

# Hearing Screening

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## Objectives

After completing this article, readers should be able to:

1. Describe the prevalence and burden of hearing impairment.
2. Delineate the "gold" and "proxy" gold standards for measuring hearing impairment.
3. Describe the screening tests currently used to detect newborn hearing impairment.
4. Identify the advantages and disadvantages of universal newborn hearing screening.
5. Describe appropriate hearing screening methods after infancy.

## Introduction

A number of authors suggest that the critical period for development of the auditory system and speech commences in the first 6 months of life and continues through 2 years of age. Specific linguistic experience in the first 6 months of life, before meaningful speech begins, affects infants' perception of speech sounds and their capacity to learn. Moderate-to-severe hearing impairment in the first year of life is believed to compromise speech and language acquisition as well as cognitive and social development. Mild or unilateral hearing deficits also are considered to affect language development and behavior of children. Early intervention (following detection of hearing impairment in those younger than 3 months of age) reduces the age for access to effective medical and habilitative intervention for many infants. Intervention for those younger than 6 months of age also is believed by many to improve speech and language development and cognitive outcomes, diminishing the need for special education and improving quality of life. The evidence for these effects currently is limited in quantity and quality; most studies are retrospective and have significant limitations.

The prevalence of moderate through profound hearing impairment<sup>‡</sup> in newborns, including both sensorineural (SNHL) and conductive hearing loss (CHL), is in the range of 1 to 3/1,000. Previously published reports are believed to reflect an underestimate of the true prevalence. If infants who have mild SNHL are included, the prevalence would at least double. Furthermore, the prevalence of hearing impairment is increased substantially in newborns who have specific risk indicators (Table 1).

Universal newborn hearing screening (UNHS) programs are mandated in at least 32 states in the United States and are being initiated in 20 health districts in the United Kingdom. Prior to these developments, the average age of identification of hearing impairment was about 30 months, with individual cases being diagnosed at a much later age. In addition, children who had mild or moderate hearing losses frequently

## Abbreviations

<b>AABR:</b>	automated auditory brainstem response
<b>ABR:</b>	auditory brainstem response
<b>CHL:</b>	conductive hearing loss
<b>CPA:</b>	conditioned play audiometry
<b>DPOAE:</b>	distortion product otoacoustic emissions
<b>EHDI:</b>	Early Hearing Detection and Intervention
<b>OAE:</b>	otoacoustic emissions
<b>SNHL:</b>	sensorineural hearing loss
<b>TEOAE:</b>	transient evoked otoacoustic emissions
<b>UNHS:</b>	universal newborn hearing screening
<b>VRA:</b>	visual reinforcement audiometry

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Dr Hyde has a patent application (coapplicant with Dr Y. Sininger, House Ear Institute, Los Angeles, CA) on a statistical method of automatic hearing screening in infants.

<sup>‡</sup> Hearing loss severity is defined as: mild, 6 to 40 dB; moderate, 41 to 55 dB; moderately severe, 56 to 70 dB; severe, 71 to 90 dB; profound,  $\geq 91$  dB hearing level (HL)

### Table 1. Risk Indicators Associated With Neonatal Hearing Impairment

- An illness or condition requiring admission of  $\geq 48$  h to a neonatal intensive care unit
- Stigmata or other finding associated with a syndrome known to include a sensorineural or conductive hearing loss
- Family history of permanent childhood sensorineural hearing loss
- Craniofacial abnormalities, including those that have morphologic abnormalities of the pinna and ear canal
- In utero infection, such as cytomegalovirus, herpes, toxoplasmosis, or rubella

Source: Joint Committee on Infant Hearing, Year 2000 Position Statement: Principles and Guidelines of Early Hearing Detection and Intervention Programs

were not identified until school age. Infants may be referred for testing when a risk indicator for hearing impairment is present (Tables 1 and 2) or if parents, health workers, or other caregivers suspect a hearing deficit. However, it is widely reported that targeted high-risk screening can identify at best about 50% of children who have significant prelingual hearing impairment.

