Converse: Chapter 54

Craniofacial Microsomia

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Among the congenital otocephalic syndromes, the term "first and second branchial arch syndrome" designates, in the United States, a characteristic congenital malformation which is usually unilateral but occasionally bilateral. In the German literature, the deformity has been termed "dysostosis otomandibularis". Caronni (1971) has coined the term "auriculo-branchiogenic dysplasia". Stark and Saunders (1962) referred to a similar clinical association of physical findings as the first branchial arch syndrome or the oral-mandibular-auricular syndrome.

The term "first and second branchial arch syndrome" is not entirely satisfactory in differentiating specific syndromes, for there are also other malformations derived from maldevelopment of these branchial arches. Gorlin and Pindborg (1964) reviewed the various names by which the condition has been described and advocated the term "hemifacial microsomia", which implies that the deformity is exclusively unilateral and spares the cranium. Pruzansky (1971) used the term "otocraniocephalic syndromes" to describe aberrations in the development of the first and second branchial arches.

At the Center for Craniofacial Anomalies of the Institute of Reconstructive Plastic Surgery of the New York University Medical Center, the term "hemicraniofacial microsomia" is preferred for the unilateral form, and the designation "bilateral craniofacial microsomia" is reserved for the bilateral type.

Differential Diagnosis

The deformity of craniofacial microsomia, whether unilateral or bilateral, is characterized by varying degrees of a regional hypoplasia affecting the temporomandibular and pterygo-mandibular complexes (skeletal and neuromuscular structures). The bilateral form must be distinguished from mandibulofacial dysostosis (Treacher Collins syndrome) (see chapter 55), which shows a well-defined genetic pattern (Franceschetti and Klein, 1949) with the deformity being transmitted in an irregular dominant fashion; the gene is unstable and has a weak degree of penetrance. The genetic component in hemicraniofacial microsomia is less apparent, and the syndrome may be considered to be genetically heterogeneous - ie, as having more than one cause responsible for the defect. Tessier (1976) has pointed out that craniofacial microsomia has features in common with mandibulofacial dysostosis (Treacher Collins syndrome), such as the temporozygomatic defect and orbital deformities. Specific characteristics of hemicraniofacial microsomia are the malformations of the ramus and facial paralysis due to the involvement of the temporal bone in the malformation. Tessier (1976) has described three types of clefts that are present in mandibulofacial dysostosis (see Chapter 46).

Cervical spine abnormalities have been noted in many of the affected patients. The Goldenhar syndrome (oculo-auriculo-vertebral dysplasia (1952)) shows similar characteristics with, in addition, one or more epibulbar dermoids; it may be considered as a variant of
craniofacial microsomia. According to Tessier (1976), a frontozygomatic cleft (No 8) is a characteristic of Goldenhar syndrome (see Chapter 46).

A jaw malformation similar to that seen in bilateral craniofacial microsomia may be observed in patients who have suffered postnatal trauma or infection which has affected the condylar cartilage and, as a result, mandibular growth. It has been suggested, both experimentally and clinically, by several authors that impaired growth of the condyle may result in an underdevelopment not only of the mandible but also of the craniofacial osseous complex on the affected side (Sarnat and Engel, 1951; Brodie, 1964; Sarnat and Laskin, 1964). It is, however, usually relatively easy to distinguish postnatal deformities from those resulting from prenatal (genetic or intrauterine environmental) factors.

In postnatal traumatic deformity, the deformity is restricted to the jaws and the auricle is spared; the soft tissues are not deficient, and there is no temporal bone deformity. The earlier the traumatic insult occurs, the more severe the deformity, and there is often associated temporomandibular joint ankylosis.

In contrast, in craniofacial microsomia the deformity is more widespread, with involvement of the temporal bone, middle ear, mastoid process, external ear, and base of the skull, and often soft tissue hypoplasia on the affected side.

Exceptions to these differentiating features exist. A jaw malformation characteristic of hemicraniofacial microsomia may be present from birth with minimal auricular or temporal bone development. The authors have observed two such cases of hemicraniofacial microsomia in which audiograms showed a normal level of hearing in both ears and no deformity of the temporal bone could be detected by roentgenography. These patients represent variations of hemicraniofacial microsomia rather than a separate syndrome (mandibular dysostosis, Nager and de Reynier, 1948).

Patients with severe orbital clefts (see Chapter 46) and hypoplasia of the maxilla also have an occlusal slant and a short mandibular ramus but the condylar deformity, which is the hallmark of craniofacial microsomia, is lacking. In craniofacial microsomia, the mandibular anomaly is a developmental deformity that is secondary to a hypoplasia of the temporal bone; the hypoplastic maxilla is secondary to the mandibular hypoplasia.

**History**

Grabb (1965) cited the teratological tablets written in approximately 2000 BC, by the Chaldeans of Mesopotamia as the earliest recordings of malformations of the first and second branchial arches in man. Bartholinus (1654) described a child with absence of the external auditory canal, and Lachmund (1688) reported a female with microtia and agenesis of the external auditory canal. Thompson (1845) drew attention to the fact that the clinical expression of the syndrome was in structures derived from the first and second branchial arches and the intervening cleft.

In recent times the publications of Kazanjian (1939), Altman (1951), Meurmann (1957), François (1961), Stark and Saunders (1962), Gorlin and Pindborg (1964), Grabb (1965), Pruzansky (1969), Longacre, DeStefano, and Holmstrand (1963), Obwegeser (1970,
1974), and Converse and associates (1973a, b, 1974) have focused attention on the clinical findings and the reconstruction of the individual deformities.

**Clinical Spectrum of the Syndrome**

**Incidence.** In a study of birth records at several hospitals in the United States, the birth incidence was determined to be 1 in 5642 births (Grabb, 1965). Poswillo (1973) cited an incidence of 1 in 4000. Following the administration of thalidomide to pregnant women in Germany between the years 1959 and 1962, 2000 less severe cases of first and second branchial arch malformations were reported (Kleinsasser and Schlothane, 1964).

**Sex.** In the series of 102 patients reported by Grabb (1965), 63 were males and 39 were females.

**Unilateral vs. Bilateral.** In a series of 74 patients, Meurmann (1957) reported eight patients with bilateral involvement; Dupertuis and Musgrave (1959) noted a unilateral to bilateral incidence of 6 to 1; Grabb (1965) cited bilateral involvement in 12 of 102 patients; Converse, Wood-Smith, McCarthy, Coccaro, and Becker (1974) studied 15 patients with bilateral craniofacial microsomia out of a total series of 280 patients. The pathologic findings and treatment of bilateral craniofacial microsomia are discussed in a subsequent section of the chapter.

