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Prevalence and aetiology of profound deafness in the general population of Sichuan, China

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Abstract

People with profound deafness were surveyed in the general population of Sichuan. The results showed that the overall prevalence in Sichuan was 0.186 per cent (236/126 876) of the general population, males 0.212 per cent (135/63 741), females 0.159 per cent (101/63 135) ($p \leq 0.05$). Two hundred and fifteen (92 per cent) cases were older than 10 years, but 227 (96 per cent) lost their hearing under the age of 10 years, 192 (80 per cent) under five years, and 104 (44 per cent) were congenital. There was no significant difference in prevalence of deafness between the population located in urban and rural areas. However, the population living in the flat area and mountain area had a much higher prevalence than that of the hill people ($p \leq 0.05$). Amongst the nationalities investigated, there existed significant differences in prevalence. Both the population of the Hui and the Lisu presented a significantly higher prevalence of profound deafness than that of the others. In aetiology, genetic factors may account for 43 per cent (101/236) of the cases, amongst which, autosomal recessive (AR) deafness is 92 per cent (95/104) and autosomal dominant (AD) 5.4 per cent. Environmental factors were thought to be responsible for 35.6 per cent (84/236) of cases, including infectious diseases (31 per cent), ototoxic antibiotics (2.6 per cent), injury (1.7 per cent), and asphyxia (0.4 per cent). The number of cases of unknown origin were 20.3 per cent. It is suggested that, in China, the importance of genetic factors in hearing impairment should be emphasized. In addition, infectious diseases and ototoxicity play an important role in causing deafness.

Key words: Deafness, profound; Prevalence, aetiology

Introduction

Hearing loss is a major community health problem and creates a serious medico-social problem in China. There have been many investigations on population prevalence and aetiology of profound deafness in industrialized countries, but few studies have been undertaken in these aspects of profound deafness in third world nations including China. However, such studies can provide valuable information about the prevalence and aetiological factors of hearing loss, and can provide a meaningful basis for the planners of medico-social policies.

Sichuan is the most populous province in China, with 104 million people (1987). It is quite representative of all China, in economic, cultural and demographic aspects. During 1986–1987, a large-scale genetic-epidemiological survey was carried out (Zhang *et al.*, 1990). This covered 100 million people, a sampling frame population of 11 168 000 and a sample population of 126 876 people. More than 400 doctors, secondary medical personnel, epidemiologists, statisticians and computer workers joined the field, clinical or laboratory work. As a part of this project, people with hearing impairment were surveyed, and the prevalence and aetiology of hearing loss in the general population was estimated.

Materials and methods

Sichuan is a large province in southwest China. Ninety-seven per cent of its population are Chinese (Han). The rest are Yi, Tibetan, Qiang, Lisu, etc. Figure 1 shows the uneven population distribution and the referral sites from which epidemiological and aetiological data of hearing loss was obtained. The referral sites were selected according to the stratified random sampling method considering the density, and geographical and urban–rural distribution of the population, so that the results would be as representative for the whole province as possible. Details about the selection of the sites (see Appendix) has been published (Zhang *et al.*, 1990).

Screening

A three-stage protocol was adopted. In the first stage all inhabitants of a referral site were questioned and examined using a standardized comprehensive questionnaire and examination scheme by one of the team of doctors and secondary medical personnel. The examinations designed to screen for hearing loss in this stage included whispered voice testing ($\geq$ 6 years), and behavioural tests ($<$ 6

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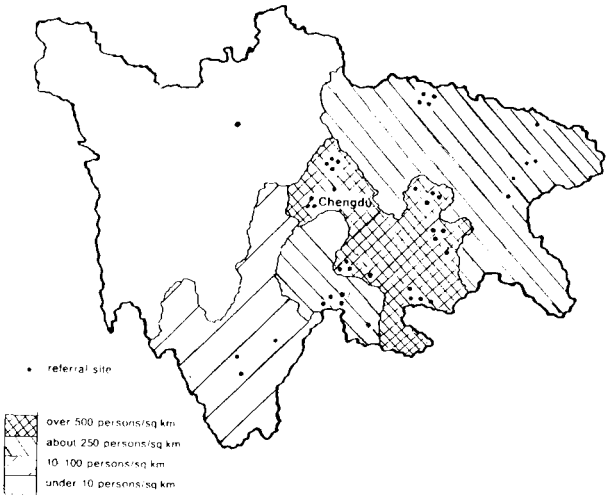

FIG. 1
Population distribution in Sichuan and location of referral sites.

years). In total, 126 876 sample persons (63 741 males and 63 135 females) were investigated and recorded.

