PREVALENCE AND ETIOLOGY OF CHILDHOOD SENSORINEURAL HEARING LOSS IN RIYADH

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Epidemiological data on hearing impairment are indispensable in order to provide effective audiological services. .A random sample survey was performed on 6421 Saudi children to evaluate the prevalence. degree and etiology of sensorineural hearing loss (SNHL). The overall prevalence of SSHL was 2.6% (16816421). The prevalence of severe to profound bilateral SNHL was 0 4% The causative factors were: heredofamilial (66. 1%), perinatal adverse factors (10.1%). meningitis (8.94). rubella (2.4%). mumps (2.4%). and nonhereditary syndromes (1.2%). *The etiology was deemed to be unknown in 8.9% of the cases* The study showed that SNHL is more prevalent in Saudi Arabia than in most of the developed and dev eloping countries. Great efforts and resources are needed for prevention and treatment of this major health problem. *Ann Saudi Med 1996;16(3):262-265.*

Sensorineural hearing loss (SNHL) in children is a health problem which has immense psychosocial as well as economic impact. The psychosocial implication of SNHL consists of the response of the individual. the affected family. society in general. and the deaf community in particular.[1] The economic impact focuses on the cost involved in the identification. habilitation. and education of the hearing-impaired. Information about the prevalence. degree and etiology of SNHL are essential to evaluate the extent of these effects; and to institute the appropriate audiological care. Unfortunately, such epidemiologic data are not available where they are most needed. The purposes of this study were to identify the prevalence of childhood SNHL in Riyadh. including determination of hearing thresholds, and to obtain information on the causative factors.

Material and Methods

A random sample survey of 6421 Saudi infants and children between six months and 12 years of age was carried out in Riyadh. Saudi Arabia. from May 1988 to September 1990. The design was essentially a three-stage stratified random sampling using age and sex as stratifying factors in the final stage. The city was divided into 93 administrative areas and these areas were distributed into six strata according to socioeconomic homogeneity. One-fifth of the areas in each stratum were chosen by a simple random method. Each area was further subdivided into roads and the latter were subsequently divided into smaller blocks of approximately equal sires: a sample of each block was randomly selected. Within each selected block, a sy stematic process was used, whereby a random starting point was chosen and a predetermined zigzag route followed. calling at every other household encountered. There were two survey teams, each comprised of an otolaryngologist. a social worker. and a field supervisor. The team established whether there were any children in the household and if so. after obtaining a consent from the family, a history was taken. An ear examination was then carried out by the otolaryngologist.

Children considered to be at risk of hearing impairment were identified. This included children who complained of hearing impairment or gave a history of a possible or probable cause of deafness (e.g., positive family history. intrauterine infection. perinatal insult. meningitis). These children were referred to King Abdulaziz University Hospital for hearing evaluation and supplementary diagnostic investigations. Hearing was evaluated by pure tone audiometry or auditory brain stem responses (ABR). depending upon the mental age and the cooperation of the child. The type of hearing impairment was determined by pure tone audiometry or by analyzing the abnormalities in the latency level curve extracted from ABR pattern.* In this paper. only children with SNHL will be reported. The degree of SNHL was determined by ABR threshold or by the pure tone bone conduction threshold averaged over 500, 1000, and 2000 Hz. A threshold of less than 20 dB was considered normal. In bilateral cases, the threshold of the better ear was determined. Ophthalmological, genetic and serological (antibodies against rubella, toxoplasmosis,

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Accepted for publication 25 October 1995. Received 18 April 1995.

threshold is probably generated by conventional pure tone audiometry, properly calibrated to international standards.[5] This test, however, is not applicable to assessment of infants, some young children or many handicapped patients. Therefore, ABR may be used for these children: and is generally accepted to be highly sensitive and reproducible for hearing loss greater than 30 dB.[6] However, some unresolved difficulties relate to the extent to which ABR can provide frequency-specific information and can distinguish between conductive hearing loss and SNHL.[5]

A major contribution to the high overall prevalence of SNHL in this study is the very high number of hereditary SNHL cases. This accounted for 66.1% of all SNHL cases, with a prevalence of 173/100,000. This is much higher than the prevalence range of 8/100,000[7] to 54/100,000[8] seen in the literature. This could be attributed to the consanguinity[9] which increases the risk of transmission of both the autosomal recessive and the polygenetic (multifactorial) inheritance.[10] The majority of hereditary deafness (70%-80%) is known to follow an autosomal recessive inheritance pattern.[10]

Perinatal adverse factors were found to be the most common nongenetic etiological cause for SNHL (10.1%). In a review paper, Davidson et al. found that, on average, 13% of SNHL is caused during the perinatal period.[5] It is widely accepted that premature neonates are more at risk for perinatal hazards than the full-term infants. Van Zanten et al. reviewed the literature and demonstrated 10-100 times higher prevalence of SNHL in the preterm infants than in the normal neonates.[11] The prevalence variation is attributed to the variation in the selection criteria and to the methods used for hearing evaluation. It is logical to assume that improvement of obstetric and neonatal care will diminish the incidence of such hearing impairment.

The single most common postnatal etiological cause of SNHL in these children was meningitis (8.9%). This is consistent with most published studies. Meningitis accounts for between 3% and 10% of severe to profound SNHL in the developed countries, and for up to 30% in the equatorial zone of Africa.[4] In a literature review, Nodal noted that 5% to 35% of survivors of bacterial meningitis were reported to develop SNHL.[12] He stated that the higher figure may be closer to the actual incidence because of the difficulties in detecting partial and temporary hearing loss. Early diagnosis and treatment of meningitis is not enough to diminish the frequency of postmeningitis hearing loss. Prevention of meningitis (by vaccination) is the only means of reducing this complication.

