

A contribution to the pathology of congenital deafness / by Edward Cock.

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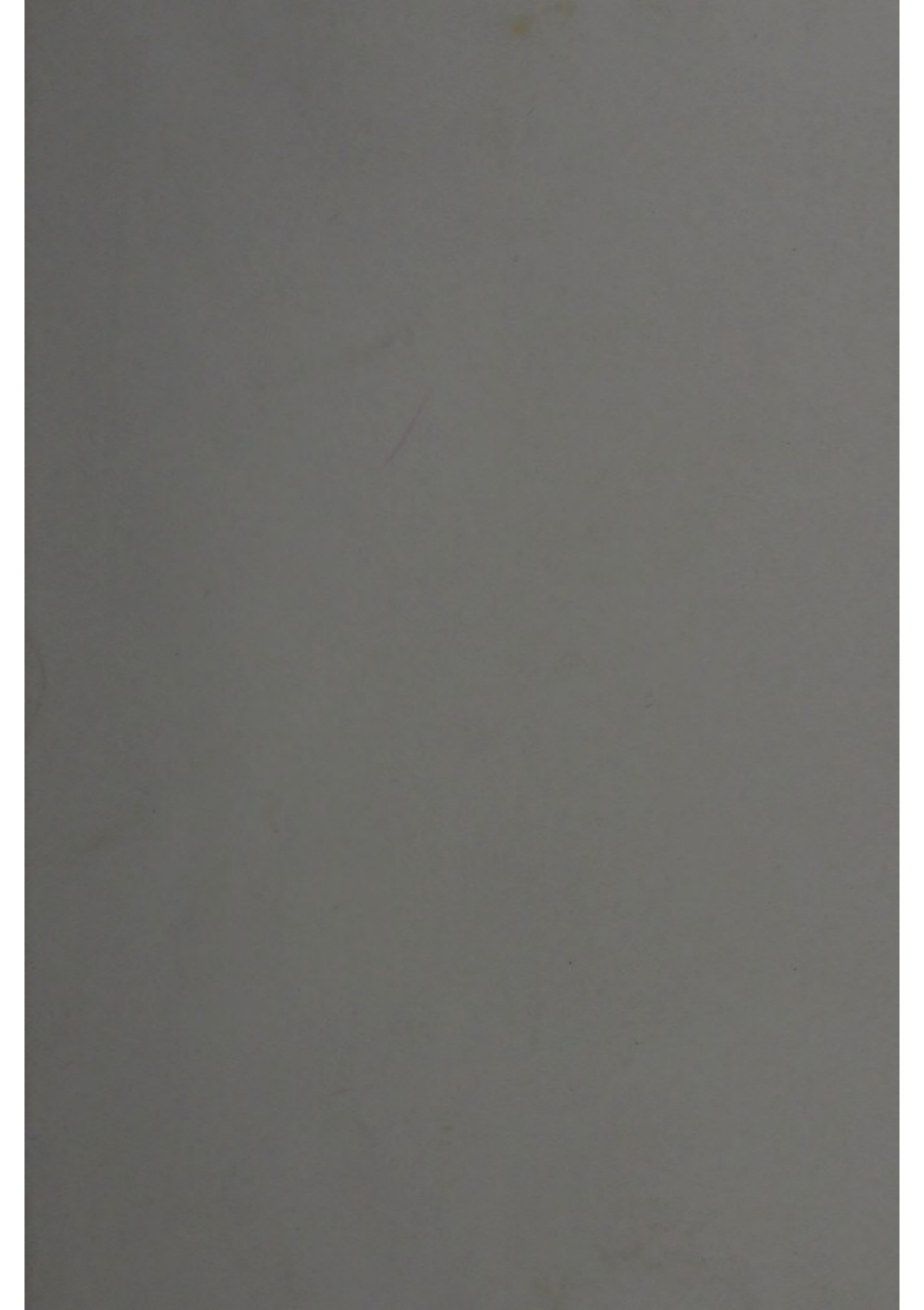
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A CONTRIBUTION
TO
THE PATHOLOGY
OF
CONGENITAL DEAFNESS.

BY EDWARD COCK.

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A CONTRIBUTION
TO
THE PATHOLOGY
OF
CONGENITAL DEAFNESS

BY EDWARD C. C. C.

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IN glancing at the history of Medicine and Surgery from the earliest ages down to the present time, we shall find, that while every other department has continued to claim the attention of scientific and intelligent men, the pathology and treatment of diseases of the ear has, until lately, been almost wholly neglected;—that while our knowledge of disease affecting every other organ and tissue of the body has expanded with a steadiness and certainty commensurate with the skill and labour which have been brought to bear on the subject, the ear still remains one of the strong-holds of the empiric, its morbid alterations but little understood, and their remedial treatment, for the most part, rather avoided than studied by those who would be most competent to undertake their management on scientific and rational principles*.

Within the last few years, the works of Itard, in France, and Kramer, in Germany, have thrown much light on the diseases of the ear: and as the subject is now beginning to claim some attention in our own country, I am induced to contribute my mite of information on that branch of aural pathology which hitherto seems to have escaped the attention, or eluded the research, of both earlier and contemporary investigators: I mean, the condition of the ear, in cases of congenital deafness.

* It is much to be regretted, that the few aurists who in this country form honourable exceptions to the above observation, have not been induced to publish the result of their experience and practice in this obscure and difficult branch of the profession.

The cause of congenital deafness has been almost universally referred to some supposed abnormal condition of the auditory nerve, inducing paralysis; although there are certainly but very few cases upon record in which the nerve has been found altered in its size or texture, unless through the agency of tubercles, hydatids, or some other cause producing mechanical pressure, or lesion of its substance.

Saunders, in his book "*On the Anatomy and Diseases of the Ear*," gives but one case as affording an explanation of the cause of congenital deafness. In this instance, the labyrinth was occupied by a soft cheesy substance; although I think it may be questioned whether this was an original formation, or a subsequent deposit of scrofulous matter*.