In the second stage, those suspected of hearing loss were referred to otologists and other specialists (including a paediatrician, obstetrician, ophthalmologist, dermatologist, neurologist, surgeon and physician) to confirm the causes of hearing loss and to identify the associated organ abnormalities. Pure tone audiometry and oto-immittance measurements were conducted wherever possible. All were otoscopically examined. Non-audiological examinations included: T3, T4 levels; urine tests for proteinuria and haematuria; ECG; skull X-ray; and also chromosomal analysis.

The last stage involved investigating those cases where no environmental factors were responsible for their hearing impairment. Initially, a detailed medical history was obtained with emphasis on family history of deafness and other associated congenital anomalies, and a pedigree including at least three generations was collected. Secondly, routine otological examinations (including pure tone test and oto-immittance audiometry) and physical examinations were carried out in all available family members. Finally, segregation analysis was applied in those families with parents of normal hearing (Liu and Xu, 1988).

Results

Amongst 126 876 people examined, 236 were found to have profound deafness as defined by a hearing loss of greater than 90 dBHL. The overall prevalence was 0.186 per cent of the general population with a significant difference between males (0.212 per cent) and females (0.160 per cent; $p \leq 0.05$). The distributions of various ages and affected ages are shown in Tables I and II.

Although there is no significant difference in prevalence of deafness between populations located in urban and rural areas (0.167 and 0.192 per cent, respectively), there exists significant differences between geographical areas (Table III). The prevalence in different nationalities is shown in Table IV. In 79.7 per cent of cases a causative factor could be identified while in 20.3 per cent the cause of the hearing impairment was unknown, as shown in Table V.

TABLE I
DISTRIBUTION OF AGE IN 236 PROFOUNDLY DEAF CASES

Age (years)	No. examined	No. of cases	Percentage	Prevalence (percentage)
0	8 915	10	4.24	0.112
5	8 826	11	4.66	0.125
10	16 416	18	7.62	0.110
15	38 231	40	16.95	0.105
30	26 613	106	44.92	0.398
45	16 454	39	16.52	0.237
60	11 421	12	5.09	0.105
Total	126 876	236	100	0.186

TABLE II
DISTRIBUTION OF AFFECTED AGE IN 236 PROFOUNDLY DEAF CASES

Age (years)	No. of cases	Percentage
Congenital	104	44.09
0	88	37.29
5	35	14.83
10	2	0.85
15	3	1.27
Unknown	4	1.69
Total	236	100.00

Discussion

Prevalence of profound deafness

Hearing impairment is a common sensory problem. Some epidemiological studies have been undertaken in industrialized countries, from which, information about prevalences has been obtained, but usually prevalence frequencies are different because different identifying standards and screening methods have been used (Davis, 1983; Fraser, 1976).

Estimates of the prevalence of hearing impairment in China are not accurate. In the studies carried out so far (Hu, D. N. *et al.*, 1987; Jiang, S. Z. *et al.*, 1988), the authors selected groups of people in schools for the deaf or in certain geographical locations. Therefore, this sampling represents a biased view which is not representative of the population as a whole. In order to estimate the actual prevalence of hearing loss in the population, it is necessary that people be examined and tested from different geographical regions. In addition, the audiological tests must be standardized to encompass all ages and degrees of hearing loss. The present survey aims to provide an accurate measurement of the prevalence of profound deafness in the general population of Sichuan.

