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ATRESIA AURIS CONGENITA.

ILLUSTRATED BY THREE CASES (TWO FROM THE CLINIC OF DR. ST.CLAIR THOMSON).

BY HUNTER TOD, M.B. CANTAB., F.R.C.S. ENG.,

Late Senior Resident Medical Officer at the Throat Hospital, Golden Square, London.

THE literature on this subject is so abundant and complete, thanks chiefly to our German *confrères* (Bezold, Joel, Meyer, Schwartze, Stetter, Ruedi, Landauer), and Vali, Gradenigo, and others, that one begins to doubt if this deformity is so rare as has been hitherto supposed. Lately I have had the opportunity of examining three such cases, and, although they do not differ from many hitherto published, yet this paper may perhaps be excused on the ground that in perusing the literature I have been able to collect as many as 114 cases. Unfortunately, many are merely cited without further description, but the number is sufficiently large to enable one to draw from them a fairly accurate clinical picture.

I have only quite recently had the opportunity of reading Ruedi's thesis on this subject (fifty-seven cases); and as we have taken many of the cases we cite from the same sources, I have, to save repetition, only appended those which do not appear in his thesis, *i.e.*, fifty-seven other cases.[1]

[1] Thomas Ruedi, Inaugural Dissertation, Basel, 1899: "Microtie mit Atresia Auris Congenita."

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Author's Three Cases.

The following three cases, two of which are bilateral, are good illustrations of atresia auris congenita. They are similar in that they possess no other deformity and that the labyrinth is not affected.

After their description, I have ventured to give a *résumé* of the main points in connection with this deformity which I have been able to gather from perusing the literature at my disposal.

The first of my cases is that of a man of exceptional intelligence, whose hearing-power I could accurately test. He affords evidence of the fact that operative treatment is useless, no matter how favourable the conditions may appear; and also that the hearing-power in some of these cases may be surprisingly good.

Case I. (*under Dr. St.Clair Thomson*).—E. H——, twenty-nine years. His previous history is recorded by Mr. Field (whom I have to thank for permitting me to make use of this abstract).[1]

In 1883 Field operated on both sides, hoping to produce an artificial external meatus. Before the operation the voice could be heard if loud, the tuning-fork perfectly; and there was no difference if mouth was open or shut.

In describing the operation, Mr. Field says: "I began by dissecting the posterior portion of the auricle, an assistant drawing the ear forward. I found a small aperture in the bone, into which I could pass a good-sized probe. I carefully made an opening, and fixed in a silver speculum, which I had made for the purpose, fastened with plaster, so that it could not move out of place. I operated in the same way on the other ear, and in three days' time the ears had completely healed, and a good opening was left on both sides through which, when the dressings were removed, the boy could hear remarkably well; in fact, sometimes, when there was no pus in the aperture, he could hear a whisper. The difficulty was to keep the parts open. I thought of skin-grafting, but it was impossible to cover the whole canal in this way. At length the granulations increased, and although the openings remained for some time, they slowly closed, and I was obliged to abandon any hope of keeping the ears from closing up again."

Since then E. H—— has had no further aural treatment, but during the past two years several minor operations on the nose for nasal obstruction. For the same period he has suffered considerably from tinnitus, at first intermittent, now constant. Paracusis

[1] Field, "Diseases of the Ear" (London).

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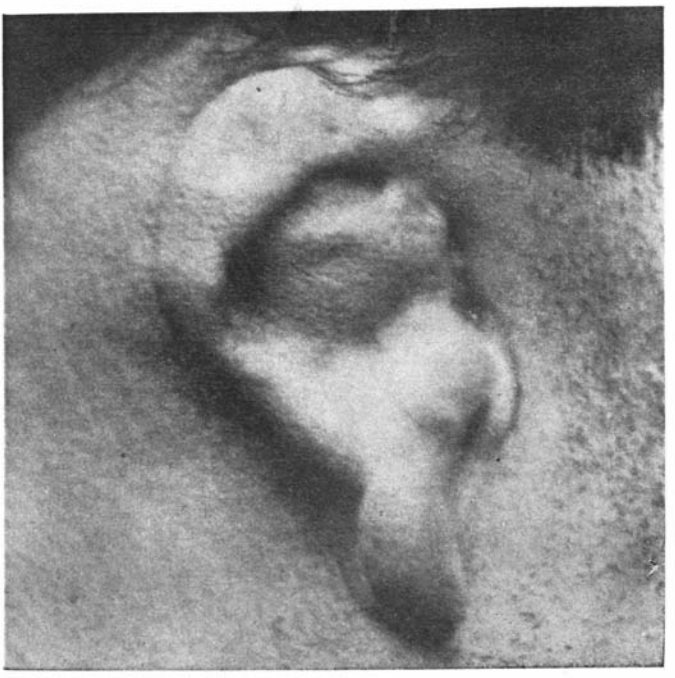


PHOTO I., CASE I. RIGHT AURICLE.

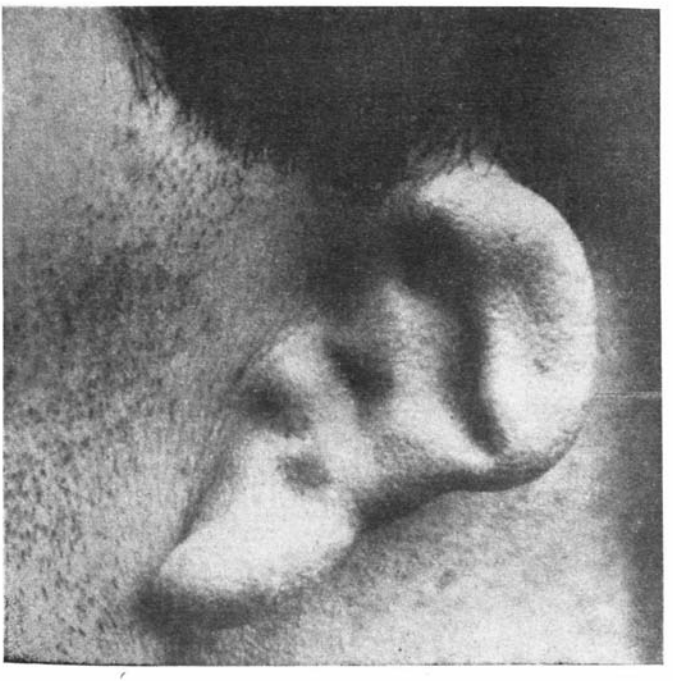


PHOTO II., CASE I.—LEFT AURICLE.

PLATE I.—TO ILLUSTRATE DR. HUNTER TOD'S PAPER ON ATRESIA AURIS CONGENITA.

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Willisii is also well marked—so much so that when travelling in a railway-carriage he often overhears the whispered conversation of others, which no one else can hear, and has unintentionally joined in the conversation, much to everyone's discomfort. He also makes the voluntary statement that he hears much better when standing opposite the speaker, especially if he has his mouth open. The use of an audophone greatly improved his hearing.

