

DOCUMENT RESUME

ED 021 384

EC 002 719

DIAGNOSIS OF HEARING LOSS.

Alexander Graham Bell Association for the Deaf, Inc., Washington, D.C.; World Federation of the Deaf, Rome (Italy).

Pub Date 67

Note-47p.; Selected papers from the Congress of the World Federation of the Deaf (5th, Warsaw, August 13-17, 1967).

EDRS Price MF-\$0.25 HC-\$1.96

Descriptors- AUDIOMETRIC TESTS, AUDITION (PHYSIOLOGY), AUDITORY EVALUATION, \*AURALLY HANDICAPPED, CHILDREN, CONFERENCE REPORTS, DEAF, ETIOLOGY, \*EXCEPTIONAL CHILD EDUCATION, HARD OF HEARING, HEARING LOSS, \*IDENTIFICATION, INFANTS, PRESCHOOL CHILDREN, RESEARCH PROJECTS

Identifiers- Waardenburg Syndrome, World Federation of the Deaf

Seven conference papers from the U.S.S.R., India, Poland, Czechoslovakia, and Yugoslavia consider the diagnosis of hearing loss. They are "Examination of Hearing of Children, Aged from 2 to 5, by Means of Playing Audiometry" by A. P. Kossacheva, "A Study of the Etiology and Pattern of Deafness in a School for the Deaf in Madras, South India" by Y. P. Kapur, "Observations on Early Discovery of Deafness" by A. Jasienska and B. Dwornicka, "Diagnosing of Deafness in Czechoslovakia" by F. Brohm, "Selective Examination Methods of Hearing in Infants" by L. Sobieszczanska-Radoszewska and D. Borkowska-Gaertig, "Some Characteristics of Waardenburg Syndrome in Defective-Hearing and Normal Children" by H. Siedlanowska-Brzosko, and "Results of Audiological Examinations of the Deaf in Belgrade" by M. Simonovic. (JD)

U.S. DEPARTMENT OF HEALTH, EDUCATION & WELFARE  
OFFICE OF EDUCATION

THIS DOCUMENT HAS BEEN REPRODUCED EXACTLY AS RECEIVED FROM THE  
PERSON OR ORGANIZATION ORIGINATING IT. POINTS OF VIEW OR OPINIONS  
STATED DO NOT NECESSARILY REPRESENT OFFICIAL OFFICE OF EDUCATION  
POSITION OR POLICY.

Diagnosis of Hearing Loss

Selected Papers from the  
Fifth Congress of the World Federation of the Deaf  
Warsaw 1967

compiled by  
Alexander Graham Bell Association  
for the Deaf  
1537 35th Street, N. W.  
Washington, D. C. 20007

EC002 719

ED021384

## Preface

Diagnosis of Hearing Loss is a collection of 7 papers selected from those presented at the Fifth Congress of the World Federation of the Deaf, Warsaw, 1967. These papers were collected and compiled by the Alexander Graham Bell Association for the Deaf, Washington, D. C. Other collections of papers from the Congress have been compiled and are available from the ERIC Document Reproduction Service. Other collections announced in this issue of Research in Education may be found by consulting the Institution Index under World Federation of the Deaf or the Subject Index under Aurally Handicapped. Titles of these other collections are:

Communication Methods for the Hearing Impaired  
Cultural Activities for the Deaf  
Education for the Hearing Impaired (Auditorily Impaired)  
Psychology of Deafness  
Rehabilitation of Hearing  
Sociological Aspects of Deafness  
Training and Qualifications (Teachers and Workers for the Deaf)  
The Very Young Hearing-Impaired Child

## Table of Contents

Examination of Hearing of Children, Aged from 2 to 5, by Means of  
Playing Audiometry. A. P. Kossacheva

A Study of the Etiology and Pattern of Deafness in a School for the  
Deaf in Madras, South India. Y. P. Kapur

Observations on Early Discovery of Deafness. Anna Jasińska, Barbara  
Dwornicka.

Diagnosing of Deafness in Czechoslovakia. František Brohm

Selective Examination Methods of Hearing in Infants. Lucja Sobieszczanska-  
Radoszewska, Danuta Borkowska-Gaertig.

Some Characteristics of Waardenburg Syndrome in Defective-Hearing and  
Normal Children. Hanna Siedlanowska-Brzosko.

Results of Audiological Examinations of the Deaf in Belgrade. Miodrag Simonović



A.P. KOSSACHEVA, Cand. Med. Sc.

Med. Aud. - 10

Scientific Research Institute of Ear,  
Throat and Nose of the Health Ministry

Moscow - USSR

EXAMINATION OF HEARING OF CHILDREN,  
AGED FROM 2 to 5, BY MEANS OF  
PLAYING AUDIOMETRY

/The paper/

Examination of hearing of children aged up to 4 presents great difficulties, examination of 2 to 3 year old children being the most difficult task. Rather numerous and diverse data available in the appropriate printed matter make it possible to deem that auditory acuity of children of early age can be determined with objective methods based on unconditioned reflex actions or on corresponding acquired conditioned reflexes /N.I. Krasnogorsky, 1935; Ts. P. Nemanova, 1940; V. S. Dashkovskaya, 1953; N. I. Kassatkin, N. S. Myrzoiants, A. P. Khokhitova, 1953; L. V. Neiman, 1960, and many others/.

Various physiological tests are used to objectively register hearing of children of the said age. The most widely used are plethysmography /O. S. Vinogradova and E. N. Sokolova, 1955; A. K. Chargueishvili and T. L. Tokadze, 1955; Kottmeyr, 1964; and others/; skin - galvanic reaction /Mespitiol et Gougerot, 1951; L. A. Bukham, 1957; R. A. Vaigner, 1946; Tamura, 1962; Tanieewski, 1963; Stride, 1963; and others/; pneumography /Ja. S. Tiomkin, 1947; E. K. Siirde; A. K. Iente and K. V. Guerasimova, 1957; and others/; encephalography /G. V. Guershuni, 1955, Appaix A. Avicrinos, 1954; Inaba, 1962, Taylor, 1963; Golstein, Kendall, Arick, 1963; and others/; electrocardiography /R. V. Menunarguia, 1958; Dwornicka, Jasińska, Norska, Smolarz, 1964; and others/; electrooculomotography /Inui, 1964/.

Each of the above methods has its advantages and its drawbacks. We consider it more correct and promising to determine the auditory function of children on the basis of the results of several objective methods taken together /G.V.Guezshuni, 1957; A.E.Louts, 1957; L.V.Neiman, 1960; Facchini G., Sul senti G., 1932; and others/. Without speaking of the peculiarities of the methods of objective examination of hearing, we must emphasize the fact that all these methods are rather complicated and require special equipment and certain skill of the examiner. In view of the above, wide utilization of such methods in general practice encounters big difficulties. We can state at present that the majority of the suggested methods of objective registration of hearing in children still remain the property of scientific institutions, while general practice has been still demanding simple and reliable at the same time methods of examination which could be applied by any doctor in his day-to-day practice.

Playing audiometry has a peculiar place among such methods. Its various modifications are rather widely used abroad. For example, Dix and Hollpike /1947/ have suggested to demonstrate separate pictures; Walrop /1953/ - puppet-show; Lessak /1963/ - a small house-audiometer; Blesolskii /1962/ - demonstration of toys; Metrinovich-Modzhievskaya /1965/ - filmscope. Playing audiometry was first suggested in the Soviet Union by L.V.Neiman and V.P. Lubovskii /1954/. L.V.Neiman has minutely developed the method of playing audiometry and assessed its scientific value /1960/. The method of playing audiometry should be further developed with a view of studying the possibilities of its utilization when examining hearing of children of an early age, and special emphasis must be laid on the possibility of its application not only in special institutions but in polyclinics as well.

