To

Rosalind

DISEASES

OF THE

EAR, NOSE, AND THROAT

IN CHILDREN

by

T. G. WILSON,

B.A., M.B., Litt.D., F.R.C.S.I., Hon. F.R.C.S. Edin., Hon. R.H.A., M.R.I.A.

Past President, Royal College of Surgeons in Ireland. Past President, Collegium Otorhinolaryngologicum Amicitiae Sacrum. Member, James IV Association of Surgeons. Surgeon-in-charge, Ear, Nose and Throat Departments, Royal City of Dublin Hospital, Dr. Steevens' Hospital, and The National Children's Hospital, Dublin. Honorary Member of the Scottish Otolaryngological Society.

With Illustrations by the Author

SECOND EDITION

WILLIAM HEINEMANN · MEDICAL BOOKS · LTD

LONDON

FIRST PUBLISHED 1955
SECOND EDITION 1962

Printed in Great Britain by
The Whitefriars Press Ltd., London and Tonbridge

PREFACE

THIS is, I believe, the first text-book to be written on diseases of the ear, nose, and throat in children, although Alexander of Vienna published a book on diseases of the ear in children in 1917. Since then our knowledge of the otolaryngological problems of childhood has greatly increased, and it is now perhaps timely that such subjects as congenital defects, infantile otitis media, the auditory and vocal rehabilitation of deaf children, and infections of the respiratory tract should be considered at more length than is possible in text-books dealing with otolaryngology as a whole. The character of otolaryngology has changed greatly in the last twenty years. The decline in the virulence of bacterial infections has diminished the incidence of acute sepsis, while that of acute diseases of virus origin and of chronic suppurative ear conditions seems to have increased considerably. Within otology the new science of audiology has developed : it may eventually prove necessary to ensure that the tail does not wag the dog. It is, of course, important that otolaryngologists and pædiatricians should have a sound knowledge of the problems involved in defective hearing and speech so that they can understand and direct the education and rehabilitation of deaf children. The advent of antibiotic drugs has not only controlled many infections but by preventing post-operative infection has proved a valuable adjuvant to surgery. In consequence, and contrary to what was at one time thought probable, new and exciting surgical fields are being explored. The fenestration operation in certain cases of microtia and the repair of congenital œsophageal atresia are cases in point.

I have throughout assumed that my readers are familiar with the basic facts of anatomy and physiology, and I have as far as possible confined myself to problems peculiar to children. For this reason I have not, for instance, attempted to give a complete description of the treatment of chronic suppurative otitis media and its complications, which is, of course, fully dealt with in many excellent text-books. On the other hand, I have devoted some space to allergy, which is common in children and is a difficult subject to compress. The result as a whole will, I hope, be of interest not only to otolaryngologists and pædiatricians, but also to general practitioners, whose work is so largely concerned with children.

T. G. W.

Dublin, 1955.

PREFACE TO SECOND EDITION

THE issue of a new edition of this book has afforded an opportunity to revise and enlarge it and to delete some material which has not stood the test of time. New sections have been added to the first chapter, on congenital abnormalities of the ear, although congenital abnormalities of the internal ear are still more fully dealt with in Chapter 6 in order to consider them in their clinical context. This section, which it is hoped will be of particular value to postgraduate students and candidates for higher degrees, has been considerably enlarged, as also has that on testing the hearing in infancy and childhood which now has a chapter to itself. The section on cholesteatosis of the middle ear has been largely re-written in the interests of clarity. Other parts which have been considerably added to are those dealing with osteomyelitis of the maxilla in infancy, speech defects, branchial cysts and diseases of the thymus gland. The section on neonatal asphyxia, a condition which is still of interest to the laryngologist although now most often treated by anæsthetists, has also been re-written. The guillotine operation for tonsillectomy is still practised in many large centres in these islands and on the Continent and the description of it has therefore been retained, with what is hoped is a reasoned appraisal of its advantages and disadvantages compared to the dissection operation, which is of course also fully described.

It may be emphasized that this book is directed more to clinicians than to audiologists or speech-therapists, although it is hoped that all those interested in diseases of the ear, nose and throat in children will continue to find it useful.

T. G. W.

Dublin, 1962.

Acknowledgements

Professor Wilfred Gaisford of the Department of Child Health of the University of Manchester has read the whole book and has made many important corrections and given me some excellent advice. Dr. Brian Pringle has once more been of the greatest assistance in correcting the proofs. Dr. A. R. S. Jessop has advised me about anæsthesia and Mr. Keith Shaw, F.R.C.S.I. has read the final chapters. To these gentlemen and once more to Mr. J. Johnston Abraham and Mr. Owen R. Evans my grateful thanks are due.

T. G. W.

PREFACE TO SECOND EDITION

THE issue of a new edition of this book has afforded an opportunity to revise and enlarge it and to delete some material which has not stood the test of time. New sections have been added to the first chapter, on congenital abnormalities of the ear, although congenital abnormalities of the internal ear are still more fully dealt with in Chapter 6 in order to consider them in their clinical context. This section, which it is hoped will be of particular value to postgraduate students and candidates for higher degrees, has been considerably enlarged, as also has that on testing the hearing in infancy and childhood which now has a chapter to itself. The section on cholesteatosis of the middle ear has been largely re-written in the interests of clarity. Other parts which have been considerably added to are those dealing with osteomyelitis of the maxilla in infancy, speech defects, branchial cysts and diseases of the thymus gland. The section on neonatal asphyxia, a condition which is still of interest to the laryngologist although now most often treated by anæsthetists, has also been re-written. The guillotine operation for tonsillectomy is still practised in many large centres in these islands and on the Continent and the description of it has therefore been retained, with what is hoped is a reasoned appraisal of its advantages and disadvantages compared to the dissection operation, which is of course also fully described.