The prevalence of profound deafness in the general population of Sichuan obtained from present survey was 0.186 per cent. There was a significant difference in the prevalence of profound deafness between males and females with males having a higher occurrence. However, there was no significant difference between the sexes in the congenital group (male 0.086 and female 0.078 per cent, respectively). This observation of a sex difference in the prevalence of deafness may be in part due to the fact that men are exposed to more auditory insults (e.g. working environment 'noise') than women. Two hundred and twenty-four cases (95 per cent) were identified ranging in age from 0 to 59 years (Table I). However, 227 (96 per

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TABLE III
THE PREVALENCE OF PROFOUND DEAFNESS IN DIFFERENT GEOGRAPHICAL AREAS

Geographical area	No. examined	No. of cases	Prevalence (percentage)
Flat	33 597	73	0.217
Hilly lands	52 107	64	0.123
Mountainous	41 172	99	0.240

$p \leq 0.05$.

cent) lost their hearing under the age of 10 years, and 192 (80 per cent) under five years (Table II).

At present, there are no reports about the prevalence of deafness in different geographical areas and amongst different nationalities in China. The results showed that there existed no significant difference between the population living in town and that in the country. However, the population located in the flat and mountainous areas had a significantly higher prevalence than that of the hill people (Table III). It was found that there was a significant difference in prevalence of profound deafness amongst different nationalities in Sichuan. Both the population of the Hui and the Lisu presented a significantly higher prevalence than the others (Table IV). The reason for the difference between geographical areas and between nationalities is unclear.

The aetiology of profound deafness in the population

Data on the aetiology of deafness in the general population are required in order to apply preventative measures. However, it is difficult to carry out this type of study, as there is no worldwide consensus about protocols for investigation and diagnosis, also making accurate judgments about the aetiological effect of genetic factors in hearing impairment is not easy. In the present study, special consideration was given to the effect of genetic factors.

In this investigation the aetiology of hearing impairment has been clarified in 80 per cent of the cases. In 20 per cent the aetiological factors were unknown. As shown in Table V, the main causes of profound deafness in the general population of China are genetic factors (43 per cent) and infectious diseases (31 per cent). Estimates of the percentage of cases with hereditary deafness vary widely from three to 78 per cent (Williamson and Steel, 1991), but most estimates suggest that genetic factors may be responsible for about 50 per cent of profound deafness (Fraser, 1976; Konigsmark and Gorlin, 1976). Our survey supports this conclusion. In genetic deafness, non-syndromic autosomal recessive (AR) is the most common (88 per cent). This may be explained largely by the fact there was a high incidence of consanguinity in some populations (15 per cent of the congenital deafness group in this study). Of the syndromic deafness, only Waardenburg, Pendred and Usher syndromes have been identified, which is compatible with the fact that they are the most frequent deafness syndromes (Fraser, 1976; Williamson and Steel, 1991). The prevalence of the three syndromes is: 1:125 000; 1:62 500; 1:125 000; respectively. In chromosomal defects, only Down syndrome was found in three cases – prevalence 1:41 700.

TABLE IV
THE PREVALENCE OF PROFOUND DEAFNESS IN DIFFERENT NATIONALITIES

Nationality	No. examined	No. of cases	Prevalence (percentage)
Han (Chinese)	116 534	210	0.180
Yi	2 964	7	0.236
Tibetan	2 933	1	0.034
Miao	2 157	4	0.185
Lisu	1 968	13	0.661
Hui	273	2	0.732
Others	39	0	0

Environmental factors may account for the range 20 to 50 per cent of hearing impairment (Williamson and Steel, 1991). The present investigation showed that 36 per cent of profound deafness was attributed to acquired deafness, including infectious diseases (31 per cent), ototoxic antibiotics (2.55 per cent), trauma (1.7 per cent). In the infectious diseases, the group defined as 'high fever' are those whose hearing was lost following high fever but the causes could not be distinguished in the survey. Meningitis (7 per cent) and measles (5 per cent) are two other significant causes of infection in profound deafness. The high proportion of infections in the acquired deaf group may reflect differences in aetiology between industrialized countries and developing countries. In China there is surprisingly little mention of the aetiological effect on hearing of viral factors such as rubella which is cited as significant cause of congenital deafness (Fraser, 1976; Newton, 1985). One of the probable reasons is that there are few routine serological tests for the virus in China. In the present survey, those cases resulting from rubella may have been placed in the unknown category.

