1. A nerve stimulator is placed at the stylomastoid foramen and used to measure the electrical threshold which will elicit gross facial movement. What test does this describe?
2. Electroneuronography
3. **Nerve Excitability Test**
4. Maximal Stimulation Test
5. Electromyography
6. Vestibular Evoked Myogenic Potential

**Difficulty Level: Hard**

**Explanation:** Thresholds for eliciting facial movement is compared from the healthy side to the affected side. A difference of >3.5 mA suggests there is weakness of one side compared to the other, and that the patient may possibly be a candidate for surgical decompression.

In a nerve excitability test for facial nerve function, the thresholds to elicit facial movement on the healthy and affect sides are compared, and a difference >3.5 mA is considered a significant weakness.

1. A 27 year old female presents to your office for evaluation of conductive hearing loss. She immigrated from a resource-limited country recently, and describes having chronic otologic problems for many years. Most recently, she tells you that she had a hole in her right ear drum and had a procedure to repair it. Fortunately, she brings with her an operative note from her prior surgeon. In the surgery, the surgeon describes a postauricular approach for the tympanoplasty, and noted that the incus and stapes were intact, but erosion of malleus was encountered. A fascial graft is placed in an underlay fashion onto the incus. What type of tympanoplasty is described?
2. Type I
3. **Type II**
4. Type III
5. Type IV
6. Type Va

**Difficulty Level: Hard**

**Explanation:** Wullstein created a classification scheme in 1956 identifying five basic types of tympanoplasty. Ossicular prostheses did not exist at that time, so the nomenclature can be quite confusing when his classification is memorized verbatim and not applied to this modern technology. Type II tympanoplasty repairs the TM and the ossicular chain with restoration of the lever mechanism by setting the graft onto the incus.

A type II tympanoplasty describes repair of the tympanic membrane and restoration of the lever mechanism of the ossicular chain.

1. A patient has absent acoustic reflex (AR) in the right ear with ipsilateral stimulation and an absent AR in the left ear with contralateral stimulation, but normal reflex in the left ear with ipsilateral stimulation and normal AR in right with contralateral stimulation.

Which of the following is the most likely site of the lesion?

1. Seventh nerve lesion proximal to the innervation of the stapedius.
2. **Cranial nerve (CN) VIII**
3. Stapes footplate.
4. Brainstem.
5. Lesion of the facial nerve distal to the stylomastoid foramen.

**Difficulty Level: Hard**

**Explanation:** The acoustic reflex arc involves CN VII and CN VIII. Intense sounds perceived by CN VIII are then relayed to the ipsilateral superior olive and facial motor nucleus. The superior olive complex then stimulates bilateral facial motor nuclei, resulting in contraction of the stapedius muscle. This reflex is involuntary, and believed to have a role in hearing protection.

The acoustic reflex arc involves a unilateral CN VIII and the motor function of bilateral CN VII.

1. A 6 year old male with a normal external ear is being assessed for candidacy for surgical correction of congenital aural atresia. A CT of temporal bone is obtained and reveals a well pneumatized middle ear space and mastoid, a normal oval and round window, a facial nerve, and a malleus/incus complex. There is no stapes or incus stapes-connection seen on CT. What is this patient’s score on the Jahrsdoerfer grading system and is he a candidate for surgery?
2. 5; he is not a candidate for surgery
3. 7; he is not a candidate for surgery
4. 5; he is a candidate for surgery
5. **7; he is a candidate for surgery**
6. 9; he is a candidate for surgery

**Difficulty Level: Hard**

**Explanation:** The Jahrsdoerfer grading system is used to assess candidacy for surgical correction of congenital aural atresia. The scale is based on temporal bone CT findings and assesses nine different parameters. The parameters are stapes, oval window, facial nerve position, status of the ossicles and round window, and pneumatization of the mastoid. The stapes is assigned two points and the other parameters are assigned one point each. Scores 6 through 10 range from marginal to excellent candidates for surgery, whereas a score of five or less usually points to a poor outcome. The patient in this case has a score of 7 with a well pneumatized middle ear space and mastoid, a normal oval and round window, a facial nerve, normal external ear, and a malleus/incus complex. Without a stapes, this patient does not have an incus-stapes connection and thus loses 3 points on the grading scale. He is still a candidate for an aural atresia repair with an ossiculoplasty according to the Jahrsdoerfer criteria. Of note, there have been additional suggested modifications to the Jahrsdoerfer criteria, such the height of the tegmen, volume of the middle ear space, and others to consider when evaluating a patient for atresiaplasty.

