Paparella: Volume II: Otology and Neuro-Otology

Section 3: Diseases of the Ear

Part 2: External Ear

Chapter 20: Congenital Aural Atresia:

Embryology, Pathology, Classification, Genetics,

and Surgical Management

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Congenital aural atresia is a relatively common unilateral or bilateral malformation. It is usually associated with deformity of the auricle and regularly accompanied by a developmental anomaly of the middle ear. It is combined with an abnormality of the inner ear in approximately 10 per cent of cases (Altmann, 1949; Naunton and Valvassori, 1968; Bergstrom et al, 1971; Jahrsdoerfer, 1978).

Congenital aural atresia has long been recognized. Several prehistoric skulls have this developmental defect (Hrdlicka, 1933). The deformity is described in the teratologic tables written by the Chaldeans of Mesopotamia about 2000 BC (Ballantyne, 1894).

Moderate and severe forms of congenital aural atresia are encountered in about 1 of 10.000 to 20.000 individuals (Mündnich, 1966). Bezold (1926) found this malformation in 11 of 20.408 patients with otologic problems. From 1950 to 1958, 26 of 289.444 (1 in 11.000) infants born in 35 obstetric clinics in the German Federal Republic had an isolated ear deformity (Kleinsasser and Schlothane, 1963).

Congenital aural atresia occurs more commonly in the male. It is observed much more often unilaterally. Unilateral atresia, for reasons still unknown, affects the right ear more frequently than the left (Altmann, 1965a; Jafek et al, 1975).

At one time, congenital aural atresia was considered a well defined and independent entity. However, it is now obvious that this malformation often represents but one of several malformations involving the derivatives of the posterior segments of the first and second branchial arches. This association was first recognized by Thompson (1845) and Virchow (1864). Aural atresia may also be associated with abnormalities affecting other organ systems, as the result of single gene disorders, chromosomal abnormalities or environmental teratogens.

Embryology

A brief review of the normal development of the ear is given here to facilitate comprehension of the problems encountered in congenital aural atresia.

The inner ear and the middle and outer ear develop independently. Differentiation of the inner ear begins early in the third week of intrauterine development with the appearance

of the ectoderm auditory placode at the level of the myelencephalon. The auditory placode invaginates during the fourth week to form the auditory pit, which later becomes the auditory vesicle. Around the thirty-first day, the labyrinthine recess develops from the dorsal end of the auditory vesicle. This recess gives rise to the endolymphatic duct and sac. During the fifth week a superior dorsal and an inferior ventral compartment develop from the auditory vesicle. The superior compartment differentiates into the semicircular canals and the antrum which, in turn, develop into the utricle and saccule. The lower compartment, through evagination and involution, becomes the cochlear duct. During the seventh week, the cochlear duct completes one turn; during the eight week, one and one-half turns; and during the tenth week, two and one-half turn. At about the twelfth week, the endolymphatic labyrinth is fully differentiated, except for part of the organ of Corti in the apical turn (Bast and Anson, 1960).

In the 2.7-mm long embryo a conglomeration of ganglion cells appears adjacent to the anterior wall of the auditory vesicle and gradually advances toward its medial wall. This represents the acousticofacial ganglion. The origin of this ganglion is not uniform. The acoustic ganglion originates from cells of the rostral wall of the auditory vesicle. The facial ganglion originates mainly from the ganglion crest and, to a lesser extent, from cells of the first epibranchial placode. While the facial ganglion separates from the original acousticofacial ganglion during the third week, the acoustic ganglion maintains its close relationship to the developing auditory vesicle. The acoustic ganglion divides (at the same time as the auditory vesicle) into a superior and an inferior portion. The superior portions sends nerve fibers to the utricle and to the superior and lateral ampulla. The inferior portion sends nerve fibers to the saccule and to the posterior ampulla. The remaining part of the inferior portion of the acoustic ganglion later becomes the spiral ganglion, situated at the concave side of the cochlear duct. The close relationship of this ganglion to the developing cochlear duct leads to its typical spiral arrangement. At about the same time the facial nerve comes into view. Nerve fibers grow from the neural tube into the mesenchyme of the second branchial arch. The chorda tympani nerve arises from peripheral nerve fibers of the geniculate (formerly facial) ganglion. The chorda tympani joins the developing facial nerve for a short distance and, by the fifth week, unites with the third branch of the trigeminal nerve (Altmann, 1965a).

While the membranous labyrinth is taking shape, a gradual concentration of mesenchyme takes place around it (fourth week). This surrounding mesenchyme is transformed into cartilage after the seventh week, and subsequently converted into bone to form the inner ear capsule. Ossification of the labyrinthine capsule begins at about the sixteenth week, as the endolymphatic labyrinth reaches adult form, and develops from 14 centers. Ossification begins near the round window and ends in the area of the oval window and fissura ante fenestram. It is generally completed by the twenty-third week (Richany et al, 1960).

During the third week of fetal development, while the auditory vesicle is being formed, an outpocketing develops from the foregut and becomes the first pharyngeal pouch. The dorsal end of this first pharyngeal pouch, through gradual expansion, becomes in succession the eustachian tube, tympanic cavity and extensions of the latter - namely, the epitympanic recess, antrum and mastoid air cells. Between the fourth and sixth weeks, the lower portion of the tympanic cavity begins to form. In the eight week, the upper portion is still occupied by mesenchyme. During the twenty-third week, the upper portion of the cavity begins to develop and, in the thirtieth week, the middle ear cavity is almost completely

formed. Subsequently, the mesenchyme gradually disappears from the middle ear cavity and the walls and contents become lined by endodermal epithelium of the tubotympanic recess.

The malleus and incus are derived from the first and second branchial arches. The first branchial arch forms the head of the malleus and the body and short process of the incus. The second branchial arch forms the short process and handle of the malleus, long process of the incus, and stapes, with the exception of the vestibular portion of the footplate. The vestibular portion of the stapedial footplate is a derivative of the otic capsule. The head of the malleus, body of the incus, and cartilaginous mandible are initially continuous as Meckel's cartilage of the first branchial arch. The styloid process, hyoid bone, stapedial arch and tympanic portion of the footplate, long crus of the incus, and malleus handle are initially continuous as Reichert's cartilage of the second branchial arch. Subsequently, the ossicles separate from their parent cartilages and begin an independent growth. Interference with their later differentiation may give rise to malformations occasionally encountered during explorations of the middle ear.

