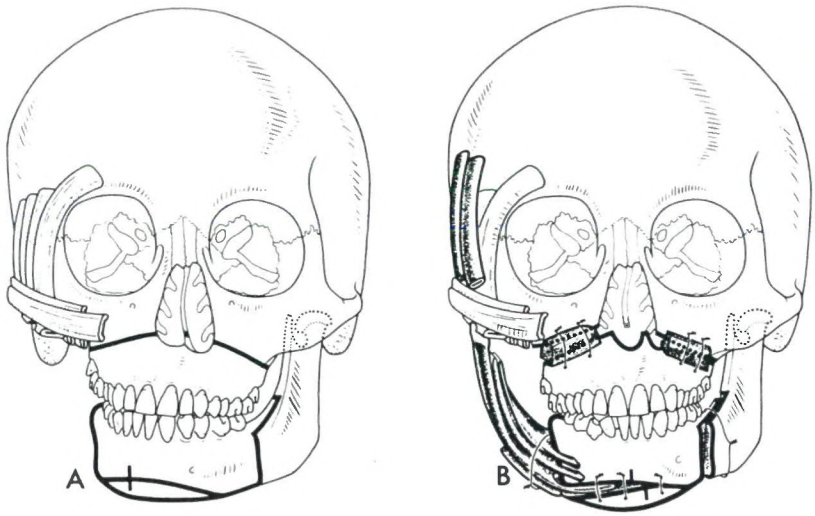


Figure 14–6. *A*, Following initial reconstruction of the missing glenoid fossa and hypoplastic temporozygomatic area, rotation of the lower half of the face is performed. *B*, Multiple rib grafts serve to reconstruct the missing condyle and ramus. The longest rib graft's cartilaginous end is inserted into the previously reconstructed glenoid fossa. Iliac bone is grafted to the maxillary osteotomy.

Unilateral Deformities

Lateral Hypoplasia

When only the correction or camouflaging of lateral hypoplasia is necessary, this may be undertaken at any stage of the patient's growth. It must be borne in mind that subsequent procedures may be necessary to keep pace with further growth.

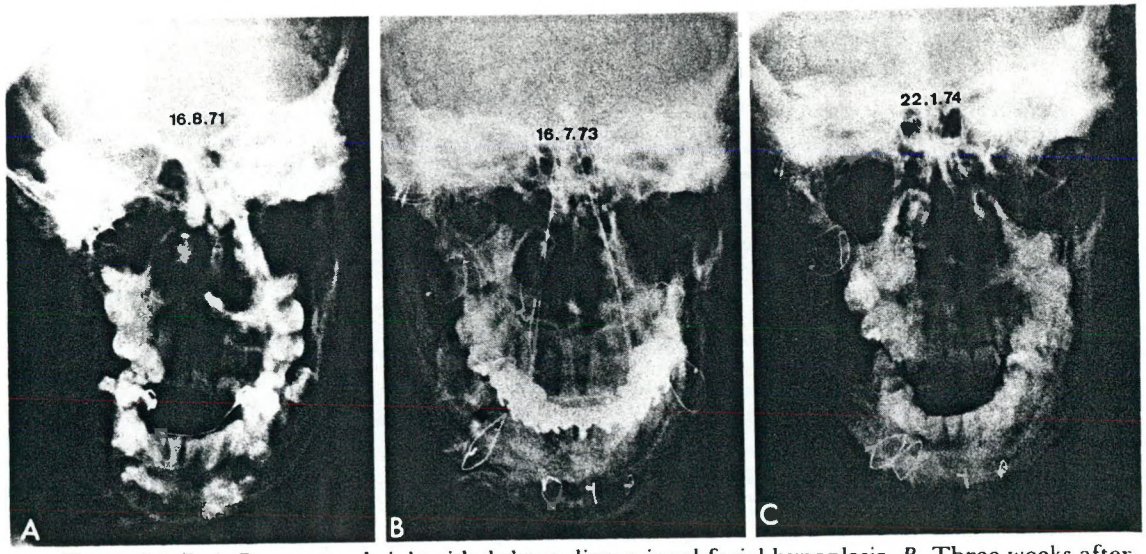


Figure 14–7. *A*, Pronounced right-sided three-dimensional facial hypoplasia. *B*, Three weeks after reconstruction and correction using the procedure shown in Figure 14–6. *C*, The same case six months postoperatively. The facial skeleton is almost symmetric.

For early camouflaging fat tranplants, autogeneic bone or allogeneic lyophilized bank cartilage onlays may be used without fear of interference with later more extensive corrective procedures in the same skeletal areas.[36] It is, however, important to complete the correction of vertical and sagittal discrepancies before attempting correction of the lateral hypoplasia.

Three-Dimensional Hypoplasia

Vertical and sagittal hypoplasia should be corrected by pivoting the lower half of the face anteriorly, inferiorly, and horizontally toward the affected side about a point located in the tuberosity region of the normal side. Anteroinferior overcorrection is recommended.[24, 26] An additional superior displacement on the normal side is necessary to prevent excessive maxillary central incisor exposure. This approach to the correction of severe forms of otomandibular dysostosis is of the essence in achieving an acceptable solution to the problem of a three-dimensional facial deformity. That solution cannot be reached as simply, rapidly, and satisfactorily by other previously published methods.