The parameters for the Jahrsdoerfer criteria are the presence of the stapes, oval window, facial nerve position, status of the ossicles and round window, and pneumatization of the mastoid. The stapes is assigned two points and the other parameters are assigned one point each.

1. The size of the auricle at age 5 years is what percent of an adult-sized auricle?
2. 30%
3. 50%
4. 65%
5. **80%**
6. 100%

**Difficulty Level: Moderate**

**Explanation:** It is an important point to consider that the reason microtia repair is delayed until at least age 5-6 is two-fold: first, the pinna is nearly (80%) the size of an adult pinna, and secondly, this is the age that children will attend school and may be subject to the psychologic effects of teasing. If using rib grafts for reconstruction, waiting until the child is older will also allow for growth of the ribs and chest.

The pinna at age 5 is approximately 80% of its eventual adult size.

1. A 35-year-old male presents to the clinic after an MRI brain ordered for a workup of seizure activity. There is an incidental discovery of a mass in the left petrous apex that is hyperintense relative to the brain in both T1 and T2 sequences. Which of the following is the most likely diagnosis?
2. Cholesteatoma
3. Meningioma
4. **Cholesterol granuloma**
5. Paraganglioma
6. Vestibular schwannoma

**Difficulty Level: Easy**

**Explanation:** It is helpful to have a general idea of the relative intensities of cerebellopontine angle and petrous apex lesions as this will help narrow the differential diagnosis.

Cholesterol granulomas and endolymphatic sac tumors are hyperintense on both T1 and T2 sequences.

1. The nervus intermedius (of Wrisberg) carries which of the following?
2. Special visceral efferent fibers, whose cell bodies are located in the geniculate ganglion, and which project to the nucleus tractus solitaries
3. General sensory afferents terminating in the spinal trigeminal tract
4. **Parasympathetic general visceral efferents to the salivary and lacrimal glands, as well as special visceral afferent fibers from the chorda and general sensory afferents from the external auditory canal and postauricular area**
5. Corticobulbar fibers originating in the somato- motor cortex

**Difficulty Level: Moderate**

**Explanation:** Within the internal auditory canal the nervus intermedius runs as a separate nerve between the facial and superior vestibular nerves. It carries secretory fibers for the lacrimal,gland as well as the sublingual and submandibular glands. It also carries afferent taste fibers from the anterior 2/3 of the tongue as well as sensation from the posterior wall of the external auditory canal. It then runs within CNVII within the temporal bone. The nervus intermedius typically merges with the facial nerve by the time it reaches the end of the internal auditory canal.

The nervus intermedius contains parasympathetic general visceral efferent and special visceral afferent fibers.

1. Which vestibular test is capable of testing each side of the vestibular system independently?
2. **Caloric testing**
3. Positional testing
4. Rotary chair
5. Posturography
6. Oculomotor testing

**Difficulty Level: Easy**

**Explanation:** Each ear is tested independently with both cold and warm water to illicit increasing or decreasing amounts of nystagmus. Because the horizontal semicircular canal is most lateral, this is what is being stimulated with caloric testing.

Caloric testing is the main test that is part of a traditional VNG that is able to test each ear independently.

1. A 38-year-old female presents to your office for evaluation of sudden left facial paralysis since last week. She reports having had an upper respiratory infection that started two days prior. She denies any prior otosurgical history, prior head trauma, medical problems, or any current otologic or audiologic symptomatology. Examination reveals a House-Brackmann IV left facial paralysis but an otherwise unremarkable examination. Which of the following is correct regarding this patient's likely diagnosis?
2. A brainstem MRI should be ordered immediately to rule out a central nervous system tumor.
3. **She has a high likelihood of returning to complete facial function.**
4. The tympanic segment of the facial nerve is the likely site of compression injury.
5. Electroneurography (ENoG) should be performed immediately and surgical decompression performed if 90% degeneration is evident.
6. Antiviral therapy alone improves the likelihood of a complete return of facial function in patients with this condition.

**Difficulty Level: Easy**

**Explanation:** Patients with Bell’s palsy demonstrating only a partial paresis upon presentation have an excellent prognosis, with most achieving a total return of facial function and only a tiny percentage having significant residual dysfunction. In general, the severity of the paresis at the nadir correlates with the severity of dysfunction and the risk of synkinesis after recovery. However, progression to complete paralysis may occur early in the clinical course. As a result, these patients should be monitored closely, and steroids (with or without antivirals) should be instituted to prevent worsening edema and injury to the facial nerve.