A recent systematic review of the evidence supporting UNHS has concluded that modern screening tests for hearing impairment can improve identification of newborns who have permanent hearing impairment, but the efficacy of UNHS to improve long-term language outcomes remains uncertain. That review formed the basis for a recent rating by the United States Preventive Services Task Force of the evidence for effectiveness of UNHS programs as “inconclusive.” The review identifies some important gaps and deficiencies in current knowledge about newborn hearing screening, and it is helpful in identifying a research agenda to place screening programs on a more evidentially secure footing. The findings of the review reflect a narrow focus on the scientific quality of evidence relating to specific questions. Such reviews are useful, but they do not encompass a broader picture of poor current identification and a substantial current burden of unidentified and unmanaged hearing disorders in infancy. The review also does not reflect the values that society may place on improvements in early identification, audiologic evaluation, information to parents, access to medical interventions, ability to hear, and

### Table 2. Risk Indicators Associated With Progressive Or Delayed-onset Hearing Loss

- Parental or caregiver concern regarding hearing, speech, language, or developmental delay
- Family history of permanent childhood hearing loss
- Stigmata or other findings associated with a syndrome known to include a sensorineural or conductive hearing loss or eustachian tube dysfunction
- Postnatal infections associated with a sensorineural hearing loss, including bacterial meningitis
- In utero infection, such as cytomegalovirus, herpes, toxoplasmosis, rubella, or syphilis
- Neonatal indicators, specifically hyperbilirubinemia at a serum level requiring exchange transfusion, persistent pulmonary hypertension of the newborn associated with mechanical ventilation, and conditions requiring the use of extracorporeal membrane oxygenation
- Syndromes associated with progressive hearing loss (eg, neurofibromatosis, osteopetrosis, Usher syndrome)
- Neurodegenerative disorders (eg, Hunter syndrome) or sensory motor neuropathies (eg, Friedrich ataxia, Charcot-Marie-Tooth syndrome)
- Head trauma
- Recurrent or persistent otitis media with effusion for at least 3 mo

Source: Joint Committee on Infant Hearing, Year 2000 Position Statement: Principles and Guidelines of Early Hearing Detection and Intervention Programs

other interventions designed to improve communication skills in infancy. These broader considerations are beyond the scope of pure evidence review, but they are within the purview of several multidisciplinary position statements that have been developed nationally and internationally in recent years that generally endorse UNHS.

### Diagnostic Tests: The “Gold Standard” and the “Proxy Gold Standard”

The gold standard for assessing hearing deficit in infants older than 6 months of age comprises behavioral tests that rely on operant conditioning, such as visual reinforcement audiometry (VRA). This involves testing an infant’s response to specific tones projected within a soundproof room from different directions. When performed correctly, VRA can yield accurate audiometric thresholds in children as young as 6 months of age who have normal neurologic development. However, in

younger infants and in those who have developmental delay or certain physical disabilities, behavioral tests of any type are unreliable and have a low specificity.

The auditory brainstem response (ABR) is the only test of auditory function accepted as a proxy gold standard for assessment of hearing sensitivity in newborns and infants. The ABR is an electrical waveform (an evoked potential) generated by neuronal activity in the auditory nerve and brainstem pathways following a transient sound such as a click. Its registration (via skin electrodes, electroencephalographic amplifiers, and computer averaging) does not require a behavioral response. The intensity and rate of stimulation primarily determine the response size, latency, and morphology. The presence of a detectable ABR is considered a proxy for perception of sound. The lowest stimulus level that evokes a detectable ABR is an estimator of the true perceptual threshold for various sounds. There is a high correlation between hearing impairment in infants and alteration in the ABR pattern. Overall, ABR testing provides a reasonable evaluation of thresholds over a broad range of hearing impairments and permits differentiation between CHL and SNHL. Many studies have demonstrated that the click ABR in early infancy is a good predictor of pure tone auditory thresholds in the 2,000 to 4,000 Hz range, although a more technically correct interpretation is that the ABR reflects the best pure tone threshold in the range of 500 to 4,000 Hz. Skill and experience are required for valid and efficient ABR testing and interpretation of results.

