

After-Condition of Patients.

The two men exhibited to-day illustrate very well the method of speaking, one by lip-movement over the small amount of air taken into the mouth—this sound proves sufficient to conduct conversation in the house, and even out of doors—the other by lip-movement over a current of air carried to the lips by a tube from the trachea; this sound is found to be sufficient to enable the man to make himself understood to a number of people.

One of these patients, Mr. B——, holds an official position as a tax collector and attends meetings of the committee. The other, Mr. T——, looks after home matters, shopping, post and so on. He might well have carried on work as a bookkeeper, having been a bank manager. A fuller account of these and others will be found in the *Lancet* for March 20, 1920.

THE ORIGIN OF SPORADIC CONGENITAL DEAFNESS.

BY JAMES KERR LOVE, M.D.

TWICE during the past ten years the writer has tried to show that hereditary deafness is Mendelian in incidence. On neither occasion did he definitely include as Mendelian, cases of "sporadic congenital deafness," although—if we exclude cases of deafness due to congenital syphilis—such sporadic cases are clinically identical with true hereditary deafness. The present paper is meant to show that such sporadic cases are not only clinically but genetically identical with the hereditary cases: that sporadic congenital deafness is hereditary and that such heredity is Mendelian. I am going to assume that the reader is familiar with Mendelian phenomena as displayed by the crossing of tall and short peas and the subsequent self-fertilisation of the resulting hybrids.

Hereditary Deafness.—It has long been recognised that deafness runs in certain families.

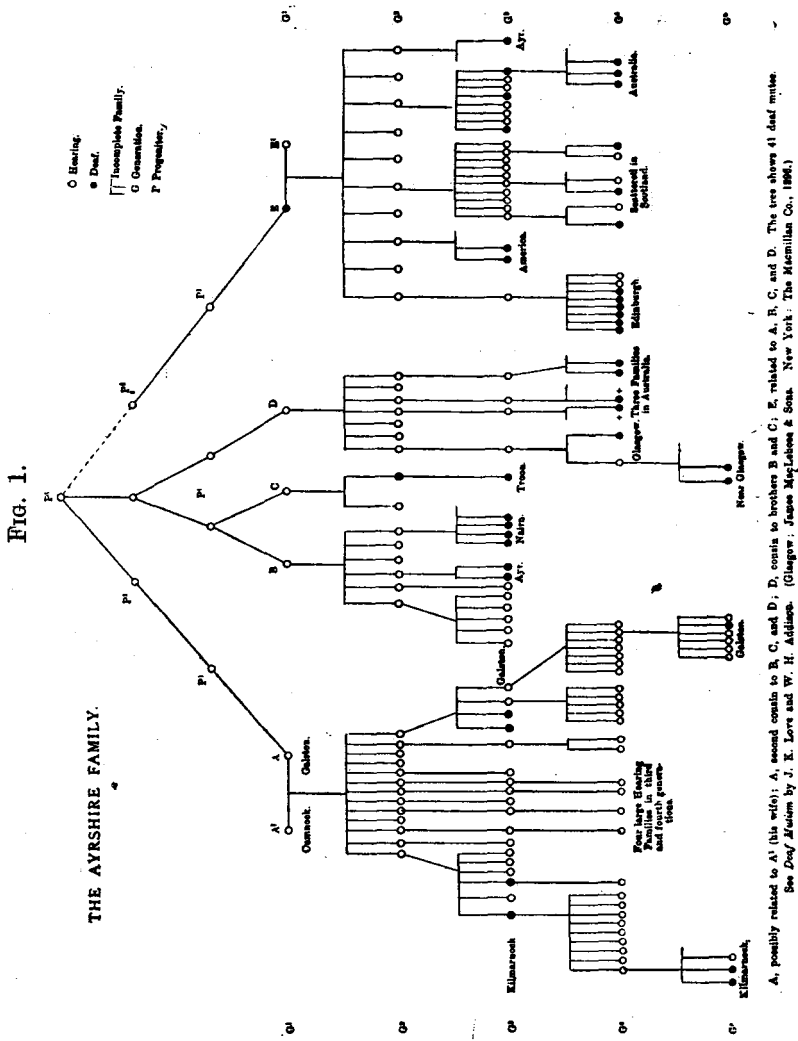
Dr. Graham Bell, of telephone fame—once a teacher of the deaf—Dr. Fay, of Washington, a teacher of the deaf, and many others have urged the heredity of many cases of congenital deafness. The present writer, nearly a quarter of a century ago, wrote similarly after an exhaustive examination of many hundreds of deaf children and an extensive review of the literature of the subject, and yet many people doubt the heredity of congenital deafness, and the deaf themselves either do not believe in the heredity of deafness, or act as if they do not believe in it. Why is this?