It may be emphasized that this book is directed more to clinicians than to audiologists or speech-therapists, although it is hoped that all those interested in diseases of the ear, nose and throat in children will continue to find it useful.

T. G. W.

Dublin, 1962.

Acknowledgements

Professor Wilfred Gaisford of the Department of Child Health of the University of Manchester has read the whole book and has made many important corrections and given me some excellent advice. Dr. Brian Pringle has once more been of the greatest assistance in correcting the proofs. Dr. A. R. S. Jessop has advised me about anæsthesia and Mr. Keith Shaw, F.R.C.S.I. has read the final chapters. To these gentlemen and once more to Mr. J. Johnston Abraham and Mr. Owen R. Evans my grateful thanks are due.

T. G. W.

PART I

DISEASES OF THE EAR

CHAPTER 1

CONGENITAL ABNORMALITIES

General Etiological Considerations

THERE is no need to stress the importance of congenital defects. Almost a quarter of the human race is said to die from congenital abnormalities at or before birth, and a proportion of the survivors are so handicapped as to be a liability to the community.

The causes of congenital abnormalities may be either hereditary or environmental.

Hereditary diseases are those inherently present in the individual from the moment of conception, and are due to a defect in the parent's germ-plasm. Environmental diseases are due to bacterial or other infections, trauma, toxins or nutritional deficiencies.

Hereditary Congenital Abnormalities. The transmission of hereditary defects, into which category practically all congenital abnormalities were placed until recently, is generally thought to be governed by Mendel's theory of genetics. In brief, it is considered that each individual has a specialized germ-cell or sex cell, the chromosomes of which contain the genes which determine the characteristics of the individual. This germ-cell may be male (the spermatozoon) or female (the ovum) and from the union of these two is formed the fertilized ovum which in time becomes a new individual. Thus half the chromosomes of the fertilized egg are derived from each parent. In this way the genes of such characteristics as the colour of the eyes and hair, the blood antigens, and physical abnormalities are transmitted to the offspring. Whether or not a particular quality is transmitted or not depends upon whether the gene is dominant ; or, if recessive, whether it is present in the chromosomes of both parents. In the case of deaf mutism consanguineous marriages have a remarkable effect on its incidence. According to Ballenger (1947) forty-seven marriages between blood-relations produced seventy-two deaf-mutes.

In the past many people thought that all congenital defects were transmitted in this way, and in consequence that the only way to avoid their occurrence was by selective breeding. While it is not disputed that hereditary factors are frequently the cause, the work of Gregg (1941) and others has conclusively shown that non-genetic causes are of greater importance, if only because they are preventable.

Environmental Abnormalities. Environmental causes may be infective, endocrine, mechanical, physical, or nutritional.

INFECTIVE. Gregg (1941), an ophthalmic surgeon working in Sydney, New South Wales, noticed a marked increase in the number of cases of congenital cataract attending his clinic during the years immediately following the pandemic

outbreak of rubella in Australia in 1939, when a large number of adults had contracted the disease. He suggested that rubella contracted by mothers during the early part of their pregnancy was the direct cause of congenital cataract and heart lesions. This hypothesis was at first received with some scepticism; however, it is now fully confirmed, and while the risk that rubella in early pregnancy will cause malformation in the infant is said to be much less than originally suspected, " deafness should be specially looked for in these cases " (Charles, 1960). Other diseases, including the common cold, morbilli, influenza, and pneumonia, may have a similar effect (see p. 79).

Rubella, and presumably also other infectious and injurious agents, attacks the part of the embryo which is most actively developing at the time of infection. In consequence, the type of defect varies with the period of gestation. Thus, Swan (1949) and others have found in a large series of cases in Australia that the average period of pregnancy for congenital cataract was 1·4 months, for heart lesions 1·5 months, and for deaf-mutism 2·3 months. This has been confirmed by experimental work.

Syphilis is also a common cause of congenital deafness. The most severe forms of aural syphilis occur during intra-uterine life, the new-born infant showing all the signs of congenital deafness and a static labyrinth which is not excitable. The proportion of cases of congenital syphilitic deafness varies amongst deaf-mutes, and has been variously assessed at from 3·5 to 25 per cent., the latter figures being arrived at by the Wassermann test, the former by clinical examination (Ballenger, 1947) (see p. 77).

KERNICTERUS may cause congenital defects (see p. 76).

ENDOCRINE. There is evidence of increased incidence of congenital defects among the children of diabetic mothers (Skipper, 1933; Hurwitz and Irving, 1937).

MECHANICAL. Denis Browne (1951) has put forward a hypothesis (the " compression baby " theory) that fœtal defects such as talipes equinovarus, spina bifida, and congenital dislocation of the hip are caused by abnormal mechanical conditions *in utero*. Some consider abnormal position of the fœtus to be the cause (Chapple, 1945). Browne considers hydraulic pressure, in addition to malposition and mechanical pressure, to be causative. In cases of maternal hydramnios, œsophageal atresia should always be looked for. There is nothing new about this theory; in *The Acts of the Apostles* iii, 2, we read of " a certain man lame from his mother's womb."

PHYSICAL FACTORS such as radiation of the maternal pelvis with X-rays or radium may produce abnormalities.

NUTRITIONAL FACTORS have been held to cause an increased incidence of abnormalities after periods of food shortage induced by war and famine.

A curious phenomenon was recorded by Record and McKeown in Birmingham in 1951. They noted that a higher proportion of anencephalic monsters was born in that city in December than in June. Doll, Hill, and Sakula (1960) found that in Scotland during the eight months from May to December 1958 the number of anencephalic stillbirths was 30 per cent. higher than normal and concluded that this might have been due to infection with the Asian strain of influenza virus.