E. H—— at present is an accountant, his chief complaint being that the tinnitus prevents him thinking, and is especially worrying when he has to deal with large figures. His deafness has been about the same as long as he can remember. Apart from his ears he has no other deformity, nor have his relatives.

On both sides the auricles are markedly deformed and smaller than normal, but by no means rudimentary; and there is complete occlusion of the meatus.

On the right side (Photo I.) the helix is inrolled, and at the site of Darwin's tubercle is fused to the antihelix. The tragus and antitragus are fused; the lobule is adherent to the skin and is turned forward. The concha is convex, and the whole cartilaginous portion of the auricle feels thin and rudimentary.

Between the tragus and antitragus, at the site of the external meatus, is a small perforation leading through to the back of the ear; this and scars, one extending from the perforation downwards, and another along the posterior border of the auricle, bear evidence of former operative treatment.

On the left side (Photo II.) the condition is very similar, only the ear lies more obliquely, and the tragus and antitragus are more prominent. By pressure between them one can feel a depression which suggests the position of the external meatus.

On each auricle, on the right side almost at the anterior superior margin of the helix, and on the left situated somewhat lower down, is a small dimple, suggestive of a fistula auriculæ congenita (to be distinguished from the ordinary fistula auris congenita).

The nasal obstruction, due to a small spur and enlargement of the posterior ends of the inferior turbinates, was removed by operation, but in no way benefited the hearing.

By posterior rhinoscopy the orifices of the Eustachian tubes appeared normal, and there were no adenoids. The palate moved normally on deglutition and phonation.

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TESTS FOR HEARING.

Voice (patient's eyes shut):

	R.	*L.*
1. Distinguishes sound	20 feet+	20 feet+
2. Simple sentences:		
(*a*) Side face	1½ to 2 feet	4 feet
(*b*) Front face (mouth open)—3 feet		
(*c*) ,, (mouth shut)—2 feet		
3. Words:		
(*a*) Side face	6 to 8 inches	2 feet
(*b*) Front face (mouth open)—3 feet		
(*c*) ,, (mouth shut) —1½ feet		

Ordinary conversation could be sustained at a distance of 10 feet if the patient made use of "lip-reading."

4. Whispering: Voice heard close to ear, but not understood.

Watch: Only on contact. Heard best at root of nose (glabella) and just behind auricles.

Tuning-forks:

	Schwabach's Test.		Air Conduction.		Rinné's Test.	
	R.	*L.*	*R.*	*L.*	*R.*	*L.*
C	+12″	+16″	Not heard		—	—
C_1	+22″	+28″	−60″	−65	−40	−45
C_2	+8″	+6″	−20″	−12	−15	−10
C_3	± ?	± ?	−20″	−15	−15	−11
C_4	—	—	−15″	−25	+8 ?	+8 ?
G_4	—	—	−6	−6	± ?	+3 ?

On passing the Eustachian catheter and inflating with air, one could hear, on holding one's ear next to the patient's, a hissing sound as if air were entering the (?) cavum tympani. The sound appeared to be quite superficial, and was better heard on the left side.

The air inflation relieved temporarily the feeling of fulness in the ears, but otherwise did not benefit.

Bougies could be passed, on each side, nearly 2 inches beyond the catheter, and were felt as if they were "sticking out of the ear."

Summary.—Congenital bilateral aural deformity, with total atresia of both meatus. Careful operation, under favourable conditions, twelve years previously, unsuccessful. Hearing sufficiently good to carry on conversation and daily occupation. Tuning-forks: (1) by air conduction not heard below C_1, but in upper half of the scale well and without intermission; (2) marked increase of bone conduction for lower tones. No other deformity. Eustachian tubes patent. Bougies can be passed nearly 2 inches. No paralysis of palate. Hears better with mouth open. Chief complaint is tinnitus. Paracusis Willisii well marked.

CASE II.—D. S——, girl, five years. One of a large family; all healthy and in no way deformed. The child's intelligence is

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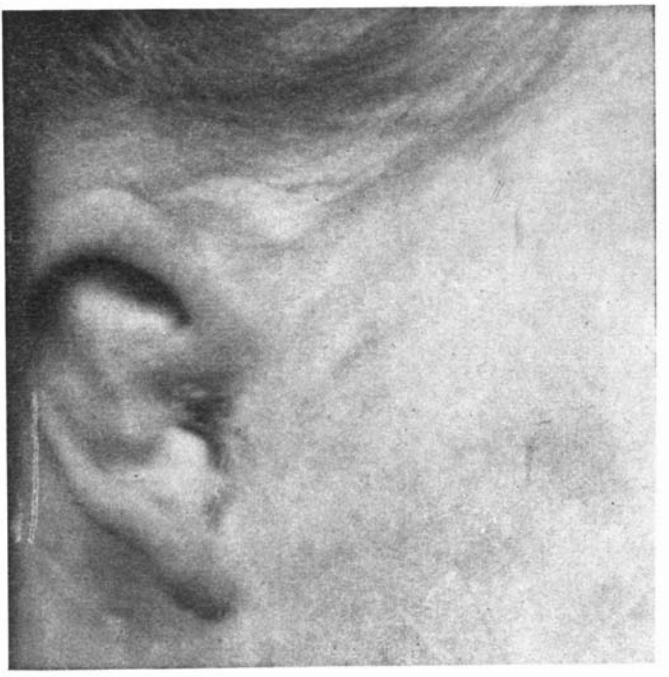


PHOTO III., CASE II.—RIGHT AURICLE.

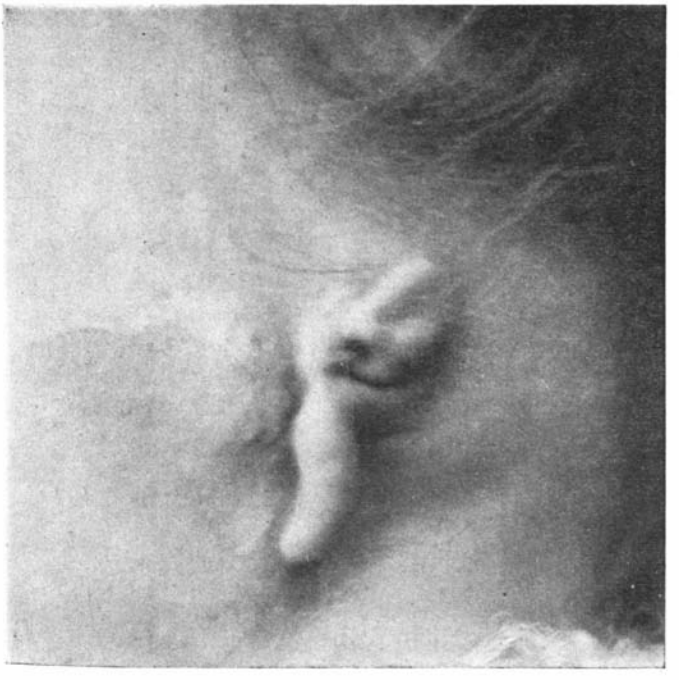


PHOTO IV., CASE II.—LEFT AURICLE.