Itard, who has published a voluminous work on the diseases of the ear, mentions two cases of congenital deafness, in which the tympanum was filled with a calcareous deposit: also two others, in which a morbid growth had taken place from the membrane lining that cavity—"végétations produites par la membrane qui la tapise": and a fifth, where a gelatinous secretion occupied not only the tympanum, but also the canals of the labyrinth. He likewise speaks of a child, where the auditory nerve was converted into a substance resembling mucus; and of a man, in whom it was shrivelled up, and reduced to a mere thread.

Pinel relates the result of dissections, in which the water of the labyrinth was altogether deficient, leaving the cavities dry and empty; but these would appear to be cases where deafness occurred in after-life, and not where the defect was co-existent with birth.

Accounts are also on record, of congenital deafness being caused by an extension of the true skin over the membrana tympani, by the presence of polypi in the meatus externus, &c.

Dr. Kramer, of Berlin, who has certainly written the most practical work on acoustic medicine and surgery, does not appear to have extended his researches to the causes of

* When a section is made through the internal ear by means of a fine saw, the mixture of the bone-dust with the water of the labyrinth produces a soft paste, which exactly resembles the cheesy matter described by Saunders; and I think it is not improbable, that he inadvertently mistook this factitious substance for scrofulous deposit in the vestibule and cochlea.

congenital deafness, and therefore throws no light on the subject.

Through the kindness of Mr. Watson and the Medical Officers of the Asylum for Deaf-and-Dumb, I have been enabled to inspect the bodies and examine the temporal bones of most of the children who have died in that institution during the last six years. The facts elicited from the dissection of the first six cases have already appeared in the Nineteenth Volume of the Transactions of the Royal Medical and Chirurgical Society. In three of these cases, I discovered in the organs of hearing such palpable deviations from the normal structure, as served to convince me that a congenital malformation does exist much oftener than has generally been supposed; and that a careful investigation of the auditory apparatus may, in many instances, throw light upon the pathology of deafness from birth. Since the publication of the Paper, several subsequent examinations have afforded some still more curious results, which may possibly open a wider field for speculation to the physiologist, as well as to the morbid anatomist. For the purpose of rendering the present article more complete, and laying before the reader, in one view, the whole series of malformations which I have discovered from time to time, I shall take the liberty of extracting from the Medico-Chirurgical Transactions the results of my earlier labours, and prefixing them to the detail of the cases which have come subsequently under my observation.

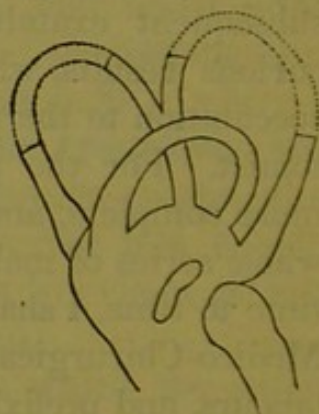
“The subjects examined were all children who died of strumous diseases of the thoracic and abdominal viscera. In three instances, one or both ears were the seat of scrofulous ulceration, affecting the tympanum and meatus externus, with partial destruction of the membrana tympani. In one case, the cavity of the tympanum, together with the mastoid cells, was completely filled with the thick cheesy deposit of scrofula, whilst a similar affection pervaded the whole cancellated structure of the petrous bone. The connexions of the ossicula auditûs were destroyed, but the bones themselves remained entire. I merely mention these facts as indicating the strumous habit of body, which I believe prevails very generally among the deaf-and-dumb: for as these affections could have existed but for a short time previous to death,

they can hardly be supposed to have had any connexion with the congenital defect in the organ of hearing.

"I may also remark, that, in all the cases examined, the petrous portions of the temporal bones exhibited more than the usual varieties of size and shape. In some, the bone was so deficient in particular spots as barely to cover the internal cavities; whilst in others there appeared a preternatural osseous development. In one instance, the petrous bone of a child, twelve years old, exceeded in size, hardness, and compactness of structure, that of any adult which I have witnessed.

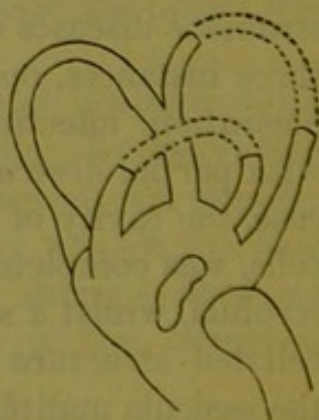
"The malformation which I discovered in two instances may be described in a few words. It consisted in a partial deficiency of two of the semicircular canals. The extremities of these tubes opening into the vestibule were perfect; but the central portions were impervious, or, rather, did not exist at all. In the first case, I had the opportunity of examining the ear from one side only*.

The vertical and oblique semicircular canals were both impervious at their central portions. The accompanying diagram will render the state of the parts easily understood; making some allowance for the false perspective which I have had recourse to, in order to exhibit all the canals in one view.



"The defective portions of the canals are traced out with dotted lines.

"In the second case, both ears were examined. On the right side, the middle portions of the oblique and vertical canals were wanting, the bone presenting an appearance like that already described. On the left side, the horizontal and vertical canals exhibited a similar imperfection. The scala tympani, likewise, was terminated, at its larger extremity, by a bony septum, which separated it from the tympanum, and occupied the situation of the membrane of the fenestra rotunda.



* This examination was made in the month of November 1832.

"In the third case, not a vestige was to be found of the fenestra rotunda on either side; the usual situation of the membrane being occupied by solid bone.

"The temporal bones of this child were exceedingly large, although soft and spongy in texture. The cavities were more than usually capacious; and the Eustachian tubes presented a remarkable development, being three or four times larger than common. On one side, the aqueduct of the vestibule readily allowed the passage of a large bristle: on the other side, the canal could not be traced through the bone, although its two extremities were more than usually expanded. Suppuration had taken place in one tympanum.

"With the exception of these malformations, and the scrofulous affections of the tympanum mentioned above, which were probably of recent occurrence, no deviation from the healthy state could be discovered in either of the five subjects examined. The Eustachian tubes were pervious; the bones, muscles, and membranes, entire and natural: the labyrinths were filled with their transparent fluid. In no instance did the auditory nerve present any peculiarity, although carefully traced from its origin to its distribution. The chorda tympani was present in every instance; but I cannot vouch for the integrity of all the little nervous fibrillæ, which pass into the tympanum, and ramify on its walls, requiring the aid of a microscope for their dissection.