Patients with Bell's palsy and incomplete paralysis have an excellent prognosis, with most achieving complete recovery or only mild residual paresis. In general, the greater the severity of the paresis at the nadir, the worse the prognosis is for the return of function and subsequent development of synkinesis.

1. A 46 year old military veteran presents to your clinic complaining of ringing in his ears. He notes that it is present all of the time and sounds like a high pitched buzzing. He feels the right ear has worse ringing to the point it is keeping him up at night. Based on this patient'™s complaint, which of the following would be the LEAST useful in this patient'™s evaluation?
2. Audiogram
3. Medication evaluation
4. Comprehensive history and physical
5. Tinnitus handicap questionnaire 1 (TH1)
6. **Vestibular testing**

**Difficulty Level: Easy**

**Explanation:** This patient is complaining of tinnitus. There may be many causes of tinnitus, both objective and subjective.

When evaluating a patient for tinnitus, it is important to evaluate for reversible causes or warning signs/symptoms. A basic work up should include a comprehensive history and physical, including medication use, noise exposure history, and assessment of how much the tinnitus is impacting the patient's quality of life, and an audiogram.

1. A 67-year-old male with diabetes and hypertension presents an eight-hour history of severe vertigo with nausea and vomiting. He was previously asymptomatic and has not had prior episodes of vertigo. He denies hearing loss but notes that the room is persistently spinning, and his symptoms are exacerbated by head movement. On examination, he has a spontaneous horizontal nystagmus with a fast phase to the left, and on head-impulse testing, there is a catch-up saccade with a right head thrust but not a left head thrust. A test of skew is unremarkable. His ear and tuning-fork exams are normal, and he has no other focal neurological deficits. How should this patient be counseled?
2. Perform outpatient magnetic resonance imaging (MRI) to assess for a brain tumor.
3. Perform urgent MRI prior to initiating stroke treatment.
4. **Symptoms will likely abate in the next few days without intervention.**
5. Will likely benefit from a vestibular nerve section in the future
6. He has a viral infection of the inner ear and is at high risk for temporary facial paralysis.

**Difficulty Level: Moderate**

**Explanation:** This patient suffers from vestibular neuritis, an inflammatory disorder of the vestibular nerve likely caused by a viral etiology or small vessel occlusion. Many patients have a viral prodrome or infection preceding their vertigo. Treatment is supportive, and symptoms should abate over the next few days. For severe symptoms, some patients require IV hydration and anti-emetics. Antivirals, vestibular suppressants, and steroids have also been employed, but with inconclusive results.

Vestibular neuritis is characterized by acute onset of vertigo without hearing loss or other neurological symptoms and does not improve with staying still. The typical clinical course is to have symptoms for several days that then improve, though it is common for patients to develop issues with chronic imbalance or positional vertigo, thus highlighting the importance of exercise and the potential need for vestibular rehabilitation in this population to help develop compensatory mechanisms.

1. A 48-year-old female presents with weekly attacks of vertigo lasting 1-2 hours followed by 4-6 hours of slowly improving nausea and dysequilibrium. During these episodes she notes aural fullness and roaring tinnitus. Which of the following is one of the requirements for a definite diagnosis of Meniere's disease?
2. One spontaneous or evoked episode of rotational vertigo lasting more than twenty minutes
3. **Tinnitus and/or perception of aural fullness**
4. Positive sorbitol stress test for endolymphatic hydrops
5. Abnormal electronystagnography testing
6. Stable sensorineural hearing loss

**Difficulty Level: Hard**

**Explanation:** Meniere's disease is diagnosed based upon clinical symptoms, but the underlying pathologic lesion is endolymphatic hydrops, which results in distortion of the membranous portions of the labyrinth. It can only be diagnosed definitively on autopsy.

Definite Meniere's disease is diagnosed based on fulfillment of the following criteria: Fluctuating aural symptoms (tinnitus, aural fullness, hearing loss) in the affected ear, audiometric confirmation of low to mid frequency sensorineural hearing loss in the affected ear, and two spontaneous episodes of rotational vertigo lasting 20 minutes to 12 hours. Other causes of vertigo must be excluded.

1. Which of the following signs or symptoms distinguishes central versus peripheral vertigo?
2. The intensity is more severe in central
3. The vertigo has fatigability in central
4. There is associated nausea, hearing loss, and sweating in central
5. **Nystagmus is enhanced with ocular fixation in central**
6. Nystagmus is horizontal and/or rotary in central

**Difficulty Level: Easy**

Nystagmus is often enhanced (or at least not diminished) with ocular fixation in central vertigo whereas the nystagmus will be suppressed in peripheral causes of vertigo.