**Variations of the Syndrome.** The deformity in craniofacial microsomia varies in extent and degree. As in many other craniofacial anomalies, it is often difficult to classify the individual deformity.

In the severe form, all of the structures derived from the first and second branchial arches are hypoplastic, while in other types, either the auricular or jaw dysplasias may be predominant.

It is in the latter cases that the associated deformities are less evident. There may be many shades of expression, according to the degree of involvement of the structures derived from the first and second arches and the involvement of the adjacent skeletal structures:

1. The ear deformity may be maximal, while the jaw deformity is not apparent on clinical examination. Roentgenographic studies, including tomography, demonstrate, however, that all cases of external auditory canal and auricular hypoplasia with middle ear deformity have mandibular changes on the affected side.

2. The ear deformity may be less severe, and the jaw deformity may not be evident; roentgenographic analysis may show a disparity of the skeletal structures. In these cases of minor jaw deformities, careful clinical examination will often show a slight deviation of the mandible to the affected side. The midsagittal plane extends through the interspace between the upper central incisors (midincisor point), but the lower midincisor point may be off center in relation to it.

3. The characteristic jaw deformity of hemicraniofacial microsomia may be present without gross auricular or temporal bone maldevelopment. These cases are difficult to
differentiate from postnatal deformities caused by injury, but the diagnosis becomes evident if the deformity was present at birth. This type of deformity may also be complicated by associated malformations such microphthalmos. The auricle may be normal in shape, protruding, or low-set, with a normal hearing mechanism.

4. "Formes frustes" or microforms are more frequent than is generally acknowledged. They must be searched for in cases of slight facial asymmetry and in auricular malformations without manifest jaw deformity. The patients shown is an example of a "forme fruste". The deformity is characterized by a soft tissue deficiency in the right parotid-masseteric area (the parotid gland was hypoplastic), a protruding right auricle (with normal hearing) and a slight degree of macrostomia involving the right oral commissure. The dental occlusal relationships were adequate. Correction was achieved by insertion of a dermis-fat graft that restored adequate facial contour, a protruding ear operation according to the technique of Converse and associates (1955, 1963), and closure of the macrostomia.

Grabb (1965) proposed a classification of six groups according to the specific anatomical abnormalities but emphasized the overlapping among the individual groups.

Embryology

The ear can be taken as a frame of reference in the otocraniocephalic syndromes (Pruzansky, 1971) because of its developmental relationship with the jaw (Table 54-1). A brief review of the phylogeny and ontogeny of the auricle and hearing apparatus is helpful in understanding the mechanism of the malformation in the craniofacial microsomia syndrome.

The two principal divisions of the organ of hearing come from different embryonic anlagen. The sensory end organ in the inner ear is derived from the ectodermal otocyst, while the sound-conducting apparatus in the external and middle ear comes from the gill structures.

The membranous labyrinth has its beginning in the 3.5 week old human embryo (Arey, 1946) as a thickening of the ectoderm on the side of the head - the otic placode (see Chapter 35). This area is enfolded to become the otic pit, then pinches off to become the otocyst. By means of a series of folds, the otocyst differentiates in the 3 month old fetus into the endolymphatic duct and sac, the semicircular endolymphatic ducts, the utricle, the saccule, and the cochlear duct, which contains the organ of Corti. By the fifth month of fetal life, the sensory end organ of the ear attains adult form and size, as the cartilaginous otic capsule ossifies.

It is speculated that our piscine ancestors swam in seas not yet as salty as today's oceans and that man's endolymph, entrapped by the enfolding otocyst, closely resembles in chemical composition the dilute salt water of that primeval sea. Our ancient aquatic forebears did not require any special mechanism to transmit sound to the inner ear. As in today's fish, sound was readily transmitted from the sea through the skin to the fluid of the inner ear.

However, when these enterprising ancestors struggled out of the seas onto dry land, a new problem appeared. A mechanical device was needed to convert air vibrations of large amplitude and small force into fluid vibrations of small amplitude and large force. The gill
structures, no longer needed for breathing, became converted into such a mechanism. The first branchial groove became the external auditory meatus and canal, while the first pharyngeal pouch became the eustachian tube and middle ear. Instead of the branchial groove and pharyngeal pouch connecting to become a gill cleft, a thin intervening layer of tissue remained to form the tympanic membrane.

The mandible, incus, and malleus developed from the cartilage of the first branchial arch (Meckel's cartilage), while the stapes (with the exception of the footplate, which originates from the otic capsule), styloid process, and hyoid bone developed from the cartilage of the second arch (Reichert's cartilage) (See Table 54-1). The large area of the tympanic membrane, connected by the lever system of the ossicular chain to the small area of the oval window, provided the ear with an effective mechanism to overcome the sound barrier between air and water.

Table 54-1. Structures Derived From the First and Second Branchial Arches and the Otic Capsule

<table>
<thead>
<tr>
<th>First branchial arch</th>
<th>First branchial groove</th>
<th>First pharyngeal pouch</th>
<th>Second branchial arch</th>
<th>Otic capsule</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maxillary process</td>
<td>Maxilla</td>
<td>Tympanic membrane</td>
<td>Facial nerve</td>
<td>Vestibular surface of stapes, internal acoustic meatus</td>
</tr>
<tr>
<td>Mandibular process</td>
<td>Palatine bone</td>
<td>Eustachian tube</td>
<td>Posterior part of auricle</td>
<td>Inner ear.</td>
</tr>
<tr>
<td></td>
<td>Zygoma</td>
<td>Middle ear cavity</td>
<td>Manubrium of malleus, long process of incus, stapedial superstructure, and tympanic surface</td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Stapedial artery, styloid process, stylohyoid ligament</td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
<td>Lesser cornu of hyoid</td>
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</tbody>
</table>

The gill structures destined to form the sound-conducting apparatus and the jaws first appear in the human embryo at 4 weeks, at about the same time as the otic pit.

By the third fetal month, the pinna has been formed from the first and second branchial arches on either side of the first branchial groove; the latter is the primary shallow,
funnel-shaped external auditory meatus. From the inner end of the primary meatus, a solid cord of ectodermal cells extends further inward, with a bulblike enlargement adjacent to the middle ear. It is not until the seventh fetal month that this cord canalizes, beginning medially to form the tympanic membrane and then extending laterally to join with the primary meatus to form the completed external auditory meatus. The external and middle ear, although capable of transmitting sound to the inner ear, are not yet of adult form and size.

