SCREENING AND EPIDEMIOLOGY OF DEAFNESS IN VERY YOUNG CHILDREN

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ABSTRACT

This paper deals with the results of systematic testing of 10,270 new-borns between 1969 and 1975. The results of that testing program had a yield of 13 or 1 in 750 live births with a hearing loss. Further analysis was completed to consider factors contributing to that hearing loss.

INTRODUCTION

Over the last 20 years there has been an attempt to diagnose deafness in very young children and to treat these hearing deficiencies from a very early age.

For over 10 years now, workers have turned their attention to neonatal screening of deafness and to the study of auditive sensory-motor reactions.

In France systematic screening equipment and techniques were developed by Paul Veit and Geneviève Bizaguet. In Lyons the research team of Professor Mounier-Kuhn continued work on infant deafness (J. C. Lafon and A. Plantier — A. Morgon and D. Charachon) established an epidemiology of deafness in very young children. In 1969 Marion Downs in Colorado made a study on deafness in 17,000 new born babies.

Now reliable statistics are available from M. P. Downs (Colorado), G. T. Mencher (Nebraska), A. Sonninen (Finland), D. Ling (Canada) and J. C. Lafon, J. Helias and A. Menegaux (Besançon).

These statistics show that at birth there is approximately one case of deafness for 1,000 births. At the age of 1 year the number of cases is between 1.2 and 1.5 for 1000 births.

All these results have shown the usefulness of systematic screening of the auditive sensory-motor reactions of babies from birth for the detection of deafness in a population, particularly in a population with an established deafness risk.

Only 5% of cases of deafness have been found to have no precise etiology.

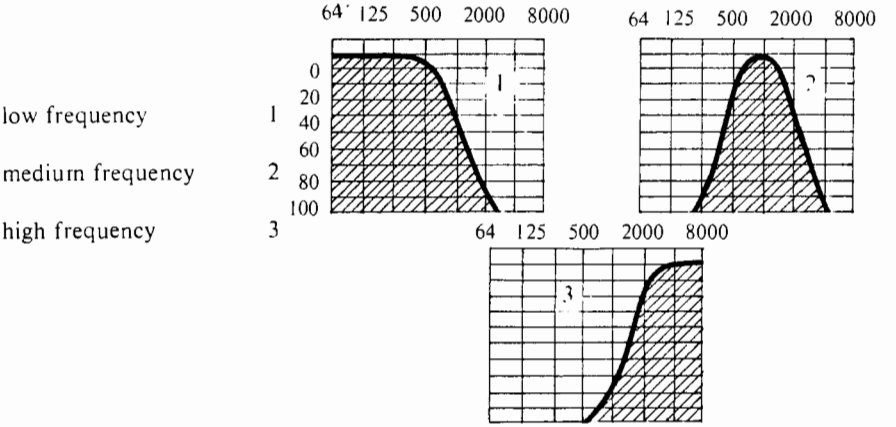
Evidence now demonstrates that systematic screening for deafness should be part of the standard neonatal and postnatal examinations. However until this does become a reality and despite the difficulty of organising such screening tests in big cities and countries, the benefit of early adaptation and education would justify the setting up of a medical-psychological structure with well trained staff.

A strategy of systematic screening of deafness at birth was started in 1969 at the Maternity Hospital and the Premature and Neonatal Pathology Service of the University Medical School in Besançon.

Several years of experience linked with the regional tradition of the Audiophonology Centre have enabled us to test a population of 10,000 new born babies from 1969 to 1975 and to analyze the result of this preventive action.

METHODOLOGY

1.1 We used the screening audiometer of VEIT-BIZAGUET which gives 4 types of signals: white noise and white noise that is filtered through low, medium and high pass bands.



1.2 The motor reactions that were observed are not specific to the sensory means used i.e. hearing, as they also exist for other sensory stimulations.

Thus, to be able to speak of **sensory motor reactions** to an auditory stimulus the tester must be sure that:

— there is a temporal relationship between the sound stimulus and the observed motor reaction
— the new born baby is not exposed to a concomittant stimulation i.e. visual, tactile, olfactory.