PLATE II.—TO ILLUSTRATE DR. HUNTER TOD'S PAPER ON ATRESIA AURIS CONGENITA.

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somewhat deficient, but may only be due to the fact that she has never learnt to speak; only mumbles indistinctly. She was operated on, eighteen months ago, on both sides. The left side was unsuccessful, but on the right side the parents affirm that the hearing was somewhat improved. From the operation report one can learn very little, except that a small opening was found on each side in the bone, at a position much lower down than was expected, and that the attempt made to keep the canal open failed.

The auricles were small and considerably deformed. The left side was rudimentary, and took the usual form of a longitudinal puckered-up ridge; there was much scarring from the previous operation. On the right side the auricle was fairly well developed. At the site of the external meatus was a small opening filled with ceruminous crusts, on removal of which a probe could be inserted, but ended blindly in a cul-de-sac. The mastoid regions felt normal. (See Photos III. and IV.)

The tests for hearing could not be satisfactorily carried out; but apparently the watch was heard on contact, as were the tuning-forks.

The voice could be heard if one spoke loudly about 3 feet behind the child's back. The higher tuning-forks also seemed to be heard through the air, but the answers were often contradictory.

Summary.—Child, aged five years. Deaf-mute. Operation unsuccessful. Tuning-forks and watch heard by bone conduction; voice and higher tuning-forks by air. No other deformity.

Case III. (*under Dr. St.Clair Thomson*).—F. H——, healthy, bright girl, seven years. Brought to hospital with complaint that she had had several attacks of deafness following a cold in the head; that she snored at night, and usually kept her mouth open.

On examination, at first sight the auricles appeared normal. Closer examination showed the left auricle to be somewhat shorter, broader, and flatter than on the right side, the lobules being on the same level, but the helix of the left side at a lower one than on the right.

On the *left side* the external meatus quickly ended in a cul-de-sac, at the bottom of which was a fine opening suggesting permeability, but which on probing was found to end blindly after 1 millimetre.

The cartilaginous meatus, as well as the mastoid region, felt normal. There was no other deformity.

Parents and other relatives normal. Adenoid vegetations diagnosed and removed. Later "politzerized." Marked improvement in hearing.

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Mother stated that there never had been any discharge from the ears, and that the left side had been closed since birth.

The right membrana tympani was retracted, but otherwise normal. A hissing sound as though air was entering the cavum tympani could be heard in both ears on "politzerizing." The uvula and palate moved normally on deglutition, phonation, and stimulation.

TESTS FOR HEARING.

	L.	*R.*
Watch	Firm contact only over mastoid; *not* over auricle.	6 inches
Weber	Markedly to left side.	
Rinné, C_2	−35 seconds	
Schwabach, C_2	+15	Normal
Air Conduction:		
Voice: (1) Whisper ...	3 inches	Normal
(2) Ordinary conversation	10 to 12 inches	Normal

Tuning-forks: Below C_2 not heard on left side; the higher tuning-forks gave indefinite result owing to right ear being normal.

Summary.—On left side atresia auris congenita, with very slight aural deformity. Brought to hospital not on account of deformity, but from symptoms arising from the presence of adenoids. Marked improvement of hearing after removal of adenoids (Dr. Thomson), but unfortunately state of hearing before operation not tested. Hearing-tests on affected side showed bone conduction to be markedly increased, and air conduction below C_2 to be absent.

Taking into consideration with these cases those which have already been published, the following are the main points in connection with this deformity:

Auricular deformity almost invariably accompanies total occlusion of the external meatus. The deformity varies exceedingly. The auricle may be entirely absent (Gradenigo[1] five cases, Binnie,[2] Cantlie[3]); it may be merely represented by a fold of skin (Corton,[4] Stiles[5]); by a row of cartilaginous tags (Randall,[6] Schwartze[7]); or by an auricle varying in its deformity to such an extent that in the one case the auricle can hardly be recognised as such, and in the other the deformity is so slight that only careful observation detects it (Case III.).

[1] *Arch. f. Ohr.*, xxxiv. 281. [2] *Annal. of Surg.*, Philad., xxiv. 201.
[3] *Brit. Med. Jour.*, 1891, i. 1223. [4] *New York Med. Times*, 1886.
[5] *Trans. Med.-Chir. Soc. Edin.*, 1898, xv. [6] *Intern. Clinic.*, Philad., iii. 328.
[7] *Path. Anat. d. Ohr.; Lehrbuch der Chir. Krankheit. d. Ohr.*

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Finally, the auricle may be quite normal (Jacobson,[1] Wolff,[2] Bonnafont,[3] Rutten,[4] and Gordon[5]).

In over 50 per cent. the auricle is described as "rudimentary," in 40 per cent. as being "deformed."

The deformity is more or less consistent, in spite of its varying degree. In slighter cases the auricle appears turned forward, and the helix may be fused to the tragus, with or without there being actual atresia of the external meatus (Stetter[6]). In the more severe cases the auricle appears crumpled up, and takes the position of a longitudinal ridge.

The consistency of the deformity can only be explained (Gradenigo[7]) by referring the cause to arrest of development.

His states that at the end of the second month of embryonic life the ear can be recognised in detail. In the third month its upper posterior part projects from the skull and turns forward on to the face, where it remains two to three weeks, and then returns to its former position, thus setting free the antihelix. If the ear remains in the "turned-forward" position, development is arrested. Vali[8] agrees that this may be so in cases where there is also maldevelopment of the external and middle ear; but in cases where only the auricle is deformed he is inclined to agree with Moos[9] that it may be due to external causes, as pressure of the umbilical cord occurring later in embryonic life.

As is well known, *fistula auris congenita* occurs in ears otherwise normal. In cases of atresia with auricular deformity these fistulæ are remarkably rare. I can only find four such cases, described by Bückner,[10] Vali,[11] Grunert,[12] and Doran.[13] As emphasized by Grunert, these fistulæ should not be confused with fistulæ auriculæ congenitæ (compare Case I.).

That these fistulæ occur so rarely in cases of atresia and so much more frequently in normal auricles is surely in itself suggestive that they are probably due to incomplete development in the tissue round the first branchial cleft, from which the auricle is derived, and not connected developmentally with the branchial cleft itself, which later on is represented by the external and middle ear and the Eustachian tube.

Auricular appendages in association with atresia auris congenita are nearly as rare as the fistula. In seven cases which I have

[1] *Arch. f. Ohr.*, xix. 34. [2] *Ibid.*, xxxv. 131.
[3] *Annal. de la Chir. franç.*, 1843, viii.
[4] *Rev. de Laryng.*, 1893, xiii. 615. [5] *Brit. Med. Jour.*, 1888, i. 812.
[6] *Arch. f. Ohr.*, xxi. 92. [7] *Ibid.*, xxxiv. 281. [8] *Ibid.*, xxxiii. 28.
[9] *Zeit. f. Ohr.*, xi. 267. [10] *Arch. f. Ohr.*, xxii. 200. [11] *Arch. f. Ohr.* xxxiii. 28.
[12] *Arch. f. Ohr.*, xlv. 10. [13] *Trans. Obstet. Soc. Lond.*, xxxiii. 199.

