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ABSTRACT

The audiological assessment of 50 deaf blind children, 6 months to 14 years of age, in an outpatient setting is described, as are testing procedures and results. Etiological factors are given which include maternal rubella (accounting for 27 children), meningitis, prematurity, neonatal anoxia, and Rh incompatability. Discussed are the following testing procedures: pure tone audiometry (which is not appropriate for children with minimal or no hearing and vision); play audiometry, such as dropping a block in a bucket, a procedure said to be useful for children above 2 years of age but to have limitations in an outpatient setting because extensive training sessions are required; conditioned orientation response audiometry, which was effective for 25 children who perceived the light stimulus; impedance audiometry, involving use of an electroacoustic bridge for obtaining data from both ears (22 of 24 children tested showed middle ear involvement); and behavior observation audiometry, for detection of overt responses such as startle reflexes or cessation of an activity in very young or otherwise untestable children. Some of the results indicated that 22% of the children had hearing sensitivity within the range of normal limits, but had problems other than blindness that precluded language and speech development; that two children were untestable; and that of 37 children with significant hearing loss, 62% were in the severe to profound range (10 were rubella children). (MC)

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AUDIOLOGICAL ASSESSMENT OF DEAF-BLIND CHILDREN

by

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INTRODUCTION

Since 1966, the Callier Hearing and Speech Center has evaluated 50 children who were classified as deaf-blind. These patients were not in a residential setting; they were evaluated on an outpatient basis. The diagnosis of deaf-blind was given either by the results of extensive visual and auditory testing or on the basis of a known visual impairment, such as cataracts, and the inability of the child to develop normal speech and language. Figure 1 shows the ages of subjects at the time of the initial evaluation. The patients ranged in age from 6 months to 14 years, and the mean age was 4 years 6 months. Table I is a summary of the etiological factors causing the hearing and visual impairments. The various etiologies included: maternal rubella, meningitis, prematurity, neonatal anoxia and Rh incompatibility. The primary etiological factor was maternal rubella, accounting for 27 of the 50 children or 54% of the population. Specific etiologies could not be established for 10 children.

The present paper will describe the methods of audiological assessment used in evaluating these children, impression of the efficacy of these testing procedures and an overview of the audiometric results that were obtained from this population.

AUDIOLOGICAL ASSESSMENT

In the audiological assessment, both formal and informal tests were employed. Formal testing included: (1) standard or conventional pure tone audiometry, (2) play audiometry, (3) conditioned

