

Chapter VIII: Hearing Impairment

Causes

• *In Infants:*

- o Risk factors for congenital or perinatally acquired hearing loss include:
 - Family history of hereditary childhood sensorineural hearing loss;
 - Parental consanguinity;
 - 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;
 - Birth trauma;
 - Findings associated with a syndrome known to include hearing loss.

• *In children*

- o Hearing loss in the preschool and school-aged group is largely related to:
 - Meningitis, encephalitis, mumps and measles.
 - Trauma.
 - Acute otitis media (AOM).
 - Chronic otitis media with effusion (OME).

• *In adults*

- o Hearing loss acquired between adolescence and age 50 may be due to:
 - Meniere's disease.
 - Trauma.
 - Otosclerosis.
 - Ototoxic drug exposure.

- Eighth cranial nerve tumors.
- Noise-induced hearing loss.
- Secondary to hypertension, diabetes and atherosclerosis.

Screening tests

- o Pure-tone audiometry
- o Auditory brainstem response (ABR)
- o Evoked otoacoustic emission (EOAE)

Recommendations for prevention

• *In infants and children*

- o Routine hearing screening of asymptomatic children beyond age 3 years is not recommended.
- o There is insufficient evidence to recommend for or against routine screening of asymptomatic neonates for hearing impairment using evoked otoacoustic emission (EOAE) testing or auditory brainstem response (ABR).
- o Recommendations to screen high-risk infants may be made on other grounds (see clinical intervention).
- o Clinicians examining infants and young children should remain alert for symptoms or signs of hearing impairment.

• *In adolescents and working-age adults*

- o There is also insufficient evidence to recommend for or against routinely screening asymptomatic adolescents and working-age adults for hearing impairment.
- o Recommendations against such screening, except for those exposed to excessive occupational noise levels, may be made on other grounds (see clinical intervention).

• *In old adults*

- o 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.
- o There is insufficient evidence to recommend for or against routinely screening older adults for hearing impairment using audiometric testing (see clinical intervention).

Burden of Suffering

- o Prevalence estimates for hearing impairment vary depending on age and the criteria used to define the various causal conditions.¹

In Infants

- o For severe congenital and prelingually acquired losses, estimates range from 1 to 3/1,000 live births.^{1-5,12,20,25}
- o Moderate and severe hearing losses in early infancy are clearly associated with impaired language development.⁶⁻⁸
- o Factors that increase the risk for congenital or delayed-onset sensorineural hearing impairment include:^{2,5,9}
 - Family history of hearing impairment,
 - Congenital or central nervous system infections,
 - Ototoxic drug exposure,
 - Prematurity,
 - Congenital head and neck deformities,
 - Trauma,
 - Several other factors associated with admission to an intensive care nursery.

In Children

- o Uncorrected permanent hearing loss will have deleterious effects on personality and psyche of the child, in addition to low schoolartc achievement.
- o Chronic and recurrent acute otitis media is commonly associated with temporary hearing loss in infants and school-aged children.
- o 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.¹⁰
- o 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.¹⁰

In Adults

- o Hearing impairment creates further difficulties in adulthood, and has been correlated with:^{2,11,12,14}
 - Social and emotional isolation,
 - Clinical depression, and
 - Limited activity.
- o Hearing loss acquired between adolescence and age 50 may be due to:
 - Meniere's disease,
 - Trauma,
 - Otosclerosis,
 - Ototoxic drug exposure,
 - Eighth cranial nerve tumors.
 - Noise-induced hearing loss, which is a common cause of sensorineural hearing impairment in this age group.

- Secondary to hypertension, diabetes and atherosclerosis.

In Old persons

- o The prevalence of hearing impairment increases after age 60 years, with presbycusis being the most important contributor to this increase.
- o Approximately 25% of patients between ages 51 and 65 years have hearing thresholds greater than 30 dB (normal range being 0-25 dB) in at least one ear.¹³
- o Older persons with hearing impairment are particularly prone to suffering the associated social and emotional disabilities described earlier.^{16,17}

Accuracy of Screening Tests

Pure-tone audiometry

- o Cooperative children and adults are usually tested with pure-tone audiometry.
- o 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.²⁹
- o Audiometric results are, however, subject to error due to:^{10,30}
 - Improper calibration of the equipment,
 - Improper technique,
 - Background noise in the test area,
 - Unintentional or intentional misreporting by the subject.
- o Evaluation of neonates and infants below the age of 2-3 years with audiometry is more difficult; it therefore usually requires some form of electrophysiologic more than behavioral testing.

Auditory brainstem response testing (ABR)⁵¹

- o The Auditory brainstem response testing (ABR) is reliable, objective, noninvasive and painless test.
- o ABR is currently viewed as the standard for physiologic testing in infancy and the most accurate method available for determining hearing function.^{1,2,5,18}
- o Sensitivity rates have been reported to be 97-100% and specificity rates to be 86-96% in comparison with behavioral testing measures.^{2,5,19}
- o The ABR test is a useful diagnostic tool for measuring hearing when more conventional hearing tests cannot be used.
- o Electric potential wave activity in the auditory centers of the brain stem is recorded in response to a series of clicks presented to each ear. Thus, the ABR test indirectly estimates the level of hearing in the peripheral auditory system (middle ear and inner ear).
- o Because of the nature of this testing procedure, the ABR test does not rely on any form of subjective response from the individual being tested.
- o It is virtually unaffected by the use of anesthesia or sedation, or whether the patient is asleep or attentive.