"In addition to these three cases of malformation, I may state a fourth; which was dissected by my friend Mr. Dalrymple, and is now in his possession. In this instance, the aqueduct of the vestibule was so large as to admit the passage of a small probe; whereas, in the natural state, a fine hair can with difficulty be introduced into the canal."

In the same Number of the *Medico-Chirurgical Transactions* will be found a communication from Mr. Thurnham; which I read with considerable interest and satisfaction, since it describes a similar malformation of the semicircular canals discovered in the ear of a deaf-and-dumb person. I am not aware of any other investigations having been made in this country.

I have regretted, that, in the examination of the different ears which afforded the above results, the cochlea was not in every instance traced out with all the care that might have

been bestowed upon it: for, as in the outset of my researches, my attention and views were more particularly directed to the tympanum and its appendages, it was not until I became aware of the abnormal condition of the semicircular canals that I determined, in every future case, to undertake a deliberate and patient investigation of the labyrinth. It is therefore possible, that certain deviations in the organs may have been overlooked, which would not now escape my observation.

Since the year 1835, the Asylum for Deaf-and-Dumb has furnished me with seven additional post-mortem examinations, the particulars of which I will now proceed to describe.

CASE 1.

A boy, 12 years of age, who had sunk rapidly under fever, attended by acute cerebral symptoms.

Great vascularity throughout the brain and its membranes: effusion between the arachnoid and pia mater, and within the ventricles. The origin of the auditory nerve presented nothing unusual.

The left temporal bone.—The tympanum was completely filled by spongy and highly-vascular granulations, which appeared to grow from the mucous membrane lining the cavity; and to derive their vessels, which were numerous, large, and injected with blood, from the same source. This adventitious growth also occupied the mastoid cells, extended into the Eustachian tube, and adhered closely to the membrana tympani and the chain of small bones: it had no malignant character, but bore considerable resemblance to the exuberant granulations which are so abundantly produced after fracture of the skull, attended with lesion of the brain, or after-abscess of the testicle. The petrous bone was remarkably ill shaped, very narrow from before to behind: the jugular fossa exceedingly large and excavated, so as to encroach upon the tympanum and Fallopian canal; the latter of which was laid open towards the fossa, when the periosteum was removed. The groove for the superior petrosal sinus was very broad and deep: in fact, the petrous portion was so scanty in its dimensions, and presented so little solid bone, that it was evident, on viewing its exterior, that the semi-

circular canals could not exist in their perfect state. The meatus auditorius internus was represented by a narrow slit; and the portio mollis became pulpy and transparent, as it descended into the canal. On cutting open the bone, the labyrinth presented the following appearances. The auditory canal, instead of being terminated by the cribriform plate forming the base of the modiolus, opened at once into a cavity of a somewhat conical shape; communicating with the vestibule by a very large aperture; and also with the tympanum, by means of the fenestra rotunda; through which last, the vascular granulations already described had found their way. This cavity, in fact, represented the mere external shell of a cochlea, but without a vestige of modiolus, spiral lamina, or scalæ: the auditory nerve entered it, and apparently expanded on its walls. The vestibule, or what answered to it in situation, was rendered irregular; on the one hand, by forming a continuous cavity with the imperfect cochlea; and on the other, by extending itself outwards, so as to include that portion of bone usually embraced within the concavity of the horizontal semicircular canal. Not a trace, however, existed of either the horizontal or the oblique canal: the anterior opening of the vertical canal alone was present; but the canal itself suddenly stopped, after having completed about half its natural course. There was no aquæductus vestibuli. (Vide Plate, Fig. 1.)

The right temporal bone.—The tympanum was occupied by an adventitious growth, precisely similar to that on the left side. The petrous bone was more regular in its external configuration. The aquæductus vestibuli consisted of a very large funnel-shaped canal; terminating, in the vestibule, by an oval opening, sufficiently wide to admit the eye of a small probe. The semicircular canals were perfect; and the vestibule natural as to size and form, but communicating, by a very large opening, with a cavity somewhat resembling that on the left side, and representing the shell of a cochlea which contained the rudiment of a modiolus (that is to say, the base shutting out the cavity from the meatus internus, and allowing the passage of the auditory nervous filaments), and an attempt at the formation of a spiral lamina, which, however, did not make a complete turn, and consisted chiefly of

membrane, which disappeared on the parts becoming dry from exposure. The fenestra rotunda could hardly be said to exist. (Vide Plate, Fig. 2.)

CASE 2.

A boy, about twelve years old, who had been deaf and dumb from birth, and subject to epileptic fits.

All the vessels of the body, but particularly those about the head, were remarkably distended with fluid blood, which flowed copiously from every incision. The brain was exceedingly hard and firm, resembling cheese in its consistence: its vessels were strongly injected, and the ventricles contained a considerable quantity of fluid; but no tumour, or other unnatural appearance, could be discovered within the cranium. The mesentery was studded with innumerable bony deposits, contained between its layers; but which, apparently, had not interfered with the function of the lacteal absorbents, as the lad was remarkably well grown, and stout for his age. The temporal bones were exceedingly large, massive, and heavy; and in this respect would have been remarkable, had they belonged to an adult instead of a child. The cancellated structure which generally exists in the petrous portion was entirely deficient; and the bone presented, throughout, a dense compact texture, of almost iron hardness, and resembling the closest-grained ivory. The meatus externus, on both sides, was partly filled with dry inspissated cerumen, and the position of the membrana tympani was nearly horizontal.

