CONGENITAL DEAFNESS*

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WHEN a child is born with a malformation, a cleft palate, an anencephaly or other awful phenomenon, every one is puzzled at the events, that must have taken place, secretly and hidden, during development.

Congenital deafness belongs also to these mysterious events, but deafness has not such a spectacular character as the other congenital anomalies though it is essentially of the same order. This deafness is not observed immediately after birth, but only suspected afterwards, especially in a first-born, when the child has already reached an age, at which its companions begin to react to sounds, and are already chattering some unintelligible words. At the examination of the child several questions arise such as: What is the meaning of congenital deafness? What is the cause of this deafness? Is the child really deaf? Is it possible to prevent this deafness? In many cases it will not be possible to answer several of these questions and we must agree, that the whole problem of congenital deafness is rather confusing.

What is the meaning of the expression congenital deafness? The expression itself says only that deafness is present at birth but it does not explain in any way the etiology and nature of the affection by which the symptom deafness has appeared and the word congenital shrouds it in a haze of mystery that covers our ignorance about its origin.

Formerly congenital was used as equivalent to hereditary. Indeed, many times the congenitally deaf child will have an inherited type of deafness, but congenital must not be confused with heredity. In this respect three possibilities must be taken into account:

1. Deafness may be congenital but not hereditary as in deafness caused by rubella during pregnancy.
2. Deafness may be hereditary, but not congenital as in otosclerosis.
3. Deafness may be congenital and hereditary at the same time as in sporadic recessive deafness.

The meaning of the word deafness is also not well-defined. In the real sense deafness means total loss of hearing, but Bezold has already shown that many deaf-mutes may have quite important remnants of hearing. Where must the limit be fixed between deafness and hearing? Congenital deafness may be defined as a more or less pronounced loss of hearing, present at birth, leading to mutism and transmitted by heredity or

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acquired by chemical, physical or mechanical influences in the prenatal period. So it is apparent that congenital deafness is a collective noun for pathological deviations from different origin and different degree of the organ of hearing and that for their understanding, as for diseases in the adult, the genetics, the anatomy, the physiology, the pathology, the pathologic anatomy of the individual during development must be known. We are still far away from this general knowledge of fœtal life.

The Causes of Congenital Deafness

Nowadays it is clearly recognized, that congenital deafness is not only defined by hereditary factors, but that environmental influences also play a part in its origin. These factors are always interacting and it will often be difficult to disentangle or to distinguish them. In recent times this is clearly demonstrated by the clinical observations of Gregg (1941) and Swan who has pointed out the importance of a virus infection such as rubella as a source of congenital deafness. Hitherto this cause of deafness was unknown and considered to be transmitted by inheritance and as such included in the statistics as sporadic recessive deafness. Therefore congenital deafness is of dual origin. For a clear understanding it is necessary to discuss these two factors of its origin separately.

In heredity the fate of the creature in being is irrevocably destined when the sperm has penetrated into the egg. Then the nuclei of the two sex cells are fused and a change in the composition of the cell will not any more be possible. All the hereditary characters of the new individual and its descendants are assured from generation to generation. In man at fertilization 24 paternal and 24 maternal chromosomes are joined. These chromosomes differ in size and shape, but each special form of chromosome is combined with its identical companion. By the first division of this single cell the development of the new individual starts. The chromosomes are composed of genes, the bearers of the genetical characters of the individual. Their total number is numerous and estimated to be in man "somewhere between 5,000 and 120,000". Each pair of genes has its special and fixed place in the chromosome. By its biochemical action it probably produces materials such as enzymes or catalysors all of identical but different composition, controlling or producing a chain of chemical reactions, which in the end determine the visible character. Consequently one pair of genes is allotted for the developement of the organ of hearing. Now it will depend on the chemical construction of such a gene if there will be a normal or a pathological organ of hearing. If the two genes are absolutely identical and normal or homozygote a normal ear will develop; if they are different in construction or heterozygote a more or less altered organ will arise when the abnormal gene is dominant. When the abnormal gene is recessive the organ of hearing will seem to be sound. Only if at fertilization these

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two abnormal recessive genes come together deafness will appear. In hereditary congenital deafness the chemical construction of the produced enzymes of these genes must in some way be different, though closely related, because different kinds of lesions of the organ of hearing exist and also several other organs may be affected. It might also be that the abnormal products of these genes influence the surrounding genes, so that by their interaction several organs in the neighbourhood are damaged. Because of the heredity of these pathological conditions the abnormal gene must have been changed in composition or, in other words, the original gene must have undergone mutation. The moment of mutation must have taken place while the gametes were formed and ready to couple. What a small chance of mutation of this gene must there have been!