In the seventh fetal month, pneumatization of the temporal bone begins, with excavations lined by mucous membrane extending out from the middle ear cavity while the jellylike mesodermal tissue in the middle ear cavity begins to resolve. At birth the eustachian tube inflates; the fetal mesodermal tissue in the middle ear and antrum continues to resorb until the epithelium lies close to the periosteum, and pneumatization of the temporal bone proceeds.

The external auditory meatus, entirely cartilaginous at birth (except for the narrow incomplete ring of the tympanic bone), deepens by growth of the tympanic bone to form the adult osseous meatus. Except for some pneumatization of the petrous apex that may continue into adult life, the external and middle ear finally reach adult form and size in late childhood (in contrast to the inner ear, which becomes adult in fetal life). It is generally accepted that the first branchial arch furnishes the anterior part of the auricle (see Table 54-1); the second arch provides the structures of the remaining external ear (Wood-Jones and Wen-I-Chuan, 1934). The maxilla, palatine bone, and zygoma develop from the maxillary process of the first branchial arch, the mandible from the mandibular process.

Meckel's cartilage, the primary jaw of lower vertebrates, represents the temporary skeleton of the first pharyngeal arch; the two symmetrical cartilaginous bars in early fetal life describe a parabolic arch that serves as a model and guide in the early morphogenesis of the mandible.

Three main regions of Meckel's cartilage should be considered: (1) the distal portion, which becomes incorporated into the anterior part of the body of the mandible; (2) a middle portion, which gives rise to the sphenomandibular ligament and contributes to the mylohyoid groove of the mandible; (3) the proximal or intratympanic portion, which differentiates into the malleus, the incus, and the anterior malleolar ligament.

Embryology of the face is discussed in more detail in Chapter 53.

Etiopathogenesis

Unlike mandibulofacial dysostosis, the genetic component in craniofacial microsomia is ill-defined, and there is no evidence of genetic transmission except in a few patients. Grabb (1965) reported that only 4 of 102 patients studied had one sibling or one parent with maldevelopment of the first and second branchial arch syndrome. Hanhart (1949) described a form of inheritable auricular hypoplasia, and Rogers (1968) listed several family studies by other authors which suggested a possible hereditary basis for auricular deformities. Summitt (1969) reported a pedigree of the Goldenhar syndrome which was compatible with autosomal dominant transmission.
A thorough search for minor deformities (such as preauricular tags or a receding chin) in other members of the family may provide a clue that the affected genes are not absent in the patient. As Fisher (1961) has stated, "A high index of suspicion based primarily on a careful family and maternal history, and understanding of the anomaly's causative factors and keenness of observation are important in identifying congenital abnormalities - many of which can be detected only after one has searched for them."

Consequently current etiopathogenic theories favor an intrauterine factor (or factors) which affects the embryo and has the following three characteristics:

1. The factor varies in its intensity and penetrance.

2. The factor strikes at varying periods in the course of prenatal development - from the first to the seventh month.

3. The damage is produced in varying loci in the fetus or embryo, along any point of the developing first and second branchial arches; the damage may be localized (segmental) or widespread.

The etiopathogenesis has not been adequately explained to date. The theory of mesodermal deficiency of Hoffstetter and Veau has been involved by Stark and Saunders (1962).

Others have suggested that vascular defects of the stapedial artery may account for maldevelopment of the first and second branchial arches (McKenzie and Craig, 1955). The stapedial artery, a temporary vascular supply for the primordia of the first and second branchial arches, appears as a collateral of the hyoid artery and anastomoses with the pharyngeal artery; it is ultimately replaced by the finite external carotid system. Willie-Jorgensen (1962) felt that abnormalities of the latter artery were responsible for anomalies of the first and second branchial arches. A decreased heat pattern over the region of the external maxillary artery has also been demonstrated and is interpreted as evidence of a vascular deficiency (Ide, Miller, and Wollschlaeger, 1970).

Poswillo (1973) has produced phenocopies of craniofacial microsomia following the administration of triazene to the mouse and thalidomide to the monkey. Embryonic hematoma formation with spreading hemorrhage prior to the formation of the stapedial artery was demonstrated. The extent and size of the hematoma correlated with the size of the anomalous defect. In the mouse experimental model, all of the variations found in the human syndrome could be reproduced. The spectrum of defects was broad: a small aural hematoma producing a residual deformity of only the external ear and auditory ossicles; and larger hemorrhagic lesions affecting the condyle, mandibular ramus, and zygoma.

As mentioned earlier, Kleinsasser and Schlothane (1964) reported a large number of newborns with first and second branchial anomalies following the widespread use of thalidomide during pregnancy.
Pathology

The deformity in hemicraniofacial microsomia usually has the three major features of auricular, mandibular, and maxillary hypoplasia. The hypoplasia, however, also involves adjacent anatomical structures: the zygoma, the pterygoid processes of the sphenoid bone, the temporal bone (the middle ear and less frequently the internal ear; the mastoid process is small, and on the roentgenogram is seen to be acellular), the facial nerve, the facial musculature of expression, the muscles of mastication, the parotid, the cutaneous and subcutaneous tissues, and the tongue, soft palate, pharynx, and floor of the nose.

While the jaw and ear deformities are the most conspicuous in the majority of patients, the development of the first and second branchial arches and the structures derived therefrom are intimately interlinked with those of the chondrocranium and membranous bones of the skull; associated deformities of the temporal bone and other cranial bones are inevitable. In extreme forms of the dysplasia, extensive hemicraniofacial involvement is evident. As Pruzansky (1971) has stated, maldevelopment in one area may trigger a "domino effect", with involvement of the entire craniofacial bone community.

In addition to the skeletal deficiency, varying amounts of soft tissue hypoplasia, microphthalmos, anophthalmos, cranial nerve palsy, cleft lip and palate, and lateral facial clefts are frequent accompanying deformities.

**The Jaw Deformity**

The most conspicuous deformity of hemifacial microsomia is the hypoplasia of the mandible on the affected side. The ramus is short or virtually absent, and the body of the mandible curves upward to join the short ramus. The chin is deviated to the affected side; the body of the mandible on the unaffected side does not follow its usual curvature but assumes a flattened contour with a straightened gonial angle.

Ramus and condyle malformations vary from minimal hypoplasia of the condyle to its complete absence in association with hypoplasia or agenesis of the ramus. In all patients, condylar anomalies can be demonstrated, and this finding may represent the hallmark of the syndrome. As a consequence, the spatial relationships of the distorted, deformed, and/or deficient anatomical parts, as well as the neuromuscular components identified with these parts, become of paramount importance in the diagnosis and the planning of treatment.

