

Sensory Disorders

Clara J. Richardson, MSN, RN-BC

HEARING DISORDERS

Description

Hearing disorders in children are defined as hard of hearing: loss of 25–70 dBHL (decibel hearing level); and as deaf: loss of more than 70 dBHL. The loss may be unilateral or bilateral.

Etiology

A conductive hearing loss is due to dysfunction of the external or middle ear. Sensorineural loss is caused by impairment of the cochlea or auditory nerve. A mixed loss has both sensory and conductive components.

Causes of conductive loss are malformation of the outer or middle ear and, more often, infections of the middle ear. Sensorineural loss is caused by hereditary factors in about half of cases. Other causes include infections, anoxic brain injury, physical trauma, prematurity, excessively loud noises, or ototoxic medications.

Incidence and Demographics

The incidence of permanent bilateral hearing loss is 0.6–2.6 per 1,000 children. Numerous genetic syndromes are associated with hearing loss and account for about 50% of cases. Sensorineural hearing loss not associated with a syndrome is more prevalent in Ashkenazi Jews, people of northern European descent, those of Japanese descent, and people from Ghana. In the United States, hearing loss is more common in males and in Blacks.

Risk Factors

- See causes above
- Cleft palate
- Neonatal hyperbilirubinemia
- Intracranial hemorrhage
- Maternal infections during pregnancy
 - Rubella
 - Cytomegalovirus (CMV)
 - Toxoplasmosis
 - Herpes virus
 - Syphilis
- Bacterial meningitis
- Down syndrome
- Chronic otitis media with effusion
- Head injury
- Ototoxic antibiotics
 - Kanamycin (Kantrex)
 - Gentamicin (Garamycin)
 - Vancomycin (Vancocin)
 - Tobramycin (Nebcin)
- Cisplatin, Carboplatin (chemotherapy)

Prevention and Screening

- Newborn screening
- Screening of children
 - Multiple episodes of otitis media with effusion
 - Cognitive disability
 - Cleft palate

Assessment

History

- Inability to localize sound by 6 months
- Delayed consonant-vowel babbling by 7 months
- Failure to respond to verbal instructions not accompanied by gestures by 16 months
- Delayed comprehensible speech by 24 months

Physical Exam

INFANTS

- No reaction to loud noise
- Delayed developmental communication milestones

OLDER CHILDREN

- Use of gestures rather than words
- Asking for repetition
- Decreased response to verbal expression
- Avoidance of social interaction
- Confused or inattentive facial expression

DIAGNOSTIC STUDIES

- Electrophysiological testing for sensorineural loss
 - Evoked otoacoustic emissions (EOAE)
 - Screening auditory brainstem response (SABR)
- Behavioral hearing tests to distinguish sensorineural from conductive loss
 - Behavioral observation audiometry (BOA) for infants younger than 8 months
 - Visual reinforcement audiometry (VRA) for those 8 months to 2½ years
 - Conditional play audiometry (CPA) for children older than 2½ years
 - Speech audiometry for those older than 2½ years
- Tympanometry to evaluate middle ear function

Management

- Invasive: Cochlear implant surgically implanted in the inner ear

Nonpharmacologic

- Amplification with hearing aids or assistive listening devices

SPEECH-LANGUAGE THERAPY

- Oralism
- American Sign Language (ASL)
- Total communication

ASSISTIVE TECHNOLOGY

- Teletypewriter
- Telecommunication devices

STRATEGIES TO ENHANCE COMMUNICATION

- Get child's attention before speaking
- Position: close, eye level, and in front of child
- Speak clearly, at even rate, and in short sentences
- Use facial expression

Pharmacologic

- No pharmacologic management indicated

Patient/Family Education

- Disability
- Amplification options
- Communication options

Outcomes and Follow-Up

- The child will have early identification of hearing impairment.
- The child will participate in an early intervention program.
- The child will utilize amplification devices.
- The child will show increased hearing with cochlear implant.
- The child will experience optimal communication.
- The family will demonstrate strategies to enhance communication with child.
- The family will verbalize understanding of disability and treatment options.

VISION DISORDERS**Description**

Vision disorders in children range from low vision (partial sight), with visual acuity better than 20/200 but worse than 20/70, to blindness, with acuity of 20/200 or worse.

