A case of acquired total deafness, the result of inherited syphilis; with post-mortem / by Walker Downie.

From "THE PRACTITIONER" *for March*, 1896.

A CASE OF CONGENITAL MALFORMATION OF THE EARS.

BY WALKER DOWNIE, M.B., F.F.P.S.G.,

Lecturer on Diseases of Throat and Nose, Glasgow University; Hon. Aurist, Royal Hospital for Sick Children, etc.

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The following case came under the observation of Dr. Ferguson, of Perth. The child, a male, which was born at full term on September 3rd, 1895, was the result of the mother's fourth pregnancy. Dr. Ferguson, who has attended the mother at each of her confinements, says that hitherto there had been no departure from the normal configuration in any of her offspring. There is no consanguinity between the parents, and both are healthy and well formed; and there is no evidence of physical deformity in the more extended family history. The child at birth was well nourished, of average size, and, apart from the malformations, fully developed. When examined by me on October 15th, he was much emaciated, and cried almost continuously, the chief exception being when he was feeding at the breast.

Both auricles were abnormal.

On the *right side* (Fig. 1) there was a fully-developed auricle of fully average size, with the various elevations and depressions well marked; but, in addition to this, there was a "blurred image" of this auricle in miniature, springing from the site of the tragus, and with its helix directed forwards. This supernumerary auricle seemed to consist of what might be described as a "reflection"—or, perhaps better, mainly of skin and fat, with a small portion of cartilage on its inner aspect. In this supernumerary auricle might be traced a helix and an anti-helix, with a shallow linear depression representing the fossa of the helix, a well-marked concha, and a lobule in common with the lobule of the normal auricle. Immediately in front, and in part undermining this supernumerary structure, was a deep rounded depression, just as if at some stage of development the tissue here had been scooped out to form the supernumerary auricle.

The external auditory meatus was narrow and slit-like, but the tympanic membrane appeared to be normal.

Left Side (Fig. 2).—Here the auricle was represented by a mass of soft tissue, irregular in shape and deficient in cartilage. The helix was represented by two curves, as if the posterior border of the helix at its most prominent part had been pushed forwards. The anti-helix was incorporated

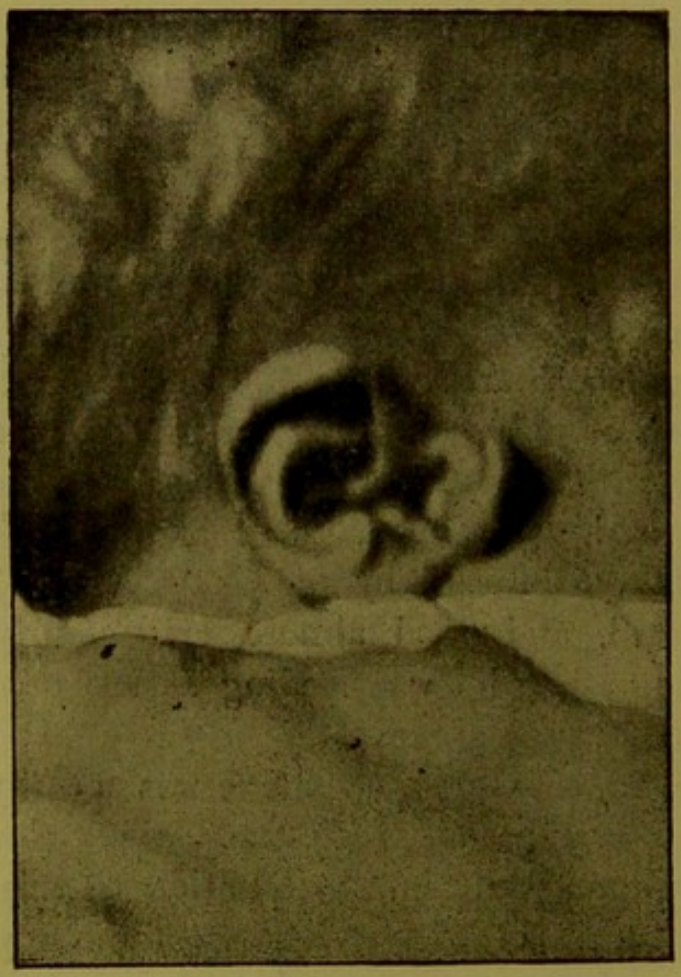

Fig. 1.

with the helix, there was no fossa between them, and, in place of a cup-shaped concha, this hollow was represented by a deep linear depression. At the upper part of that which represented the lobule there was a very tiny circular depression, confined to the skin apparently. There was no depression or sinus representing the opening of the external auditory meatus, and no depression in the bone could be felt near its usual site.

