

35. Screening for Hearing Impairment

RECOMMENDATION

Screening older adults for hearing impairment by periodically questioning them about their hearing, counseling them about the availability of hearing aid devices, and making referrals for abnormalities when appropriate, is recommended. There is insufficient evidence to recommend for or against routinely screening older adults for hearing impairment using audiometric testing (see *Clinical Intervention*). There is also insufficient evidence to recommend for or against routinely screening asymptomatic adolescents and working-age adults for hearing impairment. Recommendations against such screening, except for those exposed to excessive occupational noise levels, may be made on other grounds (see *Clinical Intervention*). Routine hearing screening of asymptomatic children beyond age 3 years is not recommended. There is insufficient evidence to recommend for or against routine screening of asymptomatic neonates for hearing impairment using evoked otoacoustic emission testing or auditory brainstem response. Recommendations to screen high-risk infants may be made on other grounds (see *Clinical Intervention*). Clinicians examining infants and young children should remain alert for symptoms or signs of hearing impairment.

Burden of Suffering

Prevalence estimates for hearing impairment vary depending on age and the criteria used to define the various causal conditions.¹ For severe congenital and prelingually acquired losses, estimates range from 1 to 3/1,000 live births.^{1-5,12,22,30} Moderate and severe hearing losses in early infancy are clearly associated with impaired language development.⁶⁻⁸ Factors that increase the risk for congenital or delayed-onset sensorineural hearing impairment include family history of hearing impairment, congenital or central nervous system infections, ototoxic drug exposure, prematurity, congenital head and neck deformities, trauma, and several other factors associated with admission to an intensive care nursery.^{2,5,9} Chronic and recurrent acute otitis media is commonly associated with temporary hearing loss in infants and school-aged children. Prevalence rates for otitis media are 12% before age 3, 4-18% for ages 4-5, and 3-9% for ages 6-9 years.¹⁰ At any given time, about 5-7% of children ages 5-8 have a 25-dB hearing

loss, usually a self-limited complication of otitis media with effusion.¹¹ Only a small proportion of episodes of otitis media occurring in school-aged children result in serious long-term complications, usually due to chronic middle ear effusion or previously undetected sensorineural deficits.¹¹ The uncertainties of the population occurrence rates and causes of infant and childhood hearing loss have been emphasized.⁸¹

Hearing impairment creates further difficulties in adulthood. Adult hearing impairment has been correlated with social and emotional isolation, clinical depression, and limited activity.^{2,12,13,16} Hearing loss acquired between adolescence and age 50 may be due to relatively uncommon causes such as Ménière's disease, trauma, otosclerosis, ototoxic drug exposure, and eighth cranial nerve tumors. Noise-induced hearing loss is a common cause of sensorineural hearing impairment in this age group. This is particularly true for the estimated 5 million Americans with occupational exposure to hazardous noise levels.¹⁴ The prevalence of hearing impairment increases after age 50 years, with presbycusis being the most important contributor to this increase. Approximately 25% of patients between ages 51 and 65 years have hearing thresholds greater than 30 dB (normal range being 0–20 dB) in at least one ear.¹⁵ An objective hearing loss can be identified in over 33% of persons aged 65 years and older and in up to half of patients ages 85 years and older.^{16,17} Older persons with hearing impairment are particularly prone to suffering the associated social and emotional disabilities described earlier.^{18,19}

Accuracy of Screening Tests

Multiple methods of audiologic testing are potentially suitable for evaluating possible hearing deficits. Test selection is usually dictated by patient age and occasionally by type of hearing loss in question (i.e., conductive vs. sensorineural). Cooperative children and adults are usually tested with pure-tone audiometry. With pure-tone thresholds in audiometric test booths used as a reference criterion, this technique has a reported sensitivity of 92% and a specificity of 94% in detecting sensorineural hearing impairment.³⁷ Comparable results have been obtained in recent studies using hand-held audiometers.^{16,37} Audiometric results are, however, subject to error due to improper technique, background noise in the test area, and unintentional or intentional misreporting by the subject.^{11,38} Efforts have been made to devise a sufficiently accurate test utilizing the pure-tone audiometer that is briefer and less costly than standard pure-tone audiometry, but clinical efficacy is not yet confirmed.³⁵

