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VARIETIES OF NERVE-DEAFNESS.[1]

BY DR. DUNDAS GRANT, F.R.C.S.

IF we may judge by recent otological literature, the general tendency has been to confine attention exclusively to the surgical treatment of suppurative disease of the middle ear and petrous bone in relation to their dangerous sequelæ. Consideration seems to have been almost entirely withdrawn from diseases of the ear as an organ of hearing.

This is not altogether surprising when we remember how much can be done in the former direction, still less so when we realize how many of the latter class of diseases are not susceptible of successful treatment. The feeling has, however, gone too far, and the diseases in question have lacked the careful study they deserve. While admitting that the proportion of incurable cases is considerable, I am convinced that in many the effect of treatment has been marred by erroneous diagnosis. I have detected this as made by myself as well as by others. I do not refer here simply to obscure cases in which certainty of diagnosis is scarcely attainable, but fairly ordinary cases in which more detailed examination leaves little room for doubt. It is only right to remember that in investigations in which the tests are so largely subjective the patient's faulty observation and inaccurate evidence are often largely responsible for the erroneous decision.

[1] Opening paper of a discussion on "Varieties of Nerve-deafness," British Laryngological, Rhinological, and Otological Association, March 14, 1902.

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By nerve-deafness I mean dulness of hearing produced by disease of the auditory nerve, its nuclei, the cortical centres or the strands connecting them.

The auditory tract commences in the hairs of the auditory nerve-cells, and continues through the spiral ganglion, the cochlear nerve (in the internal auditory meatus and the cranial cavity), the accessory auditory nucleus, the trapezoid body, a tract parallel with the lateral fillet, the internal geniculate body, the subthalamic portion of the tegmentum cruris, the internal capsule, and the corona radiata, to the cortical centre in the superior temporo-sphenoidal convolution.

The pathological possibilities are obviously extremely numerous, and some extremely obscure. I shall only refer to a few of them.

I shall not take up your time by reviewing in detail the *tuning-fork tests*, as to the value of which we are fairly agreed; I shall, however, make the reservation that I assume that the evidences they afford are reasonably well marked, and that we interpret their results with a careful regard to their recognised limitations.

Diminished bone-conduction, negative Weber and positive Rinne, when met with in association, afford the typical, unquestionable evidence of uncomplicated nerve-deafness, if the dulness of hearing is at all marked.

When nerve-deafness accompanies middle-ear disease Rinne is negative, Weber may be positive, and we may have to depend on diminution of bone-conduction, or, I believe, absence of the increase of bone-conduction, which should accompany pure obstructive disease.

In simple unilateral nerve-deafness we may have a negative Rinne, because in the test the air-conduction of the affected ear is compared with the bone-conduction of the sound one. The result may be checked by means of Weber's test, which would be "negative," and is therefore, in unilateral cases, extremely valuable. I leave you to judge of the signs obtainable when there is obstructive deafness on one side and nerve-deafness on the other.

A "positive" Rinne does not, however, exclude a minor degree of obstructive deafness, unless the time during which the given fork is heard at the meatus after ceasing to be heard on the bone is the same as it is in the normal individual.

It is to be noted that if a very high-toned fork is employed it is impossible to prevent it being heard through the air by the opposite ear if the latter is normal (or even affected with obstructive disease), even though this is stopped by any means available, and air-conduction appears longer than it really is.

The same is true with regard to the high-pitched tones of

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Galton's whistle, and therefore, to make sure that it is actually the affected ear—and not the sound one—we are testing, we stop up the diseased ear as well, and note whether the sound ceases to be heard: then we open the ear again and note whether the sound is again audible. This is the check devised by Dennert.

Again, with a low-toned fork, which is necessarily massive, the patient is apt to mistake the mechanical vibration excited by the instrument for acoustic stimuli, and an apparent increase of bone-conduction is produced. A fork of about 250 vibrations per second should, therefore, be used, and preferably one which in the normal individual is heard for about fifteen seconds through the air after ceasing to be heard through the bone.

There is one test which deserves more credit than it generally receives—namely, Gellé's centripetal pressure. In carrying out this test a vibrating tuning-fork is placed on the vertex, while the tip of a Politzer-bag is in the auditory meatus. When the bag is compressed the sound should instantly decrease to a very marked extent if the stapes is mobile, because this bone is driven in so as to exercise pressure on the labyrinthine contents. Should this decrease not take place, the stapes is assumed to be ankylosed. In the former case Gellé's test is "positive," in the latter "negative." It is obvious that if the membrane is extremely relaxed or if it is largely destroyed the pressure cannot be so effectively exercised, and its results are, in such circumstances, of little value. The cases in which it is of special value are those in which there is, with a practically normal membrane, a high degree of deafness, and the diagnosis lies between ankylosis of the stapes and nerve-deafness.