### Screening Tests: Advantages and Disadvantages

Current screening technologies include: automated auditory brainstem response (AABR), transient evoked otoacoustic emissions (TEOAE), and distortion product otoacoustic emissions (DPOAE). The screening device objectively and automatically detects the response to sound (either an evoked potential or an otoacoustic emission), and the outcome is designated as a “pass” or “fail” (“refer”) by the automated analyser. In the screening mode, the three screening methods indicate presence or absence of a response at a specific stimulus level; they neither quantitatively estimate the severity of the hearing impairment nor distinguish conductive from sensorineural hearing impairment.

The AABR is a modification of conventional ABR testing, usually involving a single stimulus level and automated response detection. Typically, a series of click stimuli at a level of approximately 30 to 40 dB nHL (normal hearing level is the threshold of audibility of the

clicks in normal young adult listeners) is delivered. The electrical signals from at least three or four electrodes on the head are amplified and computer-processed in an attempt to extract the minute ABR from the ongoing electromyogenic and electroencephalic activity that is unrelated to the stimulus. The key technique is averaging of the waveforms recorded after several thousand stimuli, delivered very rapidly. The resultant waveform is tested statistically to determine whether it is a genuine evoked response or merely random electrical noise.

Using statistical response detection eliminates the need for waveform interpretation by a highly trained professional. This is important both to reduce screening manpower costs and to increase the accuracy and consistency of response detection. A variety of automated ABR screening instruments is commercially available.

TEOAE are elicited by click stimuli delivered by a probe transducer in the external ear canal. The emission or “echo” from the inner ear is a very faint sound with a complex waveform that is recorded by a sensitive, miniature microphone in an external ear probe assembly. Some method of signal enhancement, such as signal averaging, is necessary to distinguish the otoacoustic emissions (OAE) from ambient sound.

TEOAE presence implies integrity of sound transmission through the outer and middle ear structures and functional integrity of the outer hair cells, which are the primary sensory transducers with the organ of Corti in the cochlea and are believed to be the site of emissions generation. Low ambient noise level, a clear external auditory meatus and middle ear, probe stability, appropriate choice of stimulus intensity, later postnatal testing, and cochlear maturation all improve the specificity of TEOAE screening. Because the original patent on the TEOAE method only expired recently, the variety of commercially available TEOAE screening devices is limited, although this is changing rapidly.

DPOAE are an alternative form of cochlear emission, also having their origin in the outer hair cells of the cochlea. The stimulus is two simultaneous sustained pure tones (primary frequencies of  $f_1$  and  $f_2$ ) typically in the 50 to 70 dB intensity range and with a frequency ratio of about 1.22. Under these conditions, a nonlinear stimulus interaction occurs within the cochlea, and a tonal distortion product at a frequency of  $2f_1 - f_2$  is generated and radiates back to the external ear. Just as for the TEOAE, the DPOAE are detectable in the external meatus. The frequency-specific nature of the DPOAE may provide more precise information than with the TEOAE, but poor recording conditions may result in inaccurate measurements.