1. What malformation describes membraneous aplasia of the cochlea and saccule?
2. Alexander aplasia
3. Mondini aplasia
4. **Scheibe aplasia**
5. Michel aplasia
6. Enlarged vestibular aqueduct

**Difficulty Level: Moderate**

**Explanation:** The bony labyrinth and the superior portion of the membranous labyrinth, including the utricle and semicircular canals, are normally differentiated in patients with Scheibe aplasia. The organ of Corti is generally poorly differentiated with a deformed tectorial membrane and collapsed Reissner’s membrane, which compromises the scala media. Scheibe aplasia is the most common histopathologic finding in patients with congenital deafness.

Scheibe aplasia describes membranous aplasia of the cochlea and saccule (pars inferior).

1. A 32 year old is brought to your clinic for evaluation of hearing loss. Evaluation of the patient shows a flattened midface with an overall syndromic appearance. The patient indicates a long standing history of arthritis despite his young age as he shows you how he can bend his wrist and let the back of his palm touch his arm. As he talks you notice that his speech sounds off a little and he indicates he had a cleft palate repaired as a child. What is the most likely diagnosis?
2. **Stickler syndrome**
3. Van der Woude Syndrome
4. Treacher Collins syndrome
5. Loeys-Dietz syndrome
6. Non syndromic cleft palate

**Difficulty Level: Moderate**

**Explanation: T**his patient has hypermobile joints, hearing loss, cleft palate, and a flattened midface. These are all characteristics of a patient with Stickler syndrome which is a collagen disorder that is generally inherited in an autosomal dominant manner. Other symptoms would include myopia (near sightedness) and some patients may have the Pierre Robin sequence of cleft palate, large tongue, and retrognathia. Genes associated with this syndrome include COL11A1, COL11A2, COL9A1 and COL2A1.

Stickler syndrome is a genetic disorder typically resulting in micrognathia leading to Pierre-Robin sequence, joint hypermobility, early onset arthritis, myopia, retinal detachment, cataracts and sensorineural hearing loss. There are 4 types, and 3 of the 4 are autosomal dominant with the last being autosomal recessive. It is caused by mutations in COL2A1 (type 1), COL11A1 (type 2), COL11A2 (type III) and COL9A1, resulting in defective type II, IX and XI collagen. Type 3 is unique in that there are no ocular abnormalities.

1. A 28 year old pregnant female presents to the ENT clinic with 3 months of progressive bilateral hearing loss. The patient denies any recent infections and loud noise exposure. She does note that her mother had early hearing loss requiring hearing aids. On physical exam the tympanic membranes are intact and the middle ear spaces are well aerated. On the right there is a red blush seen on the promontory. An audiogram shows a bilateral moderate severe conductive hear loss with absent acoustic reflexes. What is the most likely diagnosis?
2. Tympanic paraganglioma
3. **Otosclerosis**
4. Osteogenesis imperfect
5. Paget's disease
6. Congenital fixation of the stapes

**Difficulty Level: Easy**

**Explanation:** Otosclerosis is a disease that is unique to the otic capsule bone that can cause a conductive hearing loss, mixed hearing loss, or (rarely) a purely sensorineural hearing loss. With conductive hearing losses in otosclerosis, there is an artificial drop in the bone line at 2k Hz (the Carhart notch). It is an autosomal dominant disease. Patients with otosclerosis present with progressive hearing loss and for an unknown reason the disease is accelerated during pregnancy. The physical exam finding of a red hue can be seen at the promontory (Schwartze sign), though this is seen in only a minority of patients.

Otoslcerosis often presents with a conductive or mixed hearing loss with a drop in the bone line around 2k Hz, and there is often a positive family history of early hearing loss. Schwartze sign may be seen in a minority of patients.

1. All of the following tests have the potential to be abnormal in a patient with isolated Meniere’s disease EXCEPT:
2. Audiogram
3. **Tympanogram**
4. Electrocochleography
5. Vestibular Evoked Myogenic Potential
6. Glycerol Dehydration Test

**Difficulty Level: Easy**

**Explanation:** Meniere’s Disease manifests as a sensorineural hearing loss, not a conductive loss, and the tympanogram is expected to be normal.

ECochG, VEMP and audiometric testing is commonly abnormal in patients with Meniere's disease, however tympanometry should be normal.