About the endogenous causes of mutation nothing is known. As an exogenous cause cosmic radiations are mentioned, but experiments, made to prove this suggestion, were without results. Mutations may be induced by irradiations of the sex cells by X-rays and radioactive substances as is carried out in experiments on drosophila, the well-known fruit-fly. They may also be produced by physical actions such as heat-stroke or chemical substances such as mustard gas or formaldehyde. In natural circumstances the rate of mutation is very small (1 : 100,000 germ cells in man). But since in a given individual the total number of genes is so great, mutation is by no means uncommon (Crewe). It may be but that amongst all these mutations the gene for the organ of hearing will be affected, will be very rare.

Types of Congenital Hereditary Deafness

There are two types of congenital hereditary deafness, namely dominant hereditary and recessive sporadic deafness.

A. *Dominant inner-ear deafness* is rarely congenital. Usually it develops at middle age; the deafness has a strong progression with often lengthy pauses. There is mostly a loss of hearing for all frequencies. According to its dominance it will be present in every generation with a chance of 50 per cent. of the offsprings according to the Mendelian laws. This type could because of its late appearance, nearly be neglected as a congenital type. However Waardenburg describes extensively in 1951 a hitherto unknown hereditary syndrome as a combination of six chief characters of which one is congenital dominant deafness. These characteristics are summed up in the order of their occurrence:

1. Lateral displacement of the medial canthi and lacrimal points of the eyelids and blepharophimosis.
2. Hyperplastic broad high nasal bridge.
3. Growing together of the eyebrows with hypertrichosis of their medial portion.

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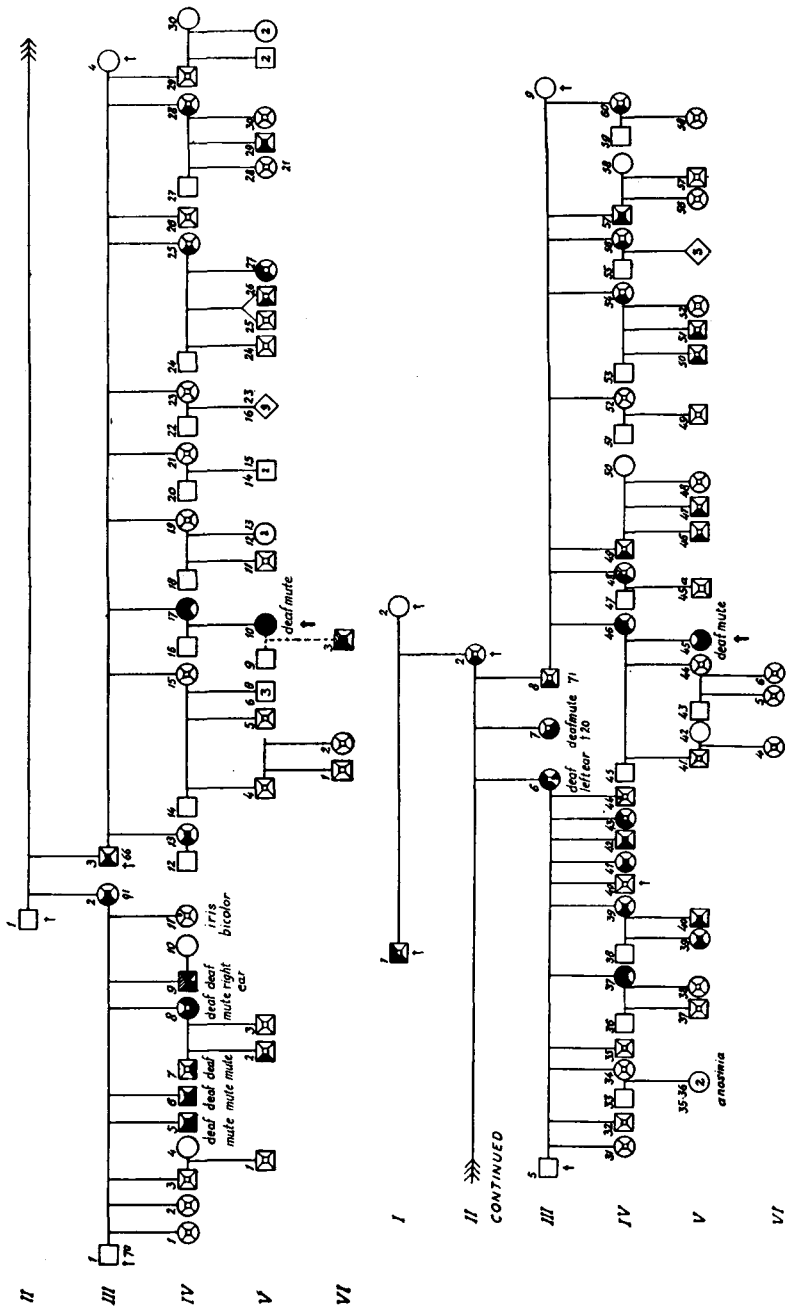