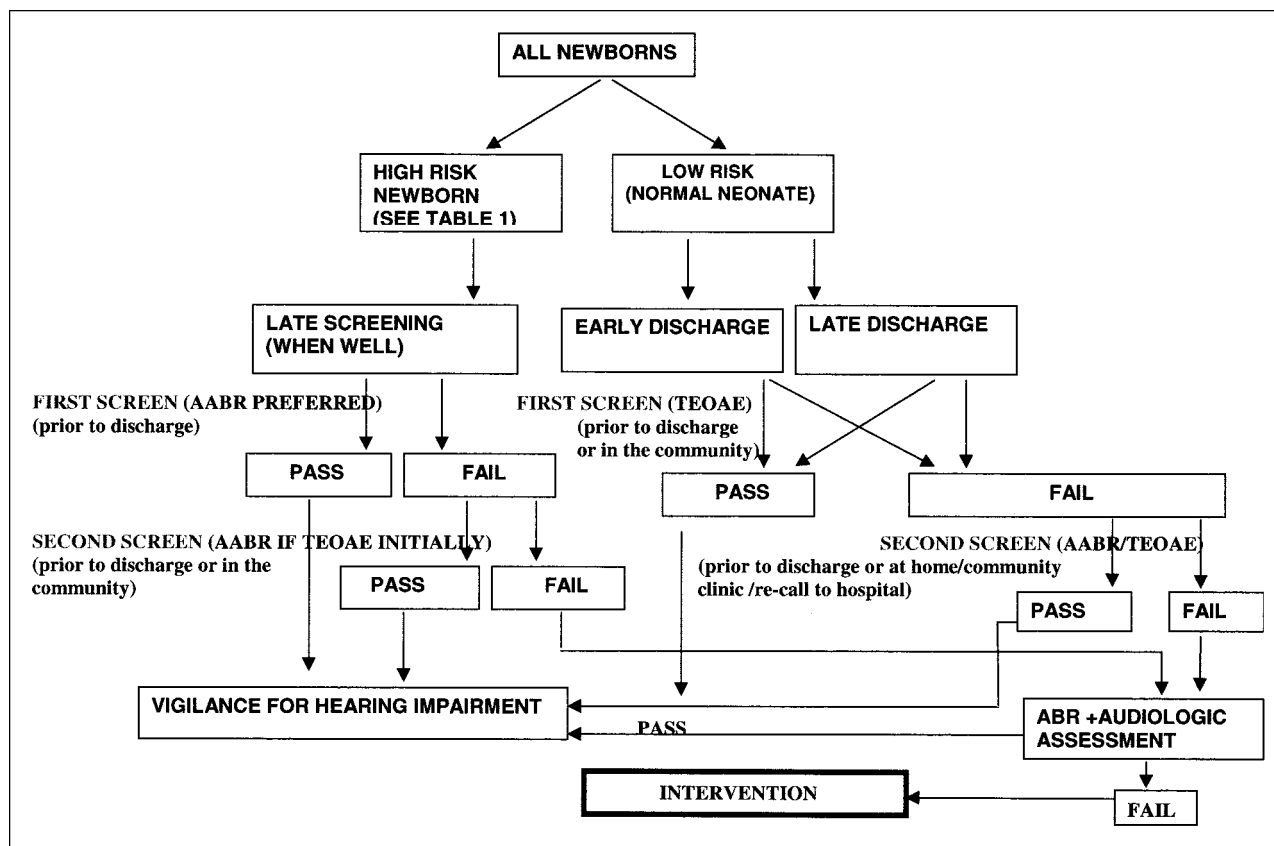


Figure. Suggested universal hearing screening protocol. AABR=automated auditory brainstem response, TEOAE=transient evoked otoacoustic emissions, ABR=auditory brainstem response

Any factor that interferes with the registration of a clear ABR or OAE will cause false-positive screening outcomes. The specificity of the AABR, TEOAE, and DPOAE improves when screening takes place at a later postnatal age and with cleaning of the external auditory meatus. This difference is believed to be due to cochlear maturation, clearance of middle ear fluid after the first 48 hours of life, or improved tympanic membrane mobility. Excessive environmental noise also decreases the specificity of TEOAE and DPOAE.