The concrete aim of our work was to develop such a modification of the method of playing audiometry which

could be used for determining hearing of children aged from 2 to 3 and could be applied in usual hospital and outpatient practice. The method used consisted in registering hearing of children on the basis of conditioned motivational reaction corroborated with various electrical toys. The toys were chosen in advance taking into account interest shown by children of certain age. Examinations were carried out in a specially equipped, big and light room with pictures on the walls to attract children's attention, with gaudy window-curtains, furnished with special furniture for children. The surroundings made for the cheerful mood of a child and helped to establish the necessary contact with him. To carry out the examination proper children were put in a special soundmuffling chamber and due regard was given to the necessity of concentrating child's attention on the objects of examination. The apparatuses used included a semiautomatic audiometer, model AA-OI, an "Elza" sound audiometer and a speech audiometer "Melodia". In all cases auditory thresholds were determined by air and bone sound conduction and special telephones were used to this effect. All the apparatuses were installed outside the chamber. Child's behavior was observed through a special window. However, in certain cases the presence of the person carrying out examination or of his laboratory assistant in the chamber is indispensable.

Having studied the child's medical record and his disease and having established a contact with him, the examiners passed to tentatively determine his hearing with the help of sounding toys /reed-pipes, accordions, guitars, etc./, their sounds being preliminarily measured in respect of frequency and intensity. Such preliminary acquaintance permitted to choose a tone perceived more easily by children hard of hearing and, this being done, to pass on to playing audiometry.

At the moment of passing of an acoustic stimulation through an air or bone telephone the patient pressed the button wired with the electrical toy. This procedure was

first demonstrated by the doctor /or the laboratorian/ himself using gestures, mimicry or together with a child already acquainted with the method of examination. Such demonstrations are most convincing and easy to understand. The patient begins to imitate. When his response to a certain sound was invariable, they started to practise his motor reaction to a sound of a different frequency. This combination of sounds with an entertaining game made it possible to reach an increased alertness of a child who get interested in the procedure. During subsequent visits the child willingly performed the assignments himself showing exactly the same data as those of the previous days.

Usually the motor - conditioned reaction remained stable steady and it is only after a prolonged interval that it began to extinct. Accoustic stimulation was passed from a tone heard to a tone unheard, i.e. from a more intense tone to a less intense tone. The sounding was gradually diminished by not more than 5 db. Such arrangement of the examination procedure, although it does not meet the requirements of audiometry with adults, is indispensable when examining hard-of-hearing children and especially deaf-mute children. In order to avoid any tiredness of the children, examinations of left and right ears were made in turns and necessary intervals between successive accoustic stimulations were observed so as not to learn the rhythm. Each accoustic stimulation lasted 2 - 3 seconds. In order to excite children's curiosity in the procedure the electrical toys were changed from time to time. As a rule it took 2 - 3 days to get acquainted in general with a child and to establish a contact with him, including preliminary examination with the help of sounding toys and practising the conditioned motor reaction.

It took usually 10 to 20 repetitions to master the reaction as required for hearing determination.

Apart from individual examination we have used the so-called "group" method with which a group of 3 or 4 children of the same age was seated opposite the child being examined

who had already acquired the necessary habits. Such arrangement facilitated considerably the task of preliminary training of each child in the group. With this method it was already during the first day that the child comprehended and performed the assignement given to him, at first by way of imitating with a certain amount of uncertainty and the efficiently reacting to sound with a high degree of continuity.

Four to eight combinations were sufficient to get such reactions with the "group" method. However during such examinations it is most important to avoid overstrain and compulsion, otherwise the contact established with the child can be upset and his negative attitude to the examination can deepen.

In all 280 children were examined by us. Observations were carried out in the surdologopedical department. With a view to comparing the data received with the playing audiometry with those of the usual threshold audiometry, parallel examinations of 80 children were carried out, the age of the children examined making it possible to apply both usual audiometry and playing audiometry. The curves of auditory sensitivity based on the results of the two methods were characterised by the values which were so close that it was possible to say that the results of the two methods were identical. The remaining 200 children were examined only with the help of playing audiometry. Of 2 hundred children examined 63 children aged from 2 to 3, 65 - from 3 to 4 and 72 - from 4 to 5.

There were 110 boys and 90 girls.

The children were sent to the clinic with a diagnosis of deafmutism or grave hardness - of-hearing. 186 children had not any verbal contact. Only 14 children spoke but they were hard of hearing as a result of a neuritis of auditory nerves.

As it is known, absence of hearing or reduction of hearing of some children may be caused by psychoneuropathological and other disorders. Thus to get more proper idea

about the condition of hearing we have carried out complex examination of the children with the help of otolaryngologist, pediatrician, logoped, and psychoneurologist.

As a result of examination it was found the basis of the disorders in hearing were the following factors: 27 children - defects of uterine development;  
18 children - premature birth;  
30 children - child birth traumatism.

Besides different diseases children have suffered at the early age from serious infections, measles, mumps, grippe, meningitis, middle otitis - 69 children. 8 children have disorder of hearing because of hemolytic jaundice; 23 have lost the hearing as a result of the use of streptomycin in a great quantity. As to 20 children we failed to find out the reason of deafness; 3 children had hereditary deafness; investigations carried out according to the methods described allowed us to find out that two children who were previously considered to be deaf, have normal hearing throughout the whole range of frequencies.

As a result of complex examination it was found out that these children suffered from alalia, their age: one was of 2 years and 6 months, another was 3 years old. The rest 198 children sent with diagnosis of deafness as the examination shown have suffered from deafness of different degree: 40 - children - deafness of the second degree; 51 - deafness of the third degree, as to 107 remains of the auditory function were only found.

Thus, the method of playing audiometry used by us without special complicated apparatuses allows to define with a sufficient degree of accuracy the condition of hearing of children of 2-4 years old.

The application of this method gives an opportunity to find out that children who were previously considered deaf, have the hearing. Timely definition of the deafness degree gives an opportunity to begin the teaching of children in special children's creche and kindergartens, to get normal general development and to prepare the child for school.



Y.P.KAPUR, M.D.

Med.Aud-18

Professor of Otolaryngology and  
Director of Deafness Research Project  
and Hearing and Speech Center  
Christian Medical College and Hospital  
Vellore - 1 - SOUTH INDIA

A STUDY OF THE ETIOLOGY AND PATTERN OF DEAFNESS  
IN A SCHOOL FOR THE DEAF IN MADRAS, SOUTH INDIA  
/The paper/

A survey of the children admitted to the C.S.I. School for the Deaf in Madras City, South India was carried out with the following objectives in view:

- 1/ There are about 58 Schools for the Deaf in India. These schools do not have adequate facilities for evaluating hearing loss in the children who are brought to the schools for admission. Assessment is made on the basis of the parents' history of severe hearing loss and the lack of development of speech. It has been suspected by a number of observers that there are many children in the schools for the deaf in India who did not have a hearing loss severe enough to warrant their admission to such a school. This survey was to investigate if this a valid suspicion.
- 2/ The second objective of this study was to determine the types of hearing loss among children in the School for the Deaf.  
Van Egmond quotes Dahlberg<sup>1</sup> as stating that the number of cases of congenital deafness by heredity comprises about half to one third of all the deaf, recessive deafness strongly predominating.  
Lindenov<sup>2</sup> studying the deaf-mute population of Denmark, concluded that 45.5% of cases of all congenital deafness had hereditary deafness.

Fraser<sup>3</sup> in a preliminary statistical segregation analysis showed that 40-45% of children with severe hearing loss suffered from recessive deafness.

Konigsmark and McKusick<sup>4</sup> have, in their recent survey, observed that one-third of the students in schools for the deaf, in three states in the United States of America, were hereditarily deaf.

This study was to determine the number of children who were hereditarily deaf, the mode of inheritance and if there were other abnormalities associated with the deafness. In those cases who were suspected to have acquired deafness the etiological factors responsible for causing deafness were investigated.

- 3/ Another objective of this study was to study the role of consanguinity in producing deafness. There is a very high incidence of consanguinity in this region.

Centerwall and Savarinathan<sup>5</sup> have found that one in three of the marriages in this region are consanguineous marriages.