Evaluation of neonates and infants below the age of 2–3 years with audiometry is more difficult or not feasible because it depends on developmental ability; it therefore usually requires some form of

electrophysiologic and/or behavioral testing. Auditory brainstem response testing (ABR) is currently viewed as the standard for physiologic testing in infancy and the most accurate method available for determining hearing function.^{1,2,5,20} Sensitivity rates have been reported to be 97–100% and specificity rates to be 86–96% in comparison with behavioral testing measures.^{2,5,21}

In order to detect congenital or postnatally acquired hearing loss, some form of newborn screening performed prior to hospital discharge has been recommended as most efficacious for ensuring early identification and proper follow-up and treatment of hearing loss.^{5,6,20,22} As a universal *screening* test, ABR (or modified ABR) is probably unsuitable because of the need for costly equipment and trained operators in all community hospitals and birthing centers. Another screening modality for neonates is the high-risk register (HRR), a specific list of clinical risk factors associated with higher rates of neonatal and infant hearing impairment.^{23,24} Those who meet criteria then undergo more objective hearing evaluation, usually ABR. The HRR identifies 50% or more of unselected infants with hearing loss²⁴ and 75–80% of hearing-impaired neonates in the intensive care nursery.⁵ Behavioral testing techniques have also been used for infant hearing screening, including the “crib-o-gram,” auditory response cradle, and distraction testing.^{29–32} The limited specificity and sensitivity of behavioral testing, as well as specialized equipment and training requirements, renders these methods less desirable than physiologic testing procedures.

Evoked otoacoustic emission (EOE) testing is a relatively new screening method suitable for neonatal and infant screening.^{22,25–28} Otoacoustic emissions are sounds generated by normal cochlear hair cells and detectable with relatively simple instrumentation.⁶⁷ Data concerning normative standards and reproducibility are now becoming available.^{68–73} Using a cutoff of 30 dB to designate hearing impairment, EOE testing has an overall agreement rate with ABR of 91%, with a sensitivity of 84% and specificity of 92%.^{74,75} Statewide neonatal auditory screening programs have been devised using EOE, and the logistical issues of operating such a program have been described.⁸⁶ Studies of EOE testing suggest a high rate of false-positive screens relative to true-positive results, which would be expected when testing for a low-prevalence condition, and some failures of testability, necessitating retesting with EOE and ABR.⁸³ In one screening study, only 15% of positive EOE screening tests were confirmed on repeat EOE testing 4–6 weeks later; the proportion of infants with confirmed screening tests who actually had hearing loss is unknown, since the results of diagnostic follow-up tests were not available.⁸⁸ Based on the authors' estimates of true population prevalence, more than 90% of the positive neonatal EOE screening tests were false positives. Problems such as ambient noise in the newborn nursery and other factors that affect the techni-

cal conduct of EOE require solution before this technique can be applied widely.^{83,84}

The majority of children with congenitally or neonatally acquired losses are identified by age 4–5 years.¹ Hearing loss in the preschool and school-aged group is largely related to acute or chronic otitis media with effusion (OME), of which the majority of cases resolve uneventfully.¹¹ Routine audiometry can often detect the mild conductive hearing loss associated with OME.³³ Accuracy for detecting hearing loss associated with OME by audiometry may be variable in this age group, however, because of the mild and changing nature of the conductive loss, varying patient cooperation, conditions that make testing difficult (e.g., mental retardation), and the fact that middle ear conduction deficits may be superimposed on previously undetected sensorineural hearing loss due to other conditions.

Routine screening of working-age adolescents and adults is usually limited to those in high-risk occupations involving exposure to excessive noise levels. Among older persons, however, in whom the rate of hearing impairment is high, recommended screening methods for detecting hearing loss have included written patient questionnaires, clinical history-taking and physical examination, audiometry with a hand-held device, and simple clinical techniques designed to assess for the presence of hearing impairment.^{15,16,35,36} These screening tests have not been fully evaluated, however. For example, the whispered voice test is one simple clinical technique used to assess hearing. Reported sensitivities and specificities have been 70–100% using pure-tone audiometry as the reference standard, but there are inadequate data on interobserver variability.¹⁶ The free-field voice, tuning fork, and finger rub tests have been criticized on similar grounds.¹⁶ Self-assessment questionnaires to identify hearing impairment probably represent the most rapid and least expensive way to screen for hearing loss in the adult. Depending on audiometric criteria, these questionnaires are reported to be 70–80% accurate for identifying patients with hearing loss defined by pure-tone audiometry.^{16,36,82}

Effectiveness of Early Detection

Assessing the effectiveness of screening for hearing impairment depends upon the evidence that (a) hearing loss leads to decreased function and affects the quality of life, (b) screening leads to earlier detection of hearing abnormalities than spontaneous clinical presentation or observation, (c) various forms of hearing loss can be treated effectively, and (d) effective treatment leads to improved function and well-being.