Embryology

Abnormalities of the middle ear are frequently found in association with meatal atresia, while the internal ear may be normal. This is because the internal ear has a separate development from the external and middle ear. The epithelium of the internal ear is developed from the ectoderm (Holmes, 1949) and is first seen in the 2-mm. embryo as a thickened plate lying on the surface of the head just dorsal to the second branchial cleft. This plate invaginates and forms the otic vesicle, which differentiates into the cochlea and semi-circular canals. At six weeks the cochlea consists only of the cochlear duct, in the form of a short curved tube. At seven weeks it has developed into a single complete turn, and the scala tympani and scala vestibuli have been formed out of the

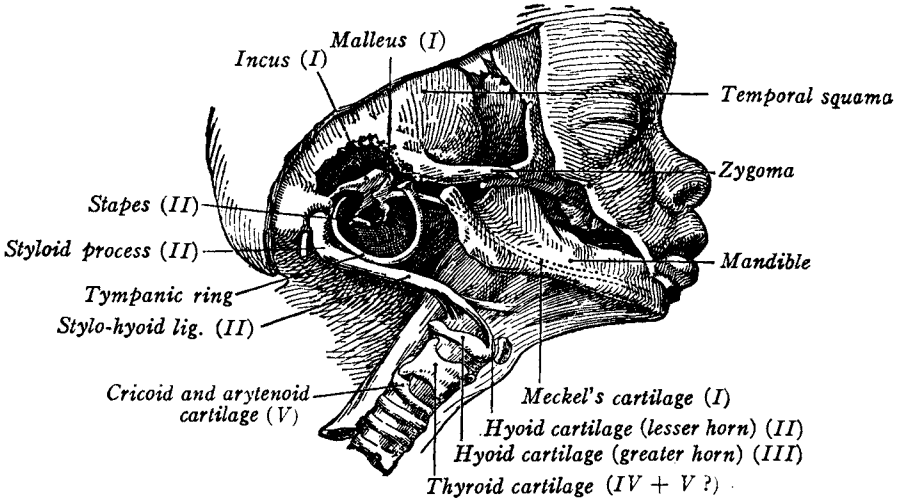

FIG. 1. Derivatives of the human branchial arches, demonstrated in a lateral dissection of the neck.

(*Arey, after Kollman.*)

surrounding mesenchyme. The cochlea reaches its full two-and-a-half turns by the ninth or tenth week and growth continues until the fifth month of pregnancy when it ceases.

The epithelium of the cochlear duct begins to differentiate in the basal turn at about eight weeks, and is followed by the middle and apical turns. The organ of Corti and tectorial membrane are recognizable in the basal turn by the twelfth week and the spiral ganglion and auditory nerve have now linked-up with the sensory end-organ. At four months the cochlea is almost in its adult form and by six months development is complete.

The auditory tube and tympanic cavity are derived from the tubo-tympanic recess between the first and third visceral arches. The inner part of the recess is narrowed to form the auditory tube, while the outer part is subsequently differentiated into the tympanic cavity. The tympanum is surrounded by loose connective tissue in which the ossicles develop : even in adult life the ossicles are enveloped in mucous membrane. The mastoid antrum appears during the

sixth or seventh month : the mastoid air-cells do not begin to form until the end of fœtal life.

The ossicles are developed from the condensed mesenchyme of the first and second branchial arches, the malleus and incus coming from the inner end of Meckel's cartilage, which is contained in the first arch and becomes separated from the mandible when ossification begins ; the stapes is similarly derived from the end of Reichert's cartilage which is contained in the second branchial arch. These facts are of practical importance, as will become evident later.

The external auditory meatus is developed from the dorsal end of the hyo-mandibular cleft. The ventral portion of this groove is the primary meatus, a funnel-shaped tube from which the cartilaginous meatus and a small portion of the osseous meatus are formed (Gray's *Anatomy*). From the funnel-shaped tube a solid core of epidermis extends inwards along the floor of the tubo-tympanic recess. This core hollows out to form the inner portion of the meatus, the secondary

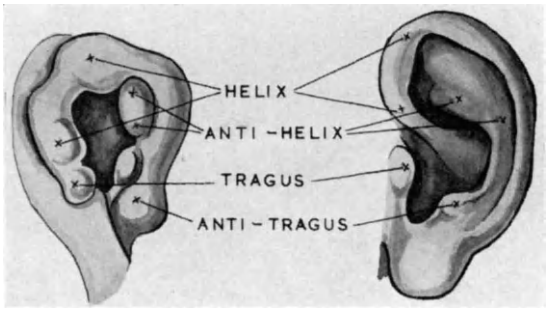

FIG. 2. The development of the human auricle, showing the elevations on the mandibular and hyoid arches which take part in its formation.
(*Copied from Arey, "Developmental Anatomy," IVth Edn. W. B. Saunders Co.*)

meatus, while the blind end forms the outer epidermal layer of the tympanic membrane. The fibrous layer of the membrane is formed by the mesenchyme and the inner layer by the entoderm of the tubo-tympanic recess.

The auricle arises from the outer part of the first pharyngeal, visceral, or hyo-mandibular cleft where six tubercles appear. Of these, three are situated on the mandibular and three on the hyoid arch. Of those on the mandibular area, the most ventral comes to form the tragus, the identity of the remainder being lost. The rest of the auricle develops from the mesenchyme of the hyoid arch. The lobule, corresponding to the end of the auricular fold, is the last part of the ear to develop. In spite of the authoritative statement of Wood-Jones and Wen (1933–34) it is derived partly from the mandibular although principally from the hyoid arch (Wilson, 1959).

Classification. Congenital abnormalities of the ear may be classified as follows :—

(1) Abnormalities of the auricle.
    (*a*) Minor variations in shape.

(*b*) True deformities : vertical and horizontal fissures ; macrotia ; microtia (which may be accompanied by atresia of the external meatus) ; anotia ; melotia ; accessory auricles ; and polyotia.

(*c*) Congenital fistulas ; mandibulo-facial dysostosis ; congenital arterio-venous fistulas.

(*d*) Congenital tumours.

