INFANT HEARING SCREENING: THEORY AND PRACTICE

by

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ABSTRACT

The "Year of the Child" provides a unique opportunity for the allied health field to focus professional and public attention on the urgency of identifying hearing loss in infants. This article reviews the development of infant screening programs, discusses contributions made by the United States Joint Committee on Infant Hearing, the Nova Scotia and Saskatoon Conferences on Early Identification of Hearing Loss and outlines the results of an infant screening program currently under way at the Grace Maternity Hospital, Halifax. Concluding remarks are directed toward the establishment of a Canadian Joint Committee on Childhood Hearing Impairment, the further development of Canadian professional cooperation and the responsibility of our profession to become advocates in the establishment of infant screening programs.

As part of our professional recognition of this designated "Year of the Child", the focus of this article is directed toward recent and continued advances which have been made in identifying hearing loss in infants.

Since the inception of neonatal auditory screening programs, the early detection and identification of hearing impairment in infants has seen monumental development (Wedenberg, 1963; Downs and Sterritt, 1964, 1967; Hardy, 1967). Due perhaps to a perceived urgency, the first neonatal programs were plagued with a number of innate difficulties which drew justifiable criticism (Goodhill, 1967; Ling, Ling and Doehring, 1970; Goldstein and Tait, 1971; Borton and Stark, 1962; Northern and Downs, 1974). More recently, however, through the efforts of longitudinal clinical research, the emergence of the United States Joint Committee on Infant Hearing in 1970 and two related international conferences, held in Canada, a number of positive steps have been taken to develop and strengthen screening programs by refining the procedures to be utilized in the early identification of hearing loss. It is the intent of this paper to discuss a number of developments which have contributed to this cause.

THE MODEL PROGRAM

The United States Joint Committee, which consists of representatives from the American Academy of Ophthalmology and Otolaryngology, the American Speech and Hearing Association, and the American Academy of Pediatrics issued two policy statements which markedly altered the philosophy and approach to neonatal hearing screening. The initial statement in 1970 urged increased research efforts in the area of behavioral screening programs, specifically, in terms of five variables which will affect

the procedure (e.g. test stimulus, infant response, scoring criteria, pretest state, and test environment). A second statement which was issued in 1973 and only after a number of longitudinal research studies had been completed (Downs and Hemenway, 1969; Mencher, 1972; Alexander et al., 1976; Altmann, 1969; Feinmesser and Tell, 1971) recommended the use of a "high risk register" for hearing impairment. The criteria for identifying a newborn as "at risk" includes the presence of one or more of the following:

a) History of hereditary childhood hearing impairment in first cousins or closer;

b) Rubella or other nonbacterial intrauterine fetal infection (e.g. cytomegalovirus infections, herpes);

c) Defect of ear, nose or throat. Malformed, low set or absent pinnae; cleft lip or palate (including submucous cleft); any residual abnormality of the otorhinolaryngeal system;

d) Birth weight less than 1500 grams;

e) Bilirubin level greater than 20 mg/100 ml serum.

After the Joint Committee statements were issued, a great deal of effort continued toward the development of a behavioral protocol as an additional screening procedure. In 1974 the Nova Scotia Conference on Early Identification of Hearing Loss presented a series of recommendations which were subsequently adopted by the Joint Committee. Among those recommendations, which are still current, are the use of the five item high risk register presented earlier and an additional statement which reads as follows:

> "As a supplement to the high risk register, an agency may employ behavioral screening tests as in the appended model."

The model referred to may be seen in Table 1. It represents a defensible recommendation for a behavioral evaluation of neonatal hearing as a supplement to the high risk register. The behavioral procedures approved by the Joint Committee were accepted only after intensive research involving the five variables referred to in the 1970 statement. For a comprehensive review of the literature focusing on that research, including an extensive list of references, the reader is referred to Mencher, Jacobson, and Seitz (1978).