Etiology**Cortical Visual Impairment (CVI) Due to Damage of the Visual Cortex in the Occipital Lobe**

- Hypoxia
- Infection of the central nervous system
- Traumatic brain injury

Retinopathy of Prematurity (ROP)

- Incomplete growth of the retinal blood vessels, which is not complete until 9 months gestation
- Blood vessels grow abnormally, die, and form scar tissue
- Scar tissue can cause retinal detachment and loss of vision

Optic Nerve Hypoplasia

- Nerve is small and thin and transmits impaired information to brain
- Results in sensory nystagmus, jiggling movement of the eyes

Strabismus

- Malalignment of eyes or deviation of one eye
- Results in amblyopia, reduced visual acuity in one eye

Incidence and Demographics

One-half to two-thirds of children with developmental disabilities have a significant vision disorder. Approximately 25% of newborns weighing less than 2,500 grams will have some degree of retinopathy of prematurity.

Risk Factors

- Prematurity
- Central nervous system (CNS) infection
- Traumatic brain injury
- Eye trauma

Prevention and Screening

Prevention

- Early prenatal care
- Avoid high concentrations of oxygen in premature infants
- Rubella immunization for all children
- Safety education to prevent eye trauma
- Compliance with treatment of vision impairment

Regular Vision Screening With Age-Appropriate Screening Tool

- Assessment of normal vision parameters in infants and young children
- Acuity charts

Assessment

- History of risk factors

Physical Exam

- Infant does not fixate on face or follow objects
- Eyes wandering or nystagmus
- Eyes that gaze in one direction
- No reaction to bright light or movement of object toward eye
- Focusing only on bright light

Diagnostic Studies

- See screening section above

Management

Invasive

- No invasive management indicated

Nonpharmacologic

- Textured, sound-producing toys
- Verbal cues before touching, moving from space to space
- Orientation to environment
- Self-care education
- Optical aids to enhance vision
- Braille education
- Computer training, voice recognition software
- Books on tape

Pharmacologic

- No pharmacologic management indicated

Patient/Family Education

- Description of child's specific defect
- Strategies to promote normal development
- Information about nonpharmacologic management above

Outcomes and Follow-Up

- The child will participate in an early intervention program.
- The child will develop independence of self-care activities and mobility.
- The child will develop skills in other senses to compensate for lack of vision.
- The child will demonstrate socialization at home and school.
- The family will maintain a safe environment for the child.
- The family will orient the child to surroundings.
- The family will demonstrate strategies to promote optimal development.

CONJUNCTIVITIS**Description**

Conjunctivitis is inflammation of the conjunctiva, the membrane lining the eyelid and covering the exposed surface of the sclera.

Etiology

The two common causes of bacterial conjunctivitis in children are *Streptococcus pneumoniae* and *Haemophilus influenzae*. Newborns may become infected during the birth process with *Chlamydia trachomatis* or *Neisseria gonorrhoeae*.

Incidence and Demographics

- About 15% of the population will have an episode of conjunctivitis at some time

Risk Factors

- Exposure to causative organism

Prevention and Screening

- No prevention or screening indicated

Assessment

- History of maternal infection

Physical Exam

- Crusty, purulent drainage
- Redness of the conjunctiva
- Swollen eyelid
- Burning sensation
- Itching
- Tearing

Diagnostic Studies

- Diagnosis based on symptoms
- Culture if no improvement

Management

Invasive

- No invasive management indicated

Nonpharmacologic

- Warm, moist compresses to loosen crusty drainage
- Cold compresses to decrease swelling
- Avoid eye makeup during infection
- Frequent handwashing

Pharmacologic

- *S. pneumoniae* and *H. influenzae*: Ophthalmic treatment
 - Erythromycin
 - Bacitracin/polymyxin B
 - Polymyxin B/TMP
- *C. trachomatis*
 - Erythromycin orally
- *N. gonorrhoeae*
 - Ceftriaxone (Rocephin) IM or IV
 - Cefotaxime (Claforan) IM or IV

Patient/Family Education

- Prevention of spread of infection
- Medication administration

Outcomes and Follow-Up

- The child will show decrease in redness, swelling, and drainage.
- The child and family will verbalize understanding of strategies to prevent spread of infection.
- The family will demonstrate medication administration.