1. Because as a rule congenitally deaf parents have hearing children, and—

2. Because hearing parents often have deaf children.

It was all very puzzling, and however convinced one is in his own mind can he blame the deaf?

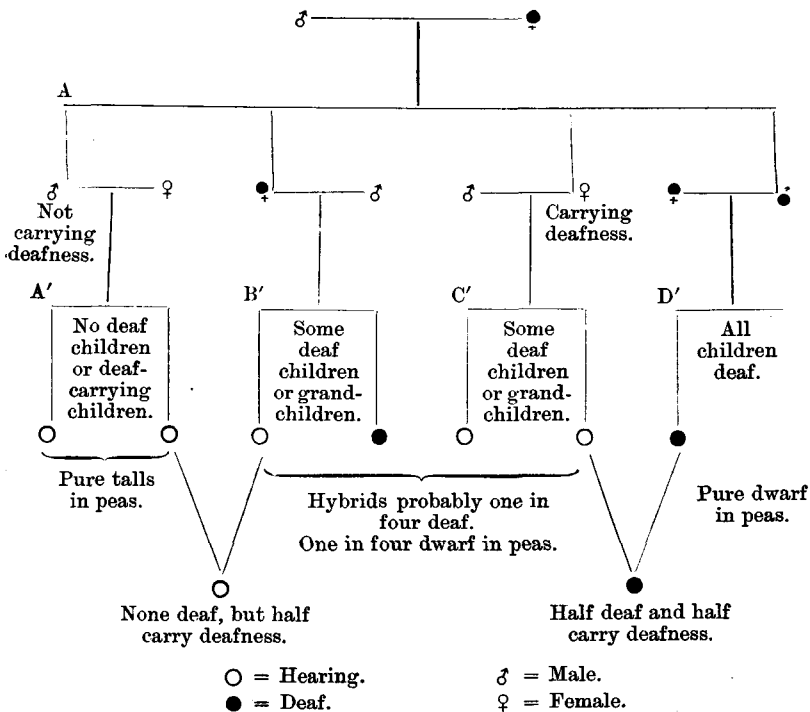
Take the Ayrshire family, a copy of whose family tree is given here (Fig. 1). Amongst over forty deaf-born children, in only two cases can deaf parents be shown. In every other instance the deaf children come from hearing parents. There is no doubt about the fact that deafness belongs to the family. Why and how this unexpected distribution? We do not know why. But we begin to know how. That is the



position with most sciences. From astronomy, which is an old and fairly exact science, to psychology, which is a new and a rather nebulous one, we have no answer to why, but a good deal of reply to how. So in the science of heredity, an important part of the still newer science of eugenics, we begin to answer how, but cannot tell why.

Let us glance at the accompanying tree, which is imaginary, but every fact of which is present in the Ayrshire tree or on its supplement (see "Mendelian Tree of Hereditary Deafness," Fig. 2). A hearing husband marries a hereditarily deaf wife and two deaf children result. Two hearing

FIG. 2.—MENDELIAN TREE OF HEREDITARY DEAFNESS.



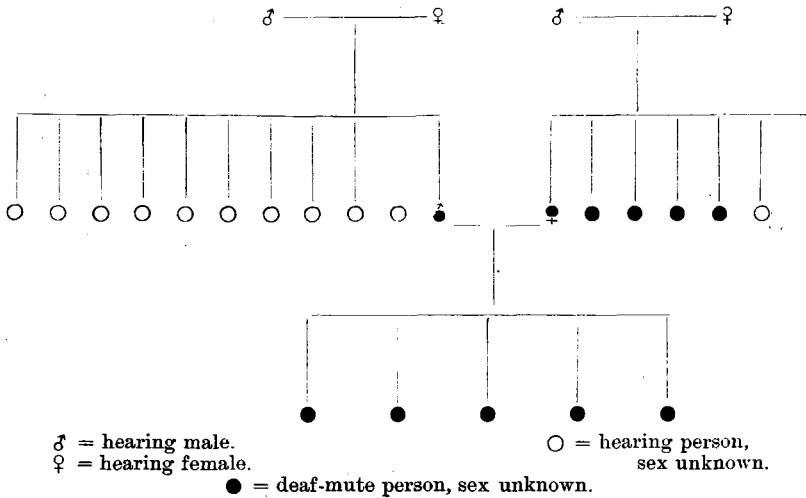
members complete the family. (It might quite well have happened that all four were hearing and that the deafness appeared in grand-children.)