The conclusion arrived at was that, in an early stage of development, the first branchial cleft, which at an early stage

is converted into the external auditory meatus, the tympanum, and the Eustachian tube, had become completely closed, and that there was complete absence of those parts—the external ear, the middle ear, and the Eustachian tube—on the left side. There were no fistulæ representing remains of branchial canals or otherwise.

In addition to the aural abnormalities, the mouth was

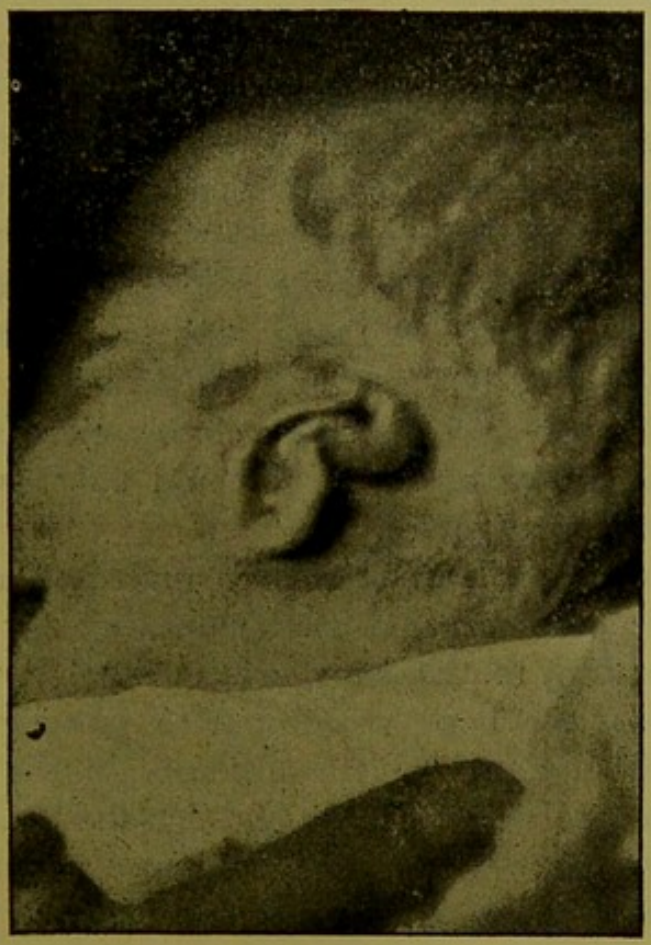

Fig. 2.

kept almost constantly widely open, suggesting a dislocation of the lower jaw, and the lower lip drooped, presenting a fish-like or V-shaped outline when separated from the upper lip. In the oral cavity the most prominent feature was the abnormal size of the subglottic tissue. This was so shaped and so prominent as to suggest the presence of a second tongue, and it seriously interfered with both respiration and deglutition. From an early period after birth there were frequent attacks as if the child was becoming asphyxiated, and it was observed that laying him over the arm, face

downwards, or pulling the tongue forward, enabled him to breathe freely again. This difficulty in breathing, however, robbed the child of sleep. He died of exhaustion when a few days over six weeks old.

Within six hours of death, Dr. Ferguson made a partial *post-mortem* examination. He was restricted to an inspection of the left ear, which, however, was the part of special interest. He reports as follows :—

"Dissecting off the skin which included the abortive auricle and reflecting it forwards, I found on the under surface no attempt at perforation such as would indicate the site of an external auditory meatus; the whole deep surface of this integument was continuously and equally smooth. Lying between the skin and the bone was an oval, flat, well-defined pad of soft tissue, pink in colour, somewhat like fat to the eye, but yielding to the finger more like coagulated blood; yet, when more closely examined, it was proved to be neither of these. I could only suppose it to be embryonic in character.