Hopkins, Guilder and Macklin<sup>6</sup> of the Clark School for the Deaf in America have found cousin marriages among parents of the deaf children to be more common than the population in general. The incidence of cousin marriages is 8% in these families whereas it is only 0.2% in population at large.

De Wilde<sup>7</sup> stated that 7% of deaf children who are deaf-mute originate from consanguineous marriages.

Johnsen<sup>8</sup> found severe high tone sensorineural deafness in 8.3% of the sibships having deaf children resulting from consanguineous marriages compared with the consanguineous marriages rate of 2.1% in Denmark.

Stevenson and Cheeseman<sup>9</sup> suggested that the rate of consanguineous marriages in Northern Ireland is between 0.5 and 3.3% and if limited to first cousins it was 0.1 and 0.4%. They felt that there is no doubt that the matings

producing deaf-born offspring included an undue proportion of consanguineous matings. In 309 matings of persons who were not born deaf but who had born deaf offspring 11.7% were consanguineous and the proportion for first cousin matings only was 6.1%.

Thus consanguinity is known to play a role in causing severe hearing loss. One of the objectives of the study was to determine the consanguinity rate among parents of children admitted to the school for the deaf.

#### PROCEDURE:

The C.S.I. School for the Deaf was chosen for this study because the children admitted to this school adequately represent the socioeconomic background of the population in the region.

The total enlistment of the school at the time of our study was 158.

Protocols were designed to record the information to be obtained. Special emphasis was placed on the prenatal and postnatal history, family history of hearing loss, hearing loss in siblings, consanguinity, milestones, childhood illnesses, age at which hearing loss was first noticed and any other associated abnormalities. Histories were recorded by personal interviews with the parents or near relatives who were familiar with the child's background. Only those children whose parents or nearest relatives familiar with the child's background could be interviewed, were considered for the study. In 29 children out of the 158 on the roll this interview could not be carried out as parents and relatives lived at great distances and were not available for personal interviews. These children were excluded from the study. Thus a total of 129 children were admitted to the study.

The children examined were between 5 years and 19 years.

A complete general physical examination was carried out to determine if there were any other congenital abnormalities. A complete otolaryngological examination was carried

out followed by individual puretone audiograms.

Selected families who had more than one deaf member in the family had as complete pedigree as possible made.

FINDINGS:-

The findings are shown in Table I. The children were divided into three categories as below following the scheme by Van Egmond and Danish:

- 1/ Acquired Deafness
- 2/ Congenital Deafness /non-hereditary/
- 3/ Congenital Deafness /hereditary/

The criteria used for classifying children into these categories are given below:

- 1/ ACQUIRED DEAFNESS: There were 22 children with acquired deafness out of the 129 children admitted to the study /17.2%/. These children did not have hearing loss at birth and according to the parents had evidence of hearing and speech development before the onset of the illness in question. Parents have a tendency to blame a particular illness for causing the hearing loss in their children when such a loss may have already been in existence. For this reason emphasis was placed on the age at which the illness occurred and the duration of the hearing loss. In none of these cases was the hearing loss known to be present at birth. The age at which the illness occurred was also considered important for if these diseases occurred at an age when a existing deafness may be difficult to recognise there could be doubt on such a deafness being an acquired type. The illnesses thought to be responsible for causing hearing loss in these children with acquired deafness are given in Table II.

The majority of the diseases thought to be responsible for causing acquired deafness occurred after the first year of birth /17 out of the 22 cases/. This is considered important as the parents were able

to notice hearing and speech development in the child before the onset of the illness.

2/ CONGENITAL NON-HEREDITARY DEAFNESS: There were six cases in this group /4.7%. In these cases there was a prenatal factor which could have been responsible for causing hearing loss.

3/ The majority of the cases fitted the category of Hereditary congenital deafness. 100 cases out of the 129 cases admitted to the study /77.3%. In this group the majority of the children had no evidence of hearing or speech development at any time and their histories did not indicate any prenatal or neonatal factors which may have caused the deafness.

In a number of these cases there were sporadic cases of deafness in the families and some children in this group had other siblings with deafness since birth.

Each group of cases will be discussed under their headings.

The audiograms obtained according to their characteristics fell into five groups - conductive group, mixed hearing loss group, gentle sloping group, residual group and a group in which there was no hearing. Slides showing the typical audiograms will be shown.

Fisch<sup>10</sup> in the discussion on congenital deafness audiometric patterns has attempted to associate different types of audiometric patterns with certain types of pathological lesions. However, he has pointed out that there are still a number of other unknown factors and variables to be considered as many audiograms do not fit any pattern.

Fraser<sup>11</sup> mentioned that audiograms cannot provide conclusive evidence of the etiology of deafness in the individual case. He is of the opinion that if no response at all takes place at the maximum intensities of the audiometer over the whole frequency range the deafness is likely to have been acquired.

Our findings indicate there were no characteristic audiometric patterns among the deaf children in the three categories.

DISCUSSION:

ACQUIRED DEAFNESS: The diseases thought to be responsible for causing deafness in the acquired deafness are shown in Table II. It is of interest to note that measles was responsible for causing deafness in six cases /27.3%/ of cases with acquired deafness. Measles is known to cause otitis media in a large number of cases /Krugman and Word <sup>12</sup>/. However, Danish et al<sup>13</sup> found measles to be responsible for causing hearing loss in 12 out of the 145 cases with acquired deafness.

Bordley<sup>14</sup> has pointed out that measles is responsible for causing hearing loss in a large number of children and this should be investigated further. He stated that of all the diseases that are contracted in infancy measles offers the greatest threat to hearing. In a review of 485 children with hearing loss measles was thought to be responsible for 48 cases. Thus measles appears to be illness which does offer a serious threat to hearing.

There were five cases /22.7%/ in which there was a history of very high fever of a few days duration followed by a hearing loss. This is common in this region. A large number of children are frequently seen in hospitals with severe hearing loss and with a history of high fever of unknown origin of 3-4 days duration. One case had conductive hearing loss with evidence of bilateral serous effusion.

Audiograms: There was no characteristic audiogram pattern. Slides of the audiograms will be shown.

CONGENITAL NON-HEREDITARY DEAFNESS: Toxaemia of pregnancy, attempted abortion, prematurity, congenital syphilis and severe shock during pregnancy were considered the etiological factors among the seven cases with congenital non-hereditary deafness.

In one case of attempted abortion drugs were used by the mother. However, the mother was not able to tell

us the nature of the drugs used. In another case the mother had severe shock during pregnancy. In three cases mothers had severe toxæmia during pregnancy.

Audiograms: There was no characteristic audiometric pattern in this group. Slides of the audiograms will be shown.

CONGENITAL HEREDITARY DEAFNESS: The majority of the cases appeared to have congenital hereditary deafness - 100 out of the 129 cases /77.3%/.

This is a very high figure compared with the figures in the United States<sup>4</sup>, Holland<sup>1</sup>, Denmark<sup>2</sup>, and United Kingdom<sup>3</sup> which vary between 33% to 45%.

The most common form of transmission of hereditary deafness is on a recessive basis. In such situation, each of the parents has contributed an identical defective gene to their offspring. Since the genetic make up of relatives is more likely to be similar, the frequency of recessive deafness would be higher among offsprings of related parents.

In South India normally about one out of every three marriages is between relatives<sup>5</sup>. In the present study it was found that out of the 100 children with hereditary deafness 56 children or 56% were born of consanguineous parents /Table III/. Thus the consanguinity rate among parents of these children was very high which is very significant.

Fraser<sup>3</sup> has found that in his study the large group of recessive hereditary deafness includes familial cases and isolated cases where consanguinity of parents make it likely that recessive inheritance is involved.

Pedigree charts were made on 35 families which covered as many as 66 children in this group. Analysis of these pedigree charts showed that of the 66 children with hereditary deafness as many as 63 of them inherited the deafness by recessive transmission. In 23 out of the 35 of the families examined there was consanguinity among parents. In only three cases there was possible dominant inheritance.