There is good evidence that the TEOAE, DPOAE, and AABR are accurate tests for detection of significant hearing impairment in neonates and infants. A two-stage screening protocol tends to yield lower false-positive rates (with specificity >94%) without substantial reduction in sensitivity. Each technology is affected by environmental conditions and the age at which the screen takes place, with the OAE methods affected more than the AABR. There is more variability in the specificity with the TEOAE and DPOAE than with the AABR. That difference is reduced when a two-stage screening proce-

dures is used and the AABR is used for the second stage of the screen. Any of the three screening technologies may be used in a two-stage procedure to detect hearing impairment in newborns. The tests are noninvasive, brief, and inexpensive.

### UNHS Programs: Advantages and Disadvantages

Current screening protocols typically employ either a one-, two-, or three-stage screen, with up to two screens prior to discharge from the birth admission, follow-up screening in the community, or both. A higher rate of false-positive findings immediately postnatally (attributed to resolving middle-ear conditions) may be addressed by multistage screening with an outpatient rescreen. Children failing the screening protocol should undergo prompt confirmatory and diagnostic hearing assessment with manual ABR or VRA (preferably including ear-specific and frequency-specific techniques), tympanometry, acoustic reflexes, and other audiologic tests. A possible screening protocol is outlined in the Figure.

The current American Academy of Pediatrics guidelines suggest universal identification of hearing impairment by 3 months and commencement of intervention by 6 months of age. Because high referral rates result in increased stress on audiologic services and may cause psychological stress on families of those testing positive, a maximum false-positive rate of 3% is widely endorsed and considered feasible for hearing screening programs. A population coverage benchmark of 95% has been proposed, and a target of 100% sensitivity has been suggested. Realistic values will be determined by the sensitivity and specificity of practicable screening protocols and the outcome cost-benefit structure.

The accuracy of screening tests may not be identical in high-risk and low-risk groups. Potential sources of variation include different amounts of progressive or early-onset (ie, noncongenital) pathology in the two groups, confounding of behavioral hearing test outcomes by cognitive deficits, and differences in the distribution of hearing impairment for the two groups. Furthermore, it is easier to achieve behaviorally satisfactory test conditions (ideally, a sleeping baby) in low-risk babies, who generally are less distressed.

Important practical issues for UNHS programs include follow-up of both children who fail in-hospital screening and those who are not accessed or successfully screened before discharge. Low rates of screening are of concern and are believed to be due to shorter hospital stays and poor compliance for follow-up of first-stage screening failures. For each additional year of experience with screening, most current programs report increasing participation and follow-up of children who fail the screening. However, ongoing surveillance of infants who pass the neonatal screening but are at risk for progressive or early-onset hearing impairment generally is poor. The yield of UNHS programs is limited by hearing impairment possibly being acquired by both pediatric intensive care unit graduates and well babies (targeted or nontargeted screening). A pass in the UNHS program may give parents a false sense of security that their infant has normal hearing and may lessen parental or professional vigilance for detection of acquired hearing loss. Acquired hearing impairment due to both congenital (eg, cytomegalovirus) or acquired infections (eg, meningitis), acquired conductive hearing impairment (due to recurrent otitis media), or auditory neuropathy will not be detected by UNHS programs.

There are concerns that a false-positive screen will result in unnecessary parental anxiety, with a negative effect on the parent-child relationship. Questionnaires of parents whose children underwent UNHS did not con-

firm this concern. The screen was considered to be quick and painless for the infant. UNHS was considered to be a measure of security except in infants who had unilateral or mild deficits that required no intervention. In addition, high false-positive rates may increase the burden on diagnostic services. High-quality screening programs that have a maximum false-positive rate of 4% (after a two-stage screen) should minimize this effect.

Overall, UNHS programs have demonstrated earlier identification of hearing impairment, earlier diagnosis, and earlier intervention, whether by hearing aids or other interventions. It is critical that UNHS activities be followed by timely, appropriate, and well-integrated subsequent steps in the overall process that lead to delivery of effective, efficient, culturally sensitive, and family-centered hearing health care. The overall system is commonly referred to as an Early Hearing Detection and Intervention (EHDI) program.