Interior of the right ear.—The cavity of the tympanum was completely filled with dense fleshy granulations, adhering most firmly to the walls, completely enveloping and concealing the bones, adapting themselves to all the crevices and irregularities, and extending into the mastoid cells and the commencement of the Eustachian tube. A small quantity of thin purulent fluid escaped, when the roof of the tympanum was removed. The membrana tympani was exceedingly convex, being drawn inwards until it nearly touched the promontory; thus diminishing the size of the cavity, and pushing the bones out of their place, so that the extremity of the manubrium of the malleus and the long crus of the incus were in close contact, and carried to the edge of the

fenestra ovalis. I could not discover any vestige of the sides of the stapes, although the plate of bone forming its base still continued to occupy the foramen ovale. I should, however, remark, that the extreme difficulty of removing the adventitious growth which surrounded the bones rendered the precise state of the stapes somewhat uncertain; and it is just possible that it might have been broken up, in the attempt to clear the tympanum from the granulations which filled it. The vestibule was spacious, and contained the usual limpid fluid; the semicircular canals were pervious and natural; the entrance to the cochlea presented the ordinary appearance, when viewed from the vestibule, but terminated suddenly, in a blind extremity about a line from its commencement. Of the cochlea itself no trace existed, its situation being occupied by solid hard bone. The fenestra rotunda, when traced from the tympanum, terminated in a minute cavity; which might be considered as the rudiment of a scala tympani, and into which the granulations already alluded to had entered. The auditory nerve was remarkably hard and small, and was entirely distributed to the vestibule; that portion, which in the normal condition pierces the modiolus to supply the cochlea, being, of course, deficient.

Interior of the left ear.—The tympanum presented nearly the same appearance as on the right side; with this addition, that the membrana was closely adherent to the promontory, producing a still greater diminution in the size of the cavity, and a greater distortion of the bones from their natural situation. The incus was ankylosed to a little spicula of bone, which projected from the inner wall of the tympanum, just above the Fallopian canal. The labyrinth presented nothing unusual, if we except, that the modiolus of the cochlea was smaller than natural, and that the granulations from the tympanum had passed through the fenestra rotunda into the commencement of the scala tympani. The auditory nerve was small and hard, by far the greater portion of it going to the vestibule.

CASE 3.

A boy, between 12 and 13 years of age, who died of mesenteric disease, accompanied by extensive ulceration of the mucous membrane lining the cæcum and ascending colon.

The temporal bones were well formed, as regarded their external configuration; and the internal organs of hearing presented no appreciable deviation from the normal condition.

CASE 4.

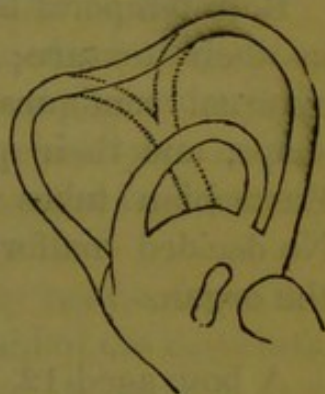
A girl, 13 years of age, who died of phthisis.

The right ear.—The tympanum partly filled with soft vascular granulations; a muco-purulent fluid occupying what remained of the cavity. The Eustachian tube very large, and containing a quantity of semi-inspissated mucus. Bones, muscles, and nerves, perfect. Vestibule spacious, semicircular canals perfect, with very large ampullæ. The communication between the vestibule and cochlea wider than usual. The cochlea was natural, as regarded its external configuration or shell; but its interior was very deficient. Thus, on tracing the scala tympani from the fenestra rotunda, it was found to take about half a turn round the base of the modiolus, and then terminated in a blind extremity. The scala vestibuli, on the other hand, formed about one-third of a turn; and then opened into a cavity which constituted the remaining part of the cochlea, as represented by its external shell. In fact, the base of the cochlea alone was perfect, the remaining portion consisting merely of the cavity just mentioned: not only was the greater portion of the spiral canals deficient, but the blind extremity of the scala tympani prevented any continuity or communication between them.

The petrous bone, more especially in the neighbourhood of the tympanum, was unusually loose and cellular, presenting numerous irregular cavities, which apparently communicated with the tympanum, and were filled with the same muco-purulent fluid: they seemed a sort of extension of the mastoid cells.

The left ear.—Petrous bone irregular in shape; the meatus externus remarkably narrow, representing an elongated ellipse, when cut across. No external appearance of aquæductus vestibuli. The lining membrane of the tympanum thickened, but not granular; the cavity, together with the mastoid cells and Eustachian tube, filled with sopy, adhesive, and slightly purulent mucus. The interior of the vestibule presented but four openings of the semicircular canals,

instead of five; that which is common to the posterior extremity of the vertical, and the superior extremity of the oblique, being deficient. On tracing the vertical canal from its anterior opening, it was found to terminate, not by returning to the vestibule, but by becoming continuous with the upper part of the oblique canal, which latter opened below, as usual. The two canals, thus united, formed one irregular tube, which extended across the superior and posterior part of the petrous bone, and presented the appearance indicated in the accompanying diagram. The cochlea presented much the same appearance as on the right side; the lamina spiralis making about half a turn round the base of the modiolus, and separating the scala tympani (which reached to about the same extent) from an irregularly-shaped cavity into which the scala vestibuli opened soon after leaving the vestibule. (Vide Plate, Fig. 3.)



CASE 5.

A boy, 11 years old, who died of tubercular phthisis.

The entire temporal bones, but more especially the petrous portions, were most remarkable for their enormous size. In development, solidity, and weight, they exceeded any that I have ever seen. They were totally destitute of the usual cancellated structure; and the different cavities lay deeply imbedded in a thick mass of compact bone, the excessive hardness of which rendered the examination a work of great difficulty and labour. The external meatus was small, and contracted on both sides. The jugular fossæ were deep and large. The fossæ for receiving the semilunar ganglia of the fifth pair were also much excavated. On the left side there was a separate canal for transmitting the motor portion of the nerve. On the right side, instead of a complete canal for the passage of the carotid artery, there was merely a groove, which was completed into a canal by the sphenoid bone. On the left side there was neither canal nor groove: the artery must therefore have traversed the sphenoid bone, in order to reach the cranium. The Eustachian tubes were

more circular than common, contracted in size, and very irregular as regarded their bony parietes. This seemed to depend on the great accumulation of bone around them, which encroached on their calibre. The only remarkable deviation from the normal structure observed within the organ was the absence of the stapedius muscle on the right side.

CASE 6.

A lad, aged 16, who died of tubercular phthisis.

