CHAPTER 1

Hearing Loss

Dr. Naeem K. Makhdoom

A 40-year-old female who has been working as a telephone operator for the past 10 years comes to visit your clinic. She says, "For the past 3 months, I have not been enjoying my life and music anymore, and I get embarrassed at social gatherings because I do not understand what people are talking about. It sounds like music is distant, and I cannot hear the snap of some treble sounds. I also cannot hear "S" or "T" sounds well. It is making my life really terrible. I am frustrated, and I have been withdrawing from social activities and feeling depressed. I am not sure what is happening to my ears. Is it something related to my job? Something wrong that is affecting my hearing ability? What structures are found in the ear, and how does hearing happen? Please help! I can't hear properly anymore!"

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Q1. What is hearing loss/deafness?

Deafness, hearing impairment or *hearing loss* is a partial or total inability to hear sound in one or both ears.

Hearing loss occurs when there is diminished sensitivity to sounds that are normally heard. The term hearing impairment is usually reserved for people who exhibit relative insensitivity to sound in the speech frequencies. The severity of hearing loss is categorized according to the degree of the increase in the hearing threshold (i.e., the quietest sound intensity that a patient can perceive).

Q2. What are the anatomical parts of the ear?

The ear is divided into three major parts (Figure 1.1):

- The outer ear (external ear)
- The middle ear
- The inner ear

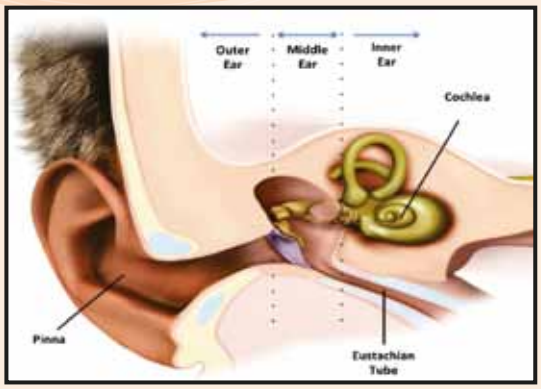

Figure 1.1: *Diagram showing the anatomy of the ear. (Courtesy of Med'el Co.)*

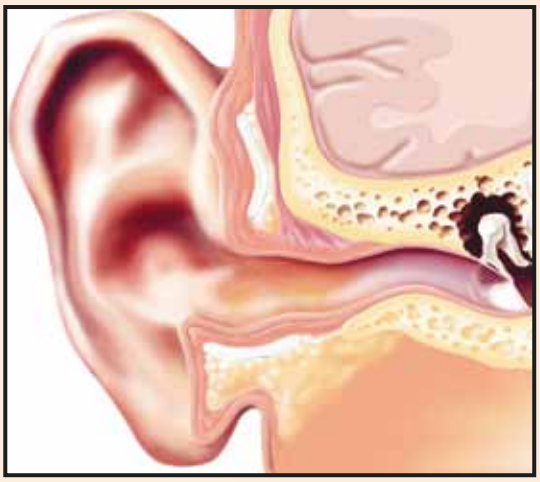

Figure 1.2: *Diagram showing the anatomy of the outer ear (Courtesy of Oticon)*

Q.3 What are the components of the external ear and its important innervations?

The external ear has two parts: the auricle and the external auditory canal (Figure 1.2).

The auricle or pinna is the trumpet-shaped protruding part of the ear. It is formed from a plate of corrugated elastic cartilage (except for the lobule, which is composed of fat and fibroareolar tissue) that is covered by skin.

The external auditory canal extends from the center of the pinna to the tympanic membrane. It is divided as follows:

a. The lateral one-third, which has a cartilaginous wall, is covered by skin that contains hair follicles, ceruminous glands and sebaceous glands.

b. The medial two-thirds, which has a bony wall, is covered by skin that is free of hairs and glands.

The external ear serves as a sound collector and directs sound toward the tympanic membrane.

Q4. What is the eardrum/tympanic membrane?

The eardrum or tympanic membrane (Figure 1.3) is a membranous structure that stretches across the inner end of the external auditory canal. It is divided into two parts: the major part is the pars tensa, and the much smaller part is the pars flaccida. The pars tensa is formed from four layers: an outer squamous cell layer, two fibrous middle layers and an internal mucosal layer. The pars flaccida is formed from only two layers because it is devoid of collagen fibers.

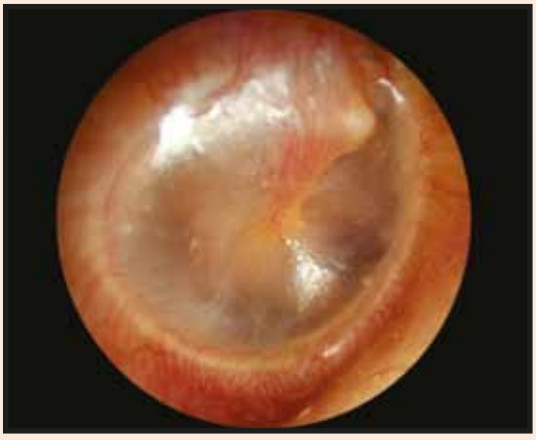

Figure 1.3: *Picture of the eardrum.*

Q5. What are the components of the middle ear cleft?

The middle ear cleft (Figures 1.4 and 1.5) is an air-containing cavity that is lined by mucosa located within the temporal bone between the external and inner ears. It consists of the Eustachian tube, the tympanic cavity (middle ear cavity), the aditus and the mastoid antrum and air cells.

The Eustachian tube is approximately 3 cm in length. It is composed of a proximal bony region (1/3) and a distal cartilaginous region (2/3). It connects the nasopharynx to the tympanic cavity. The tympanic cavity lies between the tympanic membrane laterally and the inner ear medially. Its roof is formed from thin bone (the tegmen tympani), which separates it from the middle cranial fossa. The floor is separated by a thin bone from the jugular bulb. Laterally, the middle ear is bound mainly by the tympanic membrane. The medial wall of the tympanic cavity is the lateral wall of the inner ear. In it, there are two small windows, the oval and round windows. In front of these windows lies the promontory, which is the bony covering of the cochlea. The facial nerve crosses the medial wall above the oval window.

The bony lateral semicircular canal lies posterosuperior to the facial nerve. The aditus is a small opening connecting the superior part of the tympanic cavity (attic or epitympanum) to the mastoid antrum, which is a large air cell lying behind the tympanic cavity within the mastoid bone. The antrum is connected with the mastoid air cells, which are highly variable in size and number.

The middle ear houses the ossicular chain (malleus, incus and stapes), the intratympanic muscles (stapedius and tensor tympani) and nerves (the facial and its chorda tympani branch as well as the tympanic plexus, which is formed by a branch of the glossopharyngeal nerve).

The middle ear amplifies and conducts the sound waves to the inner ear. The tympanic membrane is at its most efficient when the middle ear pressure is equal to the atmospheric pressure. This pressure is maintained by the Eustachian tube.

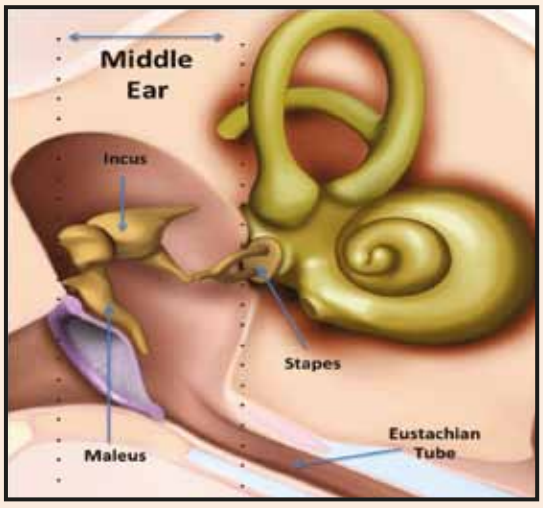

Figure 1.4: *Diagram showing the anatomy of the middle ear. (Courtesy of Med"el Co.)*

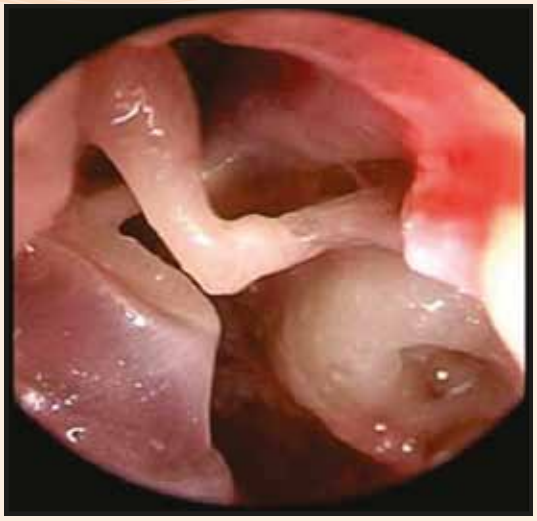

Figure 1.5: *Endoscopic picture of the middle ear from the mastoid showing that the tympanic membrane is retracted. The malleus is impeded by the eardrum anteriorly and is attached posteriorly to the tensor tympani. The incus is attached to the stapes, which covers the oval window and is attached posteriorly to the stapedial tendon. The socket under the stapes is the site of the round window, while the bulging over the stapes is the facial nerve canal.*

Q6. Name the three ossicles in the middle ear (Figure 1.6).

1. The malleus, which is laterally attached into the tympanic membrane.
2. The incus, which is related to the malleus laterally and to the stapes medially.

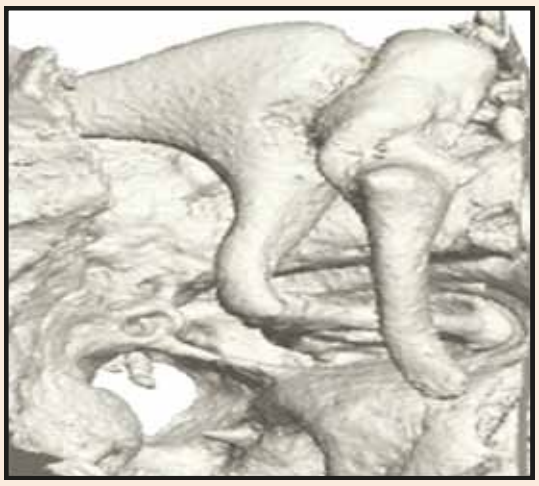

Figure 1.6: *Reformatted high-resolution CT scan showing the ossicles of the middle ear.*

Q7. Describe the sensory innervation of the external and middle ear.

1. C2 and C3 (great auricular and lesser occipital nerves): Post-parotidectomy patients are advised to cover their ears to prevent frostbite.
2. CN-V (auriculotemporal nerve of the mandibular branch): Dental diseases, sinonasal disorders and temporomandibular joint diseases can cause referred earache.
3. CN-VII: Acoustic neuroma can cause anesthesia of the ear (*Hitzelberger sign).*
4. CN-IX (tympanic or Jacobson nerve): Tonsillitis and tonsillectomy can cause referred earache.
5. CN-X: (auricular or Arnold's branch): A cough reflex occurs when the ear is stimulated. Cleaning the ear can cause vasovagal syncope.

Q8. What is the inner ear?

The inner ear consists of a membranous labyrinth embedded in a bony labyrinth (Figure 1.7). The osseous labyrinth consists of the bony cochlea anteriorly, the vestibule in the middle and the bony semicircular canals posteriorly. It contains perilymph and the membranous labyrinth. The membranous labyrinth contains endolymph as well as the auditory and

vestibular end organs. The auditory receptors that detect sound are present in the cochlear duct, which is enclosed in the bony cochlea. The vestibular end organs that sense acceleration and gravitational forces are present in the saccule and utricle, which are present in the vestibule and in the membranous semicircular glands that are present in their corresponding bony canals.

Figure 1.7: *Diagram showing the anatomy of the inner ear (courtesy of Med"el Co.)*

Q9. What is the cochlea?

The cochlea is a snail-shaped structure with two and a half turns that is divided into three compartments along its entire length (Figure 1.8):

- Scala vestibuli (superior)
- Scala media (middle)
- Scala tympani (inferior)

Reissner's membrane separates the scala vestibuli from the scala media.

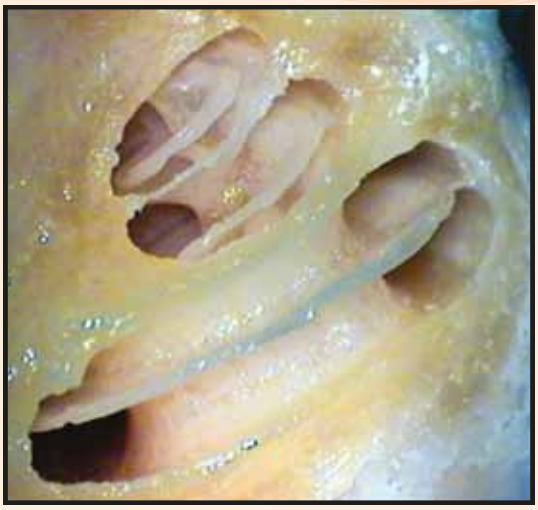

Figure 1.8: *Picture of drilled cochlea showing the partition between the scala tympani and the scala vestibuli. (Photographs courtesy of the personal collection of Dr. David P. Morris FRCS (ORLHNS) MD.)*

The basilar membrane separates the scala media from the scala tympani. The perilymph is a fluid similar in composition to the extracellular fluid that fills the scalae vestibuli and tympani. The endolymph is a fluid similar in composition to the intracellular fluid that fills the membranous labyrinth.
The scala media houses the organ of Corti.

Q10. What is the organ of Corti?

The organ of Corti is the sensor for hearing in the inner ear. It is an energy transducer or transformer that converts the hydraulic energy of acoustic waves into bioelectrical energy to be transmitted through the auditory nerve. It consists of the following parts:

- A single row of inner hair cells
- Three rows of outer hair cells
- Supporting cells

It has two walls:

- Superior (tectorial membrane)
- Inferior (basilar membrane)

Q11. How do we hear?

Upon the impact of sound waves on the tympanic membrane, vibrations are transmitted to the ossicular chain until the stapes impacting on the oval window triggers a series of travelling waves in the perilymph. Sound is transmitted along the scala vestibuli and the scala tympani to stimulate the hair cells in the organ of Corti. The sound energy is transformed by the organ of Corti into action potentials that are received by the afferent nerve endings and transmitted to the cochlear nuclei, superior olivary complex, lateral lemniscus, inferior colliculi and medial geniculate bodies to reach the auditory cortex in the temporal lobe, with significant crossover between the left and right sides. As the impulses reach the brain, we experience the sensation of hearing (Figure 1.9).

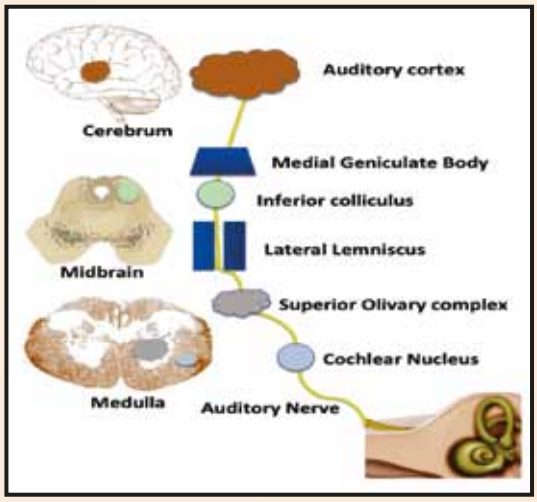

Figure 1.9: *Diagram showing the hearing pathway.*

Q12. What are the components of the peripheral vestibular organ?

It consists of five distinct end organs:

- Three cristae are present in the three membranous semicircular canals: the superior, lateral and posterior semicircular canals (positioned at right angles to each other).
- Two otolith organs (maculae) are present in the utricle and the sacule.

Q13. What is the difference in function between the semicircular canals and the otolith organs?

The semicircular canals detect angular accelerations (head rotations), while the otoliths detect linear accelerations (e.g., gravitational forces).

Q14. What is the vestibulo-ocular reflex (VOR)?

It is a reflex that stabilizes images on the retina during head movement. The VOR moves the eyes in the direction opposite of the head movement, which preserves the image in the center of the visual field.

CHAPTER 2

Congenital Ear Deformity

Prof. Jose N. Fayad

A mother comes to see you with her 2-month-old son, who was born with bilateral congenital deformities of the pinna. This is the first time these deformities have occurred in the family, she says. She is distressed and has many questions about why this happened and what can be done to fix it. The child failed a newborn screening test for hearing, and the mother has been asked to follow-up with her ear doctor. She tells you that her son is scheduled to undergo a CT scan in a few days. Otherwise, the baby is healthy, and does not have any other abnormalities or congenital malformations (Figure 2.1).

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Figure 2.1: *Shows microtia and atresia.*

Figures 2.2: *Baha with band.*

Q1. What is the highest single priority in this case at this point in time?

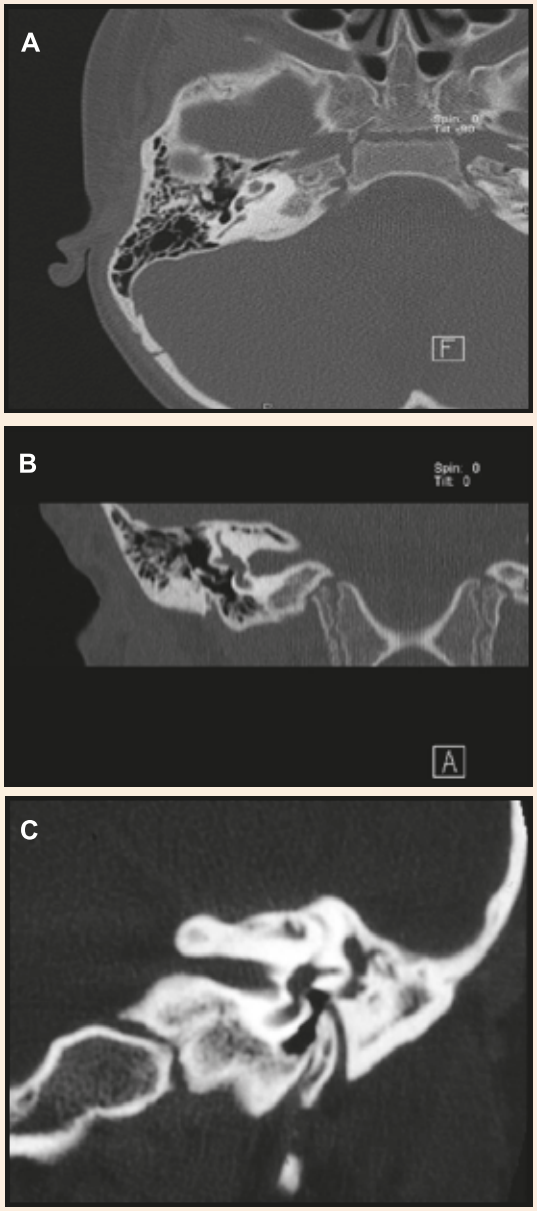
The highest single priority in this case is to refer the patient to an audiologist and fit the child with a bone-conduction hearing aid as soon as possible without delay. A BAHA band (Figure 2.2) will provide the patient with the hearing amplification needed for speech development. You explain to the mother that the auditory nerves are usually healthy in this type of disorder and that hearing aids will stimulate the inner ear and bypass the ear canal and the middle ear.

Q2. The mother asks you if she should proceed with the CT scan that her family doctor has advised?

There is no need to proceed with radiological evaluation at this point in time because no surgical intervention will be performed before 3 years of age at the earliest and perhaps not until 5 or 6 years of age. A CT scan can deliver a significant dose of radiation. Therefore, a CT scan should not be performed until surgery is being considered. You ask the mother to cancel the CT scan of the temporal bones and tell her that you will only perform a CT if there are signs of a complication such as drainage from the ear, redness, swelling around the ear or severe pain (Figures 2.3A, B and C).

Q3. What would you do differently if the child had a unilateral microtia / atresia?

The treatment would be the same. The priority would be to fit the child with a bone-conduction hearing aid (i.e., a BAHA band). X-ray examination is not necessary unless there is a complication or a suspicion of cholesteatoma on the atretic side (Figure 2.4A and B). Cholesteatomas are more frequent in cases of severe stenosis of the ear canal than the atresia of the ear canal. The incidence is approximately 15% in cases of severe stenosis of the ear canal.