FIG. 1
Pedigree 11, Family de Jo (Waardenburg).

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4. Partial or total heterochromia iridum.
5. Congenital deafness or partial (unilateral deafness).
6. White forelock as a form of partial albinism.

Not everyone of the affected persons have all the characteristics, but 20 per cent. of them are congenital dominant deaf. Waardenburg has gathered 16 pedigrees of this anomaly. It is estimated that about 1·43 per cent. of all deaf mutes in the Netherlands (3,500) owe their deafness to the irregularly expressed dominant form occurring in this syndrome. Fig. 1 shows pedigree eleven published by Waardenburg, in which all these peculiarities may be seen.

B. Congenital sporadic recessive deafness so called because often without known cause only one child in a seemingly healthy family is deaf. If consanguinity exists the chance of deafness rises. This fact was already mentioned by Fay in 1889, therefore long before the laws of inheritance were known. Our countryman de Wilde relates in 1913 that about 7 per cent. of all deafmute children originate from consanguineous marriages.

In what manner such sporadic cases of deafness arise and may be influenced by consanguinity is demonstrated in Fig. 2, taken from a figure

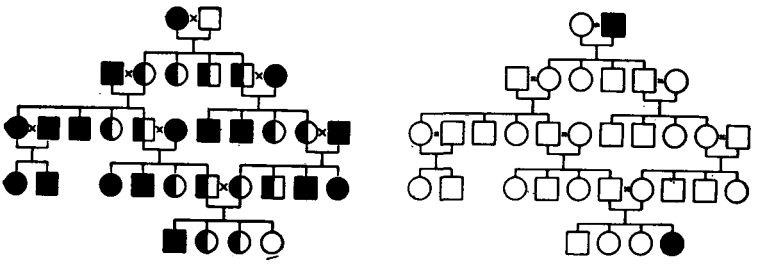


FIG. 2

Pedigree of a recessive human trait. Black is the *normal* dominant phenotype; black-white, the heterozygous individuals. White is the recessive trait, always homozygous if visible. (This is not the usual way of drawing a recessive pedigree, but rather one meant to make the facts at once visible. The usual way is to draw the abnormals black and all normals white, i.e. in this case, all would be marked white except the 2 white persons in the figure, the male at the top and the female at the bottom, the 2 homozygous recessives, who would be marked black.)

of Goldschmidt, beside which the usual way of recording a pedigree is shown. Recessive deafness may be combined with other disorders, disturbed vestibular function, retinitis pigmentosa, feeble-mindedness and hereditary ataxia. According to Hammerschlag all these defects are the outcome of one and the same hereditary disorder. According to Albrecht these disorders are completely independent.