Attempts at classification of the mandibular anomaly have been made (Pruzansky, 1971):

**Grade I**: Hypoplasia is minimal or slight.

**Grade II**: The condyle and ramus are small; the head of the condyle is flattened; the glenoid fossa is absent; the condyle is hinged on a flat, often convex, infratemporal surface; the coronoid process may be absent.

**Grade III**: The ramus is reduced to a thin lamina of bone, or it is completely absent.
The posterior wall of the glenoid fossa is partially formed by the tympanic portion of the temporal bone which provides the bony portion of the external auditory canal in the normally developed ear. When there is hypoplasia of the temporal bone, the posterior wall of the glenoid fossa of the temporomandibular joint may be absent; on occasion, a distinct fossa cannot be identified. The infratemporal surface is flat, and the hypoplastic ramus is often hinged on this flat surface at a point anterior to the contralateral unaffected temporomandibular joint.

Mandibular growth deficiency is usually closely related to the degree of hypoplasia of the condyle. In the more severe conditions, there is considerable disparity in condylar growth between the affected and the unaffected sides.

Facial asymmetry increases progressively during the formative years as the growth disparity between the affected and the unaffected side causes the mandible to deviate laterally and upward toward the affected side. The cant of the occlusal plane (higher on the affected side, lower on the unaffected side) is caused by the short hypoplastic ramus and by hypoplasia of the zygoma, maxilla, and its dentoalveolar process on the affected side. The downward growth of the maxilla on the affected side is restricted by the short ramus.

The skeletal asymmetry is clinically demonstrated by the high occlusal cant on the affected side. The floor of the maxillary sinus and of the nose on the affected side is at a higher level; in some patients, it was noted that the base of the skull was elevated on an inclined plane similar to the inclined occlusal plane.

Anteroposterior and superoinferior dentoalveolar and skeletal dimensions are shorter on the affected side. Development and eruption of the molar teeth appear to be latent. Crowded dentition, with a characteristic tilt of the anterior maxillary and mandibular teeth toward the affected side, is often noted.

With growth in the width of the bimastoid and bicondylar areas, it is noted that the associated lateral positioning of the condyle and ramus on the affected side fails to keep pace with the movement laterally of the temporal bone and the glenoid fossa. The medial position of the condyle and ramus often places the body and dentoalveolar arch of the mandible in a position lingual to the maxillary arch, with a resulting crossbite.

Other Skeletal Deformities

Craniofacial bones other than the mandible or maxilla can be involved, especially the tympanic and mastoid portions of the temporal bone; the petrous portion is usually remarkably spared. The styloid process is frequently smaller and shorter on the affected side. The mastoid process can have a flattened appearance in the postauricular region, and there can be partial or complete lack of pneumatization of the mastoid air cells. The petrous portion, which houses the inner ear, might escape underdevelopment because of the protection against spreading hematoma afforded by the precartilaginous otic capsule (Poswillo, 1973).

The zygoma can be underdeveloped in all of its dimensions, with flattening of the malar eminence. A decrease in the span of the zygomatic arch results in a decrease in the length of the canthal-tragal line on the affected side.
Grabb (1965) reported that 11 of the 102 patients in his series had malformations of the vertebrae (hemivertebrae, fused vertebrae, spina bifida, scoliosis) and/or ribs.

**Muscles of Mastication**

There is a tendency to consider the deformities of craniofacial microsomia as being only osseous. However, there is an associated muscular hypoplasia which involves the muscles of mastication - the masseter, medial and lateral pterygoid, and temporalis - with a strong secondary influence on skeletal development (Wolff's law, 1892). It has been shown that resection of the temporalis muscle in the young animal modifies the architecture of the mandible (Horowitz and Shapiro, 1951).

Muscle function, especially that of the lateral pterygoid muscle, is impaired in many of these patients. The right muscle is responsible for the lateral movement of the mandible to the left, while the left muscle controls movement to the right. Both sides act synergistically in executing protrusive opening movements. In patients with hemicraniofacial microsomia, a severe limitation of protrusive and lateral movements due to hypoplasia of the lateral pterygoid muscle is observed.

The impact that this factor has not only on the developing musculature but also on the morphologic character of the attached bones is apparent. An alteration in mandibular movements (opening, lateral, and protrusive) comparable to the degree of anomaly of the condyle, ramus, and body is frequently observed.

When the patient opens his mouth, the deviation toward the affected side is produced not only by the skeletal asymmetry but also by the minimal or absent contribution of the ipsilateral medial and lateral pterygoid muscles in countering the opposing actions of the muscles on the unaffected side. The condyle on the unaffected side is displaced abnormally downward and laterally when the mandible is depressed, almost to the point of dislocating the condyle. No discernible condylar movement can be elicited on the affected side during opening and protrusive movements of the mandible. Thus, in testing for lateral pterygoid muscle weakness, one finds an absence of ability to shift the jaw laterally toward the unaffected side and deviation of the midline of the chin toward the affected side during opening and during forceful protrusion.

When there is condylar hypoplasia or agenesis, the lateral pterygoid muscle on that side is often totally absent. In many cases, the coronoid process is not present, and there is no evidence of a temporalis muscle tendon.

**Ear Deformity**

Auricular malformations are a usual manifestation of the syndrome. Meurmann (1957) proposed a classification of the auricular anomalies based on the studies of Marx (1926): *Grade I*, distinctly smaller malformed auricles with most of the characteristic components; *Grade II*, vertical remnant of cartilage and skin with a small anterior hook and complete atresia of the canal; *Grade III*, auricle almost entirely absent except for only a small part, such as a deformed lobule.
In a comprehensive study, Pruzansky (1971), using air and bone conduction audiometry and temporal bone tomography, evaluated 57 patients with craniofacial microsomia. It was observed that the degree of auricular deformity as classified above does not correlate exactly with hearing function. The type of hearing loss, although usually assumed to be conductive in origin, can be determined only by audiometry. Tomography, not auricular morphology, is the only indicator of middle ear structure. In addition, the unaffected ear may also harbor abnormalities in structure and function and should be evaluated. There was, however, a direct relationship between the degree of severity of auricular malformation and ipsilateral mandibular deformity. Reconstruction of the auricle and middle ear in patients with craniofacial microsomia is discussed in Chapter 35.

Deformities of the Nervous System

Nervous system abnormalities in patients with craniofacial microsomia have received little attention in the medical literature (Gorlin, Jue, Jacobsen and Goldschmidt, 1963).