Figures 2.3A, B and C: *Absent external ear canal.*

Figures 2.4A: *Right ear discharge with microtia, leading to a suspicion of cholesteatoma.*

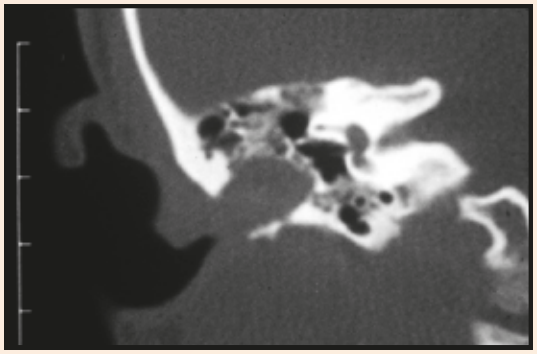


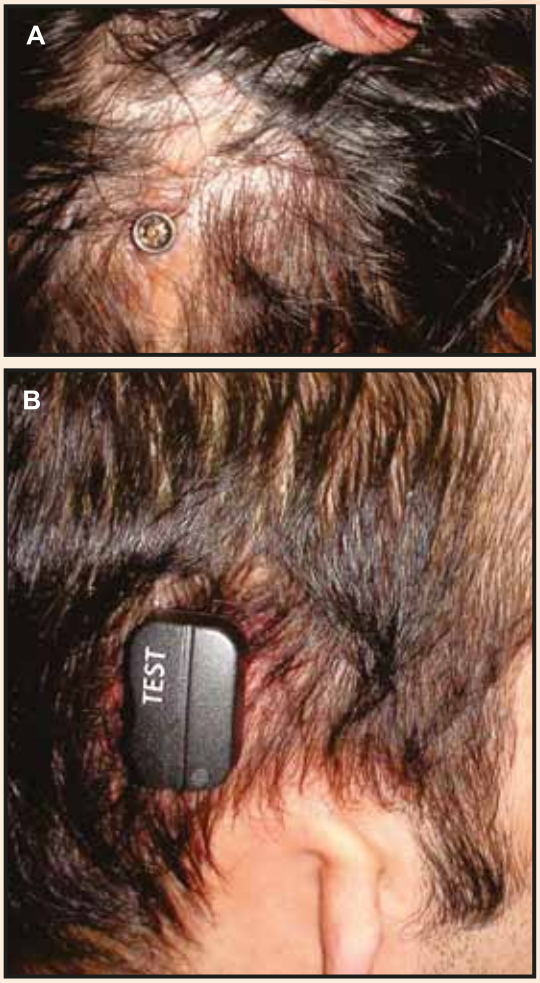
Figure 2.4B: *CT scan shows destructive lesion in the external auditory canal, most likely cholesteatoma*

Q4. The mother asks you, "What other test is important to determine that the child has no hearing deficit and good nerve conduction?"

An auditory brainstem response (ABR) test will be performed to verify objectively that nerve function is sufficient. Audiological evaluations can be difficult because of masking conditions. An ABR test with reversed polarity can overcome these difficulties. Children may exhibit profound sensorineural hearing loss (SNHL) in one of their ears.

Q5. What must be done between now and when it is time to consider surgery?

The child must wear the device consistently to develop proper speech. In the meantime, the parents should learn more about the BAHA device and possibly call or meet surgeons who are familiar with BAHA surgery, atresia repair and microtia repair. They should educate themselves in order to have a fruitful discussion with the surgeons concerning the best treatment for their child (Figures 2.5A and B).



Figures 2.5A and B: *(A) implant in place and (B) BAHA processor in place.*

Q6. Are there devices other than the BAHA?

The BAHA is currently the standard implant for atresia. It bypasses the absent outer ear canal and the abnormal middle ear and stimulates the inner ear directly via bone conduction to fully close the gap (Figure 2.6). Other implantable electromagnetic devices can also be useful. However, electromagnetic devices are not compatible with MRI examinations, and they are also more expensive. The surgery can be challenging depending on the severity of the malformation.

Figure 2.6: *BAHA sound bypasses the outer and middle ears.*

Q7. What will be involved in the surgical rehabilitation?

There are two essential parts to the surgery. First, a plastic surgeon will reconstruct the pinna. An otologist will then create a new ear canal and reconstruct the hearing mechanism. These surgeries are typically performed at separate times. Plastic surgery usually precedes the ear canal reconstruction.

Q8. How is the pinna reconstructed, and what is the timing of the surgery?

Three options are available. First, the pinna can be reconstructed using a rib cartilage graft (Figure 2.7). This surgery must precede the external auditory canal reconstruction and is typically performed at 6 years of age when the cartilage has reached a sufficient size to be harvested to reconstruct the pinna. Alternatively, an artificial material called Medpor may also be used to reconstruct the pinna (Figure 2.8).

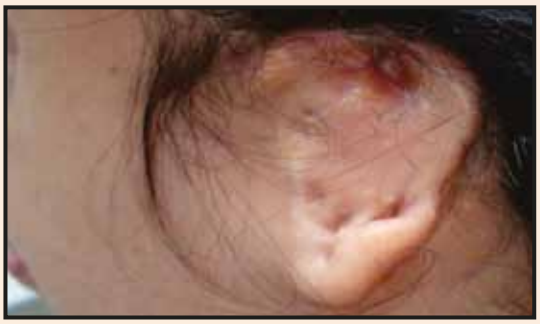


Figure 2.7: *Rib cartilage graft in reconstructed pinna.*

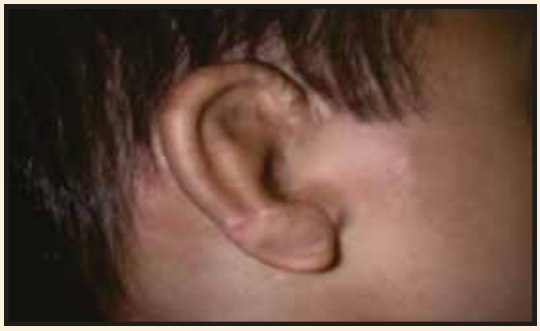


Figure 2.8: *Medpor pinna.*

The third option is to create a prosthesis that matches the normal ear pinna (Figures 2.9A and B).

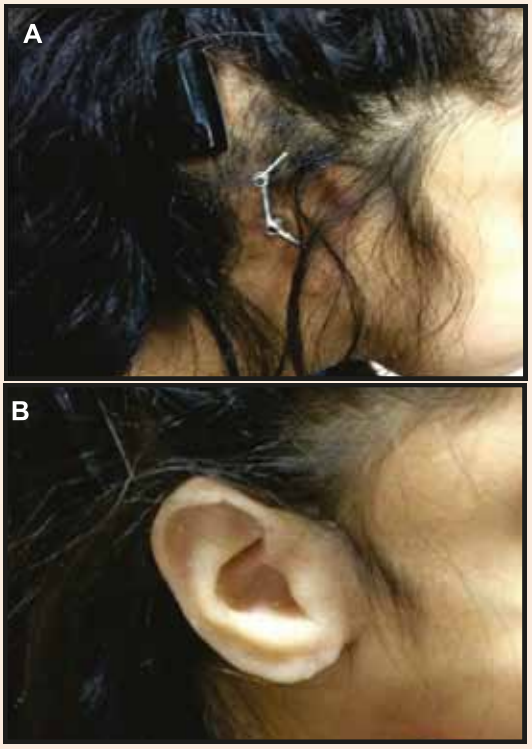


Figure 2.9A and B: *Osseointegrated screws hold a silicon prosthesis. Notice that the patient also has canal atresia and uses the BAHA for hearing.*

Each of these procedures has advantages and disadvantages. The parents should meet with surgeons who are well versed in these different procedures and choose the best option for their child.

Q9. What are the advantages and disadvantages of canaloplasty?

In favorable cases, the canaloplasty operation will allow restoration of hearing through a newly reconstructed ear canal. At the same time, middle ear reconstruction procedures can be performed as well. If surgery is successful, then hearing can be restored to near-normal levels. In partially successful cases, a hearing aid can be used to achieve more amplification if needed. However, canaloplasty is a challenging operation. The most common complication after canaloplasty is restenosis of the ear canal and facial nerve injury.

Q10. What is the timing of the canaloplasty in relation to pinna reconstruction?

When rib cartilage is used to reconstruct the pinna, the canaloplasty follows the pinna reconstruction surgery. When Medpor is used, the plastic surgeon sometimes prefers the otologist to operate first. This order allows the plastic surgeon to center the pinna correctly around the newly reconstructed ear canal.

Q11. If the patient has unilateral atresia, will the treatment be different?

Many surgeons believe that there is no need to rush to ear canal reconstruction when the atresia is unilateral and the other ear is normal. They feel strongly that the patient, as an adult, should be involved in the treatment decision.

CHAPTER 3

Earache

Dr. Hesham Hasan

A male infant was brought to you with a 1-day history of irritability with poor oral intake. The mother reported a history of a runny nose and fever over the last few days. The otoscopic examination performed by the doctor revealed the image in Figure 3.1.

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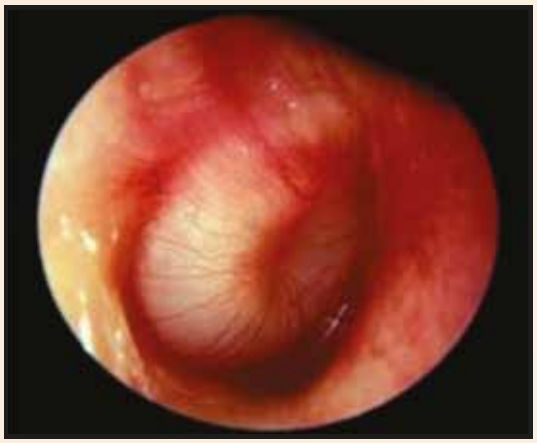


Figure 3.1: *Picture of the right ear. (Courtesy of Dr. Munahi Al-Qahtani)*

Q1. What are the common symptoms of the ear?

1. Earache
2. Drainage
3. Fullness
4. Hearing loss
5. Tinnitus
6. Itching
7. Vertigo

Q2. Are all earaches due to problems in the ear?

No. The ear is a common organ for referred pain due to its rich innervation. Did you know that auriculotherapy is based on the theory that the entire body is treatable by stimulation of the surface of the ear exclusively?

Q3. Does asking detail questions about the ear discharge help to locate the lesion?

Yes, asking more detail questions can help to locate the lesion.

- Wax is part of the normal ear cleaning mechanism.
- Mucus comes from the middle ear because there are no mucus glands in the outer or inner ear.
- Clear fluid from the ear could be a CSF leak and should be considered serious.

Q4. Can ear fullness indicate the site of the lesion?

In fact, all ear parts can give the sensation

of ear fullness including wax in the outer ear, middle ear effusion and inner ear hydrops (Meniere's disease).

Q5. What are the common abnormalities of the auricle?

The auricle is basically composed of skin and cartilage. Therefore, any illness of these tissues can occur in the auricle. The common congenital anomalies are the following:

- Prominent ear or bat ear (Figure 3.2)
- Microtia (small or partially developed ears)
- Preauricular sinus (small opening present at birth in front of the auricle)
- Accessory auricles (prominent skin at the front of the ears) (Figure 3.3).

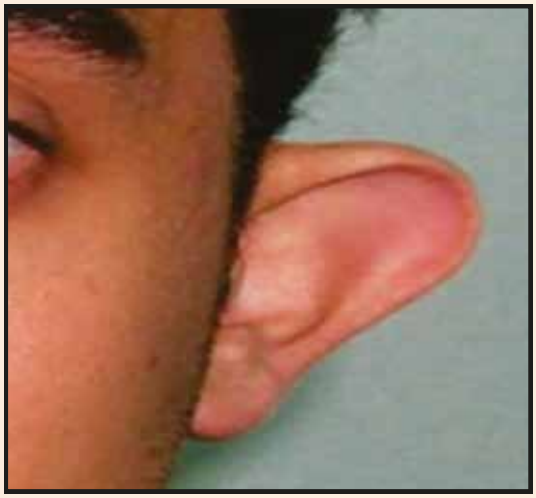


Figure 3.2: *Bat ear. (Courtesy of Dr. Munahi Al-Qahtani)*

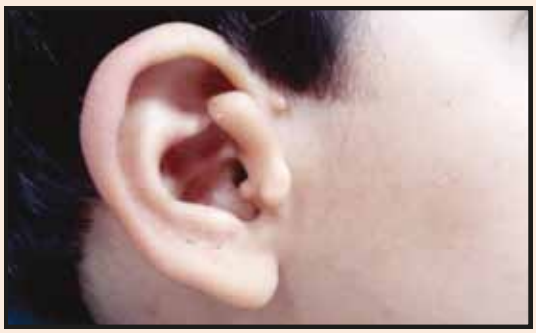


Figure 3.3: Accessory auricles

Q6. What instruments are used to examine the ear canal and eardrum?

Otoscopy (Figure 3.4) is the most common tool used to examine the ear. It consists of three parts:

- Handle, which contains the batteries to power the light source
- Head, which contains the light bulb and magnifying lens
- Cone, which is inserted into the ear canal

Figure 3.4: *Otoscope*

Some otoscopes have a rubber bulb and are called pneumatic otoscopes. These otoscopes facilitate the determination of the mobility of a patient's tympanic membrane in response to pressure changes. Decreased movement can indicate the presence of fluid in the middle ear.

Ear endoscopy (Figure 3.5) offers visualisation of fine detail of the ear canal and eardrum superior to the resolution of most standard otoscopes. Endoscopy also has the advantage of displaying an image on the monitor for the patients and allows the images to be recorded and replayed. Ear endoscopy gives a wide-angle and close-up view as well as behind the corner in cases of a perforated eardrum to exclude cholesteatoma. The heat produced by the light may cause dizziness.

Figure 3.5: *Ear endoscope*

Ear microscopy (Figure 3-6) is very useful in examining the ear. It gives binocular vision, which is very helpful in determining depth during the examination and when cleaning the ear. It also allows for two-handed manipulations of the ear.

Figure 3.6: *Microscopy of the ear in ENT clinic*

Q7. How can you easily examine children with an otoscope?

Otoscopic ear examination is completely painless but requires a great deal of skill and experience to obtain a clear assessment within a short time. Otoscopic evaluation is a very important skill for all doctors to master regardless of their specialty, but otoscopy is particularly important for doctors who work with children.

The child's head should rest against an adult's chest preferably on a parent's lap with one hand holding the child and the other hand holding the head to the opposite side.

The examiner should gently pull the auricle down, back and outward to straighten the ear canal. The tip of the otoscope will be placed gently into the ear. The speculum's tip is moved in different directions to see the inside of the ear and eardrum without touching the skin of the canal (Figure 3.7). Earwax is very common in children and an expert physician should be capable of examining the ear with some wax present.

Figure 3.7: *Ear examination in a child*

Remember, straighten the ear canal in adults by pulling the auricle up, back and outward (Figure 3.8).

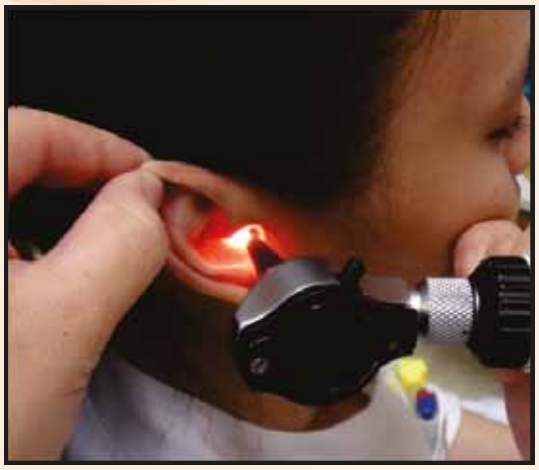


Figure 3.8: *Ear examination in an adult*

Q8. If the patient has a painful ear examination with the otoscope, what does this indicate?

Either the patient has otitis externa or the examiner incorrectly used the otoscope. To differentiate between these two, always ask the patient if just moving the ear is painful. Otitis external causes pain when touching the tragus. Remember, talking to your patient during the examination relieves much of their anxiety and encourages better cooperation.

Q9. What are the most common mistakes when using an otoscope?

All doctors should know how to properly use a stethoscope as well as otoscope. The following are the most common mistakes made by non-expert physicians:

1. The examiner's hand does not rest against the cheek of the patient.
2. Blind introduction of the speculum into the canal.
3. Using poor light.
4. Not using the largest speculum that comfortably fits within the ear, which gives better visualisation and is more comfortable.
5. Examining only the ear with the complaint and not both ears or starting the examination with the diseased ear.
6. Making sure the lens is clean.
7. Not examining the canal well.

Q10. What are the most common aspects missed during an ear examination?

1. Beginning with a careful inspection of the auricle and postauricular skin.
2. Testing the facial nerve.
3. Using a tuning fork.

Q11. If a patient has wax build-up, what should you do?

If the patient is cooperative, wax can be removed by suction, syringing or curettage (Figure 3.9). These methods should not be painful and should not cause any harm to the patient. If the wax is hard, the application of eardrops to dissolve it may be necessary.

Figure 3.9: *Wax hook for ear curettage*

Q12. How useful is a tuning fork during an ear examination?

Tuning fork tests are easy, fast and very reliable.

The Rinne Test

The tuning fork is held next to the external auditory meatus to test air conduction. Then, when the sound is no longer heard, the fork is pressed against the mastoid process.

If the sound is reported to be louder by air conduction than by bone conduction, no conductive hearing loss is indicated and the result is referred to as a positive Rinne test (Figure 3.10).

If the sound is reported as louder by bone conduction than air, the results indicate conductive hearing loss, which is referred to as a negative Rinne test (Figure 3.11).

In the case of total SNHL, the bone conduction is transmitted across the skull and perceived by the patient's contralateral ear. This is called a false-negative Rinne test.

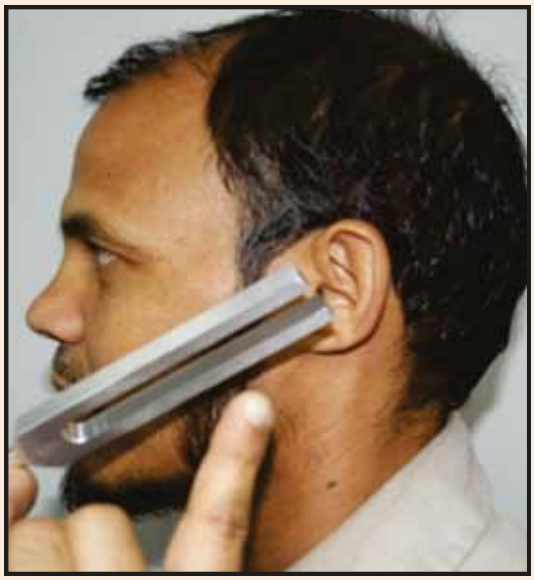


Figure 3.10: *Positive Rinne test*

Figure 3.11: *Negative Rinne test*

The Weber test

This test is used to complement the Rinne test to determine whether unilateral hearing loss is conductive or sensorineural. The vibrating tuning fork is placed in the midline of the forehead and the patient is asked if the sound is heard better on one side or in the middle of the head.

In a patient with symmetrical hearing, the Weber test is centralising.

If the tuning fork lateralises to the same side as the ear with poorer hearing, the hearing loss is conductive. If the tuning fork lateralises to the side opposite the ear with poorer hearing, the hearing loss is sensorineural (Figure 3.12).

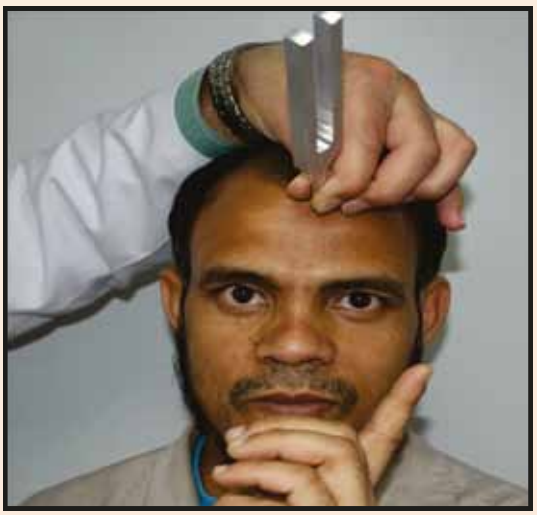


Figure 3.12: *Weber test lateralises to the side with better bone conduction.*

CHAPTER 4

My Child Is Not Speaking

Dr. Khalid A. Hadi

A mother visits the ear, nose and throat clinic with her 3-year-old daughter and is concerned about her speech. The only incoherent words the daughter can say are "ma" and "ba". This is her first child, and she does not know whether she should worry or whether she is exaggerating the issue. Her mother-in-law told her that many children have delayed speech until the age of 5 and speak normally thereafter and that no treatment should be done at this age. She is married to her cousin, and there is no family history of any illness, including hearing loss. The pregnancy was uneventful. The child had jaundice at birth, which required hospitalization for one week; she has had no significant illnesses since that hospitalization. The consultant asks questions about hearing loss, but the mother denies any history. The mother was reassured that her child passed a hearing test at birth called the otoacoustic emissions (OAE) test. Upon examination, the child is hyperactive and difficult to examine. No clear dimorphic features are present, and the ear exam appears normal.