Thus in 95.4% of cases in this group /66 children/ were suspected to have had the homozygosity for a recessive gene, while only 4.6% of cases belonged to dominant type. This again confirms that hereditary deafness is of recessive in character in the majority of the cases.

There was no case of sex linked deafness. In two cases there was evidence of congenital thyroglossal cyst and in two others there was evidence of congenital heart diseases. There were no other abnormalities in other organs nor were there any other syndromes present.

There were six cases in this group who had moderately severe bilateral sensorineural deafness. One case had a conductive hearing loss with normal tympanic membranes. This appeared to be a bilateral otosclerosis case.

Audiograms: A study of the pattern of audiograms in this group of children showed, that except in the case with otosclerosis, no characteristic pattern. The majority had either a gentle sloping curve or evidence of residual hearing at 250-500 cycles per second.

REFERENCES:

- 1/ Dahlberg: Quoted by A.A.J. Van Egmond, Congenital Deafness, Journal of Laryngology and Otology, 68:429-443, 1954.
- 2/ Lindenov, H.: The Etiology of Deaf-Mutism, Copenhagen, 1945.
- 3/ Fraser, G.R.: A Study of causes of deafness amongst 2,355 children in Special Schools, Research in Deafness in Children Blackwell Scientific Publications, page 13, 1964.
- 4/ Konigsmark, Bruce W. and McKusick, Victor A.: Hereditary Deafness, The Volta Bureau, 1966.
- 5/ Savarinathan, Gemma; Sundar Rao, P.S.S.; and Centerwall, W.R.: The Relationship of Consanguinity to the Occurrence of Cleft Lip and Palate in South India - unpublished report.
- 6/ Hopkins, L.A. a.o. Laryngoscope, 56:570, 1946.
- 7/ De Wilde, P.A.: Ned. T. Geneesk, 61, 225, 1910.
- 8/ Johnsen, S.: Heredity of perspective deafness, Acta Otolaryngologica, Stockholm, 42:539, 1952.
- 9/ Stevenson, A.C. and Cheeseman, E.A.: Hereditary Deaf Mutism, with particular reference to Northern Ireland, Annals of Human Genetics, 20:3:177-231, 1956.
- 10/ Fisch, L.: The Aetiology of Congenital Deafness and Audiometric Patterns, Journal of Laryngology and Otology, 69:479:1955.
- 11/ Fraser, G.R.: Profound Childhood Deafness, Journal of Medical Genetics, I:118-151:1964.
- 12/ Krugman, S. and Ward, R.: Infectious Diseases of Children: Measles, The C.V. Mosby Company, St.Louis, 1960.
- 13/ Danish, Jacob M.: Tillson, J.Karetas; and Levitan, Max: Multiple Anomalies in congenitally deaf children, Eugenics Quarterly, 10, 12-21, 1963.
- 14/ Bordley, John E.: The Problem of the Preschool Deaf Child, Laryngoscope, 62:514-520, 1952.

TABLE I

CLASSIFICATION<sup>‡</sup> OF DEAFNESS AMONG 129 CHILDREN IN THE  
C.S.I. SCHOOL FOR THE DEAF, MADRAS

Type of Deafness	Number	% of children examined
1. Acquired Deafness	22	17.2
2. Congenital non-hereditary deafness	6	4.7
3. Congenital hereditary deafness	100	77.3
4. Congenital or Hereditary deafness	1	0.8
Total	129	100.0

<sup>‡</sup> As explained in text.

TABLE II

REPORTED CAUSES IN 22 CASES OF ACQUIRED DEAFNESS

Reported etiology	Number	
	Age at detection of deafness 1st year	After 1st year
Measles	-	6
Typhoid	1	-
Chicken pox	-	1
Whooping cough	-	1
High fever /unknown origin/	1	4
Small pox	1	-
Dysentery	-	1
Ear infection	1	-
Head injury	-	1
High fever /	1	-
Dysentery /		
Whooping cough /		
High fever /	-	1
Whooping cough /		
Measles /		
Diarrhoea /		
Measles /	-	1
Small pox /		
Whooping cough /		
Whooping cough /	-	1
High fever /		
Total	5	17

TABLE III

CONSAINGUINITY RATES AND RELATIONSHIP BETWEEN THE TYPES OF DEAFNESS

	Types of Deafness		
	Acquired	Congenital non-hereditary	Congenital hereditary or hereditary
With Consanguinity	5	---	56
With no consanguinity	17	6	44
Total	22	6	100
Consanguinity rate	22.7%	nil	56.0%
			1
			nil



**Anna JASIENSKA, Barbara DWORNICKA**

**Medical Academy of Silesia  
Otolaryngological Clinic**

**Zabrze - Poland**

**Med.Aud. - 7**

**OBSERVATIONS ON EARLY DISCOVERY OF DEAFNESS**

**/The paper/**

For the last few years in the world literature are more frequently published reports concerning the need of early detection of hearing impairment and deafness in children. This is in a strict relation with the necessity of early rehabilitation.

In 1963 at the II Congress of Children's Laryngology Prof. Międoński underlining the significance of audition for the child's development cited Sutermeister's sentence: "I venture to assure, that no other infirmity induces such important effects as deaf-mutism. No other bodily failure causes such moral handicap".

Appreciating the social meaning of the problem, we took into consideration the question of audition investigation in the earliest childhood. Our interest in early discovery of deafness dates from 1962, when we presented at the II Congress of Children's Laryngology in Warsaw the report: "Attempt to determine the foetus reaction to sound stimuli". The present work brings the results of studies performed on 31 pregnant women in the last month of

pregnancy. Our researches were based on the electrocardiographic method for the first time used in Poland, to define the reaction of the foetus puls rate to sound stimuli. The mother's and the foetus heart-rate was continuum registered by the 3-leads phonocardiograph of Hellige with direct tracing. Simultaneously sound was emitted with the Peters' audiometer SPD-2 through an air-conducting earphone, with a rubber cuff, applied on the mother's symphysis pubis above the child's head. Frequencies of 1000 c/s and 2000 c/s of 100 db-intensity were emitted during 5 sec. The foetus heart-rate was registered before-during-and after the sound emission, until the restitution of the primary heart-rate. The heart-rate of the foetus often rose or decreased significantly already during the sound emission or just after the emission. Frequencies of 1000 c/s gave an average acceleration of heart rate of 7 heart-beats per minute /5 to 13/. With 2000 c/s a greater acceleration was noted - in average 11 heart beats per minute /4-20/. Simultaneous registration of the mother's heart-rate permitted to state that she showed no emotional reaction related with the sound and tactile stimuli, which would be manifested by heart-rate acceleration. Therefore it may be admitted, with great probability, that the stimuli transmitted by the mother don't influence the foetus puls acceleration reaction. Time relation between puls rate acceleration and sound emission shows an undoubted relationship with the acoustic stimulus activity. "Acta 1964 is of great interest.

The present work has been a preliminary one to follow-up researches in early discovery of deafness in newborns. In further investigations the Ecg technique applied in human foetus has been used for hearing examination in newborns. The results of our investigations have been presented at the Congress of Czechoslovakia's Laryngologists in Praha in 1964 and published in Archives of Otolaryngology 1967: "Attempt in objective examination

of hearing in newborns by EKG". We have examined 36 newborns aged from 1 to 10 days. The heart reaction of the newborns has been observed at sound emission of 500 and 3000 c/s of 90 db intensity, during 5 sec. The heart rate was traced continuum by 1-lead electrocardiogram, like in foetus cases before - during - and after the sound emission. Simultaneously has been observed the newborns general reaction of muscle reflexes. The average change in puls rate following the two sounds was equal and in average showed 11 heart-beats per minute. In one case the puls-rate nor the muscle reaction showed no noticeable changes. The audition investigation method involving the use of ear-cardiac reflex with the aid of EKG method is a very sensitive and objective evaluation of the newborn's audition reaction. This method, however, requires special equipment and a trained staff, which is rather possible in clinical centres.