Theoretically, the greatest benefit from hearing screening comes from detection of moderate to severe hearing impairment between birth and age 3 years. Auditory stimuli during this period appear to be critical to de-

velopment of speech and language skills,^{2,46} although other factors undoubtedly also play an important role. If screening for hearing deficits is performed near the time of birth, followed by definitive diagnosis, the choice of treatment and treatment success will depend on the etiology of the hearing loss. For sensorineural impairment, depending on the degree of loss, treatment may range from amplification in the majority of cases to cochlear implantation in profoundly deaf children. In both cases, speech and hearing therapy has been promoted as a key component of treatment and the efficacy of such therapy has been claimed.^{75,76} Cochlear implant technology continues to evolve for treatment of profound deafness in children. Several studies have demonstrated improved language development and communication skills in deaf infants following cochlear implantation.^{47,48} Several nonrandomized, prospective studies have also demonstrated superior communication performance in prelingually deafened children who received implants as compared to similar children using more traditional tactile or acoustic hearing aids.^{49,75,76}

Although the benefits of various treatments for hearing loss seem manifest, no controlled clinical trials have evaluated the effect of early screening on long-term functional and quality-of-life outcomes. Rather, studies of treatment efficacy are generally observational and retrospective, consisting of clinical series or case-control studies of highly selected patients, often with heterogeneous causes of hearing loss, and incompletely defined treatment regimens or protocols of uncertain compliance. Additionally, important confounders such as other patient characteristics (e.g., race or ethnic group, socioeconomic status, level and laterality of hearing loss, the presence of co-morbidity, disability, or developmental delay due to various causes), family characteristics, and the presence and nature of other therapeutic interventions are often not considered in the analysis. Thus, despite widespread professional opinion of general treatment efficacy, much more information is needed on the existence and level of treatment protocol efficacy. In many instances, however, it may understandably be deemed inappropriate to withhold any customary type of treatment in the research setting despite the limited evidence of treatment efficacy.⁸⁵

Conductive hearing loss in the preschool-age group is most commonly due to self-limited cases of otitis media with effusion. Multiple studies have concluded that hearing impairment in infancy due to chronic or recurrent otitis media with effusion can impair language development.³⁹⁻⁴¹ Although these studies have come under methodologic criticism,^{42,43} several authors believe that available evidence is adequate to substantiate this relationship.^{8,44} Auditory thresholds in hearing-impaired children can be improved through amplification with hearing aids and frequency modulation radio devices. Auditory and language training can also improve communication skills.^{12,75,76} While early detection and treatment of such

hearing loss would therefore appear to be beneficial, there are no controlled studies comparing outcome of hearing-impaired persons identified through screening to those not screened. The fewer than 5% of infants with chronic otitis who do not respond spontaneously or with medical management are at further risk for more significant pathology including middle ear fibrosis or adhesions and cholesteatoma.¹¹ Myringotomy and pressure-equalizing tube placement can resolve the conductive loss and prevent reaccumulation of middle ear effusion.⁵⁰ No randomized or otherwise well-controlled study exists, however, demonstrating that infants or young children screened *with routine hearing tests* for chronic middle ear disease have a better outcome than those not screened in this manner. Nevertheless, if hearing loss is detected as part of the routine diagnosis or management of chronic OME, management of either sensorineural or conductive losses by standard regimens is indicated.

In older children, otitis media with effusion is responsible for the majority of hearing loss identified through screening.^{3,11} As is the case in infants and toddlers, however, there is little evidence that asymptomatic children receiving hearing screening have better functional outcomes than those not screened. In fact, several studies of preschool and school-aged children who underwent audiometric screening demonstrated no significant difference in future audiometric performance between screened and unscreened children⁵¹ nor any preventive benefit from screening.⁴ Most hearing loss detected under these circumstances is self-limited and related to acute otitis media with effusion that resolves spontaneously within 6–8 weeks.^{3,11} Since the critical period of language development has passed at this age, these individual episodes would appear to have little impact on educational performance. Studies have been unable to provide consistent evidence that clinical interventions for chronic OME (e.g., antibiotics, myringotomy, tympanostomy tubes) are able to achieve sufficient long-term improvement in hearing and language skills to justify the risk of complications.^{41,43,51,53} A small portion of children routinely screened for hearing loss will demonstrate a protracted hearing impairment due to previously undetected, less severe, sensorineural losses as well as chronic and recurrent middle ear disease. These children may be at risk for educational and language problems,^{1,52,53} although the evidence for this contention has been challenged.⁴²

