PREVENTION OF HEARING HANDICAP

FRANCIS I. CATLIN

Many problems in audiology are not solved because superficial appearances are taken at face value, and little attempt is made to ferret out the principles which lie beneath. The prevention of hearing handicap is such a problem.

Prevention means a 'coming before' or anticipatory action which prohibits a sequence of events from taking place. Note that prevention implies direct action. The ultimate event which we wish to avoid is the development of a hearing impairment which is handicapping to the child. By handicapping, we mean an impairment which interferes with normal development and places the child at a disadvantage relative to other children of similar socio-cultural environment. Our first principle, therefore, is this: *prevention implies direct action.* What direct actions prevent hearing handicaps? Are preventive actions needed?

Some understanding of the magnitude of the problem may be obtained from a recent report published by the Subcommittee on Human Communication and its Disorders to the National Advisory Neurological Disease and Stroke Council of the National Institute of Neurological Diseases and Stroke (1969). In this document entitled 'Human Communication and its Disorders: An Overview', the Committee estimates that there are approximately 40,000 deaf individuals between the ages of 5 and 18 years in the United States (The committee defines the deaf as 'those in whom the sense of hearing is non-functional for the ordinary purposes of life'). In the same report, the task force committee states that approximately 360,000 United States citizens under 17 years of age have auditory problems of one type or another which are less severe than deafness, but which impair communication and hence social efficiency.

Our second principle is no less important: *effective prevention requires defined objectives.* We havejust considered prevalence data defined in terms of non-functional hearing, impairment of communication and social efficiency. Are there other definitions of hearing handicap which need to be considered? In particular, with children, we must be concerned with those hearing handicaps which affect the ability of the child to develop normal communicative skills and to acquire an education. In addition, we should consider those conditions which may not be associated with a hearing handicap at present, but carry a high probability that the child will develop a hearing handicap in the near future.

Objectives are defined in other ways than communicative needs; cost and effectiveness are frequently the Siamese twins of prevention, although not always related. Can we be satisfied if our programme for a prevention is effective for, say, 50 per cent of the population, or 75 per cent? Will we be humanitarians and

Dr Francis I. Catlin, Deptt. of Otolaryngology, Baylor College of Medicine, Houston, Texas, USA.

be satisfied with no less than preventive measures for 100 per cent of the population? Can we afford it? Are there alternatives, when we consider the estimate by the Subcommittee on Human Communication and its Disorders that cost for educating young people with severe acoustic handicaps in the United States of America is at least one hundred and five million dollars a year and that the annual expense of conducting formal educational programmes for acoustically impaired children, including special teachers to meet their needs, is in the order of one hundred and thirty million dollars per year? Unfortunately, preventive procedures are often tempered in quantity and quality by financial considerations because this cost—'big picture' is not seen at the community level. Surely, part of our job, if we take our public health responsibilities seriously, is to make people aware of the value of preventive health maintenance.

The third principle is our discussion of prevention states that *the effectiveness of current preventive measures varies with each specific cause of the hearing handicap.* For some underlying causes, no preventive measures are known, others are quite responsive to preventive procedures, if the latter can be brought into action. Before we consider preventive measures, we must, therefore, know something about the causes of hearing handicap and the frequency with which these causes are encountered.

Consider the problem of incidence: Childhood disease is conventionally divided into a temporal sequence of occurrences: prenatal, perinatal and postnatal, —with the latter category subdivided into infancy (under one year) and childhood. Differentiation between 'congenital' and 'postnatal' hearing impairment illustrates some of the difficulties encountered by the diagnostician, since the diagnosis of hearing impairment is often made only when speech fails to develop normally during the first two years of life, or even later, Fraser adds: 'Furthermore, congenital and postnatal deafness cannot be equated simply with genetical and acquired forms. Congenital deafness may be due to maternal rubella, and, owing to diagnostic difficulties in infance, evidence is lacking as to whether genetical childhood deafness is in fact usually congenital or rapidly progressive in early life' (1964), the latter being a common situation with one strain of recessive deaf mice. Fraser also notes that the degree of hearing impairment varies, mild losses being occasionally encountered with syndromes usually associated with profound childhood deafness. He adds: 'There is even no absolute correlation between educational segregation and degree of hearing loss. Thus, severe hearing loss in infancy is sometimes compatible with normal education, and special schooling may occasionally be necessary with a relatively mild deafness. Other, as yet ill-understood, factors may condition the extent of the child's adaptation to the hearing loss'.