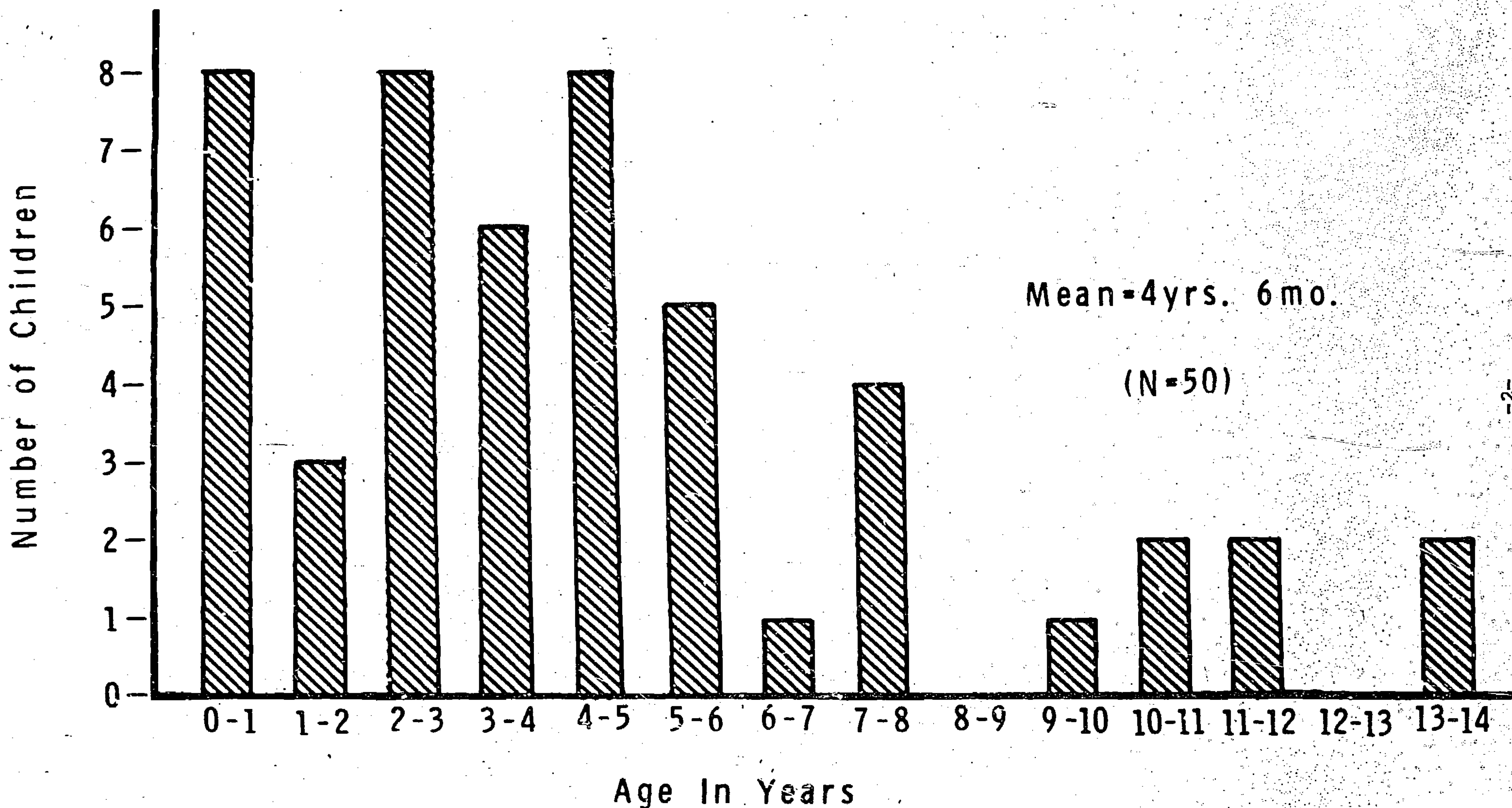

Number of Children
8
7
6
5
4
3
2
1
0
0-1
1-2
2-3
3-4
4-5
5-6
6-7
7-8
8-9
9-10
10-11
11-12
12-13
13-14
Age In Years
Mean=4yrs. 6mo.
(N=50)

TABLE I

ETIOLOGICAL FACTORS

Etiological Factor	N	%
Rubella	27	54
Meningitis	3	6
Prematurity	6	12
Neonatal Anoxia	3	6
Rh Incompatibility	1	2
Unknown	10	20
	50	100%

orientation response or COR audiometry and (4) impedance audiometry. Informal testing was accomplished using behavior observation audiometry, commonly referred to as BOA. It should be noted that electrophysiologic methods, such as evoked response audiometry, psychogalvanic skin response audiometry and electrocochleography, were not evaluated.

The procedure generally employed when using conventional pure tone audiometry is to instruct the patient to make an overt response, such as raising his hand or pressing a signal button to the presence of the sound stimulus. In addition, a brief demonstration is generally provided. It is obvious that conventional audiometric techniques had limited applicability to this deaf-blind population. The primary factors which negated the general use of conventional audiometry with our population were: (1) the young chronological age of many of the patients, (2) the inability of most of these children to follow verbal instructions and (3) the inability to demonstrate the task due to the visual impairment. Only four from the sample could be tested using conventional audiometry. All four of these children were above the age of four years and had sufficient speech and language abilities to enable them to follow the instructions. These children had mild to moderate hearing losses.

Play audiometry, the second technique used with this population, was accomplished using standard responses, such as dropping a block in a bucket or stacking blocks. This technique is efficient in testing young children and is probably the technique most often employed with the pediatric population generally above the age of 2 to 2½ years. It is a more objective testing method than BOA or COR. Results with this population seemed to indicate that play audiometry had limited applicability to this deaf-blind population. As compared to children with no visual deficit, results indicated that more training sessions were required to condition the deaf-blind child to respond appropriately than were feasible. Therefore, the primary factor that precluded the use of play audiometry with this population was the number of sessions required. It would seem that the best method of applying play audiometry for difficult-to-test populations, such as the deaf-blind, would be to schedule daily test training sessions. This procedure was not possible, however, due to clinical scheduling limitations and the distances travelled by some of the patients to come to Callier Center in Dallas.