The oldest boy (A) hears and does not carry deafness (like the pure tall pea). Deafness never appears in this family so long as no member marries into a deaf family. The second child (B), a deaf girl, and the third, a hearing girl carrying deafness, marry hearing partners, and all have some deaf children or grandchildren. They are hybrid, and were the families large enough about one in four would be deaf. In such small families this ratio cannot be expected. In any individual family all may hear or all may be deaf, but on the whole the ratio is observed, and accounts for the fact that hereditary deafness forms a pretty

continuous stream from one generation to another without much tendency either to increase or diminish. The average families of deaf fraternities are five or six and all do not reach adult life, and of these who do all do not marry. All this I see can be paralleled in the Ayrshire tree. Look now at the fourth child D. A hereditarily deaf man marries a hereditarily deaf woman as in the C—g family of Edinburgh and all the children are deaf (Fig. 3).

We are not done with the parallelism of the pea and the child. A pure tall A' may be crossed with hybrid tall B' and then all are tall though half are hybrid, and as long as hybrids meet any pure tall only tall will result. But whenever a hybrid meets another hybrid or

FIG. 3.—C—G FAMILY (Edinburgh).



For the above case I am indebted to Mr. Ilingworth, of Edinburgh. No doubt all the children are deaf. But even if they be pure recessives, which is quite uncertain, they must each marry pure recessives if no hearing children are to follow.

a dwarf then the dwarfs appear. So it probably is with deafness. So long as hearing people carrying deafness meet pure hearing people, no deafness results. But if by any chance—and the chance is bound to come—two hybrids, hearing people carrying deafness, get together, deaf children must follow. This would happen were B' and C' to unite. Lastly C' and D' may unite; in this case half the peas will be dwarf and the other half tall but carrying deafness. In the case of children, half would be deaf and the other hearing but carrying deafness (hybrid). The importance of these two classes of union will appear when we come to study sporadic congenital deafness.

At the risk of a charge of reiteration let me now tabulate these results.

There are six possible combinations in the pea family. T. means tall, D. dwarf.

Table of Unions between Tall and Dark Peas.

| No. | Nature of union. | | Result. | | |
|-----|--|-----------------------|----------------|---------------|---------|
| | The double letter shows the nature of the parents responsible for the individuals here united. | | Pure talls. | Hybrid talls. | Dwarfs. |
| 1. | T. T. × T. T. | Pure talls | 100 | — | — |
| 2. | D. D. × D. D. | Pure dwarf | — | — | 100 |
| 3. | T. T. × D. D. | | — | 100 | — |
| 4. | T. D. × T. D. | Hybrid tall | 25 | 50 | 25 |
| 5. | T. D. × T. T. | | 50 | 50 | — |
| 6. | T. D. × D. D. | | — | 50 | 50 |

The above table is altered from the late Mr. A. D. Darbishire's book, "Breeding and the Mendelian Discovery."

Corresponding Unions between Deaf and Hearing.

Look now at the possible union of hearing and hereditarily deaf people.

H. means hearing, D. means carrying deafness of the latter; some are deaf, some hearing. Of the former all hear and none carry deafness.

| No. | Nature of union. | | Result. | | |
|-----|--|--------------------------|------------------|--------------------------------------|-------|
| | The double letter shows the nature of the parents responsible for the individuals here united. | | Pure hearing. | Hybrid hearing carrying deafness. | Deaf. |
| 1. | H. H. × H. H. | Pure hearing | 100 | — | — |
| 2. | D. D. × D. D. | Pure deaf | — | — | 100 |
| 3. | H. H. × D. D. | | — | 100 | — |
| 4. | H. D. × H. D. | Hybrid hearing | 25 | 50 | 25 |
| 5. | H. D. × H. H. | | 50 | 50 | — |
| 6. | H. D. × D. D. | | — | 50 | 50 |

All these unions actually occur in the human family with the kind of result above shown.