Extensive work has to be done for the recognition of the hereditary character of these cases of deafness. As many as possible of the relatives of the affected persons must be examined. For a proper judgment the pedigrees are usually too short, three or four generations, and it is only from

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information or annotations in family bibles and so on that we may obtain more or less reliable data of the course of heredity in still more generations. The first examinations in this field were done by Albrecht in 1921. The most extensive number of pedigrees are those of Lindenov (1945) in Denmark and of Hopkins, Guilder, Macklin of the Clark School of the deaf in America (1946). The last have calculated out of their material of 310 pedigrees that one recessive gene is responsible for this type of deafness. They found that cousin marriages among the parents of these children are more common than among the population in general, the incidence of cousin marriages being about 8 per cent. in these families, whereas it is only 0·2 per cent. in the population at large. They discuss also the question whether there are not one, but more recessive genes for deafness, because in some of their pedigrees the children of two deaf parents had normal hearing. This might be possible if one of the parents had an acquired deafness. However if both parents have only recessive genes, several kinds of recessive deafness must exist, so that the tendency to deafness of the children is not the same. In this case one gene might interfere with the nerve cells of the cochlea, the other with the fibres of the cochlea nerve. Notwithstanding this extensive and accurate research concerning their pupils uncertainty of the cause of deafness often remains. It must always be kept in mind that these solitary cases of deafness may be not inherited but may be acquired during pregnancy and sometimes also shortly after birth by a non-recognized infectious disease or other factors before the child was talking.

How much easier is it for genetists to study heredity in Drosophila with its 40 generations in one year, each of which with 200-300 offspring. Man needs for the same number of generations a thousand years and their offspring is scanty.

The next figure shows one of the pedigrees of these authors, a pedigree of 11 generations, that must be over a period of nearly 300 years.

The number of cases of congenital deafness by heredity is presumably half to one third of all the deaf, recessive deafness strongly predominating. According to Dahlberg two thirds of the congenital deaf would be deaf by heredity. From these data the importance of heredity as a cause of congenital deafness is clear.

The anatomical data of the two groups of hereditary deafness procure (a noteworthy survey with extensive literature was given by Fraser in 1921 in this Society) an insight in the symptoms of these affections. In recessive sporadic deafness the total acoustic nervous system, that is to say the nerve fibres, the nerve trunks, the central nuclei and tract are deficient, whereas the cochlear bone is intact. In dominant hereditary deafness the development of the cochlear bone is arrested with subsequent defects of the nerve tissue of the spiral and malformations of the membranous connective tissue. In both groups the organ of Corti is more or less

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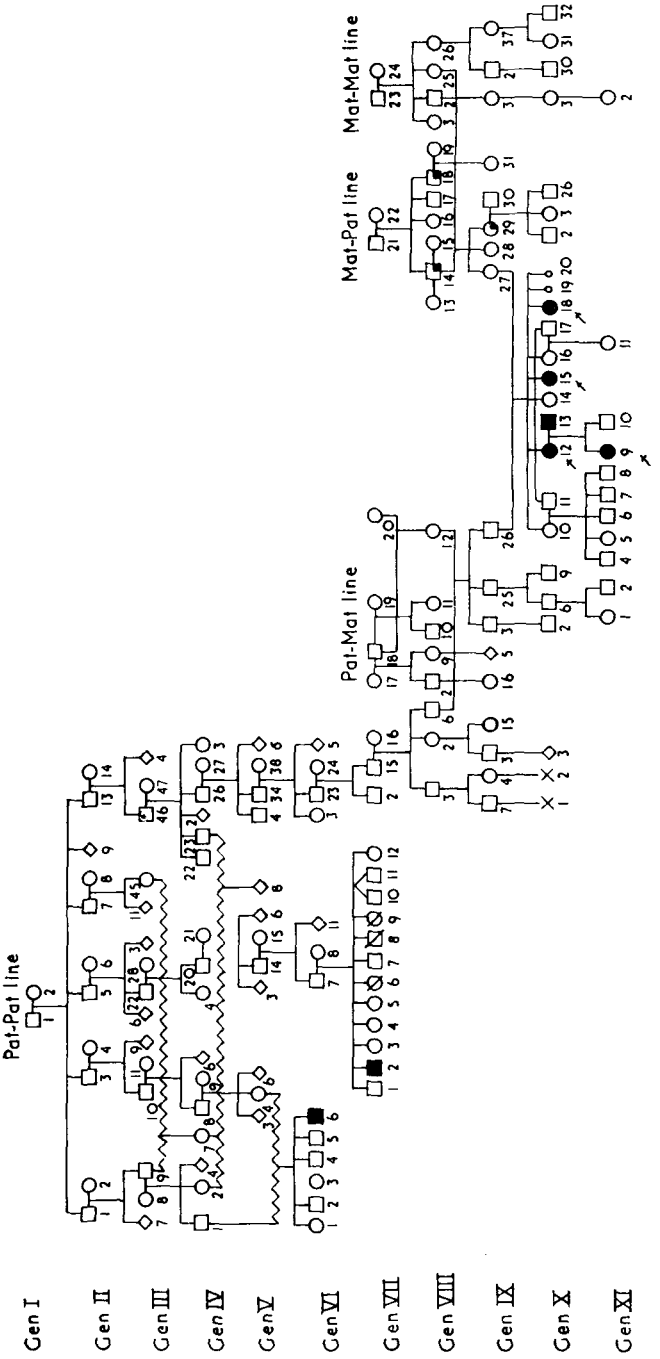