* * * * *

Q1. Why does the consultant continue to ask about hearing loss?

Congenital hearing loss is a very common disease. In many instances, it is a silent and hidden handicap. It is hidden because children, especially infants and toddlers, cannot tell us that they are not hearing well. It is a very serious handicap in children because if it is left undetected and untreated, it will adversely affect speech and language development as well as cause serious social and emotional problems and academic failure. Fortunately, many of the negative results of deafness in babies can be prevented or substantially lessened through early identification and treatment. Many research studies have demonstrated that early intervention with hearing-impaired children results in improved language development, increased academic success and increased lifetime earnings.

Q2. Should she worry about her daughter or she is exaggerating things?

She should definitely worry. By 3 years of age, a child should be able to make short sentences. Deafness in infants is a serious concern because it interferes with the development of language. The longer a child's deafness goes undiscovered, the worse the outcome is likely to be. Children whose hearing loss is not identified until 2 or 3 years of age may suffer from permanent impairments of speech, language and learning.

Q3. Is her mother-in-law correct?

This is a common form of bad advice given to parents from friends and family. Many useful measures can be performed at this age. The diagnosis of hearing loss can be made accurately within the first days of life. Treatments, which might include hearing aids, rehabilitation and cochlear implants, lead to better outcomes if initiated early.

Q4. The mother mentioned that a hearing test was performed in the nursery. What are the hearing screening techniques that can be performed in the nursery?

- The OAE test uses a sensitive microphone to measure an echo or emission response from the cochlea.

- The auditory brainstem response (ABR) is an EEG-like response from the auditory nerve for sounds presented to the ear (Figure 4.1).

- Two-stage screening (OAE + ABR)

Figure 4.1: *ABR performed with electrodes on the head*

Q5. Is it possible that the child has hearing loss even though she passed the OAE?

The OAE test, which is part of universal newborn hearing screening, is a test of cochlear function. It is still possible that the child may have hearing loss due to other reasons:

1. The child may have nerve problem not discovered by OAE.
2. The child may develop hearing loss later in life.
3. The test may give false-negative results.

Q6. If the child is found to have hearing loss based on the ABR, what do the mother and child need and when?

Parents need extensive counseling regarding hearing loss, and children need immediate speech rehabilitation with hearing aids in both ears.

Q7. What are the most important histories in a child with hearing loss?

It is advisable to repeat hearing testing if hearing or language concerns arise in any child in general and particularly in children with any of the following:

- Caregiver concerns for hearing, speech, language or development
- Family history of hearing loss, especially in the presence of consanguinity
- Neonatal intensive care unit (NICU) stays >5 days or any of following (regardless of the length of stay): mechanical ventilation, ototoxic medications (e.g. gentamicin, tobramycin), loop diuretics (e.g. furosemide), hyper-bilirubinemia requiring exchange transfusion
- In utero infections such as cytomegalovirus (CMV), herpes, rubella, syphilis or toxoplasmosis
- Craniofacial anomalies
- Physical findings (e.g., white forelock) (Figure 4.2)
- Syndromes involving hearing loss, e.g., neurofibromatosis, osteopetrosis, Usher, Waardenburg, Alport, Pendred as well as Jervell and Lange Nielson syndrome JLNS
- Neurodegenerative disorders, e.g., Hunter syndrome or sensory motor neuropathies such as Frieidreich's ataxia and Charcot-Marie-Tooth
- Culture-positive postnatal infections associated with hearing loss, e.g., herpes, varicella or meningitis
- Head trauma (basal skull or temporal bone)
- Chemotherapy

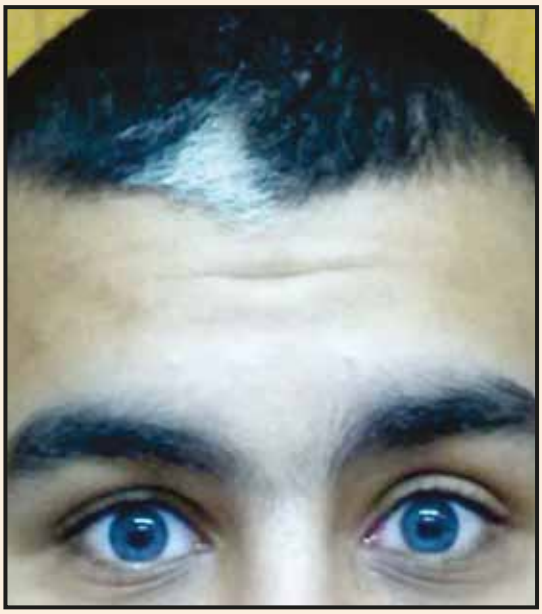


Figure 4.2: Waardenburg syndrome. Notice the white forelock & blue eyes.

Q8. How do you work-up this child?

1. Obtain a complete perinatal and family history.
2. Perform a physical examination for abnormalities.
3. Refer to audiology and radiology for temporal bone CT scan and MRI.
4. Refer to genetics and ophthalmology.
5. Other: Perform CMV test, electrocardiogram (EKG) and developmental evaluations.

Figure 4.3A: *PTA is performed inside a special soundproofed booth.*

Q9. If the mother has hearing loss, what tests can be performed on her?

Many hearing test can be performed to assess adult hearing:

Pure-tone audiometry (PTA) is a subjective hearing sensitivity test performed inside a special soundproof booth (Figure 4.3A). The test is performed at different frequencies (500, 1000, 2000, 4000 and 8000 Hz) to determine the pure-tone threshold (PTT), which is the softest sound audible to an individual at least 50% of the time. The results are plotted as an audiogram. This test involves the peripheral and central auditory systems (Figure 4.3B).

A speech audiogram provides information

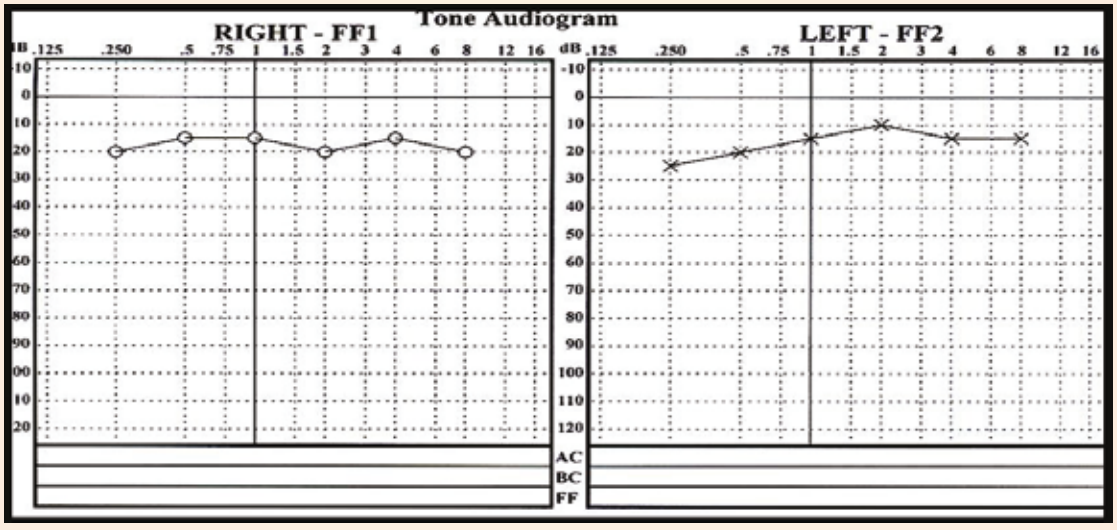


Figure 4.3B: *PTA shows bilateral normal hearing.*

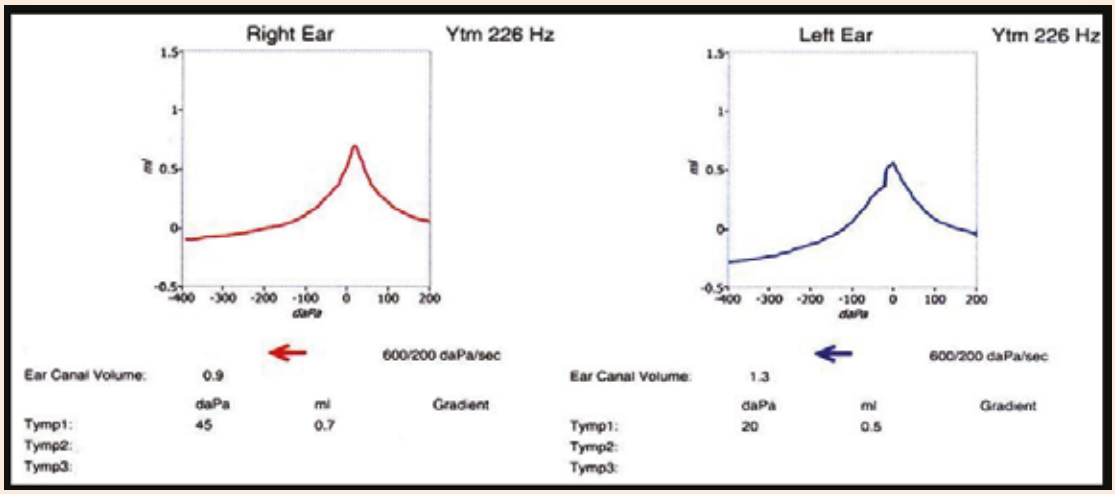


Figure 4.4: *Tympanogram type A bilateral.*

regarding discomfort or tolerance to speech stimuli and word recognition abilities.

Impedance testing determines the status of the tympanic membrane and middle ear via tympanometry to evaluate their shapes and volumes (Figure 4.4). It also evaluates the acoustic reflex pathways, which involve the hearing pathway through the auditory brainstem and cranial nerve VII.

Q10. Because PTA testing is subjective, is it possible to obtain a false result?

Yes, although this outcome is rarely due to malingering. Rather, crossover of the sound from one ear to the other can occur at 50 dB via air conduction and at 0 dB via bone conduction. To overcome the cross response, masking is performed by applying noises to the non-tested ear.

Q11. How can PTA differentiate between conductive and sensorineural hearing loss?

The audiogram is calibrated to measure bone conduction. It is performed when In conductive hearing loss, the threshold of bone conduction is normal, but the air conduction threshold is elevated (Figure 4.5).

The difference between the air and bone thresholds is called the air–bone gap. In sensorineural deafness, both the air and bone thresholds are elevated by almost the same degree (Figures 4.6 and 4.7). In mixed hearing loss, both the air and bone thresholds are elevated, but the bone conduction is better than the air conduction with an air–bone gap (Figure 4.8).

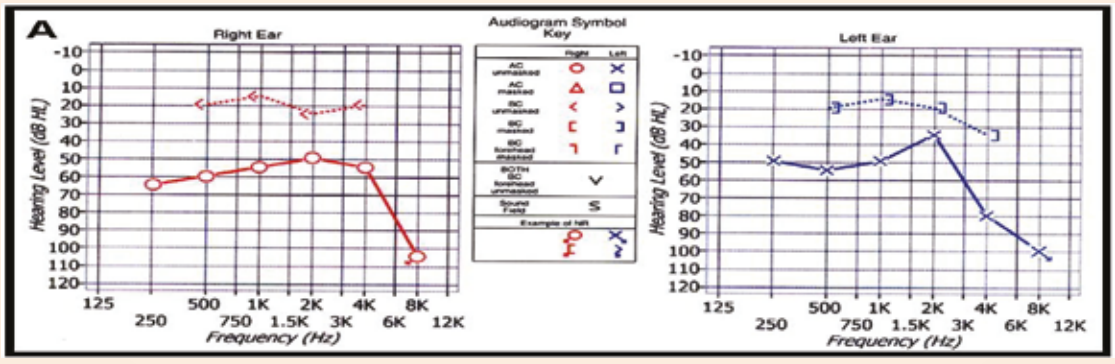


Figure 4.5: *Bilateral conductive hearing loss*

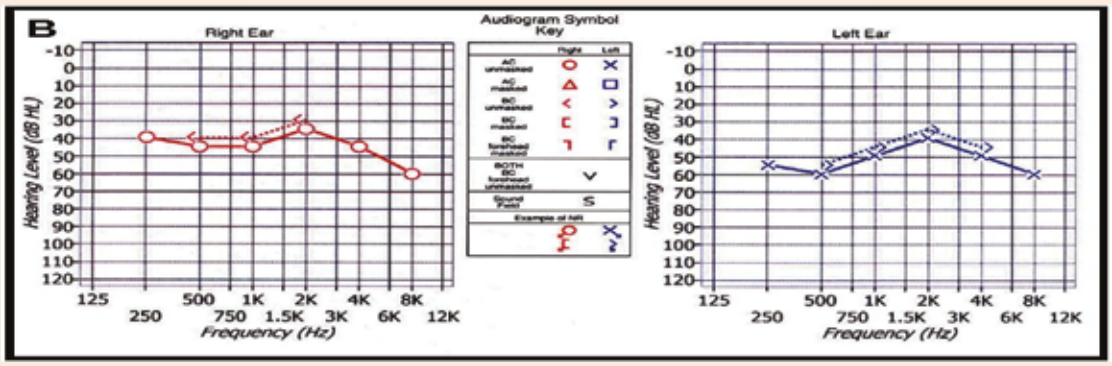


Figure 4.6: *Bilateral sensori neural hearing loss.*

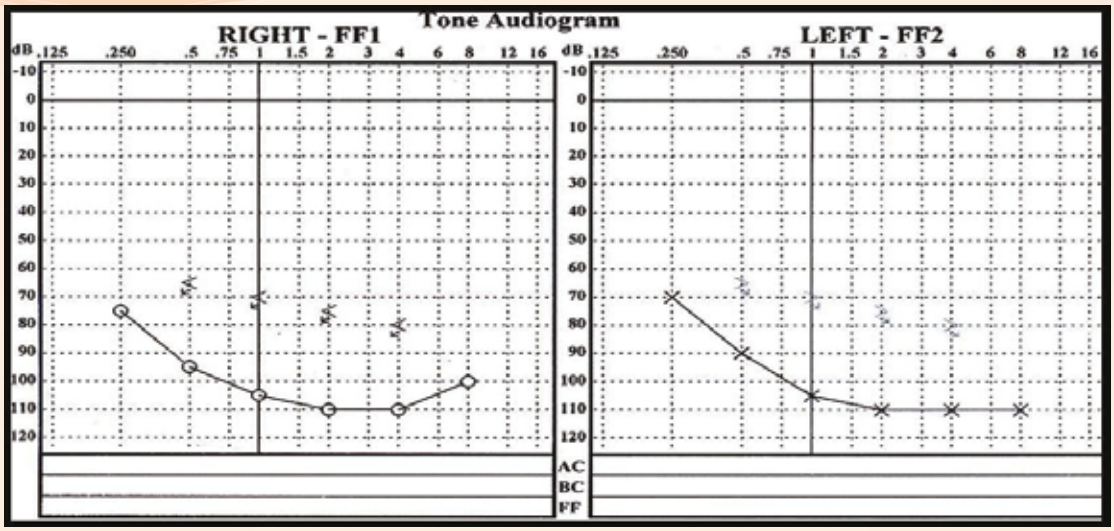


Figure 4.7: *PTA results showing bilateral profound hearing loss.*

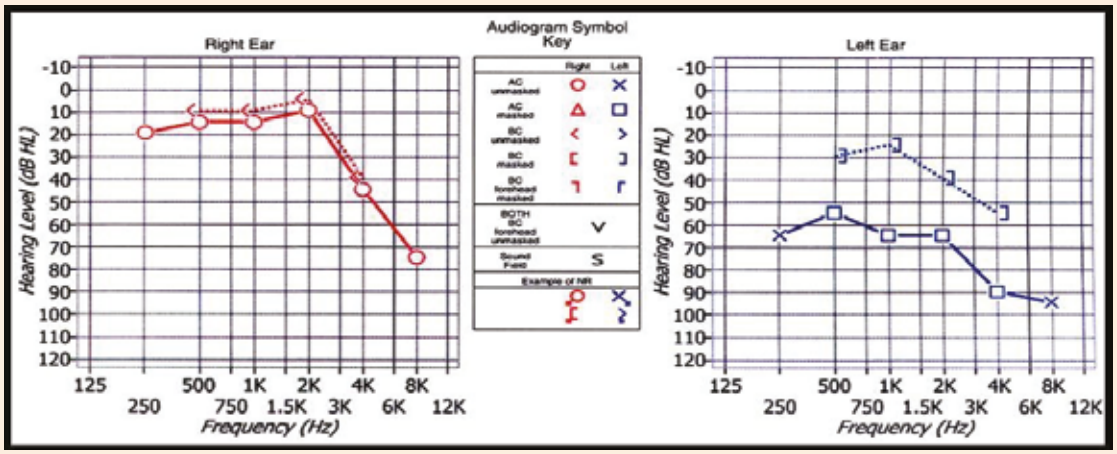


Figure 4.8: *Audiogram Right ear sloping sensori neural hearing loss and audiogram left ear mixed hearing loss.*

Q12. The mother was told that her daughter has 20% hearing loss. What does that mean?

A percentage is not an accurate measure for a hearing assessment because humans can hear many frequencies, and each frequency can be subjected to a different degree of loss. Therefore, the audiogram will allow a better understanding of the hearing loss (two dimensions). The speech audiogram provides even better understanding (three dimensions). However, for simplicity, the following guide can be used:

- Normal (0-25 dB)
- Mild hearing loss (26-40 dB)
- Moderate hearing loss (41-55 dB)
- Moderate-severe hearing loss (56-70 dB)
- Severe hearing loss (71-90 dB)
- Profound hearing loss (>90 dB) (Figure 4.8)

CHAPTER 5

My Father Is Not Communicating!

Dr. Ghada Bin Khamis

A 68-year-old male presents to your clinic with his son. The patient's son is very concerned about his father. His father was previously a very active and social individual, but he started to slowly withdraw, and he has become totally isolated and depressed. This withdrawal started approximately 4 years ago when he started misunderstanding things said to him and started raising the volume on the television. As his hearing got worse, he began to withdraw and stopped attending family gatherings or visiting friends until he became completely isolated, and he now spends his time mainly sitting in his room reading. The son also reports that they tried a hearing aid 3 years ago, but his father rejected it at that time and insisted that he did not need it. He is an otherwise healthy male with no major health concerns.

When you talk to the old man, you have to raise your voice, but he still is not able to understand what you are saying. However, he insists that he is fine and does not understand why his family keeps telling him that he cannot hear or why his son brought him to see a doctor.

* * * * *

Q1. What changes occur in the aging auditory system that must be considered when treating elderly patients?

Changes in the external ear include thinning of the skin, dehydration, a decrease in fat and a decrease in collagen. Additionally, the hair becomes thicker and longer. These changes lead to t he feeling of itchy ears, an increased likelihood of trauma to the ear canal and reduced tolerance for hard materials.

Changes in the middle ear involve the tympanic membrane, which becomes thicker, dull and less elastic, and the ossicular chain, which becomes calcified and undergoes arthritic changes in its synovial joints. In addition, calcification of the cartilaginous support of the

Eustachian tubes occurs. These changes lead to an increased probability of middle ear effusions in association with upper respiratory tract infections (URTIs), which are common in older individuals due to changes in the respiratory system.

Changes in the inner ear include the loss of cochlear outer and inner hair cells, stiffening of the basilar membrane, thickening of the capillaries in the stria vascularis, degeneration of spiral ganglion cells and loss of cochlear neurons. These changes lead to reduced hearing acuity, reduced tolerance of loud sounds and decreased ability to understand speech, especially in the presence of background noise.

Changes in the central auditory connections include the loss of neurons, changes in neuron size, decreased dendrite branching and increased white matter lesions. Additionally, an overall reduction in the size of the brain occurs. These factors lead to a reduction in the speed of signal processing, greater difficulties understanding conversation in the presence of competing signals, an increased likelihood of auditory processing disorders, changes in secondary memory, increased difficulty and effort when learning new things and decreased ability to process and integrate different sensory modalities.

Q2. What are the impacts of untreated hearing loss in older adults?

Hearing loss has a detrimental effect on social well-being, communication, cognitive function and functional health status. These effects include the following:

- Avoidance and withdrawal from situations that require communication and situations in which communication is difficult, such as family gatherings, restaurants and speaking on the telephone. This withdrawal may ultimately lead to social isolation, sadness and depression.

- Embarrassment, fatigue, irritability, tension, stress, depression, negativism, paranoia, loneliness, avoidance of or withdrawal from social activities, danger to personal safety, rejection by others, reduced general health, decreased awareness of the environment, impaired memory, decreased adaptability to learning new tasks, decreased ability to cope and reduced overall psychological health.

- Stress in relationships with others because family, friends and co-workers experience frustration, impatience, anger, pity and guilt when interacting with a person with hearing loss.

- Misunderstandings in the physician –patient relationship regarding his/her diagnosis, treatment and medication.

- Increased likelihood of developing dementia.

