The Primary Causes of Deafness

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THEY GROW IN SILENCE

Suspicion that a child may be born deaf may arise during pregnancy if inherited deafness or maternal rubella (German measles) are involved. Severe neonatal jaundice due to the incompatibility between the blood of the mother and her infant may produce damaging conditions causing hearing loss before or just after birth. In such cases, physicians may apprise parents of the possibility of deafness. Hearing loss may also be due to a severe infection in later childhood that affects the brain or its covering membranes. If the onset is prior to the time the child has started speaking in sentences--about eighteen months to two years--the loss may not be immediately recognized. If it occurs after this age, when the child has begun to communicate verbally, the deafness will be apparent immediately.

Disease Processes Causing Deafness Prior to Birth

There are two major causes of congenital deafness: genetic deafness and rubella. Deafness in the offspring is anticipated most often when one or both parents are deaf, presumably on a genetic basis. Deaf adults often do not know the cause of their own deafness. Members of the current community of deaf adults, at the time of this writing, were born at a time when there was less focus on birth defects. Establishing the cause of deafness at that time was a much more random process. It is likely that many cases formerly thought to be inherited were due rather to subclinical maternal rubella.

If there are profoundly deaf individuals in the lineage of either of the parents of a deaf child, the child's deafness is most likely to have a hereditary cause. Nondeaf parents who have deaf grandparents or other close relatives may have some concern that their child will be born deaf; if most of their relatives have normal hearing, their concern need not be great.

Heredity

Genetic factors have been the leading cause of deafness throughout this century except during certain epidemic periods of rubella. Generally, 50 to 60 percent of all deafness is attributed to genetic factors.

It is often surprising to professionals in deaf habilitation and rehabilitation work--and to parents--to discover that 90 percent of genetic deafness is carried by a recessive gene. This fact is especially surprising in cases where there may be no known deafness in the immediate family. The lottery of genetics is such that, although this recessive gene for deafness is present in approximately one out of ten persons, only six children per ten thousand are deafened genetically. The recessive gene in one parent has to be matched by one in the other parent if a recessive trait is to appear.

Children deafened through heredity causes are less likely to have other defects than children deaf from nongenetic causes. As a group, the genetically deaf do better in school. There is also evidence to suggest that, as a group, they may have slightly higher intelligence test scores than deaf children generally have.

Paradoxically, parents are generally loathe to accept the fact that their child's deafness is of genetic origin. They react to such a diagnosis as if it stigmatizes them. In view of the commonness of recessive gene carriers and the excellent achievement record of genetically deaf children, this attitude, though understandable, is certainly unnecessary.

Where genetic deafness is suspected or established, the family should take two important steps. First, competent genetic counseling should be sought in order that all family members can be made fully aware of the probabilities for deafness in future offspring. Second, regular and complete ophthalmological examinations should be sought because, of the fifty-seven known forms of genetic deafness, there are ten that involve both hearing loss and visual problems.

Rubella

Between 1963 and 1965, a rubella epidemic raged across the United States. It resulted in the birth of more handicapped children than did the thalidomide disaster. Educational programs for deaf children are at present hopelessly overwhelmed by the number of children from this epidemic. Whereas rubella usually causes about ten percent of children's deafness, those preschools serving youngsters born during the 1963-65 period report a 40 to 80 percent prevalence of postrubella cases of deafness. This means that over one-half of all deaf children under the age of six were deafened by rubella.

Rubella is an incipient disease. A mother will be aware of the illness if she has developed a rash, swollen lymph nodes in her neck, and low-grade fever, but often the disease does not come to actual clinical definition. A study was made of mothers of infants who had rubella virus cultured from the infant's urine at birth. The study revealed that 50 percent of these mothers were unaware that they had been infected during gestation. When the rash occurs, it is typically evanescent, and when the mother appears for examination by her physician two or three days later, it may have already disappeared.

Although rubella virus usually causes only a mild illness, it can seriously affect a developing fetus. Deafness is most frequent, but visual problems (cataracts and/or retinal damage), lowered intelligence, or heart defects can also occur. Studies at Johns Hopkins Hospital indicate that 85 to 90 percent of postrubella infants suffer significant physical damage during gestation.

The period of greatest danger to the fetus is the first three months of pregnancy, but damage can occur if the mother is infected even within a few weeks prior to conception, or as late as the eighth or ninth month. Rarely is there significant damage to infants infected after birth.

The rubella virus is not necessarily eliminated from the infant's tissues after birth. It has been cultured from infants as old as nine months. It can remain within various cells of the body and continue to cause cellular damage for, as yet, an undetermined period of time.

Vaccines have been developed and are being distributed that give promise of eventually eliminating rubella as a major cause of deafness. Primary concern remains as to the effectiveness of these vaccines to control epidemics without undesirable effects to the recipients.