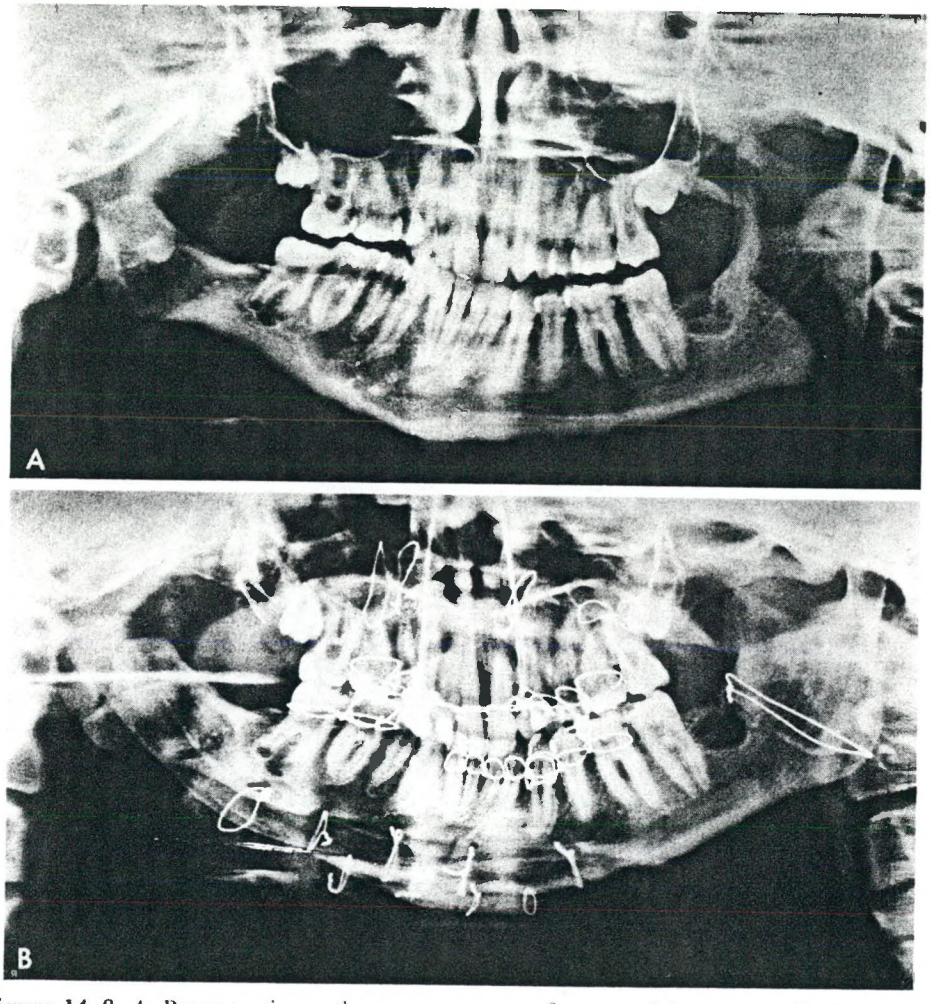


Figure 14–8. *A*, Preoperative orthopantomogram of severe right otomandibular dysostosis. *B*, Initial temporomandibular joint reconstruction was followed by rotation of the lower half of the face and genioplasty. Autogenous rib and iliac bone were used.



Figure 14–9. *A* and *B*, Preoperative views demonstrating the facial asymmetry of the patient whose radiographs are shown in Figure 14–8. *C* and *D*, Postoperative views of the same patient following rotation of the lower half of the face and free-fat grafting of the right cheek.

In order to accomplish this rotation and repositioning of the lower half of the face, a Le Fort I osteotomy and a bilateral sagittal split osteotomy of the mandibular ramus, together with a genioplasty, are performed in one operating session and via intraoral incisions only (Figs. 14–8 and 14–9).[19–23]

Late results after rotation of the lower half of the face, with and without simultaneous reconstruction of the ramus, prove the value of the Zurich approach.[37]

The mobilized maxilla may be positioned relative to the as yet unmobilized mandible by means of a prefabricated acrylic occlusal splint constructed on a plaster cast model mounted in a semiadjustable articulator, as described for the preoperative preparation. Following osteosynthesis of the maxilla, the mandible is mobilized and relocated relative to the newly repositioned maxilla.

When the acrylic occlusal wafer is not used, the fully mobilized maxilla and mandible are fixed in the preoperatively planned occlusion by means of continuous-loop wire splinting and intermaxillary fixation.[18] The interposition of a thin acrylic occlusal wafer is necessary only when a stable interocclusal interdigitation proves difficult to establish exactly.

Additional superior repositioning of the maxilla on the unaffected side in order to correlate the maxillary incisor–lip line is achieved by removing the required amount of bone from the maxillary base along the osteotomy line on the unaffected side, or in some cases by merely impacting the maxilla superiorly on this side.[37] Resultant bony defects are bridged with transplanted autogeneic pelvic bone. Autogenous bone is also wedged into the retromaxillary space between the maxillary tuberosity and the pterygoid plates when the maxilla has been moved anteriorly.[22]

Osteosynthesis and bone graft fixation are achieved by means of direct bone wiring, supported in turn by a zygomatic buttress or lateral inferior orbital margin and piriform aperture skeletal suspension if possible. The Kufner suspension method may prove to be an adequate alternative.[12] A short screw is fixed in the forehead in the frontal sinus region, and attached suspension wires extend subcutaneously to the oral cavity in the canine area bilaterally. Skeletal suspension from a reconstructed zygomatic arch is obviously not advisable. An asymmetric or retropositioned chin may be simultaneously corrected by a one- or several-step sliding genioplasty or the propeller genioplasty.[17, 19, 20, 38]

Temporomandibular Joint Reconstruction

In patients in whom the syndrome of otomandibular dysostosis finds only rudimentary expression, the glenoid fossa and mandibular condyle may appear to be radiographically normal. Anterior translocation of the condyle is, however, either impossible or extremely limited, resulting in a distinct deviation of the mandible toward the affected side when the mouth is opened. Surgical mobilization of the joint followed by postoperative mouth-opening exercises gives good results provided the mandibular elevator musculature is normal.

An insufficiently developed glenoid fossa, inadequate for the support of a reconstructed ramus and condyle, may exist in conjunction with either lateral (horizontal) hypoplasia so mild as not to require correction or sufficiently pronounced to necessitate at least zygomatic and temporal bone augmentation.