"This removed, the bony surface underneath was found quite smooth; there was neither an opening nor any rough spot corresponding to the bony canal. Further, passing the finger forwards, the bone was found to present a rounded end anteriorly, with complete absence of the zygomatic process. Into the depression caused by the absence of the zygoma the finger dipped easily, and in front the edge of the malar bone, to which the zygoma normally articulates, was felt quite free. In the depth of this depression, however, notwithstanding these marked defects in the temporal bone, the lower jaw was found articulating in the normal position."

Remarks. — Defects in the formation of the auditory meatus are usually combined with defects in the auricle, with partial or complete absence of the middle ear, and often with arrest of development of the bones of the head.

Sometimes, as here, all signs of the auditory canal are absent; occasionally there is a shallow depression or a short blind canal in its place; and Politzer reports a case which he examined, in which a fibrous cord took the place of the canal. He says that in unilateral absence of the auricle

combined with atresia of the meatus examination of the movements of the velum palati during phonation should never be omitted. If the movement of the palatine arches is symmetrical, the development of the Eustachian tube and the middle ear is probably normal. If, on the other hand, the half of the palate on the malformed side exhibits diminished movements, it may justly be assumed that the tympanic cavity and Eustachian tube are defective in so far as they are combined with a defective development of the muscular apparatus of the palate and tube.

Any attempt to verify by this method the opinion formed as to the absence of the tympanum and Eustachian tube was futile in a child so young as the one under consideration.

Note on Development.—The primary auditory vesicles (one on each side), which constitute the origin of the organ of hearing, appear each as an involuted follicle from the superficial epiblast of the head within the first few weeks of embryo life, and for a time each is open to the surface. In its beginning this otic vesicle has no connection with the nervous centres, though it forms the rudiment of the labyrinth of the ear. It is situated immediately above the upper or proximal end of the two first post-oral plates, and is, consequently, on a level with the first post-oral cleft.

The outer opening of the follicle very soon contracts and becomes entirely closed, and, sinking down towards the basis of the cranium, becomes embedded in the formative mesoblastic tissue and undergoes chondrification and ossification at a very early period. The position of the primary otic vesicle, which thus goes to form the osseous and membranous labyrinth, is in close proximity to the first branchial cleft, which latter is afterwards converted into the external meatus the middle ear or tympanum, and the Eustachian tube.

The auricle or pinna is of integumental origin, being gradually developed in the second post-oral bar—that is, on the posterior margin of the first visceral cleft.

A CASE OF ACQUIRED TOTAL DEAFNESS, THE RESULT OF INHERITED SYPHILIS; WITH POST-MORTEM.

By WALKER DOWNIE, M.B., F.F.P. & S.G.,

LECTURER ON DISEASES OF THROAT AND NOSE, GLASGOW UNIVERSITY; HON. AURIST, ROYAL HOSPITAL FOR SICK CHILDREN, ETC.

In October, 1891, a lad seventeen years of age, who was said to have become completely deaf during the previous six years, was brought to me at the Ear, Throat, and Nose Department of the Poliklinik.

His mother reported that he had been a weakly boy at birth and had continued delicate until five years of age. When seven years old his eyes became "inflamed," for which condition he was under treatment as an out-patient at the Glasgow Eye Infirmary for over two years. He was sent to school when eight, but before the session had closed the teacher recommended that he should be kept at home until his eyes were well and until his hair, which had begun to fall out, leaving bare white patches, had grown again. At eleven his scalp was free from patches, his hair was healthy and well grown, and his eyes were well and strong—that is, the redness and tenderness due to the inflammatory process had subsided, but his sight was bad, chiefly from the presence of corneal opacities.

While this improvement was taking place his hearing was becoming impaired; the left ear appeared to be first affected, but soon both were implicated, and at the end of six months he was totally deaf. The transition from the state of hardness of hearing to that of total deafness appeared to have taken place very suddenly. His mother described how she heard him sobbing bitterly one morning while lying in bed, and as she approached him he exclaimed, "It 's awa' a' together!" and from that time he was totally deaf.