1. A 25 year old male presents to the emergency room after a motor vehicle collision. A CT scan of the head is concerning for a left temporal bone fracture. Physical exam demonstrates a House-Brackmann VI facial nerve paralysis on the left side. Where is the most likely site of facial nerve injury?
2. Internal auditory canal
3. Meatal segment
4. **Perigeniculate region**
5. Second genu
6. Mastoid segment

**Explanation:** The site of injury of the facial nerve in temporal bone fractures is in the perigeniculate region in 80-93% of patients. If surgical decompression is warranted, this area can be reached by a translabyrinthine or transmastoid/supralabyrinthine approach. The other areas are all possible locations of injury, with mastoid segment being the second most common; but perigeniciluate region is by far the most common. This includes the labyrinthine portion of the fallopian canal, which is the shortest and most narrow portion.

1. A 57 year old male with a long standing history of Meniere'™s disease presents for further treatment. He has been on a low salt diet since diagnosis. He takes daily hydrochlorothiazide. He has severe to profound hearing loss in the affected ear. He is complaining that he continues to have weekly disabling symptoms and is looking for further treatment. He undergoes vestibular testing showing normal vestibular function in the contralateral ear and 70% weakness in the affected ear. All of the following are reasonable treatment options EXCEPT?
2. Labyrinthectomy
3. **Transtympanic neomycin injection**
4. Transtympanic gentamicin injection
5. Intratympanic dexamethasone injection
6. Endolymphatic sac shunting

**Difficulty Level: Moderate**

**Explanation:** This patient has persistent symptoms despite previous medical therapy and has borderline to non-serviceable hearing. Therefore, his choice of vestibulotoxic procedure is not limited by the need for hearing preservation. Aminoglycosides are both vestibular and ototoxic. The key to knowing the correct answer here is knowing which is most selective for the vestibular system. Gentamicin is a vestibulotoxic aminoglycoside, which is why it is used in a transtympanic injection to selectively destroy the vestibular system. It does have cochleotoxic properties and can result in significant hearing loss, but it primarily exerts its effects on the vestibular system. Neomycin is primarily ototoxic and would not be a good obtion for ablative therapy.

Prior to considering any ablative procedure of the vestibular system vestibular testing must be performed. This not only confirms the vestibular weakness is in the affected ear but also verifies the contralateral system is present. Lack of all vestibular function can lead to very severe disequilibrium which is why the presence of the contralateral system must be confirmed.

Neomycin is a primarily ototoxic aminoglycoside, and is not used in the treatment of Meniere's disease.

1. A 59 year old male undergoes MRI of the brain due to concern for a cerebellopontine angle mass. Using the adjacent gray matter as a reference, which of the following correctly describes the appearance of a vestibular schwannoma on MRI?
2. **Isointense or hypointense on T1, slightly hyperintense on T2, strongly enhances with contrast**
3. Isointense on T1, variable intensity with T2, mild to moderate enhancement with contrast
4. Hypointense on T1, hyperintense on T2, nonenhancing, no restriction on DWI
5. Hypointense on T1, hyperintense on T2, nonenhancing, DWI showing restriction.
6. Hyperintense on T1, hypointense on T2, hypointense on T1 with fat suppression

**Difficulty Level: Moderate**

**Explanation:** Vestibular schwannomas are by far the most common cerebellopontine angle tumor in adults. On MRI they typically are found to be iso- or hypointense on T1, are slightly heterogeneously hyperintense on T2 and have marked enhancement with gadolinium contrast. They do not have dural tails.

Vestibular schwannomas have an iso or hypointense appearance on T1, slightly hyperintense appearance on T2 and marked enhancement with contrast.

1. A patient presents with a conductive hearing loss at age 35. He has a notch in the bone hearing loss at 2,000 Hz. His father had a similar onset of hearing loss which improved with “some stapes surgery.” The patient is undergoing a laser stapedotomy but once the tympanic membrane is elevated, a blood vessel is seen coursing through the stapes foot plate. What is this structure derived from?
2. 1st branchial arch
3. **2nd branchial arch**
4. 1st branchial groove
5. 2nd branchial groove
6. 1st branchial pouch

**Difficulty Level: Easy**

**Explanation:** This patient has a persistent stapedial artery. In normal development, this structure involutes. Persistence is rare and can lead to the middle meningeal artery arising from it which can lead to hypoplasia of the foramen spinosum in the skull base. Most patients are asymptomatic but it may present with pulsatile tinnitus and hearing loss. Persistence of this artery may necessitate abortion of the surgery. It is a derivative of the second branchial arch.