Patients in the series at the Institute of Reconstructive Plastic Surgery have been investigated by complete neurologic examination, skull radiography, computerized axial tomography, and electromyography (Aleksic and coworkers, 1975a, b, c, d).

Cerebral Anomalies. A wide variety of cerebral anomalies exists in craniofacial microsomia and may include ipsilateral cerebral hypoplasia (Aleksic and associates, 1975a), hypoplasia of the corpus callosum (Timm, 1960), hydrocephalus of the communicating (Timm, 1960; Aleksic and associates, 1975c) and obstructive (Herrmann and Optiz, 1969) types, intracranial lipoma (Gaupp and Jantz, 1942; Aleksic and associates, 1975c), and unilateral hypoplasia of the brain stem and cerebellum (Mathies, 1966; Aleksic and associates, 1975c). Other associated abnormalities include mental retardation (Gorlin and coworkers, 1963), epilepsy, and electroencephalographic findings suggestive of epilepsy (Franceschetti, Klein and Brocher, 1959; Christiaens, Walbaum, Farriaux and Fontain, 1966; Aleksic and associates, 1975c).

Cranial Nerve Anomalies. Cranial nerve abnormalities are frequent in hemicraniofacial microsomia and can range from arhinencephaly of the bilateral (Virchow, 1864; Timm, 1960) and unilateral (Aleksic and associates, 1975a) types to unilateral agenesis and hypoplasia of the optic nerve with secondary changes in the lateral geniculate body and visual cortex (Aleksic and associates, 1975d), congenital ophthalmoplegia and Duane's retraction syndrome (Bowen, Collum and Rees, 1971), hypoplasia of the trochlear and abducens nuclei and nerves (Aleksic and associates, 1976), congenital trigeminal anesthesia (Sugar, 1967), and aplasia of the trigeminal nerve and motor and sensory nucleus (Aleksic and associates, 1975b). The most common cranial nerve anomaly is facial paralysis secondary to agenesis of the facial muscles, aberrant pathway of the facial nerve in the temporal bone (Bellucci, 1972; Sekhar and associates, 1975), or hypoplasia of the intracranial portion of the facial nerve and facial nucleus in the brain stem (Aleksic and associates, 1975c). Congenital hearing loss may be due to a malformed inner ear (Aleksic and Sekhar, 1976), hypoplasia of the cochlear nerve and brain stem auditory nuclei (Aleksic and associates, 1975c), or hypoplasia and impaired function of the ninth through the twelfth cranial nerves (Mathies, 1966; Berkman and Feingold, 1968). In conclusion, any cranial nerve can be clinically involved in patients with craniofacial microsomia, and it is likely that hypoplasia or agenesis...
of a portion of the entire cranial nerve trunk and corresponding brain stem nuclei represents the pathoanatomic substrate of the clinical dysfunction.

Electromyographic abnormalities have been briefly described in the literature (Grabb, 1965). Several of the authors' patients showed diminished or low-normal motor conduction velocity of the facial nerves, usually unilateral, while electromyographic abnormalities ranged from absence of the muscle to long polyphasic potential with incomplete interference pattern on active innervation (Aleksic and coworkers, 1975c). No cases of fibrillation potentials have yet been documented. The interpretation of the electrodiagnostic data has been somewhat hampered by the fact that many patients have undergone earlier surgical procedures in the region of the mandible and auricle.

Soft Tissue Deformities

In addition to the anomalies of the muscles of mastication and nervous system, there is often a generalized soft tissue hypoplasia which involves the skin, the subcutaneous tissue, and the facial musculature of expression. The musculature of the soft palate and of the tongue is occasionally less developed on the affected side. The not infrequent hypoplasia or aplasia of the parotid gland, previously noted by Entin (1958), places the branches of the facial nerve in a superficial and vulnerable position. The latter finding has obvious surgical implications. The shortness of soft tissues on the affected side is made evident by the shorter distance between the mastoid process and the angle of the mouth or the lateral canthus of the eye. The skin and subcutaneous tissue also show varying degrees of atrophy, particularly in the parotid-masseteric and auriculomastoid areas.

In the series reported by Grabb (1965), 10 per cent of the patients had malformations of the eye and eyelids and/or palate.

Transverse facial clefting ranging from macrostomia to a full thickness defect of the cheek can be present (Converse and coworkers, 1974). The clefts probably result from failure of fusion of the maxillary and mandibular processes. In embryologic development, the lateral commissure of the oral fissure is initially situated at the point of bifurcation of the maxillary and mandibular processes. With fusion of the latter and development of the muscles of mastication, the original broad mouth is reduced in size. In addition, the parotid glands, originally located near the embryonic oral commissure grow laterally toward the developing ear, but the parotid duct papillae remain in their more medial position (Converse and coworkers, 1974).

Treatment

Corrective Jaw Surgery

Correction of the jaw deformity in craniofacial microsomia has been attempted either by expansion for contour restoration or by osteotomies to change the jaw asymmetry and crossbite.

Contour Restoration Operation. Early expansion of local contour to maintain the growth of the soft tissues on the affected side has been achieved by serial autogenous split
rib grafts (Longacre, De Stefano and Holmstrand, 1961; Longacre, 1968), by allografts of cartilage or bone (Stark and Saunders, 1962), and by preserved cartilage allografts and inorganic implants (Brown, Fryer and Ohlwiler, 1960).

Other techniques for improvement of contour in the adult include correcting the deficient mandible either with a transplant taken from the lower portion of the unaffected half of the mandible (Gorski and Tarczynska, 1969) or by bone graft onlays (Converse and Shapiro, 1954; Longacre, De Stefano and Holmstrand, 1961). The only long-term report of results obtained by these contour-restoring procedures is that of Longacre (1968), who reported maintenance of contour in craniofacial dysostoses following serial autogenous onlay bone grafts.

Such surgery, however, fails to improve dental occlusion or function and also fails to align the facial skeleton in a position conducive to subsequent growth.

**Osteotomies of the Jaws.** Correction of the jaw asymmetry and crossbite by osteotomies and bone grafts has been done in children and adults, and the timing of the surgery has been the subject of controversy.

Poswillo (1974) cautioned against reconstruction of the facial bones before age 12. He felt that the trauma associated with surgery could destroy the functional matrix (Moss, 1968) of part of the facial skeleton and profoundly interfere with subsequent facial growth.

Obwegeser (1974) also advocated postponement of definitive jaw surgery until growth of the facial skeleton has ceased, as he felt osteotomies performed during childhood interfered with growth of the facial skeleton.