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Family History :

Inquiries into the patient's family history elicited facts illustrative of the terrible ravages resulting from the introduction of syphilis by a spouse.

The mother of the patient was fifty years of age when she came to consult me regarding her son.

She was married when twenty-three, and the following particulars of the results of her fourteen pregnancies are interesting :

First—A healthy male child born at full time. This boy died when six years old from inflammation of the lungs. Shortly after the birth of this child, the mother appears to have contracted syphilis, the early constitutional symptoms of which seem, from her own account, to have been severe and well marked. While so affected she become pregnant a second time ; and within fifteen months of her first confinement was delivered of a still-born female child at the seventh month.

Third—A male child born near the end of the seventh month, and which lived ten hours.

Fourth—Male twins in the eighth month, one still-born, the other survived two days.

Fifth—A female child still-born in the seventh month.

Sixth—A female child at full time. She is still alive and apparently in good health.

Seventh—A miscarriage in the third month.

Eighth—A female child still-born in the seventh month.

Ninth—A miscarriage just over the third month.

Tenth—The patient, now seventeen years old.

Eleventh—A female child at full time, still alive and in fairly good health.

Twelfth—A miscarriage in the third month.

Thirteenth—A female child at full time who died from measles when two years old.

Fourteenth—A miscarriage in the fourth month—this occurring five years ago.

Patient's condition when first examined.—The lad, who was described as having formerly been bright and smart, was listless and stupid, and he was thin and anæmic. His forehead was somewhat prominent, his nose well formed, the cornea of both eyes was of ground-glass appearance generally, with several patches of distinct opacity, his incisor teeth were serrated, his

palate high and narrow with prominent transverse ridges, and his voice was monotonous.

When his hearing became first affected he complained of severe pain in both ears and of loud noises in the ears and head, but these had long since disappeared. He never had discharge from either ear, nor any history of middle-ear disturbance.

To the sound of the voice, the tuning-fork, and loud bell he was totally deaf. Occasionally in his eagerness he said he heard the tuning-fork when placed over his mastoid, but this occurred quite as often when the tuning-fork was at rest as when vibrating.

Both meatuses were normal in form and appearance, and both tympanic membranes were intact, slightly indrawn, and somewhat whiter than the normal.

Over the right parietal eminence there was a prominent swelling, smooth, hard, and painless, the base of which measured an inch and a half long by fully one inch broad. It was explained that two years previously he was struck by a piece of wood on the right side of the head. Considerable pain followed the receipt of the injury and a swelling resulted which had gone on increasing in size. Several similar, though smaller, swellings over other parts of the head had been observed in the interval, but these had disappeared, in some cases after days, and in others after weeks.

The use of gray-powder and the regular application of mercurial ointment were prescribed, though with but small hope of benefit accruing. At the end of six weeks he returned and his mother reported that he could hear the rumbling of carts passing along the street, and he himself, from this fact, felt hopeful of further improvement. No change, however, could be discerned on using the various tests, but as his general health had markedly improved, continuance of the treatment was advised.

While he remained under observation his general health steadily continued to improve, and he was able to do some work in connection with his father's bakery. Increase in the size of the swelling (gumma) over the right parietal region was noted from time to time. At the end of six months from date of first observation there was no doubt but that he could hear the sound of a vibrating tuning-fork by osseous conduction for about one fourth of the length of time it was audible to a normal ear.

I did not again see him until requested to meet the family attendant on the 12th of June, when I learned that shortly after

his last visit to the dispensary (March) the swelling on the head began to discharge watery fluid, and then to break down and give rise to a highly fetid discharge; and for this reason he had been kept at home.

On the morning of the 10th of June he had worked for some hours "baking and firing," and having finished he washed himself and sat in front of the kitchen fire to read. While so engaged the book fell from his hands several times, and on his mother's correcting him for his apparent carelessness she noticed that his hand twitched and he became convulsed on the left side and would have fallen from the chair had she not caught hold of him. When laid in bed it was found that he had paralysis of the left arm and left leg. On the following night he had a second seizure in which his convulsive movements were less marked and chiefly affected the face.