FIG. 3.

Pedigree 14.

VI 6. Born deaf. Died unmarried at 68.
VII 2. Born deaf. Died unmarried at 23.
VIII 14, 18. Hard of hearing since late 60's. Arteriosclerosis and Bright's disease.
IX 29. Hard of hearing in one ear due to "catarrh". Cerebrospinal meningitis as a baby
X 12. Born deaf.
13. Deafness attributed to scarlet fever at three. No deafness in family as far as known.
15. Born deaf.
18. Born deaf.
IX 9. Born partially deaf.
10. Born partially deaf.

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damaged, through which the several degrees of hearing loss can be explained. In both groups are many differences and transitions in the anatomical picture so that a standard image cannot be given. The significance of the results of the examination of the petrous bone is limited because only in a few casesalso has the central nervous system been investigated. Siebenman-Bing (1904), Quix-Brouwer (1910) and Nager (recent) described several changes in the acoustic nuclear region. Saxen (1934) reported in two cases changes in the VIIIth nerve and the nuclear cells. Gauger (1942) examined systematically the central nuclear region of 16 normal persons and compared them with 6 pathological cases of deafness of different origin. In one case of dominant hereditary deafness he found besides the changes in the peripheral organ an extensive deficiency in the development of the central nuclear region, the only case of this kind hitherto described. In 3 cases of recessive deafness the acoustic nuclei were normal, contrary to expectation. Besides the relatively small number of human specimens the petrosa of several deaf animals cat, dog, mouse, etc., were examined by several authors to give more information, all with interesting data and a many-coloured image. From these numerous investigations in man and animal of which an excellent survey with extensive bibliography is given by Altmann (1950) a rather good idea of the anatomical changes in hereditary deafness is obtained. The missing of a standard image is not essential, if we assume (and many facts point to this direction) that the histological deviations are acquired in, and dependent on, different stages of development of the individual.

Atresia Auris Congenita

Another malformation that may produce congenital deafness if both sides are affected is the atresia of the external meatus, often attended with misshaped auricles and chiefly with changes in the middle ear, though sometimes the inner ear also may be damaged. According to Altman three groups of this anomaly can be distinguished. In the first group only changes such as an incomplete annulus tympanicus and a rudimentary drum, unimportant in respect to the loss of hearing exists. In the second group the middle ear is badly malformed. The lateral wall of the tympanic cavity is essentially formed by a bone, called the atresia plate. The stapes plate may be grown together with the fenestra, causing a considerable impairment of hearing, that leads to mutism if both ears are affected. In the third group the inner ear may also be affected. In this case a severe grade of deafness exists, that cannot be cured. Ombrédanne, who has the largest experience in operating on these cases (far more than 100) accepts only the second form. Other anomalies may be also present in this defect. Thus we had in our clinic at the same time two children with a mandibulofacial dysostosis, each of them with an atresia of both ears. In one of them,

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a boy of 6 years, the differential diagnosis between feeble mindedness and not well-developed intellect by deafness was very difficult. The psychologist thought the mental faculties to be quite normal. After opening of the middle ear by Dr. de Wit (very difficult because only a small cell above the round window existed) a fenestration was performed. The boy seemed to hear after the operation. At present we are trying to teach him to speak and to educate him.

