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INFANT HEARING SCREENING PROGRAM: HIGH-RISK FACTORS FOR HEARING LOSS

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ABSTRACT—It is important to identify early risk factors for hearing loss because there are significant advantages of early identification for hearing improvement and significant consequences for late identification of hearing impairment. A great deal of discussion has evolved as to which children up to age five should be tested and when the testing should occur. Various screening techniques have been developed and these are important for the direction of future research in the diagnosis and management of hearing impairment in infants and children.

KEY WORDS: Risk factors, early identification, screening

In March 1993, the NIH issued a consensus statement on the early identification of hearing impairment in infants and children. The NIH Consensus Development Conference On Early Identification of Hearing Impairments made the following recommendations:

1. Screen all infants admitted to the NICU for hearing loss prior to discharge.
2. Do universal screening of hearing for all infants within the first 3 months of life.
3. Test failures of the evoked otoacoustic (EOAE) emissions test, a suggested model for screening, with auditory brain response (ABR).
4. Make comprehensive intervention and management programs an integral part of a universal screening program.
5. Do not replace ongoing surveillance throughout infancy and early childhood with universal neonatal screening.
6. Educate primary caregivers and primary health care providers on the early signs of hearing impairment.

The results the 1993 NIH consensus statements are intended to advance our understanding of the technology of diagnosing hearing impairment and to be useful to health professionals and the public. In addition to screening all babies in the NICU, the consensus statement strongly

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recommends that universal screening be implemented for all infants within the first 3 months of life.

Previously, the application of the high-risk criteria developed by the Joint Committee on Infant Hearing in 1990 identified approximately 9% of newborns as needing screening. Previous programs only diagnosed 50% of the children subsequently found to have hearing impairment. The primary disadvantage of high-risk screening is that 50% of newborns with hearing defects are not in the high-risk registry group and thus are missed by screening procedures. Presently, 70% of children with acquired hearing impairments are initially identified by parents, and the diagnosis of hearing impairment is not made in children before the age of 2 or 3 years, which is after the critical period for speech and language development.

High-risk groups for hearing impairment include children who have speech delay, congenital infections, meningitis (especially of the bacterial type), significant head trauma, congenital head/neck anomalies, or ototoxicity.

Risk factors included a family history of congenital or delayed-onset childhood sensorineural hearing impairment, as well as congenital infection known or suspected to be associated with sensorineural hearing impairment, such as toxoplasmosis, syphilis, rubella, cytomegalovirus, and herpes (Torch). Cranial-facial anomalies, including morphologic abnormalities of the pinna and ear canal, an absent philtrum, and a low hairline, are additional important risk factors. It has been known for many years that a birth weight of less than 1500 grams (Robertson, Snuve, & Christianson, 1994), as well as hyperbilirubinemia at a level exceeding the indication for exchange transfusion, are significant risk factors. Ototoxic medications, including, but not limited to the aminoglycosides, that have more than a 5-day period of usage put patients at risk for hearing impairment. For example, gentamicin, tobramycin, kanamycin, and streptomycin are all antibiotics implicated in ototoxicity. In addition, these antibiotics given together with loop diuretics (Lasix) are important factors. Bacterial meningitis has an important association with sensorineural hearing loss.

Hearing loss has been associated with severe Apgar score depression at birth. This may include infants with Apgar scores of 0–3 at 5 minutes, those who fail to initiate spontaneous respiration by 10 minutes, and those with hypotonia persisting up to 2 hours. In addition, prolonged mechanical ventilation for a duration of 10 days or more can be considered to put infants at risk for hearing impairment.

In our institution, the incidence of bilateral sensorineural hearing loss in the NICU is 1–3%. The addition of other neonatal high risk criteria (HRC) adds significantly to the identification of hearing impairment. Therefore, it is recommended that all infants admitted to the NICU should be screened for hearing loss prior to discharge. However, use of an HRC limits the population being screened and excludes approximately 50% of infants with hearing impairment.

Previously reported, large series of hearing screenings in the neonatal period include those of Mencher (1972); Feinmesser, Tell, and Levi (1982); and Barr (1976). In all of these series, more than 10,000 children were tested, and a severe sensorineural hearing loss was identified in approximately 1%. False-positive rates in these series were between 0.76 and 2.1%, and false-negative rates ranged from 3.3 (Mencher, 1972) to as low as 1.5% (Barr, 1976).

During the past year (which ended in December 1995), 561 patients were admitted to the Loyola NICU and were evaluated for hearing screening. Fifty-seven babies died before they left the NICU (10.2%); this left 504 as a potential test population. One hundred and sixty-six babies were discharged prior to testing. These children usually had been in an NICU for less than 24 hours, or they were at a gestational age of less than 33 weeks at the time of discharge. We therefore tested 436 babies (86.5%), each of whom were screened at least once. Of the 436 babies tested, 365 (83.7%) passed the initial screening examination. Seventy-one (16.2%)

were referred for further tests. Sixty (84.5%) of those referred were rescreened. Of these patients, 37 (61.7%) passed a subsequent screening examination and 23 (32.4%) failed in the re-screening. All of the screening tests in our institution were done by automated ABR (ALGO^2). Of the 23 babies who failed their second ABR screen, 18 (78.3%) had ENT and audiologic follow-up. Of these, 8 (44.4%) had middle-ear effusion, 3 (16.7%) had normal hearing following removal of ear canal occlusion, 5 (27.8%) had sensorineural hearing loss, and 2 were still inpatients.