Shortly after the appearance of the primordium of the stapes (sixth week), a condensation of the mesenchyme signifies the beginning development of malleus and incus. The incudomallear and incudostapedial articulations are already established during the eight week when chondrification begins. Ossification of malleus and incus begins during the sixteenth week and is largely completed by the thirtieth week (Richany et al, 1954).

A spherical condensation of mesenchyme during the fourth week marks the beginning of the development of the stapes. This mesenchymal mass is perforated by the stapedial artery during the fifth week, assumes a ring-like form during the sixth week, approaches the periotic mesenchyme during the seventh week, and fuses with that portion of the periotic mesenchyme that forms the stapedial lamina. Chondrification of the stapes begins during the eight week, and ossification around the eighteenth week. Originating in the center of the tympanic portion of the footplate, ossification spreads along the crura and reaches the capitulum by the twenty-first week (Richany et al, 1960), by which time the stapes has reached its ultimate dimensions. At this stage, the crura are bony cylinders, with a thin cortical layer of bone and a central marrow space. The footplate consists of an inner and outer layer of bone, a small central marrow space, and an inner layer of cartilage that covers the vestibular and articular surface. After the twenty-fourth week, an excavation process transforms the cylindrical crura into inward-concave semicanals, transforms the capitulum into an empty cylinder open towards the footplate, and removes the outer, tympanic, osseous layer of the footplate (Bast and Anson, 1960). This final remodelling process of the stapes is completed by the thirty-eight week.

The annular ligament of the oval window develops between the tenth and thirteenth week by dedifferentiation of the cartilaginous stapedial lamina of the otic capsule into mesenchyme, and subsequent differentiation into fibrous tissue. The annual ligament is fully developed by the sixteenth week.

The round window area is recognized during the eleventh week as a condensation of embryonic connective tissue between the scala tympani and the fossula of the cochlear fenestra, which itself is filled with mesenchyme. At this stage, the round window extends directly to a portion of the wall of the periotic duct. Subsequent differentiation of the

respective areas into cartilage, bone, and fibrous connective tissue results in the formation of the round window niche and round window membrane by the twenty-sixth week.

The tensor tympani muscle and tendon are derivatives of the first branchial arch; their development begins between the eight and eleventh weeks. The stapedius muscle begins to develop at eight and one-half weeks from a group of mesenchymal cells located adjacent to the interhyal. The stapedius tendon in turn develops from mesenchymal cells in close proximity to the head of the stapes. The anterior wall of the fallopian canal and the pyramidal process are formed by fusion of the laterohyal with the otic capsule. The laterohyal and tympanohyal both participate in the formation of the descending portion of the fallopian canal. As a result of their malformation, the facial nerve may take an abnormal course and leave the middle ear in a more anterior and lateral direction (Altmann, 1965b).

As the inner ear is being developed, the middle and outer ear are also taking shape. On the outside of the embryonic head, between the first and second branchial arches, a branchial groove develops opposite a corresponding outpocketing of the pharynx. The epithelium at the medial end of the groove is for a time in contact with the entoderm of the first pharyngeal pouch. Later, during the enlargement of the head, mesoderm grows between and separates these two epithelial layers. Near the close of the second fetal month, the branchial groove deepens again into a funnel-shaped tube. This tube, which subsequently becomes supported by the cartilages of the outer ear, represents the outer one third of the external acoustic meatus. The medial extremity of this funnel then produces a solid core of epithelial cells which grows inward to meet the outer epithelium of the pharyngeal pouch again until only a thin seam of connective tissue remains to separate the two epithelial layers. This seam of connective tissue, which is derived from the mesenchyme, becomes the tunica propria of the tympanic membrane.

The core of epithelial cells remains solid until near the end of fetal life. Meanwhile, the connective tissue around the margin of the tympanic membrane begins to ossify at about the third month. It gives rise to the tympanic ring which serves as a support for the tympanic membrane. Finally, in the seventh month of fetal life, when all other structures of the outer, middle, and inner ear are well differentiated, the solid core of epithelial cells splits, first in its deepest portion to form the outer surface of the tympanic membrane, and then extending and opening outward to joint the lumen of the primitive meatus. This tube, with the surrounding connective tissue and expanded bony tympanic annulus, becomes the inner two-thirds of the external acoustic meatus - that is, the bony external auditory canal. Therefore, congenital atresia of the meatus may occur with a normally differentiated and functioning tympanic membrane and ossicular chain, as well as with various malformations of these structures, depending on the age at which development was delayed or arrested.

The auricle is formed by growth of mesenchymal tissue flanking the first branchial groove. At about the sixth week, six knoblike hillocks appear from the mandibular and hyoid arch tissues. The coalescence of these tubercules, by the third month, and their further development mold the auricle (Bast and Anson, 1960). Whereas the tragus derives from the first (mandibular) arch, all the auricle except the tragus develops from the second (hyoid) arch.

Failure in differentiation of the first and second branchial arches may affect the auricle and result in microtia, anotia, or malposition of the pinna, or the middle ear ossicles may be malformed, fused, or aplastic. Malformation of the stapedial footplate, on the other hand, results from the total lack of differentiation, incomplete differentiation, or failure of the lamina stapedialis to separate from the otic capsule (Lindsay et al, 1966).

Failure of the first branchial groove to develop may result in stenosis or atresia of the external meatus. A disturbance in differentiation of the first pharyngeal pouch will affect the architecture of the eustachian tube and middle ear, as well as the extent and character of the pneumatization of the mastoid process.

Since the membranous labyrinth derives from the ectodermal otocyst and develops independently from the rest of the ear, which is mainly a derivative of the branchial apparatus, combined malformations of the inner, middle, and outer ear should theoretically not be expected to occur frequently; however, a significant number of combined malformations have been documented (Altmann, 1965a; Bergstrom et al, 1971).

Pathology

The more severe forms of congenital auricular malformation are always associated with meatal atresia, whereas meatal atresia may, in a rare instance, be seen in patients with a normal pinna.

Atresia of the meatus may be membranous or osseous. Membranous atresia is much less common and is characterized by a rudimentary cartilaginous canal, separated from the middle ear by a dense structure of connective tissue. In the osseous form of meatal atresia, the bony portion of the external meatus is absent, and a more or less solid mass of compact bone forms the lateral wall of the middle ear cavity. Osseous atresia is regularly accompanied by a malformation of the cavity and structures of the middle ear. These malformations vary in degree. The origin, formation, and location of the atresia plate, and anomalies involving the middle ear, will be discussed later.