The cause of this anomaly is unknown. It must be of different origin. The embryologic development of the inner and the middle ear is not the same, the labyrinth being formed from the otic vesicle, the tympanum and the auricle from the second branchial arch and groove. The data about the heredity of this affection are scanty, but it seems to be dominant hereditary (Potter in five generations (*J. Hered.*, **28**, 255).) In most cases, however, an intra-uterine acquired development is probable.

According to the anatomical deviations there is a conductive deafness, that in many cases can be improved by fenestration.

Erythroblastosis fetalis

may be also a cause of congenital deafness. This affection is the result of an intravital hemolysis caused by an anti-body-antigen reaction in the new born evoked by the antagonism of the different Rh-factors in the blood of mother and child. A few days after birth the child may show an icterus gravis and evidence of damage to the central nervous system, which may result in a neurological symptom complex, called kernicterus, because of an elective icteric discoloration of the central nuclei of the brain. The main neurological symptoms of this affection are athetosis, rigidity and mental defect. Other symptoms may also be present, depending on the localization of the cerebral lesion. It may be localized also in the nuclei of the cochlear pathways resulting in a more or less severe impairment of hearing. The first case of deaf mutism of a child with rigidity of the lower extremities, who had had icterus gravis is described by Cocquet (1944), Crabbé and Gerrard (1950), who published 4 cases of deaf mutism after kernicterus. Gerrard in 1952 published two other cases of children with kernicterus who were totally deaf. Erythroblastosis seems to be an important factor as an originator of congenital deafness and its importance lies also in the fact that serological reactions may cause a severe degree of hardness of hearing and that in future sufficient attention must be paid to them as a cause of deafness of obscure origin.

Environmental Factors

The evolution from the fertilized egg till full-grown fruit may be compared to a long road covered in a relatively short time. A single cell with a weight of about 0·005 mgr. is, after nearly

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30 divisions, developed in 40 weeks into a highly differentiated organism of 5,000 grams. This is a very important period for the individual especially the first 8-12 weeks of pregnancy during which all the organs and organ systems have nearly acquired their defini ape and the embryo has obtained all the features of a little human b In this