For adults between the ages of approximately 18 and 50 years, unrecognized hearing impairment is uncommon except for high-risk groups such as persons in occupations at risk for noise-induced hearing loss.^{54,55} The incidence of hearing impairment, predominately due to presbycusis, rises quickly beyond age 50, however. No controlled study has proven the effectiveness of screening for hearing impairment in the adult population.¹⁶ Two reviews cite numerous studies documenting the benefits of

hearing amplification in these patients.^{16,55} A 1990 randomized controlled trial demonstrated a measured improvement in social, cognitive, emotional, and communication function from hearing aid use in a group of elderly veterans with previously documented hearing loss.⁵⁶ The issue of patient compliance with recommendations to obtain hearing amplification has been raised as it relates to hearing screening,^{15,55} but compliance rates of close to 40–60% can be achieved in some settings.¹⁶ Patients receiving hearing aids have demonstrated improvement in communication and social function, as well as emotional status.⁵⁶

Recommendations of Other Groups

The Joint Committee on Infant Hearing 1994 Position Statement, developed and approved by the American Speech-Language-Hearing Association (ASHA), American Academy of Otolaryngology-Head and Neck Surgery, American Academy of Audiology, American Academy of Pediatrics (AAP), and Directors of Speech and Hearing Programs in State health and welfare agencies, endorses the goal of universal detection of infants with hearing loss before 3 months of age.⁵⁹ When universal screening is not available, the committee recommends testing infants with indicators associated with sensorineural and/or conductive hearing loss, by 3 months of age. The high-risk indicators are similar to those described under *Clinical Intervention* (see below). The Bright Futures guidelines recommend hearing screening for all newborns prior to 3 months of age.⁶⁰ The National Institutes of Health recommends universal screening of all infants before age 3 months using evoked otoacoustic emission testing.⁷⁷ The Canadian Task Force on the Periodic Health Examination recommends regular assessment of hearing during well-baby visits during the first 2 years of life using parental questioning and the clap test.⁸⁰ The American Academy of Family Physicians (AAFP) recommends screening high-risk infants for hearing impairment; high-risk criteria are similar to those described under *Clinical Intervention* (see below).⁶⁵ The recommendations of the AAFP are currently under review.

The AAP recommends periodic historical inquiry regarding hearing throughout infancy and childhood and objective testing at ages 3, 4, 5, 10, 12, 15, and 18.⁶¹ The Bright Futures guidelines recommend hearing screening at ages 3–6, 8, and 10, and yearly from ages 11–21 if the adolescent is exposed to loud noises, has recurring ear infections, or reports problems.⁶⁰ In 1990, ASHA reaffirmed its recommendation for annual audiometry for all children functioning at a developmental level of 3 years through grade 3 and for all children in high-risk groups.^{62,63} ASHA also added tympanometry to their screening protocol for this age group as well as for any other patient undergoing screening audiometry up to age 40.

The Canadian Task Force on the Periodic Health Examination recommends against routine preschool screening for hearing problems.⁸⁰ The AAFP does not recommend routine hearing screening in children after age 3 years;⁶⁵ this recommendation is under review.

Recommendations for adults vary and also depend on age. Although ASHA proposes a screening protocol applicable to young adults, no guidelines are given regarding exactly who should be screened or what are optimal times for screening.⁶² In the U.S., federal law mandates baseline and annual audiometry for workers of any age exposed to hazardous noise levels.¹⁴ The Canadian Task Force recommends risk assessment for hearing loss by history and physical examination at age 16 and thereafter during clinical visits for any other reason.⁶⁴ The AAFP recommends screening for hearing impairment in adolescents and adults regularly exposed to excessive noise in recreational or other settings;⁶⁵ this recommendation is under review.

The Institute of Medicine recommended audiometric testing once each during ages 40–59, 60–74, and 75 and over.⁶⁶ The Canadian Task Force on the Periodic Health Examination recommends screening the elderly for hearing impairment, using a single question about hearing difficulty, whispered-voice out of the field of vision, or audioscope.⁸⁰ The AAFP recommends evaluation of hearing in persons aged 65 years and older, and hearing aids for patients found to have hearing deficits;⁶⁵ this recommendation is under review.