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collected the appendages were found in four cases on both sides, although in three cases the atresia was only present on one side, showing the close relationship of these appendages to the other aural deformities.

In Cantlie's[1] case, where on the right side there was total absence of the auricle and external meatus (the skin over the region of the external meatus being quite smooth), accompanied by further deformity of the face, an auricular appendage was situated $1\frac{1}{2}$ inches in front of, and 1 inch below the tip of the mastoid process; on the left side, where the auricle and external meatus were normal, two appendages were found identical in position and appearance to that on the right side.

In Walker Downie's[2] case the left auricle was rudimentary and there was no sign of the external meatus, and on the right side, in front of the otherwise normal auricle and meatus, was a "blurred image" in miniature of an auricle, springing from the tragus, with the helix directed forwards. "This supernumerary auricle seemed to consist of what might be described as a reflection."

Other deformities may accompany congenital atresia of the external meatus, and are explainable by the arrest or maldevelopment of the structures in connection with the first and second branchial bars.

The most common deformity is asymmetry of the face, due to the affected side being smaller, and often paralyzed (ten cases).

In several cases where a post-mortem could be obtained the zygomatic process was found to be absent (W. Downie[3]). This is of interest in that some authors consider the zygomatic process to be the homologue of the "quadrate bone" of the lower vertebrates, which unites the lower jaw to the basis cranii and periotic capsule.

Other deformities cited are: absence of the mastoid process, sacro-coccygeal fistula, deformity of mouth, fistula in cheek, asymmetry of the palate, and obstruction of the choanæ.

The hearing-power in these cases is of more practical interest, and questions may be asked as to what is to be expected from these patients, and also if their condition can be improved by operative or other treatment.

Ruedi gives a table of twelve collected cases where the hearing-power was more or less carefully tested. In addition to these cases, I find twenty-three where mention is made of the hearing, and in fourteen of these (including two of my cases) the actual state is given.

[1] *Brit. Med. Journ.*, 1891, i. 1223.
[2] *Practitioner*, London, lvi. 261.
[3] *Ibid.*

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From these cases we learn that there is very rarely complete deafness. Moos describes three cases as "deaf-mute," and Doran one case as being "stone-deaf." Several cases are said "to hear sufficiently well to understand." In the cases where actual tests have been made, the hearing-power appears to vary, as regards air-conduction, to a very great extent, and to have no apparent relationship with the deformity. The extreme cases may be taken as exceptional.

For instance, Binnie[1] quotes a man in whom the hearing, even when the normal ear was stopped, seemed to be perfect even for conversation carried on in low tone, in spite of having "no auricle and external meatus on the right side, the surface of the head in the ear region being marked as if a hot flat-iron had been passed over it. This had often been tested by his friends."

And Robb[2] relates a case where, although on neither side trace of the meatus could be found, whispering could be heard 6 feet, and loud conversation up to 15 feet.

On the other hand, in Bonnafont's[3] well-known case, where there was only a membranous septum in the external meatus, the watch could not be heard on contact before operation, although over 1 metre afterwards.

As a rule, the watch is heard on contact or to a distance of a few centimetres. "Whispering" may or may not be heard, but ordinary conversation to a distance up to 1 to 2 feet, rarely so well as in Case I. (4 feet). If the patient has learnt lip-reading, he may appear to hear very much better than he really does.

The bone conduction is markedly increased in the lower half of the scale, but the air conduction is so diminished that below C_1 the tuning-fork is no longer heard. As the scale ascends the air conduction improves, and there is usually no break in the tone sequence.

This condition, as Bezold[4] pointed out, being the same as occurs in cases of middle-ear disease without accompanying labyrinthine disease, in these cases of congenital atresia shows that the labyrinth (as one would expect, seeing that its development is quite independent from that of the external and middle ears) is probably normal.

In several cases, as in Case I., the hearing-power was found to be increased if the mouth was kept open, this presumably being due to the conduction of the sound to the cavum tympani by the Eustachian tube.

In Mussey's case, quoted by Meyer,[5] where there was no com-

[1] *Annal. Surg.*, Philad., xxiv. 201. [2] Ref., *Arch. f. Ohr.*, xviii. 220.
[3] *Annal. de la Chir. franç.*, 1843, viii. [4] *Zeit. f. Ohr.*, xxvi. 11.
[5] *Langenbeck's Arch. Klin. Chir.*, xxix. 488.

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munication between the Eustachian tube and the cavum tympani, there was no difference in the hearing-power whether the mouth was kept open or shut. Politzer[1] considers that paralysis of the palate in these cases points to obstruction and maldevelopment of the Eustachian tube.

In other cases the hearing was much improved by the use of ear-tubes (Vali[2]) and by the audophone (Knapp,[3] and Case I.).

Is operative interference justifiable in these cases? The answer is an emphatic No. Schwartze[4] expressed this opinion in 1885; Joel in 1888 confirmed it by his pathological and clinical observations, and said that such cases should be for the surgeon cases of *noli me tangere*. Repeated attempts, however, have been made to make an artificial external meatus, in the hope of improving the hearing, but the results have been failures.

From a study of embryology, we learn that the external and middle ear and the Eustachian tube are derived from the cleft between the first and second branchial arches, from whose cartilaginous bars the ossicles are developed. When the maldevelopment of the branchial arches is so great that the cleft between them (at least, the outer half) ceases to exist, surely it is too much to expect the ossicles to be normal, even if they exist, to say nothing of the fenestræ ovalis and rotunda and of the membrana tympani.

The papers by Bezold[5] and Joel,[6] which give a record of sixteen cases examined post-mortem, together with five more cases quoted by Ruedi, confirm what the embryologists suggest. In no case was the middle ear found to be normal. In two cases the cavum tympani was absent, and in fifteen deformed and much smaller than normal; in no case could a membrana tympani be found. The ossicles were either rudimentary, ankylosed or absent. In only one case (Kiesselbach[7]) were both fenestræ mentioned as being normal; in that case the stapes was absent (probably lost during the dissection), so that one cannot tell if the stapes was fixed during life by firm connective tissue or was normally movable.

In all the other cases either one or both fenestræ, when mentioned, were abnormal or absent, or else the stapes was firmly fixed by connective tissue or by bone in the foramen ovale.

Only in Lucae's[8] oft-quoted case was the labyrinth found to be markedly defective, and in this case, during life, there was absolute deafness to voice and tuning-forks on the affected side.

But supposing for the moment that the labyrinth is found

[1] *Lehrbuch*, 700. [2] *Arch. f. Ohr.*, xxxiii. 28.
[3] *Zeit. f. Ohr.*, xi. [4] *Chir. Krankheit. d. Ohr.*, s. 80.
[5] *Zeit. f. Ohr.*, xxvi. 11. [6] *Ibid.*, xviii. 278, 1888.
[7] Gerlach's "Beiträge zur Morphologie," i., 1883. [8] *Virch. Arch.*, xxix.