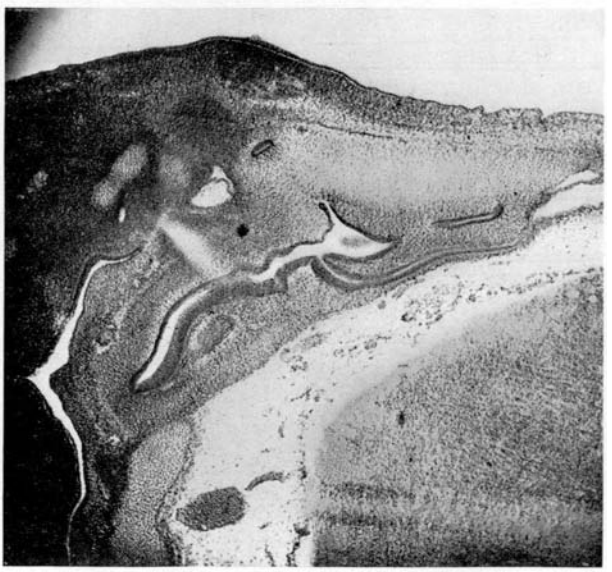


FIG. 4a



FIG. 4b

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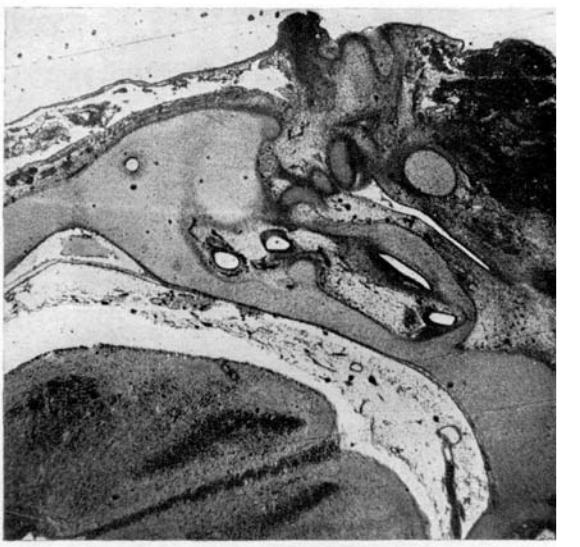


FIG. 4c

Organopoetic period of the organ of hearing, fig. 4a human embryo of 22 mm., fig. 4b and c human embryo of 37 mm.

organopoetic period the still undifferentiated cells are exceedingly vulnerable and exogenous influences may give rise to all kinds of defects. Many characteristics of the individual may be changed by environmental factors, and often in such a way that they cannot be distinguished from hereditary disease. An example of such a deviation of the genotype or such a phænocopy is the congenital deafness after rubella of the expectant mother. The conformity of this deafness with the sporadic deafness may be explained by the analogous chemical structure of agents, influencing the young dividing cells. The question arises if there is an essential difference between the influence of a gen and a virus. For both a virus and a gen consist of a protein-compound to which a rubinucleid is added. The affinity of the metabolics of the recessive gen and the rubella virus may give rise to similar changes in the still undifferentiated groups of cells in the ductus cochlearis. In the pathologico-anatomical structure of both affections there seems to be some similarity. In both affections the development of the organ of Corti is arrested. In recessive deafness this atrophy of the sense elements is either a primary one or a secondary one following a degeneration of the stria vascularis. In rubella, of which only very few specimens are available, Carruthers describes, among other structural changes a degeneration of the stria. Nauta quoted by Miss Kamerbeek, announces an arrest of development of the organ of Corti and a particular precipitate seemingly deformed blood cells in the scala vestibuli, Schall and Lurie and Keleman observed

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in three petrous bones of human embryos of 15½ weeks, 118 mm. in length (obtained by producing abortus in mothers with rubella, so that it may be assumed that the embryo also was infected), besides hæmorrhages in the scalae, an arrest of development of the organ of Corti, which they ascribe to a lack of nutrition caused by structural changes of the blood vessels. This observation would be in agreement with Grüneberg, who assumes that the organ of Corti, in embryos receives its nutrition, not from the stria, but from the vas spirale. How it may be, in several cases of histological examination of recessive deafness it is difficult, as to the ductus cochlearis, to distinguish between hereditary and acquired deafness.

Other infectious diseases such as measles, scarlatina, whooping cough, influenza, etc., seem rarely to lead to congenital deafness. The likelihood of catching these diseases during pregnancy is, with the exception of influenza, very small because many people have already suffered from them in childhood. But it might be that the metabolites of these diseases have not such an affinity for the hearing organ as rubella. In every case prudence in judgment must be exercised, for, after an inquiry, Kamerbeek reported after mumps three cases of congenital anomalies, one of which was congenital deafness and Taylor in 1934 recorded an increase in the number of congenital deaf children, after the influenza epidemic in 1918. He ascribes this increase to the great use of drugs like quinine and salicylates in this period, but at this time the deafness caused by rubella was still unknown.

Syphilis, mostly acquired by the fœtus in the later months of pregnancy may be also a cause of congenital nerve deafness, though the onset of deafness in congenital lues is generally observed later on, at the age of 8-12 years.

To the other defects of endemic *cretinism* congenital deafness also belongs. Formerly this deafness was considered to be of a dominant hereditary type. After the discovery that this disease was evoked by a pre-natal deficiency of iodine and since it is considerably decreased after sufficient iodine supply to the population it is evidently a familiar instead of a hereditary affection. An example again how difficult it may be to make a distinction between hereditary and environmental factors in the origin of congenital deafness.