Despite these sources of error, Fraser estimates a prevalence of 1:1,000 for profound childhood deafness. Proctor appears to concur with an estimate of the prevalence of hereditary deafness between 1:2,000 and 1:6,000 births, adding that one-third to one-half of these cases of congenital deafness seem to be due to

hereditary factors (1967). Perhaps of more interest to us is the distribution of incidence among the various causes. Fraser (1964) apportions the causes of profound childhood deafness according to Table 1. His figures are expressed as percentages of the total and are equivalent to actual numbers per 100,000 births in the general population.

TABLE 1

(From Fraser)

Causes of Profound Childhood Deafness

			Percentage
Genetical	Recessive	With retinitis pigmentosa	7.5
		With goiter	3.0
		With abnormal ECG	1.0
		Others	26.0
	Dominant	With pigmentary anomalies	2.5
		Without pigmentary anomalies	10.0
	Sex-linked		1.5
Acquired	Prenatally		6.0
	Perinatally	(including excess of prematurity)	10.0
	Postnatally	(after the end of the prematurity period)	30.0
Congenital Malformations			2.5
		(Total)	(100.0%)

According to Fraser, 51.5 per cent of profound childhood deafness is related to hereditary factors. In spite of the large number of syndromes associated with dominant nerve deafness, such deafness is inherited as a dominant trait in 12.5 per cent and is sex-linked in only 1.5 per cent. The majority of human inherited deafness cases are of recessive type, being 37.5 per cent in Fraser's study. Fraser's study reflects populations of Great Britain and Europe in which relative incidences may reflect the effects of older cultures and significant inbreeding. Therefore, these statistics are not strictly applicable to the United States but serve as a frame-of-reference to which we shall refer again.

More recently, Konigsmark (1971) has reviewed the data on hereditary severe congenital deafness from his own clinic and others within the United States of America. Konigsmark states that the more than 60 types of hereditary deafness known include 17 known types of congenital severe hereditary neural deafness syndromes. These account for more than one-half of all the cases of congenital deafness, and may be divided into four main groups as in Table 2.

The next group of hereditary hearing problems are those in which the hearing loss begins somewhat later in childhood. Proctor (1967) describes several of these syndromes in his classification shown in Table 3.

TABLE 2. Hereditary Congenital Service Deafness

I. ***Hereditary*** *Congenital Severe Deafness with no Associated Abnormalities*

(A) Majority of congenital hereditary deafness syndromes

(B) All show severe hearing loss at birth

(C) Six types:

(1) Dominant congenital severe deafness
(2) Dominant unilateral deafness
(3) Recessive congenital severe deafness
(4) Recessive early-onset neural deafness
(5) Sex-linked early-onset neural deafness
(6) Sex-linked hereditary deafness (recessive)

II. *Hereditary Congenital Severe Deafness Associated with Integumentary System Disease*

(A) Five syndromes with skin or hair pigmentary change. One syndrome with skin thicken" ing over joints and with abnormalities of nails.

(B) All show severe hearing loss at birth.

(C) Seven syndromes:

(1) Waardenburg's Disease (dominant and var. penetrance)
(2) Dominant Albinism and congenital deafness.
(3) Leopard Syndrome (dominant and var. penetrance).
(4) Hereditary piebaldness and congenital deafness (probably recessive).
(5) Sex-linked pigmentary abnormalities and congenital deafness.
(6) Dominant keratopachydermia, digital constrictions and deafness.
(7) Recessive oxychondystrophy and deafness.

III. *Hereditary Severe Congenital Deafness Associated with Skeletal Disease*

(A) Severe congenital hearing loss.

(B) Two syndromes:

(1) Recessive absence of tibia and deafness.
(2) Recessive split-hand and foot syndrome.

IV. *Hereditary Severe Congenital Deafness Associated zvith other Abnormalities*

(A) Severe congenital hearing loss

(B) Abnormality of eye, thyroid, heart, or ear.