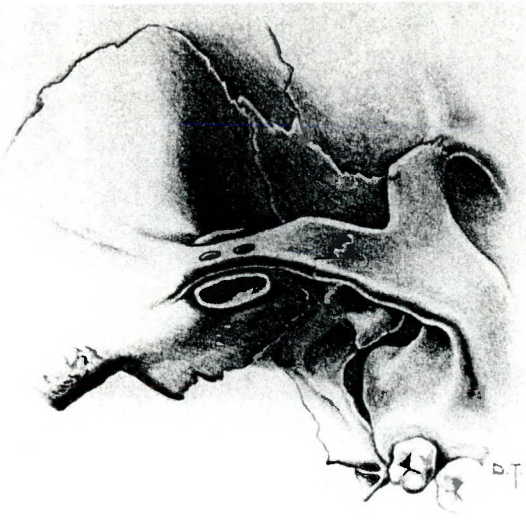
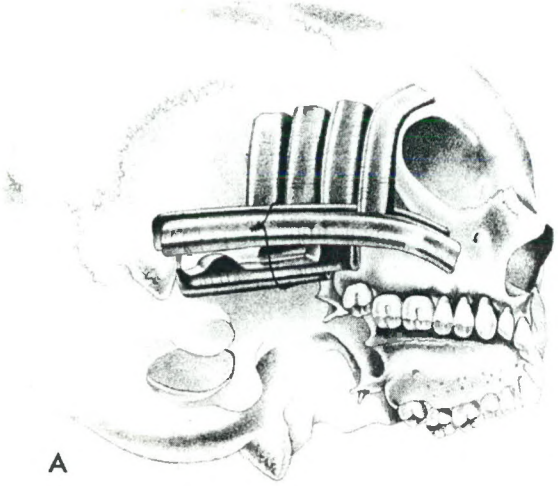
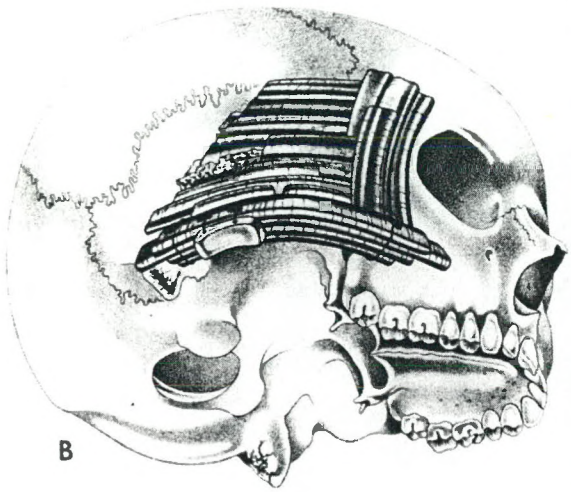


Figure 14–10. Reconstruction of the glenoid fossa in mild horizontal hypoplasia in the temporomandibular joint area. A piece of rib graft with attached cartilage is inserted between the temporal bone and zygomatic arch and held in place by bone spicules or wire.





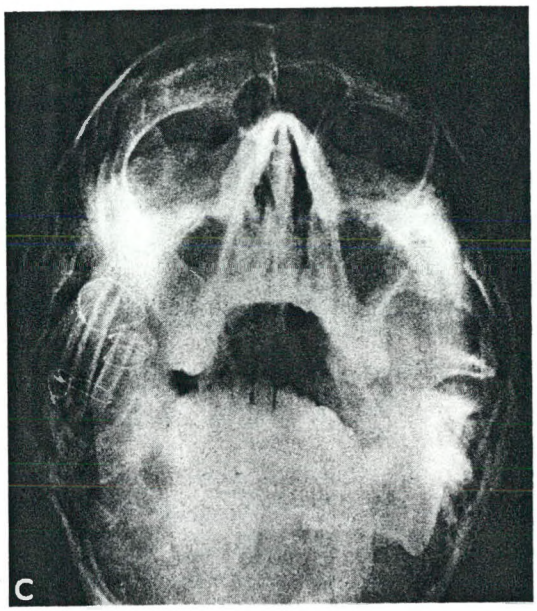


Figure 14–11. *A*, Reconstruction of the glenoid fossa, zygoma and zygomatic arch, temporal bone, and lateral orbital rim in moderately severe otomandibular dysostosis. *B*, Reconstruction in severe cases. *C*, Postoperative radiograph of reconstruction by the method depicted in *A*.

Reconstruction of the glenoid fossa without concomitant zygomatic and temporal bone augmentation must be accomplished prior to subsequent vertical and sagittal repositioning of the lower half of the face. This may be achieved by fixing a piece of rib graft with attached cartilage to the zygomatic arch, utilizing a tragus border incision with temporal extension (Fig. 14–10). At least three months should then elapse before the ramus is reconstructed. Unfortunately the fossa thus produced is usually positioned too far anteriorly and medially, resulting in asymmetric, medial displacement of the ramus and necessitating additional lateral correction with bone or cartilage onlays.

In cases of pronounced deformity, reconstruction of the glenoid fossa with concomitant zygomatic and temporal bone augmentation allows for a more desirable positioning of the fossa, and hence it is possible to construct a symmetric ramus. The zygomatic arch (and possibly the zygomatic bone and the temporal and mastoid area) is built up with layered, halved ribs correctly shaped, to which is secured a section of rib with attached cartilage carved to resemble a glenoid fossa (Figs. 14–11, 14–12, and 14–13).