TABLE 1

SUGGESTED PROTOCOL FOR A BEHAVIORAL HEARING SCREENING TEST

1. Test Stimulus:

A random noise having a low frequency attenuation of 30 dB or more per octave below 750 Hz; a maximum of 90 dB sound pressure level at the pinna; a rise-decay time of five (5) milliseconds or more; a duration of 0.5 to two (2) seconds; an interest interval minimum of fifteen (15) seconds.

2. Infant Response:

Any generalized body movement which involves more than one limb and which is accompanied by some form of eye movement.

3. Scoring Criteria:

Controlled by one of two methods:
a) Scorer does not know when a test stimulus is actually present, or
b) Two observers score and infant's responses independent of one another.

Furthermore, two (of eight maximum) stimulus responses should be positive to score as a "pass" and a "failure" should be retested at least once (with a commulative positive response score of more than 20%) before being considered a true test "failure".

4. **Pretest State:**

The pre-test behavioral state of an infant is an important determinant in governing the initiation of a response and must be controlled or described in specific terms. This protocol calls for a sleeping infant (eyes closed, no observable body movement for at least fifteen (15) seconds prior to stimulation.

5. **Test Environment:**

The ambient noise level at the time of the typical test should be measured and reported.

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* From the recommendations of the Nova Scotia Conference on Early Identification of Hearing Loss. For reference see G. T. Mencher, Editor, **Early Identification of Hearing Loss,** (S. Karger: Basel, Switzerland), 1976.

In 1978, a second conference on the early identification of hearing loss was convened in Saskatoon. Emanating from that conference was a series of fourteen recommendations related to early diagnosis. Included in the resolutions was an endorsement and reaffirmation of the principles of early identification and the specific recommendations of the Nova Scotia conference. In addition, a new category based on the significant number of hearing losses associated with anoxia at birth was added to the high risk register. It is defined as, ". . . significant asphyxia associated with acidosis" (see Table 2).

TABLE 2

STATEMENT RE: SIGNIFICANT ASPHYXIA

In the past many neonates with fetal distress and low Apgar scores have been examined with a low incidence of hearing loss. However, as regional neonatal intensive care units expand and greater numbers of ill infants are transferred for special care, those term infants requiring neonatal intensive care because of antenatal and/or birth anoxia or birth trauma are now more readily identified. Those infants with hypoxic encephalopathy with persisting abnormal neurological signs or neonatal convulsions secondary to anoxia have a high incidence of neurosensory hearing loss as well as other developmental disabilities.

A specific level of acidosis (Arterial pH) is not given here as an indicator of subsequent handicap as in some areas it may alter the pH level prior to measurement. A pH level of 7.3 associated with asphyxia has been associated with hearing loss.

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* From the recommendations of the Saskatoon Conference on Early Diagnosis of Hearing Loss. For reference, see Gerber, S. E., and Mencher, G. T., Editors, **Early Diagnosis of Hearing Loss,** Grune and Stratton, New York, 1978.

Other resolutions dealt with methods for the confirmation of the presence and/or degree of hearing loss within the first six months of life. Chief among the new methods was the formal inclusion of evoked response audiometry as a test procedure, advancing that technique from research to clinical application. Further, since middle ear effusion may occur and persist in infants, procedures for identification and management of that problem were also presented (see Tables 3 and 4).

TABLE 3

STATEMENT RE: ELECTRIC RESPONSE AUDIOMETRY (ERA)

1. **Auditory Brainstem Response**

Two forms of the auditory brainstem response test are recognized; both of them test only the peripheral auditory mechanism. The simpler test uses unfiltered clicks as stimuli and test primarily the sensitivity of the ear to frequencies above 1500 Hz. It is particularly applicable as an early follow-up test of babies at six (6) weeks of age. A more elaborate test, still under development, used filtered clicks and yields a four frequency (500, 1000, 2000, and 4000 Hz) audiogram. The test is applicable to all ages; for both of these tests sedation is not usually required for infants below three months of age.