These responses are varied in their latency, their type and their intensity and the ones that are most frequently used in clinical examination are:

— **motor reflexes** (very short latency — approximately 1/10th sec.)
— auro palpebral reflex
— moro reflex
— motor reflex
— limb tonerity reflex.
— **Postural reflexes** (latency 0.5 to 1 second)
— cephalic acoutrop reflex
— ocular acoutrop deviation
— cephalic acoutrop reactions reflex
— hyper extension of the head.

1.3 The acoustic test was given as follows:

— **First examination** between 24th and 48th hour: if the test is given earlier there is a high percentage of false negative results due to the obstetrical shock, if the test is given later further necessary examinations can not be given easily because both mother and child will have left the hospital.
— **The best moment** is determined by the state of awareness of the child which is extremely unreliable: this is usually during the half hour which precedes feeding when the new born baby wakes up from a sleep that corresponds to the transition between state 2 and state 3 of Prechtl and Beintema's classification.

1.4 The sensory motor behaviour of the new born baby is normal when it responds to three reactions:

— reaction to the emission of two acoustic signals: white noise and high pass band noise

— reaction to an intensity threshold that is below or equal to the safety threshold fixed at 15 dB of background noise that is measured with a sonometer (dBC)
— bilateral reaction.

When one or more of these elements is missing the new born baby is considered to be "suspect" and must be seen again while he is still in the hospital. The babies that leave the hospital without showing normal auditory reactions are tested again at 3 months.

1.5 Our data was obtained from three sources:
— systematic screening of 7,000 new born babies in a maternity hospital
— screening applied to 470 high risk new born babies selected according to pediatric criteria in a neonatal pathology service
— screening of 2,800 new born babies in hospital in the same neonatal pathology service.

RESULTS

For 10,270 new born babies examined we found 13 with bilateral hearing losses whose etiologies are as follows:

2.1 screening at the maternity hospital
— 4 profound auditory deficiencies (a loss of 90 dB according to the B.I.A.P. classification) .057% or 1/1750
— 1 case of genetic origin found at birth and diagnosis confirmed at 4 weeks
— 2 cases were judged to have satisfactory auditory reactions.
— However, further tests were made because both cases presented a potential risk
  a. one diagnosis made at 2 months (encephalopathy)
  b. the other at 1 year (genetic origin)
— 1 case was not recognized at screening as the reactions were normal at birth. This child was re-examined in our service at the age of 2: a case of indetermined etiology.

2.2 Screening in the Neonatal Pathology Service

In this service we found 4 children with hearing loss: 1 profound, 2 severe and 1 moderate (according to the BIAP classification) among 2,800 children tested: .143% or 1/700
— 2 cases were found at birth:
  a. one case was without an auditory response, profound hearing loss was confirmed at 18 months and no previous history was found except for a premature birth
  b. one case where there were positive reactions, but the threshold was too high (100 dB); severe hearing loss which was confirmed at 2 years
— 1 case of profound hearing loss who had normal reactions at birth: this case was closely followed because both parents were deaf and dumb, and deafness appeared at 6 months
— 1 case of severe hearing loss who had normal reactions at birth. This child came back to the hospital at 2 years 3 months and was diagnosed as a case of deafness probably due to maternal rubella despite a normal test result at birth.

2.3 Screening of children from high risk pediatric service

According to the pathological criteria 470 children were selected from the Premature and Neonatal Pathology Service and here we found 5 cases of hearing loss (4 profound, 1 severe): 1.06% or 1/94.

— 3 cases of profound hearing loss due to maternal rubella: at neonatal screening the reactions were abnormal and the control examinations confirmed the diagnosis
— 2 unrecognized cases:
  a. one child with a disturbed behaviour pattern in a pathological context (meningeal hemorrhaging, multiple apnea, severe jaundice) followed by a return to normal auditory reactions with a normal neurological examination. Examined again at 1 year when in hospital and auditory reactions were found to be normal. At 20 months during a simple rhinopharyngeal infection the child had problems walking and a profound hearing loss became apparent.
  b. one other child also had no initial reactions but then showed normal reactions. At the age of 3 a profound hearing loss was diagnosed. The neonatal origin was probably anoxia and an ototoxic treatment.

2.4 Statistical summary

For 10,270 children screened at birth we found 13 cases of hearing loss: .127% or 1 for 750 births.