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clinically to be functionally normal, that according to the various hearing-tests only an external and middle-ear obstruction exists, what success could be expected, however slight, from an operation? Those who have read Schwartze's[1] papers on "Acquired Atresia and Stricture of the Meatus, and its Treatment," cannot help appreciating the difficulty of keeping open the artificial meatus after operation in these cases. The tendency for the meatus to gradually close, and for complete atresia to recur, is so great that only the most careful and prolonged after-treatment can insure success. And how much more difficult are the cases of congenital atresia! Not only the cartilaginous but the bony meatus as well is completely obliterated. What hope is there, even if one could form a canal, of keeping it open?

And granted for a moment that an artificial canal could be obtained, what is the state of the cavum tympani? Ossicles at best rudimentary—at any rate useless, as there exists no membrana tympani. The stapes probably fixed in the foramen ovale by connective tissue or bony ankylosis, or perhaps the foramen may be absent.

In twenty-one (nine bilateral) collected cases where operation was attempted, success was only obtained by Bonnafont; but here the conditions were more favourable, the atresia being due, not to a bony, but a membranous, septum in the external meatus.

Meyer[2] in one case laid bare a membrane, which he thought might be the membrana tympani, and the hearing was said to be improved whilst the opening could be kept open.

Knapp[3] cites a case where the atresia was due to soft tissue, and not to bony obstruction, and where a silver tube 3 centimetres long could be inserted into the opening; but owing to occurrence of hæmophilia suppuration occurred and the wound closed.

Bremer[4] also quotes two cases where Meier attempted to make an artificial opening by puncturing the depression where the supposed external meatus was situated, and then dilating with trocar or laminaria. In one case the opening remained three months, and hearing was said to be improved meanwhile.

Results of Operation.—Briefly stated, from these cases we learn that—

1. The operation has never met with permanent success (Bonnafont's case excepted).

2. In all the other cases the atresia was due to bony obstruction, with the exception of Knapp's case.

[1] *Arch. f. Ohr.*, xlvii. and xlviii.
[2] *Langenbeck's Arch. Klin. Chir.*, xxix. 488.
[3] *Arch. f. Otol.*, New York, xi. [4] *Arch. f. Ohr.*, xiii. 276.

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3. In five cases there was no trace of the external meatus, although dipping in of the skin at the situation of the external meatus suggested it.

4. In nine cases either a depression of bone or fissure was present, suggesting a canal.

5. In several cases hearing was said to be better whilst the opening existed; but the opening invariably closed, and the final result was nil.

The method of operation formerly carried out consisted, as in Meier's[1] cases, in puncturing the skin at the point where a depression suggested the external meatus, and then dilating and attempting to keep open the canal by means of the cautery or tubes.

The more recent method, advocated by Kiesselbach[2] and Vali,[3] is to cut down on to the bone at the site of the depression, or make an incision behind the ear and turn it forward, and then seek for the fissure in the bone, which, if existent, will probably be found lower down than one would expect.[4] If there be a fissure, it is carefully enlarged by chiselling away the surrounding bone, a probe being inserted into the opening. In this way one hopes to reach the middle ear and obtain an artificial external meatus.

Assuming, then, that operative treatment is recognised as useless, *can anything be done for these cases?* Unfortunately, very little.

In Case III., for example, removal of the adenoid growths caused considerable improvement in the hearing, but presumably because the atresia was only unilateral. Still, we have seen that in some cases the hearing-power is increased if the mouth be kept open, due to the conduction of the sound by the Eustachian tube: and so probably any pathological condition, such as adenoids, post-nasal catarrh, etc., which could in any way, either directly by pressure or indirectly by setting up a tubal catarrh, bring about obstruction of the Eustachian tubes, would probably help to further diminish the hearing-power. Therefore, in order to give the patient every chance, these cases should be most carefully examined in early childhood, and adenoids, if present, should be removed, and treatment especially directed to obtain a normal post-nasal space.

As in these cases the labyrinth is rarely affected, and the hearing-power is present, no matter how slight it may be, it is only right to insist that everything possible should be done in the way of oral instruction and lip-reading, and as early as possible, to prevent the patient remaining a deaf-mute.

[1] *Arch. f. Ohr.*, xiii. 276. [2] *Ibid.*, xix. [3] *Ibid.*, xxxiii.
[4] Welcker, *Arch. f. Ohr.*, Bd. i.

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In several cases quoted the hearing was remarkably good and the speech was normal; in others the speech was thick and nasal, whilst some were stated to be deaf-mutes. Lastly, owing to the bony conduction being so good, much benefit was obtained in some cases by the use of the audophone.

The ætiology of this condition is obscure. Many theories are advanced, but which is correct? Certainly the deformity is not hereditary. I can find no published case where any near or distant relatives were similarly affected. The female sex seems to preponderate slightly—55 per cent. to 45 per cent. males.

Hitherto the right side has been considered to be almost invariably the one affected, but it is not so. Out of 95 cases, the right side was affected 38 times, the left 23, and both sides 34 times.

Syphilis, to which so much is traced, is considered by some to be the primary cause of this deformity. It may be so, but in no case is there any history given of any other sign of inherited syphilis: and more often than not the parents and their other children are stated to be healthy.

"Maternal impressions" need not be taken seriously. Is there any woman alive who is not firmly convinced that the congenital deformity which her child may be suffering from was due to some maternal impression?

The umbilical cord also has been accused of bringing about this deformity, and Moos[1] quotes two cases (both deaf-mutes, with atresia and aural deformity) where the cord was in each case said to be wound round the head.

Kiesselbach[2] believes that, if arrest of development of the auricle could be due to pressure of the umbilical cord, the occlusion of the meatus might also occur, owing to the position of the membrana tympani in the embryo being almost horizontal, and thus permitting of adherence of the walls of the meatus. This view might be supported by a specimen of Stiles's,[3] where the external auditory canal was obliterated in consequence of the tympani plate being flattened up against the squamo-zygomatic element of the temporal bone.

Virchow[4] considers congenital anomalies of the ear and its immediate neighbourhood to be due to early disturbance in the closure of the first branchial cleft, but not so much a simple developmental defect as an irritative inflammatory process, which may give rise to induration, adhesions, and scar retraction.

[1] *Zeit. f. Ohr.*, xi. [2] *Ibid.*, xxvi.
[3] *Monthly Journ. of Med. Sci.*, Edinburgh, vii. [4] *Archiv.*, Bd. xxx., xxxii.

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But if this be so, the process must be an exceedingly early one, as, according to Kolliker, the cartilaginous framework of the ossicles is complete at the end of the second month, whilst the first branchial cleft closes during the fifth week of intra-uterine life.

Joel[1] also points out the striking analogy between the pathological conditions found in the cavum tympani of these cases of congenital atresia and those of otitis media chronica suppurativa et adhesiva.