(2) Abnormalities of the external auditory meatus and middle ear. Microtia with atresia of the external auditory meatus.

(3) Abnormalities of the Eustachian tube.

(4) Abnormalities of the tympanic membrane.

(5) Abnormalities of the middle ear and mastoid process.

(6) Abnormalities associated with malformations of the skull.

(7) Dermoids and cholesteatomata of the middle ear and mastoid.

(8) Abnormalities of the internal ear.

Abnormalities of the Auricle

(*a*) **Minor Variations in Shape.** The external ear or auricle varies widely in size and shape as might be expected from the manner of its development. The majority of these variations may be regarded as being within normal limits. Very many boys of preparatory school age have " bat-ears " of truly remarkable dimensions. These appendages, however, like the feet, appear to shrink as the child's development proceeds, and by the time adult life is reached they no longer appear abnormal. These cases do not, therefore, come truly into the category of abnormally large ears, a condition which is known as macrotia.

An unusual anomaly sometimes found is the abnormal adherence of the upper part of the auricle to the head. While the lower half of the auricle stands out normally, the upper part appears to disappear into a pocket of skin, and the retro-auricular fold is absent at this point (Altmann, 1951). The detachment of the auricle from the head posteriorly normally occurs during the fourth month of development ; in these cases for some reason the detachment of the auricle is incomplete in the upper part. As a rule the " relief," or pattern of folds, grooves, and hollows of the auricle is normal.

Variations in the shape and relief of the auricle are, however, numerous. One of the commonest is persistence or presence of Darwin's tubercle, which is homologous to the tip of the mammalian ear and is to be found at about the junction of the upper third and lower two-thirds of the helix. A flattened area above this is also frequently seen : like unduly prominent ears, it is a sex-limited recessive most often found in males. Sometimes the crus helicis extends far back, and may join the antihelix to separate the cymba from the cavitas conchæ. In " Wildermuth's ear " the antihelix is more prominent than the helix ; in " Mozart's ear," which is named after the musician, the enlarged portion of the antihelix is continuous with the helix. Another anomaly is the " cat-ear," in which the auricle forms a hood over the meatus. A vertical cartilaginous bar, the so-called crus cymbæ may form a vertical ridge in the concha. The lobule of the ear may be abnormally large, absent, or adherent : the latter peculiarity

being especially common in women, Germans, negroes, and bushmen. In rare instances the lobule may be bifid, when it must be distinguished from traumatic division due to wearing heavy ear-rings. According to Aprile, Scalori and others this anomaly is due to imperfect fusion of the portions of the mandibular and hyoid arches which join to form the lobule (Wilson, 1959) (*see* embryology). Hypertrichosis of the auricle is a disfiguring abnormality which occurs occasionally, in which, according to Tommasi, the whole of the lower half of the auricle may become invested in long black hair. He described it as having occured in five generations of males. Another type of anomaly is found in " Potter's facies "

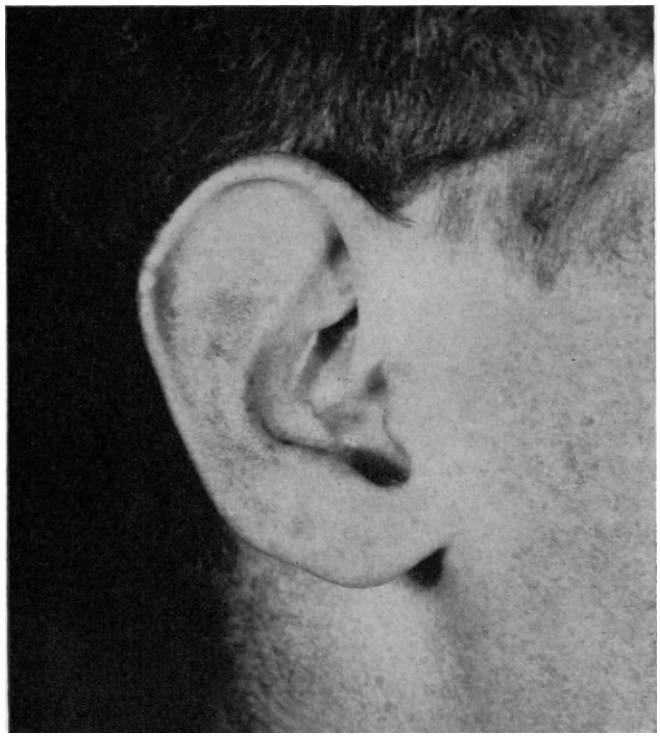

FIG. 3. Darwin's tubercle.

which occurs in association with complete absence of both kidneys (Potter, 1946). In these cases the ears are approximately normal in shape but are large, low-lying and somewhat deficient in cartilage. Dr. Potter states that in these cases complete bilateral renal agenesis can be diagnosed from the facies alone. There is slight increase in the width between the eyes, a prominent fold of skin arising at the inner canthus and proceeding downwards and laterally to below and beyond the outer canthus, accompanied by some flattening of the nose, mild retraction of the lower jaw and large low-lying ears with incomplete cartilage development.

(*b*) **True Deformities of the Auricle.** As Altmann (1951) points out, true malformations are often found in combination with other anomalies of the ear or face.

The auricle may be divided by VERTICAL or HORIZONTAL FISSURES, which are explained by anomalies in the closure of the first branchial cleft and by failure of union of the first and second branchial arches.

MACROTIA is general enlargement of the ear when it is enough to amount to deformity.