- o ABR can be used:
 - As a screening procedure for newborns who are at risk for hearing loss.
 - As a diagnostic tool to identify infants and small children with a hearing loss.
 - To estimate hearing levels in difficult to test patients, i.e., mentally retarded, autistic, developmentally delayed.
 - To evaluate patients with suspected retrocochlear pathology.
 - Intra-operative monitoring (Neurological patients).
- o Test Procedure:
 - The ABR test is performed on an individual who is resting quietly or in a sleep state.
 - Mild sedation may be used, when necessary, under a physician's supervision.
 - A series of clicks are presented to each ear through special earpieces inserted into the ear canals.
 - The audiologist can vary the intensity of the clicks.
 - Skin electrodes are fixed on the individual to record appropriate brain stem wave activity.
 - The averaging computer in the ABR unit then analyzes the information and it is compared to normal responses.
 - This procedure does not cause any discomfort to the person.
 - The test takes approximately one hour.
- o 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.
- o 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.

Evoked otoacoustic emission (EOAE)

- o Evoked otoacoustic emission (EOAE) testing is a screening method suitable for neonatal and infant screening.^{20,21-24}
- o Basis of otoacoustic emissions:⁵²
 - Otoacoustic emissions are acoustic signals generated by the normal inner ear, either in the absence of acoustic stimulation (spontaneous emissions) or in response to acoustic stimulation (acoustically-evoked emissions) or electrical stimulation (electrically-evoked emissions).
 - Acoustically evoked otoacoustic emission testing allows the audiologist to understand how the outer hair cells of the inner ear are working.
 - There are three types of Otoacoustic emission testing:
 - Spontaneous,
 - Transient (TEOAE), and
 - Distortion Product (DPEOAE).
- o Using a cutoff of 30 dB to designate hearing impairment, transient EOAE testing has an overall agreement rate with ABR of 91%, with a sensitivity of 84% and specificity of 92%.^{46,47}

Screening procedures

Infants and Children

- o The majority of children with congenitally or neonatally acquired losses are identified by age 4-5 years.¹
- o 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.¹⁰
- o Routine audiometry can often detect the mild conductive hearing loss associated with OME.²⁶
- o 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),
 - The fact that middle ear conduction deficits may be superimposed on previously
 - Undetected sensorineural hearing loss due to other conditions.

Adolescents, adults, and old persons

- o Routine screening of working-age adolescents and adults is usually limited to those in high-risk occupations involving exposure to excessive noise levels.
- o Self-assessment questionnaires to identify hearing impairment probably represent the most rapid and least expensive way to screen for hearing loss in the adult.
- o Depending on audiometric criteria, self-assessment questionnaires are reported to be 70-80% accurate for identifying patients with hearing loss defined by pure-tone audiometry.

Effectiveness of Early Detection

- o Assessing the effectiveness of screening for hearing impairment depends upon the evidence that:
 - Hearing loss leads to decreased function and affects the quality of life,
 - Screening leads to earlier detection of hearing abnormalities than spontaneous clinical presentation or observation,
 - Most of the forms of hearing loss can be treated effectively, and
 - Effective treatment leads to improved function and well-being.

In infancy and childhood

- o Theoretically, the greatest benefit from hearing screening comes from detection of moderate to severe hearing impairment between birth and age 3 years.
- o Auditory stimuli during this period appear to be critical to development of speech and language skills,^{2,35} although other factors undoubtedly also play an important role.
- o 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.

- o 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.
- o 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.^{49,50}
- o Several studies have demonstrated superior communication performance in prelingually profoundly deafened children who received cochlear implants as compared to similar children using more traditional tactile or acoustic hearing aids.^{38,47,48}

Conductive hearing loss in the preschool-age group

- o Conductive hearing loss in the preschool-age group is most commonly due to self-limited cases of otitis media with effusion.
- o Hearing impairment in infancy due to chronic or recurrent otitis media with effusion can impair language development.³¹⁻³³
- o Auditory thresholds in hearing-impaired children can be improved through amplification with hearing aids and frequency modulation radio devices.
- o Auditory and language training can also improve communication skills.^{11,47,48}
- o 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.
- o Myringotomy and pressure-equalizing tube placement can resolve the conductive loss and prevent reaccumulation of middle ear effusion.³⁹
- o 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.
- o In older children, otitis media with effusion is responsible for the majority of hearing loss identified through screening.^{3,10}
- o 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.
- o 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 children⁴⁰ nor any preventive benefit from screening.⁴
- o 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.^{3,10}
- o 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.^{1,41,42}