Converse, Horowitz, Coccaro and Wood-Smith (1973b), however, recommended surgery during childhood with the following objectives: (1) improving the symmetry of the mandible by bilateral vertical osteotomies of the ramus during the period of mixed dentition; (2) providing for downgrowth of the maxilla by lengthening the shortened mandibular ramus; (3) restoring the adequate dental occlusion; and (4) expanding the facial skeleton at an early age to fill out the soft tissues on the affected side of the face.

**Osteotomies in the Adult.** Correction of the jaw asymmetry and of the crossbite by mandibular osteotomy and bone grafts has been performed in the adult (Limberg, 1928; Converse and Shapiro, 1952; Dingman and Grabb, 1963, 1964).

Obwegeser (1970) has obtained satisfactory results in adults by a combined Le Fort I osteotomy of the maxilla and a bilateral sagittal section of the mandibular ramus. This procedure cannot be employed in the patient during the period of mixed dentition, as the maxillary osteotomy would injure or destroy the unerupted teeth.

Obwegeser (1974) has advocated that in severe cases the temporomandibular joint should be initially reconstructed with a costochondral rib graft. Any associated hypoplasia of the temporal bone, zygomatic arch, and lateral orbit rim should also be corrected. At a subsequent operation, the lower half of the facial skeleton can be rotated inferiorly and
anteriorly by a combination of a Le Fort I osteotomy, a bilateral sagittal split osteotomy of the mandible, and a double-step horizontal advancement osteotomy.

**Osteotomies in Childhood.** A metatarsal head transplant, as a substitute for the missing growth center of the mandibular condyle, has been advocated (Glahn and Winther, 1967). Others who used this procedure earlier found little or no growth of the grafts and no particular improvement in joint function (Dingman and Grabb, 1964; Freeman, 1965).

Converse and Rushton (1941) operated on a 12 year old patient with the jaw deformity of craniofacial microsomia. A horizontal osteotomy of the ramus was performed above the inferior alveolar foramen, and iliac bone graft was inserted between the fragments after the placement of a bite-block to open the occlusion on the affected side.

A similar procedure was performed by Osborne (1964) in patients with craniofacial microsomia and in those with asymmetrical growth resulting from injury. Osborne advocated surgery before the age of 6 years in order to give the maxilla an opportunity to develop after the release of the upward pressure exerted on the affected side by the hypoplastic mandible. Longitudinal studies of two of Osborne's cases of congenital origin were done by Knowles (1966), who was able to document maxillary growth.

Delaire (1970) recommended even earlier intervention between the ages of 4 and 6 years. He elongated the shorter ramus with an inverted L osteotomy and inserted costal bone grafts into the gap formed at the horizontal branch of the L.

Converse and his associates (1973b) advocated a two-stage procedure during childhood to correct the mandibular asymmetry, to allow expansion of the constricted maxilla, and to prevent further contraction of the overlying soft tissue.

The **first stage**, usually done when the child is 8 or 9 years old and in the mixed dentition period when mandibular growth is less active, corrects the vertical and horizontal hypoplasia of the malformed half of the mandible. A bilateral vertical osteotomy through the ramus is performed, permitting a forward, lateral, and downward movement of the malformed mandibular body toward the unaffected side. The rotation and displacement result in a bony gap in the deficient ramus on the affected side, reflecting the amount of downward and medial displacement of the mandibular body. Interposition iliac bone grafts are placed in the defect and maintained in position by wedging them between the fragments and by securing them with interosseous stainless steel wires. The iliac bone grafts are removed according to the technique of Crockford and Converse (1972).

The **second stage** of the corrective treatment is undertaken during adolescence and completes the treatment. Although the first stage achieves satisfactory occlusal relationships between the maxillary and mandibular dentoalveolar arches, the mandible usually remains asymmetrical because of the disparity in shape of the two halves of the bone. The disproportionate growth between the affected and the unaffected halves of the mandible also causes varying degrees of asymmetry and malocclusion. It is during the second stage of the treatment that final symmetry, definitive occlusion, and mandibular contour are achieved.
Planning the Treatment. Prior to surgery, casts of the dentition are made, and an occlusal bite-block is fabricated which will be used to maintain the sectioned mandibular segment in the most desirable mediolateral and superoinferior position. The plaster cast ("model") of the mandibular teeth is placed in the position the mandible will occupy after the corrective surgical procedure. The occlusal plane is to be lowered on the affected side, and after the bilateral vertical osteotomy of the ramus, an interocclusal space (open bite) will be established on the affected side. The surgically created void permits growth of the mandibular and maxillary dentoalveolar process and facilitates the eruption of the permanent dentition. Moreover, it minimizes or eliminates the preexisting severe cant of the occlusal plane (resulting from condylar growth disparity).

The bite-block is fixed into position during the surgical procedure, and intermaxillary wire fixation is employed. In addition, the lengthening procedure provides spaces to realign the teeth by orthodontic means, thus eliminating the crowded condition of the dentition characteristic of hemicraniofacial microsomia.

The purpose of the vertical section through the unaffected ramus is to facilitate the rotation of the mandible without unduly disturbing the temporomandibular joint.

It is not unusual that on the affected side the ramus is slender and obliquely directed downward and forward; the coronoid process is usually absent. On occasion, the posterior aspect of the body rests on the infratemporal surface because the ramus so short. The latter finding is unusual, and in nearly all of the patients, the size of the affected ramus is sufficient for a vertical osteotomy.

If the tooth bud of the third molar sits in the projected path of the osteotomy, it should be extracted.

Operative Technique. Both mandibular rami are approached through submandibular incisions placed in one of the natural skin folds of the neck. The dissection is extended subcutaneously upward until the mandibular angle and the posterior portion of the mandibular body are exposed. Care is exercised to avoid injury to the marginal mandibular branch of the facial nerve.

On the unaffected side of the mandible, the insertions of the masseter muscle are raised subperiosteally from the lateral surface of the ramus, and the posterior border of the ramus is freed of its muscular and ligamentous insertions. The neck of the condyle, the sigmoid notch, and the coronoid process can be easily identified on the unaffected side. On the medial aspect of the ramus, the medial pterygoid muscle is raised from its area of insertion on the posterosuperior portion of the bone. A vertical osteotomy is performed, with the line of the osteotomy extending from the posterior portion of the sigmoid notch downward to a point anterior to the mandibular angle.