Having diagnosed the existence of nerve-deafness as such, we have to decide on the *site and the nature of the lesion* on which it depends.

In regard to the site we have to depend on the information afforded by the state of the neighbouring nerves and other parts of the nervous system, and also by the determination of the part of the acoustic scale for which the hearing is most defective. The nature of the lesion is chiefly revealed by its mode of onset and course, and by the associated symptoms of disease of the system in general or other organs in particular.

Sudden onset is characteristic of hæmorrhage, embolism, or nervous shock.

Rapid onset, with pyrexial disturbance, indicates acute inflammation. Without inflammatory disturbance a rapid onset is very characteristic of syphilitic disease of the labyrinth.

A *gradually-developed nerve-deafness* results generally from

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degenerative, chronic inflammatory, or slow nutritive changes, or from the formation of a new growth.

In all cases in which nerve-deafness is diagnosed or suspected the hearing for the highest-pitched tones should be tested by means of Galton's whistle or Koenig's steel rods. The former is probably sufficient for clinical purposes.

It may be taken as a general rule that in a case of nerve-deafness as detected by means of the tuning-fork, if the loss of hearing is specially marked for the highest tones, the disturbance is most probably situated in the labyrinth; if not, it is probably in some other part of the central auditory tract, and in many cases is functional. The check referred to in connection with high-pitched tuning-forks is equally important in testing with Galton's whistle.

Should no obvious lesion of other nerves (we shall consider the vestibular nerve as being part of the auditory) *or the general nervous system be present*, we are guided by the results of testing the range of hearing, and if hearing is most impaired for the highest tones we may very safely assume that the lesion is in the *labyrinth*.

Should hearing not be mainly defective for the highest tones, then the lesion is situated most probably in some other part of the auditory tract, such as the cortical centre, and is often functional in nature.

The methodical examination of the *cranial nerves* from before backwards would be the proper step for the clinical student, but we may for practical purposes save time by taking them in the following order: facial, vago-accessory, hypoglossal, sixth, and continuing forwards to the first.

Facial paralysis alone, conjoined with nerve-deafness, is in suppurative cases usually due to tuberculosis of the petrous bone, while without suppuration it is more likely to arise from a lesion in the internal auditory meatus.

Involvement of the vago-accessory, as evidenced by paralysis of the corresponding half of the palate and of the vocal cord of the same side, indicates a lesion deeper than the internal meatus—say, a gummatous pachymeningitis on the under surface of the medulla. The same is true of the hypoglossal nerve.

Paralysis of the trunk of the sixth occurs in basic meningitis, involving it in its long exposed course on the under surface of the pons, characterized by paralysis of the external rectus of the corresponding eye. When with it the internal rectus of the opposite eye is affected, the lesion is situated in the nucleus.

Anæsthesia of one-half of the face alone would indicate a local lesion of the sensory nucleus of the fifth, but as found in associa-

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tion with nerve-deafness it is usually combined with hemianæsthesia of the side of the body, for special as well as general sensation.

Regarding involvement of the central nervous system in general the following points: Cross-paralysis, hemiplegia, hemianæsthesia, "word-deafness," ataxy, optic neuritis, ophthalmoplegia.

Cross-paralysis indicates a lesion in the pons below the decussation of the facial tracts. Hemiplegia points to the opposite hemisphere, probably the internal capsule; hemianæsthesia may arise from disease of the sensory tract in the posterior part of the internal capsule, or else from an affection of the cortical or subcortical sensory areas, generally hysterical. In the former case there is loss of the corresponding halves of the fields of vision of *both* eyes; in the latter there may be loss of acuteness in the whole of the field of vision of the opposite eye (crossed amblyopia), coupled sometimes with colour-blindness in that eye (achromatopsia or dyschromatopsia). This is well shown in the case reported below.

I have already referred to the partial ophthalmoplegia arising from disease of the sixth nucleus—namely, paralysis of the external rectus of the same and internal rectus of the opposite eye. Complete ophthalmoplegia would point to disease in the nuclei and longitudinal bundle in the tegmentum below the level of the aqueduct of Sylvius—apt to be encroached upon by tumours of the quadrigeminal bodies.

Testing the vision and determining the fields of vision is of importance in connection with hemianæsthesia.