MICROTIA may be of much more serious import, particularly when bilateral, as it is often accompanied by atresia of the external auditory meatus and maldevelopment of the middle ear causing severe deafness. Defects of the skull, palate, and mandible frequently co-exist. Marx (1926) distinguishes four degrees

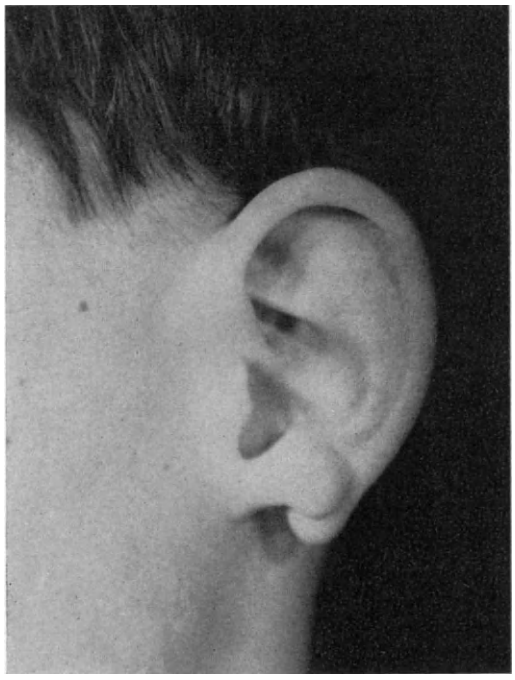

FIG. 4. Bifid lobule due to imperfect fusion of the mandibular and hyoid arches.

of microtia. In the first degree the auricle is normal in shape but smaller than usual : this is of little significance or importance. In the second degree the helix is represented by a longitudinal rudimentary ridge somewhat resembling its proper form. In the third degree the rudimentary ridge is irregular and bears no resemblance to the helix, while in the fourth degree the auricle is completely absent. This is regarded as very rare, only seven cases having been recorded so far.

Complete absence of the auricle (as above) is known as ANOTIA.

MELOTIA is the caudoventral displacement of the auricle, which results from the frequently co-existing hypoplasia of the mandible.

ACCESSORY AURICLES are common and most usually unilateral. They occur

in 1·5 per cent. of the population (Altmann). A degree of hereditary predisposition is present. These tubercles are most often situated just in front of the tragus or ascending part of the helix, and they vary from the size of a match-head to that of a pea or occasionally of a hazel-nut. According to Jones and Wen (1934) they usually lie in front of a line from the temple to the anterior margin of the crus helicis and the inter-space between the latter and the upper part of the tragus, thence into the concha to the external auditory meatus, and out again between the lower part of the tragus and the antitragus to the junction between the concha and the cheek. This line corresponds to the line of junction between

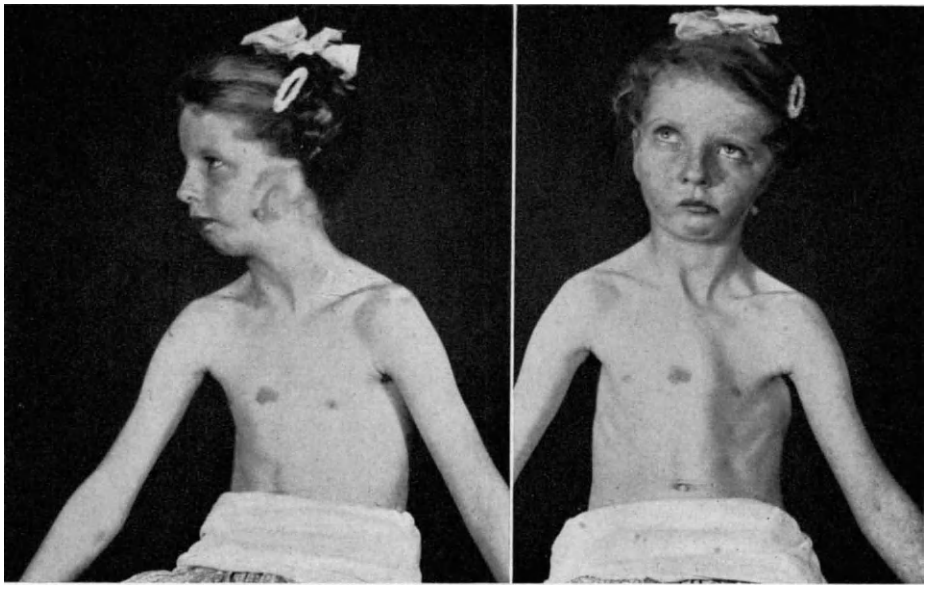

FIG. 5. Dysostosis mandibularis in a girl of eight years, showing the associated microtia and macrostomia. Agenesis of the left lung causing deformity of the chest wall is also present. See also Fig. 129. (Author's case.)

the hyoid and mandibular folds, and the accessory auricles result from excessive growth of the mandibular folds in this line. Auricular appendages in this area are usually found in association with normal ears. When, however, these appendages are found farther out in the cheek they are often associated with microtia, melotia, or transverse fissure of the cheek. They are evidently due to disturbances in the union of the maxillary and mandibular processes of the first branchial arch. Auricular appendages normally contain elastic cartilage which may extend deep into the tissues.

POLYOTIA is the occurrence of more than one auricle on the same side. In certain rare cases auricular appendages are grouped together so as to resemble several separate if deformed auricles ; this, however, is not true polyotia. Bol and de Kleyn (1918) have reported what is probably the only true case so far recorded. This occurred in a baby who possessed two well-formed auricles facing each other like mirror-images along a line extending from the incisura

anterior to the incisura antitragica. On the other side two auricular appendages were present beside a normal auricle.

Treatment. The ears of babies often appear to be unduly prominent before the hair of the head has grown, and young mothers are apt to be very sensitive about the appearance presented. Consequently they very often press for active treatment, but in most cases this should be withheld. Babies' ears can be flattened by constant bandaging, but this is a wearisome and cruel procedure which is seldom pursued to a successful conclusion. It is unwise to make a diagnosis of macrotia until growth has ceased, and plastic surgery should not be contemplated until the patient has reached the age of ten or twelve. Occasionally one ear is much larger than the other, and in these circumstances operation during early childhood may be permissible. Young ladies who are beginning to grow up very often present themselves for correction of protruding or " lop " ears because they wish to wear their hair off the ears.