If it was felt that the child could be conditioned and frequent sessions could be scheduled, the child's teacher and/or parent could help prepare the child for assessment. This was accomplished by instructing them to condition the child to perform a specific task to the presentation of a stimulus known to be perceived by the child, such as a tactile, visual or intense auditory stimulus. The parent or teacher was counselled to use short sessions of 10 to 15 minutes frequently during the day. Progress was monitored periodically to determine the stage of conditioning. Even with this method, play audiometry required 6 months to 1 year before definitive testing was accomplished in 9 of the 14 children where play audiometry was applied. Play audiometry was used as an initial diagnostic tool only after other techniques attempted were not successful. The play technique was also used to confirm results from other techniques as the children matured.

A technique that might be considered as having little or no application to the deaf-blind population is COR audiometry. In this method, a visual stimulus is used to reinforce appropriate localization responses. It has been shown that the COR technique is an efficient tool in the assessment of hearing sensitivity for children between the ages of 1 and 3 years (Suzuki and Ogiba.) In addition, the COR technique has been shown to have application to the mentally retarded population (McPherson, 1960 and Fulton, 1965.) The obvious difficulties in applying this method to this deaf-blind population were: (1) 8 children were below the age of 1 year and (2) the apparent lack of visual sensitivity to perceive the light stimulus. However, this method proved to be of considerable value in testing the population at Callier Center. It was found that only 4 children in the population above the age of 1 year were unable to perceive the light stimulus used. Parenthetically, one of these children had an ophthalmological diagnosis of cortical blindness; and the other three were the only children with a diagnosis of retrolental fibroplasia. Of the 34 children above one year of age who had sufficient visual sensitivity to perceive the light stimulus, 25 successfully responded to the COR technique. There does not seem to be a simple explanation as to why the remaining 9 children could not be tested by this technique because they seemed to demonstrate similar overt behavior to those children who were successfully tested using COR.

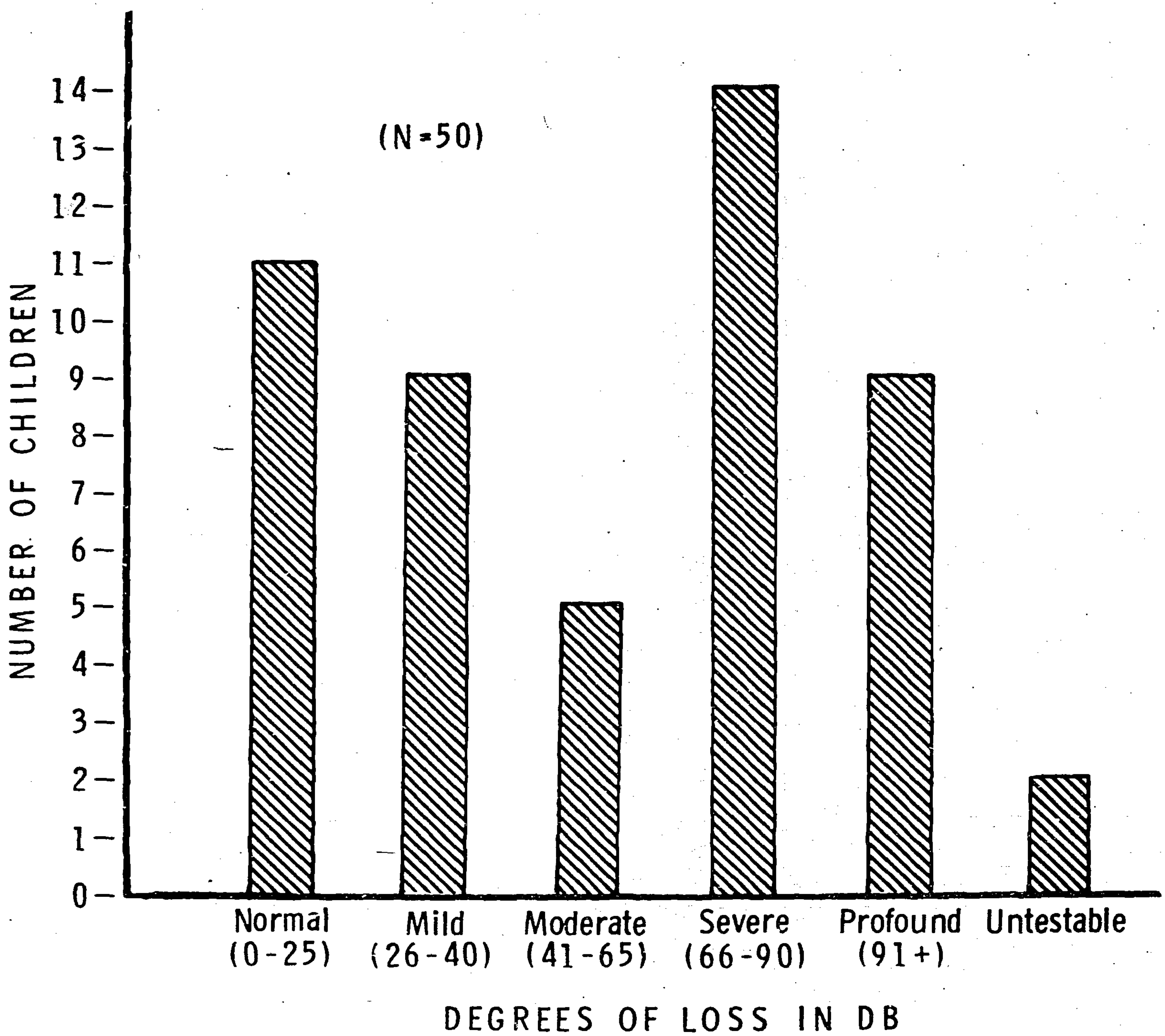
Impedance audiometry has been used as a routine procedure in Callier Hearing and Speech Center since late 1969. Therefore, impedance results were not obtained on all of the deaf-blind children in this study. Impedance audiometry, using an electroacoustic bridge, was successful in obtaining data from both ears for 22 of the 24 children tested with this technique. Decreased cooperation in two children caused incomplete findings. Results from electroacoustic impedance measurements revealed that 10 out of 22 children had apparent middle ear involvement. Subsequent otolaryngological examinations were commensurate with impedance findings in 9 of these 10 cases. A significant point seems to be that middle ear pathology could not have been detected by the routine method of air and bone conduction comparison due to the severity of the hearing losses and the difficulty in obtaining threshold responses. Results of these findings indicate that impedance audiometry, using an electroacoustic impedance bridge, is a valuable part of the audiometric test battery, especially for difficult-to-test populations, including deaf-blind children.