In 1966 during the III-rd Days of Laryngology in Childhood in our report: "Necessity of performing routine auditory investigations in newborns" we have proposed to undertake auditory mass-examinations in newborns in the whole country. These investigations ought to be performed in all obstetric departments and obstetric centres. This would be a first selection which could permit early discovery of newborns with suspected hearing impairment. These investigations, as mass-examinations ought to be unified, easy to perform and to interpret, so as to be effected even by midwives and nurses. In the Obstetric Clinic of Zabrze since April 1966 hearing investigations have been started in all newborns. The investigations are based on the observation of the newborn's muscle reflexe to spoon and cup rattling and to two musical instruments. From the available percussion instruments we have chosen "cymbals" and "marakesh", as their acoustic spectrum has a preponderance of high frequencies. Many

authors report, that newborns have a better hearing sensitivity for high frequencies. Wessen explains that in the middle ear may be foetal-water and makes the low frequencies conduction difficult. The above described method doesn't necessitate an expensive equipment nor a trained staff and therefore it may be used in hearing routine examinations in newborns.

Involving now the use of EKG response technique we are elaborating the problem of development of auditory reaction in foetus according to the month of pregnancy. This will be a modest contribution to the few works concerning auditory physiology in human foetus.



Prof. František BROHM M.D.  
Brno - CZECHOSLOVAKIA

Med. Aud-2

DIAGNOSING OF DEAFNESS  
IN CZECHOSLOVAKIA

/Summary/

Two facts are known:

- 1/ the rehabilitation of a deaf person leads to successful result only when started early enough
- 2/ For various reasons, the parents of deaf and semi-deaf children go to the doctor too late.

This is why we have started a few years ago preventive examinations of children with the view to detect deafness the soonest possible. The point is to establish an exact diagnosis towards the end of the second year of age and to start rehabilitation on time.

Children undergo periodical preventive examinations. The first symptoms are reported already at the maternity hospital immediately after birth. All children with an established or suspected deafness or partial loss of hearing are referred to an otolaryngologist or a phoniatriest for detailed examination.

Chronological programme of the examination:

1st examination:

The maternity hospital registers all children whose

- a/ story of the family is positive,
- b/ pregnancy was pathological,
- c/ there is incompatibility of the Rh factor in parents,
- d/ childbirth was prolonged or pathological and could result in a lesion of the central nervous system.
- e/ there are visible congenital anomalies of the ear.

2nd examination carried out between the 2nd and 4th month.

3rd examination between the 10th and 12th month.

4th examination at the age of three years.

Apart from these periodical tests, examinations extend to all children who:

a/ suffered of an inflammation of the central nervous system,

b/ after the use of ototoxic antibiotics and eventually other drugs,

c/ after lesions of the head and other diseases capable of causing deafness or partial loss of hearing,

d/ when the parents suspect deafness or partial loss of hearing.

Examinations are carried out at pediatrics centres by one paediatrist or a nurse. These are informational examinations. In case when the child is suspected of deafness, examinations are repeated many times and if they are again negative, a detailed examination is carried out at the otolaryngological or phoniatrist ward. All children with an established diagnosis are registered. Rehabilitation and the use of auditory prosthesis are started immediately.

For the informational examination we use various acoustic toys, the clinking of a spoon against a pot, the sound of a drum or a trumpet. All toys and other objects are acoustically measured and according to the intensity of the sound and its frequency it is possible to get a sufficiently exact image of the hearing ability. The toys form a sort of scale from weak sounds to the strong sounds of the drum and the trumpet. The difference in frequencies makes it possible to assess the degree of hearing loss.

We register the conditioned and unconditioned reflexes, from the cochlea-palpebral reflex to the /Moro/ shock reflex, orientation and investigation reaction up to the execution of verbal command.

On the basis of such data, we are able to:

- 1/ establish the necessary diagnosis and start treatment on time,
- 2/ start on time rehabilitation and apply auditory prosthesis,
- 3/ plan with a relatively high degree of accuracy the number of places necessary in schools and institutes for deaf and semi-deaf children.



Dr Lucja SOBIESZCZANSKA-RADOSZEWSKA  
Dr Danuta BORKOWSKA-GAERTIG  
Mother and Child Institute  
Children Otolaryngology Clinic  
Warsaw, POLAND

Med.Aud.-27

SELECTIVE EXAMINATION METHODS OF HEARING  
IN INFANTS

/A paper/

Analysis of questionnaire polling on deaf and hard-of-hearing children which was published in press in 1963, shows that about 97% deaf cases and about 66% cases of serious hearing disorders originated, or were recognized by parents occurred under 2 years of age. Finding such frequent disorders in infants presents to audiologic service an important task. One of more important problems, according to international pedo-audiologic committees, is conducting selective examinations of hearing in infants.

For the purpose of early recognition of auditory defects in children - selective examination of hearing in all infants 2 months old and later 9 and 10 months old was undertaken in Czechoslovakia within pediatric consultation service./2/. Similar examinations were undertaken in some parts of England, Sweden and the United States.

It is known that by orientational auditory screening of infants it is possible to recognize only if the child hears but detailed estimate of degree and place of disorder of hearing organ is impossible /5, 12, 13/ while reactivity of child to acoustic stimuli depends not only on its state of hearing but also on the degree of its psychomotor development connected with its age /5, 14/.

As infant grows its reaction to loudness of sounds changes and from 4th month of life the reaction begins to be fixed on stimulus of intensity of about 40 - 55 dB, and at about 6 months on stimulus of 30 dB if examination was in silenced interior. Out of different reactions of an infant to sound stimulus passing as it grows, the longest remains reaction of search for the source of sound. This kind of response to stimulus appears sometimes very early and begins to get fixed at about 4th month of life and between 6th and 12th month is considered as steady reaction. Such response can be obtained in children up to 24 months old and even older. It has been found also that depending from infant's development age not only the kind of reaction changes but also frequency of responses to sound stimuli. According to investigations by Miller and de Schweinitz /7/ infants of 3 months of age react in 15% to an acoustic stimulus, of 4 months of age in 44%, of 6 months in about 50%, and between 8 - 10 months of age in 75% cases. Findings quoted above concern development of children reactivity to acoustic stimuli irrespective of sex, race and place of living. However the factor requiring differentiation in hearing examinations in infants is the kind of acoustic stimulus. According to many authors especially Ewing /1958/ /4/ an infant differently from grown-ups must be tested with sounds known to it from its surrounding in order to incite its interest with known situation and related with sound and to cause impulsive seeking for the source of the sound. Acoustic stimulus ought to be changed with infants growth and fitting to its age. Therefore during infant age rattles, squeaking or clattering toys ought to be used as well as striking with spoon against a cup or plate, rattling with milk bottles used for feeding through nipples, rustle with paper, mother's or woman's voice.

In every country those sounds are included within a range of frequency from 60 to 16000 c/s, they differ how-

ever with colour and melody of sound characteristic to environment what in infancy conditions remembering and recognition of given acoustic signal. All those problems are connected with selective examination of hearing in children with well developed central nervous system and correct hearing.

Children with defective hearing and disorders in registering sounds from surrounding in central nervous system require other methods of examination. In case when only hearing organ is impaired such children may react in a manner proper to age but to louder stimuli. In hard-of-hearing and retardates such reaction appears to increased acoustic stimulus in a manner proper to developmental age and not to one as per birth certificate. In children socially deaf and not registering sounds of everyday life of intensities lower than 70 or 80 dB examination method of reaction by means of known sounds becomes useless because a child living in continuous silence would react to every loud acoustic stimulus which suddenly enters its consciousness. The kind of reaction will depend on the level of development of central nervous system.

#### Own research

In order to discover early auditory defects, 3 kinds of tests were worked out for orientational examination of hearing in infants of 8 to 12 months of age, taking under consideration their state of hearing and general development.

Test adapted to open therapeutics in such way that every physician could apply them and them preliminary screening of hearing might constitute a part of general pediatric examination of every infant in 8 to 12 months of age.