Infants with cranial-facial anomalies, a family history of hearing loss, and a diagnosis of intrauterine infection comprise a special high-risk category. These groups should be screened based on the same protocol for follow-up as is the NICU population.

For a further evaluation of the currently known risk factors, it is necessary to review the embryology of the inner and middle ear. The external ear is formed at 4–5 weeks of gestation, but the middle ear forms at a later time, at 6–18 weeks, and the inner ear, at between 3–10 weeks. Thus, all of these events occur in the first trimester. The inner ear, once it is formed at 10 weeks, has almost fully developed. Therefore, most injuries to the fetus occur in the first trimester.

Downs and Silver stated in 1972 that all infants with cranial-facial anomalies have a high incidence of hearing loss. A current classification includes the following mesodermal abnormalities (Grundfast, 1983):

1. Gargoylism syndrome
2. Marfan's chondrodystrophy
3. Alport's syndrome
4. Pendred and Jervell-Lange syndrome

Cranial-facial malformations include the following:

1. Cleft palate with micrognathia and glossoptosis, the so-called Pierre-Robin anomaly
2. Trisomy 21, Down syndrome
3. Trisomy 13, 15 (Patau syndrome)
4. Mandibular-facial dysostosis
5. Treacher Collins syndrome
6. Oculo-auriculovertebral dysplasia (Goldenhar's syndrome)
7. Acrocephalosyndactyly (Alport's syndrome)
8. Gonadal dysgenesis (Turner syndrome)
9. Osteopetrosis (Albers-Schoberg disease)
10. Achondroplasia (Parrot's disease)
11. Mucopolysaccharidosis, Hunter-Hurler syndrome
12. Oral facial digital syndrome, MOHR syndrome, MOHS syndrome
13. Cranial-facial dysostoses (Crouzone's disease)

The aplasias include the Michel type, which is total aplasia of the cochlea; the Mondini type, which indicates a cochlea that has not fully developed, resulting in 1-1/2 or 2 turns; the Scheibe type, which is cochlear saccular degeneration; and lastly, the Alexander Bing type. Examples of the ectodermal type are:

1. Waardenburg's syndrome
2. Usher syndrome
3. Von Recklinghausen syndrome (neurofibromatosis I).

The Mondini type of dysplasia can result in significant bilateral sensorineural hearing loss. These patients have increased pressure in the endolymphatic system, and the cochlea is incompletely formed, with less than the usual 3-1/2 turns. There is significant hair cell degeneration in these patients, which is the cause of the hearing loss. The Scheibe type of loss results in cochlear and saccular degeneration, with the cochlear hair cells absent. The cochlea has the normal number of turns; therefore, the findings on CT scans in Scheibe patients would be normal. The Jervell-Lange and Waardenburg syndromes result in significant hair cell degeneration as well as loss in the vestibular system.

Treacher-Collins disorder can result in significant middle-ear disease with low-set pinnae. Patients with trisomy 13 (Baty, Blackburn, & Carey, 1994) have low-set ears, a cleft lip or palate, and micro-ophthalmia,

and hemangioma and aplasia of the optic nerve are noted. Significant inner and middle ear changes are present. Losses of cochlear hair cells are found, resulting in profound sensorineural hearing impairment. Trisomy 18 (Baty et al., 1994) results in low-set ears, abnormal pinnae, microophthalmia, slanting eyes, and cleft lip. In these patients, significant hair cell damage and cochlear damage are also noted. Viral infections that lead to sensorineural hearing loss at birth include cytomegaloviral disease, which results in loss of the stria vascularis. Measles can cause significant changes in the stria vascularis, saccule, and spiral ganglion, as well as loss of hair cells. Mumps, which usually results in unilateral hearing loss, involves damage to the organ of Corti and changes in the stria vascularis. Rubella usually leads to damage to the stria vascularis.

Hall described the "charge syndrome" (Morgan et al., 1993), which results in coloboma, heart defect, choanal atresia, retarded growth, genital hypoplasia, and ear anomalies. Affected children have low-set pinnae and significant sensorineural hearing loss in approximately 35% of the cases. Facial palsy is not unknown.

Waardenburg syndrome results in lateral displacement of the medial canthus, a prominent nose root, hyperplasia of the medial eyebrows, heterochromia of the iris, congenital deafness, and a white patch in the hair.

Low birth weight (below 1500 grams) has been noted to cause a significant hearing loss. In a large series by Bergman (1985), 9.7% of patients with low birth weight had a significant sensorineural hearing loss. This rate goes up to 28% if the low birth weight is associated with a seizure disorder.

