OTOLARYNGOLOGY

Volume 2

EAR

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PREFACE

Volume 2, *Ear,* is intended to be used in conjunction with the other two volumes of OTOLARYNGOLOGY and especially Volume 1, Basic Sciences and Related Disciplines. Sections of related interest in Volume 1 include Embryology and Anatomy, Physiology, Histology and Pathology, Physical Diagnosis, Audiology and Communication Disorders, and so forth. An example of a topic of interest in Volume 3 is Otoplasty.

This volume is devoted to clinical otology with an emphasis on pathophysiology. Although ear diseases are emphasized, principles and aspects of surgical otology are also presented.

These three volumes of OTOLARYNGOLOGY are graduate textbooks and Volume 2, *Ear,* demonstrates very well the variety of opinions that can exist regarding etiology and management of otologic disease. Some overlap between various chapters written by different authors was considered desirable bv the editors.

Otology has made spectacular advances in the past two decades. This scientific and clinical progress has resulted from the genius of famous contemporary otologists and scientists in related disciplines on both sides of the Atlantic and Pacific Oceans, as well as the technical contributions made largely with the aid of the operating microscope. The current status of our knowledge, including important contributions in tympanoplasty, stapedectomy, surgery for Meniere's disease, and acoustic tumor removal, is recounted in this volume. We are well advised, however, to recall that such progress rests on a sound historical foundation, the scientific contributions of the famous European otologists around the turn of the century.

With this rich historical background, we can optimistically look to the future for a scientific renaissance in otology and the solution of the many clinical otologic problems which are so commonly manifest in society.

MICHAEL M. PAPARELLA, M.D.

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DISEASES OF THE EXTERNAL EAR

Chapter 1

CONGENITAL AURAL ATRESIA: ANATOMY AND SURGICAL MANAGEMENT

by George T. Nager, M.D.

Congenital aural atresia comprises a group of relatively frequent uni- or bilateral malformations of the external and middle ear. These malformations may, in some instances, be associated with anomaly, or even agenesis, of the inner ear (Altmann, 1965a).

Aural atresia has long been recognized. For instance, several of the prehistoric skulls found in this country reveal this defect (Hrdlicka, 1933).

Moderate and severe forms of aural atresia are encountered in about one out of 10,000 to 20,000 individuals (Mündnich, 1966). In the period 1950 to 1958, 26 of 289,444 (1:11,000) children born in 35 obstetrical clinics of the German Federal Republic revealed an isolated ear deformity (Kleinsasser and Schlothane, 1963). Bezold (1926) found this malformation in 11 of his 20,408 patients with otologic problems.

Males are affected as frequently as females. Atresia is unilateral four times as often as bilateral. Unilateral atresia, for reasons still unexplained, seems to affect the right ear more often than the left (Altmann, 1965a).

At one time aural atresia was considered a well defined and independent entity. As time went on, however, it became obvious that it often represents but one of several malformations involving the derivatives of the posterior segments of the first and second branchial arches. It also became apparent that it can be associated with malformation syndromes affecting other parts of the skeleton, that it can occur in certain chromosomal aberrations, that it may in some instances be inherited and that it may be found in embryopathies.

ATRESIA IN ASSOCIATION WITH OTHER MALFORMATION SYNDROMES

Aural atresia may be observed with cranial, facial, mandibulofacial, mandibular and acrofacial dysostoses. Among these dysostoses, the craniofacial dysostosis (Crouzon's disease) (Crouzon, 1912) and the mandibulofacial dysostosis (Treacher Collins and Franceschetti syndrome) (Treacher Collins, 1900; Franceschetti et al., 1944, 1949) are best known to plastic surgeons and otolaryngologists. The characteristics of craniofacial dysostosis include cranial synostosis involving the coronal, sagittal and lambdoidal sutures; hypertelorism, exophthalmus and external strabismus; parrot-beaked nose, short upper lip, hyoplastic maxilla and a relative mandibular prognatism; frequent association with microtia, aural atresia, anomalies of the middle ear and hearing loss; and autosomal dominant inheritance.