Test 1 permits to select properly hearing children and of psychomotor development regular for the age. This test cannot be passed by children with defective hearing and retarded in development. Evaluation of hearing is based on localization reaction of acoustic stimulus using so called sounds from child's surrounding. For examination such sounds are employed as noise of rapping toys e.g. tambourine, rattles,

striking a spoon against a cup, rustle of tissue paper and woman's voice of colloquial speech pitch. Intensities of particular acoustic stimuli oscillate within the range of 35 - 45 dB. Due to simple and cheap technique of examination this test is suitable for selectional screening in pediatric consulting centres.

Test 2 permits selection of children with slight impairment of hearing or rather considerable retardation of psychometric development. This test cannot be passed by children with serious auditory disorder and very retarded psychomotor development. Evaluation of hearing is based on localizing reaction or on auricular and eyelid impulse applying acoustic stimulus in form of so called sounds from child's surrounding but of greater intensity because within the range of 55 - 70 dB. For this test sounds of drum, whistle, trumpet or woman's voice are applied. The test is to be used in children's otolaryngologic consultation centres.

Test 3 is for children with hard auditory disorders and remarkably retarded psychomotor development. This test cannot be passed by children totally deaf or with impaired auditory reactivity in central nervous system. Evaluation of hearing is based on localization reaction or auricular and eyelid impulse while using acoustic stimuli caused by musical instruments such as drum, castanets, trumpet, maraca and cymbals. Intensity of applied sounds oscillates within the range from 60-105 dB. This test is for otolaryngologic consultation centres.

#### Standardization of Tests

258 children of 8 - 12 months of age were examined. Examinations were carried out at Consultation Centre D<sub>1</sub>, Infant-Home, and at Clinics: Healthy Infant, Otolaryngologic, Surgical and Pediatric of Mother and Child Institute. In all the cases auditory screening and laryngologic examination were carried out. Psychomotor development of every infant was checked by psychologists of the Clinic. Test were standardized for 4 groups of infants specially selected:

- 1/ well hearing and of good general development,
- 2/ hard-of-hearing due to inflammation of internal ears,
- 3/ with retarded development or with disorders of central nervous system,
- 4/ serious auditory defects - considered to be deaf.

Group 1 was composed of 192 children in which localization to acoustic stimuli was obtained within ranges between 30 - 50 dB. Laryngologic examinations did not disclose any inflammatory changes. 55% children in that group slightly retarded psychomotor development was found.

Groups 2 and 3 were composed of 58 children in which localization reaction acoustic stimuli was obtained only at intensities between 50 - 80 dB - and in 10 children it was unilateral. Laryngologic examination disclosed serious nasal infection with inflammation of auditory meatus or purulent inflammation of internal ear. 10 among those children had preponderant unilateral changes in ears. 14 children of the Group had no changes in noses, throats and ears but psychologic examinations disclosed retarded psychomotor development. The Group included also children with disorders of central nervous system which might have been probably connected with the lack of proper reactions to acoustic stimuli.

Group 4 included 8 children in which reaction in form of cochlear-eyelid impuls to acoustic stimulus of intensity about 90 - 100 dB. 3 among these children after frequently repeated examinations have shown serious auditory impairment. Laryngologic examination disclosed no inflammatory changes. 5 of remaining children have shown serious retardations in psychomotor development. These were the children with disorders of central nervous system in which the lack of reactions to acoustic stimuli was connected not with auditory defects but with considerable retardation in psychomotor development.

#### Discussion and Summary

Preparation of adequate tests for conducting selective examinations of hearing in infants in Poland was based

upon results and experiments of authors conducting research on auditory organ of infants for many years /1,3, 6,8,9,10,14,15,16/.

To make selective examinations of hearing cover all the infant population of a country - pediatricians ought to include them in general medical examinations. Taking under consideration large number of examined children and non-specialistic service of pediatricians, test for selectional examinations of hearing in infants were elaborated in a manner that in a best and possibly universal way protect both examined and examining against making errors.

For the above reasons:

- an age group was selected composed of infants from 9 - 10 months old because at that time of life since 3 - 4 months directional impuls of reaction to sound is already fixed;
- toys and everyday objects used in home surrounding were chosen to make the test easy and interesting as their sounds were well known to the children;
- out of selected sounds tones of low and high pitch were considered to form a base of some differentiation of eventual hearing disorder;
- besides the test for properly hearing children and generally well developed 2 additional test were prepared which in an out clinic of a laryngologist would permit examination of a child suspected by pediatricist that its hearing is impaired already by first testing. These tests enable to differentiate hard hearing from grave auditory impairment and deafness;
- taking into consideration influence exercised by psychomotor development in relation to acoustic stimuli it was accepted as positive reaction not only symptoms of localizational reaction but also auriculareyelid impuls;
- standardization of tests was undertaken in average conditions of not silenced out-clinic by means of level pitch audiometer in order that introduced standards were

at best measurable and to have results obtained in future in particular out-clinics all over the country to be comparable.

- scheme of selectional hearing examination in infants was elaborated with consideration of kind of examination, frequency of transmitted sound and its pitch at fixed distance from the source of sound.

Standardizing investigations of tests proved their intended usefulness for children properly hearing, hard-of-hearing and with serious auditory impairment.

Those tests ought to be checked by experiments on selected ground in order to disclose and correct their errors before admitting them into general use in Public Health Service.

LITERATURE:

1. Barr Bongt: Pure tone audiometry for pre-school-children.
2. Brohm F., Supacek I., Tieha H.: Metodika vyso-trovani sluchu kojeneu u malych deti pri depristazi slu-ehovych vad. Ceskoslovenska Pediatric-Praha XVI-6-551-56-1961.
3. Bystrzanowska T.: Audiologia kliniczna PZWL 1963.
4. Ewing J. and Ewing A.: 1944 J.Laryngol.Otol. 59,309.
5. Hardy W.G., Hardy J.B., Brikner Ch.H., Frazier T.M., Dahyherty A.: Auditory screening of infants Annals of Otology, Rhinology and Laryng. Sept.1962, Vol.No.3.
6. Miller June B., de Schweinitz Louise: Is it Deafness? Journal of the Kansas Medical Society, Dec.1962 p.522-524-530 Vol.LXIII, W.XII. -
7. Miller June B., de Schwei-nitz Louise: Indentification audiometry in young children. Preceedings of the Thirty-Fourth Meeting Conference of Executive of American Schools for the Deaf. Austin Texas, April 1.6.1962.
8. Miller June B., de Schweinith Louise, Goetzinger Cornelis P.: How Infants Three, Four and Five Months of Age respond to Sound. Exceptional children Vol. 30, N.4, Dec.1963.
9. Mitrynowicz-Modrzejewska: Fizjolo-gia i patologia glosu, sluchu i mowy PZWL 1963.
10. Peiper Albert: Die Eigenart der Kindlichen Hirntätigkeit 1956, Veb Georg Thieun Leipzig.
11. Robinson Geoffrey C.: Hearing loss in infants and young pre-school children. The Volta Review Vol 66 N 6 June 1964.
12. Ross Labora-tories Columbus 16 Ohio Currents in public health Oct. 1963 Vol.N.9.
13. Seeman M. und Petvik M. - Praha - Klanganalyse und Hörprüfung: Sonderdruck aus Zeitschrift für Phonetic Sprachw,issenschaft und Kommunikationsfors-chung Band 16, 1963. Heftl.3.
14. Scheridan screening tests for young children and retardates. British Medical Journal Saturday October 25-1958.
15. Silverman S.R.: Symposium on disorders of communication Hearing. The Journal of Pediatrics Vol.62 Febr.1963.N.2 -
16. Weden-berd Erik: Objective auditory tests on non-cooperative children. Acta Otolaryngologia Suppl. 175 Stockholm 1963.