OTITIS MEDIA AND OTITIS EXTERNA

Description

Acute otitis media (AOM) is a viral or bacterial infection of the middle ear. Otitis media with effusion (OME) is fluid in the middle ear without acute infection. Chronic suppurative otitis media is persistent infection resulting in perforation of the tympanic membrane. Otitis externa (OE), commonly called swimmer's ear, is bacterial infection of the outer ear.

Etiology

Obstruction of the Eustachian tube allows fluid to accumulate in the middle ear. The fluid may be contaminated from the nasopharynx, resulting in acute otitis media. If the fluid does not drain, an otitis media with effusion results. The effusion may last for weeks or months after the infection has resolved. Otitis media is often caused by *Streptococcus pneumoniae*, *Haemophilus influenzae*, and *Moraxella catarrhalis*. Most cases of chronic suppurative otitis media are due to penicillin-resistant *S. pneumoniae*.

The outer ear canal is ordinarily protected from infection by cerumen, but when the canal is altered by water, humidity, insufficient cerumen, or use of foreign object to clean canal, bacteria invade. Common causative organisms of otitis externa are *Pseudomonas*, *Enterobacteriaceae*, and *Proteus*.

Incidence and Demographics

Each year in the United States, 13.6 million pediatric office visits are for acute otitis media, and there are approximately 2.2 million cases of otitis media with effusion. The incidence of otitis media peaks in children between 6 and 24 months of age. It is more prevalent in Native Americans, Alaskans, and Canadian Inuit children. Otitis externa affects about 4 in 1,000 people annually. It peaks in children between 7 and 10 years of age.

Children are more prone to develop otitis media than adults for several reasons. The child's Eustachian tube is shorter, wider, lies more horizontal, and consists of underdeveloped cartilage. Children have larger adenoids that prevent drainage, immature humoral defense mechanisms leading to increased infection risk, and spend more time lying down which allows fluid to pool at the back of the throat with easy access to the Eustachian tube.

Risk Factors

Acute Otitis Media

- Bottlefeeding instead of breastfeeding
- Feeding in flat position rather than with head elevated
- Frequent contact with multiple children
- Exposure to tobacco smoke and air pollution
- Frequent pacifier use
- Previous episode of AOM
- Allergic rhinitis
- Cleft palate
- Down syndrome

Otitis Externa

- High humidity and warm temperatures
- Swimming
- Trauma to the ear canal
- Hearing aid use

Prevention and Screening

- No screening is indicated, but some prevention is possible.

Acute Otitis Media

- Breastfeeding
- Decrease use of pacifier in infants older than 6 months
- Avoid propping bottle
- Avoid exposure to tobacco smoke and pollution
- Avoid forceful nose blowing

Otitis externa

- Avoid using cotton swabs to clean ear canal
- Use hair dryer on lowest setting to dry canal after swimming or bathing
- Prophylaxis with acidifying or alcohol ear drops before swimming

Assessment

History

ACUTE OTITIS MEDIA WITH/WITHOUT EFFUSION

Upper-respiratory infection

OTITIS EXTERNA

Water in ear

Physical Exam

ACUTE OTITIS MEDIA

- Pain evidenced by ear pulling, head rolling, irritability in infants
- Immediate relief of pain if tympanic membrane ruptures
- Fever, rhinitis
- Decreased appetite, vomiting, diarrhea
- Postauricular and cervical lymph gland enlargement
- Tympanic membrane appears red, bulging, with no visible landmarks or light reflex

OTITIS MEDIA WITH EFFUSION

- Intermittent ear discomfort
- Feeling of fullness, popping, fluid motion
- Conductive hearing loss
- Tympanic membrane appears dull gray, slightly injected, with obscured landmarks and visible fluid level

CHRONIC SUPPURATIVE OTITIS MEDIA

- Signs of effusion

OTITIS EXTERNA

- Pruritus
- Pain
- Erythema
- Grayish, greenish, cheesy discharge
- Swelling with conductive hearing loss

Diagnostic Studies

- Tympanometry
- Tympanocentesis with aspiration and culture of middle ear fluid after multiple antibiotic failures