Mandibulofacial dysostosis includes the following features: antimongoloid slant of the palpebral fissure, coloboma of the lower eyelid, absence of puncta and lack of cilia in the medial third of the lower eyelid; hypoplasia of the malar bone, infraorbital ridge and zygomatic arch; hypoplasia of the mandible, micrognathia and macrostomia; microtia, aural atresia, malformations of the middle ear and hearing loss; and dominant transmission.

The following anomalies may be associated with mandibulofacial dysostosis: clefts of the lip and palate, malocclusion, and nasal deformities. In the mandibulofacial dysostosis (Treacher Collins' type) with limb anomalies (Nager's acrofacial dysostosis) (Nager and de Reynier, 1948) the limb deformities consist of absence of radius, radio-ulnar synostosis and hypoplasia or absence of the thumbs. The abnormalities of the auditory system include microtia and aural atresia, occasional anomalies of the inner ear, and hearing loss. All reported cases are sporadic, but one suggests a dominant mutation.

Aural atresia can also be associated with facial, labial and palatal clefts; hypoplasia of the palate and the parotid gland; agenesis of the anterior pillar and tonsils; teratogenic tumors of the tonsils; and aberrant thymus tissue in the middle ear (Altmann, 1965a). Aural atresia can occur with other skeletal anomalies (for instance, poly- or hypodactyly), visceral anomalies, Fanconi's recessive panmyelopathy, oculoauricular dysplasia, and so forth (Goldenhar, 1952; Schwarz and Becker, 1964; Altmann, 1965a).

According to some authors, the damage to the posterior (rhomboencephalic) cranial organization center that is held responsible for cranial malformations and aural atresia must have taken place much earlier than was hitherto assumed. In many instances there is evidence of simultaneous, but lesser, damage to the anterior cranial organization center. The damage may result from a genetic defect or a biochemical disorder (Altmann, 1965a).

ATRESIA IN ASSOCIATION WITH CHROMOSOMAL ABERRATIONS

Aural atresia may be observed with chromosomal aberrations; for instance, trisomy of the chromosome groups 13, 14, 15, 17, 18 and 21 (Ferguson-Smith, 1961; Schwartz and Becker, 1964). It can be associated with Turner's syndrome, which is characterized by the XO genotype (Jones, 1967). In Turner's syndrome the female patient exhibits the two cardinal findings: sexual infantilism and retardation of growth. The patient may reveal a host of additional somatic anomalies. Those of special interest to the otolaryngologist are misplacement and malformation of the ears with impairment of hearing, micrognathia, high palatal arch, epicanthal fold and mental retardation.

HEREDITY

For many years aural atresia was considered to be mostly or exclusively a hereditary disorder. Certain forms of atresia undoubtedly reveal a regular dominant autosomal inheritance (Johannesmeier, 1948; Hanhart, 1949; Goldenhar, 1952; Schwarz and Becker, 1964). In one family, for instance, microtia manifested itself through four generations exclusively in males, suggesting a Y-chromosomal inheritance. Another example of dominant transmission with variable penetrance is the syndrome of hereditary ear malformations and conductive deafness described by Wildevanck (1962) and McLaurin et al. (1966). The characteristics of this syndrome include uni- or bilateral flat ears in about 40 per cent, preauricular pits in about 40 per cent, preauricular appendages in about 20 per cent, conductive deafness in about 15 per cent and microtia, aural atresia or preauricular fold in about 20 per cent of affected individuals. The syndrome of familial malformed low-set ears and aural atresia, plus mental retardation is found in 50 per cent of affected individuals (Mengel et al., 1967). Thus, the mode of inheritance, in the genetically controlled form of aural atresia, may be either recessive or dominant.

EMBRYOPATHIES

Aural atresia does not often affect more than one member of a family; therefore, there must be other etiologic factors that can induce this anomaly. For instance, during the past two decades it has become obvious that a medicament (thalidomide) and the rubella virus can also produce the syndrome of aural atresia.