Q3. What tests should be included in the audiological test battery?

- Pure-tone audiometry
- Immittance measurements, including tympanometry and stapedial reflexes (thresholds and decay both ipsilaterally and contralaterally)
- Speech audiometry, including speech recognition thresholds and word recognition / discrimination scores (if hearing loss permits)

Q4. The audiological testing revealed bilateral high frequency moderate SNHL sloping to profound (Figure 5.1). What are your recommendations?

- Hearing aids for both ears. Binaural amplification is essential for better hearing and better understanding.
- Cochlear implant work-up may be needed later. Hearing aids may not be sufficient for this patient because of the degree of hearing loss in the high frequencies.

Q5. The patient is resistant and is still not interested in hearing aids. What can you do?

- Counsel about the importance of hearing aid use.
- Counsel about the potential negative impacts of untreated hearing loss (from Q2).
- Counsel about the hearing aid adjustment process and let the patient know that he may initially feel uncomfortable and that the sound quality through the hearing aids may be strange, but he will adjust to the hearing aids' sound quality after using them for some time.
- Enlist family members in the counseling and treatment process because the patient will need their support.
- Make sure the patient is aware that the medical team (physician, audiologist and speech - language pathologist) will be working with him throughout the process.

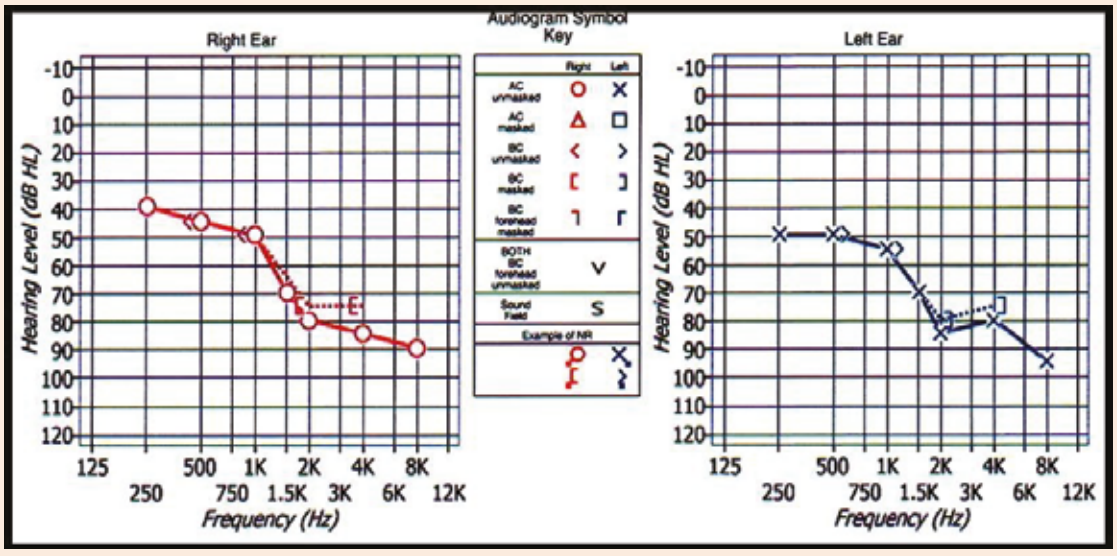


Figure 5.1: Bilateral high frequency moderate SNHL sloping to profound.

- The audiologist will work with him and continue re-programming and adjusting his hearing aids until he is able to adapt to them.
- The speech-language pathologist and the hearing therapist will work with him on auditory training and aural rehabilitation to adjust to his hearing aids.
- The patient may need time to accept treatment, and the medical team must be patient and work with the patient and his family to reach their goals.

Q6. His son is wondering how his father could suffer hearing loss and at the same time exhibit intolerance to noise?

This is common feature in sensorineural hearing loss when the cochlea is the site of the pathology. The phenomenon is called recruitment, which is the perception of disproportionate loudness. The cochlea works as an energy converter, amplifier and filter for the sound.

Q7. Why would you consider a cochlear implant for this patient?

- Hearing aids transmit sound using the ears' natural transduction process. The impaired auditory system presents limitations in audibility, comfort with loudness (recruitment), clarity and intelligibility. These limitations reduce the benefits of hearing aids.
- Cochlear implants directly stimulate the spiral ganglion of the auditory nerve with electrical stimulation and do not require the hair cells. A cochlear implant is also not affected by cochlear dead regions, which affect intelligibility. Cochlear implants avoid surges of neural activation, which lead to loudness via recruitment by activating the auditory nerve fibers in a manner that is limited by gain controls.

Q8. What referrals are necessary and why?

- Cochlear implant surgeon: to initiate the cochlear implant medical work up.
- Cochlear implant audiologist: for cochlear implant audiological evaluation and to ensure proper hearing aid fitting, hearing aid use and hearing aid benefit.
- Speech language pathologist and / or hearing therapist: Auditory training and aural rehabilitation should start as soon as the hearing aids have been fitted. This is essential for the patient to achieve maximum benefit from his hearing aids, to evaluate the amount of benefit from the hearing aids, to evaluate whether a cochlear implant is recommended and to prepare the patient for the post-cochlear implant rehabilitation.

- Psychologist and / or psychiatrist: to assist the patient and his family in coping with the hearing loss and to manage the patient's depression.

Q9. Will the patient's age affect the outcome of the cochlear implant?

Age has little effect on the outcomes of a cochlear implant in the post-lingually deafened. The duration of deafness is more important. For children, the cochlear implants should be places as early as possible (Figure 5.2). Minimum benefits can be expected after 5 years old, which is the cut-off age between pre-lingual and post-lingual social interactions.

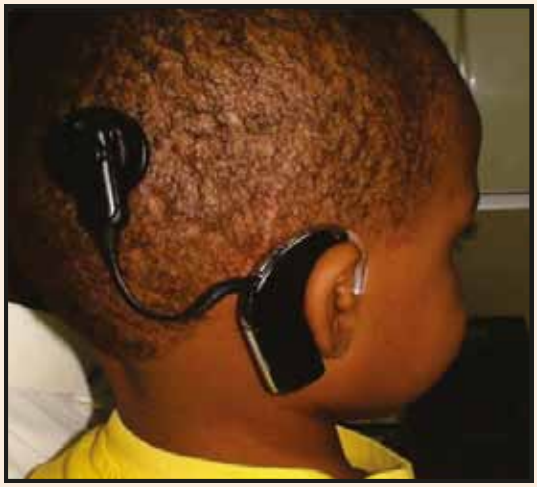


Figure 5.2: *Child with a cochlear implant.*

Q10. Do older adults perform well with cochlear implants?

Yes, older adults show significant improvements after receiving cochlear implants compared with their pre-implant performance. However, they do not perform as well as younger adults.

Q11. Is the audiogram sufficient to decide whether a patient is a cochlear implant candidate?

The audiogram alone is not enough. It is possible for the audiogram to indicate that hearing aids are suitable when they are not. Questions you may ask include the following:

- Is your patient able to talk on the phone easily with people he does not know?
- Is your patients' inability to understand conversation (even with the hearing aids) limiting his interactions with others or his social life?
- How is his hearing loss affecting his life?

Q12. Would you consider bilateral cochlear implants for this patient?

Bilateral cochlear implants could be an option. However, bimodal hearing (i.e., a hearing aid in one ear and a cochlear implant in the other ear) may be a better option for this patient because he has some aidable residual hearing in the mid and low frequencies.

Q13. What are the criteria for a cochlear implant?

Bilateral severe-to-profound SNHL

- Minimal benefit from bilateral powerful hearing aids
- Post-lingual deafness (preferably less than 5 years of age for pre-lingual patients).
- Fit for surgery

- Normal cochlea and auditory nerve
- Appropriate expectations of outcomes

Q14. If the patient has a bilateral acoustic neuroma that requires resection, is their any option for the patient to hear?

Yes, the patient still can be helped by implanting electrodes in the brainstem, which is called brainstem implant (ABI) (Figure 5.3).

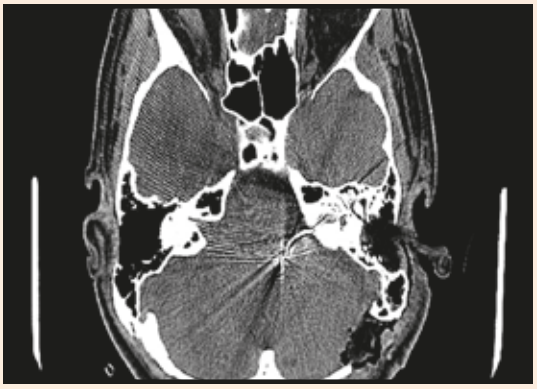


Figure 5.3: *CT scan showing ABI electrodes in the brainstem.*

CHAPTER 6

Recurrent Dizziness

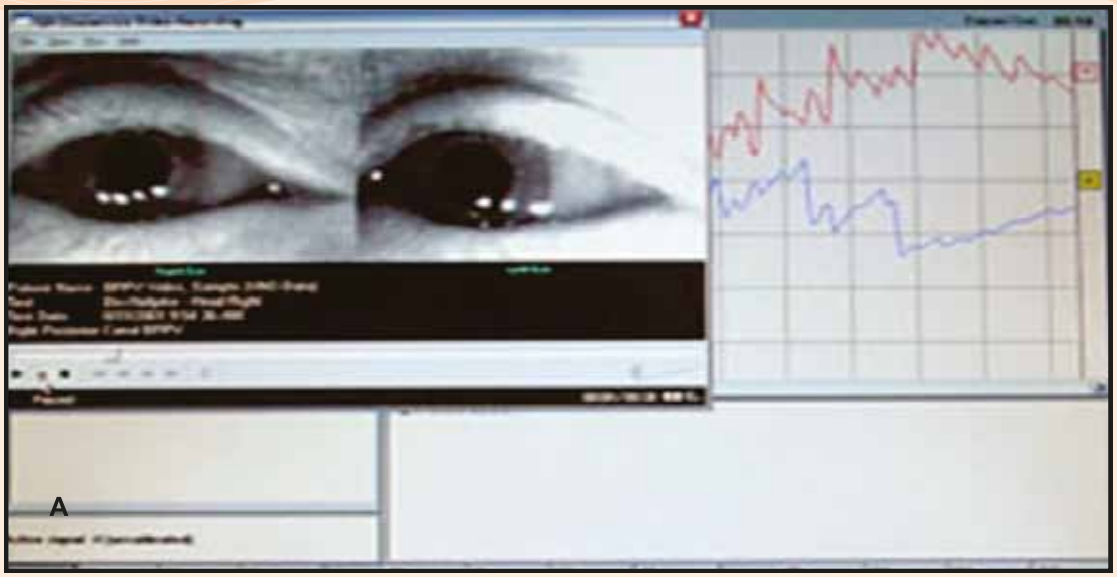
Dr. Murad Al-Momani

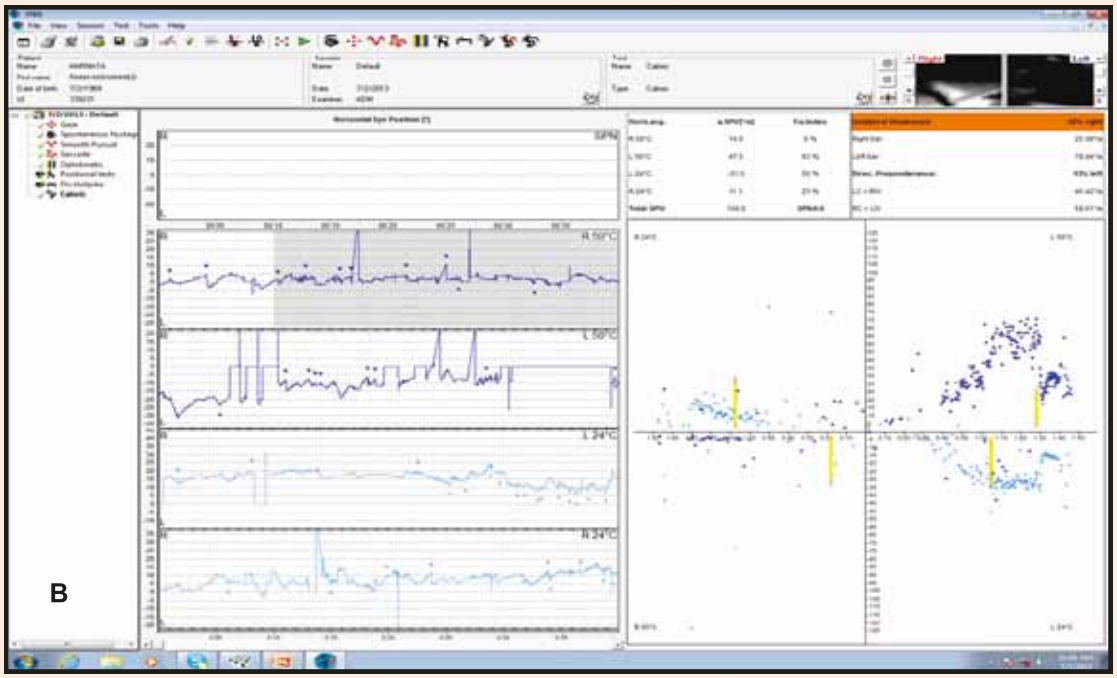
A 28-year-old female presents to the ENT clinic complaining of nausea, ear fullness and dizziness. She says, "I feel like things are moving around me," and this feeling lasts for several hours before it improves and then disappears. She also reports that her hearing is gradually worsening, especially in her right ear. The hearing loss in the right ear becomes more severe when she gets the dizzy spells. Moreover, she reports hearing a "roaring" sound in the right ear, which increases in intensity just before and during the dizzy spells. She dates the start of these symptoms to approximately three years ago. Initially, the symptoms were mild and infrequent, but with time they have increased in frequency and severity. Initially, she used to experience these symptoms on average every 6 months, but the frequency has been gradually increasing to a current average of every other week.

She has been treated by her family practitioner, who diagnosed her with migraine and prescribed Advil (ibuprofen) for the headache and Reglan (metoclopramide) for the nausea. However, no improvement was noticed by the patient. A recent examination by an ENT doctor revealed normal external and middle ears. She was then referred to an audiology clinic for a vestibular and hearing assessment. Hearing tests revealed normal middle ears bilaterally and mild-low frequency SNHL in the right ear. The vestibular assessment included vestibulonystagmography (VNG) and caloric testing (irrigation of the ear using cold and warm air or water stream). The VNG was performed using goggles covering the eyes with small cameras to record the eye movements during the testing (Figure 6.1). The caloric testing showed abnormal right-sided weakness (42%). The results of the VNG are shown below (Figures 6.2A, B and C).

* * * * *

Figure 6.1: *VNG testing performed with goggles*





C. VNG

Test	Result
Spontaneous nystagmus	Absent (normal)
Saccadic eye movements	Normal
Smooth pursuit eye movements	Normal
Optokinetic eye movements	Normal
Positional nystagmus	Absent (normal)
Dix-Hallpikenystagmus	Negative bilaterally (normal)

Figures 6.2A, B and C: *VNG recording of eye movements and analysis of the tracings.*

Q1. What is the most likely cause of the patient's symptoms?

The patient's history and the test results indicate Meniere's disease in the right ear.

Q2. What does the term dizziness mean?

Dizziness is defined as disorientation in space, a sense of unsteadiness, a feeling of movement within the head such as giddiness or a swimming sensation, light-headedness or a whirling sensation. Dizziness changes the sense of balance and can increase the risk of falling.

Q3. What are the common types of dizziness?

Vertigo (the illusion of movement of the self or nearby objects), pre-syncope (light-headedness or faintness), disequilibrium (unsteadiness on the feet) and "other" (usually a floating sensation).

Q4. What are the causes of dizziness?

Because the mechanisms for maintaining balance are complex, determining the exact cause of dizziness is often difficult and requires input from several medical specialties. Dizziness can be caused by various problems associated with the inner ear, brain or heart.

Q5. What are the common causes of vertigo?

- Meniere's disease is associated with the fluid balance-regulating system in the inner ear resulting in an increase in the endolymph within the membranous labyrinth. Symptoms include episodic attacks of tinnitus (ringing in the ear), ear fullness, hearing loss and attacks of vertigo of variable intensity and duration that may be accompanied by nausea and vomiting. This condition can be treated with dietary changes and medications. Surgery may be recommended if medical treatment is not successful.
- Benign paroxysmal positional vertigo (BPPV) is a disorder in which changing the head's position with respect to gravity leads to sudden vertigo, which is a feeling that the room is spinning. The vertigo can vary in its intensity from mild to severe; usually lasts for less than 1 minute; and may be accompanied by other symptoms, including dizziness, light-headedness, a sense of imbalance, nausea and vomiting.
- Vestibular neuritis (viral labyrinthitis) is caused by inflammation of the vestibular nerve, most likely due to a virus infection. The main symptom is the sudden onset of sustained vertigo that lasts for a few days or weeks. Characteristically, there is no hearing loss. Treatments include medications to relieve the symptoms of dizziness and nausea and to restore balance.

- Autoimmune inner-ear disease is more common in people who have other autoimmune diseases, such as lupus and arthritis. Symptoms include sudden hearing loss in both ears. Medical treatment includes tapered steroid therapy over 1-2 months.
- Perilymph fistula may occur after a sudden change in barometric pressure, such as the change experienced during flight. The change in pressure can cause a rupture of the oval or round windows membranes that normally separate the middle and inner ears. Symptoms include hearing loss, vertigo, light-headedness and ear pressure. Often, the rupture heals spontaneously. If it does not heal, surgery may be required.

Q6. What are the central causes of dizziness?

They include acoustic neuroma, brainstem glioma, multiple sclerosis, brainstem infarction, migraine headache and neurofibromatosis.

Q7. What is nystagmus?

Nystagmus refers to uncontrolled eye movements. Nystagmus usually involves quick, jittery movements made by both eyes, either horizontally or vertically.

Q8. What is Meniere's disease?

Meniere's disease is thought to result from an increase in the inner-ear fluid called endolymph. The disease is sometimes called "endolymphatic hydrops." This increase in pressure causes symptoms such as vertigo, hearing loss, ear fullness and tinnitus (ringing or roaring in the ears). The cause of this condition is not known with certainty (idiopathic), but several causes have been suggested, including autoimmune processes, genetic disorders and viral infections.

Q9. Is Meniere's disease curable?

Spontaneous recovery may occur in some cases. Certain medications are used to control the symptoms, such as Valium and Betaserc (betahistine). Although these treatments might help in mild cases, they are not effective in more severe cases. A low-salt, low-caffeine diet decreases the severity of the symptoms. Other treatment modalities include intratympanic application of gentamicin or steroids and surgical procedures to drain the endolymph or vestibular nerve neurectomy in selected cases.

Q10. How can the vestibular and audiological tests help in diagnosis?

They can help to determine the possible site of the lesion. Although it is not always possible to precisely determine the site of the lesion, it is usually possible to differentiate between peripheral and central causes of dizziness using vestibular and audiological test results (Figures 6.3, 6.4 and 6.5).

Figures 6.3A and B: *Irrigation of the ear using warm and cold air during caloric testing. The recording involved the use of video goggles to record eye nystagmus.*

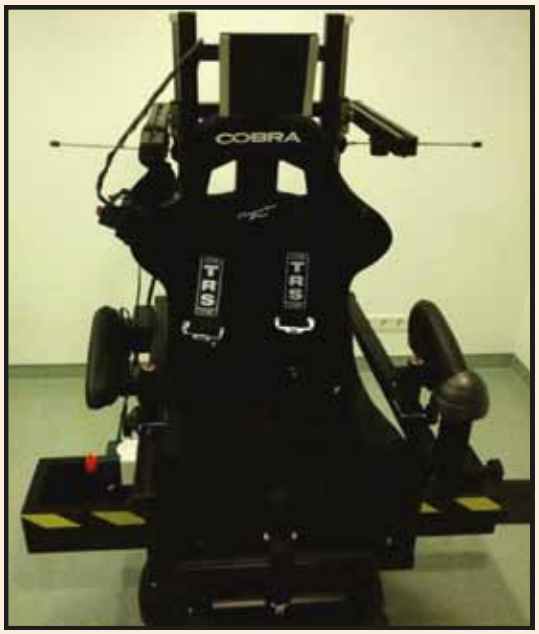


Figure 6.4: *Rotational testing using a computerised rotary chair and recording the eye nystagmus via the video goggles*

Figure 6.5: Posturographic testing that involved computer-controlled movable foot plates to record the response of the body to different degrees and directions of movement.

11. What other tests might be used to help in diagnosis?

The ENT doctors may ask for imaging studies such as a CT or MRI to rule out central causes including tumors. Other testing may involve a complete blood count (CBC) and other blood work to screen for other possible causes such as infection.