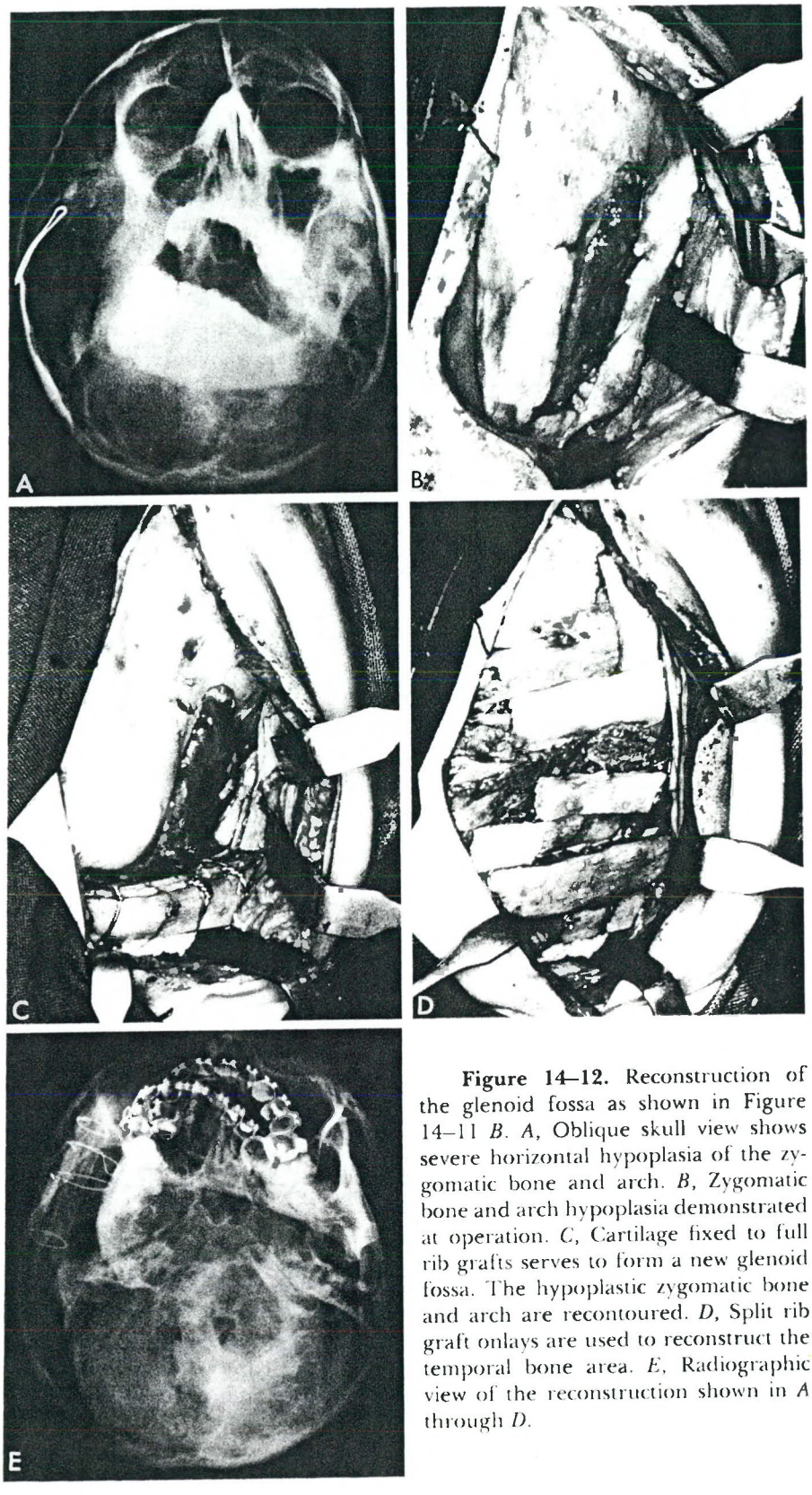


Figure 14–12. Reconstruction of the glenoid fossa as shown in Figure 14–11 *B*. *A*, Oblique skull view shows severe horizontal hypoplasia of the zygomatic bone and arch. *B*, Zygomatic bone and arch hypoplasia demonstrated at operation. *C*, Cartilage fixed to full rib grafts serves to form a new glenoid fossa. The hypoplastic zygomatic bone and arch are recontoured. *D*, Split rib graft onlays are used to reconstruct the temporal bone area. *E*, Radiographic view of the reconstruction shown in *A* through *D*.



Figure 14–13. Facial views of the patient before, *A*, and after, *B*, the reconstruction of the missing glenoid fossa and horizontal hypoplasia of the temporozygomatic area shown in Figure 14–12.

Reconstruction of the Mandibular Defect

Sagittal splitting of the affected mandibular ramus may prove difficult, depending on the degree of hypoplasia present. Splitting a hypoplastic ramus sagittally and lengthening it may result in loss of contact between the segments. When this occurs, a long thick segment of autogeneic pelvic bone or rib graft, lightly decorticated and appropriately shaped, should be inserted, spanning the gap in the ramus, attached to both segments and passing from the chin prominence to the reconstructed fossa superiorly (Figs. 14–14 A and B and 14–15).

An extremely hypoplastic ramus that precludes sagittal splitting is invariably accompanied by severe deviation of the chin prominence to the ipsilateral side. A

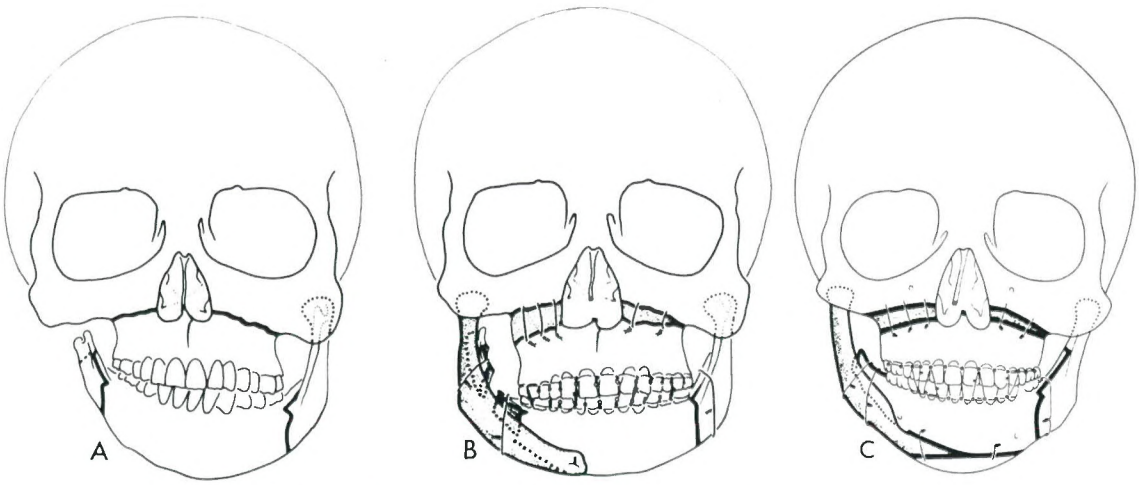


Figure 14–14. *A* and *B*, When the lower half of the facial skeleton is rotated downward and forward after sagittal splitting of the ramal stump, the bone graft may be onlayed to the mandible to reconstruct the missing portion. *C*, The missing mandibular part may be reconstructed by onlaying the bone graft to the mandible after splitting or totally freeing the ramal stump. The distal portion of the graft is inlayed between the mandibular body and the rotated chin prominence.



Figure 14–15. *A*, Radiographic view of the facial skeleton before reconstruction. *B*, Postoperative radiograph showing correction achieved by using the procedure shown in Figure 14–14 *A*. *C*, Preoperative appearance of the patient. *D*, Postoperative photograph of the patient following a free fat graft to the cheek and chin augmentation with lyophilized bank cartilage.

unilateral sagittal split osteotomy of the unaffected ramus and freeing of the ramal stump allow for mandibular repositioning. The deficient ramus and condyle are made good by means of bone grafting, as previously described, with the graft wedged anteriorly into the defect between the detached chin prominence and the body of the mandible (Figs. 14–14 C and 14–16 A, B, and C). Dead space between bone segments is filled with bone chips. It is essential that the ramal bone grafts be as thick and as long as possible, allowing for correction of both lateral hypoplasia and vertical length of the ramus as well as recontouring the mandible to bring its lower border more inferiorly on the affected side.