Hearing impairment is the single most common abnormality resulting from congenitally acquired rubella.[5] In the western world in nonepidemic years, less than 7% of severe-profound childhood SNHL is attributed to congenital rubella,[13] but up to 60% in epidemic years.[14] In this study, rubella was found to account for only 2.4% of childhood SNHL. This low prevalence may reflect a high level of natural immunity, or may be due to failure to recognize the disease because the diagnosis was based largely on the history. The presence of other stigmata of fetal rubella may supplement the diagnosis, e.g., the highly specific retinal changes.[3] The introduction of vaccination programs in several countries has reduced the number of hearing disabled children.[7]

Four cases (2.4%) of SNHL in this survey were attributed to mumps. The hearing losses were unilateral and severe-profound in all cases. Mumps remains a significant cause of acquired unilateral deafness in countries where routine mumps vaccination is not practiced. Deafness occurs in approximately 1/20,000 cases.[15]

In this survey, causative factors for SNHL could not be found in 8.9% of the cases. In several reported series of SNHL in children, approximately 40% are placed in a category of unknown cause.[5] Retrospective diagnosis of some causes of deafness cannot be made from the history, clinical examination and laboratory investigation. For example, elevated CMV antibodies may represent merely reactivation of acquired infection, which does not adversely affect the fetus. The etiological diagnostic evaluation of SNHL is a dynamic process. A systemic follow-up utilizing audiological and nonaudiological test procedures may achieve a reduction in the number of unknown etiologies.

In conclusion, this study demonstrates that childhood SNHL is a major health problem in this region. The greatest advance in the treatment of this disorder will be its prevention, e.g., by educating the public regarding the hazards of consanguinity, immunization against meningitis and rubella, and appropriate perinatal care.

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syphilis, cytomegalovirus and herpes simplex) information was obtained.

Results

A total of 6421 children were recruited; 55% males and 45% females. The age distribution is show II in Table I.

On the basis of the history and clinical examination at home, 1256 children were considered to he at risk of hearing impairment. Auditory brain stem response was performed on 879 and pure tone audiometry on 377 children.

The number of children found to have 20 dB or more SNHL was 168 (2.6% of the total surveyed children). The male/female ratio of the hearing impaired was I .2: I. Of these hearing Impaired children, 18 had unilateral SNHL. and 155 had bilateral SNHL. The degree of the bilateral SNHL is shown in Table 2.

An etiology of the hearing impairment could hc proposed in 91.14 of the children (143/168). Table 3 shows the distribution of the SNHL cases to rhe etiological factors. The diagnosis of hereditary hearing impairment was based on positive history of deafness in a parental sibling of direct parental ancestors. regardless of the parental hearing status. Children were placed in the category of perinatal complications if the birth weight was less than 2500 grams or if there was any history which indicated that the condition at birth required resuscitation. Hearing impairment was considered postmeningitis if the disease had been diagnosed and treated at a hospital or a health center. The diagnosis of the four cases of rubella was based on a history of rash and lymphadenopathy during early pregnancy. I n three cases. ophthalmological examination showed the highly specific rubella retinopathy, i.e., fine or coarse salt and pepper appearance located in the macular area or fundus periphery.[3] Rubella antibodies were demonstrated in all cases. The four cases of postmumps deafness gave a history of the disease followed by unilateral hearing loss. Two children w ith Down syndrome had the specific features and the chromosomal aberration.

TABLE 1. *The age distribution of the 6421 children who entered the study.*

Age	Number	Percent
6-12 months	1004	15.6
1-2 years	851	13.2
2-3 years	683	10.6
3-4 years	676	10.5
4-5 years	638	9.9
5-6 years	651	10.1
6-7 years	374	5.8
7-8 years	347	5.4
8-12 years	1178	18.3
No data	19	0.3
Total	6421	100

TABLE 2. *Degree of bilateral SNHL.*

Degree	Number	Proportional prevalence (%)	Overall prevalence (%)
Mild (20-40 dB HL)	96	61.9	1.5
Moderate (40-70 dB HL)	32	20.6	0.5
Severe (70-90 dB HL)	18	11.6	0.3
Profound (90-105 dB HL)	9	5.8	0.1
Total	155	100	2.4

TABLE 3. *Causes of sensorineural hearing impairment among 6421 Saudi children.*

Cause	Number	Proportional prevalence (%)	Overall prevalence (%)
Hereditary	111	66.1	1.73
Perinatal complication	17	10.1	0.26
Meningitis	15	8.9	0.23
Rubella	4	2.4	0.06
Mumps	4	2.4	0.06
Nonhereditary syndromes	2	1.2	0.03
Unknown	15	8.9	0.23
Total	168	100	2.60

Discussion

The overall prevalence of SNHL was 2.6%. The prevalence of bilateral SNHL higher than 40 dB in the better ear was 0.9%. and the prevalence of were-profound bilateral SNHL was 0.4%. These prevalences are much higher than figure\ giv en by most researchers. Davidson et al. reviewed IO studies from 14 countries and found a prevalence range of 0.56 to 2.3/1000 for hearing loss greater than 40 dB bilaterally in the w estern w orld.[4] They also found that prev alence for the less dev eloped countries tends to he slightly higher. nith figurer, between 2.0 and 4.2/1 000.

Regional variations of SNHL prevalence are expected because of the effect of environment and the level of medical care. However. inconsistency in data collection and analysis technique play an important role in these variations. For example. there is no internationally accepted definition as to what constitutes a "significant" hearing loss. As a result. hearing levels are chosen arbitrarily by the researchers. Almost every possible level has been used. f r o m 20 dB unilaterally t o 90 dB bilaterally.[5]

Another feature which complicates valid interpretation of results is the lack of standardization for methods used for hearing evaluation. The "best" estimate of true hearing

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