CHAPTER 7

I'm Spinning

Prof. Abdulrahman Hagr

A 36-year-old female complains of positional dizziness that began 3 weeks before evaluation in the ENT clinic. Her symptoms occur when she rolls over in bed to the right side and are precipitated by specific head movements (neck extension and rotation). These brief episodes last for less than one minute. There are no other neurologic or otologic symptoms. Her history is significant for a 4-day episode of severe dizziness, nausea and vomiting that occurred 2 months before evaluation and was preceded by a head cold. She never had any ear symptoms. A family physician at that time diagnosed her with labyrinthitis and prescribed promethazine, which the patient is still taking on an irregular basis. Following this acute episode, the patient has experienced several days of nausea and severe imbalance, and she was unable to practice her work as a dentist for approximately one week. Since returning to work, she reports a sense of mild disequilibrium and unsteadiness during quick head movements. These symptoms have gradually improved, and she is almost entirely asymptomatic at rest, though she still complains of positional vertigo. The family history is non-contributory.

The general examination is normal. Neurologic examination reveals no nystagmus or impaired tandem gait. Her cranial nerves, cerebellar function and ear examinations are normal.

* * * * *

Q1. She is wondering whether she has Meniere's disease like her sister.

She does not have the classic symptoms of Meniere's disease. This disorder is characterized by episodes of vertigo, ear fullness, tinnitus and fluctuating hearing loss during the attacks, which may last for more than 20 minutes.

Q2. What is the most likely diagnosis?

1. Vestibular neuritis 2 months prior
2. Benign paroxysmal positional vertigo (BPPV) for her persistent symptoms

Q3. What additional examinations help in this patient's diagnosis?

- Dix-Hallpike manoeuvres (Figure 7.1) The induced vertigo and nystagmus of BPPV are characterized by having a latent period and being fatigable. The direction of the nystagmus (quick component) is toward the lower ear.
- Examination of the patient's eyes while the patient is wearing magnifying glasses (Frenzl) (Figure 7.2), which abolish fixation to demonstrate nystagmus.

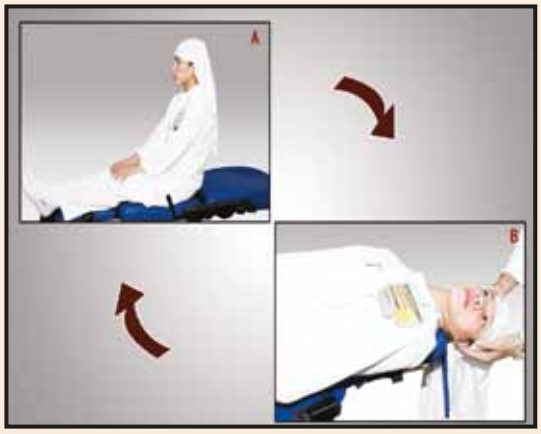


Figure 7.1: *The Dix-Hallpike test of the right ear. In Panel A, the patient is sitting upright with the legs extended, while the examiner stands behind the patient and rotates the patient's head 45° toward the tested ear. In Panel B, with the eyes open, the examiner moves the patient from the seated position to supine with the right ear down. The patient's eyes are then observed for at least 1 minute because there is a characteristic period of latency prior to the onset of nystagmus.*

Figure 7.2: *Magnifying and illuminating (Frenzl) glasses can be very useful in illustrating nystagmus, fixation is removed as the patient can hardly focus through magnifying glasses on a dark room.*

Q4. What are the appropriate tests?

1. Pure-tone audiogram
2. Tympanogram
3. Stapedial reflex
4. Electronystagmogram (ENG) or videonystagmography

If the patient has BPPV, only an ENG can show the nystagmus; the other tests are useful for excluding other causes.

Blood tests and CT or MRI scans of the ears and brain do not reveal any characteristic features in this disease. They are only performed if there is an atypical presentation to rule out other differential diagnoses.

Q5. Is it possible to have more than one diagnosis for vertigo in the same patient?

Yes, patients may exhibit vestibular neuritis followed by Meniere's disease or BPPV.

Q6. What are the pathophysiological sites of BPPV, Meniere's disease and vestibular neuritis?

BPPV: The utricle and the semi-circular canals (usually the posterior canal)

Meniere's disease: Membranous labyrinth

Vestibular neuritis: Vestibular nerve

Q7. What are the treatment options for BPPV?

1. Repositioning maneuvers (Epley's maneuver) (Figure 7.3)
2. Vestibular suppressant medications.

Figure 7.3: *Epley manoeuvre. Patient is quickly and passively forced down backwards by the clinician into a supine position with patient's head rotated until the left ear is down (Panel B). Once the nystagmus ceases, the head and body are rotated until the head is face down (Panel C). Finally, the patient is slowly brought up to an upright sitting posture (Panel D).*

Q8. What is the natural history of BPPV?

BPPV is a self-limited condition that resolves spontaneously over weeks to months.

Treatment manoeuvres can result in an immediate resolution of the symptoms, but it may recur.

Q9. What are the major categories of dizziness?

1. Non-vestibular
2. Vestibular (peripheral or central)

Q10. What symptoms suggest peripheral vestibular system dizziness?

Hearing loss, tinnitus, ear fullness, ear drainage, previous ear surgeries and ear trauma.

Q11. What symptoms should be recognised as emergent cases necessitating urgent test?

Loss of consciousness, severe headache and visual, sensory or motor losses.

Q12. What are the main central vestibular disorders that cause dizziness?

1. Migraines
2. Transient ischemic attacks (TIAs)
3. Cerebrovascular accidents (CVAs)
4. CNS vasculitis
5. CNS tumors

Q13. What is the difference between vertigo and dizziness?

Vertigo is the illusion that one's body or the surrounding environment is spinning. Dizziness is non-specific; it frequently refers to disequilibrium, unsteadiness or light-headedness.

CHAPTER 8

Ear Discharge

Prof. Leila Telmesani

A 24-year-old male presents to the clinic complaining of non-offensive right-ear discharge and right-side decreased hearing. His symptoms started in early childhood. Since then, he has experienced recurrent ear discharge whenever he develops an URTI or when water gets into his ear. There is no history of tinnitus or vertigo. His past medical and surgical histories are negative.

The general examination is normal. An ear examination shows a moderately sized central perforation of his right tympanic membrane with a thick edematous middle ear mucosa and mucopurulent discharge from the perforation. There is no tenderness of the tragus or the mastoid bone. The left ear is normal. Rinne's test is negative in the right ear and positive in the left ear. Weber's test lateralizes to the right side.

* * * * *

Q1. The patient wants to know if he has an infection in the external ear canal (otitis externa).

Discharge from the external ear can be purulent or watery but never mucopurulent because the skin of the external ear canal does not have mucous glands. Otitis externa is usually associated with itching and ear pain (otalgia) that worsens with mastication or movement of the auricle. Generally, otitis externa is not associated with hearing loss unless the external canal is completely obstructed. From the history, this patient's condition is unlikely to be due to otitis externa.

Q2. What is the most likely diagnosis?

Acute suppurative otitis media is not a likely diagnosis because the onset of the symptoms was insidious and because of the long duration. Additionally, CSF otorrhea is not a likely diagnosis because the discharge is not watery, not subsequent to trauma (e.g., motor vehicle accident (MVA) or ear surgery), and does not worsen with straining. The most likely diagnosis in this patient

is the tubo-tympanic type of chronic suppurative otitis media (CSOM) with acute exacerbation (Figure 8.1). It is not the atticoantral (cholesteatoma) type because the discharge is not offensive and not constantly present. In addition, the examination revealed a central perforation.

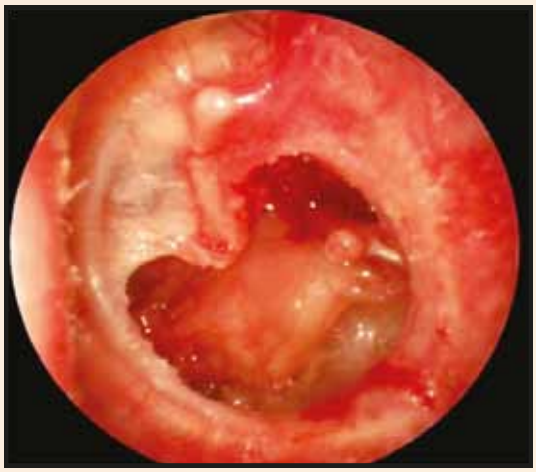


Figure 8.1: *Central perforation with ear infection.*

Q3. What are the possible predisposing factors that contributed to this condition?

Most likely, the patient had an acute suppurative otitis media that started in childhood and failed to heal, producing a permanent perforation in the pars tensa due to one or more of the following factors:

- High-virulence organisms such as β -hemolytic streptococci
- Poorly treated acute suppurative otitis media due to inappropriate antibiotics or patient non-compliance
- Reduced immunocompetence due to poor socio-economic status or another illness

Q4. Why do we call this condition the tubo-tympanic type?

The pathology is in the mucosa of the Eustachian tube, middle ear and tympanic membrane (the anterior and inferior parts of the middle ear cleft). It is also referred to as the "safe" type of chronic suppurative otitis media (Figure 8.2).

Figure 8.2: *Inflammation of the mucosa of the Eustachian tube, middle ear and tympanic membrane is considered relatively safe.*

Q5. Why does the patient experience decreased hearing?

The conductive hearing loss in CSOM depends on the site and extent of the pathology:

- If the CSOM involves the tympanic membrane, the hearing loss is usually mild conductive deafness (Figure 8.3).
- If the CSOM involves almost all of the tympanic membrane (subtotal perforation) exposing the right window, the

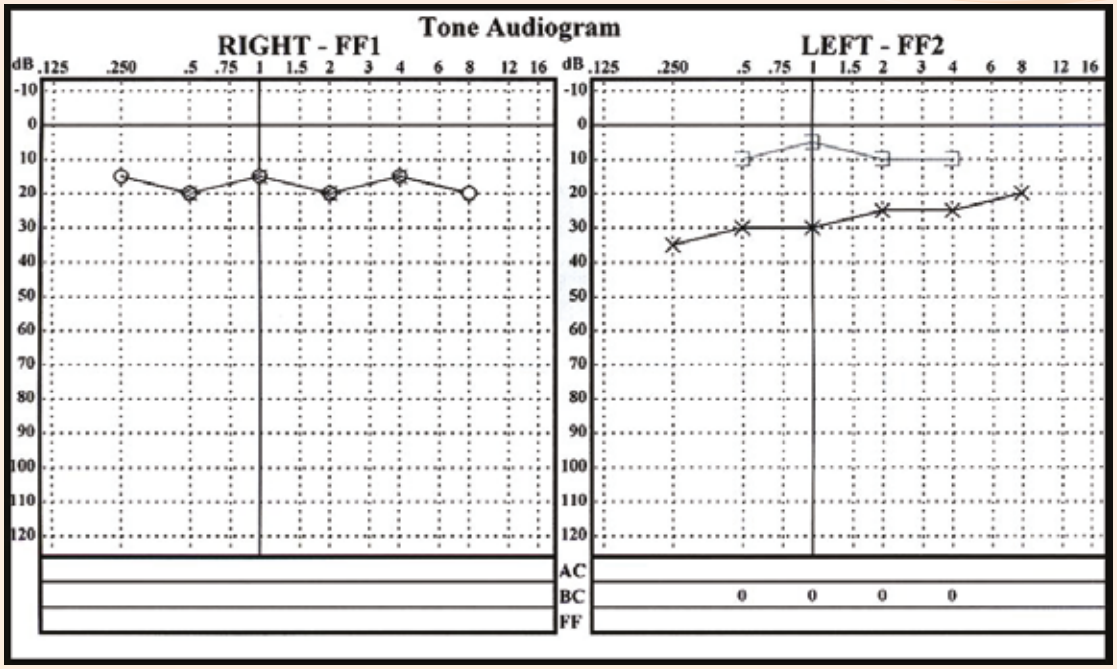


Figure 8.3: *PTA showing unilateral conductive hearing loss with a small air-bone gap.*

condition can cause moderate conductive hearing loss.

- If the CSOM has caused disruption of the ossicular chain (the long process of the incus is generally affected first due to its poor blood supply), the patient will experience severe conductive deafness.

Q6. What is the cause of the recurrent ear discharge?

Activation of the infection usually follows either

1. URTIs
2. Introduction of water into the external ear canal

Q7. What tests should be performed in this patient?

1. After proper microscopic aural toilet, a swab from the ear for culture and sensitivity may show aerobic organisms such as *Pseudomonas aeruginosa*, *Proteus* spp., *Escherichia coli* and *Staphylococcus aureus* or anaerobes such as *Bacteroides fragilis* or anaerobic streptococci.
2. Audiological evaluation of the patient's hearing should be performed, including a pure-tone audiogram; speech reception thresholds; and speech discrimination scores to determine the type and degree of hearing loss, which rarely exceeds 50 dB.

The hearing loss is usually conductive, but with long-standing infections, mixed hearing loss can occur due to toxin absorption into the inner ear.

Q8. What is the treatment for this condition?

A. Conservative Treatment

1. Aural toilet and appropriate systemic antibiotics for acute infections.
2. Treat the contributing factors (e.g., allergies, rhinosinusitis, infected tonsils and adenoids).
3. Advise the patient to keep his ear dry by using ear plugs when showering or swimming.

B. Surgical Treatment

1. If the patient is bothered by the inconvenience of taking precautions to prevent water entry into his ear or if he has significant deafness, advise the patient regarding:
   1.1 Myringoplasty: grafting only the tympanic membranes
   1.2 Tympanoplasty: inspecting the middle ear, removing any pathology, reconstructing the ossicular chain and grafting the tympanic membrane
2. Surgical treatment of the contributing factors (e.g., a deviated nasal septum)

Q9. What are the possible complications of the surgery?

Complications are rare, but the following can happen:

a. Failure of the graft
b. Persistent hearing loss
c. Facial nerve palsy (very rare)
d. Sensorineural hearing loss (rare)

Q10. What are the consequences if the condition is left untreated?

Serious complications are rare. Adhesions (Figure 8.4) in the middle ear might develop along with tympanosclerosis. The latter is observed as a white deposit in the remnant of the tympanic membrane or on the promontory, ossicles or oval window (Figure 8.5). The condition develops secondary to hyalinization and subsequent calcification of the subepithelial connective tissue. Both pathologies might lead to further decrease in hearing.

Long-standing cases (recurrent infections) and the absorption of toxins through the round window and oval window can damage the cochlea and result in SNHL.

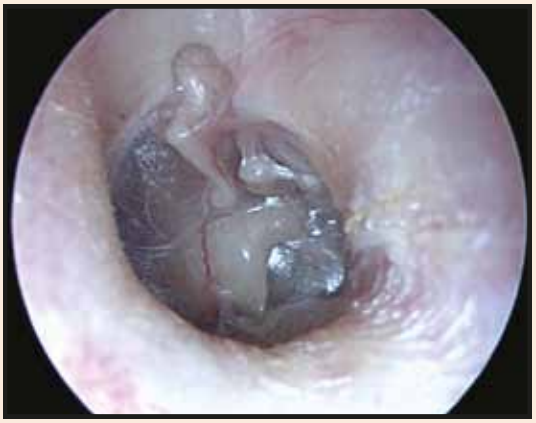


Figure 8.4: *Adhesion of the eardrum to the ossicles and the medial wall of the middle ear. (courtesy of Dr. Munahi Al-Qahtani)*

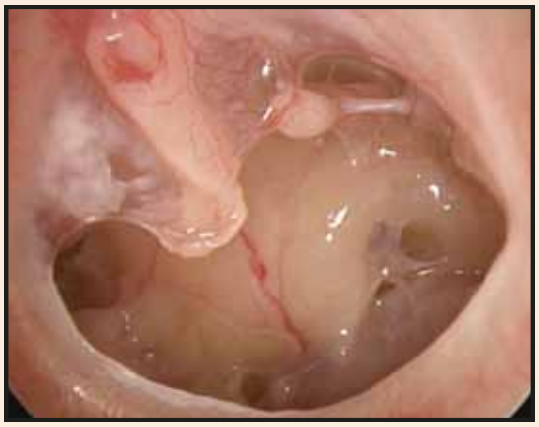


Figure 8.5: *Tympanosclerosis in the remaining part of the eardrum. Note the large inferior perforation and the clear middle ear mucosa. The long process of the incus is absent. (Courtesy of Dr. Munahi Al-Qahtani)*

Q11. What is the management of patients with frequent and persistent ear discharge despite water precautions and the use of proper antibiotics?

In this case, the patient might have a reservoir of infection in the mastoid (chronic mastoiditis). A CT scan of the mastoid will confirm the diagnosis and provide insight into the detailed anatomy and extent of the disease (Figure 8.6).

The treatment includes the following:

- Cortical mastoidectomy (clearing disease from the mastoid antrum and mastoid air cells) to establish communication between the mastoid antrum and the middle ear).
- Tympanoplasty or myringoplasty can be performed simultaneously or after the mastoidectomy.

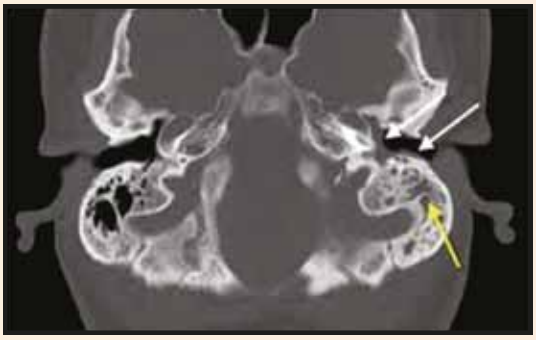


Figure 8.6: *CT scan showing temporal bone axial cut showing opacity of the mastoid air cells and thickening of the external canal and middle ear lining.*

CHAPTER 9

Ringing In The Ears

Dr. Abdulmonem Al Sheikh

A 33-year-old female who is 24 weeks pregnant presents to the clinic with worsening right ear tinnitus and impaired hearing. The problem started 2 years prior, with a slowly progressive hearing impairment that rapidly worsened during pregnancy. She has had no vertigo or ear discharge. She has not travelled recently and has not experienced any infections or trauma. She reports hearing better in a noisy environment. The patient appears well on examination. Her right ear examination shows a normal external ear and tympanic membrane, but there is redness noted at the promontory medial to the tympanic membrane. Her left ear is completely normal. The Rinne test was negative on the right side and positive on the left, while the Weber test lateralized to the right side. The rest of the head and neck examination is normal.

* * * * *

Q1. What is the next measure to confirm your diagnosis?

Pure tone audiogram and tympanometry. The audiogram is shown in Figure 9.1

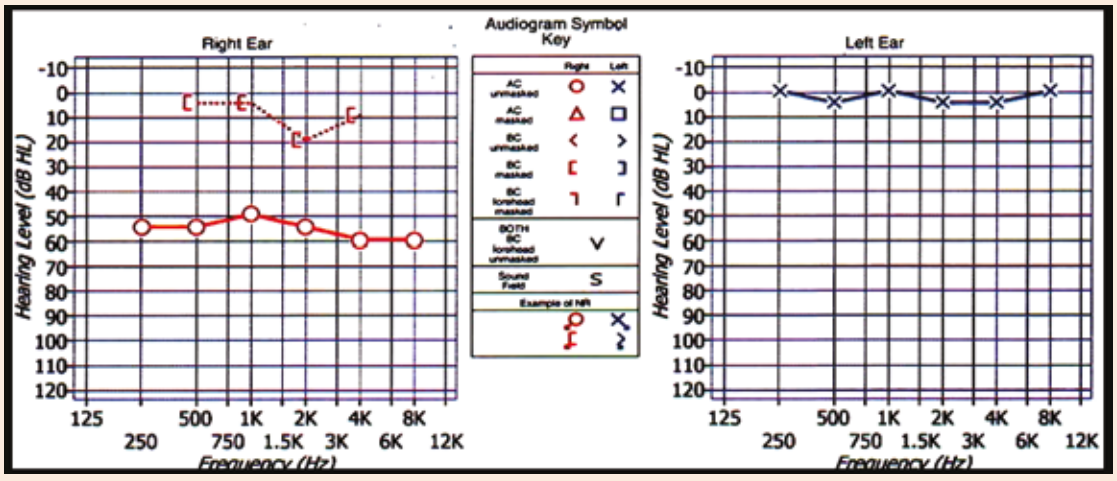


Figure 9.1: *Pure tone audiogram showing conductive hearing loss in the right ear*

Q2. What is the most likely diagnosis?

The most likely diagnosis is right-sided otosclerosis.

Q3. What is otosclerosis?

Otosclerosis is a disease of the bony otic capsule characterised by abnormal replacement of the mature bone of the otic capsule by woven bone of greater thickness, which will eventually result in stapes Figure 9.2) fixation.

Figure 9.2: *Stapes (Photographs courtesy of the personal collection of Dr. David P. Morris FRCS (ORLHNS) MD).*

Q4. What are other differential diagnoses?