I found him much emaciated, conscious, and intelligent, though he spoke with difficulty. The area occupied by the swelling was now nearly as great as the right parietal bone itself, and the swelling was fungus-like, composed of broken-down inflammatory tissue, together with sloughing scalp and brain tissue. I recommended his removal to the hospital, which, however, was not accomplished till the 16th. On the morning of the 17th June he was put under chloroform, the scalp thoroughly cleansed, and the sloughing material cleared away. I then removed a considerable part ($1\frac{1}{2}'' \times 2''$) of the right parietal bone, wholly necrosed, and this permitted a large quantity of very fetid pus to escape. In the ward journal it is reported that "paralysis of the left arm and leg and left side of the face, present on admission, became more marked shortly after operation. There is also during renewal of dressings considerable protrusion of more or less disorganized brain-substance through the opening in the skull. He continued fully sensible and fairly well till the 22d, when swallowing and breathing became difficult and he died somewhat suddenly in the afternoon of that day. He had no convulsions after the operation."

A **post-mortem examination** was made next day and the following is an abstract of the report:

"The surface generally is pale and the body very spare. There is a gaping partly incised wound near the summit of the head to the right of the middle line, the anterior border of which is immediately above the external meatus.

This wound is occupied in its middle part by a soft hemorrhagic mass which projects through an opening in the skull. There is a considerable gap in the *duru mater* to which the brain-substance is partly adherent. A corresponding gap is in the brain-substance, which may be in general described as involving the ascending parietal convolution, except its lower extremity, and a considerable part of the parietal lobe. At the base there is considerable purulent infiltration of the soft membranes extending somewhat into the Sylvian fissure.

"Along the right border of the liver numerous deep cicatrices, but no actual gumma, were found. The thoracic and abdominal organs generally were found to be in a highly amyloid condition.

"The right temporal bone was removed and placed in an acid solution for section.

"On examining the bone externally, the small size of the mastoid process was the only feature in which it appeared to deviate from the normal.

"The bone was then bisected, cutting through the long axis of the petrous portion, thus exposing the external meatus throughout its length, the tympanum, the anterior portion of the cochlea, and the internal auditory meatus.

"The external auditory meatus was found normal in appearance; the tympanic membrane thin and translucent and free from adhesions. The malleus was healthy and normal in size and form. The incus, which was displaced during section of the bone, was healthy; but the base of the stapes was incorporated with, or ossified to, the border of the foramen ovale and so had become immovably fixed to the inner wall of the tympanum. The lining membrane of the tympanic cavity was intact and the Eustachian tube patent.

"The tympanum was of average size, the attic well developed, but there were no interstices in the bone posteriorily *i. e.*, the mastoid bone which as stated was unusually small was solid.

"The internal auditory meatus was next examined. At its inner extremity it was of average normal calibre, and both the auditory and facial nerves contained therein were healthy. But tracing it outward at a distance of 1 *cm* from

the inner opening, the upper wall became suddenly thickened, encroaching on the canal and at a farther distance of 3 *mm* the canal was almost completely obliterated.

"The vestibule was so greatly encroached upon as to make it doubtful whether any portion of this space remained. The cochlea was readily examined and was of average size, but the modiolus and lamina spiralis ossea were so thickened as to occupy an unusually large proportion of the cavity of the cochlea. Of the semicircular canals only a trace of the external (horizontal) one could be found, the remaining portion of this part of the labyrinth being lost in a mass of dense bone of ivory hardness.

"The chronic inflammatory process which had led to such extensive new-formation of bone-tissue was, I think, undoubtedly syphilitic in character. It resulted in solidification of the usually spongy mastoid, in the obliteration of the outer third of the internal auditory meatus, with consequent destruction of the vitality of the auditory nerve, and it not only thus cut off the internal auditory meatus from the internal ear, but it resulted in the obliteration of a large portion of the labyrinth itself, and all this up till the end with no other result than complete loss of hearing."

HÆMORRHAGE FOLLOWING TONSILLOTOMY.

BY

J. WALKER DOWNIE, M.B., F.F.P.S.G.,

LECTURER ON DISEASES OF THE EAR AND THROAT, WESTERN MEDICAL SCHOOL, GLASGOW; SURGEON, THROAT DEPARTMENT, ANDERSON'S COLLEGE DISPENSARY, ETC.

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