It was stated earlier that unilateral atresia is more common than bilateral atresia. In unilateral atresia it is not unusual for the other ear to show a less severe malformation, with a narrow meatus and hypoplastic tympanic membrane or abnormal middle ear ossicles. The mechanisms by which developmental anomalies of the branchial apparatus occur are not clear. Developmental failure in any particular instance depends on which portion of the branchial apparatus was involved, and at which embryonic stage normal development was arrested.

Case histories of 241 patients with congenital aural atresia seen at the Johns Hopkins Hospital between 1955 and 1975 were reviewed by Jafek and colleagues (1975). Analysis revealed the following:

1. Seventy patients showed bilateral involvement; an incidence of 29 per cent.

2. The deformity occurred more commonly in the male (61 per cent), and affected the right ear more frequently (57 per cent) than the left.

3. A positive family history of a congenital ear deformity affecting either a sibling or another relative was obtained in 14 per cent.

4. Additional malformations were found in 56 per cent of patients. A specific malformation syndrome could be recognized in 35 patients (15 per cent). Fifteen of these patients had mandibulofacial dysostosis (Collins, 1900; Franceschetti and Zwahlen, 1944; Franceschetti and Klein, 1949), three had craniofacial dysostosis (Crouzon, 1912), three exhibited a first and second branchial arch syndrome (Grabb, 1965), three had the rubella syndrome, and two had achondroplasia. Congenital aural atresia was also found associated, in one instance each, with Goldenhar's syndrome, thalidomide embryopathy, Mobius syndrome, Noonan's syndrome, the otopalatal-digital syndrome, the oral-facial-digital syndrome, the Pierre-Robin anomaly, and von Recklinghausen's neurofibromatosis. However, the association of congenital aural atresia with many of these malformation syndromes may have been fortuitous, and further studies are necessary.

5. In the presence of congenital aural atresia, a careful search for other abnormalities in the head and neck as well as in other organ systems is indicated.

6. Although the degree of the deformity of the external ear is generally proportional to the malformation of the middle ear, no characteristic correlation patterns could be identified radiologically, and each case has to be evaluated individually.

Classification of External and Middle Ear Malformations

Developmental anomalies of the external and middle ear may be conveniently classified into the following three groups, depending on the degree of malformation and the structures involved: minor, moderate, and severe malformation (Altmann, 1965b).

Minor External and Middle Ear Malformations (Group I)

In this group the auricle can be normal, but frequently it reveals some intimation of malformation. Whereas the auricle may reveal no abnormalities in overall dimensions and position it may display minor variations in relief, resembling the later developmental stages of the pinna. Many of these variations show a hereditary tendency. The external auditory canal can also be normal; it occasionally is hypoplastic in its entire length. In a rare instance it is present only in its medial portion. The tympanic membrane can be normal in the presence of a regular canal. More often it is thickened and appears opaque. A normal tympanic membrane, however, does not preclude the presence of anomalies of the middle ear. Such anomalies can include a hypoplastic middle ear cavity, malformation of the malleus and incus, abnormalities of middle ear muscles and nerves, and agenesis of the oval and round windows. The handle of the malleus is often deformed and in an abnormal position. In a rare instance the inner ear is abnormal. The semicircular canals can be hypoplastic, the lateral being the most frequently involved (Mündnich, 1966). Minor external and middle ear malformations are commonly seen in craniofacial dysostosis (Crouzon, 1912).

Moderate External and Middle Ear Malformations (Group II)

This group encompasses the majority of ear deformities. The auricle is rarely normal and is usually represented by a small rudimentary soft tissue structure of variable size, often in an abnormal, anterior, and inferior position. Anomalies of the auricle have been divided into three types: microtia types I, II, and III (Marx, 1922). In microtia type I the different parts of the rudimentary auricle are still recognizable. In microtia type II the auricle is represented by a vertical, longitudinal, somewhat curving ridge, which usually includes some cartilage and resembles a primitive helix. In microtia type III, the rudiment of soft tissue no longer has any resemblance to any portion of the auricle. In a rare instance the auricle may be absent; however, this severest expression of the malformation, known as anotia, is usually found in severe external and middle ear malformations (group III).

The external auditory canal in Group II malformations is either hypoplastic or aplastic, or it may end blindly with one or occasionally two fistulous tracts leading toward a rudimentary tympanic membrane. The fistulous tract can be associated with an epidermoid inclusion cyst (cholesteatoma).

The tympanic bone can be either present or absent. If present, it is invariably severely malformed and usually represented by an osseous plate whose central portion may consist of connective tissue. This atresia plate forms the lateral wall of the middle ear cavity. If the tympanic bone is absent, the upper portion of Reichert's cartilage may show anomalies, both in configuration and position. It can form the principal portion of the atresia plate. In such instances the chorda tympani nerve courses laterally to it (Marx, 1922; Altmann, 1965b). The atresia plate, therefore, is medial to the normal location of the tympanic membrane and the middle ear cavity represents only the medial half of a normal cavum tympani. If the tympanic bone is absent and if the upper portion of Reichert's cartilage is very hypoplastic, bony processes from the squamous portion of the temporal bone and hypotympanum join in the formation of a lateral osseous wall of the tympanic cavity. In such instances the chorda tympani nerve also courses laterally to the atresia plate. The atresia plate is again medial to where the tympanic membrane would normally be encountered, but less medial than when formed by Reichert's cartilage (Altmann, 1965b).

The cellular development of the mastoid process varies greatly. It can be normal, limited, or absent. In most instances it is restricted. Differentiation of the antrum and periantral cells is directly related to the degree of pneumatization. Normal pneumatization is occasionally associated with fairly good differentiation of middle ear structures.

Ossicular malformations are regularly encountered in group II malformations. In general, they are more frequent and more severe than in group I. The malleus and incus frequently form a conglomeration, or have bony unions with the atresia plate and walls of the epitympanic recess. Infrequently the incus lacks a connection with the stapes. The stapes may be malformed or even absent. The stapedial arch may be fixed by an osseous connection to the fallopian canal or to the promontory, or by a bony structure that takes the place of the stapedius tendon. The stapedial footplate can be fixed as a result of incomplete differentiation of the annular ligament, or total lack of it. The middle ear muscles may be anomalous, supernumerary, and malpositioned.