The differential diagnosis should include most of the causes of conductive hearing loss, including

1. Tympanosclerosis
2. Post-traumatic dislocation or fracture of the ossicles
3. Adhesive otitis media or otitis media with effusion
4. Congenital anomalies of the middle ear
5. External canal lesions: wax, atresia, foreign body and tumor (Figure 9.3)

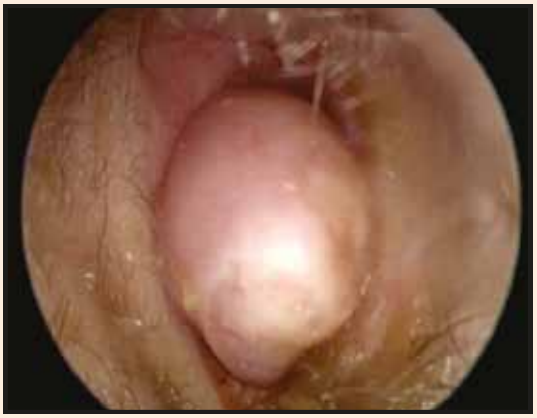


Figure 9.3: *Tumur (osteoma) blocking the ear canal and causing conductive hearing loss. (courtesy of Dr. Munahi Al-Qahtani)*

Q5. What are the common symptoms and signs of otosclerosis?

The most common characteristic and symptoms include the following:

1. Female gender, as women are generally more frequently affected than men
2. Slowly progressive hearing loss that is more commonly bilateral
3. Tinnitus
4. Paracusis Willisii
5. A positive family history in many patients

The most common signs include the following:

1. Normal ear examination
2. Schwartze's sign: increased vascularity of the promontory may be observed through the eardrum
3. Negative Rinne's test
4. Weber's test lateralizes to the affected side in unilateral cases
5. Type As tympanogram

Q6. What is paracusis Willisii?

It is defined as the ability to hear better in noisy surroundings.

Q7. What does a negative Rinne's test indicate?

It indicates that bone conduction is greater than air conduction using a 512 Hz tuning fork and indicates conductive hearing loss with an air–bone gap of at least 15 dB.

Q8. What diagnostic tests would help in the work-up of an otosclerosis patient?

The diagnosis of otosclerosis is based on the history, physical examination, audiological tests and CT scan to exclude other diagnoses. However, the definitive diagnosis is usually achieved during exploration of the middle ear.

Q9. What are the audiological findings in otosclerosis?

1. Air-bone gap greater in low frequencies
2. Carhart's notch
3. Good speech discrimination
4. Type As tympanogram

Q10. What is Carhart's notch?

It is an elevation of the bone threshold at the frequency of 2 kHz due to changes in middle ear mechanics. It is reversible by successful surgery.

Q11. What is the effect of pregnancy on this disease process?

Pregnancy adversely affects the disease process due to hormonal changes, although this phenomenon has not been consistently documented in the literature.

Q12. Once the diagnosis of otosclerosis has been established, what are the treatment options?

The treatment options can be divided as follows:

1. *Surgical*

Stapedectomy or Stapedotomy: These operations involve removing the stapes superstructure (head and crura), fenestration or partial removal of the foot plate with a microdrill or CO_2 laser and insertion of a piston-like prosthesis (Figure 9.4) between the incus and the oval window.

2. *Non-surgical*

a. *Observation*: This option can be chosen for patients with mild hearing loss who are not concerned about the hearing loss. In most cases, however, the hearing loss will progress and necessitate another modality of intervention.

b. *Amplification*: This is the second option if the patient does not want or if there are contraindications to surgery. Although it is non-invasive, the procedure has its own drawbacks, including cosmetic problems, cost and demanding maintenance.

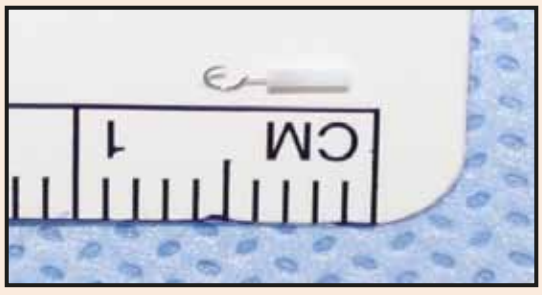


Figure 9.4: *Stapes prosthesis, around 0.5 cm in length.*

Q13. What are the potential complications of surgery?

1. Tympanic membrane perforation
2. Sensorineural hearing loss
3. Facial or chorda tympani nerves injury
4. Vertigo
5. Short-term or long-term failure

CHAPTER 10

Itchy Ear

Dr. Hassan Al-Shehri

A 55-year-old male presents to the emergency room with bilateral ear pain for 2 days. The pain is severe enough to prevent him from sleeping. There is an itching sensation and scanty purulent discharge. He is a swimmer, and he recalls that several days before the pain started he felt itching in both ears, which he tried to relieve by using cotton buds to clean his ears. He is diabetic and on insulin.

Examination shows that the skin lining of the external canal is swollen with redness and covered by scanty yellowish discharge. Otherwise, he has a clear head and neck examination.

* * * * *

Q1. What is the most likely diagnosis?

Acute otitis externa (AOE)

Q2. What are the factors that have predisposed the patient to this problem?

The risk factors are swimming and the habit of using cotton buds to clean the ears. Swimming is a predisposing factor because water causes alkalinization of the external canal, which is the optimum condition for bacteria to grow. Cleaning the ear using cotton buds (Figure 10.1) may cause repeated microtrauma to the skin of the auditory canal, predisposing it to infection.

Q3. What are the most common organisms that cause otitis externa?

– Bacteria (most common, ~90% of AOE): *Pseudomonas aeruginosa*, *P. vulgaris*, *E. coli*, and *S. aureus*

– Fungi (second most common): *Candida albicans* and *Aspergillus niger*

– Viruses: herpes zoster virus

Figure 10.1: *Acotton swabis the most common object people use to harm their ear.*

Q4. Do otitis externa patients complain of deafness?

Generally, otitis externa is not associated with hearing loss unless the external canal is completely obstructed.

Q5. What is the treatment for bacterial otitis externa?

Ear canal toilet is a very important step in the management of otitis externa. The best means to clean the ear is by suction, preferably under the operating microscope (Figure 10.2). Topical drops containing antibiotics and/or acidification drops (acetic acid) may be used. Insertion of an ear wick (Figure 10.3) is a useful measure to decrease edema and allow the medication to reach deeper into the ear canal. The patient may be given analgesia and advised to avoid getting water in the ears.

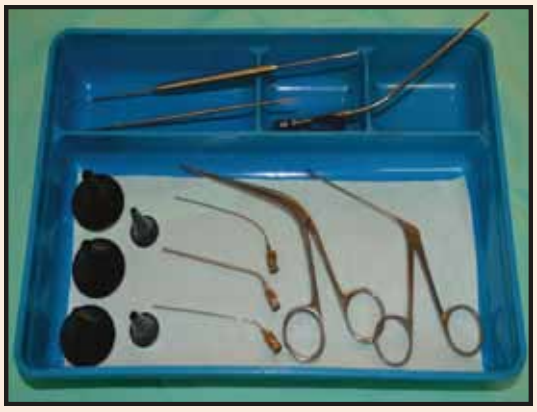


Figure 10.2: *These are the instruments used to clear the ear under a microscope.*

Figure 10.3: *An ear wick swells with antibiotic eardrops and fills the canal.*

Q6. What is the name of otitis externa that is caused by fungal infection?

Otomycosis.

Q7. How do you diagnose otomycosis?

The symptoms are similar to bacterial otitis externa. However, otoscopic examination usually demonstrates the presence of fungi (Figure 10.4). The diagnosis may be confirmed by examination of an ear swab in the microbiology lab.

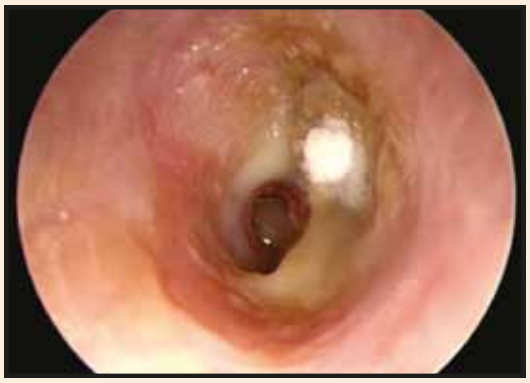


Figure 10.4: *Fungal infection of the ear canal. Notice the perforation of the eardrum and the middle ear secretion. (courtesy of Dr. Munahi Al-Qahtani)*

Q8. What is the treatment for otomycosis?

Frequent ear canal cleaning (e.g., by suction) and topical antifungal medications. Patients may be given analgesia and advised to avoid getting water into the ears.

Q9. What other conditions can cause otitis externa?

Allergies, a humid climate, swimming and frequent rubbing of the ears.

Q10. How does a patient with allergic otitis externa present?

Usually with chronic ear itching, mild discomfort, erythema of the canal skin and scaling.

Q11. What are the causes of allergic otitis externa?

Eczema and chronic irritants such as topical medications, soaps, swimming pool chloride and hearing aid mold.

Q12. What is the treatment for allergic otitis externa?

Discontinue the use of irritants and apply topical steroids.

Q13. If the infection is not controlled and the patient develops facial nerve paralysis on the affected side, what should you suspect?

Malignant otitis externa (osteomyelitis of the skull base).

Q14. Who is at highest risk of malignant otitis externa?

Elderly diabetic and immunocompromised patients.

Q15. How does a patient with malignant otitis externa present?

Classically, these patients present with the 4 Ds: discharge, discomfort, diabetes and dysfunctional cranial nerves.

The patient's general condition may deteriorate.

Granulation tissue may be present at the bony cartilaginous junction of the external canal (Figure 10.5).

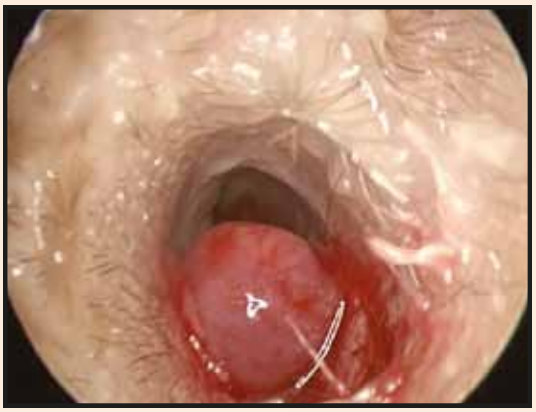


Figure 10.5: *Granulation tissue in the external ear canal in a diabetic patient should be taken seriously. (courtesy of Dr. Munahi Al-Qahtani)*

Q16. What is the most frequent causative organism of malignant otitis externa?

Pseudomonas aeruginosa causes 99% of cases.

Q17. How can malignant otitis externa be diagnosed?

- History and clinical examination
- Positive culture for *Pseudomonas aeruginosa*
- Imaging studies: CT scan, MRI, gallium and technetium scans

Q18. What is the treatment for malignant otitis externa?

- Antibiotics (antipseudomonals)
- Hyperbaric oxygen chamber therapy
- Diabetes control
- External canal debridement

CHAPTER 11

Night Crying

Dr. Rabea Mohammed Al Sabilah

An otherwise healthy 12-month-old boy has a cold that is accompanied by 2 days of rhinorrhea, cough and fever. On day 5, he becomes fussy and wakes up crying multiple times during the night. The following day he is febrile (temperatures up to 39°C). Examination of the ears shows bilateral red, bulging tympanic membranes. One day later, after visiting his family physician, his mother notices bilateral mucopurulent ear discharge, which is initially blood stained. Following the discharge, the child is less irritable, and his fever subsides.

* * * * *

Q1. The mother presents to your clinic and is concerned about the ear discharge. What is the most likely diagnosis?

Acute otitis media (AOM) (Figure 11.1).

Q2. You tell the mother that her child has AOM, and she asks you, "What is AOM?"

AOM is inflammation of the mucus membranes of the pneumatic spaces of the temporal bone, including the middle ear, mastoid air cells and Eustachian tube.

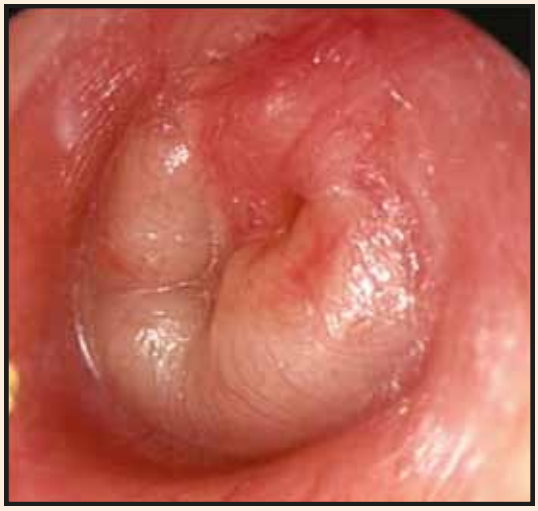


Figure 11.1: *AOM with bulging of the eardrum due to pus.*

Q3. The mother asks you, "Is my child's hearing going to be affected?"

Some degree of hearing loss may be present, but the majority of AOM cases usually resolve spontaneously or with treatment, and the short duration of the disease does not have a severe impact on the child's hearing.

Q4. Are there any tests that you need to do for the child?

Usually, no test is required because AOM is a clinical diagnosis. Culture of the discharge from the ear may be indicated in cases of chronic or recurrent discharge. Audiometry should be performed if chronic hearing loss is suspected, but not during the acute infection.

Q5. What are the risk factors for developing AOM? ABCDEF

A. Allergy

B. Bottle-feeding

C. Cigarette smoke exposure

D. Day care

E. Enlarged adenoids

F. Facial abnormalities

Q6. Name the most common pathogenic organisms.

1. *Streptococcus pneumonia* causes approximately 25-50% of cases.
2. *Haemophilus influenzae* causes approximately 25% of cases.
3. *Moraxella catarrhalis* causes approximately 12% of cases.

Viruses are responsible for the infection in approximately 40% of children with AOM. The most common viruses are respiratory syncytial virus (RSV), rhinovirus, adenovirus and influenza.

Q7. What is the etiology/pathophysiology of otitis media?

An URTI leads to congestion of the mucosa and then obstruction of the Eustachian tube. The viruses and/or bacteria that colonize the upper respiratory tract reach the middle ear by direct continuity and probably by insufflation as a result of the negative middle ear pressure. The microbial growth in the middle ear mucosa induces an acute inflammatory response, resulting in the formation of exudates and pus, which accumulate in the lumen and exert pressure on the tympanic membrane, causing intense pain and fever, lethargy and irritability. The tympanic membrane may then rupture, causing otorrhea but resulting in relief from the earache and fever. In most cases, the ear dries up, and the perforation heals spontaneously.

Q8. What is the epidemiology of AOM?

Acute otitis media is a disease of early childhood. The incidence of AOM peaks in children at 6-18 months of age. The AOM affects over 80% of all children before they reach the age of 3 years. Recurrent AOM (three or more episodes in

6 months or four or more episodes in 12 months) affects 20-30% of children.

Q9. What would you do if the previously mentioned child had symptoms of pronounced vomiting and seizures?

Meningitis should be excluded along with other intracranial complications of AOM: epidural abscess, brain abscess, otitic hydrocephalus and sigmoid sinus thrombophlebitis.

Q10. What would you do if the above child had symptoms of pronounced vomiting and deafness?

Labyrinthitis should be excluded along with other intratemporal complications of AOM: mastoiditis and facial nerve paralysis.

Q11. What antibiotic should you prescribe for the child, and what other medications can be given?

- Amoxicillin remains the drug of choice, and a dose of 80 mg/kg/day is recommended for 7 days.
- Azithromycin is an alternative antibiotic if the child has a documented allergy to penicillin (treat with standard doses for 3 days).
- Amoxicillin/clavulanic acid is not necessary in the initial treatment of AOM. It is a suitable second-line treatment if the initial treatment is unsuccessful.
- Pain relief should be offered. Acetaminophen and ibuprofen are the recommended analgesics.

Q12. What are the indications for surgical intervention?

Simple myringotomy is indicated if the patient has red and bulging tympanic membranes to relieve the pain and produce a clean incision that is more likely to heal than a spontaneous perforation. Insertion of ventilation tube "grommets" (Figure 11.2) is indicated in the following circumstances:

1. The presence of any complication
2. Recurrent infection: three episodes of AOM in 6 months or four episodes in a year, especially if associated with an effusion
3. Persistence of the infection
4. Deafness for more than 3 months

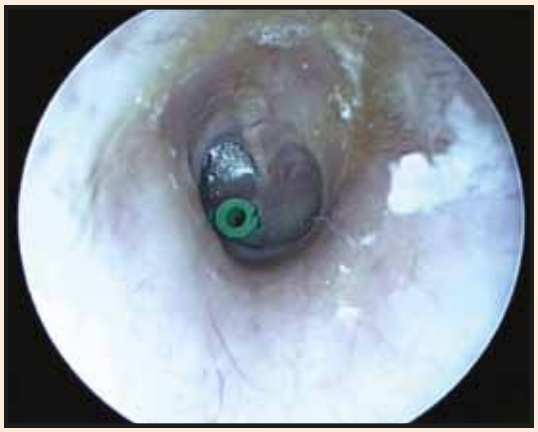


Figure 11.2: *Pressure-equalising tube (grommet) in the eardrum. (courtesy of Dr. Munahi Al-Qahtani)*

CHAPTER 12

Poor Attention

Dr. Saleh S. Alamry

A mother brings her 2-year-old daughter with Down syndrome with a pacifier in her mouth to the clinic, and she reports a history of poor attention over the prior 4 weeks. The inattention was noted after an URTI for several days, which resolved spontaneously without the use of medication. The daughter has no history of ear discharge or otalgia. The child attends day care for 6 hours per day. She has previously experienced recurrent otitis media, which were frequently treated with antibiotics. The mother reports that the daughter exhibits poor attention during ear infections, but this time, the child does not have a fever or an earache. The mother says that the biological father is contributing to the recurrent ear infections as a result of the cheap day care he provides for the daughter rather than the smoking of the stepfather, which she thinks is ridiculous as a suggested cause. Upon examination, the patient is malnourished and underweight and has poor hygiene. She is very scared of any examination and cries persistently.

* * * * *

Q1. What is the most likely diagnosis?

Child neglect should be seriously considered in this case. However, this is a classic case of otitis media with effusion (OME) with no symptoms except hearing loss presenting as poor attention, which might be followed by poor speech in children. Otitis media commonly occurs following URTIs in children attending day care. However, other causes of poor attention and hearing loss should not be overlooked.

Q2. What is the key diagnostic tool for OME?

Conventional otoscopy is the standard procedure used for diagnosing OME. It is easy, fast and accurate, and the instrument is available in every clinic. In OME, the tympanic membrane is retracted, has an amber yellow or pinkish or bluish discoloration and may display an air-fluid level or air bubbles (Figure12.1) in the middle ear. Pneumatic otoscopy (Figure 12.2) is useful

in demonstrating the diminished movement of the tympanic membrane. Tympanometry may help the diagnosis if the examination is equivocal. An affected patient will show a type B tympanogram (Figure 12.3).

Q3. How common is OME?

Extremely common; approximately 60% of children experience OME by 6 years of age.

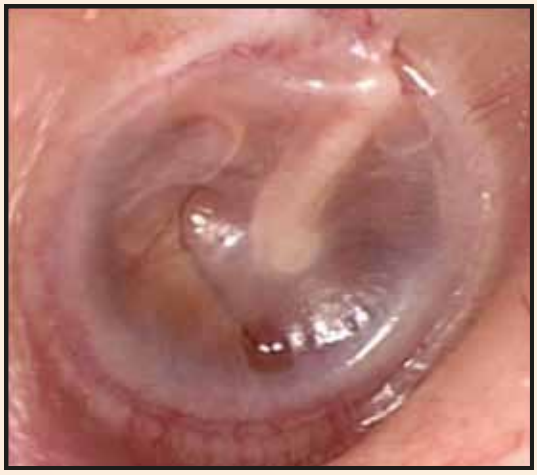


Figure 12.2: *Otitis media with effusion in the right ear. Notice the air-fluid line that resembles a hair medial to the eardrum.*

Figure 12.1: *Pneumatic otoscopy aids in diagnosing middle-ear effusion.*

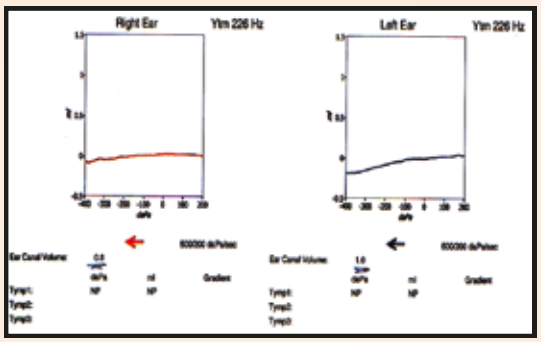


Figure 12.3: Tympanometry shows a type B pattern with a normal ear canal volume.