In addition, there now is some indirect evidence that early intervention improves speech, language, cognitive ability, and personal-social skills through amplification with hearing aids or cochlear implants or other communication development programs. Evidence is based largely on retrospective data from cohorts that may not be entirely representative of a universal hearing screening program.

### Surveillance for Progressive and Late-onset Hearing Loss

Infants who are at risk for postnatal hearing loss that may present after neonatal screening should be rescreened periodically. Intervals of about every 3 to 6 months for at least 3 years have been suggested, but such a schedule is likely to prove impractical. For some risk factors, such as perinatal cytomegalovirus infection, there is evidence of continued postnatal expression over even longer periods. Specific postnatal events such as bacterial meningitis or head injury should be followed systematically by hearing screening. The screening technology most practical for widespread use by nonaudiologist personnel is probably automated OAE, either DPOAE or TEOAE, but because of the increased prevalence of hearing impairment in the at-risk group, the use of a more accurate test such as AABR may be indicated. Infants who fail any such screening, whether periodic or driven by a risk event, should receive full audiologic and otologic examination.

It is likely that 5% to 10% of newborns manifest one of the risk indicators for progressive or late-onset hearing loss defined by the Joint Committee on Infant Hearing (Tables 1 and 2), so the total amount of screening activity needing to be directed at progressive and late-

onset hearing impairment is substantial. The proportion of children who have hearing impairments at 5 years of age that actually are congenital is not yet well understood, and reported ranges vary widely. This is predictable because epidemiologic patterns of postnatal risk and perinatal management practices affect the distribution of impairment.

The key practical aspects of screening young infants relate to the behavioral state of the child and to the environmental noise levels. Whether OAE or ABR methods are used, accuracy will be poor if the child is not resting quietly (preferably sleeping). The primary problem associated with OAE is physical movement of the stimulus probe in the ear canal; for the AABR, electromyogenic interference associated with gross body movement may decrease sensitivity. A similar deterioration of specificity is expected if environmental noise levels are too high, and a limit of about 55 dBA (55dB sound pressure level with a so-called “A-weighting” of energy at various frequencies that approximates the sensitivity characteristics of the human ear) has been suggested.

The requirement of a sleeping infant means that accurate, objective, physiologic audiometry by OAE or ABR methods is increasingly difficult to obtain in a child older than 6 months of age. If the risk of hearing impairment is substantial, testing under mild sedation or light general anesthesia may be considered. Sedation or anesthesia probably would be used for full, objective diagnostic assessment immediately following screening failure in infants older than about 6 months or in younger infants who are found to be untestable in natural sleep.

It is fortunate that behavioral screening by VRA or related methods such as conditioned play audiometry (CPA) is increasingly feasible for many children older than 6 months of age who have no substantive cognitive deficit. The skill and experience required for accurate and consistent VRA and CPA are substantial. Informal office behavioral screening using various kinds of noisemakers and observing behavioral response is notoriously inaccurate and has little if any place in a high-quality system for childhood hearing health care. Definitive audiologic assessment of children who have significant cognitive disabilities can be a long-term challenge that requires careful longitudinal integration of various types of objective, physiologic evidence (otoacoustic emissions, evoked po-

tentials, middle ear muscle reflexes) as well as behavioral data from formal tests and real-world observations.

### Screening After Infancy

One goal of screening children in the age range of 3 to 5 years is to identify preschoolers who may have developed hearing impairment that is likely to interfere with communication and educational development. In the context of an integrated system for EHDI, such impairments are likely to be late-onset, progressive, or adventitious (hearing loss associated with diseases or traumatic events occurring in early childhood such as meningitis or head trauma). Risk indicators include family history, specific infections, trauma, and parent/caregiver or clinician concerns about hearing, speech, language, or developmental delay.

