Some remarks on mal-formation of the internal ear, being the result of post-mortem investigations performed in five cases of congenital deafness / By Mr. Edward Cock. Communicated by Dr. Bright.

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BEING THE RESULT OF

POST MORTEM INVESTIGATIONS

PERFORMED

IN FIVE CASES OF CONGENITAL DEAFNESS.

By MR. EDWARD COCK,

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COMMUNICATED BY

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READ DECEMBER 9TH, 1834.

PERHAPS there is no part of the human body which has so little engaged the attention of the pathologist, or which has afforded such slender encouragement to his research as the ear. The minute delicacy and complicated nature of the organ, the difficulty which attends its examination, and the patience required for a thorough investigation of its different parts, will perhaps account for the great obscurity which at present envelopes the pathology of congenital deafness.

When structures so numerous and diversified, are found assembled within a small portion of bone of compact texture and ivory hardness,—when bones, ligaments, joints, muscles, membranes, secreting tissues, vessels, nerves, and fluids are compressed within so small a compass,—it may be conceived how much time and labour must be expended, and how many ears must be dissected, before we can gain even a superficial and imperfect knowledge of this organ in its healthy state, and how difficult to appreciate every morbid change, or every congenital mal-formation which possibly may occur in its structures.

Under these circumstances, it is not surprising that congenital deafness should have been almost universally ascribed to paralysis of the auditory nerve, although I believe there is scarcely a case 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.

At the request of Dr. Babington, who is physician to the Asylum for the Deaf and Dumb, I have, within the last two years, taken the opportunity of examining the temporal bones of five children who died in that institution, and in two of these have detected such palpable deviations from the normal

structure, as would indicate that a congenital malformation does exist oftener than is generally supposed, and therefore that to this cause, many cases of deafness may reasonably be ascribed.

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.

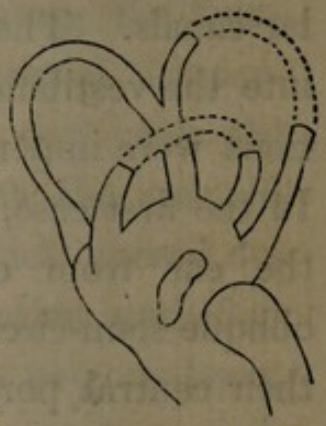
The mal-formation which I discovered in two instances, may be described in a few words. It consisted in a partial deficiency of two of the semi-circular canals. The extremities of these tubes opening into the vestibule were perfect, but the central portions were impervious, or rather did not exist at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

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

ing 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.



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 two cases of mal-formation I may state a third, 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.

The present state of our knowledge, respecting the function which the different parts of the ear exercise in the appreciation of sound, is so vague and limited, that it is impossible to hazard even a conjecture, as to the effect likely to be produced by the mal-formations I have just described, for, until we can assign a probable office to the various divisions of this complicated organ, it is useless to attempt more than a plain statement of facts. Little doubt, however, can be entertained of the importance of the semicircular canals, 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.

The earliest formation of an acoustic apparatus is found in the crustaceous animals, and consists of a membranous sac filled with fluid, and containing a little bone or some cretaceous matter, on which the auditory nerve becomes distributed. Such a structure is seen in the crab and lobster, and appears to correspond with the vestibule of the mammalia.

If we ascend the scale of organization, the next

class, fishes, present a development not only of vestibule, but of three semicircular canals, which in some of the tribes are very large.

The reptiles are furnished with an organ in most respects similar to the fishes, but in some of them, a faint trace of a rudimentary cochlea and tympanum becomes apparent.

The succeeding class, or the birds, have the vestibule and semicircular canals perfectly developed; they likewise possess a cochlea approaching in its form to the spiral canals which are found in the highest orders, together with a tympanum and external meatus.

Lastly, the class mammalia exhibits, with slight modifications, the perfect development and elaborate construction which characterize the ear of man.

From this brief and consequently general sketch of the ears of animals, it will be seen that there is only one class (and that the lowest in which an auditory apparatus can be traced) which does not possess semicircular canals: from which we may be led to infer, that, with the exception of the vestibule, they are, of all the parts composing this complicated organ, those most essential to the appreciation of sound.

With respect to the last case of mal-formation,

which I quote upon the authority of Mr. Dalrymple, perhaps something like an explanation of the possible cause of deafness may be ventured upon. The aqueductus vestibuli may probably 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, as in this case, 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, through the agency of the water, ought to be propagated throughout the whole extent of the labyrinth, will reach no further than the vestibule itself.

St. Thomas's Street,
October, 1834.

Since the foregoing remarks went to the press, another post-mortem examination of a child, from the Deaf and Dumb Asylum, has furnished results, which I think tend to throw additional light on the pathology of congenital deafness.

In this 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 effect of such a mal-formation would probably be, to prevent the vibratory impression received on the membrane of the fenestra ovalis from being propagated through the vestibule and the canals of the cochlea; for if we consider the labyrinth of the ear as a long osseous tube, commencing at the fenestra ovalis, and terminating at the fenestra rotunda, (and thus closed at both extremities by membrane,) then if a solid structure be substituted for the yielding material, which, in the natural state, closes one extremity of the canal, viz., the round opening, the motion imparted by the ossicula to the membrane of the fenestra ovalis, would no longer produce that undulation of the fluid through the labyrinth, which appears essential to the appreciation of sound.

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.

Aug. 10, 1835.

G. Woodfall, Printer, Angel Court, Skinner Street, London.