The facial nerve may be hypoplastic or, in a rare instance, absent. Its course from the geniculate ganglion can be straight downward over the promontory. The nerve may course uncovered by bone above or below the oval window, or even through the stapedial arch. On rare occasions it may bifurcate around the oval window or divide into three portions distal from it. The greater and the lesser superficial petrosal nerves and the chorda tympanic nerve can be absent (Altmann, 1965b).

Severe External and Middle Ear Malformations (Group III)

In this group the auricle generally is severely malformed; it can even be absent. The external auditory canal is aplastic. Strands of connective tissue and epidermis, or a fistulous tract, are occasionally encountered as a rudiment of an external meatus. Very often the mastoid process is hypoplastic and not pneumatized. The middle ear and antrum may be absent or may be represented by a slit-like lumen. Such a rudimentary middle ear cleft may contain embryonic mesenchyme, mature connective tissue, or osseous trabeculae. Anomalies of the facial nerve are the rule. The middle ear ossicles are frequently absent. Malformations of the inner ear are most frequently encountered in this group. The inner ear malformations can involve one or several semicircular canals and the vestibule, as well as the cochlea. Severe external and middle ear malformations are often associated with the severe cranial malformations. They are regularly encountered in mandibulofacial dysostosis (Collins, 1900; Franceschetti and Zwahlen, 1944; Altmann, 1965b). They have been commonly observed in thalidomide embryopathy.

Atresia in Genetic Disorders

Despite the frequency of congenital aural atresia, little is known about its genetics as an isolated trait, or in conjunction with abnormalities of the pinna, middle ear, and inner ear or in association with abnormalities in other organ systems. Family histories have rarely been taken, and the results of complete physical examinations are only occasionally reported. Therefore, it is difficult to delineate more than a few disorders in which a genetic etiology is significant. The paucity of well-documented reports indicate that complete evaluation, including a family history, of all individuals with external ear malformations is necessary to determine the role of heredity in the development of abnormalities of the ear.

Microtia, Meatal Atresia, and Hearing Impairment

Unilateral or bilateral external ear abnormalities, including atresia of the external auditory meatus and microtia, have been reported in association with hearing loss as a recessive disorder by Konigsmark and co-workers (1972). The pinnae may be almost completely absent. Ellwood and colleagues (1968) and Dar and Winter (1973) have described another, similarly affected family. Both conductive and sensorineural hearing losses have been noted. Middle ear abnormalities, consisting of absence of or malformed ossicles, and reduction in size of the middle ear cavity, have been described. The oval window, semicircular canal, cochlea, and internal auditory canal are normal (Konigsmark et al, 1972). Meurman (1957) described two sibs, one with bilateral aural atresia without microtia, and the other with both unilateral atresia and microtia; these differences may represent variation in expression of the same disorder. No mention was made of the patients' hearing status or whether the parents were similarly affected, and it is possible that these two patients had the same disorder as

described by Konigsmark and associates (1972).

Conductive hearing loss, microtia, and meatal atresia have been described by Kunkel (1959) in a family in which the inheritance pattern was compatible with a dominant disorder. Balci (1974) reported a father and son with aural atresia and microtia. The father had a unilateral mixed hearing loss of unknown degree and had a history of noise exposure; hearing in the other ear was normal. No audiometric studies were performed on the son (Balci, 1978).

Mandibulofacial Dysostosis (Treacher Collins Syndrome)

Mandibulofacial syndrome (Treacher Collins syndrome) is an autosomal dominant disorder characterized by bilateral and, in most cases, symmetric abnormalities. Distinguishing features include flattening of the frontonasal angle, antimongoloid obliquity of the palpebral fissure, colobomas of the lower eyelids with absence of eyelashes medial to the colobomas, and hypoplasia of the maxillary sinuses and malar bones. Hypoplasia or absence of the zygomatic processes, mandibular hypoplasia, increased mandibular angle, and prominent antegonial notch are common. In some cases, the palate may be cleft, lachrymal puncta and meibomian glands may be absent, blind fistulae may be present between the ear and oral commissures, and there may be macrostomia (Gorlin et al, 1976a).

Otologic abnormalities are a prime feature of the disorder and include microtia, external auditory canal atresia, and moderately severe conductive hearing loss (Hutchinson et al, 1977). However, hearing may be normal in some cases (Axelsson et al, 1963; Farrar, 1967). The malleus, incus, and stapes have been reported to be malformed (McKenzie and Craig, 1955; Harrison 1950-1951; Stovin et al, 1960; Holobrow, 1961; Herberts, 1962; Axelsson, 1963; Edwards, 1964; Sando et al, 1968; Sugar and Berman, 1968; Hutchinson et al, 1977). Inner ear structures have been reported to be present and normal by some investigators (Böök and Fraccaro, 1955; McKenzie and Craig, 1955; Herberts, 1962; Behrents et al, 1977; Hutchinson et al, 1977), and abnormal by others (Mann and Kilner, 1943; Livingstone, 1959; Herberts, 1962; Sando et al, 1968; Hutchinson et al, 1977). The middle ear cavity is usually hypoplastic or absent (Hutchinson et al, 1977).

Most patients are of normal intelligence. Those reported to be mentally retarded are probably "retarded" secondary to hearing impairment.

Variation in expression among affected individuals is marked. That is, both severely and mildly affected individuals may be present in the same family (Leopold et al, 1945; Straith and Lewis, 1949; Böök and Fracaro, 1955; Wildervanck, 1960; Axelsson et al, 1963; Maran, 1964; Rovin et al, 1964; Farrar, 1967; Fazen, 1967; Hall, 1969). The extent of the variation emphasizes the importance of evaluating all relatives of an affected individual for minor stigmata, so that accurate genetic counseling may be provided. Examinations should include a clinical evaluation of the craniofacies, roentgenographic study of the facial bones and mandible, and audiologic evaluation. Several reports have described affected sibs with this syndrome, although the parents were presumed normal (Stovin et al, 1960; Holobrow, 1961; Herberts, 1962; Axelsson et al, 1963). However, careful examination of the parents was not performed. The high number of new mutation cases reported may in reality reflect inadequate investigation of relatives of affected individuals.