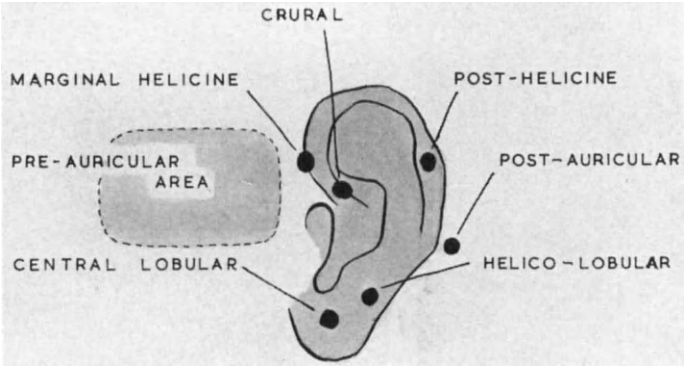

FIG. 6. Sites of opening of congenital fistulas.
(*Modified from Millar, J. B., and Moore, P. M.* (1950), *Arch. Otolaryng.*, 51, **2**, 245.)

Operation under these circumstances is simple and satisfactory. For protruding ears a falciform portion of skin $\frac{1}{2}$ in. or more wide and about $1\frac{1}{2}$ in. long is excised from the retro-auricular region : this crescent of skin contains in its centre the retro-auricular fold. The underlying cartilage is excised for a similar or slightly larger area, as indicated by the deformity, and the incision carefully sutured. The ear is then firmly bandaged for a day or so.

In some cases the antihelix is absent, and it may be desired to cure the deformity. This requires a somewhat more elaborate operation, and it may be advisable first to make a model of the auricle in latex to practise upon (Brown, 1948). The correction is made by incising the cartilage along the line of the future antihelix and then suturing it in either a back-to-back or overlapping position.

The plastic correction of macrotia in most cases consists simply in the removal of a wedge or segment of the auricle, but the procedure outlined above may be carried out at the same time if it is thought necessary. Accessory auricles are usually excised without difficulty, and deformities such as splits of the lobule

are also easily corrected. The plastic repair of microtia, anotia, and polyotia is more complicated and requires the utilization of tubed pedicle grafts and the later insertion of autogenous cartilaginous supports. The result in these cases is seldom satisfactory. The use of rubber or other artificial prostheses has obvious drawbacks ; and if the patient is a girl, it may be better to advise her to conceal the defect with her hair : if a man, to forget about it.

(*c*) **Congenital Auricular Fistulas.** Congenital fistulas of the ear are relatively common and of considerable practical importance. PRE-AURICULAR FISTULAS, narrow tubes ending blindly, are found along the line of predilection of Fig. 7, Wood-Jones and Wen, described above.

Congdon, Rowhanavongse, and Varamisara (1932) divide these groups according to the site of their occurrence. About 90 per cent. are found close to the anterior border of the ascending limb of the helix. Next in order of frequency

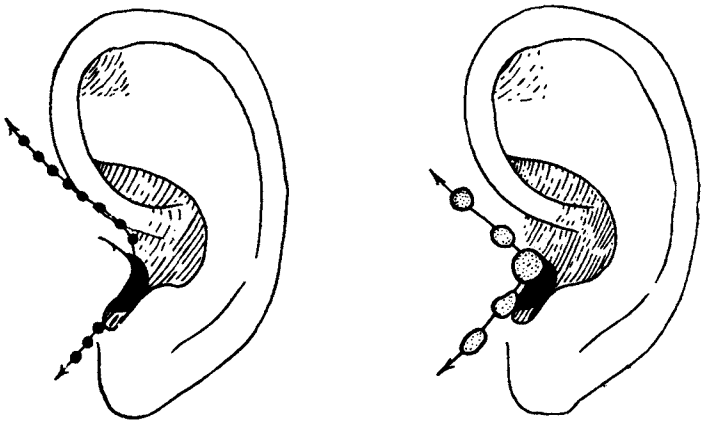

FIG. 7. A. the distribution of pre-auricular fistulas. B. The distribution of pre-auricular appendages (from F. Wood-Jones and Wen I-Chuan).

are the pre-auricular fistulas which lie in a line extending from the lower border of the crus helicis to the angle of the mouth. They are often associated with congenital scars, and a small pigmented mole may be found near the fistulous opening. Other less common situations are in the crus helix, in the helix descendens, and at the junction of the helix and the lobule. Posterior auricular fistulas are likely to be of infective rather than congenital origin. Finally, central lobular fistulas are pits in the middle of the lateral surface of the lobule, sometimes associated with another similar pit on the medial side. These fistulas may be indistinguishable from ear-ring punctures, particularly when they communicate, as occasionally happens. These are the typical sites for fistulas : others may be found in the cavitas conchæ, the fossa triangularis, and the antitragus. These fistulas may be deep, tortuous, and extensive.

COLLAURAL FISTULAS between the ear and the neck form another group : they have two openings, the upper lying in the external auditory meatus or in the incisura intertragica, the lower behind the angle of the mandible in front of the

sternomastoid muscle. It is possible for the upper end of the fistula to open into the middle ear.

Auricular fistulas are not normally associated with other malformations, but many cases have been recorded in which other abnormalities, such as microtia, auricular appendages, branchiogenetic fistulas of the neck, thyroglossal cysts, hare-lip, cleft palate, and microphthalmia have been present.

MANDIBULO-FACIAL DYSOSTOSIS is a syndrome first described by Treacher Collins (1900) and later by Franceschetti and Zwahlen (1944). Many cases had been described before the syndrome was recognized as such. According to Franceschetti and Klein, quoted by Altmann, its principal features are :—

Palpebral fissures sloping down laterally (antimongoloid) with a coloboma in the outer part of the lower lids, and, more rarely, in the upper lids.