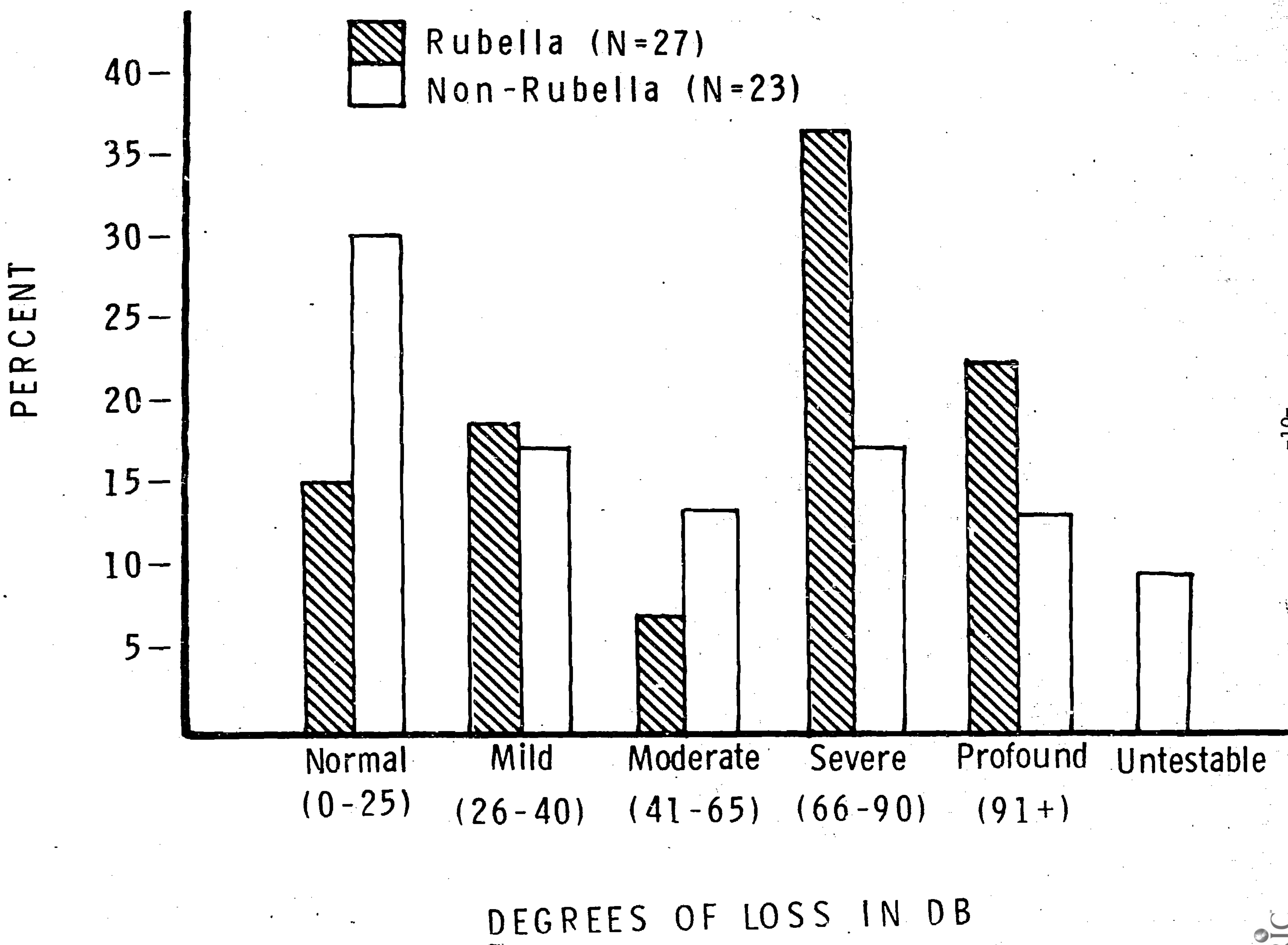
Behavior observation audiometry (BOA), the informal testing method, involved the observation of overt responses in the presence of auditory stimuli. The types of responses noted were: (1) startle reflexes, (2) rudimentary localization responses, (3) cessation of activity, such as "listening behavior," and (4) instigation of activity, such as laughing, crying and eye blinking. In all cases, two observers scored the responses independently to try to control for the inherent subjectivity of this method. This technique proved to be the only successful behavioral method used with the younger deaf-blind child, generally a year of age or younger, and with the older deaf-blind child when all other behavioral methods failed.