The ophthalmoscope should be in frequent requisition, though it is only very rarely that in our practice it reveals positive pathological changes. The existence of optic neuritis or choked disc should obviously not be overlooked. The following case was only elucidated by my finding very typical albuminuric retinitis, my observation being confirmed by the expert to whom I referred the case:

Mr. P——, aged fifty-three, was referred to me by Dr. Lauchlan on account of deafness and singing noise in the right ear; this was of three weeks' duration; he had suffered from vertigo and vomiting; the tuning-fork evidences were those of nerve-deafness. Galton was not heard above the mark 5·5, as checked by Dennert's method; the ophthalmoscope revealed well-marked albuminuric retinitis, and there was a scarcely detectable trace of albumin in the urine.

The patellar tendon reflex or knee-jerk gives us often important information, its loss suggesting locomotor ataxy or post-diphtherial paralysis, its exaggeration, apart from other serious evidences,

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pointing to heightened nervous excitability, as in neurasthenic conditions.

Disturbance of equilibrium, taking the form of definite vertigo, is in by far the largest number of cases due to aural lesion, and when this is associated with nerve-deafness the loss of hearing for the highest tones usually locates the lesion as in the labyrinth. Rarely, however, disease of the cerebellum or the peduncles may produce definite rotation. In cerebellar disease there is usually staggering like a drunken man if the middle lobe is affected; paresis of the upper extremity of the same side if a lateral lobe, even if the characteristic vomiting and optic neuritis are not yet *en evidence*.

The *nature of the lesion* is, as I have already stated, mainly indicated by the mode of onset and the course. I shall refer to a few types.

Suddenness of onset generally indicates a hæmorrhagic effusion into the labyrinth, rarely an embolism, not very unfrequently a sudden nervous shock. Hæmorrhage into the labyrinth is the lesion producing the most severe and typical examples of Menière's disease; it generally involves the semicircular canals as well as the cochlea, and is therefore characterized by violent disturbance of equilibrium and vomiting; the hearing tests reveal absolute deafness or else dulness of hearing, most marked for the highest-pitched tones. Many cases of Menière's disease are subsequently reported as severe deafness of sudden origin dating back from what had been looked upon as a severe "bilious attack." In most instances attacks of Menière's disease have been traceable to exposure to cold after being in a hot place, and occasionally to suppression or sudden stoppage of the menses; it is more liable to take place when there is increased arterial tension with weakened vessels or in the various blood diseases which favour the occurrence of hæmorrhage in various parts, such as leucæmia. In sudden functional deafness due to a nervous shock vertigo is not usually present—at all events, to any great degree—and the dulness of hearing is not markedly greater for the highest-pitched tones than for others.

Those forms having an *acute and rapid onset* are generally inflammatory, and occur frequently in the course of infectious diseases, particularly mumps, the rapidly-developing and incurable deafness occasionally occurring in the course of this disease being an acute labyrinthitis. The following case (brought by Dr. Abercrombie) illustrates the condition:

B. C——, aged eighteen, absolutely deaf in both ears, suffered

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from mumps ten years ago. Towards the end of the first week of the disease vomiting set in; she became completely deaf in one ear; then, in the course of two days, equally deaf in the other one, the vomiting lasting for three days.

Apart from obvious feverish disturbance, such a rapid onset rarely occurs except as the result of syphilis, either hereditary or acquired. In the hereditary form it usually develops about puberty, and is, as a rule, preceded by interstitial keratitis, and other physiological evidences, such as the notched and pegged "Hutchinson" teeth, are usually present. In a case (also brought by Dr. Abercrombie) deafness developed unusually early, namely, at seven years of age; the keratitis was extremely slight.

Of the forms of nerve-deafness which are *gradual in development* the following are important types:

The *anæmic* and the *congestive* are to be carefully distinguished, and the following points will enable us to do so in reasonably well-marked cases: In *congestion* there are usually collateral evidences of plethora; the patient's symptoms are worse on lying down or after the ingestion of food, or before the occurrence of the menses; they are made worse by the inhalation of nitrite of amyl, and are relieved by compression of the vertebral arteries. In *anæmia*, on the other hand, all these signs are reversed, and the constitutional signs of this condition are present, though sometimes to a very slight degree. In some cases the findings are those of *anæmia at one time and congestion at another;* I attribute this to irregular nerve control of the vascular supply, due as a rule to neurasthenic conditions. The treatment required follows as the obvious sequence of the diagnosis.

The most typical forms of anæmia occur in widows in the poorer class, who have to earn a meagre subsistence in return for laborious work, and who frequently present themselves for hospital treatment on account of their aural trouble.