Hypoplasia of the facial bones, especially the malar bones and the mandible.

Malformations of the external ear and occasionally of the middle and internal ears. The auricle usually occupies a low position. (See p. 20.)

Macrostomia, high palate, abnormal position and malocclusion of the teeth.

Blind fistulas between the angles of the mouth and the ears.

Atypical hair-growth, in the form of tongue-shaped processes at the hair-line, extending towards the teeth.

Other anomalies such as facial clefts and skeletal deformities may be present.

The disease dates from a developmental disturbance occurring at the sixth or seventh week of intra-uterine life.

These auricular fistulas are lined by stratified squamous epithelium and may contain epithelial structures such as hairs, sweat glands and cartilage. They may contain purulent infected debris, the exudation of which may cause eczematous dermatitis around the orifice. Should the latter become blocked an abscess may result.

CONGENITAL ARTERIO-VENOUS FISTULAS, or cirsoid aneurysms, resulting from anastomoses between the internal carotid artery and jugular vein, or between smaller arteries and veins, may be found in the auricular area, the external auditory meatus, or the middle ear.

Treatment. Congenital fistulas secrete a serous or purulent exudate which may be offensive. When the orifice becomes occluded a cyst or abscess forms. For these reasons surgical ablation is desirable, but it may prove very difficult. The injection of caustics into the tracts has often been tried, with little success.

The extent of the fistula and its possible ramifications are first outlined by the injection of a dye such as methylene blue, after which the whole tract must be completely dissected away. Surgical diathermy, the tract being laid open with the diathermy knife and left to granulate, is the most satisfactory method when feasible, and has the advantage of leaving a smooth flexible scar.

(*d*) **Congenital tumours** may be found in the auricle. They include lymphangiomas, hæmangiomas, papillomas, epidermoids, and dermoids. True dermoids are usually connected with the periosteum and are found above or behind the auricle. In rare cases they may be found in the auricle itself or in the preauricular area. Typical glomus tumours have been found in this area (Sannicandro, 1936 ; Ertl, 1943 ; Capps, 1952).)

Tumours are removed following general surgical principles, each case requiring separate consideration.

Congenital Abnormalities of the External Auditory Meatus and Middle Ear

Microtia with Atresia of the External Auditory Meatus. Congenital atresia of the external auditory meatus is usually associated with microtia. Heredity is not a marked factor in its incidence. It appears more often on the right side

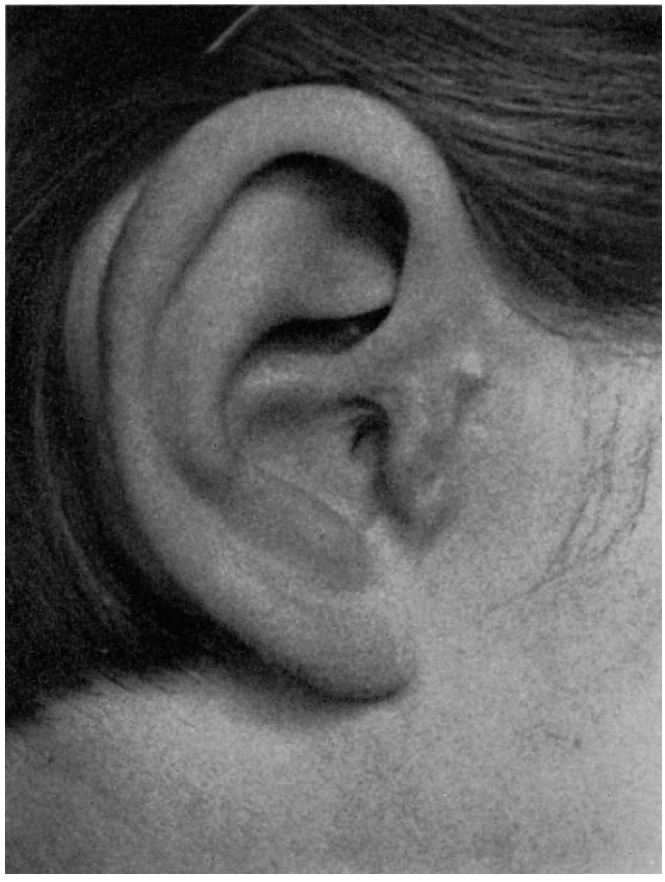

FIG. 8. Atresia of the external auditory meatus with accessory auricles.

and in males. The atresia is usually bony, but may be cartilaginous. The meatus may be represented by a blind pit, the bottom of which is occluded by bone covering the deformed middle-ear cavity. This condition is most often accompanied by a normal internal ear, as demonstrated by hearing tests in co-operative children or adults. Hearing for bone-conduction will be found to be normal, while a loss of 50 to 60 decibels for air-conduction is usually present.

In these cases, as there is no external meatus, the tympanic membrane (which is normally formed by thinning of the mesenchyme at the blind end of the meatus)

is of necessity absent, being replaced by bone which overlies the entire middle ear. The incus and malleus are almost invariably deformed and often fused together, although this is not always the case (Woodman, 1952). The malleus may be fused to the bony plate which replaces the drum membrane. The stapes, which is separately derived from the second branchial arch, may be normal and mobile in the oval window. The internal ear, which is developed separately from the auditory plate of neurectoderm, is also usually normal.

Another alteration from the normal anatomy is in the shape of the tubo-tympanic cavity, the tympanum being smaller than usual and the fenestras partly occluded by fibrous tissue. The Eustachian tube is sometimes normal, but may be malformed (Altmann, 1951). It may be entirely absent, narrowed, or may contain no cartilage. In the mastoid process cellular development varies from total absence of air-cells to complete pneumatization. Finally the facial nerve, while it may be normal in size and course, may alternatively be small and may take a very abnormal course. Having run from the geniculate ganglion between the horizontal canal and the stapes, instead of proceeding down and out through the stylomastoid foramen, it may proceed in a circle anteriorly in the position normally filled by the annulus tympanicus, and finally emerge in the region of the glenoid fossa. It may cross the tympanic cavity uncovered by bone.