For reconstructing the ramus following earlier temporomandibular joint reconstruction, an extraoral incision placed in the old scar extending from the tragus border along the upper attachment of the ear and an intraoral vestibular incision over the external oblique ridge of the mandible suffice to expose the previously reconstructed and more laterally situated joint adequately as well as the augmented temporal bone

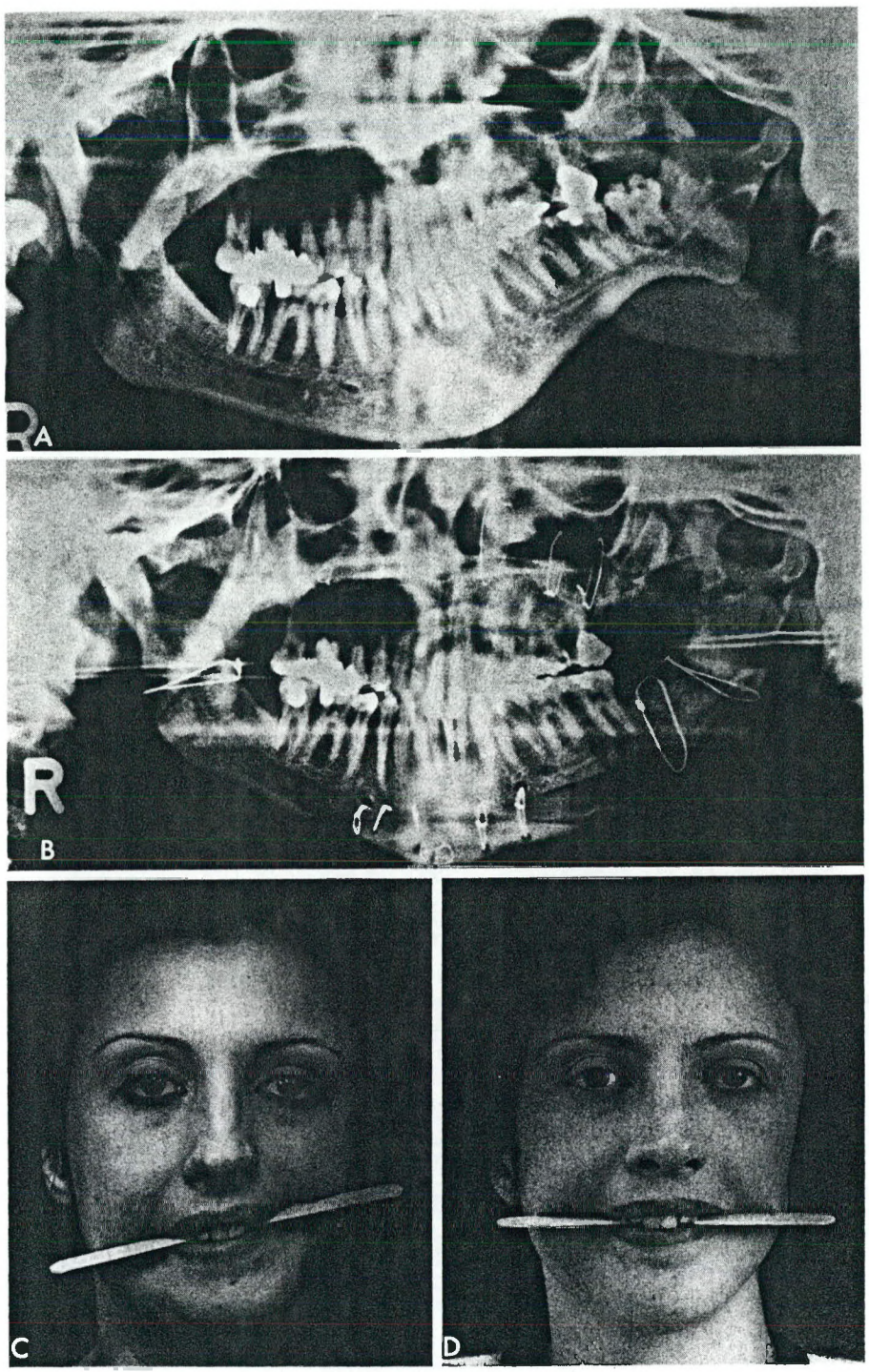


Figure 14–16. *A*, Preoperative orthopantomogram showing moderately severe left otomandibular dysostosis. *B*, Postoperative radiograph following reconstruction as depicted in Figure 14–14 *C*. *C*, The patient preoperatively. *D*, Postoperative photograph shows improved facial symmetry prior to free fat transplantation to the left cheek.

region and the affected ramus. Blunt dissection between the two incisions creates a soft-tissue pocket for the reception of the bone graft.

A lightly decorticated rib with attached costal cartilage may be shaped to extend from the cartilage-lined reconstructed joint to the affected ramus. The rib graft extends anteriorly into the gap between the repositioned chin segment and the mandibular body (see Fig. 14–6 B).

Understandably, in cases with such extreme mandibular hypoplasia, an appreciable amount of transplant bone is required to offset the deficit. Numerous long rib segments are thus imperative. Lightly decorticated whole and halved rib segments are closely adapted to each other and to the mandible with wire, and at this stage one may supplement previous augmentation of the temporal and zygomatic areas if necessary (see Fig. 14–6).

Wound closure must be total and tension-free. Extensive mobilization of the soft tissue and periosteum about the mandible ensures not only satisfactory wound closure, but also allows for some readjustment of the facial soft tissues upon the newly reconstructed facial bony framework, particularly in the chin region. Postoperative drainage of both the temporal and mandibular regions is recommended. Intermaxillary fixation is maintained for six weeks.

Intensive postoperative mouth-opening exercises with interocclusal appliances are essential to combat trismus induced by scarring of the soft tissues and masticatory musculature and to lengthen congenitally hypoplastic muscles. (See Chapters 1 and 2 for further details.)

Chin Correction

Prior to ramal splitting, the chin prominence is detached and moved anteriorly and to the normal side (see Figs. 14–14 C and 14–16 B). The defect thus created between the repositioned chin prominence and the hypoplastic mandibular body is filled by wedging the anterior end of the mandibular graft into the gap. Smaller defects are then filled with medullary bone chips. This may be accomplished at the same time as one- or two-step advancement genioplasty (see Figs. 14–5 A and B and 14–14 C).[17, 19, 20]

Following detachment of the chin prominence, the lower border, if its contour is unsymmetric, may be sectioned vertically. Bone is then removed medially from the larger of the two segments, making them of equal size. Apposition and fixation in the midline of these two segments produces a symmetric lower border contour (Fig. 14–17).