If the cause be due to an early irritative or inflammatory process, it is a remarkable fact that in after-life one never sees any recurrence of these inflammatory processes. No case has hitherto been published where a patient suffering from congenital atresia has also had the signs or symptoms of an inflammatory trouble in the middle ear or the mastoid process. Why is this? Does this mean that the ordinary road for infection is via the patent external meatus, and not from the Eustachian tube, as usually supposed, or is it only due to the altered conditions of the mucous membrane and tissues of the middle ear?

Conclusions.

One may sum up the condition extremely tersely:

1. The deformity is not hereditary, and the cause is not known.
2. It occurs rather more often in females, and is more often unilateral than bilateral.
3. One may get accompanying deformities, chiefly due to maldevelopment of the parts in connection with the first and second branchial arches.
4. The labyrinth is rarely affected. The hearing varies, but is present to some extent, though slight. Hearing-tests give practically the same results as those in an uncomplicated middle-ear affection, but more marked.
5. Embryological, pathological, and clinical observations prove operation to be useless.
6. Something more, perhaps, can be done by careful non-operative treatment and by early and assiduous instruction in speaking and lip-reading.

In conclusion, I beg to thank Dr. StClair Thomson most gratefully for having permitted me to make use of Cases I. and III., which came from his clinic, and also for his kindly suggestions and valuable assistance.

[1] *Zeit. f. Ohr.*, xviii.

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ABBREVIATIONS USED IN TABLES.—*M*, male; *F*, female; *R*, right; *L*, left; *B*, bilateral; *U*, unilateral.

TABLE I.

OPERATIONS—21 CASES (9 BILATERAL).

No.	*Case.*	*Reference.*	*Operation.*	*Result.*	*Remarks*
1	Bilateral.	Randall, *Intern. Clinics*, Phil., 1896, iii.	Bilateral exploratory incision in front of auricle. Right side, no canal. Left side, partial external meatus, but deeper parts impermeable.	Failure.	Second operation attempted later; also unsuccessful.
2	8 weeks.	Bishop, *Intern. Med. Cong.*, Berlin, 1890, iv.	Incision at culde-sac, where external meatus thought to be. Only a depression in bone.	Operation abandoned.	
3	5½ years. Bilateral.	Welcker (Blasius), *Arch. f. Ohr.*, 1864. i.	Incision behind ear, and auricle turned forwards. No canal found.	Operation abandoned.	
4	F., 7 years. Bilateral.	Bezold (Rother), *Zeit. f. Ohr.*, xxvi.	Exploratory incision. Articular process of inferior maxilla seen. No canal.	Operation abandoned.	
5	Bilateral.	Vali, *Arch. f. Ohr.*, xxxiii.	Incision behind auricle, which was pulled forward No sign of external meatus. Bone very hard.	Operation abandoned.	
6	(?)	Hedinger, *Arch. f. Ohr.*, xiii. 305.	Exploratory incision. No external meatus found.	Operation abandoned.	
7	7 years. Bilateral.	Skunlanski, *Gazetta lekarska*, 1895, i.	Exploratory incision where dipping in of skin pointed to a meatus.	Failure.	? Heard better whilst opening remained; but afterwards tests for hearing proved complete deafness to speech and tuning-forks.
8	1½ years.	Grunert, *Arch. f. Ohr.*, xlv.	Operation to improve the ear deformity and explore for external meatus. Bone exposed. No canal found.	Operation abandoned.	Aural deformity considerably diminished as a result of the operation.
9	M., 30 years. Bilateral.	Knapp, *Zeit. f. Ohr.*, xi. 13.	Operation 13 yrs. previously, but failure.	Failure.	

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TABLE I.—*continued.*

No.	*Case.*	*Reference.*	*Operation.*	*Result.*	*Remarks.*
10	F., 19 years. Bilateral.	Mazzoni (Gradenigo), *Arch. f. Ohr.*, xxxiv. 281.	Incision at point of depression, corresponding to entrance of external meatus.	Final result nil.	Hearing said to be better whilst opening existed
11 12 13	Quoted by Stetter, "The Congenital and Acquired Deformities of the Ear," Jena, 1898.		Operation attempted.	All failures.	
14 15	Cases I. and II. (already described).				
16	F., 15 years. Right side.	Meyer, *Langenbeck's Arch. Klin. Chir.*, xxix.	Bone exposed. Fissure found. A membrane like the membrana tympani laid bare.	Opening gradually closed.	Heard better so long as the opening kept open. Final result, a failure.
17	F., 8 mths. Right side.	Kiesselbach, *Arch. f. Ohr.*, xix. 227.	Incision behind ear. Fissure found. Bone carefully chiselled away around it.	? Heard better, but at best only a few sounds. Previously completely deaf.	Mother took child from hospital 12 days after operation because she was so satisfied.
18	Left ear.	Knapp, *Arch. f. Otol.*, New York, xi.	Obstruction due to soft tissue. Cul-de-sac of external meatus cut through. Silver tube, 3 cm. long, inserted.	Hæmophilia. Suppuration. Wound closed. Result nil.	Knapp thinks that but for the bleeding, the operation might have been successful.
19	Left ear.	Meier (Bremer), *Arch. f. Ohr.*, xiii. 276.	Depression behind ear pierced with needle, then with trocar. Attempt to keep canal open by crucial incision and cauterizing with silver nitrate.	Failure.	? Improvement during the short period when the wound was open.
20	Right ear.	Meier (Bremer), *Arch. f. Ohr.*, xiii. 276.	Small depression between rudimentary tragus and antitragus. Punctured. Dilated with knife. Laminaria bougies.	Failure. Canal kept open three weeks, and then closed.	? Improvement during period when canal existed.
21	Left ear.	Bonnafont, *Annal. de la Chir. franç.*, viii.	Membranous septum in external meatus opened by trocar, and kept open by cauterizing.	Successful. Membrana tympani seen after operation.	Before operation, watch on contact; one month after, 1 metre +.

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TABLE II.

ABSTRACT OF CASES COLLECTED.

No.	Reference.	Side.	Sex.	Age.	Auricle.	Other Deformities.	Hearing.	Remarks.
1	Randall, *Intern. Clinics*, Phil., iii. 328.	B.			Rudimentary. Longitudinal ridge. Total atresia.		Can hear sufficiently to understand; better, if mouth be open.	Operation (Table I.).
2	Moure, *Mém. et Bull. Soc. de Méd. et Chir. de Bordeaux*, 1886, 135.	R.	F.	12.	Rudimentary. Nearly complete absence. Total atresia.	Left ear normal. No other deformity.	Bone perception good. Tuning-fork, la^3, better heard on right side and Rinné.	
3	Knapp, *Zeit. f. Ohren.*, xi.	B.	M.	30.	Rudimentary. Crumpled-up, hook-like protuberances. Dimpling, as if presence of external meatus. Total atresia.	No other deformity.	Watch, on skull, $\frac{3}{5}\frac{4}{4}''$. General conversation, 1 to 2 feet. Loud conversation, 5 to 6 feet. Music well heard. Audophone, hears better. No difference if mouth be open or shut.	Catheter, air enters middle ear freely (see Table I.).
4	Knapp, *Arch. f. Otology*, New York, xi.	L.	F.		Congenital closure of entire external meatus by soft tissue.	No other deformity. Right side normal.	Bone conduction +. Weber to left. Voice, $\frac{1}{6}\frac{5}{0}''$ to $\frac{3}{6}\frac{0}{0}''$ both sides.	Catheter, air heard in both ears (see Table I.).
5	Heaton, *Journ. of Laryng.*, vi. 149.	R.	F.	15.	Misshapen. Two supernumerary auricles in front and below auricle. Total atresia.	Small supernumerary auricle in front of left ear. Lower half right side of face smaller. Mouth to left. Right jaw ill-developed.	Acoumeter: right, 13 cm.; left, 14 feet. Tuning-fork: right, $-15''$; left, normal. Bone conduction: right, $+10''$; left, normal.	Valsalva's experiment. air felt in left ear.
6	Meyer, *Langenbeck's Arch. Klin. Chir.*, xxix. 488.	B.	M.	13.	Rudimentary. Total atresia.	Nil.	Watch, 5 to 6 inches overhead. Contact only, if over ears. Best heard on contact with skull. Tuning-fork, best on skull. Not so well in ear region.	