Acrofacial Dysostosis (Nager Syndrome)

The facies of patients with acrofacial dysostosis (Nager syndrome) are similar to those with mandibulofacial dysostosis. Several reports have appeared (Nager and de Reynier, 1948; Franceschetti and Klein, 1949; O'Connor and Conway, 1950; McNeill and Wynter-Wedderburn, 1953; Altmann, 1955; Borcher and Klein, 1960; Chou, 1960; Fernandez and Ronis, 1964; Marden et al, 1964; Jones, 1968; Genée, 1969; Pfeiffer, 1969; Neidhart, 1968; Klein et al, 1973; Vatre, 1971; Bowen and Harley, 1974; Walker, 1974; Herrmann et al, 1975; Burton and Nadler, 1977; Lowry, 1977; Meyerson et al, 1977). However, individuals with acrofacial dysostosis have preaxial upper limb defects, consisting of hypoplastic or absent thumbs, radioulnar synostosis, radial aplasia, and abnormalities of the carpals, metacarpals, and phalanges. The humerus may be shortened.

The inheritance pattern of acrofacial dysostosis has not been determined, since most cases are sporadic. Recessive inheritance has been suggested; consanguinity was noted in the family reported by Burton and Nadler (1977), and two affected children were born to presumed normal parents in the family reported by Walker (1974). Other reports suggest autosomal dominant inheritance because of advanced paternal age (Lowry, 1977).

Abnormalities of the ear similar to those in mandibulofacial dysostosis have been noted; microtia, atresia of the external auditory canal, and malformation of the auditory ossicles. Investigation of the inner ear has rarely been performed; Franceschetti and Klein (1949) reported an underdeveloped lateral semicircular canal, although Lowry (1977) reported the semicircular canals to be normal. Hearing loss, both conductive and mixed, may occur, although many reports have not mentioned the hearing status.

First and Second Branchial Arch Syndromes (Hemifacial Microsomia: Lateral Facial Dysplasia)

This group of disorders consists of a spectrum of craniofacial malformations with considerable variation in expression from one patient to another. The variation and sporadic occurrence of the disorder mark the role of hereditary factors in its etiology. However, Grabb (1965) reported three patients with the first and second branchial arch syndrome, each of whom had an affected relative, and one patient who had an affected parent. In general, affected individuals are characterized by asymmetric facies, and unilateral abnormalities may occur (Ross, 1975).

The external ear may be absent or malformed, the external auditory canal atretic, and hearing impaired. Tissue tags may be present along a line from the tragus to the oral commissure. The mandible may be small, and its ramus and condyle hypoplastic or absent. In contrast to mandibulofacial dysostosis, there may be coloboma of the upper eyelid. Hypoplasia of the malar bones, macrostomia, and cleft palate may also be present. The zygomatic arch, sphenoid and temporal bones, temporomandibular joint, maxilla, oral and facial neuromusculature, muscles of mastication, tongue, and parotid gland may be abnormal (Grabb, 1965; Coccaro et al, 1975; Ross, 1975). Associated abnormalities of the cardiovascular, renal, and nervous systems have been noted (Gorlin et al, 1976b).

Oculoauriculovertebral dysplasia (OAVD) (Goldenhar's syndrome) (Gorlin et al, 1963) may represent a variant of the first and second branchial arch syndromes. In addition to abnormalities characteristic of the first and second branchial arch syndrome, individuals with OAVD have epibulbar dermoids or lipodermoids, ear tags and preauricular pits or fistulae, and abnormalities of the vertebral column, primarily hemivertebrae. Mental retardation and congenital heart disease have been noted (Shokeir, 1977). Both sensorineural and conductive hearing impairment have been reported (Budden and Robinson, 1973; Darling et al, 1968).

Craniofacial Dysostosis (Crouzon's Disease)

Patients with distinctive craniofacial deformity have premature fusion of the cranial sutures, ocular hypertelorism and proptosis, midface hypoplasia, and beak-shaped nose (Cohen, 1975; Gorlin et al, 1976c). Premature synostosis of the sagittal, lamboidal, and coronal sutures results in brachycephaly and acrocephaly; however, no definitive calvarial form can be said to be diagnostic of craniofacial dysostosis, since head shape is in part related to the sequence in which skull sutures fuse prematurely (Bertelson, 1958). Nausea, vomiting, and headaches are frequent symptoms. As a result of shallow orbits, proptosis may be severe. There may be divergent strabismus, loss of vision, papilledema, and optic atrophy (Koziak, 1954).

A hyperostotic ridge may be palpable in the area of the metopic suture (Parks and Costenbader, 1950), and a bony prominence may be present in the region of the anterior fontanelle. The mandible is prognathic relative to the maxilla, there is malocclusion, and the palate is V-shaped.

On radiographs of the skull, premature suture closure, thinning of the skull, and prominent convolutional markings are noted (Arce and Arce, 1942; Parks and Costenbader, 1950; Pinkerton and Pinkerton, 1952; Koziak, 1954; Vulliamy and Normandale, 1966).

External auditory canal atresia has been reported by several investigators (Aubrey, 1935; Nager and de Reynier, 1948; Wiegand, 1954; Blodi, 1957; Baldwin, 1968). Hearing loss has been noted (Aubrey, 1935; Parks and Costenbader, 1950; Blodi, 1957; Meyerson, 1957; Boedts, 1967; Baldwin, 1968; Kreiberg and Jensen, 1977), but formal audiometric studies have not been done in most cases to document the type of hearing impairment accurately.

The absence of hand malformations in craniofacial dysostosis serves to differentiate it from acrocephalosyndactyly type I (Apert syndrome) (Gorlin et al, 1976d), in which premature craniosynostosis is associated with osseous and soft tissue syndactyly of the hands and feet, and from acrocephalosyndactyly type V (Pfeiffer's syndrome) (Gorlin et al, 1976e), in which premature craniosynostosis is associated with brachydactyly, soft tissue syndactyly, and broad thumbs and toes. Other forms of craniosynostosis from which craniofacial dysostosis should be distinguished have been reviewed by Cohen (1975).

Hypertelorism, Microtia, and Clefting

Bixler and co-workers (1969), reported two female sibs with mental retardation, microcephaly, short stature, orbital hypertelorism, primary telecanthus, cleft lip and cleft

palate, and hypoplasia of the thenar eminences. In addition, one patient had a pelvic kidney; the other had crossed ectopia of the left kidney which was positioned in the right lumbar region.

Abnormalities of the ears in these girls included microtia, atresia of the external auditory canal, and hypoplasia of the auditory ossicles. The vestibule, cochlea, semicircular canals, and internal auditory canals were normal. Hearing loss was noted, but the type and degree were not specified.