Thus for .127% of cases of hearing loss .058% resulted from the neonatal acoustic test and were then diagnosed as such.

.029% came from high risk children given audiological attention despite the fact that their neonatal reactions were normal.

.039% unrecognized by screening.

DISCUSSION

3.1 We have compared our results with studies made on populations of over 10,000 cases.

	Downs M.P. Hemenway W.G. (2)	**Ling D.** (3)	**Mencher G.T.** (4)	**Sonninen A. Pyorala T. Klemetti A.** Finland (6)	**Lafon, J.C. Helias J. Menegaux A.** Besançon France	Total
Population tested	17,000	20,000	10,000	17,407	10,270	75,000
Cases of hearing loss	17	11	9	23	13	73
	.1%	.055%	.090%	.132%	.127%	.097%
Diagnosis following screening	15	4	7	11	9	46
Unrecognized during screening	2	7	2	12	4	27

3.2 The results remain variable and depend on the method chosen, but probably even more on the later screening made on the original population. It is obvious that A. Sonninen (6) who is working at a provincial level can check the validity of these results as non-diagnosed deaf children are sent to this Centre — this is also our case, but

our region is much smaller. The non-diagnosed cases screened at birth are listed in the comparative diagram. A certain amount of time is required to follow the cases in our population. We noted that the percentage of deaf children went from .097% to .127% when one more year was taken into account (5).

Thus, we feel that the results of D. Ling (3) must be treated with some caution.

3.3 The comparison of our results with other studies leads to two conclusions:
nearly half the cases of hereditory deafness do not exist at birth, but develop during the first year in association with benign pathological problems or even without an apparent reason, as if there were a cochlear fragility. In these cases there is often deafness within the family. Thus neonatal screening does not dispense with the need for further checks.
If all the cases of deafness are studied together with family background, difficult pregnancies, births and anomalies apparent during the first week only one case of deafness in ten thousand appears to be without any suspicious elements.

3.4 These results are confirmed when an etiological exploration is undertaken. A study made on 90 deaf children regularly attending our centre has shown that the percentage of unknown etiologies goes down to 5%.

3.5 These results pose the problem of systemizing neontal screening for deafness. Should this be generalized or be used only on certain groups of children with a risk factor?

This concept of "high risk" children is developing with the progress of genetic and neonatal studies. Several English speaking authors have tried to develop a strategy for screening infirmities; Lindon — 1961 hoped to discover 70% of infirmities with a protocol based on 20% of the child population during the first year, but a list of risks put forward by Mary Sheridan (1960) showed that 70% of children born alive could be considered as risk children.

90 CASES OF AUDITORY DEFICIENCY
(Severe & Profound Deafness)

	Etiology	No. of cases	% found
GENETIC DEAFNESS	— true genetic deafness	20	22.22
	— congenital deafness probable genetic factor	8	8.89
ACQUIRED DEAFNESS	**PRENATAL DEAFNESS** — Maternal Rubella — other embryopathy — foetopathy (hemorrhaging during pregnancy)	 18 1 2	23.33
	NEONATAL DEAFNESS — prematurity — prematurity with anoxia — anoxia	 6 4 5	16.67
	INFANTILE DEAFNESS — meningitis — mumps — encephalopathy	 6 4 1	12.22
	SEVERAL ETIOLOGIES	10	11.11
	UNKNOWN ETIOLOGY	5	5.56

Genetic alone	5	31.11
Genetic associated with acquired (prenatal, neonatal, infantile)	28	5.56
Acquired alone	47	52.22
Several acquired	5	5.56
Unknown	5	5.56

In the case of the risk of hearing loss our results are against the practical value of neonatal screening outside premature and neonatal pathology centres where it is essential in view of the frequency of deafness and in particular deafness that is associated with other pathologies.

However, it must be pointed out that the etiological diagnosis of deafness studied in this article (screening and etiology) are both etiological diagnoses made when there was a suspicion of deafness and diagnoses made by the pediatricians who knew the auditory risk of symptoms which require an audiological investigation.

We believe that at the present time the systematic screening for hearing loss is of interest and that increased attention must be given to children with a potential risk.

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