The purpose of the osteotomy through the unaffected ramus is to prevent a rotation of the condyle within the glenoid fossa when the mandible is shifted laterally - an essential requirement for correction of the deformity. The osteotomy through the unaffected side also permits bone grafting of the defect resulting from the osteotomy or overlapping the fragments as required.
A similar vertical osteotomy is done through the ramus on the defective side. The mandible is then rotated toward the unaffected side and the teeth are maintained in intermaxillary fixation according to the preoperative plan. After intermaxillary fixation has been established according to the correction occlusion, a bite-block is inserted to correct the cant of the occlusal plane. The gap between the fragments of the ramus on the deficient side is filled with bone grafts.

While split rib grafts have been used in the past, iliac bone is preferred and is removed from the area below the anterosuperior spine without disturbing the cartilaginous crest (see Chapter 13). Additional bone grafts for contour are usually added over the lateral portion of the ramus and along the lower border of the body and angle of the mandible.

Bone grafts are occasionally required on the contralateral side if advancement of the mandibular body results in a gap between the fragments of the unaffected ramus. In other cases it may be necessary to overlap the fragments of the unaffected ramus when considerable lateral rotation of the mandible is indicated.

The cutaneous wounds are closed in a two-layer fashion. Suction drains are not routinely employed.

Interval Between the First and Second Stages. After a period of 10 to 12 weeks, the bite-block is removed and a second occlusal guide plane is placed over the biting surfaces of the molar teeth on the affected side to serve two important functions: (1) to maintain the newly acquired mandibular position and to counter any muscular pull which tends to deviate the mandible toward the unaffected side, particularly during protrusion and elevation, thus enhancing muscular activity on the deficient side; (2) to establish a necessary interocclusal stop on the unaffected side, ie, to maintain the interocclusal space which was established on the affected side by the osteotomy.

During this interim period of growth and development, orthodontic bands and archwire appliances are placed on the molar and incisor teeth of the maxilla and mandible. They serve to correct incisor relationships, to maintain adequate arch length, and to permit management of the bicuspids as they emerge. Thus they facilitate the movement of the teeth to more effective levels of occlusion during the transition from deciduous to permanent teeth.

The orthodontic therapy between the first and second surgical stages has three objectives - guiding, functional, and corrective: guiding, to improve dental relationships and take advantage of maxillary and mandibular growth; functional, to enhance growth potential on the affected side and contribute toward maintaining the mandible in the midline; corrective, to alter intradental and interdental relationships as needed and to establish an interocclusal and muscular balance anteroposteriorly and superoinferiorly between the maxilla and mandible on both sides.

Second Stage. The second surgical stage is completed during adolescence and requires one or more operative procedures. Although the osteotomy of the first stage may have achieved a degree of symmetry, one cannot expect symmetrical growth of both halves of the mandible. The disparity of growth between the bone-grafted half and the unaffected half will require additional surgery to achieve adequate contour.
1. Onlay bone grafts may be placed over the defective side of the mandible or maxilla at any time during the period of growth, as demonstrated by Longacre (1968). Onlay bone grafts may also be needed to restore contour of the flattened "unaffected" half of the mandibular body.

2. A horizontal advancement osteotomy of the anteroinferior portion of the body of the mandible is usually indicated. In addition to anterior advancement, the segment of bone must also be shifted toward the unaffected side to place the mental symphysis in the midline. Despite late appositional growth over the symphysis of the mandible in males, an effort to expand the soft tissues in this area should be considered, and an earlier operation may be indicated. The horizontal advancement osteotomy may need to be combined with placement of iliac bone as onlay grafts over the deficient half of the mandible. Onlay bone grafts to restore contour may also be placed over the temporal bone and zygoma, as emphasized by Obwegeser (1974).

3. Because of the disparity in growth between the grafted half of the mandible and the unaffected half, elongation by a second bilateral osteotomy of the rami with bone grafting may be required.

A Le Fort I osteotomy, not feasible at the first stage because of the presence of tooth buds of the secondary dentition in the maxilla, becomes a practical surgical procedure during adolescence. The downward tilt of the maxilla on the defective side leaves a defect in the body of the maxilla which must be filled with bone grafts.

**Restoration of Soft Tissue Contour.** Soft tissue hypoplasia is a characteristic feature of the syndrome. The soft tissue hypoplasia is usually not as severe or diffuse as in hemifacial atrophy (Romberg's disease). It is usually most conspicuous in the parotid-masseteric and auriculo-mastoid areas. Hypoplasia of the facial musculature of expression or congenital facial paralysis caused by agenesis of the facial nerve results in a generalized deficit soft tissue contour. Improvement in the soft tissue deficiency has been obtained by the insertion of a tube flap in the preauricular area (Gillies and Millard, 1957), a de-epithelized flap (Brown and McDowell, 1958), or a dermis-fat graft introduced subcutaneously (Davis, 1968). Additional improvement in contour can be obtained by limited use of silicone fluid (see Chapter 15).

De-epithelized microvascular free flaps of dermis and fat have been employed by Fujino, Rytinzaburo and Sugimoto (1975) for correction of severe hemifacial atrophy using deltopectoral tissue, and by Wells and Edgerton (1977) using groin tissue based on the inferior epigastric vessels. The external maxillary vessels were anastomosed to the vessels of the transplanted tissue by microsurgical techniques.

Restoration of hearing and reconstruction of the microtic ear are discussed in Chapter 35.

**Longitudinal Studies.** Many of the patients who have undergone reconstruction by the above technique have been followed for up to 15 years at regular intervals.
Early Versus Late Reconstruction

Decisions for early surgery must be based upon the character of the anatomical deformity (the spatial relationship of the condylar process of the mandible to the temporal bone and the pterygoid process of the sphenoid bone) and the function activity of the muscles attached to these bones. Severe alterations of condyle-fossa relationships have a direct impact upon the length and direction of the lateral pterygoid muscle. The anatomical and functional activity of the lateral pterygoid muscle contributes significantly to condylar development and, in turn, to the growth of the mandible; with adequate muscular function, the mandible is well developed and reflects the musculature balance to produce facial symmetry. The severe deviations in facial asymmetry of the hemicraniofacial microsomnia patient appear to be directly related to the abnormal bony structures of the temporomandibular complex and to the imbalance of the functioning muscles attached to these bones.

Another important consideration for early surgery is the problem of soft tissue resistance to mandibular advancement. Deficient cutaneous tissues resisting the surgical projection of the mandible constitute an indication for early surgery.