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TABLE II.—*continued.*

No.	*Reference.*	*Side.*	*Sex.*	*Age.*	*Auricle.*	*Other Deformities.*	*Hearing.*	*Remarks.*
7	Meyer, *Langenbeck's Arch. Klin. Chir.*, xxix. 488	R.	F.	15.	Rudimentary. Total atresia.	Nil.	Tuning-fork heard best on affected side.	Operation (Table I.).
8	Massey (Meyer),				Rudimentary. Total atresia.	No communication of Eustachian tube with cavum tympani. Presence of ost. pharyng. of tub. Eust.	Could hear well enough to work. No change on opening or shutting mouth.	
9	Robb, ref. *Arch. f. Ohren.*, xviii. 220.	B.			No trace of any external meatus on either side. Auricles ?		Whispering, 6 feet. Loud conversation. 15 feet.	
10	Gradenigo, *Arch. f. Ohren.*, xxxiv. 282.	R.	M.	38.	Rudimentary. Longitudinal ridge. Small opening, 5 mm. long, above and in front of ridge.	Right lower jaw ill-developed.	Air conduction: whispering, nil; watch, nil. Watch better heard on right than on left mastoid process. Bone conduction: all tuning-forks heard. Lower tuning-forks (up to C_2) not heard through air.	
11	Vali, *Arch. f. Ohren.*, xxxiii. 28.	B.	M.	12.	Right: small, ill-developed. Small orifice, 3 mm. deep, at upper part (? = fist. auris congenita). Tragus & antitragus fused. Behind tragus dimpling, as if external meatus. Left: cartilage felt.	Left side of palate thicker than right. Does not move on phonation or stimulation. Uvula asymmetrical. Left half better developed.	Watch: right, 23 cm.; left, 23 cm. Best on auricle. Conversation: right, 1·3 m.; left, 1·1 m. Whispering: behind ear, at back. Tuning-fork *a*: better over ears than at mastoid or vertex; C^1, right, 54 cm.; left, 48 cm.; C^2, right, 60 cm.; left, 52 cm. Galton, good. Rinné: right, +; left, + (?). Hears	Operation (Table I.). On Politzering, air felt, but not heard in ears.

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TABLE II.—*continued.*

No.	*Reference.*	*Side.*	*Sex.*	*Age.*	*Auricle.*	*Other Deformities.*	*Hearing.*	*Remarks.*
					Greatly thickened helix, markedly inrolled. Auricle rudimentary. Total atresia.		better (1) with mouth open, (2) with ear-tubes.	
12	Gordon, *Brit. Med. Journ.*, 1888, i. 812.	R.	M.	11.	? Normal. ½ inch from auricle is complete atresia. Bony ring round meatus at the blind end.		Watch, $\frac{\frac{1}{2}''}{60}$.	
13	Bonnafont, *Ann. de la Chir. franç.*, 1843, viii.	L.			Normal. Congenital membrane simulating membrana tympani.		Before operation: Watch, not even on contact. After operation: 1 m. +.	Operation (Table I.).
14	Rutten, *Rev. de Laryng.*, 1893, xiii. 613.	L.	M.	10.	Normal. Congenital membranous septum at junction of cartilage and bony meatus. Punctiform depression in centre.		Tuning-fork heard (on vertex) equally well on both sides.	
15	Blower, *Liverpool Med.-Chir. Journ.*, vii. 206.	L.		11.	? Normal. ¾″ long cul-de-sac. Bone at blind end—stated definitely as being congenital.		Very deaf, especially in left ear.	Stenosis and chr. supp. ot. med. on right side.
16	Stiles, *Trans. Med.-Chir. Soc. Edin.*, 1895, xv. 54.	B.		6 mths.	Rudimentary, both sides. Small reduplication of skin. Two small pedunculer outgrowths	Right half of face smaller. Right jaw smaller. Chin to right side. Paresis right frontalis and	Mother stated child could hear loud noises.	

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TABLE II.—*continued.*

No.	*Reference.*	*Side.*	*Sex.*	*Age.*	*Auricle.*	*Other Deformities.*	*Hearing.*	*Remarks.*
					in front of rudimentary auricle on left side.	orbicularis palpebrarum muscles. Right cornea opaque.		
17	Bishop, *Intern. Med. Cong.*, Berlin, 1890, iv.	U.	F.	8 wks.	Rudimentary. Ear doubled on itself. Total atresia of external meatus			Operation (Table I.).
18	Bishop.	U.		14 mths.	Rudimentary. Total atresia of external meatus.			
19	Skunlanski, *Gazetta lekarska*, 1895, i.	B.		7.	Both auricles rudimentary. Total atresia.		Complete deafness to tuning-forks and speech.	Operation (Table I.).
20	Wagenhauser, *Arch. f. Ohren.*, xxi. 269.	L.		9 mths.	Narrow, crumpled-up cartilaginous protuberance. Total atresia.	Left aural region much flatter than the right side.		
21	Wagenhauser, *Arch. f. Ohren.*, xix. 55.	R.	F.	61.	Rudimentary. Total atresia.	Right side of face is completely developed.		
22	Gruber, *Bericht d. K. K. Algem. Krankenhaus.*, Wien, 1870.	B.		4 mths.	Left: upper part normal; lower half ill-developed and small. External meatus 1 cm. long. Right: same as left side; auricle more attached to head. External meatus 1 cm. long.	Choanæ obstructed posteriorly. Sound only passes 3 cm. into nose. Right eyelid fused. Jaws normal.		