Both temporal bones were exceedingly massive, and hard in their texture; and very ill-formed, as regarded their external configuration. The tympana were irregular in shape, and their parietes more than usually rough. The Eustachian tubes were small, and irregularly contracted. No decided malformation could be detected in any part of the organ.

CASE 7.

A boy, aged 12.

The temporal bones were unfortunately subjected to the action of dilute muriatic acid, for some time previous to examination. This was done for the purpose of softening them, and rendering the dissection more easy. It, however, had the effect of so completely altering and obscuring the interior of the ear, as to render an accurate investigation impossible. The auditory nerves were healthy and natural, and the external configuration of the bones presented nothing unusual. The tympana had partly undergone the granular alteration so often alluded to, but were otherwise in a normal condition. The vestibula and semicircular canals were perfect. The cochleæ were so completely altered by the acid, as to preclude the possibility of detecting any but a very decided malformation; but I am not warranted in saying that they were otherwise than natural.

I have now concluded the detail of facts which have resulted from my inquiries; and can only regret that the physiological and practical inferences to be drawn from them are so few and so vague. I am unwilling to enter into the field of conjecture and speculation; yet, nevertheless, I may venture to throw out a few hints and observations, which may

perhaps be taken up, and turned to some account, by those whose attention has been more exclusively directed to these subjects. The present state of our knowledge respecting the exact function which the different parts of the ear exercise, in the appreciation of sound, is so obscure and limited, that it is almost presumptuous to hazard more than a conjecture as to the effect likely to be produced by the different malformations have just described; for until we can assign a probable office to the various divisions of this complicated organ, it is useless to attempt going far beyond a mere statement of facts.

The little which is known and acknowledged respecting the physiology of hearing, or which may be demonstrated by the application of certain indisputable physical laws to the anatomical structure of the ear, may be summed up in a few words.

The phenomenon of hearing undoubtedly results from the impression produced by the motion of the fluid of the labyrinth on the expansion of the auditory nerve lining its cavities, and thence propagated to the sensorium. The necessary motion, or undulation, is imparted to the labyrinthine fluid by a series of vibrations emanating from the body which produces the sound (as a bell for instance), and mechanically conveyed to the vestibule. There is no difficulty in tracing the course of these vibratory movements, if we follow the connexion of the different parts of which the ear is composed. Thus vibration is propagated through the air, from the sounding body, to the auricle; and thence transmitted, through the meatus externus, to the membrana tympani; while the oscillations produced on this last are again conveyed, by the connecting chain of bones, across the tympanum, to the membrane of the fenestra ovalis, which may be considered as the entrance to, or commencement of, the labyrinth; one of its surfaces being attached to the base of the stapes, and the other in contact with the labyrinthine fluid. It follows, therefore, that every change which takes place in the form of a bell, when it is struck so as to produce a sound, will be succeeded by a corresponding impulse on the membrane of the fenestra ovalis. Again, if we imagine the cochlea to be unfolded, or deprived of its convoluted form, we shall find that the vestibule, the scala vestibuli, and scala tympani, constitute together a

continuous canal, varying in its calibre at different parts, commencing at the fenestra ovalis, and terminating at the fenestra rotunda; and, that the sides of this canal are firm and solid, while its two extremities are shut in by yielding structures, viz. the membranes occupying the two fenestræ. The construction of the labyrinth may be familiarly explained by comparing it to a long metallic tube filled with water, and secured at either end by bladder or caoutchouc. If an impulse be given to the yielding material at the one extremity of such a tube, the impression must necessarily be conveyed to the yielding material at the other extremity, and the whole column of intermediate water will be put in motion. It has been shewn in what manner vibrations are conveyed to the fenestra ovalis; and there can be little difficulty in conceiving how the impulse produced on the membrane occupying that opening must be propagated through the vestibule, through the scala vestibuli and scala tympani, until it is received on the membrane of the fenestra rotunda. The motion of the fluid causes a concussion on the nervous expansion lining the walls of the labyrinth; which concussion, being referred to the brain, we denominate 'the sense of hearing.'

In accordance with the physiological explanation just given, the organ of hearing may be said to consist of three parts or sets of structures:—1. The external ear and tympanum, being the apparatus by which vibrations are communicated to the labyrinth, at the same time that they may be altered or modified in their intensity. 2. The water of the labyrinth, which is set in motion by the vibrations so communicated to it. 3. The nervous expansion which surrounds the water, and, appreciating the motion of the fluid, produces the sense of hearing.

It follows, that, for the due exercise of the sense, three conditions of the organ are required:—1. A capability to convey the vibratory oscillations from the sounding body to the labyrinth. 2. That the fluid should be susceptible of undulation. 3. That the nervous expansion should be in a state which enables it to appreciate the motion of the fluid, and transfer the impression to the sensorium. For the purpose of ascertaining how far the malformations, and morbid altera-

tions, which I have described in this Paper, are likely to interfere with the exercise of either of the functions, I have endeavoured to explain, we may class the different abnormal appearances under the following heads:—

1. An adventitious growth filling up, and thus obliterating more or less completely, the cavity of the tympanum, enveloping the chain of bones, and encroaching upon the openings of the Eustachian tube, mastoid cells, and fenestra rotunda.

2. Deficiency of the fenestra rotunda; thus rendering one extremity of the labyrinthine canal solid and immoveable, instead of yielding.

3. Partial or complete deficiency of the spiral canals of the cochlea.

4. Preternatural enlargement of the aquæductus vestibuli.

5. Deficiency of the semicircular canals.

6. Unnatural solidity or hardness of the temporal bone.

It will be remembered, that, in most of the cases detailed above, several of these appearances existed in combination.

With respect to the granular vegetations occupying the cavity of the tympanum, I think that they would be all sufficient to occasion deafness, by preventing the transmission of vibration from the external ear to the labyrinth; while their presence at the entrance of the Eustachian tube, and at the foramen rotundum, would certainly destroy the function of two parts which are essential to the exercise of the sense of hearing. I must, however, leave the question open, as to whether this adventitious growth was coëval with birth or the period of infancy; or whether, on the other hand, it was a subsequent formation, depending upon and accruing from a prior defect in the sense of hearing, from some unknown cause;—whether, indeed, it supervened upon deafness, and was merely an effort of nature to obliterate a cavity whose function of conveying and modifying vibration had become useless, in consequence of the previous inefficiency of that part of the organ by which the vibratory oscillations were to become converted into sound, and appreciated as such by the sensorium.