Thalidomide

Aural atresia can be induced by the tranquilizer thalidomide (German: Contergan). Ingestion of thalidomide may produce a host of malformations which 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 Germany during that period about 7000 children were born with malformations of the limbs

(Weicker et al., 1962). If malformations of the hand and fingers were included, the actual number of affected children would be 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 agenesis of the inner ear (Lenz and Knapp, 1962). According to Lenz (1963), ingestion of thalidomide during the 35th and 36th day, and occasionally during the 37th 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 38th and 45th days. The infrequent duplication of the thumb was seen following ingestion prior to the 38th day. Partial or total agenesis of both arms corresponded to the period extending from the 39th to the 44th days. Cardiac malformations, duodenal atresia and congenital subluxation of the hip occurred with ingestion between the 39th and the 45th days. Ingestion around the 50th 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 ingestions, gave birth to normal children. This proves that not all women were sensitive to this teratogenic drug.

Maternal Rubella

The introduction of polytomography led to a re-evaluation 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 supplied increasing 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).

Other Factors

Note that other factors such as local hypoxia (of the ovum or of the fetus), roentgen rays, and chemical compounds and viruses that have shown a teratogenic effect in animal experiments may also influence the normal development of the middle and inner ear in man.

CLASSIFICATION OF EAR MALFORMATION

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

Minor Malformation (Group I)

In this group the auricle and the external auditory canal can be normal, but they frequently reveal some intimation of malformation. Occasionally the canal 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 wide canal; in general, however, it is thickened and appears opaque. The handle of the malleus is often clumsy, may be greatly deformed and usually is in an abnormal position. The tympanic cavity is usually hypoplastic, and the middle ear structures (ossicles, muscles and nerves) display malformations of varying degrees. In a rare instance the inner ear is abnormal, and the lateral and even the remaining semicircular canals may be aplastic (Mündnich, 1966). Minor ear malformations are commonly seen in craniofacial dysostosis (Crouzon, 1912).

Moderate Malformation (Group II)

This group comprises the majority of ear deformities. The auricle is rarely normal and is usually represented by a rudimentary soft tissue structure. The canal is either partially or totally aplastic, or it may end blindly with one or occasionally two fistulous tracts leading toward a rudimentary tympanic membrane. Such fistulous tracts can be associated with a cholesteatoma. The tympanic bone can be either present or absent. If present, it is severely malformed and usually represented by an osseous plate whose central portion may consist of connec-

tive tissue. This atresia plate forms the lateral wall of the middle ear cavity, which is generally reduced in size. The middle ear structures display malformations of varying degrees. If the tympanic bone is absent, the upper portion of Reichert's cartilage may show anomalies both in configuration and in position and can form the principal portion of the atresia plate. The hyperplasia of Reichert's cartilage may involve the laterohyale and tympanohyale. Since both latero- and tympanohyale participate in the formation of the lower portion of the fallopian canal, the facial nerve may, when they are malformed, take an abnormal course and leave the middle ear much more anteriorly and superiorly (Altmann, 1965a).

Whenever the atresia plate is formed by Reichert's cartilage, the chorda tympani nerve courses laterally to it (Marx, 1922; Altmann, 1965a). The atresia plate, therefore, is medial from the normal location of the tympanic membrane. Thus, the middle ear cavity represents only the medial half of a normal cavum tympani.

If the tympanic bone is absent and the upper portion of Reichert's cartilage is barely hyperplastic, bony processes from the temporal squama and hypotympanum join to form a lateral bony wall of the tympanic cavity. The chorda tympani nerve, here too, courses laterally to the atresia plate. The atresia plate is again medial from where the tympanic membrane would normally be encountered, but not so much medial as when formed by Reichert's cartilage (Altmann, 1965a). An antrum and periantral cells may be either present or absent, depending upon the degree of pneumatization. Pneumatization generally is inhibited but, when well developed, it may indicate fairly well differentiated middle ear structures.

Severe Malformation (Group III)

In this group the auricle generally is severely malformed; it can even be totally absent. The canal is aplastic. Strands of connective tissue and epidermis, or a fistulous tract, are occasionally encountered as a rudiment of an external meatus. Absence of pneumatization is characteristic. Antrum and middle ear may be absent or may be represented by a slitlike lumen. Such a rudimentary middle ear may contain mesenchyme, connective tissue or osseous bridges. Anomalies of the facial nerve are the rule. The middle ear ossicles are frequently absent. It is also in this group that the malformations of the inner ear are the most frequently encountered. They reveal great variation in regard to extent and structures involved. They are frequently associated with the severe cranial malformations regularly encountered in mandibulofacial dysostoses (Treacher Collins and Franceschetti), (Altmann, 1965a). These severe forms of ear malformations have been particularly common after thalidomide ingestion.