Both sisters had congenital heart disease; however, some of their relatives had congenital heart disease without otologic or facial abnormalities, suggesting that heart disease was segregating independently.

18q- Syndrome

Individuals with 18q- syndrome have psychomotor retardation, hypotonia, short stature, microcephaly, hypoplastic midface, epicanthus, ophthalmologic abnormalities, cleft palate, congenital heart disease, abnormalities of the genitalia, and tapered fingers with an increased number of whorls (de Grouchy, 1969; Schnizel et al, 1975; Konigsmark and Gorlin, 1976). The external ear canals may be normal, stenotic, or atretic, and the anthelix and antitragus may be prominent. Of the four patients reported by Bergstrom et al (1974), three had bilateral atresia or hypoplasia of the external auditory canal, conductive hearing loss, and normal pinnae. Their middle ears were normal; polytomograms revealed atresia plates in the region of the tympanic membranes. The fourth patient, who had normal ear canals, had normal tomographic studies but had a mixed hearing loss.

Atresia in Embryopathies

During the 50s and 60s, it became obvious that a medicament (thalidomide) and the rubella virus may also produce the syndrome of congenital aural atresia.

Thalidomide

Ingestion of the tranquilizer thalidomide (West germany, Contergan) may produce a host of malformations that include deformities of the limbs, malformation of the heart, anomalies of the respiratory, digestive, and urinary systems, clefts of face, lip, and palate, eye anomalies, malformations of the ear, and transient or persistent hemangiomas of the face. Thalidomide embryopathies were observed in West Germany and England between 1959 and 1962. In West Germany during that period about 7000 children were born with malformations of the limbs (Weicker et al, 1962). If isolated malformations of the hands and fingers are included, the actual number of affected children is closer to 10.000. Since malformations of the limbs are associated with anomalies of the ears in approximately 10 per cent of the patients, it can be estimated that about 1000 of these children also had ear deformities (Kleinsasser and Schlothane, 1963, 1964; Mündnich, 1966). In these children aural atresia occurred with microtia or anotia, with sixth and seventh nerve palsy and, at times, with malformations and even agenesia of the inner ear (Lenz and Knapp, 1962).

According to Lenz (1963), ingestion of thalidomide during the thirty-fifth and thirty-sixth days, and occasionally during the thirty-seventh day after the last menstrual period, produced anotia, facial nerve palsy, and damage to the eye muscle nuclei. Lesser ear deformities occurred after ingestion between the thirty-eighth and forty-fifth days. The infrequent duplication of the thumb was seen following ingestion prior to the thirty-eighth day. Partial or total agenesis of both arms corresponded to the period from the thirty-ninth to the forty-fourth day. Cardiac malformations, duodenal atresia, and congenital subluxation of the hip occurred with ingestion between the thirty-ninth and the forty-fifth days. Ingestion around the fiftieth day produced lesser thumb malformations, rectal stenosis and anal atresia. Thus, prolonged ingestion of thalidomide by a pregnant woman obviously can, during sensitive phases of fetal development, affect several organization centers, which, in turn, can lead to a kaleidoscopic combination of developmental defects. Nevertheless, many women, in spite of regular thalidomide ingestion, gave birth to normal children, suggesting that not all women are sensitive to this teratogenic drug.

Maternal Rubella

The introduction of polytomography led to a reevaluation and revision of the original concept that malformations of the middle ear were never associated with developmental defects of the inner ear. Roentgenologic investigation as well as surgical exploration provided evidence that malformations of the middle ear are not infrequently combined with severe anomalies of the labyrinth and cochlea (Ombrédanne, 1957; Frey and Mündnich, 1957). Furthermore, it became evident that, in maternal rubella, not only the inner but also the middle and outer ear can be affected (Leicher, 1952).

General Management of Congenital Aural Atresia

Microtia and congenital aural atresia are noted at birth or soon after, and attention is focused first on the deformity rather than on the potential hearing problem. If the malformation is unilateral, the hearing problem will be of secondary importance; impairment of hearing is hardly noticed, provided hearing in the other ear is normal.

In cases not complicated by involvement of the cochlea, hearing tests reveal a conductive hearing loss. This loss may be combined with relatively good hearing for speech. Even in bilateral atresia it is not uncommon for the spoken voice to be relatively well understood and, in such instances, speech development is not severely impaired. Clinical and histologic investigations, however, have revealed that aural atresia is sometimes associated with sensorineural hearing loss because of cochlear abnormalities. The vestibular labyrinth may be abnormal, and the lateral semicircular canal may be distended, abnormally located, rudimentary, or absent. Rarely, all three semicircular canals may be aplastic. Anomalies of the vestibular labyrinth occur mainly in the malformations of groups II and III, and are rarely seen in group I.

Reconstructive surgery is justified only when there is high probability of lasting success. The sole purpose of the operation should be to give the patient serviceable hearing (average speech reception threshold less than 30 to 35 dB ANSI). The surgeon's decision is based mainly on two factors: cochlear function and anatomy of the ear.

Determination of cochlear function can be done with great accuracy at the time the child is ready for surgical treatment. In bilateral atresia the child's response to airborne sound, vocabulary, as developed by amplification methods, and responses to pure tones and to speech afford a good measure of inner ear function. In unilateral atresia, determination of cochlear function is obviously much easier.

Long before the time of surgical treatment, however, hearing may be evaluated in the infant by screening methods and evoked response audiometry and, in the older child, by standard audiologic evaluation methods for that age group. Early determination of cochlear nerve function is most important in bilateral atresia, because auditory training should begin in the first year of life. This is the period when the child with normal hearing is listening in preparation for speaking. Constant repetition of sounds enables the cortical auditory centers to develop discrimination. As a result, the auditory association areas build up a store of engrams of sounds, associated with their meanings, and gradually more complicated sound patterns such as speech are recognized and understood. During this period of listening and understanding, the child is learning to control the muscles of articulation and to imitate sounds. Thus, speech and language are being developed.

Since the ability to acquire auditory discrimination gradually diminishes as the child grows older, auditory training should begin in the second half of the first year of life in the presence of bilateral atresia with a marked conductive hearing loss. In instances in which bilateral atresia is combined with malformation of the head, additional factors such as the child's intelligence and personality must be considered. Aural rehabilitation then becomes much more complex, and its success depends greatly on the patient's mental ability.