With respect to the deficiency of the fenestra rotunda—or, in other words, the substitution of solid bone in the place of

the membrane which, in the natural state, occupies that opening—such a malformation must necessarily prevent all motion in the fluid of the labyrinth, by depriving the labyrinthine canal of one of its yielding extremities, and thus cutting off from the expansion of the auditory nerve that medium of communication (*viz.* the motion of the water) through which it receives its impression. A similar effect will be produced where the cochlea is entirely wanting; or where the scala vestibuli and scala tympani are merely rudimentary, and do not become continuous with each other.

I will not undertake to decide how far a preternatural enlargement of the aquæductus vestibuli may be in itself sufficient to render the organ of hearing useless: I can only venture upon an explanation why such a malformation may possibly be the cause of deafness. The aquæductus vestibuli may possibly serve the office of a safety-valve to the delicate structure of the labyrinth; and, under intense vibration, may suffer a small portion of fluid to escape from the vestibule, when the motion imparted to the water through the medium of the fenestra ovalis is so violent as to endanger the integrity of the nervous membrane lining the cavities*: but if the aqueduct be preternaturally large, every, even the slightest, vibration will be attended with a discharge of fluid through its canal; and thus the auditory impression which, by the agency of the water, ought to be propagated throughout the whole extent of the labyrinth, will reach no farther than the vestibule itself.

I have no explanation to offer respecting the deficiency which I have described as occasionally occurring in the semi-circular canals: I cannot even speculate on the subject. The function of these appendages to the labyrinthine canal is, as far as I am aware, totally unknown; and the effect likely to be produced by their malformation must remain equally obscure. Little doubt, however, can be entertained of their

* I have not, as yet, been able to determine, to my own satisfaction, whether the aquæductus vestibuli and aquæductus cochleæ are really what their name implies; or whether, as many anatomists suppose, they are merely canals, traversing the bone, for the purpose of transmitting vessels of the cavities with which they communicate.

importance, and that they are essential, not only to the perfection of hearing as enjoyed by man, but to the appreciation of sound itself, as possessed by inferior beings probably not endowed with the same powers of discrimination; since we find these tubes fully developed in many of the lower animals where the tympanum and cochlea are altogether wanting, or exist only as rudimentary appendages*.

In laying the foregoing facts and observations before my readers, I by no means wish that a greater degree of importance should be attached to them than they deserve. I am aware that they are far from elucidating the pathology of deafness, and that they do little more than throw a ray of light over a subject which was previously involved in almost total obscurity. They may possibly excite inquiry in others; and, eventually, be the means of adding to our stock of knowledge respecting the different forms of this disease. Much remains to be discovered, before the causes of congenital deafness can be satisfactorily explained: for I feel assured, that in some, if not in many instances, the deviations I have noticed are quite insufficient to account for a total privation of the sense of hearing; and the fact of several cases having occurred in which no malformation could be detected, proves that there must be some other defect, either organic or functional, the demonstration or explanation of which has, as yet, eluded our research. Still, although I will not venture to assert that the malformations I have brought to light have been, in any instance, the sole cause of deafness, yet I must believe and maintain, in opposition to the opinion of some that they form a part of a certain pathological condition of the organ, the seat of which is not sufficiently obvious to be detected; but whether it exists in the brain, or in the temporal bone, or in both, can only be ascertained by future investigation. I utterly reject the idea, that these malformations are mere coincidences, which ought not to be considered as having a reference to the imperfection or annihilation of the sense of hearing—that they are, in fact, analogous

* For a short sketch of the comparative anatomy of the ear, tracing the development of its different parts through the various classes of animals, see *Medico-Chirurgical Transactions*, loc. cit.

to abnormal deviations in other parts of the body, which are discovered after death, but which produced no symptoms during life, as regarded the functions of the organ in which they occurred. A reference to the series of preparations in museums, my own experience in the dissection of ears, and the evidence of those whose constant occupation it is to prepare anatomical illustrations of the organ, prove, almost to a certainty, that malformation never is found, except in cases of deafness.

Although I cannot flatter myself that the issue of my labours has afforded any information which can be attended with a beneficial result in a practical point of view;—although the establishment of a fact, which has been denied, that congenital malformation may and frequently does exist, would rather discourage us from adopting remedial measures, than serve to indicate any mode of treatment for the relief of the patient;—yet I am not without the hope, that my investigations may still prove of some value, in the hands of those who undertake the treatment of deafness on scientific principles, and with a view to the benefit of their patients as well as their own emolument. I trust they may stir up a spirit of inquiry into the symptoms and degrees of deafness; which may lead to the establishment of a more accurate and efficient diagnosis; and enable the aurist to distinguish between those cases which are irremediable by art, and those which may be expected to derive benefit from judicious treatment. There can be no doubt, that, even in cases of congenital deaf-dumbness, the derangement of the organ, whether functional or organic, may exist in different degrees of intensity; that, in some instances, the mischief consists merely of an imperfection or dulness of hearing, which, from neglect, has entailed the additional misfortune of dumbness on the sufferer; whilst in others it amounts to a total and irremediable privation of the sense. The power of discrimination can only be acquired by the intelligent and scientific aurist, and must be the result of much experience and patient investigation; but the knowledge thus acquired would frequently prove the means of rescuing the hopelessly deaf and dumb from the constitutional injury and bodily suffering consequent upon tedious and painful courses of treatment, indis-