MALFORMATIONS OF THE MIDDLE EAR STRUCTURES (Fig. 1)

Ossicular malformations are regularly encountered in Groups I and II. In Group III the ossicles are often absent. Malleus and incus frequently form a conglomeration, or have bony unions with the atresia plate and the walls of the epitympanic recess. Not rarely 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, totally 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 and, in a rare instance, it may bifurcate. The greater and the lesser superficial petrosal nerves and the chorda tympani nerve can be absent (Altmann, 1965a).

The eustachian tube may be normal, deformed, hypoplastic or even totally absent.

MALFORMATION OF THE AURICLE

Developmental anomalies of the auricle may be conveniently divided into the following three classes: minor malformation, microtia (Marx's (1922) types I to III), and anotia (Fig. 2).

Minor Malformation

This class comprises the variations involving the relief and finer differentiation of the auricle.

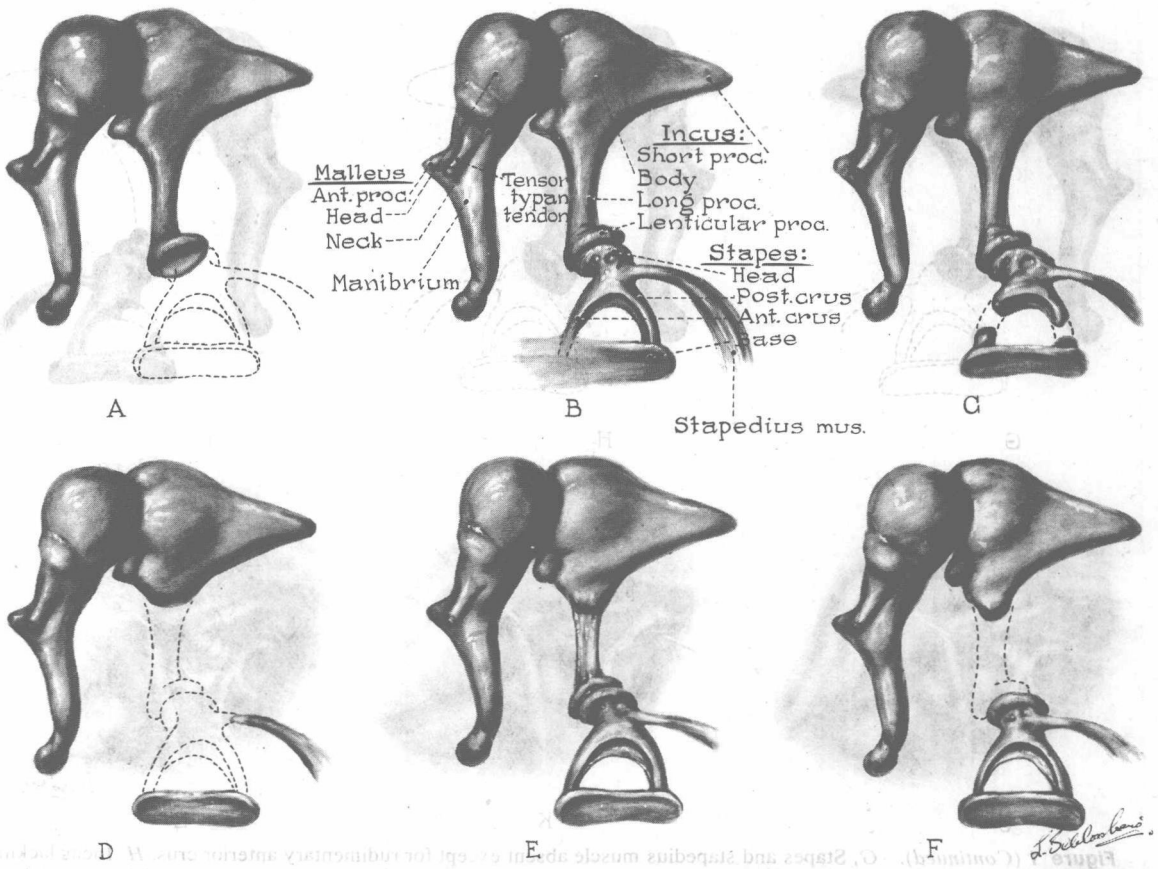

Figure 1. Auditory ossicles of aberrant form encountered in malformation of the middle ear. *A*, Stapes and stapedius muscle absent. *B*, Incomplete differentiation of oval window with anterior stapes fixation. *C*, Stapes having a partial anterior and posterior crus. *D*, Incus lacking long crus; stapes lacking crural arch. *E*, Incus with long crus consisting of strand of connective tissue but normal lenticular process. *F*, Incus lacking entire long crus.