Dr Hanna SIEDLANOWSKA-BRZOSKO  
State Institute for Deaf-mutes  
and blind.  
Warszawa - POLAND

Med. Aud. - 20

SOME CHARACTERISTICS OF WAARDENBURG SYNDROME IN DEFECTIVE-  
-HEARING AND NORMAL CHILDREN

/The paper/

In order to detect Waardenburg Syndrome 1142 children with congenital defective-hearing, from Kindergarten-schools and schools for defective-hearing, were examined. In 4 cases all the features of the syndrome were stated and in 18 cases some of the absent signs were stated in other members of the family. 1100 children attending normal schools were examined as a control group. In this group no case of Waardenburg Syndrome was stated, although some of the syndrome's features were noticed.

In 1950 the holland ophtalmologist P. Waardenburg described the syndrome of congenital defects consisting of perceptive lesion, lateral dislocation of the internal eye-corners and of the lacrimal points, enlargement of the nasal base, joint eyebrows, heterochromic irides, white curl over the forehead, defective-hearing - are the characteristic features of this syndrome.

Waardenburg /1951/ reports that this syndrome is observed in 1,43% of cases of deafness in Holland. Di George and Olmsted /1957/, report, in consequence of studies effected in the U.S.A., that the syndrome appears in 1,5% of children with congenital deafness.

According to Waardenburg, the heredity of this syndrome has a dominating autosomic character, with no relationship to the sex, and each feature of the syndrome may penetrate with a variable frequency. In many cases an incomplete syndrome of lesions is stated in one family member, whereas individual defects may appear in other family members.

The syndrome has been described by many authors, they confirmed the principles of transmission of the incomplete syndrome in several family members /Arnvig 1959, Partington 1959, Fisch 1960, Ghost 1962/. The polish medical literature published in 1965 the report of Siedlanowska-Brzosko and Blaim describing this syndrome. To investigate the onset of the syndrome in children's environment with congenital auditory disorders, 1142 children attending special schools and 1100 children from normal schools were examined to obtain a comparable material. The research was effected in 3 groups of age: 3-10 years, 11-16 and 17-18 years. As a principle to estimate the distance between the eye-corners and between the irides, the normal values given by Harley and Waardenburg were taken:

Harley's standard:

age	distance between the internal corners	distance between the external corners	distance between the irides
3-10	22-30	78-80	46-58
10-16	28-34	81-90	55-64
Waardenburg's standard:			Age= over 16 years:
women	24-37	74-92	54-72
men	26-39	77-96	58-75

Table I shows the frequency of Waardenburg Syndrome individual features in children attending special schools /group A/ and normal schools /group B/.

On the basis of investigation results in group A the following frequency of various coexisting signs of the Waardenburg Syndrome was stated.

Individual signs of the syndrome were stated in 77 children. Auditory disorders in all the cases were of perceptive character. 4 children presented all the signs of the syndrome. 13 children presented all the sign except the white curl. In 31 children beyond defective-hearing, only a marked dislocation of the internal eye-corners and of lacrimal points and the enlarged nasal base were stated. 1 child had only the enlarged nasal base, joint eyebrows and the white curl over the forehead and 14 children beyond joint eyebrows had an enlarged nasal base.

Besides in the group A other coexisting congenital lesions were stated in 14 cases.

In 92 cases other incidents of auditory deficiency were stated in the family: 42 of them concerned children with Waardenburg Syndrome-features.

On the basis of the analysis results in group A - the Waardenburg Syndrome was stated in 22 children from 17 families - therefore 1,9% of the examined ones.

Table 2 shows the frequency of individual signs of Waardenburg Syndrome in the mentioned 17 families. It must be underlined that in all the families at least 2 cases of deafness or defective-hearing have been stated.

In the control group B, children attending normal schools, 12 children had a larger distance between the internal eye corners but without dislocation of the lacrimal points and a large nasal base, 1 child had joint eyebrows and a large nasal base, in 2 cases heterochromic irides and in 1 case the white curl over the forehead were stated. Besides in 2 cases a coexisting congenital lesion was stated in the child, and in 5 cases - in other family members. Perceptive auditory deficiency was stated only in 1 case. On the whole these signs, as individual signs, were stated in 16 cases, so that none of them was qualified as Waardenburg Syndrome.

Comparing the group A and B, a visible relationship of Waardenburg Syndrome characteristics to defective-hearing is stated:

Defection of Waardenburg Syndrome by an <sup>exact</sup> examination of children suspected of defective-hearing, will facilitate the diagnosis of auditory deficiency of hereditary character.



V KONGRES ŚWIATOWEJ FEDERACJI GLUCHYCH - WARSZAWA 1967

V Congress of the World Federation of the Deaf - Warsaw 1967

V Congrès de la Fédération Mondiale des Sourds - Varsovie 1967

Miodrag SIMONVIĆ M.D.  
University of Belgrade,  
Medical Faculty  
Clinic of Oto-Rhino-Laryngology  
Belgrade - YUGOSLAVIA

Med.Aud. = 38

RESULTS OF AUDIOLOGICAL EXAMINATIONS OF THE DEAF IN BELGRADE

/The paper/

Deafness, i.e., serious hearing loss, belong among those diseases which, in spite of the progress of medicine, show the greatest rise in the number of cases in most countries. In view of a continuous rise in birth-rate of population in the world, the problem of deafness is to represent in the future one of the most serious concerns of society, since the rehabilitation can be successful only if started in time, and if carried out in a systematic way.

The task of medicine, particularly of audiology, should not be confined to a mere watching of this development, and to the undertaking of the measures for an early identification and rehabilitation of persons with hearing loss, but it should consist also of active and detailed examination of all the causes that lead to the hearing loss, and of the measures for their timely discovery, and for an adequate rehabilitation. In this respect, systematic analyses of the deaf and hard-of-hearing population can be very helpful, since it is very difficult to distinguish and precisely to delimitate these two categories of persons with hearing loss. High technical perfection and the possibilities for a further improvement of the modern hearing aids enable us to introduce through early and systematic rehabilitation into the society even such persons who

have been earlier considered as practically deaf in view of their poor residual hearing. On the other hand, if the cases of a moderate hearing loss were neglected, this could result in a gradual development of a definite and incorrigible habitus of deaf people. That is why the modern audiology distinguishes the cases of practical deafness from those of a serious hearing loss. This difference is rather a result of neglect or of mental incapacity for rehabilitation than of the actual <sup>degree</sup> of hearing loss, so that we should consider it from this angle.

The systematic analysis of the deaf population on various age levels, which has been long carried out, can offer precious data on the periods of the appearance of deafness, and on its causes. In this way, it is possible to undertake the appropriate medical measures in the case of those causes which result in numerous and increasingly frequent hearing losses, in order to prevent them whenever possible. In the same way, the watching and the establishing of the number of cases with hearing loss to be expected in the future will help us in planning the foundation of appropriate medical and social institutions for rehabilitation of persons with impaired hearing. The results of these analyses should indicate in an indirect way the kinds of treatment of many diseases belonging also to other spheres of medicine, as well as the way of life, and work of pregnant women, children and adults, in which may they will influence the hygienic and social conditions of the whole nation.

We consider that in the future preventive measures in the struggle against deafness should be our main concern, since it is only in this way that we could expect the number of cases of deafness to decrease. We cannot be satisfied with the efforts we have been investing so far, since audiologists have been little consulted in all the spheres of activity, either urban or industrial, as well as in social and even in health services.

Our clinical experience showed that the possibility for the development of hearing loss could be reduced to minimum through correct treatment of diseases and through appropriate protective measures in daily life and work. Even in the case that a hearing loss takes place, a timely surgical intervention or medical therapy can either restore the hearing, or stop the progress of deafness.

In recent years, we have established a close cooperation with a whole range of medical and social institutions, which either deal with the problems of the deaf or include in their work the cases of hearing loss. In this way, we were able to obtain a great number of patients with various causes of hearing loss for our research /hospitals for pulmonary tuberculosis, bone tuberculosis, children hospitals, neurosurgical clinics, hospitals for gynaecology, traumatological and urological clinics, neuro-psychiatric clinics, hospitals for infectious and internal diseases, institutes for labour medicine, for rehabilitation, mental health and for cerebral paralysis/. In addition to this, we examined also students of numerous schools for the deaf and for the hard-of-hearing, as well as adult deaf persons in their institutions and work units. We have carried out also numerous examinations of the workers who work under noisy conditions, as well as of the population exposed to city noise. Numerous patients with hearing loss, the etiology of which was of different kinds, as well as a great number of patients sent from various parts of country for examination, have been examined in the Audiological Department of our Clinic.