Management

Invasive

- Myringotomy with pressure-equalizing tube insertion

Nonpharmacologic

- Acute otitis media
- Application of heat or cold

Otitis Externa

- Clean canal of debris with curette
- Clean drainage from outer canal and skin around ear
- Apply petroleum jelly to skin around ear to prevent excoriation from drainage
- Gauze wick to get medication inside canal if canal is very edematous

Pharmacologic

OTITIS MEDIA

- Treatment for ear pain before initiation of antibiotic treatment
- Acetaminophen (Tylenol) or ibuprofen (Motrin)
- Analgesic ear drops such as antipyrine/benzocaine (Auralgan)
- Start antibiotics if symptoms persist
- Immediate antibiotic treatment for fever over 102.2° F (39° C), or for infant younger than 6 months of age
- First line: Amoxicillin (Amoxil, Trimox)
- Alternatives: Cefuroxime (Ceftin), cefdinir (Omnicef), cefprozil (Cefzil), azithromycin (Zithromax)
- Persistent OM: Amoxicillin/clavulanic acid (Augmentin), cefuroxime (Ceftin), ceftriaxone (Rocephin)

OTITIS EXTERNA

- Polymycin B/neomycin/hydrocortisone (Cortisporin otic drops)
- Ofloxacin (Floxin otic drops)
- Analgesics for pain

Patient/Family Education

- Diagnosis and treatment plan
- Medication administration
- Strategies for nonpharmacologic management
- Strategies for prevention
- Strategies for dealing with temporary conductive hearing loss
- Keeping water out of ears of children who have pressure-equalizing tubes

Outcomes and Follow-Up

- The child will experience effective pain management.
- The child will recover without complications.
 - Persistent hearing loss
 - Perforation of tympanic membrane
 - Acute mastoiditis
 - Delayed language development
- The family will verbalize understanding of diagnosis and treatment plan.
- The family will demonstrate medication administration, nonpharmacologic management, and strategies for dealing with hearing loss.
- The family will verbalize strategies for prevention.

RETINOBLASTOMA

Description

Retinoblastoma is a primary intraocular cancer.

Etiology

The disease begins with gene mutation. If untreated, retinoblastoma grows, causing retinal detachment and necrosis. Invasion continues into the orbit, the optic nerve, and then into the central nervous system. Sites of metastasis are lungs, bone, and brain.

Incidence and Demographics

The incidence of retinoblastoma is 1 in 20,000. It is usually diagnosed in children between 1 and 2 years of age, and onset after 5 years of age is rare.

Risk Factors

Family history

Prevention and Screening

There is no prevention, but screening for red reflex should be done at every well-child visit.

Assessment

History

- Family history
- Family often first to notice whitish glow

Physical Exam

- Leukokoria, white reflex instead of normal red reflex
- Strabismus
- Red eye
- Tearing
- Corneal clouding
- Discoloration of iris
- Blood in anterior chamber
- Glaucoma

Diagnostic Studies

- Indirect ophthalmoscopy to determine size and location of tumor
- Ultrasound
- CT scan
- MRI with contrast
- Lumbar puncture and bone scan in advanced disease

Management

Invasive

- Laser thermotherapy to destroy tumor with high temperature
- Cryotherapy to destroy tumor with extremely cold temperature
- Enucleation for advanced disease

Nonpharmacologic

- Radioactive plaques: High-dose radiation therapy
- External beam radiotherapy

Pharmacologic

- Chemotherapy to reduce tumor size before local therapy
- Vincristine (Oncovin)
- Cyclophosphamide (Cytoxan)
- Doxorubicin (Adriamycin)
- Cisplatin (Platinol)
- Carboplatin (CBDCA)
- Etoposide (VP-16, VePesid)

Patient/Family Education

- Diagnosis and treatment plan
- Side effects of chemotherapy
- Care of prosthetic eye

Outcomes and Follow-Up

- The child will recover without metastasis.
- The family will verbalize understanding of diagnosis and treatment plan.
- The family will demonstrate care of prosthetic eye.

TRAUMA TO THE EYE**Description**

Eye trauma involves injury to the orbit, eyeball, eyelids, conjunctiva, or lacrimal glands.