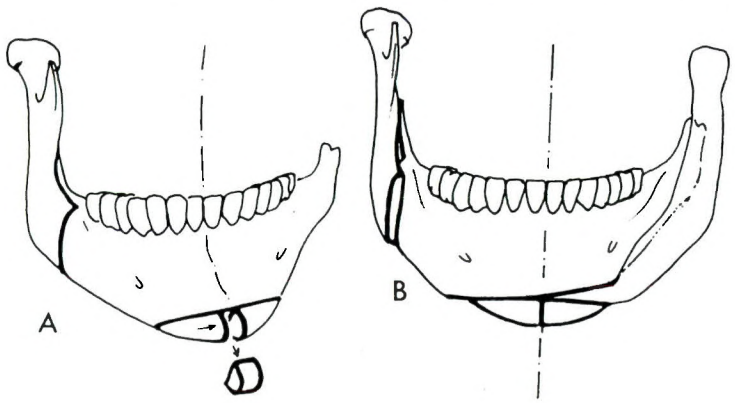


Figure 14–17. *A*, The chin prominence is detached and sectioned vertically, and a portion is removed from the medial end of the longer section. *B*, The remaining two chin segments, now of equal size and symmetric, are fixed together and rotated as a unit.

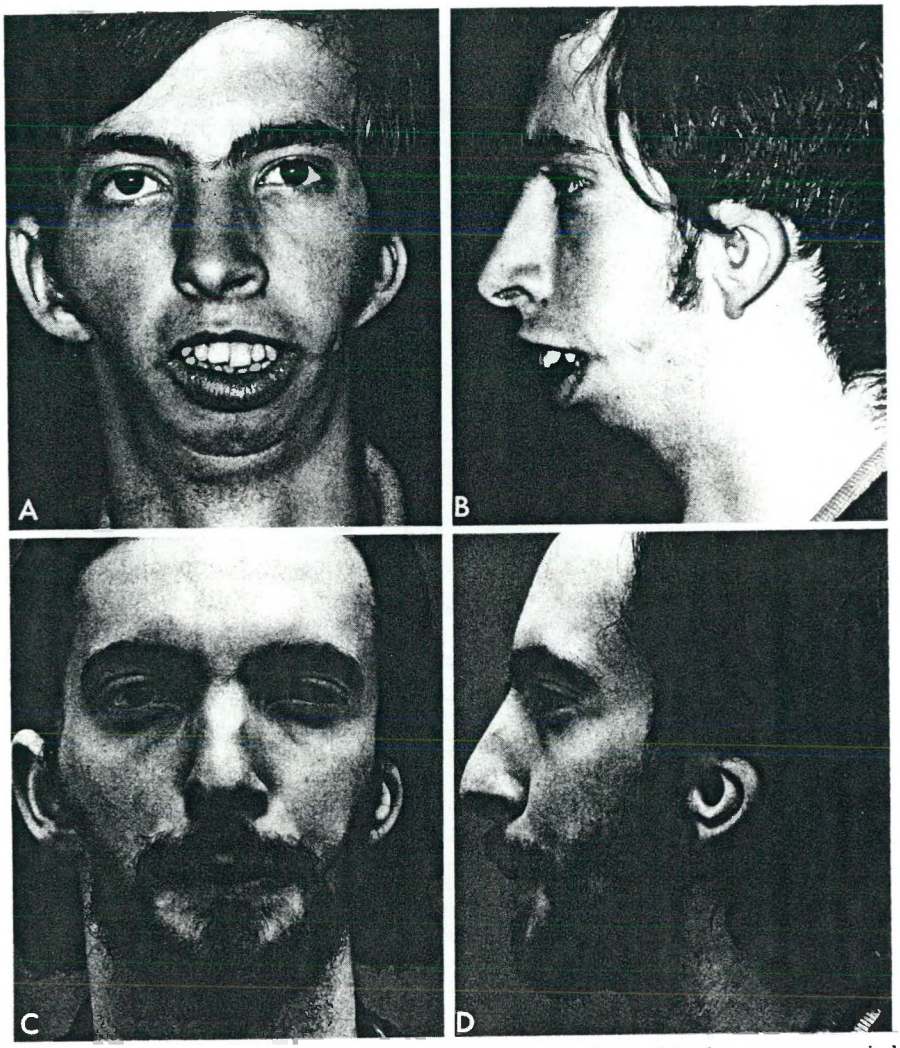


Figure 14–18. *A* and *B*, Preoperative appearance of a patient with almost symmetric bilateral otomandibular dysostosis. *C* and *D*, Appearance following retropositioning of the anterior maxillary segment, two-step chin augmentation, and multiple rib onlay grafts to the mandible.

An alternative method for correction of the asymmetric chin is the propeller genioplasty.[38] The chin prominence is detached symmetrically, and a second, cranially located, osteotomy is angled so that rotation of the muscle-pedicled bone segment through 180 degrees permits additional augmentation of the hypoplastic mandibular body as well as formation of a symmetric chin contour.

Bilateral Symmetric Otomandibular Dysostosis

Bilateral symmetric otomandibular dysostosis is rare. The lower third of the face is strikingly small but almost symmetric with pronounced distoclusion and severe chin retrusion (Fig. 14–18). The usual ear and mouth deformities are, in varying degrees, present bilaterally.

Vertical asymmetry of clinical significance is usually not encountered. Therefore,

rotation of the lower half of the face is not indicated. Contour correction by means of extensive onlays of bone and lyophilized cartilage, without extensive osteotomies, would be more appropriate.[36]

Distoclusion may be corrected by maxillary protrusion operation within the limits prescribed by good facial esthetics, and lateral (horizontal) sagittal and vertical mandibular augmentation may be accomplished by means of rib onlay grafts and a one- or several-step anteroinferior sliding genioplasty.

Bilateral Asymmetric Otomandibular Dysostosis

The bilateral asymmetric variant of otomandibular dysostosis is less rare than the symmetric form (Fig. 14–19). It presents a similar clinical picture except that the hypoplastic, retruded mandible deviates toward the side that has been more severely affected. In the case presented here there was considerable hypoplasia of the cheek on one side, less marked hypoplasia on the opposite side, bilateral macrostomia, extreme chin retrusion, and asymmetric distoclusion. A minor deviation of the mandible on mouth opening and virtual absence of forward movement of the condyle on either side were noted.