In the past four years, we have examined a total of 3,200 persons with serious hearing loss including practically all possible causes for it. So we consider that such a comprehensive analysis could enable us to draw certain conclusions that would apply to the problem of deafness. As all the above mentioned institutions are sending to us almost all the cases of serious hearing loss

for examination, we could consider to have also the appropriate ratio of causes of deafness for our statistics. In this way, we were able not only to watch the causes of deafness but also in every case the success of applied therapy. By watching the causes of deafness at different age levels and in different periods we draw the conclusions on preventive measures and treatment, and foresee the future tendencies.

In addition to a detailed case history of every individual and his family /in certain cases even the parents of the patients have been examined/, every subject has been generally examined by the otolaryngologist, after which a tonal liminar and supraliminar as well as speech audiometry has been carried out. Our analysis included only those persons, whose average hearing loss was above 60 decibels and who, consequently, belonged to the category of persons with serious or total hearing loss.

Wherever it was possible, we applied the tests for topographic diagnosis which, in addition to supra-liminar tests, included also various tests for discovering central lesions /binaural hearing, Cálearo test/. We dedicated particular attention to speech audiometry and to defective pronunciation, omitted or wrongly used letters or syllables, and to the possibilities for social contacts in general. In very serious cases, we endeavoured to establish whether the patient was able to recognize any speech elements or any number of syllables in order to ascertain whether a hearing aid could contribute to the social contact even to the smallest possible extent.

We checked the adequacy and adaptability of a hearing aid through a whole range of most modern amplifiers with different characteristics, size and forms of amplification, adapting them to the degree of hearing loss. We took into consideration for a long auditory training even those patients where was even the slightest hope for a successful use of hearing aids.

We have been supplementing our diagnoses by examining the function of the vestibular apparatus. In addition to static and dynamic functions, we paid particular attention to the caloric test and electronystagmography. Such a detailed study of vestibular and equilibrium organs enabled us more accurately to ascertain the degree, kind and place of the impairment of hearing and of vestibular apparatus.

Thanks to the use of all the above quoted tests, we were able to determine the degree and place of the hearing loss, and to recommend the most advisable methods of treatment and rehabilitation. We are going to describe our most important observations contained in this comprehensive material.

In our documents covering 3,200 cases of serious hearing loss, we had 1,785 males /56%/ and 1,415 females /44%/. It is interesting to remark here that deafness in children appeared to an equal extent in both sexes, and that the differences in frequency according to sex appeared somewhat later, which can be explained with the fact that males are more exposed to diseases, noise and injuries. We collected also a lot of evidence showing that addiction to smoking and drinks, which is more frequent among males, contributes to the frequency of cases of hearing loss.

Even though we have been endeavouring through detailed case histories and numerous tests to ascertain whether it was a question of congenital or acquired deafness in all our patients, we must admit that in all cases we were not able to do so with certainty. If we except the cases, where it has been absolutely established that the hearing loss and occurred later for various reasons, it remains still a significant group of 20% of cases in which we might not be more or less sure whether the deafness was hereditary or congenital /occured during pregnancy or in the period of birth/, or it was acquired in the first days or month<sup>th</sup> after the birth of the child. These uncertain results depend either from the fact that even parents do not remember all the details from that period, or from the possibility that there

were several diseases before, during or after the period of pregnancy, so that it is difficult to ascertain the real cause. There was also the fact that two thirds of women had during pregnancy and delivery no professional assistance, so that it is very difficult to ascertain whether there were some disturbances or not. Most parents had noticed only at the end of the first or in the course of the second year after the birth that their child did not hear or speak, while they are not able to connect this with a disease, or if they are, they do it on the basis of their belief which is not very convincing. In less than 3% of cases we succeeded in establishing with certainty the hereditary elements /deafness in parents or close relatives, the hereditary diseases that could be connected with deafness/.

There was an approximately the same percentage /3%/ which could be connected with various diseases in the period of pregnancy. We can classify here the mothers affected with tuberculosis, who have been receiving streptomycin during pregnancy. There were also comparatively frequent cases of influenza or some infectious diseases /rubeola, morbilli/, and less frequent cases of affected kidneys, heart or liver. In three cases mothers were injured during pregnancy.

For a large group of 167 persons with serious hearing loss we obtained reliable data showing that their handicap was due to various disturbances during or immediately after the delivery. In this group, the most frequent cases were those of various degrees of cerebral paralysis as a result of disturbed supply with oxygen and blood, i.e., of asphyxia of various origine, and then as a result of injuries during the delivery. 85% of those cases occurred when the delivery took place without any professional assistance and without medical help in reviving the asphyxiated child. We think that this is a significant sphere of activity, in which medicine should contribute in the

future to a reduction of deafness. In 12 cases we established the incompatibility of Rhesus factors with resulting hearing loss.

Regarding the post-delivery causes of hearing loss, we should remark that in addition to conventional infectious diseases, there were two groups of cases, on which we should dwell a little longer. First, there was a large group of 217 children, whose parents have stated that they had a high temperature only in the course of few days without any other symptoms, but that during the temperature their consciousness has been more or less troubled. These were mostly sporadic cases in remote vilages and in winter months, when professional assistance either has not been extended or has been give<sup>d</sup> with great delays. In most of these cases, subsequent careful investigations have given to us the ground to think that it was a question of epidemic meningitis or encephalitis, which result in serious hearing loss because of inadequate treatment. The second group consisted of the children who, because of various infectious diseases, have been unnecessarily receiving streptomycine. In our materials the cases of serious hearing loss ocured in this way amount to an imposing figure of 369 cases, of which a very small number can be attributed to an actual disease, while most of them resulted from the use of streptomycine.

Particular attention should be paid to such a great number of cases of hearing loss caused by streptomycine. We can notice also the family sensibility or predisposition in view of the fact that several members of the same family have often lost their hearing because of the use of streptomycine. In this case, too, as in the case of congenital deafness, both mates should be examined by a specialist before concluding the marriage. The works by Podvine<sup>©</sup> and his collaborators at our Clinic have undubitably proved that streptomycine can penetrate the placental barrier and damage the hearing of foetus. We should emphasize the

great significance of the early therapy for eliminating the consequences of serious damage caused by streptomycine.

By comparing the etiology of deafness between the lower and the higher age levels, we can see that there was a continuous decrease in the number of the cases of hearing loss as a result of ear and all infectious diseases which have been properly treated, and a rise in the number of cases of hearing loss caused by industrial and city noise, chemical and toxic causes, various injuries /primarily those which result from traffic accidents/, and the blood circulation and arteriosclerotic troubles due to an increased number of old people.

### Conclusions

The analysis of 3,200 cases of deafness and serious hearing loss showed that the number of such cases is continuously increasing, and that the struggle against this handicap can be successful only if adequate efforts are invested in the course of the whole pregnancy period and after the birth. Early identification of all the disturbances and diseases, and the undertaking of appropriate diagnostic and therapeutic measures for eliminating these disturbances represent the best prevention against deafness. The use of toxic medicaments exclusive when strictly prescribed, along with a continuous control of hearing represent also one of the measures for preventing deafness. The measures against physical and chemical detrimental elements under industrial and urban conditions can produce also precious results, while the struggle against various disturbances in blood circulation will also reduce the number of cases of hearing loss among old people.

Finally, the efforts for early diagnosis of hearing loss are not less important. The rehabilitation possibilities are now such that even the cases of a very serious hearing loss can be successfully treated if discovered in time and if an adequate auditory training has been secured, so that many hard-of-hearing people can be enabled to join the world of people with normal hearing.