(1) Recessive retinitis pigmentosa with congenital severe deafness. (Usher's Syndrome) 5-10 per cent of congenital deaf.
(2) Recessive goiter and deafness (Pendred's Disease).
(3) Recessive heart disease and deafness (Jervell and Lange-Nielsen Disease) 1 per cent of hereditary deafness.
(4) Recessive low set ears and conductive hearing loss.

TABLE 3. Hereditary Hearing loss of Postnatal onset

Dominant Types

(1) Dominant Nerve Deafness without Associated Defects (Onset 6-12 years, high frequency loss).
(2) Hereditary Nephritis and Dominant Nerve Deafness, Alports Syndrome (Onset about 10 years, progressive, bilateral).
(3) Hereditary Nephritis, Urticaria, Amyloidosis and Dominant Nerve Deafness (Onset in childhood).
(4) Dominant Bilateral Acoustic Tumors and Nerve Deafness.

Recessive Types

(1) Amaurotic Family Idiocy Tay-Sachs Disease (Juvenile form).
(2) Hepatolenticular Degeneration Wilson's Disease (Onset 5-40 years).
(3) Schilder's Disease (Onset 5 months to 14 years.)
(4) Friedreich's Ataxia +/—?
(5) Retinitis pigmentosa, Mental Retardation, Dwarfism and Recessive Nerve Deafness, Cockayne's Syndrome (Hearing loss beginning in teens).

Several other proven hereditary diseases usually result in hearing losses of conductive rather than sensory-neural type. These include otosclerosis, in which the transmission is dominant with variable penetrance. The onset occurs after 10 years of age, and predominantly about 20 years; females are affected more than males. Conductive hearing losses are also occasionally encountered in osteogenesis imperfecta.

Before we consider the causes of acquired hearing handicap, let us note that Fraser assigns an incidence of 2.5 per cent for congenital malformations. Most of these malformations are associated with faulty embryonic development of structures derived from the first bronchial arch. The role of heredity is not always clear in many of these congenital malformations. Although some relatively well-defined dominant syndromes exist, there are many intermediate forms between the various malformation syndromes which make precise diagnosis difficult.

The most common malformation, cleft palate, occurs about once in every 850 births in the United States of America. Denmark and Sweden cite an incidence of 1:750 births. In 30 reported syndromes, there appears to be a possible genetic basis. About 5 per cent of cleft palate children have a positive family history for cleft palate. Other anomalies are found in 15 per cent of cleft palate children; 2 per cent have deformities of the ear. Studies of hearing impairment with cleft palate show incidence figures ranging between 25 to 90 per cent with an average incidence of 60 per cent. There is a higher incidence of hearing loss when anomalies are present, but the relationship is not clear. Ear pathology, usually serous or suppurative otitis media, is present in almost one-half (45 per cent) of the cases; and is bilateral in 83 per cent of these. The hearing loss is primarily conductive and secondary to the otitis media.

The acquired causes of profound childhood deafness are of almost equal importance in hereditary factors. Fraser assigns an overall incidence of 46 per

cent to the acquired category, with a further division into prenatal (6 per cent), perinatal (10 per cent) and postnatal (30 per cent) occurrence. That these **figures** fluctuate may be illustrated in Table 4 which, describes the presumed aetiologies for 138 pupils of the Clarke School for the Deaf in Northampton, Massachusetts, U.S.A. (Chung and Brown, 1970).

TABLE 4. Aetiology of Hearing Impairment in Pupils of the Clarke School **1954**

Aetiology	*Percentage of Pupils*
Hereditary	26 **per** cent
Rubella (congenital)	9 **per** cent
Erythroblastosis	3 **per** cent
Birth Injury	1 **per** cert
Postnatal	26 per cfi.it
Unknown	35 **per** cent
(Number of pupils in sample)	**(138)**

In 1964, Fraser assigned an incidence of 5,6 per cent to prenatal rubella; this figure has now risen to about 1 0 per cent in the United States making this virus disease the major cause of acquired prenatal hearing loss today While the statistics for the entering class at the Clarke are not significant because only 21 pupils are reviewed—nearly one-half of these were related to proven congenital rubella infection.