The auditory brainstem response test should not be used alone, but only as part of a battery of tests that also includes impedance audiometry and behavioral tests. Furthermore, an audiogram may be generated from middle component evoked potentials using tone burst stimuli (see below).

The auditory brainstem response test, which is relatively expensive and time-consuming, need to be used only if behavioral testing is not feasible or yields equivocal results. The auditory brainstem response test must be administered by adequately trained personnel.

The simpler auditory brainstem response test has the demonstrated capability of producing diagnostic data equal to or exceeding that described in monographs by Davis (1976) and by Picton et al. (1977). The report of such a test should include, for each ear:

1. Latency of Wave V response as a function of the clicks;
2. Threshold estimate;
3. Statement of the type of hearing loss found, if any;
4. Measurement of the I-to-V interval of the brainstem responses.

The procedures and capabilities of the auditory brainstem response test are still under development. Diagnostic centers which make use of it must remain aware of developments in the state of their art.

If the auditory brainstem response test yields negative or equivocal results, electrocochleography may be considered to assess the physiological state of the cochlea.

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* From the recommendations of the Saskatoon Conference on Early Diagnosis of Hearing Loss. For reference, see Gerber, S. E., and Mencher, G. T., Editors, **Early Diagnosis of Hearing Loss,** Grune and Stratton, New York, 1978.

TABLE 4

STATEMENT RE: MIDDLE EAR EFFUSION

WHEREAS, middle ear effusion can occur in the newborn and in infants; and

WHEREAS, middle ear effusion may persist chronically for months or years producing a mild bilaterial hearing loss; and

WHEREAS, prolonged mild hearing loss may produce speech, language, educational, and behavioral problems,

RESOLVED: PARTICULAR ATTENTION SHOULD BE PAID TO THOSE NEWBORNS LIKELY TO HAVE SUSTAINED MIDDLE EAR EFFUSION. AS WITH ALL CHILDREN, THESE NEWBORNS

SHOULD BE CLOSELY FOLLOWED, AND THE EFFUSION BE CONSIDERED A PROBLEM IF IT IS SUSTAINED (MORE THAN THREE MONTHS) OR IS RECURRENT (OVER 50% OF THE TIME FOR SIX MONTHS). DIAGNOSIS SHOULD BE BASED UPON PNEUMATIC OTOSCOPY, TYMPANOMETRY, AND AUDIOMETRY. FURTHER, IF A SUSTAINED OR RECURRENT PROBLEM DIAGNOSED, EDUCATIONAL INTERVENTION SHOULD BE APPLIED IN THE FORM OF LANGUAGE STIMULATION PROGRAMS AND/OR LOW LEVEL AMPLIFICATION IN ADDITION TO ONGOING MEDICAL OR SURGICAL TREATMENT.

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* From the recommendations of the Saskatoon Conference on Early Diagnosis of Hearing Loss. For reference, see Gerber, S. E., and Mencher, G. T., Editors, **Early Diagnosis for Hearing Loss,** Grune and Stratton, New York, 1978.

THE CLINICAL PROGRAM

The newborn screening and follow-up methods recommended by the participants at both the Nova Scotia and Saskatoon meetings have been implemented in a clinical program in Halifax. The first full year of operation is completed and concern has now shifted to continued screening and the follow-up program.

As illustrated in Table 5, last year there were 4,910 babies born in the Halifax area, 523 of whom were on the five item high risk register. The procedures for obtaining information which may place a child on the register must be carefully controlled, due to the number of variables which may negate or even destroy the effectiveness of the program. For example, category "A" or affected family is a large problem area. The numbers obtained in that classification in Halifax are greater than they should be (453 of 523), no doubt due to continuing difficulty in obtaining valid information from families regarding the presence of congenital sensorineural hearing loss in first cousins or closer. Here is where interviewers must be trained to question very carefully or the number of false positives can become unreasonable.