Discussion

While congenital hearing loss is a serious health problem associated with developmental delay in speech and language function, there is little evidence to support the use of routine, universal screening for all neonates. Although screening methods have reasonable sensitivity and specificity, a substantial number of infants will be misclassified because the prevalence of hearing impairment is low. Also, screening technology is evolving, and the costs and feasibility for universal application are not fully known. Most importantly, the evidence for efficacy of early intervention is incomplete. There have been no controlled clinical trials designed to test whether devices or complex protocols lead to superior speech and language outcomes in screened children. For older children, good quality evidence suggests little benefit from screening, while for adolescents and young and middle-aged adults there is limited evidence evaluating hearing impairment and treatment. Many older adults with clinical complaints of hearing loss or documented hearing deficits, however, benefit from hearing aids or other forms of amplification.

Treating deaf children with modalities such as cochlear implants has stimulated ethical concerns from some advocates for the deaf, a full dis-

cussion of which is beyond the scope of this chapter. Attitudes held by both physicians and by society toward deaf individuals have changed over time, and various associations now offer support for individuals affected by deafness, promote their full participation in society, and seek to preserve and expand deaf awareness, deaf culture, and deaf heritage efforts.⁸⁷

CLINICAL INTERVENTION

Screening older adults for hearing impairment by periodically questioning them about their hearing, counseling them about the availability of hearing aid devices, and making referrals for abnormalities when appropriate, is recommended (“B” recommendation). The optimal frequency of such screening has not been determined and is left to clinical discretion. An otoscopic examination and audiometric testing should be performed on all persons with evidence of impaired hearing by patient inquiry. Although hand-held devices for audiometric testing (audioscopes) are also sensitive screening tools for hearing deficits, patient inquiry is likely to be a more rapid and less expensive way to screen for hearing loss in older adults. There is therefore insufficient evidence to recommend for or against routinely screening older adults for hearing deficits using audiometry testing (“C” recommendation).

There is insufficient evidence to recommend for or against routinely screening asymptomatic adolescents and working-age adults for hearing impairment (“C” recommendation). Recommendations against such screening, except for those exposed to excessive occupational noise levels, may be made on other grounds, including low prevalence, high cost, and the likelihood that hearing deficits in these individuals will present clinically. Screening of workers for noise-induced hearing loss should be performed in the context of existing worksite programs and occupational medicine guidelines.

Routine hearing screening of asymptomatic children beyond age 3 years is not recommended (“D” recommendation). It is recognized, however, that such testing often occurs outside the clinical setting. When this occurs, abnormal test results should be confirmed by repeat testing at appropriate intervals, and all confirmed cases identified through screening referred for ongoing audiologic assessment, selection of hearing aids, family counseling, psycho-educational management, and periodic medical evaluation.

There is insufficient evidence to recommend for or against routine screening of asymptomatic neonates for hearing impairment using evoked oto-acoustic emission (EOE) testing or auditory brainstem response (ABR) (“C” recommendation). Recommendations to screen high-risk infants may be made on other grounds, including the relatively high

prevalence of hearing impairment, parental anxiety or concern, and the potentially beneficial effect on language development from early treatment of infants with moderate or severe hearing loss. For many high-risk conditions, hearing testing is commonly considered to be part of diagnostic evaluation and management. Risk factors for congenital or perinatally acquired hearing loss include family history of hereditary childhood sensorineural hearing loss; congenital perinatal infection with herpes, syphilis, rubella, cytomegalovirus, or toxoplasmosis; malformations involving the head or neck (e.g., dysmorphic and syndromal abnormalities, cleft palate, abnormal pinna); birth weight below 1,500 g; bacterial meningitis; hyperbilirubinemia requiring exchange transfusion; severe perinatal asphyxia (Apgar scores of 0–4 at 1 minute or 0–6 at 5 minutes, absence of spontaneous respirations for 10 minutes, or hypotonia at 2 hours of age); ototoxic medications; and findings associated with a syndrome known to include hearing loss. ABR testing may be useful for all infants who meet at least one of these high-risk criteria or for those who fail EOE testing. High-risk infants should ideally be screened prior to leaving the hospital after birth, but those not tested at birth should be screened before age 3 months with the goal being to initiate rehabilitation by age 6 months as clinically indicated. Clinicians examining any infant or young child should remain alert for symptoms or signs of hearing impairment, including parent/caregiver concern regarding hearing, speech, language, or developmental delay.

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