As to drugs quinine, the salicylates and alcohol, several authors have, in experiments on animals, demonstrated their deleterious effects on the hearing organ of the fœtus. These drugs produced similar changes in the ear of the mother and the fœtus, with greater alteration in the ear of the fœtus. Also in the human fœtus these drugs are found in several organs and in the urine several days after birth, when they were given to the mother some time before delivery. This short survey may be a warning to be extremely cautious in administering drugs to the expectant mother.

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Congenital Deafness

Diagnosis

The diagnosis of congenital deafness in babies, immediately after birth is difficult. The most effective manner of ascertaining the hearing of the suckling is the reaction to strong sounds, with a shock or a palpebral reflex, their absence not being evidence of deafness. Later on the want of development of speaking draws the attention of the parents to the possibility of a defect in hearing of their child.

Often one tries to find an objective clinical method to establish the differential diagnosis between hereditary and acquired deafness, but none of them is conclusive for this decision. This could be expected according to the anatomical data. For though it is generally accepted that in congenital deafness only the cochlea and saccule are affected, the utricle and the canal being normal, many exceptions to this rule exist. It is evident, that the results of the rotation test and caloric test, though sometimes giving some direction to diagnosis, are mostly of little value in this respect. Langenbeck's law of symmetry, that only can be looked for in elder children, and which was some time ago popular in some regions for political aims, has appeared to be worthless, X-ray examination (incidence of Stenvers, Chaussé) may be of some value, a destruction of the labyrinth supporting the diagnosis of acquired deafness. Generally speaking the diagnosis of congenital deafness must mainly be based on anamnestic data of the parents.

Prevention of Congenital Deafness

Acquired congenital deafness may be prevented by special measures and especially in the first period of pregnancy, which is such a precarious time for the embryo. The expectant mother must be brought as soon as possible in the most favourable social and hygienic circumstances. Drugs and foods which may be harmful for the embryo must be avoided. The possibility of preventing infectious diseases must be taken into account and special attention should be paid to the prevention and cure of rubella infection, this disease being the most striking example of the cause of congenital deafness in recent times. In this respect many problems arise, such as active immunization before marriage and passive immunization during pregnancy in epidemic periods, as the induction of abortion when this disease is caught during pregnancy; all these problems are not easy to solve.

It is true to say that in the last century by social legislation, hygienic measures, the raising of the standard of life and the progress of medical science in the so-called civilized countries the frequency of many unfavourable circumstances and diseases is decreasing, so that the number of acquired pre-natal deafness is and will be reduced. On the other hand the incidence of hereditary deafness may relatively increase.

To cure hereditary congenital deafness will not be possible. Therefore

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social measures must be taken to lessen their number. There is much discussion of these eugenic problems. The most drastic measure is, we may safely say, sterilization. I will not go deeply into this matter but one thing is sure, this measure if it is thought to be in accordance with our medical conscience, if it is permitted and legally regulated ought always to be executed only with the own free will of the subject and never forced by law.

Another measure that can be taken is the prevention of a marriage between deaf persons and of consanguineous marriages.

If we are sure (but this will be seldom the case) that one of the partners has an acquired congenital deafness, then there is, of course, no reason to prevent a marriage because all the children will hear well. Before permitting this marriage we must, however, look to the occurence of congenital deafness in the relatives of both partners. For, if this is the case (this is only valid for recessive deafness) both the partners will have a good chance to be heterozygote for deafness and 25 per cent. of their children will be deaf. Generally a marriage between two deaf persons must be dissuaded.

Conclusion

In this lecture I have tried to give a general survey of congenital deafness. It stands to reason that a survey of such an extensive subject is incomplete and insufficient in many respects. Nevertheless the several aspects of the origin and the dual character of congenital deafness are indicated. The difficulty, however, of making a distinction between inherited and acquired deafness remains. Still this distinction is important in view of the prevention of this affection. Nowadays all over the world the interest in the deaf and the hard of hearing is growing and everywhere measures are being taken to cure, to educate and to rehabilitate them. To stamp out deafness as much as possible its origin must be known. Probably many causes of congenital deafness are still unknown. Rubella and erythroblastosis are a warning to be vigilant. The task for us otologists is to detect them.

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