Radiographically the condylar head on the less affected side was hypoplastic, while contralaterally a spikelike termination of the mandibular body in the region of the angle was evident. The occlusal plane was practically horizontal and parallel with the pupils.

As in bilateral symmetric otomandibular dysostosis, the predominant malformation is sagittal and lateral (horizontal) rather than vertical. Rotation of the lower half of the face is thus not always indicated. In this case, initial temporomandibular joint reconstruction, followed later by unilateral ramal sagittal splitting and mobilization of the severely hypoplastic mandibular stump accompanied by a simultaneous three-step anteroinferior sliding genioplasty with extensive rib graft augmentation, as previously described, served to establish symmetric and esthetically harmonious facial skeletal

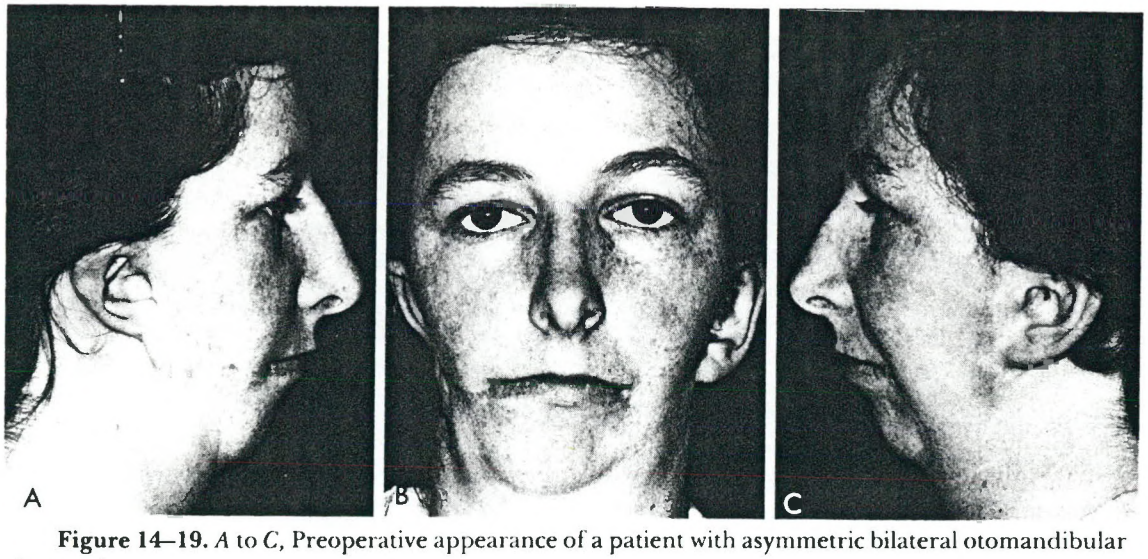


Figure 14–19. *A* to *C*, Preoperative appearance of a patient with asymmetric bilateral otomandibular dysostosis.

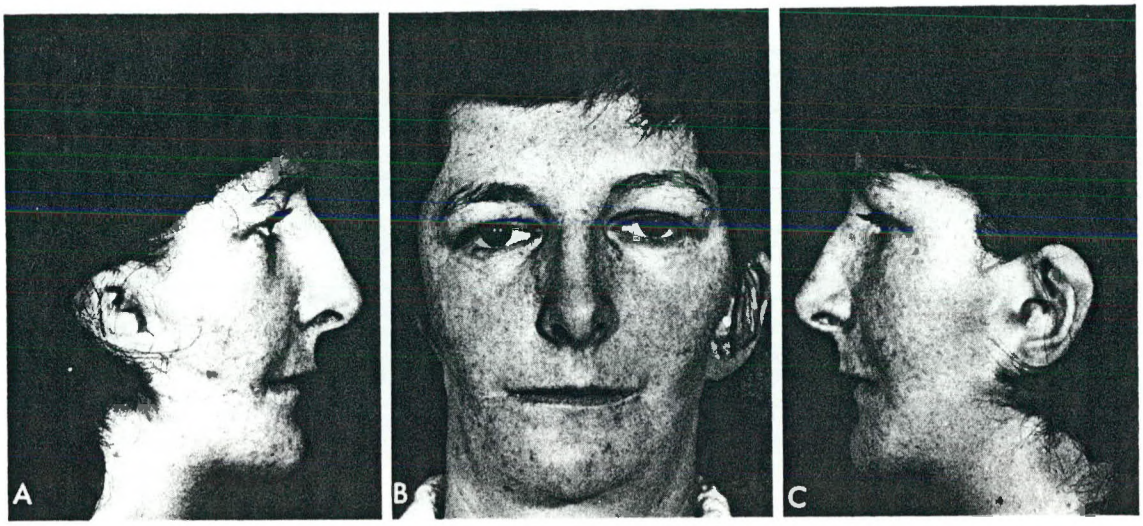


Figure 14–20. *A* to *C*, Postoperative views following three-stage reconstruction. Reconstruction of the missing glenoid fossa was followed by rotation advancement of the mandible with simultaneous reconstruction of the missing ramus and three-step chin advancement. Additional chin prominence rotation was still required.

proportions. Additional rhinoplasty, cheek contouring, and correction of macrostomia remain to be done (Fig. 14–20).

Soft-Tissue Correction

The correction of soft-tissue deformity, including ear and mouth abnormalities and cheek hypoplasia requiring free-fat grafting, should not be undertaken until all required skeletal corrective surgery has been satisfactorily accomplished (Fig. 14–21).

Procedures in Childhood

Onlays

Onlays of lyophilized bank cartilage, bank bone, or transplanted autogeneic bone may be utilized in childhood to camouflage minor deformities or partially correct more severe ones.[36] These procedures can minimize a socially unacceptable appearance at an age when the adverse opinion of a child's peers is likely to prove detrimental to stable psychologic maturation. They will not harm normal growth patterns and will not interfere with more extensive corrective procedures at a later time.

Soft-Tissue Correction

Kazanjian preferred to correct the soft tissues during childhood but was of the opinion that correction of the bone deformity should be delayed until after puberty.[11]



Figure 14–21. *A*, Preoperative view of a patient with mild right otomandibular dysostosis. *B*, Postoperative appearance following advancement genioplasty; lyophilized bank cartilage onlays to the body and ramus of the mandible and the right temporal, zygomatic, and infraorbital regions; retropositioning of anterior maxillary and mandibular segments; and a macrostomia operation. *C*, Free fat transplantation to the right cheek. *D*, Appearance following free fat transplantation.