The branchial structures include the groove/cleft, arch, and pouch. The groove/cleft is an ectoderm derivative, the arch is a mesoderm derivative, and the pouch is an endoderm derivative. The first branchial groove forms the external auditory canal, and the rest of the grooves are eventually obliterated during development. The first branchial arch includes the maxillary artery, muscles of mastication, tensor veli palatini, tensor tympani, mylohyoid, and anterior belly of the digastric. In the ear, it forms the body and short process of the incus and head and neck of the malleus (the parts in the epitympanum). The second arch includes the stapedial artery, muscles of facial expression, posterior belly of the digastric, stylohyoid muscle, and stapedius muscle. In the ear, it includes the stapes superstructure (the footplate is derived from the otic capsule). The 3rd arch forms the common and proximal internal carotids, stylopharyngeus muscle, and the greater horn of the hyoid and lower half of the body of the hyoid. The 4th arch forms the proximal right subclavian and arch of the aorta, muscles of the soft palate except for the tensor veli palatini, muscles of the pharynx except for the stylopharyngeus as well as the cricothyroid and cricopharyngeus, and helps to form the laryngeal cartilage in conjunction with the 6th arch. There are no 5th arch structures. The 6th arch forms the ductus arteriousus and proximal pulmonary arteries, intrinsic muscles of the larynx and skeletal muscle of the esophagus, and also contributes to the laryngeal cartilage formation.

The stapedial artery is derived from the 2nd branchial arch. It normally involutes during development.

1. Which of the following situations exceed the noise exposure limits set by the Occupational Health and Safety Administration (OHSA)?
2. **9 hours in a factory with a noise level at 90 dB**
3. 30 minutes of jack-hammering at a noise level of 110 dB
4. 4 hours of using a band saw at 95 dB
5. 2 hours of operating a forklift at with a noise level of 100 dB

**Difficulty Level: Moderate**

**Explanation:** The OHSA sets limits on the permissible amounts of noise exposure in the workplace to help prevent noise-induced hearing loss. The maximum allowable daily noise exposure limit has been specified as an average of 90 dB over an 8 hour period. Sound intensity greater than 90 dB can be experienced, but for shorter periods of time. For every 5 dB increase, the permissable number of hours of exposure is halved.

An easy way to remember the OSHA limits for occupational noise exposure is to remember that the scale begins at 8 hours of 90 dB noise exposure, and for every 5 dB increase in the sound level, the permissable number of hours at that level is halved. So, 8 hours at 90 dB, 4 hours at 95 dB, 2 hours at 100 db, etc.

1. A 60 year-old-male presents for preoperative discussion before removing his 2 cm left-sided jugular foramen paraganglioma. In addition to the jugular bulb, the tumor involves part of the vertical petrous carotid. You decide to employ a combined transmastoid and transcervical approach to remove the tumor. Based on the classically described Fisch approaches to the infratemporal fossa, what type of approach is most appropriate for his lesion?
2. **Type A**
3. Type B
4. Type C
5. Type A or B
6. Type B or C

**Difficulty Level: Hard**

**Explanation:** Dr. Fisch described three approaches to the infratemporal fossa (A, B, C) and their resulting exposure. All methods involve mastoidectomy and facial nerve dissection and transposition and are used to resect a jugular foramen paraganglioma (formerly referred to as a glomus jugulare tumor).

The patient described above has a jugular foramen paraganglioma necessitating a type A approach. It does not involve any deeper structures that would require a type B or C approach.

1. Which of the following is true in regard to cochlear afferents?
2. 90% originate from outer hair cells.
3. **90% originate from the inner hair cells.**
4. They first synapse in the inferior colliculus.
5. They first synapse in the lateral lemniscus.
6. They travel in the olivocochlear bundle.

**Difficulty Level: Hard**

**Explanation:** Each inner hair cell is innervated by 10-20 type I bipolar neurons and these fibers constitute 90-95% of the fibers of the cochlear nerve.

Cochlear nerve afferent fibers receive the majority of their input from inner hair cells. This sensory information is then relayed to the dorsal and ventral cochlear nuclei.

1. A 50-year-old male presents with a three day history of sudden-onset and persistent vertigo that worsens with head movement. He has had to limit his oral intake due to the associated nausea and vomiting. He denies hearing loss or other neurologic complaints. Caloric testing demonstrates a loss of response on the right side.
2. Valacyclovir
3. **Methylprednisolone**
4. Meclizine
5. Observation and vestibular therapy

**Difficulty Level: Hard**

**Explanation:** The etiology of vestibular neuritis is debateable, though the two most common hypotheses are thought to result from viral induced inflammation (specifically the reactivation of a latent herpes simplex virus infection in Scarpa's ganglion) or a small vessel occlusion. Though caloric testing is not routinely performed in the acute phase, reduction or loss of caloric response is the most consistent laboratory finding. Treatment is mostly supportive, but for severe symptoms, IV hydration and antiemetics are helpful until symptoms abate. The use of steroids can improve symptoms over the short term, though it is unclear whether it changes the clinical course over the long term. This patient has moderate symptoms, and out of all of the answer choices, a trial of steroids would be most appropriate. If symptoms were less severe, observation would be more appropriate.