Regular physical and audiological examinations are important for postrubella youngsters. Since the probability of other problems such as poor vision, heart trouble, and neurological damage are high, these possibilities must be thoroughly investigated by pediatricians, ophthalmologists, and other appropriate specialists. Sometimes, the parent must assume responsibility for seeking specialized examinations for the child because the family doctor may not always detect subtle visual or neurological difficulties.

Diseases of the Perinatal Period Causing Deafness

There are two prominent causes of deafness related to this period. These are prematurity and blood type incompatibility between the mother and the child (especially where the fetus is Rh positive and the mother Rh negative). As a consequence of blood type incompatibility, the infant becomes jaundiced during the first twenty-four hours of his life. If the jaundice is severe and an exchange transfusion is not performed soon enough, products caused by blood cell destruction are deposited in various areas of the brain. Infants with hearing loss or cerebral palsy are the common sequels.

One would not expect parents to anticipate deafness in a premature infant or one having blood type incompatibility. Some pediatricians may advise parents of the possibility, especially in the case of a jaundiced infant. Most physicians and other professionals do not wish to alarm parents unduly. Physicians, however, should consider these infants as high-risk cases and should follow them accordingly.

Premature Birth

It has recently been discovered that four times more deaf children than nondeaf children are born prematurely. Approximately 17 percent of deaf youth of school age were born prematurely. While the condition itself is rarely a direct cause of hearing loss, the association between prematurity and deafness deserves consideration. This is particularly true when the child is born prematurely and no other cause of deafness can be isolated. Conditions such as lack of oxygen and cerebral hemorrhage, which can damage the nervous system, are more common among prematures than among full-term infants and can cause deafness.

When a child is known to have been born prematurely, he should be regarded as a high risk for hearing loss. If deafness is diagnosed, it must be remembered that thorough physical examinations should be made to check for other problems, especially those involving vision, central nervous system, and the heart. The probability of these kinds of difficulties is heightened in children known to be both deaf and premature.

Complications of Rh Factor

Certain genetic combinations of blood types in parents result in blood incompatibilities between mother and child during pregnancy. One of these occurs when Rh negative mothers have Rh positive fetuses. The mother's antibodies will cross the placenta and enter the bloodstream of the fetus, destroying his red blood cells and leading to a severely jaundiced newborn baby. When this occurs, death may result at or soon after birth. Of those who survive, a large proportion are deaf. Many of these may also have cerebral palsy or problems in language development in addition to those problems due to deafness alone.

Advances in medical science may soon eliminate Rh incompatibility as a significant cause of deafness. Postnatal and in utero blood transfusions are preventing some of the effects of this incompatibility. In addition, a special gamma globulin has been developed which, when administered to an Rh negative mother after the birth of her first Rh positive child, will eliminate the destructive anti-Rh positive antibodies from her blood. This means that her second Rh positive child stands the same chance of being affected as the first child; for without the elimination of the destructive antibodies, he would be in more immediate danger of damage.

Diseases Resulting in Deafness During Later Childhood

Destructive processes during diseases such as meningitis and encephalitis can cause deafness. There may be damage to parts of the brain that are crucial to language learning. The child thus affected has special problems in subsequent language development. After the age of two, it has been observed that most children are able to develop greater language capacity at a progressive rate from that point forward. It is not a linear development, however, during the age period between two and four years.

A child freely communicating with his parents until the onset of meningitis or encephalitis, who is then deprived of this communication, has difficult adjustment problems. The fear of having almost lost his life may also accompany the loss of speech because the disease processes leading to hearing loss also threatened his life. Such may have been the parents' concern too. The near loss of a child can leave parents feeling they must now treat the child with special interest.

Meningitis

Approximately ten percent of deafness in children is caused by meningitis--an inflammation of the meninges, the protective coverings of the brain and spinal cord. The disease deafens an estimated three to five percent of the children who contract it. The chance that deafness will develop depends in part upon the organism causing the meningitis. Several organisms can infect the meninges.

A major clinical problem of meningitis is that clinical symptoms, such as headache, stiff neck, or fever cannot be readily diagnosed in infants and young children. Infants cannot specify the location of their discomfort to the doctor or their parents and can only react to the situation by irritability or tears. Hence, the disease is often not diagnosed immediately. Sometimes it is only when evidence becomes clearer--a fever above 100 degrees, a seizure, or a coma--that the child is taken to a doctor or hospitalized where the laboratory tests required for definitive diagnosis are made. Often an unavoidable delay in instituting therapy may lead to deafness. The fact that premature and very young infants are especially susceptible to meningitis compounds this problem.

The development of antibiotics has greatly improved the survival rate for most victims of the disease. But, introduction of antibiotics into clinical medicine has also changed the characteristics of postmeningitic children. Previously very young children, especially premature infants and infants less than one month of age, died from the disease. Now they survive in significant numbers and frequently have residual effects of sufficient magnitude to interfere with normal adaptation. Examples of clinically detectable effects on the nervous systems are learning disorders, muscle weaknesses, paralyses, and deafness.