Q4. What is the etiological factor of OME?

- Eustachian tube dysfunction, e.g., secondary to nose diseases
- Increased viscosity of the middle-ear fluid, e.g., otitis media
- Adenoid hypertrophy, which functions as a reservoir for the bacteria that cause AOM

Q5. What are the other symptoms of OME?

It may be asymptomatic in children. Deafness is usually the only symptom. Other less common symptoms include ear fullness, tinnitus and a fluid sensation in the ears.

Q6. Why would unilateral OME be more serious than bilateral OME?

Nasopharyngeal malignancies can present as unilateral OME. Therefore, nasopharyngoscopy and neck examination for lymph node swellings is important. A CT scan or MRI can be performed for further evaluation, and a biopsy is sometimes needed.

Q7. Her mother is worried about the prognosis of her daughter's condition. What are you going to tell her?

In children, approximately 70% of OME cases resolve spontaneously within 3 months.

Q8. What are your goals for treatment?

The first goal of treatment is to prevent OME by controlling the underlying causes. The second goal is to educate the family about the illness because the hearing loss may fluctuate. The third goal is to restore hearing and prevent complications such as ossicular damage, tympanic membrane atelectasis or cholesteatoma.

Q9. How do you treat a patient with OME?

If there is no evidence of hearing loss, speech delays, developmental disability or tympanic membrane complications, observation can be continued until the effusion resolves spontaneously. If the condition has not resolved after observation, if there is considerable hearing loss or if complications are thought to be imminent or have occurred, the insertion of a tympanostomy tube is recommended (Figure 12.4).

Q10. What condition requires a tympanostomy for all patients?

Cleft palate. Nearly all children with cleft palate will develop OME because of Eustachian tube dysfunction.

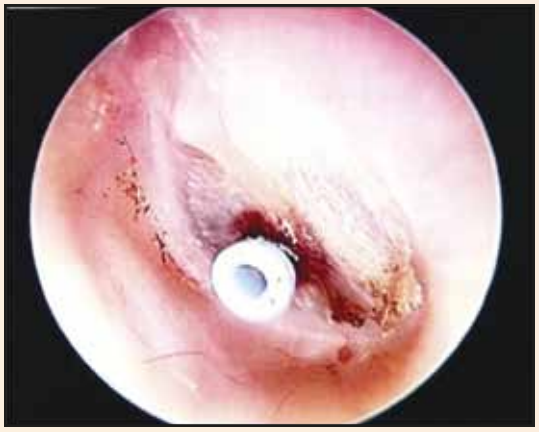


Figure 12.4: *Tympanostomy tube after 1 week.*

Q11. Why does OME have a higher incidence in Down syndrome patients?

Because of mid-face hypoplasia, Eustachian tube dysfunction and immune system immaturity.

Q12. The mother questions you, "Is it true that poor day care is the cause of my child's illness?"

This case has many underlying predisposing factors for OME, including age, Down syndrome, cigarette smoking, and day care.

CHAPTER 13

Foul Ear Smell

Dr. Khaled I. Al-Noury

A 29-year-old male is complaining of persistent left-ear discharge over the past several years. The discharge has been scanty, foul smelling and occasionally bloody. He has noticed a slight deafness in the affected side compared with the right ear. His history includes the insertion of ventilation tubes twice. He underwent these procedures at the ages of 6 and 8 years old.

The otoscopic examination (Figure 13.1) shows posterior perforation with white debris in the middle ear extending towards the attic and extending anteriorly to and under the handle of the malleus. The remaining area of the tympanic membrane is thin. Weber's test lateralizes to the affected ear. Rinne's test is negative in the affected ear and positive in the other ear.

* * * * *

Figure 13.1: The otoscopic examination. (courtesy of Dr. Munahi Al-Qahtani)

Q1. What are the important questions to ask to complete the history?

Asking about a history of vertigo, tinnitus, facial nerve weakness, headache and pain.

Q2. What is the most likely diagnosis?

The atticoantral type of chronic suppurative otitis media (CSOM).

Q3. What are the appropriate tests?

Pure tone audiogram for hearing assessment.

Ear swab for culture and sensitivity.

A CT scan of the temporal bone to evaluate the pneumatization of the mastoid and the extent of disease, particularly in relation to the facial nerve, lateral semicircular canal and middle and posterior cranial fossae.

Q4. What is cholesteatoma?

Cholesteatoma is an epidermoid inclusion cyst of the middle ear cleft, which includes the tympanic cavity, the Eustachian tube and the mastoid air cell system.

Q5. What are the types of cholesteatoma?

There are two types of cholesteatoma:

1. Congenital
2. Acquired
   - A. Primary acquired cholesteatoma arises from the pars flaccida retraction pocket.
   - B. Secondary acquired cholesteatoma arises from a marginal perforation.

Q6. What are the differences in the tympanic membrane perforations in the safe and unsafe ears?

Generally, in the safe ear, the perforation is central (Figure 13.2), while in the unsafe ear, the perforation is marginal, and the skin can migrate into the middle ear (Figure 13.3).

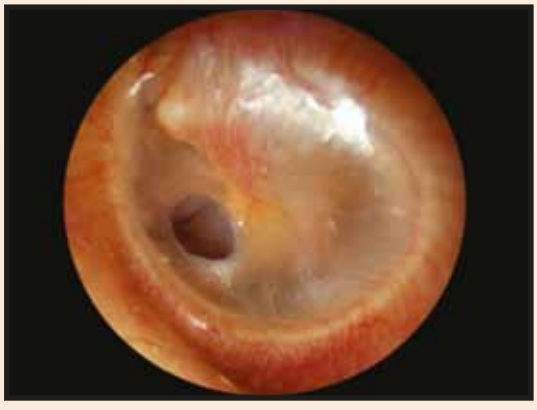


Figure 13.2: *Eardrum with a central perforation (considered a safe ear).*

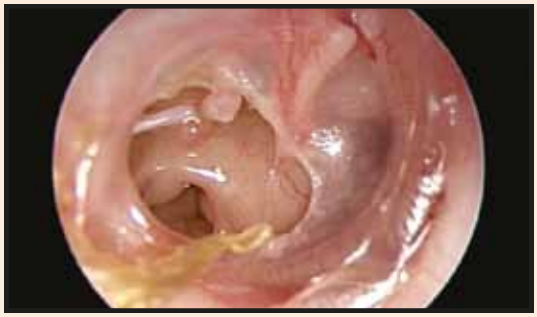


Figure 13.3: *Eardrum with a marginal perforation (considered an unsafe ear). (courtesy of Dr. Munahi Al-Qahtani)*

Q7. What are the complications of cholesteatoma?

1. Extracranial complications: otitis externa
2. Cranial complications: mastoid abscess, labyrinthine fistula, labyrinthitis and facial nerve paralysis
3. Intracranial complications: meningitis, lateral sinus thrombophlebitis, extradural abscess, subdural empyema, brain abscess and otitic hydrocephalus

Q8. What is the treatment for cholesteatoma?

The treatment is surgical removal of the cholesteatoma through a mastoidectomy.

The goals of surgery are as follows:

1. Provide safety by preventing the development of complications.
2. Maintain a dry ear.
3. Prepare the ear for possible reconstruction of the sound conduction mechanism.

Reconstruction should only be considered after controlling the disease and its complications.

Counseling the patient before surgery is crucial because the disease may not be symptomatic, and hearing may worsen after surgery.

Q9. What are the surgical approaches?

Many surgical approaches and combinations have been adopted for controlling this disease. The most well-known surgeries include atticotomy, canal wall-up mastoidectomy and canal wall-down mastoidectomy (modified radical and radical mastoidectomy) (Figure 13.4). The decision depends on the extent of the disease, the existence of complications, mastoid pneumatization, Eustachian tube function, hearing status, the reliability of the patient for follow-up and the experience of the surgeon. Hearing-restoration procedures such as tympanoplasty and ossiculoplasty can be performed simultaneously or during a second surgery. The patient needs regular follow-up exafter the surgery to care for the created mastoid cavity (if any) and to watch for any recurrence or residual disease (Figure 13.5).

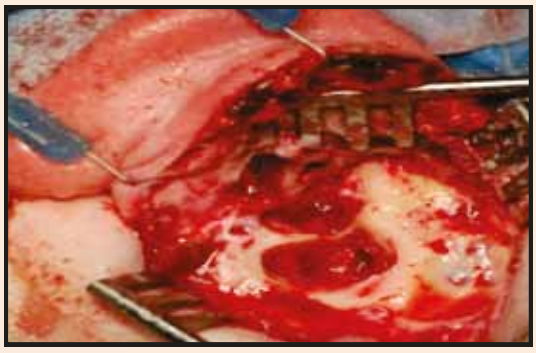


Figure 13.4: *Mastoidectomy in which the pinna was dissected and retracted anteriorly. The mastoid cavity was drilled posterior to the external ear canal. If the wall between the mastoid and ear canal is drilled, the mastoid and ear canal become one cavity. This procedure is called a canal wall-down mastoidectomy. If the wall is left intact, this procedure is called a canal wall-up mastoidectomy. (Photographs courtesy of the personal collection of Dr. David P. Morris FRCS (ORLHNS) MD)*

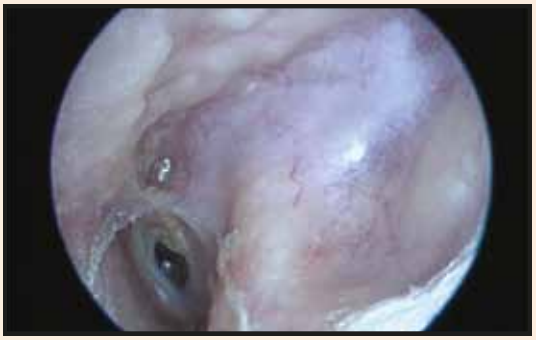


Figure 13.5: *Endoscopic picture of the ear following canal wall down mastoidectomy. Note the intact eardrum. (courtesy of Dr. Munahi Al-Qahtani)*

Q10. If the patient has persistent ear discharge when wearing a hearing aid, what other option could help his hearing loss?

The patient could undergo reconstructive surgery, such as tympanoplasty and ossiculoplasty. A BAHA device might help him because it conducts the sound to the inner ear through the bone without the need to insert anything into the ear canal that might aggravate the ear infection (Figure 13.6).

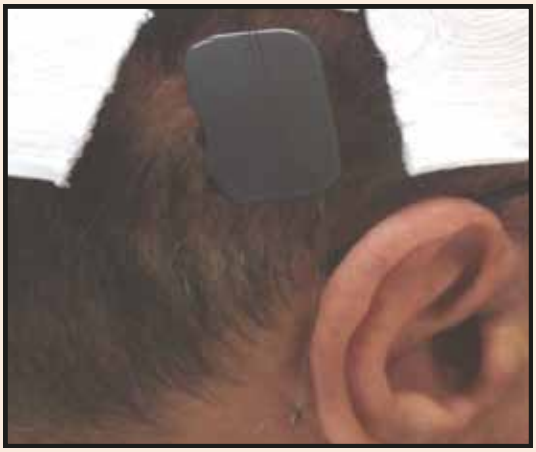


Figure 13.6: *Baha behind the ear with a cut in an Arabic hat.*

CHAPTER 14

I Woke From Sleep With Facial Asymmetry

Dr. Munahi Al-Qahtani

The emergency room doctor calls you regarding a 32-year-old male who presented to the accident and emergency room with right facial weakness for one day. He woke up from sleeping the day before with an asymmetrical smile and weak closure of his right eye. His facial weakness progressed rapidly to an obviously disfiguring facial asymmetry, and he was unable to close his right eye. He is also complaining of epiphora, dysgeusia and hyperacusis. He experienced right-side otalgia one day before the onset of the facial weakness but no other ENT complaints or neurological symptoms. His past medical history is unremarkable.

Upon physical examination, his general examination and vital signs are normal. He has a complete right-sided lower motor neuron facial palsy, but the other cranial nerves and the remaining neurological examination are normal. The ENT and head and neck examinations are normal.

* * * * *

Q1. What are the definitions of epiphora, dysgeusia, hyperacusis and otalgia?

Epiphora is watering of the eye or an overflow of tears onto the face.

Dysgeusia is an impairment or dysfunction of the sense of taste.

Hyperacusis is the abnormal perception of loudness or an over-sensitivity to certain frequencies of sound.

Otalgia is ear pain or earache.

Q2. What types of fibers comprise the facial nerve?

- Motor fibers to the stapedius, stylohyoid, digastric and facial muscles
- Secretomotor (parasympathetic) fibers to the lacrimal gland, nasal and palate glands and the submandibular and sublingual salivary glands
- Taste fibers from the anterior two-thirds of the tongue and the palate
- Somatosensory fibers from a small area in the auricle and external ear canal

Q3. What are the segments of CN VII?

The facial nerve (CN VII) has the following segments (Figure 14.1):

1. Supranuclear: from the cerebral cortex to the nuclei in the brainstem
2. Brainstem: contains various nuclei (motor, superior salivary and tractus solitarius)
3. Intracranial segment: in the cerebellopontine angle
4. Meatal segment: in the internal auditory canal (IAC)
5. Labyrinthine segment: from the fundus of the IAC to the geniculate ganglion
6. Tympanic segment: from the geniculate ganglion to the lateral semicircular canal
7. Mastoid segment: from the lateral semicircular canal to the stylomastoid foramen
8. Extratemporal segment: within the parotid gland where the nerve is divided into the temporal, zygomatic, buccal, mandibular and cervical branches (Figure 14.2)

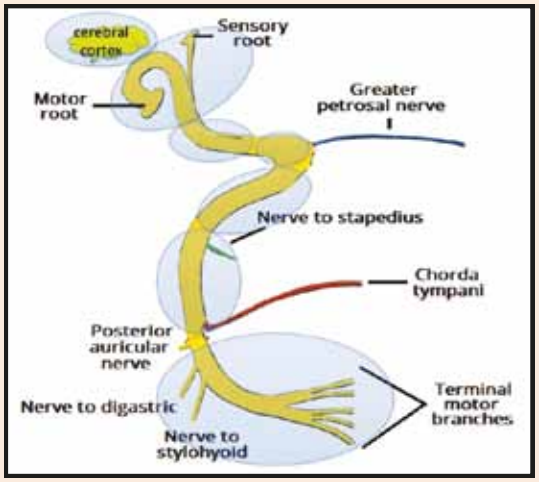


Figure 14.1: *Seven segments of facial nerve*

Figure 14.2: *Extratemporal segments of the facial nerve.*

Q4. What is the most likely diagnosis in this patient?

Bell's palsy (idiopathic acute lower motor neuron palsy) (Figure 14.3).

Figure 14.3: *The patient has Bell's palsy on his left side. Note the absence of forehead wrinkles, incomplete closure of the eye, and deviation of the angle of mouth.*

Q5. If there were vesicles over the right ear, persistent ear discharge or parotid swelling, would these factors change your diagnosis?

Yes, although Bell's palsy is the most

common cause of CN VII paralysis, the diagnosis can only be determined by excluding other causes, including the following:

- Ramsay Hunt syndrome due to varicella (herpes zoster) virus, which presents with vesicles that are observed on the ipsilateral ear (Figure 14.4), on the hard palate or on the anterior two-thirds of the tongue. The patient may present with vertigo and sensorineural hearing loss due to the involvement of CN VIII.
- Persistent ear discharge may be a symptom of acute or chronic suppurative otitis media (CSOM). Facial nerve paralysis can be a complication of any ear inflammation, but it is most commonly due to cholesteatoma. The ear must be examined under a microscope to look for signs of CSOM.
- The facial nerve and its terminal five branches divide the parotid gland into two lobes: superficial and deep. Parotid tumors that impede the neural function of the facial nerve tend to be malignant tumors.

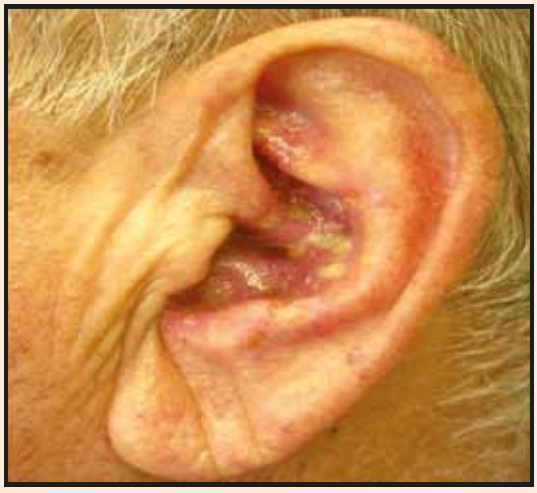


Figure 14.4: *Vesicles in the ear with facial nerve paralysis indicate Ramsay Hunt syndrome. (Photographs courtesy of personal collection of Dr. David P. Morris FRCS (ORLHNS) MD)*

Q6. How can you differentiate between upper and lower motor neuron facial nerve paralysis by physical examination?

Upper motor neuron (UMN) palsy of the facial nerve spares the forehead muscles. Asymmetry and weakness of forehead wrinkling points toward a lower motor neuron (LMN) palsy.

Q7. If there were a history of head trauma immediately preceding this patient's symptoms, what would be the most likely diagnosis, and what would be the test of choice?

The most likely diagnosis would be temporal bone fracture. Fractures of the temporal bone are classified as either longitudinal or transverse. Longitudinal fractures (70%) occur parallel to the long axis of the petrous bone, whereas transverse fractures (30%) occur perpendicular to it (Figure 14.5). Only 20% of longitudinal fractures result in facial paralysis compared with 50% of transverse fractures, which also have a higher incidence of profound sensorineuronal hearing loss and loss of vestibular function due to violation of the otic capsule.

Radiological evaluation should include a high-resolution CT scan of the temporal bone (Figure 14.5).

Q8. How can you describe the degree of facial nerve paralysis?

The most common system used for describing the degree of paralysis is the House-Brackmann scale.

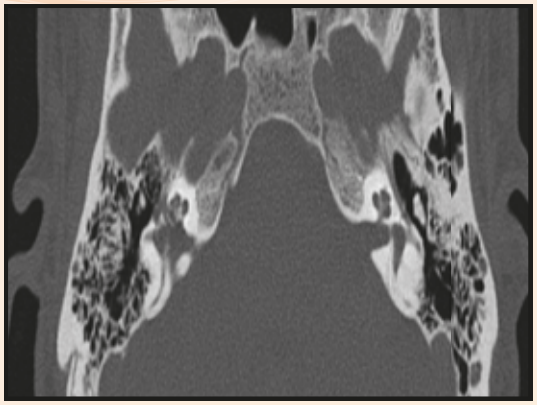


Figure 14.5: *Axial CT scan of the temporal bone showing a transverse fracture.*

House-Brackmann Scale (facial nerve palsy)

The House-Brackmann scale ranges between I (normal) and VI (no movement).

- *Grade I:* Normal symmetrical function
- *Grade II:* Slight weakness noticeable only upon close inspection; complete eye closure with minimal effort; slight asymmetry of the smile with maximal effort; synkinesis barely noticeable; contracture or spasm absent.
- *Grade III:* Obvious weakness, but not disfiguring; may be unable to lift eyebrow; complete eye closure and strong but asymmetrical mouth movement; obvious but not disfiguring synkinesis, mass movement or spasm.
- *Grade IV:* Obvious disfiguring weakness; inability to lift brow; incomplete eye closure and asymmetry of the mouth with maximal effort; severe synkinesis, mass movement or spasm.
- *Grade V:* Motion barely perceptible; incomplete eye closure; slight movement at the corner of the mouth; usually absent.
- *Grade VI:* No movement; loss of tone; no synkinesis, contracture or spasm.

Q9. How can you manage a patient with Bell's palsy?

1. Reassurance: The majority of cases resolve spontaneously.
2. Eye care: Dryness of the eye may cause irreversible corneal damage and blindness. Consider an ophthalmology referral.
3. Systemic steroids can be administered unless there are contraindications.
4. An ENT referral should be sought to exclude ear pathology (Figure 14.6).
5. A neurology consultation should be sought if the diagnosis is in doubt, the palsy is bilateral, there is a recurrence or there is no improvement after 1 month.

Remember that Bell's palsy is the most common cause of facial nerve paralysis. However, it is a diagnosis of exclusion after eliminating other causes.