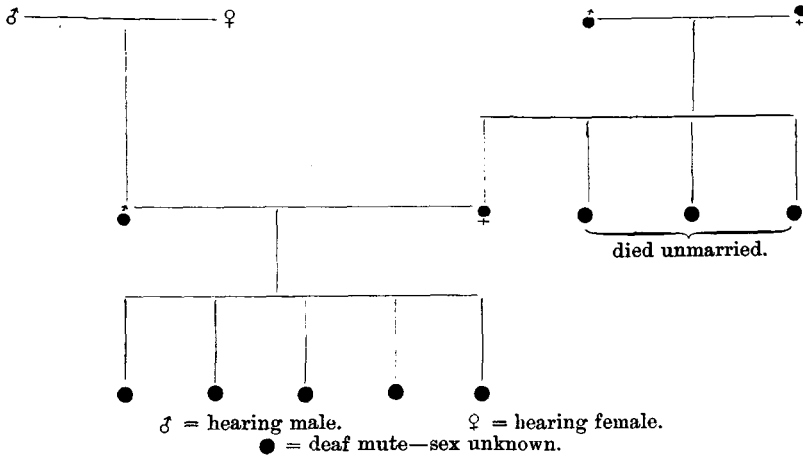
Comment on the first table is unnecessary. It is a statement of fact which comes true every time. Comment on the second table is necessary. Let us take the classes one by one.

Class 1.—A hearing man, both of whose parents heard and did not carry hereditary deafness, marries a similar woman. No deaf children can result. This is the case in far more than ninety in every hundred marriages. There is only one deaf to every two thousand of the population in England, and the half of these are born hearing and have been made deaf by disease after birth—a type of deafness with which we have nothing to do here.

Class 2.—A deaf man, both of whose parents are hereditarily deaf, marries a similar woman and all their children are deaf. This kind of union is luckily very difficult to bring about, as we shall see presently. But it has been done (Fig. 3).

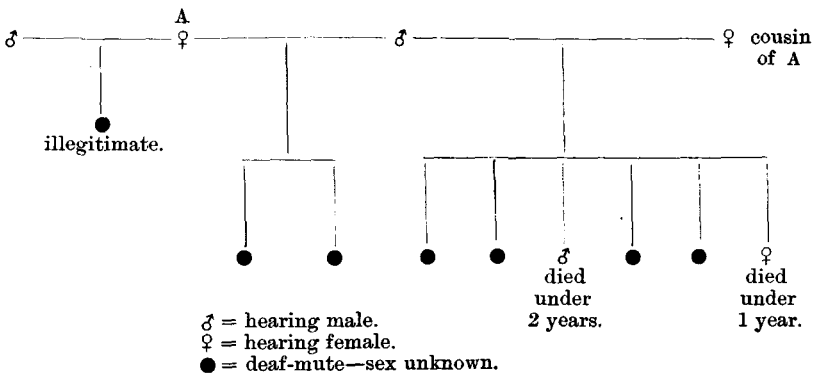
The following is an example (Fig. 4). It occurs in the records of the Doncaster Institution for the Deaf and Dumb, and was communicated to me by the late Mr. Howard, the Headmaster. It was published by me in 1896 before Mendelism had been heard of. See also Fig. 5.

FIG. 4.—G—Y FAMILY (Halifax District).



Here again deafness may be looked on as recessive. G—y, senior, or his wife carrying it, transmit it to G—y, junior, in whom it is expressed. The latter meets the same type of deafness in his wife, who comes of an entirely deaf stock, and all their children are deaf.

FIG. 5.—SUPPLEMENT TO AYRSHIRE TREE.



A and her cousin belong to the Ayrshire family. The father in this generation was hearing like the mothers, but, I suspect, carried deafness. This man makes the interesting experiment of marrying two women who are cousins, and who both carry deafness. One of these women has already an illegitimate child. All the children—except two who died so young that the condition of their hearing must have been doubtful—are deaf. In this family it is calculated that there are now about 100 deaf-mutes. There are several being educated in the Glasgow Institution now.

Class 3.—A pure hearing person marries a pure deaf-mute and all the children hear, and the deaf turn round and say, "Well, you see here is a deaf mother who has no deaf children." Yes, but look at the peas. In the first hybrid generation there were no dwarfs. Why should there be deaf children here? In Mendelian terms hearing is dominant to deafness. We need not shelter ourselves behind terms. Look at the Ayrshire tree. So far as my records go that was how the tree started in one branch at least. A hearing man married a deaf woman and all the children were hearing. But several of the grandchildren were born deaf.