Bone Correction

Both Hovell and, later, Osborne recommended that the hypoplastic mandibular ramus should be surgically lengthened in order to induce increased alveolar growth and tooth eruption to close the surgically created lateral open bite.[8, 30] Converse and associates proposed a vertical osteotomy of the hypoplastic ramus, shifting the mandible downward in order to level the bite.[2] Longacre believed that early and repeated bone augmentation is necessary to stimulate soft-tissue growth and correct contours.[13]

These procedures do not provide for the required correction of lateral and sagittal hypoplasia in the region of the mandible and the temporal–zygomatic bone complex. Notwithstanding the probable acceleration of growth, repeated osseous augmentation or additional osteotomies will almost certainly be required.[3, 28]

Soft-tissue repair and facial skeletal surgery should be delayed for as long as possible before puberty. Onlay bone or cartilage camouflaging procedures may, however, be carried out during childhood.

REFERENCES

1. Al-kayat A, Brambley P: A modified preauricular approach to the temporomandibular joint and malar arch. Br J Oral Surg *17*:91, 1979.
2. Converse JM, Horowitz SL, Coccaro PJ, Wood-Smith D: The corrective treatment of the skeletal asymmetry in hemifacial microsomia. Plast Reconstr Surg *52*:221, 1973.
3. Freihofer HP: Results after midface osteotomy. J Maxillofac Surg *1*:30, 1973.
4. Freihofer HP: Results of osteotomies in the facial skeleton in adolescence. J Maxillofac Surg *5*:276, 1977.
5. Glahn M, Winther JE: Metatarsal transplants as replacement for lost mandibular condyle (3 year follow-up). Scand J Plast Reconstr Surg *1*:97, 1967.
6. Gnoinsky WM: Die Morphologie der Dysostosis Otomandibularis. Inaugural Dissertation. University of Zürich, 1971.
7. Grabb WC: The first and second branchial arch syndrome. Plast Reconstr Surg *36*:485, 1965.
8. Hovell JH: The surgical treatment of some of the less common abnormalities of the facial skeleton. Dent Pract Dent Rec *10*:170, 1960.
9. Jackson IT, Munro IR, Salyer KE, Whitaker LA: Atlas of Craniomaxillofacial Surgery. St Louis: CV Mosby Co, 1982.
10. Jacobson A: Orthognathic diagnosis using the proportionate template. J Oral Surg *38*:820, 1980.
11. Kazanjian VH: Repair of congenital defects of the mandible. *In* Longacre JJ: Craniofacial Anomalies: Pathogenesis and Repair. Philadelphia: JB Lippincott Co, 1968.
12. Kufner J: A method of craniofacial suspension. J Oral Surg *28*:260, 1970.
13. Longacre JJ: The surgical management of first and second branchial arch syndromes. *In* Longacre JJ: Craniofacial Anomalies: Pathogenesis and Repair. Philadelphia: JB Lippincott Co, 1968.
14. Moss ML: The primary role of functional matrices in facial growth. Am J Orthod *55*:566, 1969.
15. Munro IR: Personal communication, Rome, 1982.
16. Murray JE: Personal communication, Rome, 1982.
17. Neuner O: Chirurgische Orthodontie. Schweiz Mschr Zahnheilk *75*:940, 1965.
18. Obwegeser HL: Ueber eine einfache Methode der freihändigen Drahtschienung von Kieferbrüchen. Osterr Z Stomatol *49*:652, 1952.
19. Obwegeser HL: The surgical correction of mandibular prognathism and retrognathia with consideration of genioplasty. Oral Surg *10*:677, 1957.
20. Obwegeser HL: Die Kinnvergrösserung. Osterr Z Stomatol *55*:365, 1958.
21. Obwegeser HL: Simultaneous resection and reconstruction of parts of mandible via the intraoral route. Oral Surg *21*:693, 1966.
22. Obwegeser HL: Surgical correction of small retrodisplaced maxillae. Plast Reconstr Surg *43*:361, 1969.
23. Obwegeser HL: Die einzeitige Vorbewegung des Oberkiefers und Rückbewegung des Unterkiefers zur Korrektur der extremen "Progenie." Schweiz Monat Zahnheilkd *80*:547, 1970.
24. Obwegeser HL: Zur Korrektur der Dysostosis otomandibularis. Schweiz Monats Zahnheilkd *80*:331, 1970.
25. Obwegeser HL: Deformities of the jaws. *In* JC Mustardé: Plastic Surgery in Infancy and Childhood. Edinburgh: Livingstone, 1971.
26. Obwegeser HL: Correction of the skeletal anomalies of otomandibular dysostosis. J Maxillofac Surg *2*:73, 1974.
27. Obwegeser HL: Proceedings of European Association of Maxillofacial Surgeons, Venice, 1978.
28. Obwegeser HL, Weber G, Freihofer HP, Sailer HF: Facial duplication—the unique case of Antonio. J Maxillofac Surg *6*:179, 1978.
29. Ortitz-Monasterio F: Personal communication, Rome, 1982.
30. Osborne R: The treatment of the underdeveloped ascending ramus. Br J Plast Surg *17*:376, 1964.
31. Pichler H: *In* Pichler H, Trauner R (eds): Mund und Kieferchirurgie, Band III. München: Urban & Schwarzenberg, 1948.
32. Poswillo DE: The pathogenesis of the first and second branchial arch syndrome. Oral Surg *35*:302, 1973.
33. Poswillo DE: Otomandibular deformity: Pathogenesis as a guide to reconstruction. J Maxillofac Surg *2*:64, 1974.
34. Robinson M: Congenital absence of the ramus of the mandible: Report of case. J Oral Surg *28*:302, 1970.
35. Rowe NL: Surgery of the temporomandibular joint. Proc R Soc Med *65*:383, 1972.
36. Sailer HF: Transplantation of Lyophilized Cartilage in Maxillofacial Surgery. Experimental Foundations and Clinical Success. Basel, New York: S Karger, 1983.
37. Sailer HF: Ergebnisse der Rotation der unteren Gesichtshälfte zur Korrektur der Dysostosis oto-mandibularis. Fortschr Kiefer Gesichtschir *26*, 1981.
38. Sailer HF: Propeller genioplasty—a new method for the correction of the asymmetric chin prominence. Transactions of Eighth International Conference on Oral Surgery, Berlin, 1983.
39. Ware WH: Growth center transplantation in temporomandibular joint surgery. Transactions of Third International Conference on Oral Surgery, London, 1970.