RESULTS

By using the above techniques, audiometric thresholds ranging from normal hearing sensitivity to profound hearing loss were found. The data shown in Figure 2 were based on the average threshold sensitivity for the frequencies 500, 100 and 2000Hz in either the better ear or from sound field testing. This figure shows that 22% of the population (11 children) had hearing sensitivity within the range of normal limits. These were all blind children with other problems, such as brain injury and mental retardation, that precluded the development of speech and language abilities. Two children were untestable by the procedures outlined. Of the 37 children who demonstrated significant hearing loss and could be tested, 62% fell within the severe to profound range.

Because of the large percentage of rubella children in this population, the degrees of hearing loss for this etiological factor were compared to the others in the study. From Figure 3 it appears as if more rubella children had severe hearing losses than the non-rubella population. Ten children with rubella as the etiological


NUMBER OF CHILDREN
14
13
12
11
10
9
8
7
6
5
4
3
2
1
0
(N=50)
Normal (0-25)
Mild (26-40)
Moderate (41-65)
Severe (66-90)
Profound (91+)
Untestable
DEGREES OF LOSS IN DB



Rubella (N=27)
Non-Rubella (N=23)
PERCENT
40
35
30
25
20
15
10
5
Normal (0-25)
Mild (26-40)
Moderate (41-65)
Severe (66-90)
Profound (91+)
Untestable
DEGREES OF LOSS IN DB

factor were in the severe range while 4 with other etiologies were in this category. Furthermore, the non-rubella population had more children falling within the range of normal hearing sensitivity.