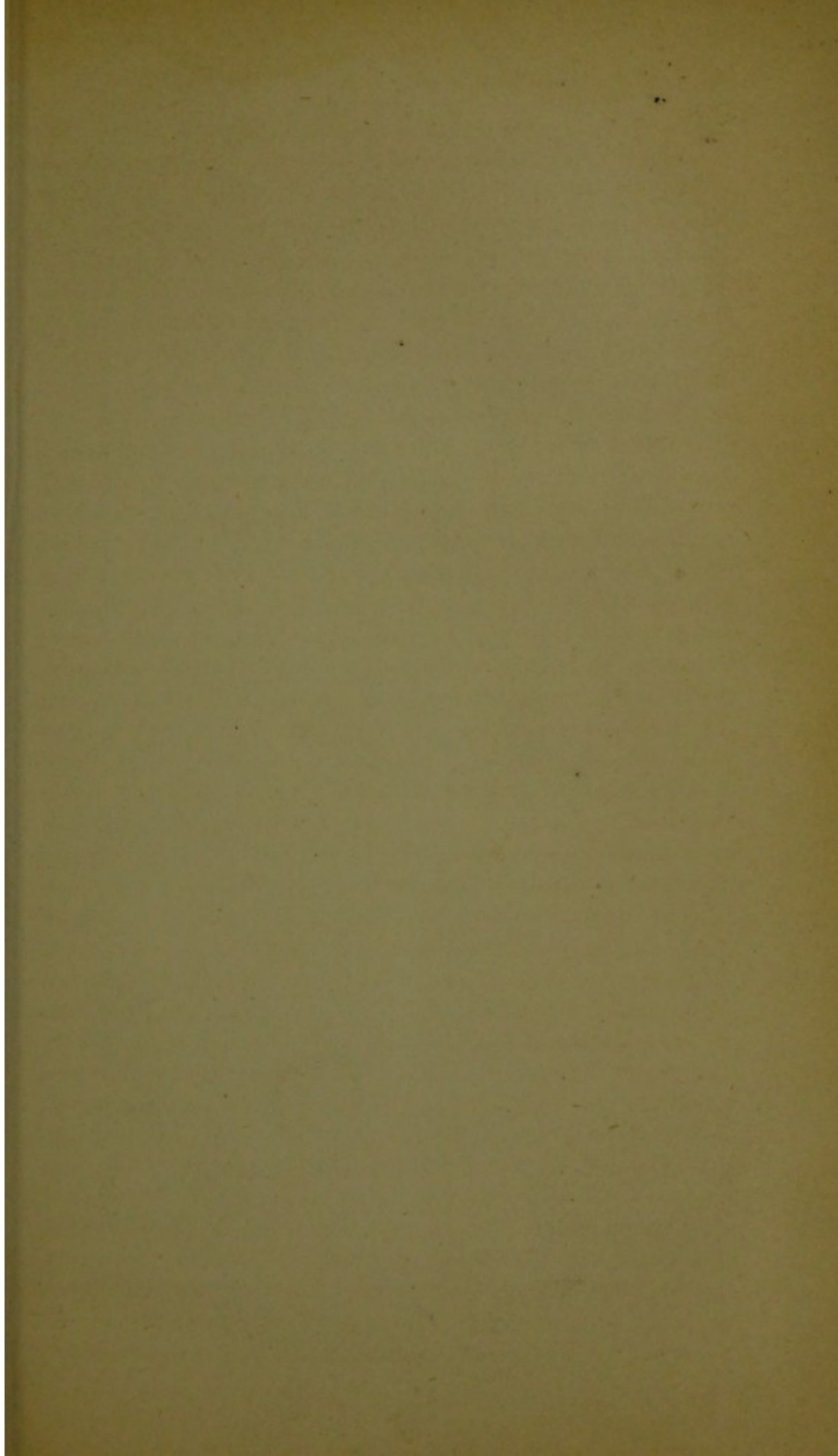


Fig. 3.

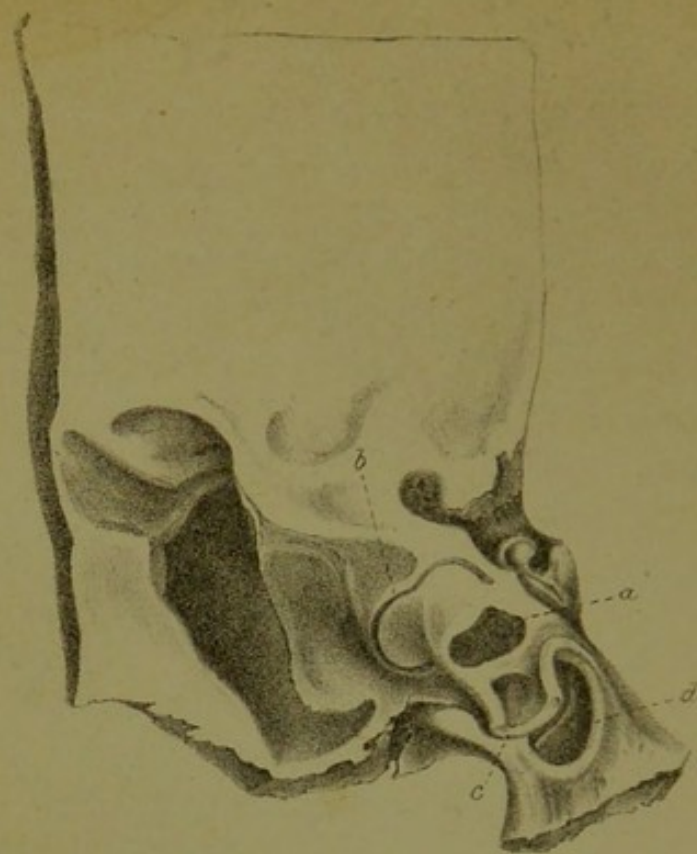


Fig. 1.