1. A 25 year old female suffers a knife stab wound to the right side of the face, and in the recovery process notes a decrease in salivation from the right parotid gland. What nerve was likely damaged?
2. Greater superficial petrosal nerve
3. **Auriculotemporal**
4. Caroticotympanic nerve
5. Chorda tympani
6. Great auricular

**Difficulty Level: Moderate**

**Explanation:** Parasympathetic fibers destined to modulate parotid gland function originate from the inferior salivatory nucleus of CN IX, and travel with the tympanic nerve to the middle ear. From the tympanic plexus, the lesser petrosal nerve exits the middle ear in the hiatus for the lesser petrosal nerve and enters the floor of the middle fossa. It then exits the middle fossa through the cannaliculus innominatus to enter the infratemporal fossa, where it synapses with post ganglionic fibers in the otic ganglion. These fibers briefly travel with the auriculotemporal nerve (CN V3) before reaching the parotid gland to provide secretomotor function.

The auriculotemporal nerve is a branch of V3 but carries parasympathetic fibers from the otic ganglion to the parotid gland.

1. Which approach to a cerebellopontine angle tumor carries the highest risk of facial nerve injury?
2. Translabyrinthine
3. Retrosigmoid
4. **Middle cranial fossa**
5. None of the above
6. All carry an equal risk of injury to the facial nerve

**Difficulty Level: Hard**

**Explanation:** Outcomes of the translabyrinthine and retrosigmoid approaches in regards to facial nerve injury are similar. The middle cranial fossa approach however is associated with a higher incidence of transient weakness in tumors with <10mm extension into the CPA and with permanent weakness in tumors with a 10 to 18mm CPA extension.

The middle cranial fossa approach to CPA masses carries the highest risk of facial nerve injury compared to the translabyrinthine or retrosigmoid approaches.

1. Which is true regarding the facial nerve?
2. The development of the extratemporal portion of the facial nerve begins in the 8th week
3. The nervus intermedius contains postganglionic parasympathetic neurons innervating the lacrimal gland
4. **The labyrinthine segment has a diameter averaging around 0.7mm**
5. The cog projects inferior to the facial nerve as it enters the tympanic cavity

**Difficulty Level: Hard**

**Explanation:** The labyrinthine segment is the narrowest portion of the fallopian canal and is thought to be the most commonly affected site of injury in conditions such as Bell’s palsy, and for this reason, is an area of interest in facial nerve decompressions.

The labyrinthine segment of the facial nerve is the narrowest portion of the fallopian canal. It is commonly implicated in Bell's palsy, where inflammation of the nerve is thought to produce a compressive neuropathy with no path of expansion.&nbsp;

1. What is the function of the GJB2 gene?
2. **It encodes a gap junction protein that transmits ions**.
3. It maintains the osmotic gradient of the stria vascularis.
4. It serves to provide micronutrients to hair cells.
5. It regulates the development of the pars inferior.
6. It causes susceptibility to ototoxicity.

**Difficulty Level: Easy**

**Explanation:** GJB2 encodes the protein Connexin 26, which, along with 5 other connexins, forms a connexon. Connexons of neighboring cells dock with eachother to form a gap junction, through which potassium ions are transmitted from outer hair cells back through the supporting cells and spiral ligament to the stria vascularis. Disruption of the function of these gap junctions prevents normal mechanosensory tranduction of hair cells, and leads to sensorineural hearing loss. Nonsyndromic hearing loss accounts for approximately 2/3 of hereditary hearing loss cases, and of these, the majority are due to connexin 26 mutations.

Mutations in GJB2 result in dysfunctional connexin 26 proteins, and subsequent congenital sensorineural hearing loss. Connexin 26 mutations account for the majority of hereditary nonsyndromic hearing loss.