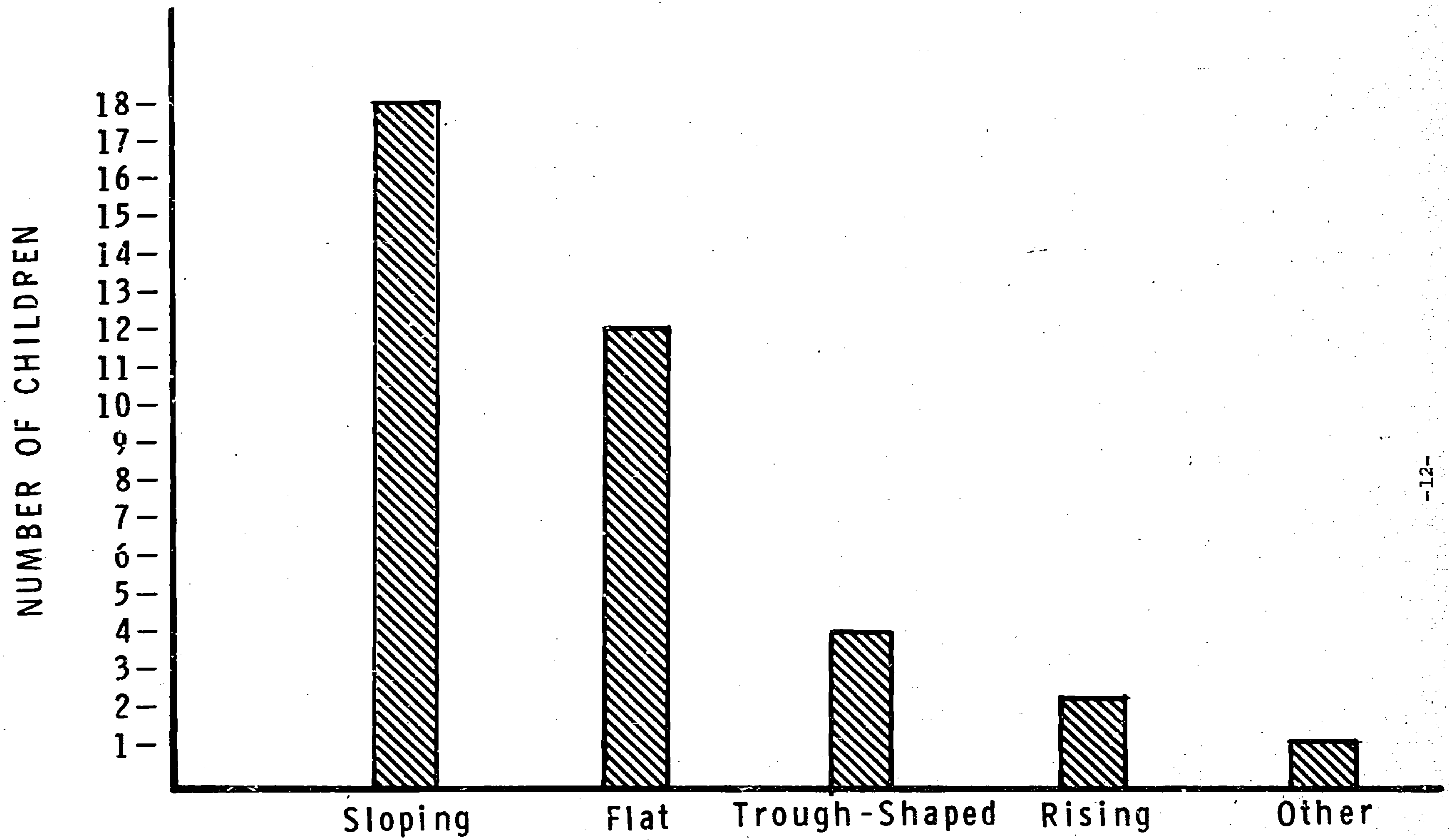
Figure 4 is a summary of the configuration of hearing losses for the 37 children who were tested and found to have significantly decreased hearing sensitivity. Of the five categories, the majority of the losses were classified as sloping. The sloping loss was defined as having an average of 5 dB or more decrease in threshold for 250-4000Hz. The second largest class into which the losses fell was flat, accounting for 12 of the 37 children.

A comparison of the configuration of the hearing losses of the rubella and non-rubella deaf-blind population was made. Two observations were: (1) only the rubella children demonstrated trough shaped hearing losses, which were losses with better hearing sensitivity in the upper and lower frequencies than the middle frequencies and (2) the rubella population had slightly more hearing losses classified as flat than did the non-rubella population.

SUMMARY AND CONCLUSIONS

(1) First, this is a challenging population to test and the techniques used on the pediatric population and mentally retarded population are not always applicable to this group.

(2) With children less than 1 to 1½ years and sometimes older, BOA appears to be the only behavioral method that can be



CONFIGURATION OF HEARING LOSS IN DEAF-BLIND CHILDREN (N=37).

applied to some deaf-blind children.

(3) The COR technique is generally a successful method for testing deaf-blind children provided that they have sufficient visual acuity to perceive the light stimulus.

(4) When testing deaf-blind children on an outpatient basis, play audiometry has limitations due to the intensive number of training sessions required.

(5) Impedance audiometry should be used as a routine procedure with all difficult-to-test populations, including the deaf-blind, due to the high incidence of middle ear pathology found in this population.

(6) The most effective approach for successfully testing these children using behavioral methods was to attempt several techniques and select the ones most suitable for the specific child.

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