Conclusion

A epidemiological survey of deafness in a 126 876 sample population was conducted in southwestern China's Sichuan province. The results revealed 236 cases of profound deafness. Prevalence was 0.186 per cent of the general population. Of the study group 96 per cent lost their hearing during childhood. Amongst different

TABLE V
DISTRIBUTION OF AETIOLOGY IN 236 DEAF CASES

Causes	No. of cases	Percentage	Prevalence (percentage)
Genetic	101	43.00	0.0796
AR (non-syndromic)	92	38.98	0.0733
AD (non-syndromic)	5	2.12	0.0039
Waardenburg syndrome	1	0.42	0.0080
Usher syndrome	2	0.84	0.0016
Pendred syndrome	1	0.42	0.0008
Chromosomal defects			
Down syndrome	3	1.00	0.0240
Environmental	84	36.00	0.0662
High fever	41	17.37	0.0332
Meningitis	17	7.20	0.0134
Measles	12	5.08	0.0095
Encephalitis	3	1.27	0.0024
Ototoxicity	6	2.54	0.0047
Injury	4	1.69	0.0032
Asphyxia	1	0.42	0.0008
Unknown	48	20.00	0.0378
Total	236	100.00	0.1860

AR: Autosomal recessive; AD: Autosomal dominant.

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nationalities and geographical areas there were found to exist significant differences. In terms of the aetiology of hearing loss, 43 per cent of cases were due to hereditary factors, 31 per cent infectious diseases and 2.6 per cent ototoxic antibiotics. It is suggested that, in China, genetic and infectious factors are the main causes of profound deafness. Early assessment of hearing in handicapped children and procedures for aural rehabilitation are urgently needed.

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Appendix

Details of population and sampling

The area of Sichuan is 570 000 square kilometres, about six per cent of the total area of China. Geographically, it consists of the West Sichuan Plateau with an average altitude of over 3000 metres and the Sichuan Basin, below which lies Chengdu Plain. The land slopes gradually downward from northwest to southeast. Three landtypes, mountainous, hilly and flat, could be distinguished with populations in the ratio 1:2:1, respectively. The population of Sichuan is about 104 million (1986). Its distribution is uneven, with the urban–rural ratio about 1:4. The density distribution of the population is shown in Figure 1.

The principle of the sampling is that the population of the referral sites in total should be representative in respect of geography and population distribution of the province. Therefore, the ratio of referral sites chosen for mountainous, hilly and flat area is 1:2:1, and the ratio of the sites in urban and rural areas is 1:4. Each referral site consisted of two to four neighbouring villages or residents' committees (in towns) with about 3000 people or 500–600 families. Selected population groups such as army units, schools, colleges and sites with massive emigration in the past 20–30 years were excluded. Forty-two villages or towns were chosen to be the referral sites which were compatible with above requirements. A large sample of people (126 876) were screened in the first stage. They were 96 per cent of the population in the referral sites. The questionnaire used in this stage included family and medical history of all diseases investigated in this survey. In terms of deafness, special emphasis was placed on: (1) identifying the presence or absence of deafness; (2) ascertaining the age at which the hearing deficit was noticed by the parents, as well as the age at which the problem was diagnosed; (3) investigating consanguinity; (4) determining the possible causes of hearing impairment. Through testing, the screening effect of the first stage for deafness it was found that sensitivity was 87.5 per cent and specificity 93.3 per cent.

In the present study the proportion of autosomal recessive cases in the unknown aetiology group with parents of normal hearing was determined by segregation analysis. This statistical technique estimates the segregation ratio (P), proportion of sporadic cases, and ascertainment probability. For our analysis we used the method of Finney (1949) as described by Vogel and Motulsky (1979). This method assumes complete ascertainment, which is appropriate because all affected individuals in the referral sites were ascertained. Analysis of multiplex sibships (more than one affected sibling) with unaffected parents gave a segregation ratio of 0.33 ± 0.07, compatible with autosomal recessive inheritance. The proportion of non-genetic cases was estimated from the proportion of simplex (one affected) and multiplex families.

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