In adults

- o 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.^{43,44}
- o The incidence of hearing impairment, predominately due to presbycusis, rises quickly beyond age 60, however.
- o There is no proof for the effectiveness of screening for hearing impairment in the adult population,¹⁴ but there are numerous studies documenting the benefits of hearing amplification in these patients.^{14,44} Patients receiving hearing aids have demonstrated improvement in communication and social function, as well as emotional status.⁴⁵

Clinical intervention

Screening

In infancy and childhood

- o Routine hearing screening of asymptomatic children beyond age 3 years is not recommended.
- o All confirmed cases identified through screening should be referred for audiological assessment and management.
- o There is insufficient evidence to recommend for or against routine screening of asymptomatic neonates for hearing impairment using evoked otoacoustic emission (EOE) testing or auditory brainstem response (ABR).
- o Recommendations to screen high-risk infants may be made on other grounds, including:
 - The relatively high prevalence of hearing impairment,
 - Parental anxiety or concern,
 - The potentially beneficial effect on language development from early treatment of infants with moderate or severe hearing loss.
- o For many high-risk conditions, hearing testing is commonly considered to be part of diagnostic evaluation and management.
- o ABR testing may be useful for all infants who meet at least one of the previous high-risk criteria or for those who fail EOE testing.
- o 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.
- o 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.

In adults

- o There is insufficient evidence to recommend for or against routinely screening asymptomatic adolescents and working-age adults for hearing impairment.

- o 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.
- o Screening of workers for noise-induced hearing loss should be performed in the context of existing worksite programs and occupational medicine guidelines.

In old adults

- o Screening older adults for hearing impairment is recommended.
- o The optimal frequency of screening older adults has not been determined and is left to clinical discretion.
- o An otoscopic examination and audiometric testing should be performed on all persons with evidence of impaired hearing by patient inquiry.
- o Although hand-held devices for audiometry 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.
- o There is therefore insufficient evidence to recommend for or against routinely screening older adults for hearing deficits using audiometry testing.

*Diagnosis and management of childhood otitis media in primary care*⁵³

- o In terms of morbidity in children in general practice, middle ear conditions are probably the most important relating to the upper respiratory tract, with 75% of all cases of Acute Otitis Media (AOM) occurring in children under the age of 10 years.
- o One in four children will have an episode of AOM at some time during the first 10 years of life with a peak incidence of diagnosis occurring between the ages of three and six.
- o The prevalence of Otitis Media with Effusion (OME) is very high (around 80% of children are reported to have OME at least once before the age of four) and this condition has important implications for child development.

Definition

Acute Otitis Media (AOM)

- o Inflammation of the middle ear of rapid onset presenting most often with local symptoms (the two most common being earache and rubbing or tugging of the affected ear) and systemic signs (fever, irritability and poor sleep for example).
- o There may be a preceding history of upper respiratory symptoms including cough and rhinorrhea.

Otitis Media with Effusion (OME)

- o Inflammation of the middle ear, accompanied by the accumulation of fluid in the middle ear cleft without the symptoms and signs of acute inflammation.
- o OME is often asymptomatic, and earache is relatively uncommon.

Clinical Assessment

- o In most situations, the GP will have to depend on history and otoscopy for diagnosing otitis media.

- o Children who require hearing assessment should be referred to an audiologist.

Diagnostic Features of AOM and OME

	Earache Fever Irritability	Middle ear effusion	Opaque drum	Congested drum	Impaired drum mobility	Hearing loss
AOM	Present	Present	Present	Present	Present	Present
OME	Usually absent	Present	Maybe absent	Absent	Present	Maybe present

Treatment for AOM

Antibiotics for AOM

- o Children diagnosed with acute otitis media should routinely be prescribed antibiotics as the initial treatment.
- o If an antibiotic is to be prescribed, the conventional five day course is recommended at indicated dosage levels.

Decongestants, antihistamines and mucolytics

- o Children with AOM should not be prescribed decongestants or antihistamines.

Analgesics

- o Parents should give paracetamol for analgesia but should be advised of the potential danger of overuse.

Oils

- o Insertion of oils should not be prescribed for reducing pain in children with AOM.

Referral for AOM patients

- o Children with frequent episodes (more than four in six months) of AOM, or complications, should be referred to an otolaryngologist.

Treatment for AOM

- In the majority of cases OME is a self limiting condition.

Antibiotics for OME

- o Children with otitis media with effusion should not be treated with antibiotics to guard against infection.

Decongestants, antihistamines and mucolytics

- o Decongestants, antihistamines or mucolytics may be used in the management of OME.

Steroids

- o The use of either topical or systemic steroid therapy is not routinely recommended in the management of children with OME.

Autoinflation

- o Autoinflation may be of benefit in the management of some children with OME.

Referral for OME patients

- o Children under three years with persistent bilateral otitis media with effusion and hearing loss of <25dB, but no speech and language development or behavioral problems, can be safely managed with watchful waiting.
- o If watchful waiting is being considered, the child should undergo audiometry to exclude a more serious degree of hearing loss.
- o Children with persistent bilateral otitis media with effusion who are over three years of age or who have speech and language, developmental or behavioral problems should be referred to an otolaryngologist.

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