Among other types are: the *senile* change which normally begins to show itself after the age of sixty, chiefly by loss of hearing for the highest-pitched tones; *occupation* deafness, the form occurring typically in artillerymen and boiler-makers; it also attacks the left ear in those who do much rifle-shooting, and results from exposure to noises. *Toxic* nerve-deafness is particularly exemplified in the dulness of hearing produced by quinine or salicylic compounds, and it is probably congestive in nature. The *neo-marital* is an occasional form of nerve-deafness comparable to the amblyopia of young husbands described by Jonathan Hutchison. *Neurasthenia* often gives rise to a form of nerve-deafness in which

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the deficiency is not especially marked for the highest-pitched tones; it is sometimes induced by over-work, excessive worry, the strain of active military service, and other analogous circumstances.

Hysterical nerve-deafness is probably sudden in onset, but when affecting only one ear the patient may at first be unconscious of its occurrence, and on its discovery may be unable to state either the mode or date of onset. In the following case of hysterical hemi-anæsthesia with crossed amblyopia this point was exemplified:

Miss H——, aged forty-one, schoolmistress, has frequently suffered from complete loss of voice and hoarseness since December, 1890; the voice had been restored from time to time by cauterization of her throat at the hands of her medical attendant. She has suffered from purulent rhinitis affecting the various sinuses, and accompanied by frontal, post-ocular, and occipital headache; both frontal sinuses were explored by me with comparatively negative results, there being no obvious retention of pus and no marked benefit from the operations. In 1895 I discovered that she had hemianæsthesia of the right side of the face and body, including the olfactory, auditory, and gustatory nerves, with diminished visual acuity on the left side. The tuning-fork tests showed the loss of hearing on the right side to be of the nature of nerve-deafness, and the loss of hearing was as pronounced for the lowest tones as for the highest. Hearing-power is now greatly diminished, but Galton's whistle can be heard up to the mark 3, practically the same as on the other side. The dimness of vision in the left eye is accompanied by complete inability to distinguish green and red from each other. The case is therefore one of hemianæsthesia with crossed amblyopia, and therefore presumably hysterical in nature. No remedies (hypnotism included) seem to have touched the condition, and she cannot be induced to undergo the "rest" treatment which is indicated in such cases. Possibly some accident or the occurrence of the menopause may break the spell.

I shall now narrate the case of a patient whom I brought before this association some years ago, as an extreme case of hysterical nerve-deafness with spontaneous recovery:

Miss A——, aged eighteen, came under my care on May 27, 1895, complaining of deafness of both ears, stuffiness in the nose, and pain in the throat. The deafness to a slight extent was of three years' duration, and it had come on gradually, but had suddenly got very much worse immediately after the extraction of eight teeth, three months before coming to me. For this operation she was

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anæsthetized with gas and ether. On examination, the hearing was practically the same in both ears. She could only hear very loud conversation, and apparently only when her hearing-power was supplemented by lip-reading. The watch was heard at 6 inches, Galton's whistle was heard up to the mark 3·8, the bone-conduction of both mastoids was diminished, and Rinne's test gave a "positive" result in both ears. There was pain over the mastoids, no discharge was present, and there was no definite history of any previous discharge. At that time she described certain indefinite attacks of giddiness, of which she has now lost all recollection. On testing her hearing for various tuning-forks by air-conduction, she was found to have completely lost the hearing for "C_2" and for "C_1," while for the other forks extending from "C" up to "C_5" the amount of hearing-power varied from 3 or 4 up to 15 per cent. A diagnosis was then made of nerve-deafness of indeterminate origin, but probably "auto-suggestive." Ammoniated tincture of valerian was ordered, blisters were applied to the mastoid process, and galvanism by means of the continuous current to the strength of 10 milliamperes, with the negative rheophores, applied to the tragi, was employed for ten minutes at a time. The treatment was varied in the usual way, and the changes were freely run on strychnia, bromide of potassium, and ultimately the liquid extract of ergot. No improvement of any moment took place, and as a natural result the patient withdrew from further treatment. She later on came back to say that her hearing was perfectly good, it having returned in the January of 1897, after a "complication" of ailments, which confined her to bed for a fortnight. While lying in bed her hearing rapidly improved, until in six months it became perfectly normal, and she can now hear a whisper by the right ear at the distance of about 14 feet, and by the left 13 feet.