Class 4.—A hearing man carrying deafness marries a hearing wife carrying deafness, and both deaf and hearing children follow. Again the opponents of the heredity of deafness object, "Why do deaf children nearly always come from hearing parents?" Because the parents were hybrids like the first hybrid generation of tall peas. Deaf children *must* follow if the family be large.

Class 5.—A hearing man carrying deafness marries a pure hearing woman and no deaf children will result. But half the children will carry deafness, and if any of these wander into classes 2, 3, 4 or 6 deaf children will result, and they are sure to wander there unless guided by the kind of knowledge Mendelism gives us. Even with that knowledge they will sometimes wander, for, as we shall see, we cannot experiment with and label children as we can peas. Did these hybrids always marry pure hearing partners no deaf children would ever follow. But I think this kind of marriage is common, and when the hybrid hearing marry the hybrid hearing many of the puzzling cases of sporadic congenital deafness may be accounted for. This kind of marriage is common because hearing hybrids are by the deaf themselves so often drawn together.

Class 6.—A hearing man carrying deafness marries a pure deaf woman. Half the children are deaf, and all the children carry deafness. This is a common type of marriage amongst the deaf. The deaf and their hearing relatives are necessarily thrown much into common society, and unions producing deaf children are the result. It will take much study of the deaf and much education of them to solve this problem. But, as we shall see, the solution is not impossible.

Sporadic congenital deafness, according to the view developed above, is due to the meeting of two heterozygotes—hearing hybrids carrying deafness. After eliminating congenital cases arising from syphilis—and these are really cases of acquired deafness—we get a simple classification of deafness as indicated in the italics which follow.

True Hereditary Deafness and Acquired Deafness due to Disease.—

The question of how to eliminate hereditary deafness would require more space than I dare expect to get within the limits of a single paper, but it is clear that any measures to be effective must be applied not only to the deaf but to the hearing hybrids or heterozygotes.

It would be interesting to know the conditions of the semicircular canals in cases of *undoubted* hereditary deafness. Looking to the comparative anatomy of the ear, it is unlikely that disturbance of the factor for hearing would involve disturbance of the balancing apparatus. There is no hearing organ in the fishes. With a view to settling the question raised here, I asked Dr. Gavin Young to test various classes of deaf-mutes—and particularly cases of *true hereditary deafness belonging to the Ayrshire family*—by rotation and by hot- and cold-water syringing.

I do not wish to anticipate his results, which will be published later, but I may note here that these are in the direction of expectation that in true hereditary deafness the balancing apparatus is intact.

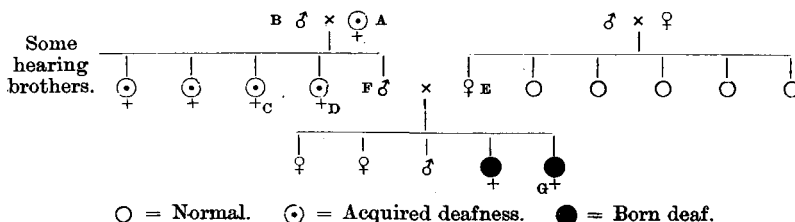
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CAN ACQUIRED DEAFNESS LEAD TO CONGENITAL DEAFNESS?

BY MACLEOD YEARSLEY, F.R.C.S.

THE following family history was obtained about eight years ago. It is interesting as an instance (the only one I am at present aware of) in which a family with a history of acquired deafness (*i. e.* deafness appearing after birth) produced offspring that were born deaf. It is important to note that the deafness was probably otosclerosis and, therefore, of hereditary character.



A, the grandmother, I did not see, but I was told by her daughter in-law (E) that she became deaf “when a young woman” and had two sisters “deaf like her.” A married B, a man without any family history of deafness. They had several children. The youngest (F) was the husband of E. He had “some hearing brothers” and four sisters who became deaf between the ages of twenty and thirty. I saw two of them (c and d), and they were cases of otosclerosis. The children of F and E were five, two girls and one boy hearing and two girls born deaf. I saw the younger (a). She was aged seventeen, and had been educated on the oral system. She had the typical “deaf” voice. Adenoids had