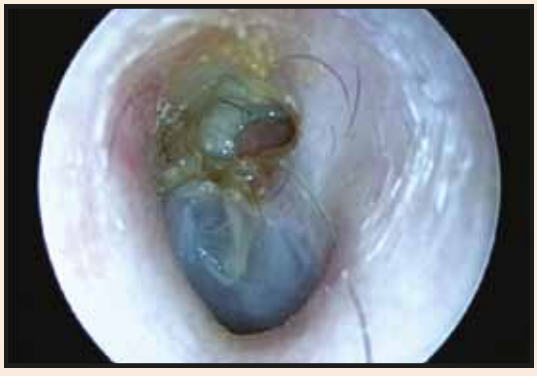


Figure 14.6: *Left ear with attic perforation, granulation tissue and purulent discharge (cholesteatoma). (courtesy of Dr. Munahi Al-Qahtani)*

CHAPTER 15

Ear Swelling

Dr. Abdulsalam Al-Qahtani

A 32-year-old female complains of fever, right ear pain and swelling of the post-auricular area for 2 days followed by right-sided facial weakness. The patient mentions that she has had many episodes of right-sided otorrhea in the past 5 years that used to resolve following treatment with antibiotics drops. However, the most recent attack of otorrhea has persisted for the last 2 months.

Examination shows right-sided pulsating mucopurulent ear discharge, soft swelling of the posterior wall of the external auditory canal and an eardrum perforation with pearly whitish debris in the middle ear. There is redness and swelling over the right mastoid area and pre-auricular area with protrusion of the right auricle. Palpation shows fluctuation of the post-auricular area with extension into the mastoid tip and along the upper posterior part of the sternocleidomastoid muscle. The Rinne test was positive in the left ear and negative in the right ear. Weber's test lateralizes to the right ear. The fistula test is negative. A facial examination showed an inability to close the right eye and an asymmetrical smile.

* * * * *

Q1. What is the most likely diagnosis? Is the facial nerve paralysis an UMN or a LMN paralysis?

The most likely diagnosis is chronic suppurative otitis media (cholesteatoma) with complications (mastoiditis, Luc's and Bezold's abscesses and facial paralysis). The facial paralysis is a right-side LMN paralysis because it involves all of the muscles of the face, including the inability to close the eye.

Q2. What further tests would you request?

A full blood count; an ear swab for Gram stain, cultures and sensitivities; a CT scan with IV contrast (Figures 15.1, 15.2 and 15.3); a pure-tone audiogram (Figure 15.4); a tympanogram; ± MRI.

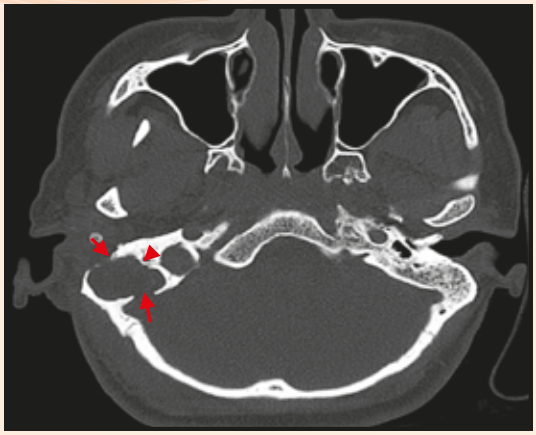


Figure 15.1: *A mastoid CT scan without IV contrast. An axial cut in a bone window shows the destruction of the right mastoid air cell septae (coalescent mastoiditis) in addition to the destruction of the posterior fossa plate and the posterior wall of the ear canal. The mastoid segment of the fallopian canal is not eroded.*

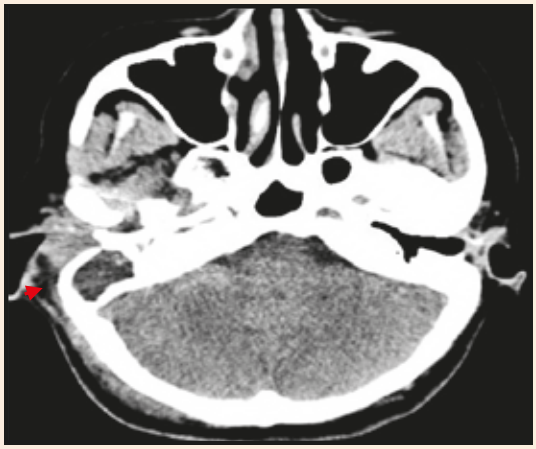


Figure 15.2: *A mastoid CT scan with IV contrast. An axial cut shows no evidence of an intracranial collection. Note the subcutaneous swelling (diffuse enhancement) in the right post-auricular area without obvious collection.*

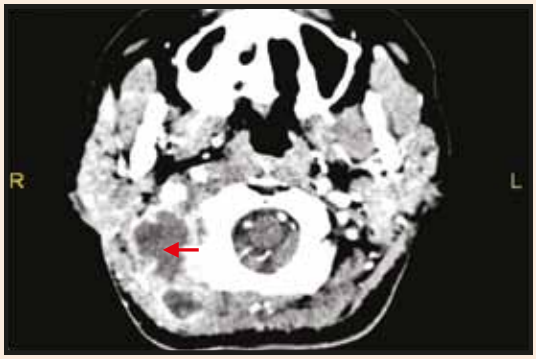


Figure 15.3: *A cervical CT scan with IV contrast. An axial cut in a soft tissue window shows two deep hypodense collections in the neck with marginal enhancement deep to the SCM muscle that favor Bezold's abscesses.*

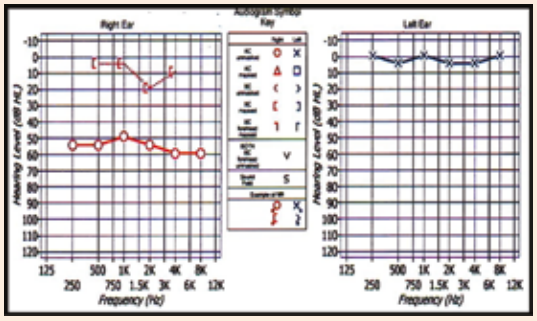


Figure 15.4: *A pure-tone audiogram shows a normal hearing in left ear. Unmasked right air conduction and masked bone conduction testing confirming the air-bone gap and conductive hearing loss.*

Q3. What are the common micro-organisms causing chronic otitis media?

Pseudomonas aeruginosa, Staphylococcus aureus, Proteus species, *Klebsiella pneumoniae*, diphtheroids and several anaerobes (*Bacteroides*, *Peptostreptococcus* and *Peptococcus* species).

Q4. What is the recommended management of this case?

Intravenous antibiotics and mastoidectomy with evacuation of the neck abscesses.

Q5. What are the complications of suppurative otitis media?

Temporal (Figure 15.5) and intracranial (Figure 15.6).

Mastoiditis (acute, chronic, coalescent and masked)
Petrous apicitis
Labyrinthine fistula
Facial nerve paralysis
Acute suppurative labyrinthitis

Figure 15.5: *Table showing temporal complications of suppurative otitis media*

Meningitis
Brain abscess
Subdural empyema
Epidural abscess
Lateral sinus thrombosis
Otitic hydrocephalus

Figure 15.6: *Table showing temporal complications of suppurative otitis media.*

Q6. What are the signs and symptoms that indicate intracranial complications of otitis media?

The following signs and symptoms are suggestive of intracranial complications:

- Fever associated with a chronic perforation
- Severe headaches
- Lethargy
- Altered mental status
- Focal neurologic signs (e.g., ataxia, oculomotor deficits and seizure)
- Papilledema
- Meningismus

The presentation of intracranial complications includes the following:

- Brain abscess – Fever, possibly seizures or focal neurologic signs, headache
- Meningitis – Fever, meningismus

Otitic hydrocephalus – Headache and signs of increased intracranial pressure

- Sigmoid sinus thrombosis – Spiking fever, edema and tenderness over the mastoid cortex, headache

Q7. What are the signs and symptoms that indicate temporal complications of otitis media?

- Fever associated with a chronic perforation
- Postauricular edema or erythema
- Facial nerve paresis or paralysis
- Retro-orbital pain on the side of the infected ear
- Vertigo
- Spontaneous nystagmus associated with sensorineural hearing loss

The presentation of the temporal complications includes the following:

- Labyrinthitis – Fever, nausea, vomiting, nystagmus and sensorineural hearing loss
- Mastoiditis with subperiosteal abscess – Fever, fluctuance overlying the mastoid area, protrusion of the pinna
- Petrositis – Retro-orbital pain, otorrhea, abducens nerve paralysis, fever

CHAPTER 16

Convulsion

Dr. Basher Abdullah

A 3-year-old boy is brought to the ER with a history of involuntary jerky movements that occurred 1 hour prior. The episode lasted approximately 2 minutes. For the past 3 days, the child has been complaining of a left-sided earache of abrupt onset associated with a high fever for which he was taking amoxicillin and acetaminophen. His earache subsided partially, but he later developed headache in addition to his high fever on the third day.

On physical examination, the child looks ill, drowsy and in pain. His temperature is 40°C. Left-ear examination shows a red bulging tympanic membrane. A CT of the temporal bones shows soft tissue opacity (with questionable fluid intensity) in the left middle ear. His CSF analysis is suggestive of bacterial meningitis.

The boy is started on ceftriaxone (100 mg/kg per day in two doses) and steroids. After 3 days, a hearing assessment (ABR & OAE) is conducted, and the results are normal. The child improves over a period of 2 weeks and is discharged home.

* * * * *

Q1. What are the characteristic features of the tympanic membrane in acute otitis media? (Figure 16.1)

- Erythema
- Bulging tympanic membrane
- Limited mobility
- Air-fluid level

Q2. The features of otitis media with effusion (OME) may resemble AOM. From the case scenario above, what features favor a diagnosis of AOM?

The acute onset in addition to the unilateral localization and the presence of pain and fever favor AOM.

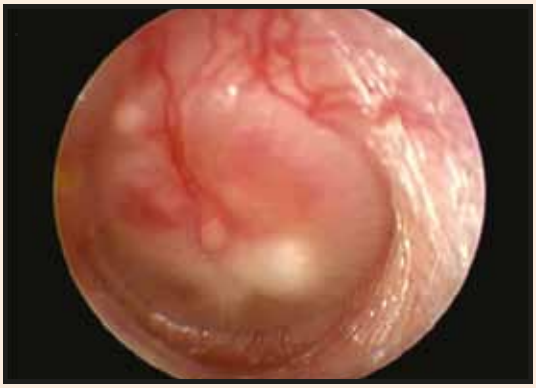


Figure 16.1: *Otoscopic findings in the boy's left ear. (Courtesy of Dr. Munahi Al-Qahtani)*

Q3. What are the three most common bacterial pathogens in AOM?

- *Streptococcus pneumoniae*
- *Haemophilusinfluenzae*
- *Moraxella catarrhalis*

Q4. What are the possible reasons for not responding to amoxicillin?

- Improper dosage
- Patient compliance
- Resistance of causative organism to amoxicillin

Q5. This boy had AOM complicated by meningitis. How does the infection spread from the middle ear to the meninges?

- Hematogenous route
- Preformed channels or fissures
- Direct extension through bony erosions
- Inner ear

Q6. In addition to meningitis, what other possible intracranial complications of AOM?

- Epidural abscess
- Subdural abscess
- Brain abscess
- Sigmoid sinus thrombosis
- Otitic hydrocephalus

Q7. In this particular case, what could be a possible cause for the boy's convulsions other than intracranial complications of AOM?

Febrile convulsion.

Q8. What important radiological test can be performed for the child?

A CT scan with contrast to the temporal bone and the brain.

Q9. Why is that particular radiologic test important in this case?

- To rule out other intracranial abscesses
- To rule out obstructive hydrocephalus
- To confirm that the ear is the underlying cause and determine whether there are any extracranial complications of the otitis media

Q10. Suppose the hearing test showed bilateral profound sensorineural hearing loss. What would you advise the child's parents?

Bilateral cochlear implants within 6 months.

CHAPTER 17

Hearing Loss In One Ear

Dr. Ibrahim M. Shami

A 43-year-old female complains of reduced hearing in her right ear that started 1 year earlier. Her hearing has gradually worsened over the previous 6 months; she has noticed that her ability to understand spoken words has diminished, particularly with telephone use. Additionally, she describes a ringing or sometimes a hissing sound in the same ear. The patient has also started to experience occasional episodes of imbalance over the prior 2 months.

Her history includes a sudden hearing loss that occurred 3 years previously. Her physician at that time diagnosed idiopathic sudden hearing loss and prescribed oral steroids for one week. She recovered within 4 weeks. The family history is non-contributory.

The general examination is normal. Neurologic examination reveals no nystagmus or impaired tandem gait. The cranial nerves, cerebellar, and ear exams are all normal. The Rinne test is positive in both ears and Weber's test lateralizes to the left ear.

* * * * *

Q1. How do you interpret a positive Rinne's test in both ears and a Weber's test that lateralizes to the left ear?

These findings suggest right-sided SNHL.

Q2. What is tinnitus and what are its main categories?

Tinnitus is the perception of sound without an external source.

Categories:

- Subjective tinnitus: the perception of sound in the absence of any acoustic, electrical or external stimulation
- Objective tinnitus: the perception of sound caused by an internal body sound or vibration

Causes of Subjective Tinnitus:

Diseases Hypertension Depression and anxiety Neurological disease (multiple sclerosis, brainstem stroke)
Medications Aminoglycosides Antihypertensive agents Aspirin / NSAIDs Others
Trauma Barotrauma Head injuries Loud noise
Hearing Loss and Otologic Disorders Hearing loss Ex: presbycusis Retrocochlear lesions (acoustic neuro mas) Ménière's disease

Causes of Objective Tinnitus:

Pulsatile	*Non-pulsatile*
Synchronous with pulse **Arterial aetiologies** Arteriovenous fistula Paraganglioma (Figure 17.1) (glomustympanicum or jugulare) Carotid artery stenosis Arterial dissection (carotid, vertebral) Persistent stapedial artery Dehiscence intratympanic carotid artery (Figure 7.2) Vascular compression of cranial nerve VIII Increased cardiac output (pregnancy, thyrotoxicosis) **Venous aetiologies** Pseudotumorcerebri Venous hum Jugular bulb anomalies **Asynchronous with pulse** Palatal myoclonus Tensor tympani or stapedius muscle Myoclonus	Patulous Eustachian tube Spontaneous otoacoustic emission

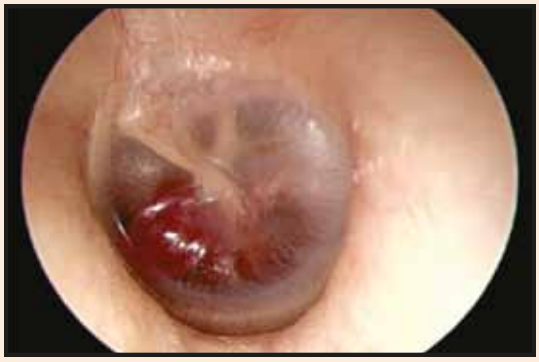


Figure 17.1: *A red lesion medial to an intact eardrum can be a vascular tumor and is commonly a glomus. (Courtesy of Dr. Munahi Al-Qahtani*

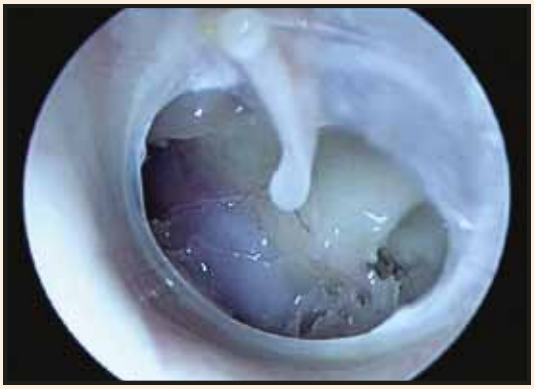


Figure 17.2: *Left eardrum perforation. Note the dehiscence of the carotid artery in the middle ear. (Courtesy of Dr. Munahi Al-Qahtani)*

Q3. This patient's pure-tone audiogram (Figure 17.3) shows unilateral SNHL, and speech audiometry shows a decreased speech discrimination score (SDS) suggesting a retrocochlear pathology. What is the most probable diagnosis for this patient?

Acoustic neuroma.

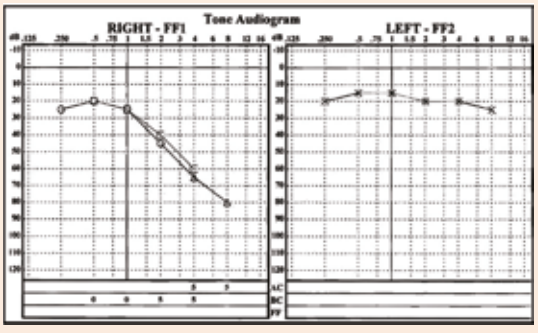


Figure 17.3: *PTA showing right-sided SNHL at high frequencies.*

Q4. What are the causes of unilateral / asymmetrical hearing loss?

Listed in table format:

External ear	Middle ear	Cochlear and retrocochlear (cranial nerve VIII and central pathway) (sensorineural)
Wax Foreign body Otitis externa Neoplasms: - Benign: exostoses and osteoma - Malignant: squamous cell carcinoma) - Congenital aural atresia	AOM Otitis media with effusion Chronic suppurative otitis media (mucosal or with cholesteatoma) Otosclerosis (may become bilateral, may have sensorineural component) Neoplasm: glomus tumor)	Trauma (physical, barotraumas, noise) Infection (labyrinthitis or meningitis) Neoplasm (vestibular schwannoma) Congenital Autoimmune Neurological (multiple sclerosis) Meniere's disease

Q5. What is acoustic neuroma (AN)?

- It is a benign schwannoma of CN VIII.
- *Origin:* Such tumors mostly develop from the Schwann cells of the vestibular nerve.
- Misnomer: Such tumors typically do not arise from the cochlear nerve and are not neuromas. Therefore, the name *vestibular schwannoma* is more accurate.
- The AN is the most common cerebellopontine angle (CPA) tumor. It represents approximately 85% of CPA tumors and 10% of intracranial tumors.

Q6. What additional tests can help to confirm the diagnosis, and what are the positive indicators of each test?

– *Routine audiometry* The tests are the initial screening tests. The *PTA* shows asymmetric SNHL hearing loss. The *speech audiometry* demonstrates a decreased SDS, and roll over (word recognition score gets worse with louder presentations). The *Stapedial reflex* shows absent or pathologic stapedial reflex decay.

– *Auditory brainstem response (ABR):* is the most sensitive and specific audiological test (10-15% false negative rate). A classical finding in ABR is an interaural latency difference for wave V of more than 0.2 ms. The test may be used to selectively investigate for asymmetrical hearing loss or vestibular weakness.

– *Vestibular testing: usually reveals* normal or ipsilateral caloric weakness.

– *MRI with gadolinium (Figure 17.4)*. It can detect tumors >2 mm that brightly enhance with gadolinium. It is the best imaging study and should be performed on any patient with asymmetrical inner ear symptoms (SNHL, tinnitus and/or vertigo).

– *CT with contrast:* indicated if MRI is unavailable or contraindicated. It suggests AN with asymmetric widening of the internal auditory canal. It may miss tumors smaller than 10 mm.

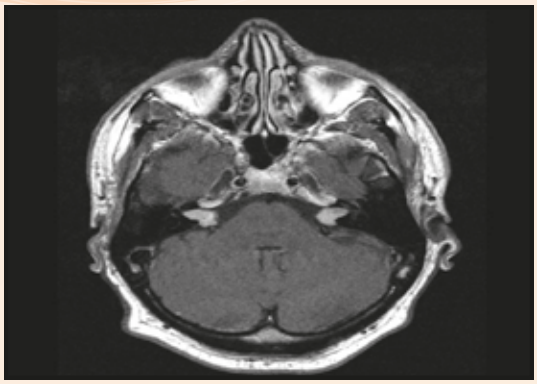


Figure 17.4: An MRI showing bilateral acoustic neuromas.

Q7. What are the symptoms of an AN?

- Hearing loss: progressive, asymmetric, high-tone SNHL
- Tinnitus
- Imbalance
- Facial weakness and numbness
- Advanced tumors may present with trigeminal nerve involvement, papilledema, occipital headaches, lower cranial nerve involvement or ataxia

Q8. How commonly does AN present with sudden SNHL?

- Approximately 10% of acoustic neuromas initially manifest with sudden SNHL.
- The prevalence of acoustic neuroma among patients with sudden SNHL is approximately 1%.

Q9. What is the natural history of AN?

- The growth rate of ANs is extremely variable.
- These tumors are generally slow growing, with an average growth rates of 0.2 cm per year. However, growth rates in excess of 2 cm per year have been observed.
- The ANs that are not treated are potentially lethal. Gradual enlargement can lead to indentation of the brainstem, increased intracranial pressure and death over the course of 5 to 15 years.

Q10. What are the other cerebellopontine angle lesions?

- Meningioma: the second most common tumor after acoustic neuroma
- Epidermoid cysts
- Non-acoustic neuroma
- Paraganglioma
- Arachnoid cyst
- Hemangioma

Q11. What are the treatment options?

Acoustic neuromas are managed using one or all of the following treatments:

- Observation
- Stereotactic radiation therapy
- Surgical excision