Almost since the development in 1945 of the first aminoglycoside, streptomycin, this class of drugs has been associated with hair cell damage in the cochlea. In addition, secondary spiral ganglion cell damage has been noted. The ototoxic aminoglycosides are currently considered to be streptomycin, kanamycin, gentamicin, tobramycin, viomycin, and amikacin. Outer hair cell loss is the usual early sign of aminoglycoside damage, especially at the basal turn. Later changes in the inner hair cells, apical turns, and loss of the stria vascularis and ganglion cells are noted in these patients. At risk for hearing impairment in the neonatal group are children who receive an aminoglycoside antibiotic for more than 5 days for any reason; they, too, have significantly elevated peak and trough levels of these drugs during the antibiotic usage. Ototoxicity in neonates is rare and ototoxicity in neonates who receive perinatal aminoglycosides is equally rare. Aminoglycoside toxicity results in outer hair cell damage. Transplacental transport of aminoglycosides has been well known. Gentamicin and kanamycin are drugs which are commonly used for treatment in the neonatal period. Also, 33% of these drugs can be transported across the placenta (Siegal & McCraken, 1981). Finitzo-Hieber (1979) reported an incidence of roughly 11% of either vestibular or cochlear loss in neonates who received gentamicin or kanamycin.

There is no known permanent ototoxicity from salicylates, and there is no histopathologic correlate or permanent auditory loss. Numerous cases of transient hearing loss have been noted. However, this type of abnormality in the neonatal group is very unusual.

Cisplatinum is a drug which is not frequently used in the neonatal period, but can be used in young adults for chemotherapy of cancer. Outer hair cell loss has been described in such patients, and the usual correlate is high-frequency hearing loss. Incidences up to 20% have been noted by numerous authors.

Sensorineural hearing loss after bacterial meningitis is well known. In a prospective study, an incidence of 6% was noted. The usual causative organisms are *Hemophilus influenzae* and meningococci. Fortunately, the former is becoming rare because a *Hemophilus influenzae* vaccine is available. The meningococcal meningitis organism can cause permanent ossification of the cochlea.

The management of otitis media with effusion in young children is important in the neonatal age group, and otitis media should be considered an important risk fac-

tor for hearing loss. In a large study by Brookhauser done at Boys' Town (unpublished data), he prospectively followed 179 children who had early otitis media and had a ventilation tube inserted before the age of 2 years. During an average follow-up period of 4.9 years, there were 8.2 episodes of otitis media per patient after the tube was inserted. The average patient had 17.4 courses of an antibiotic, with six courses of ototopicals that included medication containing neomycin. A high percentage of these children are at risk of developing high-frequency sensorineural hearing loss. Therefore, middle ear effusion has to be considered a risk factor for possible sensorineural hearing impairment.

Otitis media is the most common diagnosis in children under the age of 14 years, and there was a 150% increase in office visits for this disorder from 1975 to 1990. Currently, there are 24 million office visits by children with otitis media who are under the age of 15 years. For children under the age of 2 years, the increase in office visits in this period has increased by 224%. Otitis media at birth is recognized with difficulty. Up to the age of 3 years, it can involve early syntax and semantic problems and general inattention to language. Up to the age of 5 years, the habit of inattention can become more frequent, and, by the time the patients grow older, this can involve poor narrative and discourse skills.

There has been much discussion as to what is the purpose of doing screening and testing. Robinette (1994), at the Mayo Clinic, estimated that the average cost of testing children from kindergarten to 12th grade, in the United States, is approximately $44,000. If a diagnosis of deafness in a child is delayed and the child has to be in a self-contained classroom, the cost, on the average, is $126,000, and the cost of education in a residential school is $429,000 from kindergarten through age 12.

CONCLUSIONS

The following conclusions can be made:

1. There is approximately a 1–3% incidence of significant hearing impairment that can be diagnosed in neonatal intensive care units.
2. Routine use of the high risk registry only results in a detection rate of 50%.
3. The average age presently of diagnosing significant hearing loss in children is 2 1/2. Children with cranial-facial abnormalities are at high risk for having sensorineural hearing impairment. Hearing impairment can be associated with low apgar scores and low birth weight. Children who have intrauterine exposure to toxoplasmosis, syphilis, rubella, cytomegalovirus and herpes (Torch) are at high risk for hearing impairment in the neonatal period.

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ARTICLE FIVE

SELF-ASSESSMENT QUESTIONS

1. The use of a high-risk registry for neonatal testing can identify the following:
   (a) 10% of patients with significant hearing loss.
   (b) 30% of patients with significant hearing loss.
   (c) 50% of patients with significant hearing loss.
   (d) 70% of patients with significant hearing loss.
   (e) 100% of patients with significant hearing loss.
2. The average age for diagnosis of hearing loss in children is:
   (a) 2 months of age.
   (b) 6 months of age.
   (c) 1 year of age.
   (d) 2 years of age.
   (e) 2.5 years of age.
3. The incidence of hearing loss in neonatal population from ototoxic medications is:
   (a) less than 10%.
   (b) less than 30%.
   (c) less than 50%.
   (d) less than 70%.
   (e) less than 100%.
4. True or False: Cranial-facial anomalies can be associated with patients with significant hearing loss.
5. Which group of patients would have a higher incidence of neonatal hearing loss?
   (a) Neonatal intensive care unit
   (b) Well-baby intensive care unit