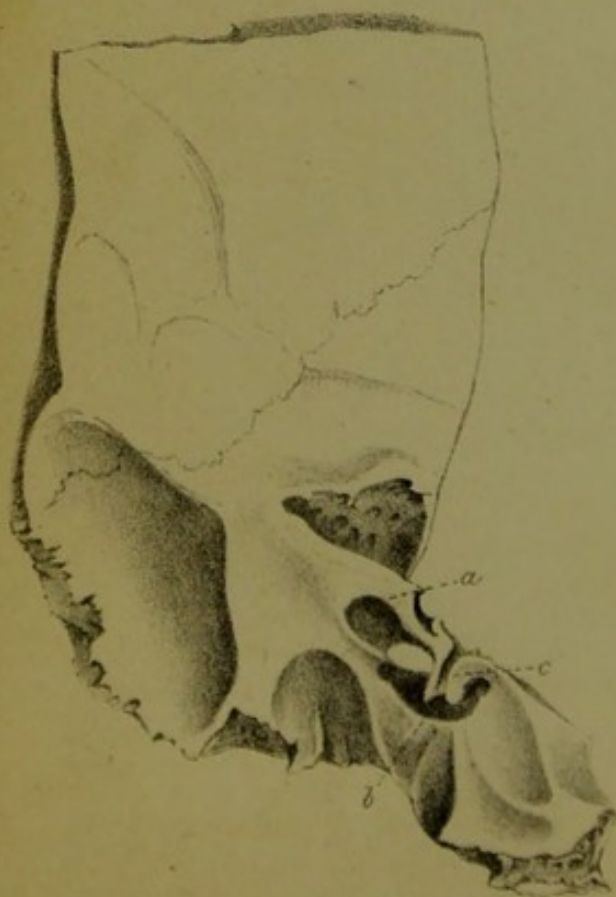
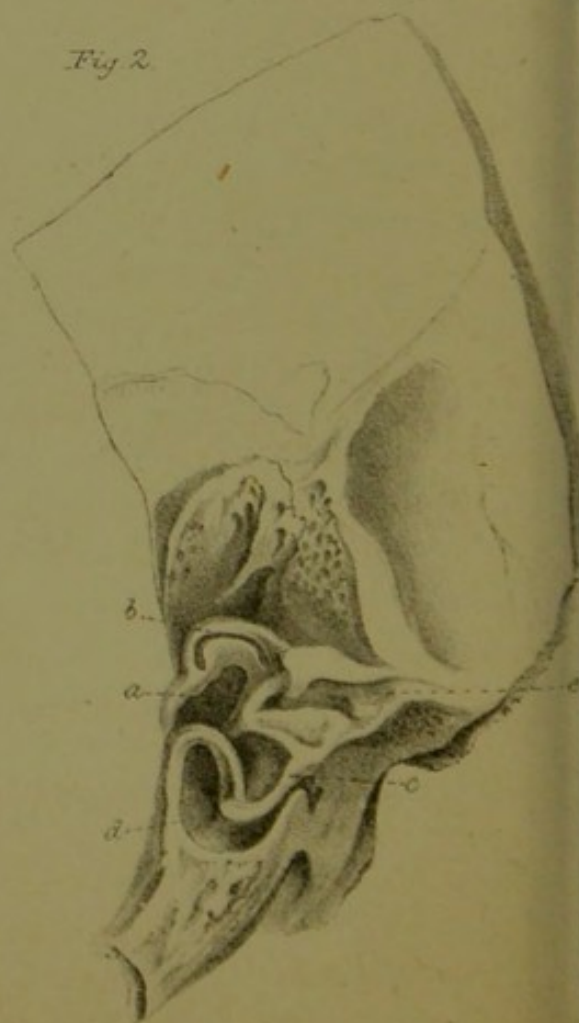


Fig. 2.



criminally adopted by the ignorant and pretending empiric. It would, in many cases, suggest to the upright and humane practitioner the more eligible plan of attending to the cultivation of the other senses through which the mind of man holds intercourse with the external world; and devoting to the mental improvement and education of the sufferer that time and attention, now too often wasted in the employment of means which only serve to impair the mental and bodily powers, while they impose on the credulity of the patient or his friends.

EXPLANATION OF THE PLATE.

Fig. I. represents the left temporal bone of Case 1. p. 294. The upper portion of the petrous bone has been removed by a section carried through the irregular cavities representing the vestibule and cochlea, both of which are thus laid open.

a The vestibule.

b The cochlea.

c The commencement of the Fallopian canal, laid open by the section.

Fig. II. represents the right temporal bone from the same subject; shewing the vestibule, vertical semicircular canal, and meatus auditorius internus laid open, together with the malformed cochlea and aquæductus vestibuli.

a The vestibule.

b Vertical semicircular canal.

c Meatus auditorius internus.

d Irregular cavity, representing the cochlea.

e Aquæductus vestibuli, preternaturally large.

Fig. III. represents the left temporal bone of Case 4. p. 298; shewing the vestibule and meatus auditorius internus laid open, together with the malformed semicircular canals and cochlea.

a Vestibule.

b Abnormal continuity between the vertical and oblique semicircular canals.

c Meatus auditorius internus.

d Irregular cavity representing the cochlea, with a rudimentary lamina spiralis.

and is entirely neglected by the ignorant and prejudiced empiric. It would, in many cases, suggest to the upright and humane physician the most eligible plan of attending to the cultivation of the other senses through which the mind of man holds intercourse with the external world; and devoting to the mental improvement and education of the subject that time and attention now too often wasted in the employment of means which only serve to impair the mental and bodily powers, while they inure to the crying of the patient or his friends.

EXPLANATION OF THE PLATE.

Fig. I. represents the left temporal bone of Case I. p. 214. The upper portion of the petrous bone has been removed by a section carried through the petrous cavity representing the vestibule and cochlear, both of which are thus laid open.

a. The vestibule.

b. The cochlea.

c. The communication of the Fallopian canal, laid open by the section.

Fig. II. represents the right temporal bone from the same subject; showing the vestibule, vertical semicircular canal, and incus and malleus in situ, together with the mallocochlear cord and the mallocochlear cord.

a. The vestibule.

b. The vertical semicircular canal.

c. The incus and malleus.

d. The mallocochlear cord.

e. The petrous cavity, representing the cochlea.

f. The mallocochlear cord, posteriorly laid open.

Fig. III. represents the left temporal bone of Case I. p. 215; showing the petrous cavity and incus and malleus in situ, together with the mallocochlear cord and the mallocochlear cord.

a. The vestibule.

b. The vertical semicircular canal.

c. The incus and malleus.

d. The mallocochlear cord.

e. The petrous cavity, representing the cochlea.

f. The mallocochlear cord, posteriorly laid open.