Other infectious diseases appear less important: Prenatal cytomegalovirus (CMV) infections are reported to have produced hearing loss, but the exact incidence is unknown at this time. Syphilis and toxoplasmosis have been documented as aetiological factors, but there is little likelihood that these diseases contribute a significant number of congnital hearing impairments in the United States today. Ototoxic drugs do not constitute a major cause of prenatally acquired hearing impairment at the present time. While some reports of hearing loss have been reported following the maternal use of streptomycin and thalidomide, these cases are few in number and thalidomide toxicity was never a problem in the United States.

Hearing loss resulting from perinatal injury has been ascribed to many factors including prematurity, multiple births, perinatal fetal distress and other unfavourable perinatal events. Fraser suggests that not less than 10 per cent of profoundly deaf children owe their impairment to perinatal misfortunes. Neonatal hyperbilirubinemia accounts for about 4 per cent of this incidence, according to Keaster (1969) and this figure is in accord with the Clarke School data. Erythroblastosis is, of course, genetically determined, although not commonly regarded as a hereditary disease.

Postnatally acquired hearing impairments arise from a number of conditions: Hereditary hearing loss of late onset and otitis media secondary to cleft palate have already been mentioned. Other sources of postnatal damage to the auditory

system include head injury, viral infections of infancy and early childhood, and pyogenic meningitis. Fraser has suggested that about 30 per cent of profound childhood deafness comes from postnatal causes. Prevalence figures for less severe hearing impairments lie between 15 to 30 per 1,000 school children. Approximately 80 per cent of these milder hearing losses are caused by the complications of upper respiratory infection involving the middle ear.

Acoustic trauma has come into the limelight as the result of increased attention to the conservation of natural and human resources. Many authorities are beginning to question the wisdom of testing infant hearing with high-intensity sounds and the advisability of using high-gain hearing aids casually on such young children (Glorig, 1971;. Trauma from amplified music, a by-product of the electronic era, presents a hazard peculiar to the young who have learned to accept loudness, tinnitus and temporary threshold shift of hearing as essential to 'real listening'.

Finally, we must consider what, for the sake of a better label, should be entitled socio-cultural causes. These include factors of hygiene, nutrition and cultural habit or community acceptability which predispose to hearing impairment. A few questions will serve to illustrate this category: why do some children consistently have hearing loss due to impacted cerumen?; there once was a popular song with a lyric that ends: 'beans in our ears'. I can't remember the number of aural foreign bodies we had to remove that year; why do some communities show much higher incidences of chronic otitis media?; why do some communities support only token hearing conservation measures?; and lastly, what can we do about it?

How Can We Prevent Hearing Handicap?

Prevention of hereditary deafness in today's world means that effective ways of genetic control need to be developed. The mode of inheritance is the key to consideration of the prevention of hereditary deafness. In dominant transmission, all offsprings will be affected if both parents have the appropriate genetic defect. If the parent carries the effected gene, the chance of the offspring being affected is usually 50 per cent. Fortunately, the dominant form is less common than the recessive form. In recessively inherited deafness, it is necessary that both parents be carriers of the particular gene, in which event, the chance of the offspring being affected is only 25 per cent. A family history of recessive deafness in each parent does not mean that the causative gene is necessarily the same, hence, the probability for hearing impairment in the offspring will vary widely.

Except in striking cases of dominant transmission, establishment of a detailed pedigree is often needed to determine the presence of hereditary factors, and at times, the results will be equivocal. At best, genetic counselling should

be made available for any person with a severe unexplained hearing loss or with a family history of hearing impairment. In cleft palate counselling, for instance, the adviser should be aware that the likelihood of a cleft palate child is 2 per cent if one parent has a cleft palate, 5 per cent if one sibling has a cleft palate and 14 percent if one parent and a sibling have the deformity (Hoopes, 1971). Inhisreview of hereditary hearing loss, Marcus (1969) comments that we have not yet realized the potential of genetic counselling programmes which, despite some practical problems of today, remain our only current satisfactory methods of genetic control.

The prevention of prenatal viral infection has already received a big boost with the availability of vaccines for mumps, measles, pertussis, and now, rubella. Some questions have arisen concerning the efficacy of the rubella vaccine; until these are settled, we should not ignore the potential risks of a maternal skin rash or unexplained fever during pregnancy.