Middle ear disorders are common in this age group, and it can be efficient to screen for both hearing loss and middle ear abnormality. If the child is cooperative, OAE screening is feasible and attractive because of its objectiv-

**When** a child fails an objective screen, one of the first questions is whether the failure is attributable to middle ear disease.

ity. The latest devices incorporate both measurement of OAE and otoacoustic immittance capabilities. This allows simultaneous detection of any hearing impairment of at least 30 dB hearing loss (with OAE) and limited differential diagnosis of the type of impairment (conductive or sensorineural).

If a skilled tester and facilities are available and the child is cooperative and responsive, many children in this age group can be screened by CPA under earphones. This technique has the advantage of assessing the full perceptual system of a child, whereas objective procedures such as OAE and AABR measure only physiologic correlates of true hearing. Children who cannot perform adequately on CPA may be able to be tested successfully with VRA. It may be useful to conduct both objective and behavioral screening, where feasible, to guard against screening errors. Failure on either screening tool normally should be followed by full audiologic assessment.

When a child fails an objective screen, such as an OAE screen, one of the first questions is whether the failure is attributable to middle ear disease. Tympanometry (im-

mittance testing) can be helpful; when results are normal, the index of suspicion for significant SNHL is increased significantly. Full and prompt diagnostic audiologic assessment is indicated. If the tympanometry result is abnormal, there is a substantial likelihood that the screening failure is attributable to middle ear disease. The presence of a sensorineural component is not ruled out by abnormal tympanometric findings.

Detailed guidelines for screening protocols for children of various age groups are published in the Joint Committee on Infant Hearing 2000 position statement and the Guidelines for Audiologic Screening developed by the American Speech-Language-Hearing Association (see Suggested Reading).

### Suggested Reading

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## PIR Quiz

Quiz also available online at [www.pedsinreview.org](http://www.pedsinreview.org)

1. A 30-year-old gravida 3, para 3 woman gives birth to an apparently healthy male infant. She is concerned about hearing impairment in her son because her 5-year-old daughter was diagnosed as having progressive sensorineural hearing loss. Which of the following statements is *most* accurate regarding newborn hearing screening?
  - A. Background noise is necessary for accuracy.
  - B. Immediate postnatal testing is associated with higher false-positive rates.
  - C. Normal neonatal screening results make further rescreening unnecessary.
  - D. Screening should be performed when the infant is awake.
  - E. Visual reinforcement audiometry is the preferred screening test.
2. Which of the following is a *true* statement regarding neonatal hearing screening programs?
  - A. A normal neonatal hearing test result ensures normal development of hearing capacity.
  - B. An infant preferably should be tested while sleeping.
  - C. Background noise level does not affect test accuracy.
  - D. Otoacoustic emission tests are more reliable than auditory brainstem response.
  - E. Testing on the first neonatal day is more accurate than on subsequent days.
3. A term infant born after an uncomplicated vaginal delivery is diagnosed as having a congenital cytomegalovirus infection. Results of gross neurologic examination appear normal. Which of the following is a *true* statement regarding management of this child?
  - A. Automated auditory brainstem response requires intact cortical function.
  - B. Hearing screening is unnecessary in an infant whose cognitive development is normal.
  - C. Otoacoustic emission tests require intact cortical function.
  - D. Periodic rescreening is necessary to evaluate for progressive hearing impairment.
  - E. Therapeutic intervention of hearing deficit is of little benefit before the age of 1 year.
4. A 1-month-old girl who has had a normal perinatal course has failed two hearing screening tests. Which of the following is the *most* appropriate recommendation?
  - A. Perform computed tomography of the base of the skull.
  - B. Reassure the parents that false-positive results are common.
  - C. Refer for auditory brainstem response and audiologic assessment.
  - D. Retest at the age of 6 months.
  - E. Retest under sedation.