The appropriate way to study the anatomic relationship preoperatively is a roentgenologic examination. The lateral (Schüller) projection of the temporal bone furnishes valuable information about the size and pneumatization of the mastoid process, and about the position and configuration of the temporomandibular articulation. If the mastoid process is well pneumatized, a middle ear cavity and ossicles are most likely to be present, although they usually are malformed. The longitudinal (Stenvers) projection of the petrous pyramid enables the configuration of the otic capsule to be evaluated. In the axial (Hirtz) projection of the skull, the regions of the external meatus, attic, and ossicles are generally well visualized. The most accurate information, however, is obtained from axial and coronal high resolution thin section computed tomography (CT) with appropriate bone windows of the temporal bone. These scans disclose the type of atresia, shape of the atresia plate, size of the middle ear, configuration of the middle ear ossicles, and differentiation of the inner ear capsule. In delineating the finer details of the cochlea and labyrinth, they may disclose associated malformations of the inner ear.

A caloric test should be performed in roentgenologically proven or suspected inner ear malformations with an intact external auditory meatus. A negative test, in the presence of an otherwise normally functioning inner ear, is not necessarily a contraindication for the rarely performed fenestration of the lateral semicircular canal.

The grade of differentiation of the auricle can, to some extent, indicate the degree of malformation to be expected in the middle ear. Anotia or microtia indicates arrest of development of the branchial apparatus prior to the third month of embryonic life;

consequently, severe dysplasia of the tympanic membrane and the middle ear ossicles is to be expected. Conversely, if meatal atresia is accompanied by a well-formed auricle, the malformation of the tympanic membrane and outer ossicles is often less severe. Microtia and anotia are generally combined with suppression or lack of pneumatization. Occasionally, however, a severely microtic auricle may be associated with normal pneumatization.

In bilateral aural atresia, reconstructive surgery may be considered when there is evidence of good cochlear function and when a middle ear cavity, ossicular structures, and if possible, both labyrinthine windows can be identified. In most instances the anatomy and cochlear function are similar in the two ears; for this reason it is immaterial which side is operated upon first. If possible, the operation should be performed on one ear when the child is about 6 or 7 years old. This, in our opinion, appears to be the earliest suitable time, for the following reasons. At birth, the tympanic cavity has almost an adult configuration, except for the position of the tympanic membrane. The mastoid process, however, does not yet exist. Its development and pneumatization begin at about the age of 1 year. By the sixth or seventh year the major portion of the developing mastoid process usually is pneumatized. Although growth of the temporal bone continues until the end of the second decade of life, the anatomy of the middle ear in general does not change significantly enough to alter a sound-conducting system constructed at the age of 6 or 7 years.

Correction of congenital aural atresia of the external and middle ear was originally recommended only in instances of bilateral malformation. Improvement of surgical techniques within the past two decades now enables properly selected patients with unilateral involvement to consider an operation. However, since unilateral aural atresia with normal hearing in the other ear does not present a severe hearing problem, surgery should be postponed until the patient reaches the age of 17 or 18 years. The prerequisite remains - namely, that one should reasonably expect serviceable hearing after surgery. It should be emphasized that if unilateral aural atresia is associated with an infected auricular fistula or a cholesteatoma, corrective surgery is performed regardless of the patient's age.

Surgical Management

Minor External and Middle Ear Malformations (Group I)

Under general anesthesia an endaural incision is made and a posterior tympanomeatal flap is raised. If the external auditory canal is stenotic, it should be enlarged. Subsequent procedures depend on the anomalies encountered in the middle ear. The primary objective is to establish free mobility of the ossicular chain, although it may be deformed. Bony synostoses connecting the ossicles to the walls of the epitympanic recess or to the promontory, or a rare ossified stapedius tendon, should be removed. If the vestibular fenestra is not fully differentiated, an oval window can be created and a stapes prosthesis inserted. These procedure generally have a good prognosis. More difficult and somewhat less promising are the techniques concerned with establishing continuity of a malformed ossicular chain. These techniques may require repositioning the malleus and incus or interposing an autograft or a prosthesis.

Moderate External and Middle Ear Malformations (Group II)

Various approaches to the problem of aural atresia have been described (Pattee, 1947; Ombrédanne, 1947; House, 1953; Meurman, 1957; Mündnich, 1965; Bellucci, 1966; Makishima and Tokunaga, 1966; Shambaugh, 1967; Derlacki, 1968; Crabtree, 1968; Linthicum, 1971; Jahrsdoerfer, 1978; Mattox and Fisch, 1986). The underlying principle of operative treatment is the construction of a sound-conducting system with an effective impedance matching mechanism. Such a system includes an air-containing middle ear cavity closed by a movable tympanic membrane that is in contact with a freely movable ossicular chain and a mobile stapes. It also requires a functioning eustachian tube. The cochlear and vestibular fenestrae must be intact.

Correction of membranous meatal atresia, in the presence of an intact tympanic membrane and ossicular chain, is not a difficult surgical problem. In contrast, correction of the more common osseous meatal atresia is technical difficult. The condyle of the mandible is more posterior than normal, and it is in direct apposition to the anterior wall of the mastoid process. The future external auditory canal must be created around its posterior aspect, through the compact and cancellous bone. The descending portion of the fallopian canal is in close proximity to the dissection and may follow an abnormal course. The majority of the patients with moderate external and middle ear malformations (group II) have an osseous type of meatal atresia. Although transmastoid approaches have been used successfully in this and other departments for many years (Nager, 1961; De La Cruz, 1985), anterior atticotomy without complete mastoidectomy is now recommended (Jahrsdoerfer, 1978; Mattox and Fisch, 1986). The advantage of this technique is that it avoids an open mastoid cavity. However, if the surgeon has any hint of becoming disoriented during the surgery, the additional landmarks of the sigmoid sinus and sino-dural angle should be sought immediately. The facial nerve is more likely to be in an abnormal position as it progresses distally; therefore, the dissection always starts with superior landmarks and proceeds inferiorly with caution.

Incision. If the auricle is in its normal position and only minimally deformed, the initial incision is similar to the usual endaural incision. More commonly, however, the auricle is anteriorly displaced over the site of the proposed external auditory canal. In this situation the rudimentary pinna is included in a "Z-plasty" incision. The pinna is encompassed in the inferiorly based anterior flap. The two flaps are elevated to expose the mastoid cortex, root of the zygoma, and cribriform area. Superiorly the temporal line may be seen, indicating the level of the floor of the middle cranial fossa. During closure the inferior flap is transposed posteriorly to line the posterior edge of the canal and provide a more normal position of the pinna behind the external auditory canal. The superiorly based posterior flap will be used to line the anterior margin of the canal.