Rh incompatibility requires sound obstetrical judgement, as do all kinds of perinatal distress. Exchange transfusions are helpful but not the final answer—as the degree and duration of Kernicterus may vary greatly. Prematurity still requires elucidation of its causes.

Ototoxic drugs require only one statement: *Do not use them unless absolutely necessary,* and then only with thorough understanding of the risks involved. Evaluate the patient's hearing periodically when possible.

Now, I am not going to suggest that we can eliminate otitis media. There will always be people who insist upon boarding aircraft with a fresh head cold!! On the other hand, there is much that we can do to reduce the incidence of otitis media and the likelihood of permanent hearing handicap; such preventive steps include:

1. Reduction of eustachian tubal obstruction by careful adenoidectomy, if needed.
2. Control of allergy.
3. Control of acute bacterial infection of the upper respiratory pathways (nasopharynx, ethmoid sinuses, especially).
4. Control of middle ear infection by myringotomy, when needed.
5. Followup to ensure that middle ear infection has subsided.

Steps 1 and 2 require that a careful health history be taken, preferably before the otitis media occurs.

The prevention of noise-induced hearing impairment in children, such as may occur with prolonged exposure to loud amplified music, poses some problems. Noise levels may be monitored at public music programmes (an unlikely procedure), but most parents do not check the loudness of the radio or phonograph used by their children. Perhaps the best way to manage this difficult problem with those 'difficult' teen-agers is to develop programmes to teach school-age children some of the known facts about the effects of noise upon hearing and how to recognize

when hazardous noise levels may be present. Such a programme could and should be a part of every school hearing conservation programme, and might be a suitable topic for parental instruction now and then.

The last group, social-cultural-economic cause, deserve considerable comment. It is hardly surprising when children accustomed to poor nutrition and hygiene show greater susceptibility to ear infection and subsequent hearing losses. The hazard is that one part of our culture now accepts chronic draining ears in childhood as a normal part of growing up; that another segment regards excessively loud noises as desirable; that another group feel that genetic research is all right, but genetic counselling is not; that many public health programmes regard hearing conservation as too expensive and probably unimportant, since hearing loss is not lethal. These cultural attitudes deserve as much attention and research as some of the primary causes of hearing handicap, for they may be equally important, if not more so.

In conclusion, let me summarize those factors which appear to be essential for the prevention of hearing handicap:

1. Identify the specific cause (s) if possible.
2. Assess the efficacy of current preventive measures.
3. Evaluate significant social, cultural, and economic factors.
4. Define the objectives to be achieved.
5. Mobilize preventive services (by direct, positive action).

The objective of modern health maintenance techniques in hearing programmes for the young is not simply the treatment of a child with a hearing handicap, but the prevention of impairment and the conservation of normal hearing.

REFERENCES

Chung and Brown, (1970); Family Studies of Early Childhood Deafness Ascertained through the Clarke School for the Deaf: *Am. J. Human Genetics;* 22, 630-644.

Fraser, (1964); Profound Childhood Deafness: *J. Med. Genetics;* 1, 118-151.

Glorig, (1971); Routine Neonate Hearing Screening: *Hear, and Speech News;* Jan-Feb., 4-7.

Hoopes, (1971); Personal communication with the author.

Keaster, Hyman and Harris, (1969); Hearing Problems Subsequent to Neonatal Hemolytic Disease or Hyperbilirubinemia: *Am. J. Dis. Child.,* 117, 406-410.

Konigsmark: (1971); Hereditary and Congenital Factors Affecting Newborn Sensorineural Hearing: Proceedings Conference on Newborn Hearing; California Department of Public Health, San Francisco, California.

Marcus, (1969); Recognition and Diagnosis of Hereditary Deafness with Tomographic Correlations: *Trans. Am. Acad. Ophth. and Otol.;* 73, 409-419.

Proctor and Proctor, (1967); Understanding Hereditary Nerve Deafness: *Arch. Otolaryngology;* 85, 23-40.

Sub-committee on Human Communication and its Disorders: (1969); Human Communication and its Disorders—An Overview: National Institute of Neurological Diseases and Stroke; U.S. Dept. Health, Education and Welfare: Bethesda, Maryland.