The spontaneous recovery seems to confirm the original diagnosis. In this case the tuning-fork test for middle tones "C_1" answered to the type of nerve-deafness, and they were sufficient to exclude middle-ear disease. The tests for air-conduction throughout the whole range of audition indicated that the maximum of loss was for deep tones. In typical cases of disease of the labyrinth the opposite would be the case, and we should expect to find the loss greatest in the uppermost part of the range, the lower part being relatively less defective. The combination, then, agrees with that described by Gradenigo as typical. on the other hand, of disease of the nervous centres, and, in fact, exactly of what he describes as occurring in hysterical nerve-deafness as set forth in his article in Schwartze's notebook. In his

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more recent publication on auditory disturbances occurring in hysteria, he describes the loss of hearing as being fairly uniform throughout the whole range, but more marked in the lower range, on account of the physical nature of tuning-fork vibrations rather than from peculiarities in the auditory nervous apparatus.

The *treatment* of nerve-deafness depends primarily on the diagnosis of the variety, and the necessity of distinguishing between the anæmic and congestive forms must again be emphasized. That valuable drug, pilocarpine, has fallen into undeserved disuse through its unfortunate effects in cases for which it was not adapted. I have no hesitation in ranking it as a most beneficial remedy in congestive conditions of the labyrinth, but most deleterious in the anæmic ones. It may be administered hypodermically or by the mouth. The initial dose by the skin is about one-twelfth of a grain. At least double this dose is necessary for internal administration. The precautions to be observed during its administration are, of course, well known by all present. The other general remedies calculated to deplete are also advisable in cases of congestion. In anæmia and conditions of exhaustion our sheet-anchor is strychnine, and when such doses have been reached as to produce twitchings or other nervous disturbances the remedy may be combined with chloral. Iron and chloride of ammonium are also of value, and in neurasthenic conditions arsenic may be added. I have a strong conviction, on experimental evidence, that quinine produces congestion of the labyrinth, and is to be accepted as the physiological corrective of anæmia of that part. In a number of cases of what I took to be well-marked anæmic nerve-deafness, treated with quinine under my own care and that of Dr. Wingrave, the benefit effected seemed to confirm this opinion most strongly.

I have perhaps not given to the electrical treatment of nerve-deafness the attention that it deserves. I have, however, seen improvement in several cases of "occupation" deafness which Dr. Lewis Jones has treated for me by means of the continuous current. In other instances the results have not been so satisfactory, though, perhaps, with a more judicious selection of cases, they might be better. In neurasthenic nerve-deafness general electrization may find a place, but supportive measures, such as the administration of cod-liver oil, malt extract, and glycerophosphates, have proved of great value. Among other useful remedies are arsenic, strychnine, and valerianate of zinc. The last remedy has been very strongly recommended to me by Dr. Bowles, who combines with it small doses of rhubarb or belladonna.

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Syphilitic nerve-deafness, occurring in the course of the secondary stage of the disease, is fairly amenable to treatment, but the results in the tertiary form are eminently desponding. Competent observers have noted very rarely an improvement as the result of injections of pilocarpine. In my experience improvement has been only temporary. I have, however, seen benefit from the administration of iodide of potassium, in doses, however, of not less than 1 drachm every four hours, continued for several weeks. On the whole, we have to trust mainly to keeping up the patient's strength by means of cod-liver oil and other supportive agents.

The above remarks are very far from exhausting the subject; but I venture to think that the most salient features have been included, and that I have dwelt mainly upon those points which are of the greatest practical importance. I trust others will amplify my description in regard to those forms of nerve-deafness with which they have had the most experience, and that they will also fill up the gaps which have been left by me.

NOTES.

The Annual Dinner of the British Laryngological, Rhinological, and Otological Association was held on March 14, the President, Dr. John Macintyre, being in the chair. The dinner was largely attended by the Fellows of the Association and their guests from distant parts of the United Kingdom. After the toast of "The King," that of "The Association" was proposed by Dr. C. O. Hawthorne, and was responded to by Mr. Lennox Browne, who spoke of the work done by the Association since its foundation fifteen years ago, under the Presidency of the late Sir Morell Mackenzie. He expressed the indebtedness of the Association to those who had to travel some hundreds of miles to take part in the work of the meetings, and mentioned that it was to Dublin—when the British Medical Association met there in 1887—that the proposition for the founding of an association specially devoted to the advancement of laryngology and otology owed its origin. The toast of "The Guests" was proposed by the President, and responded to by Sir William Dunn. Mr. Mayo Collier proposed "The Chairman," to which the President replied. During the evening some excellent music was provided by Mr. Wilhelm Ganz, with the assistance of Señor Guetary and Mr. Mervyn Dene, and Mr. Ronald Daniel gave some humorous songs.

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