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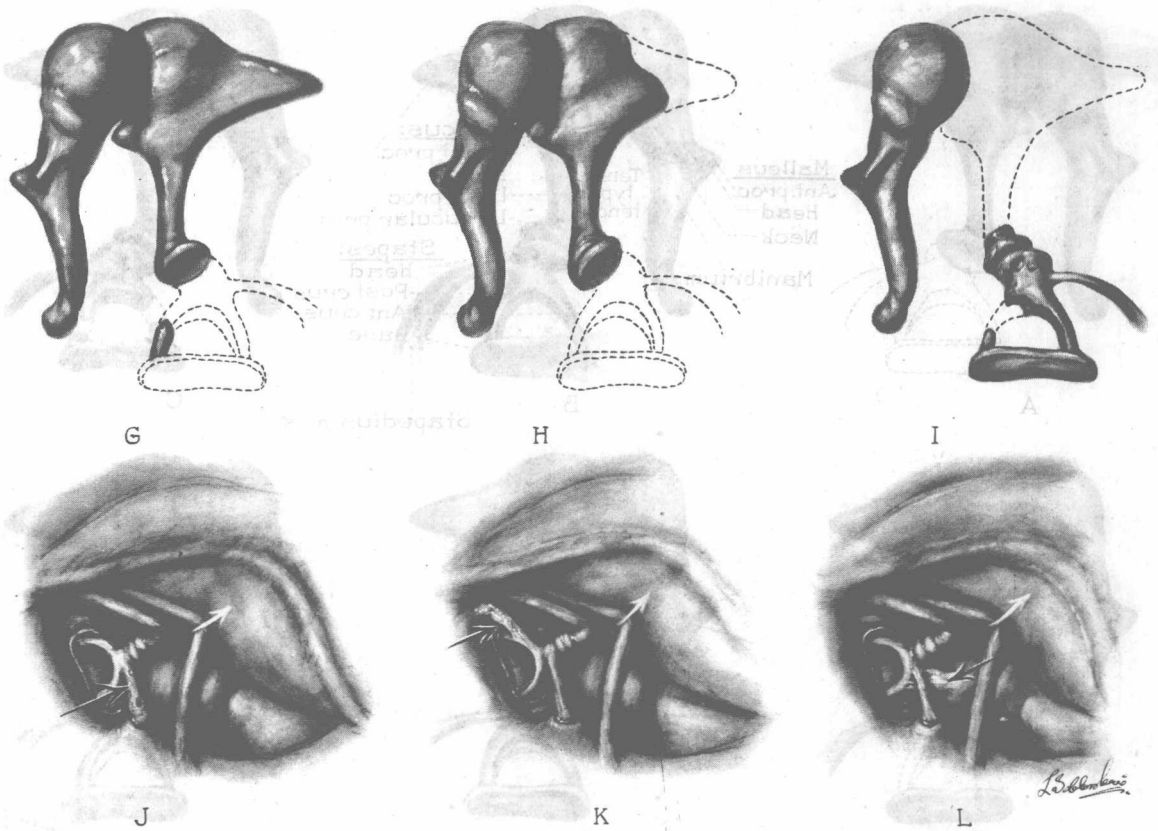

Figure 1 *(Continued).* *G*, Stapes and stapedius muscle absent except for rudimentary anterior crus. *H*, Incus lacking short process; stapes and stapedius muscle absent. *I*, Incus absent except for lenticular process; stapes having a partial anterior crus. *J*, Osseous bridge connecting stapedial arch with pyramidal process. *K*, Osseous bridge extending from anterior crus to fallopian canal. *L*, Osseous bridge connecting stapedial arch to promontory.

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