Exposure of the Epitympanum. The middle fossa dura is identified by drilling the cribriform area posterior to the condyle. The middle fossa dura is followed to the anterior epitympanic space until the ossicular mass (head of the malleus and body of the incus) is identified. The pneumatic cells of the sino-dural angle and mastoid are left undisturbed. The facial nerve is safe at this point of the dissection because it is always found medial to the ossicular mass.

Mobilization of the Ossicular Chain. All bone lateral to the ossicles is carefully removed until the complete malleus handle and incus are exposed. The ossicles are often in an abnormally anterior position. The bone over the attic is removed; however, the incus is left in place and the posterior ligament of the incus is left intact. If the mucoperiosteum beneath the atresia plate can be preserved, it provides additional support for the fascia graft; however, frequently the overlying bone cannot be dissected from it. If it is preserved, it must be reflected anteriorly to expose the ossicular chain. There usually is fixation of the malleus, either by an attachment of the tip of the long process to the atresia plate or of the head to the anterior wall of the tympanum. The ossicular chain is mobilized by separating the attachment of the ossicular mass from the atresia plate with a diamond burr. The incudostapedial joint is separated to prevent vibrations from the burr from being transmitted to the inner ear. If the incudostapedial joint cannot be seen, the ossicular mass may be stabilized with an alligator forceps while it is freed from the atresia plate. When the malleus and incus have been freed from all abnormal adhesions, the incudostapedial articulation is examined, and the mobility of the stapes within the oval window is tested. The niche of the round window, particularly its configuration, is examined, and the oval window membrane is inspected. If the vestibular and cochlear fenestrae function properly, no further surgery is necessary within the middle ear.

Annular Sulcus. A new annular sulcus is drilled in the bony external auditory canal slightly medial to the level of the ossicles. This annular sulcus is important to stabilize the fascia graft in the desired position and prevent lateralization.

The lateral portion of the external auditory canal is enlarged to as wide a funnel as possible to prevent postoperative stenosis. The limits of the enlargement are the temporomandibular joint anteriorly and the middle cranial fossa dura superiorly. Extensive opening of the mastoid air cells is avoided. Enough rim of bone should be left around the margins of the canal to allow placement of drill holes for stabilizing sutures.

Fresh temporalis fascia is harvested and placed directly over the mobilized ossicular mass. It is important that the bony annular sulcus is medial to the ossicular mass so that the grafted tympanic membrane does not lateralize from the ossicular mass.

Grafting of the External Auditory Canal. Several drill holes are made in the bony margin around the new bony external canal. Sutures (4-0 Vicryl) are placed through these holes and secure the skin flaps around the external auditory meatus. Caution should be taken not to damage the middle cranial fossa dura while drilling these holes on the superior margin of the meatus.

Split-thickness skin grafts are obtained free-hand or with a dermatome from the inner surface of the upper arm. The grafts are stabilized by placing the epithelial surface against a piece of linen soaked in antibiotic ointment. The skin graft and linen can then be easily handled and cut in any size or shape needed to line the canal. The entire canal, including the fascia graft, is lined with the split-thickness skin grafts.

Lateral Skin Flaps. The anterior margin of the new external auditory canal is rimmed with superiorly based postauricular skin flap. This flap is fixed in position with sutures that pass through the holes around the meatus. The posterior flap with the microtic pinna is

replaced along the posterior margin of the new external auditory meatus and stabilized with sutures through the holes in the margin of the meatus. The canal is packed with gelfoam and gauze packing. No hard acrylic stents are used because they have a tendency to aggravate, rather than prevent, cicatricial scar formation.

Obliteration of the Oval Window. Rarely the oval window may be absent or incompletely differentiated, with resulting fixation of the stapes. If for some reason a normally functioning oval window cannot be established, the operation is terminated.

Complications. In none of the 23 patients operated on at this institution have there been any facial nerve complications; however, the nerve was found in an abnormal location in three.

Lateralization of the tympanic membrane graft is a common cause of failure (De La Cruz et al, 1985). However, the careful drilling of a new annular sulcus medial to the level of the ossicular mass, as described above, has prevented problems with lateralization of the graft.

Stenosis of the external auditory canal is another common postoperative complication (Schuknecht, 1975; De La Cruz et al, 1985). Although postauricular flaps have been used to line the new external auditory canal (Friedberg, 1977), thin split-thickness grafts are preferred to line the cavity, as has been found in our experience and that of most others (Bellucci, 1981). , Hard acrylic stenting materials are not used because they are likely to stimulate rather than prevent cicatricial constriction.

Severe External and Middle Ear Malformations (Group III)

Operative procedures to improve hearing in this group of malformations are hardly ever undertaken, and then only after careful evaluation of the anatomy of the middle ear, inner ear, and facial nerve. The low level of the tegmen, lack of cellular development, anterior position of the lateral sinus, and hypoplasia of the middle ear ossicles not only render the operation difficult but in most instances also make it technically impossible. Whereas some authors have advocated the fenestration of the lateral semicircular canal, we prefer amplification in this group of malformations.

Reconstruction of a Congenitally Malformed or Absent Auricle

Construction of the external meatus, middle ear, and sound conducting system should be undertaken only by an otologist who is experienced in all aspects of ear surgery and is familiar with the principles of sound conduction. If the auricle is of normal size and configuration, and there is only meatal atresia, the aural surgeon merely has to create an adequate external meatus. However, if the auricle is moderately or severely malformed, reconstructive surgery of the pinna becomes an important part of the reconstruction. Rudiments of the auricle, as encountered in microtia, can be utilized and noticeably improved in structure and appearance by relatively simple plastic procedures.

Reconstruction of the entire auricle with scalp flaps, tube grafts, and implants of autogenous rib cartilage not only requires many procedures but often fails to fulfill aesthetic

expectations. Results are often less acceptable than those achieved with a good prosthesis. A notable exception to this are the marvelous results achieved by Brent (1980; 1983). The technical problems are entirely different from those associated with correction of atresia, and concern primarily plastic surgeons. These techniques, their indications, and results are outside the scope of this chapter. These cases, especially unilateral cases, should be managed in concert with an appropriately trained plastic or facial plastic surgeon. Correction of the auricular deformity, should be considered at age 5 to 6 to improve the cosmetic appearance when the child is first entering school.