Thus, four areas of analysis must be considered in any decision concerning early surgery:

1. the status of the bony structures that make up the pterygoid-mandibular complex;
2. the spatial relationships of these parts to the temporal and sphenoid bones;
3. the anatomical and functional characteristics of the muscles attached to these structures; and
4. the status of the soft tissues on the affected side compared with unaffected side.

It would follow that given adequate anatomical and functional skeletal structures within the pterygo-mandibular complex, acceptable mandibular growth might be achieved through oro-orthopedic treatment procedures, thus precluding early surgery (see Chapter 30). However, it would also follow that in the absence of adequate anatomical and functional skeletal pterygomandibular components, little or no mandibular growth can be expected through oro-orthopedic therapy and, thus, early surgery would be indicated.

It should be noted that in patients selected for early mandibular surgery, growth was observed and documented in longitudinal studies. Further studies have indicated that although notable improvement in facial asymmetry was demonstrated following osteotomies of the mandible, the desirable result was not stable. This finding emphasizes the fact that although the skeletal structures were physically repositioned, the procedures did not satisfy the adjustments needed in the functioning muscles and ligaments associated with the skeletal units. Following surgery, there still remained the muscular and ligamentous abnormalities that continued to produce the imbalance of function and development that is so closely associated with the varying degrees of facial asymmetry.
Longitudinal studies suggest that the amount of growth on the defective side is inversely proportional to the extent of the hypoplasia of the ramus.

The rationale for early surgery in severe cases where minimal growth may be expected is to obtain the beneficial effects of the altered relationships of the structures contributing to dentoalveolar growth within the maxilla and mandible on the affected side. The surgical lengthening of the ramus and the lowering of the body of the mandible on the affected side produce the necessary interocclusal space needed for the growth and development of the dentoalveolar processes. The surgery combined with the bite-block treatment procedure also assists in maintaining the mandible in the midline.

In patients in whom the disparity of growth between the affected and unaffected halves of the mandible becomes evident, a second operation during adolescence, similar to the first, will reestablish skeletal symmetry.

**Bilateral Craniofacial Microsomia**

There is little mention in the literature of the bilateral form of craniofacial microsomia. The relative incidence of the hemifacial and bilateral types was discussed earlier in the chapter.

**Variations of the Syndrome.** Converse, Wood-Smith, McCarthy, Coccaro, and Becker (1974), in a review of 15 patients with bilateral craniofacial microsomia, proposed a classification and noted that the expression of the syndrome was as variable as in the unilateral type.

Some patients show only bilateral microtia without clinically obvious jaw deformities. In this group, however, tomographic studies demonstrated mild condylar blunting or surface irregularities.

The majority of patients display bilateral microtia, mandibular micrognathia, and a class II dental occlusal relationship. While the ear deformities may not be symmetrical, both halves of the mandible appear equally hypoplastic, and there is an anterior open bite deformity.

In addition to bilateral auricular anomalies and severe mandibular micrognathia, macrostomia or transverse facial clefts are seen in other patients. The clefts range from a macrostomia to full thickness buccal clefts extending from the oral commissures to the auricular tragus. These patients represent the most complete expression of the bilateral syndrome.

**Surgical Reconstruction of the Mandible.** In the reconstruction of the micrognathic mandible in bilateral craniofacial microsomia, there are two goals: (1) restoration of mandibular size, form, and position; and (2) correction of the soft tissue deficiency in the mental region.

Restoration of mandibular size and form usually involves increasing the anteroposterior dimensions of both mandibular rami. This has been accomplished either by a sagittal split
osteotomy or by a vertical osteotomy of the rami with interposition bone grafts, as described by Converse and associates (1973b).

Vertical osteotomy of the ramus with interposition of bone grafts is particularly suited to increase the anteroposterior dimension of the ramus and also to add bone grafts to the thin condylar process and underdeveloped mandibular angle. The mandibular body is advanced to the predetermined position, and one can provide for any slight asymmetry requiring more advancement on one side than the other. Intermaxillary fixation is established, the condylar fragments are recessed, and bone grafts are interposed between the fragments and added as onlay grafts.

When the rami are of sufficient size, the sagittal split technique can be employed. The wide surface of contact between the split osseous fragments precludes the need for bone grafting.

In some cases of mandibular micrognathia in bilateral facial microsomia, one also needs to lengthen the body of the mandible by a step osteotomy of the body performed anterior to the mental foramen (Converse and Shapiro, 1952). With this technique, the mandibular incisor teeth are brought into occlusion with their maxillary counterparts. Unlike the sagittal split or vertical osteotomy techniques, there is a disruption in the continuity of the dentoalveolar arch.

Horizontal advancement osteotomy of the anteroinferior portion of the mandible may also be required to advance the mental symphysis. Horizontal advancement osteotomy alone is sufficient to correct the mandibular micrognathia if the dental occlusal relationships are acceptable and additional improvement can be obtained by orthodontic therapy. Refinements of the horizontal osteotomy include the double-tiered osteotomy (Neuner, 1965) or the addition of iliac bone grafts to the osteotomy site. The technique of the various osteotomies is discussed in Chapter 30.

A frequent complication in both hemicraniofacial and bilateral craniofacial microsomia is the soft tissue deficiency that restricts the surgical advancement of the mandible and is responsible for the long-term resorption of the advanced bone. The hypoplasia is apparent not only in the cutaneous tissues over the mandibular symphysis but also in the mucosal lining of the floor of the mouth.

Attempts have been made to counteract the soft tissue resistance by widely detaching the suprathyroid musculature through the "degloving" procedure (see Chapter 30), which exposes the entire anterior portion of the mandible in a subperiosteal plane. Postoperative forward traction of the symphysis can also be exerted by the cranial "halo" device of Georgiade and Nash (19660 and the "stick-and-carrot" outrigger appliance.

The patient shown was born with transverse facial clefts, low-set microtic auricles, deafness, and mandibular micrognathia. The facial clefts had been repaired in infancy. The problem of reconstruction required a number of operations: (1) A surgical-orthodontic premolar set-back osteotomy was performed to correct the position of the anterior maxillary dentoalveolar segment. (2) The tenuous right ramus was reconstructed by a vertical osteotomy combined with bone grafting; the left ramus underwent a sagittal split osteotomy. These
operations increased the anteroposterior dimensions of each ramus. (3) An intraoral skin graft inlay remedied the soft tissue deficiency and distended the cutaneous tissue over the mental area. (4) A bilateral step osteotomy and iliac bone grafting were used to elongate the body of the mandible. The patient has an orthodontic appliance for final alignment of the teeth.

An unusual focus of bone and ectopic dentition in the region of the pterygoid processes was resected when the patient was 7 years old.