TABLE 5

HIGH RISK INFANTS REQUIRING BEHAVIORAL TESTING

523 of 4910 (10.7%) on the High Risk Register

Category	Number	Percentage
Affected Family	453	(86.6%)
Congenital Rubella	17	(3.3%)
Defect of ENT	25	(4.8%)
Bilirubinemia	—	—
Small at Birth	28	(5.4%)

Note the absence of hyperbilirubinemia in our first year of data. In one year of operation, not one single case with bilirubin level elevated to 20 mg/ 100 ml serum was reported in Halifax. No doubt, use of intrauterine transfusions and special lights in the neonatal units have had a dramatic effect in reducing the number of cases. Perhaps, due to these new treatments, hearing loss which resulted from hyperbilirubinemia is well on its way out as a high risk factor.

Of the 523 children on the high risk register, 27 failed a supplementary behavioral screening (Table 6). The test protocol requires agreement by two independent observers in two of eight trials that they have seen an arousal response by the child. Absence of agreement is considered a preliminary screening failure. As a safeguard, the test procedure is then repeated. Only those children failing the test a second time are classed as true failures on the behavioral screening and are immediately scheduled for appointments at the Nova Scotia Hearing and Speech Clinic for audiological testing. As part of the **routine** procedure, failures are then transported to the Nova Scotia Hearing and Speech Clinic while still inpatients at the Grace Maternity Hospital. When this is not possible, appointments are scheduled as soon as possible after the baby is released from the Grace Hospital. In all cases of behavioral failure, the family physician and the public health department are notified in an effort to inform, to educate, and to enlist their aid in the follow-up habilitation services. The family physician is then expected to involve otology.

TABLE 6

HIGH RISK INFANTS BY CATEGORY

Behavioral Failure: 27 of 523 (5.2%)

Category	Number	Percentage
Affected Family	21	(77.8%)
Congenital Rubella	1	(3.7%)
Defect of ENT	1	(3.7%)
Bilirubinemia	—	—
Small at Birth	4	(14.8%)

All children who fail a second behavioral screening receive a complete audiological evaluation by a clinical audiologist. A modified pediatric test battery is administered which includes the use of white noise, narrow band noise, warble pure tones, and speech in the determination of auditory sensitivity. In addition, impedance audiometry is routine procedure whenever possible. The criteria used for positive behavioral responses are those outlined by Northern and Downs in **Hearing in Children (1978).**

Of the 27 cases (Table 7) seen for audiological evaluation, six were cleared immediately, while seven others were classed as "probably normal hearing" and rescheduled for diagnostic follow-up. The original six remaining cases are, at the time of writing, still pending appointments. This latter group consists of children recently born or a few who left the hospital before being transferred to the clinic.

Of the remaining group of 15 infants seen at the clinic on the first visit, (recall, six were cleared and six were still pending) that is, seven "probably normal hearing" and the eight outright failures, ten have already been seen for a second visit and five remain scheduled for upcoming appointments. Of the ten seen, five were cleared immediately, three were classified as "probably normal hearing", and two failed the audiological evaluation for a

TABLE 7

THE FOLLOW-UP RESULTS OF 27 HIGH RISK INFANTS — BEHAVIORAL FAILURES

	Cleared	Probably Normal Hearing	Fail	Appointments Pending
First Visit	6	7	8	6
Second Visit	5	3	2	5
Third Visit	1	1	1	2

second time. These remaining five infants were then rescheduled for a third visit. The results of that visit have confirmed one hearing loss and two with appointments pending, one of whom is likely to be our second confirmed hearing impaired infant. Undoubtedly, in the remaining twelve cases with appointments pending, there will be at least one more child with a hearing loss.

It should be noted that the Halifax program involved a budget of $6,150 for the year, which included audiological visits but excluded medical-hospital examinations. Taking into account that nearly 5000 babies were screened, the cost suggests that a program of this nature can be implemented at slightly over a dollar per child, an amenable value from an administrative point of view.