Etiology

Orbital fractures are often caused by all-terrain vehicle (ATV) crashes, paintball injuries, and fireworks. Children most commonly sustain trapdoor, hinged orbital fractures. Penetrating injuries may be due to sharp objects such as scissors or knives, propulsive objects such as firecrackers or guns, or blunt objects such as small paintballs. Nonpenetrating trauma may be caused by foreign objects, chemical or thermal burns, or large balls. Trauma to the eye can lead to hyphema, an accumulation of blood in the anterior chamber, accompanied by increased intraocular pressure. After a few days, a secondary hemorrhage may occur with further increase in pressure and poorer chance of vision recovery. This secondary injury can cause glaucoma, vitreous hemorrhage, retinal detachment, choroidal rupture, sclera rupture, or otic atrophy. Another common result of trauma is corneal abrasion from contact lenses, foreign bodies, or chemicals. Corneal abrasion may lead to ulceration and erosion of the cornea with visual impairment.

Incidence and Demographics

- There are about 2.4 million eye injuries in the United States each year.
- Children under the age of 15 years account for one-third of eye trauma hospitalizations and 43% of all sports-related eye injuries.
- The most common injury is orbital fracture, accounting for about 39% of major eye trauma.
- 33% of children under the age of 6 years with hyphema develop secondary hemorrhage.

- The risk of secondary hemorrhage decreases with age and occurs more frequently in African Americans.
- Approximately 75% will recover visual acuity after hyphema treatment.
- 14% of individuals with hyphema will have poor visual results.
- Corneal abrasion is the most common eye injury and is most prevalent in those who wear contact lenses.

Risk Factors

- Lack of protective eyewear
- Participating in activity inappropriate for developmental age
- Sickle cell disease increases risk of increased intraocular pressure

Prevention and Screening

- Protective eyewear
- Adult supervision of dangerous activity

Assessment

- History of trauma

Physical Exam

ORBITAL FRACTURE

- Intraorbital pain on eye movement
- Diplopia, double vision, or blurred vision
- Swelling
- Nausea and vomiting

HYPHEMA

- Pain
- Blurred vision
- Photophobia
- Tearing

CORNEAL ABRASION

- Pain or foreign body sensation
- Tearing
- Photophobia

Diagnostic Studies

- Visual acuity
- Radiography or CT scan for suspected fracture
- Slit lamp ophthalmic examination
- Tonometry to measure intraocular pressure

Management

Invasive

- Suturing for lacerations
- Surgical repair of fractures
- Surgical evacuation of hyphema
- Intraocular lens implantation for cataracts, a complication of trauma

Nonpharmacologic

HYPHEMA

- Eye patch and shield on affected eye
- Elevating head of bed 30° to 45° to promote settling of hyphema

CORNEAL ABRASION

- Ice compresses for 24–48 hours, then warm compresses
- Rest eyes

Pharmacologic

HYPHEMA

- Acetaminophen with or without codeine for pain
- Avoid aspirin and nonsteroidal anti-inflammatory drugs (NSAIDs)
- Topical aminocaproic acid (ACA, Amicar) to prevent secondary hemorrhage
- Topicals to decrease intraocular pressure
 - Brimonidine tartrate (Alphagan, Allergan)
 - Latanoprost (Xalatan, Pharmacia)
 - Timolol maleate (Timoptic-XE)

CORNEAL ABRASION

- Topical antibiotic to prevent infection
 - Ofloxacin (Ocuflox)
 - Polymyxin B/trimethoprim (Polytrim)
 - Ciprofloxacin (Cipro, Ciloxan)
 - Erythromycin (E-Mycin)
- Analgesics for pain
- Antibiotics to prevent infection
- Tetanus vaccine for penetrating injuries if not up to date
- Nonsteroidal anti-inflammatory eye drops
- Pain medication

Patient/Family Education

- Diagnosis and treatment plan
- Medication administration
- Nonpharmacologic management
- Prevention of future injury

Outcomes and Follow-Up

- The child will recover without permanent visual changes.
- The child and family will verbalize understanding of diagnosis and treatment plan.
- The child and family will verbalize understanding of nonpharmacologic management.
- The child and family will identify strategies to prevent future injury.
- The family will demonstrate medication administration.

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