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TABLE II.—*continued.*

No.	*Reference.*	*Side.*	*Sex.*	*Age.*	*Auricle.*	*Other Deformities.*	*Hearing.*	*Remarks.*
23	Cantlie, *Brit. Med. Journ.*, 1891, i. 1223.	R.	M.	38.	Absence of auricle. Skin smooth over region of external meatus.	Bilateral supernumerary auricles both sides. Developmental deformity of right side of face.		
24	Binnie, *Ann. Surg.*, Phil.. xxiv. 201.	B.			Left: rudimentary. Vertical row of tags. Right: not so rudimentary, but markedly ill-developed. Total atresia.	Left jaw smaller; face oblique towards left.	Hears better if mouth is open.	Depression in bone on palpation. ? External meatus.
25	Binnie, *Ann. Surg.*, Phil., xxiv. 201.	R.	M.	Adult	No trace of auricle. "Surface of ear region as if hot iron had been passed over it." Total atresia.	Nil.	"Hearing, even when normal ear stopped, seems to be perfect." Conversation can be carried on in a low tone.	
26	Doran, *Trans. Obstet. Soc.*, Lond., xxxiii. 199.	R.			Rudimentary. Longitudinal protuberance. Total atresia.	Left in anterior part of helix, almost on skin of temporal region. Fistulæ auris congenita. Cutan. sinus over sacro-coccyg. region.	Stone-deaf on right side.	
27	Kiesselbach, ref. *Arch. f. Ohren.*, xxii. 105.	U.		25.	Markedly deformed. Total atresia. External meatus (?) represented by a slight depression.			

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TABLE II.—*continued.*

No.	*Reference.*	*Side.*	*Sex.*	*Age.*	*Auricle.*	*Other Deformities.*	*Hearing.*	*Remarks.*
28	Blasius (Welcker), *Arch. f. Ohren.*, Bd. i.	B.		6 mths.	Rudimentary. Tragus fused with antitragus. Total atresia.	Nil.		Operation (Table I.).
29	Edsall, *Times and Register, N.Y. and Phil.*, i. 372.	L.	F.	8.	Helix almost absent at upper part. Tragus fused with antitragus. Ear rudimentary. External meatus shallow depression.	Nil.	Watch Politzer's acumeter not heard.	
30	Burkner, *Arch. f. Ohren*, xxii. 200.	R.	F.	8 mths.	Longitudinal ridge, upper part cartilaginous. Total atresia.	Fistulæ auris congenita just above rudimentary auricle. Small irregular appendage in front.		
31	Grunert, *Arch. f. Ohren*, xlv.	L.		Child	Rudimentary. Ear turned forwards. Total atresia.	Two fistulæ auriculæ congenita in auricle.		Deformity improved by operation (Table I.).
32	Carson, ref. *Arch. f. Ohren.*, xvi. 301.	B.		3 days.	Rudimentary. Dimple in the skin shows position of the meatus (?).			
33	Meier (Bremer), *Arch. f. Ohren.*, xiii. 276.	L.			Rudimentary. Total atresia.		Hearing not improved by operation.	(Table I.)
34	Meier (Bremer), *Arch. f. Ohren.*, xiii. 276.	R.			Rudimentary. Total atresia.		Hearing better whilst opening after operation existed.	(Table I.)
35	Moos, *Zeits. f. Ohren.*, xi. 267.	R.	M.	3.	Auricle 1½ cm. shorter and nearly a half smaller than on		Deaf-mute.	Cord wound round head at birth.

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TABLE II.—*continued.*

No.	*Reference.*	*Side.*	*Sex.*	*Age.*	*Auricle.*	*Other Deformities.*	*Hearing.*	*Remarks.*
					left side. Meatus ended in a cul-de-sac.			
36	Moos, *Zeits. f. Ohren.*, xi. 267.	R.	F.	3.	Rudimentary. Three small auricular appendages. Total atresia. Dimpling of skin in position of external meatus.	Left auricular appendage below lobule.	Deaf-mute.	
37	Moos, *Zeits. f. Ohren.*, xi.. 267.	L.	M.	7½.	Rudimentary. Longitudinal protuberance. Dimpling of skin behind tragus.		Deaf-mute.	Umbilical cord wound round head at birth. Mastoid process well developed.
38	Moos, *Arch. Otol.*, N.Y., xiii. 238.	R.	M.	30.	Atresia due to auricle being folded over tragus. Ear smaller than left side. Cul-de-sacs above and below, ending blindly.	Nil.	Hears watch well, and all tuning-forks by bone conduction.	
39	Colomiatti (Gradenigo), *Arch. f. Ohren.*, xxxiv. 281.	L.	M.	35.	Complete absence. Skin smooth. No trace of auricles.	Nil.	Bone conduction good.	
40	Gradenigo, *Arch. f. Ohren.*, xxxiv. 281.	R.		3½ mths.	Marked deformity. Lower half nearly normal. Depression pointing to external meatus.			

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TABLE II.—*continued.*

No.	*Reference.*	*Side.*	*Sex.*	*Age.*	*Auricle.*	*Other Deformities.*	*Hearing.*	*Remarks.*
41	Gradenigo, *Arch. f. Ohren.*, xxxiv. 281.	R.	M.	22.	Upper half auricle markedly deformed; lower half normal. Dimpling of skin shows (?) external meatus.		On right side, hearing-tests point to labyrinth being functionally normal.	
42	Roullard (Gradenigo), *Arch. f. Ohren.*, xxxiv. 281.	R.			Both auricles normal. Total atresia right.	Mouth large. Left side 1½ cm. from oral orifice. Dimpling of skin, surrounded by a ring of radiating hairs.		
43	Mazzoni (Gradenigo), *Arch. f. Ohren.*, xxxiv. 281.	B.	F.	19.	Absence of auricles. Total atresia.	Cartilaginous appendages on both sides.	Loud noises could be heard.	Operation (Table I.).
44	Triquet (Gradenigo), *Arch. f. Ohren.*, xxxiv. 281.	R.			Absence of auricle. Total atresia.	Between mouth and angle of jaw an appendage.		
45	Corton (Vali), *N. Y. Med. Times*, 1886.	B.	M.		Slight fold of skin represented the auricle. Dimpling showed site of (?) external meatus.			
46	Flehinger (Vali), *Med. Zeits.*, Wien, 1886.	B.			Three small elevations of skin represented auricle. Dimpling showed site of (?) external meatus.			
47	Ulhorn (Gradenigo),	B.			Auricle represented by two		Could speak and hear.	

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TABLE II.—*continued.*

No.	*Reference.*	*Side.*	*Sex.*	*Age.*	*Auricle.*	*Other Deformities.*	*Hearing.*	*Remarks.*
	Arch. f. Ohren., xxxiv. 281.				small protuberances. Total atresia.			
48	Prahl (Bremer), *Arch. f. Ohren.*, xiii. 276.	R.	M.	5.	Rudimentary. Total atresia.	Face ill-developed on right side.		
49	Polidoro Virgilius (Gradenigo).	B.			Complete absence of auricle.			
50	Lycostenus (Gradenigo).	B.			Complete absence of auricle.			
51	Stetter, "Congenital and Acquired Deformities of the Ear," Jena, 1898.							Operated on, but all failures.
52								
53								
54	My cases.							
55								
56								
57	Schwartze's Ohren. Klinik Hallé (not published).	L.	M.	1 day.	Rudimentary. Auricle represented by longitudinal ridge. Fusion of tragus and antitragus. Total atresia.	Otherwise normal.		

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