At this point in time, the identification aspects of infant screening programs appear to have clearly established their value. Criticism of the program should not focus on the newborn screening, but rather on the audiological — diagnostic follow-up of failures. While the numbers of failures are reasonable, an apparent proscrastination in drawing conclusive statements as to the meaning of "failure" is the weakest link. The most efficiently organized newborn screening program, be it high risk, behavioral, crib-o-gram or some combination thereof, breaks down if the diagnostic efforts behind it are inadequate. A major step designed to strengthen diagnostic efforts may be found in the recommendations of the Saskatoon conference. Specifically, they provided a diagnostic protocol which delienated the procedures for the physical examination of a suspect child into priority categories including Essential, Recommended, and When Indicated (see Table 8). In addition to the procedures indicated for physical examination, the participants at Saskatoon suggested that a comprehensive auditory assessment of the suspect infant should also include the following: (1) an extensive behavioral history by parental report; (2) observation of behavioral responses by appropriate auditory stimuli; (3) visual reinforcement audiometry (where age appropriate); (4) acoustic immittance measurements (includes tympanometry, acoustic reflex, and static compliance); and (5) electric response audiometry as indicated. That diagnostic protocol is now in use in Halifax. Future reports will undoubtedly evaluate their diagnostic effectiveness. Incidently, to our knowledge, the Saskatoon Conference was the first international assembly to encourage the use of brain stem audiometry as a clinical diagnostic assessment.

TABLE 8

STATEMENT RE: PHYSICAL EXAMINATION

WHEREAS, physical examination in the diagnosis of hearing loss in infants adds greatly to the total information about the child and to the understanding of the etiology of the hearing loss,

RESOLVED: A COMPREHENSIVE ASSESSMENT OF ANY CHILD SUSPECT FOR HEARING LOSS SHOULD INCLUDE THESE PROCEDURES:

A. **Essential** to the Assessment
1. Standard pediatric examination
2. Pneumatic otoscopy and/or otomicroscopy
3. Fundoscopic examination
4. Appropriate observations for specific physical abnormalities (see Appendix C)

B. **Strongly Recommended** in the Assessment
1. General Laboratory examinations
2. Appropriate serology examination for toxoplasmosis, rubella, cytomegalovirus, and Herpes
3. Urinalysis
4. Family audiograms

C. Include **When Indicated**
1. Thyroid Function
2. Polytomography of middle and inner ear (Except in established cases of antenatal infections)
3. Electrocardiogram
4. Chromosomal study
5. Fluorescent trepanemal antibody (FTA) — absorption test for syphilis
6. Appropriate testing for Mucopolysaccharidosis

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* From the recommendations of the Saskatoon Conference on Early Diagnosis of Hearing Loss. For reference, see Gerber, S. E. Mencher, G. T., Editors, **Early Diagnosis of Hearing Loss,** Grune and Stratton, New York, 1978.

COMMENTS

While efforts in the development of more accurate screening and diagnostic methods are to be continued, additional support must also be given to the planning and coordination of screening programs in Canada.

Recently, inter-disciplinary efforts in Canada have resulted in the establishment of a Canadian Joint Committee on Childhood Hearing Impairment. The Committee is composed of members of the Canadian Speach and Hearing Association, the Canadian Otolaryngological Society, and the Canadian Pediatric Society. The authors hail this move. The efforts of this inter-disciplinary committee will hopefully lead to successful and conscientious monitoring and guidance of newborn screening programs, ensuring consumer protection while providing good quality health service to the Canadian public.

The move to organize a Joint Committee is further seen as a positive step in the continued development of Canadian professional cooperation. What better opportunity

to help commemorate the International Year of the Child than by focusing professional and public attention on the need for identifying crippling auditory impairments in children. The year 1979 offers our profession a unique opportunity to initiate and develop programs oriented toward the early identification and diagnosis of hearing problems.

As professionals with a common goal, it is not only our responsibility to contribute, but also to be advocates. Now is the time to implement, not procrastinate. The "Year of the Child" should also be the year of action by not only our professional community, but all allied health professions.

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