Treatment. Until very recently, otologists usually had little to offer in the way of treatment for bilateral atresia of the auditory conductive mechanism, an exception being provided by the occasional case in which the stenosis is caused by a narrowing closed by fibrous tissue at the junction of the cartilaginous and osseous portions of the meatus, the middle ear being normal. Here a plastic reconstruction of the meatus is sufficient to provide a cure. Unfortunately, these cases are rare.

In the more usual grossly deformed cases surgeons in the past approached the problem with extreme caution, generally advising against operation even in bilateral cases, although Kiesselbach first attempted surgical cure as long ago as 1883. The deafness is usually severe and crippling, resulting in defective speech and inadequate education. Hearing aids, although useful, may not raise the threshold to a satisfactory degree of social adequacy. Recently, however, the surgical work of Ombredanne (1947), Pattee (1947), Vogel (1949), Siirala (1949), Sharp (1952), De Graaf Woodman (1952), Shambaugh and others has done much to improve the prognosis.

Operative Indications and Conditions

The condition must usually be bilateral. The operative hazards, particularly as regards the facial nerve, are so great that it is seldom justifiable to operate on a unilateral case. Shambaugh (1952), however, considers that unilateral cases can be benefited enough to justify surgery. The internal ear should be normal, as indicated by normal bone conduction. It follows that operation should usually, in spite of educational problems, be deferred until the child is old enough to co-operate in audiometry. Radiographs must always be taken, and should show a normal internal ear shadow. They will also give valuable information as to the degree of pneumatization of the mastoid process. The ideal case for operation is

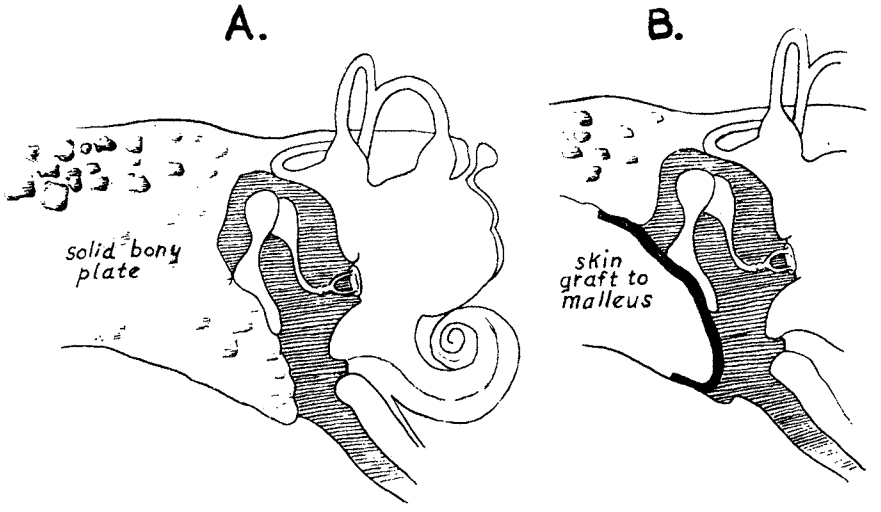

FIG. 9. A. Tympanic bony plate with mobile ossicular chain. B. Skin-graft to malleus.
(Victor Goodhill (1960), *The Modern Educational Treatment of Deafness*, Manchester University Press.)

one with a well pneumatized mastoid: Holmes does not usually advise operation in sclerotic cases because of the added operative difficulties. He has found an abnormally placed facial nerve in two cases with well pneumatized mastoids.

Operation. The operation is one of extreme delicacy and should not be attempted except by surgeons of considerable experience and skill. In brief, it consists in making and maintaining a canal down to the inner tympanic wall. The nature of the further operative procedures will depend upon the condition of the middle ear, following the well-established principles of tympanoplasty. The usual procedure is to explore the mastoid and define the mastoid antrum and the

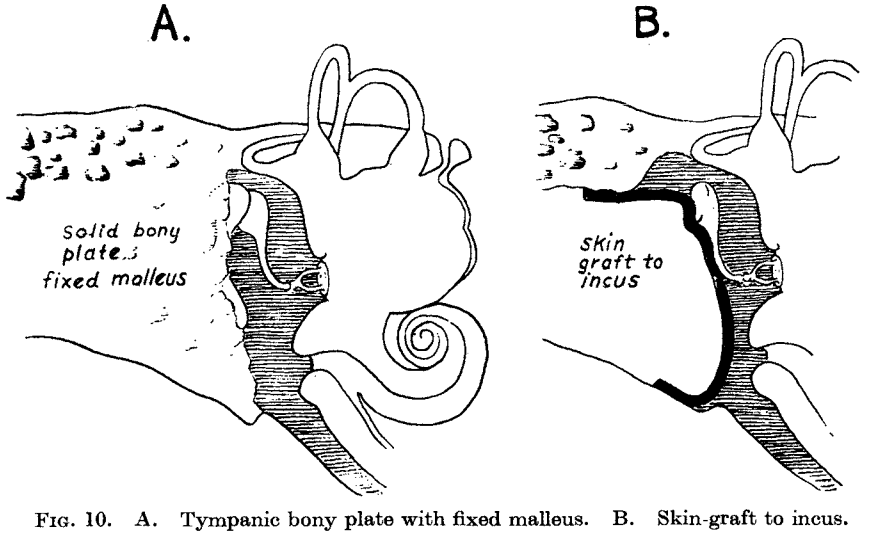

FIG. 10. A. Tympanic bony plate with fixed malleus. B. Skin-graft to incus.
(Victor Goodhill (1960), *The Modern Educational Treatment of Deafness*, Manchester University Press.)