1. Which of the following is an advantage of monoaural hearing compared to binaural hearing?
2. Dynamic range reduction.
3. Summation.
4. Sound localization.
5. Squelch.
6. **None of the above.**

**Difficulty Level: Moderate**

**Explanation:** A. Patients with sensorineural hearing loss in which, due to loudness recruitment, the patient’s subjective experience of loudness increases disproportionally compared to someone with normal hearing. Dynamic range represents the range of volumes at which a sound can be perceived, from barely audible to maximum loudness without discomfort. Dynamic range reduction or dynamic compression algorithms used in binaural hearing aids can improve hearing.

1. A 24-year-old female presents with a two-day history of dizziness. When she woke up this morning and sat up, she felt the room spinning. The episode lasted about one minute. A second episode was associated with nausea. She reports an episode of viral gastroenteritis that resolved one month ago. She denies any headache, tinnitus, hearing deficits, vision changes, weakness, or gait abnormalities. Her history is significant for allergic rhinitis, treated with fluticasone nasal spray. Vital signs are normal. Cardiac, pulmonary, and neurological examinations are unremarkable. The Dix-Hallpike maneuver is performed, which causes upward torsional nystagmus after about 30 seconds. After the nystagmus resolves, the patient sits up, and it recurs in the opposite direction. What is the diagnosis?
2. Vestibular neuronitis
3. Meniere disease
4. Cerebellar infarction
5. **Benign paroxysmal positional vertigo**

**Difficulty Level: Easy**

**Explanation:** Vertigo is a common complaint in primary care, specialty, and emergency medicine settings. Therefore, it is essential to differentiate between different causes of vertigo and treat it appropriately. Both duration of vertigo and the results of a Dix-Hallpike maneuver can help to differentiate peripheral from central vertigo. Central positional vertigo is characterized by static downbeat nystagmus that persists as long as the provocative position is maintained. The torsional component seen in peripheral vertigo is another differentiating factor. In peripheral disease, there is a latency of nystagmus and fatiguability, with the duration of nystagmus typically <1 min. The most common cause of peripheral vertigo is benign paroxysmal positional vertigo (BPPV). This is caused by otoliths moving around in the semicircular canals. The classic symptom is that vertigo is induced by a quick turning of the head, such as rolling over in bed or looking over one’s shoulder. The direction of nystagmus depends upon the type of BPPV, but posterior canal BPPV is the most common. This patient’s Dix Hallpike maneuver supports posterior canal BPV as the nystagmus is upward and torsional, and there is a 30-second delay after the maneuver. The Epley maneuver can cure BPPV rapidly.

Vertigo is a common complaint with multiple possible causes and exists in central and peripheral forms. The most common form of peripheral vertigo is BPPV, and the Dix Hallpike maneuver can be diagnostic.

1. Within the internal auditory canal which of the following is located superior to the transverse crest and anterior to Bill's Bar?
2. **CN VII**
3. Cochlear nerve
4. Superior vestibular nerve
5. Inferior vestibular nerve

**Difficulty Level: Easy**

Cranial nerve VII is in an anterior superior position within the internal auditory canal. The position of the contents of the IAC can be remembered using the mnemonic “7-Up, Coke down” which corresponds to the following diagram (see image below).

1. A 48 year-old female presents for the evaluation of left hearing loss for several years. She has a history of a large left tympanic membrane perforation that failed a previous attempt at underlay tympanoplasty in the past. She currently reports no otalgia, otorrhea, or vertigo. Examination reveals a near-total perforation of the left tympanic membrane. You decide to proceed with tympanoplasty with a lateral grafting technique. Which of the following helps to decrease the incidence of post-operative blunting?
2. Avoiding placement of any canal packing into the anterior sulcus.
3. Removal of the anterior canal wall bulge.
4. Avoid direct placement of the graft over the anterior canal bone.
5. All of the above.
6. **B and C only**

**Difficulty Level: Hard**

**Explanation:** The lateral graft tympanoplasty is usually reserved for near total, total, or marginal perforations, or those that have failed prior medial grafting techniques. The advantages of the lateral graft technique include better exposure, improved graft take rate, and applicability to all types of perforations. The disadvantages of the lateral grafting technique include prolonged healing time, graft lateralization, blunting of the angle at the anterior sulcus, and the formation of epithelial cysts from in-turning of skin flap edges during surgery. Blunting of the angle at the anterior sulcus typically occurs due the failure to preserve the tympanomeatal angle, thus creating a dead space for scar tissue to form in and creates a more obtuse angle than the physiologic norm. Both blunting and lateralization can lead to significant post-operative persistent conductive hearing loss due to loss of the vibratory surface area of the tympanic membrane. To help prevent blunting, the anterior canal wall bone should be removed, opening up the anterior sulcus. If possible, it is important to ensure overlapping of the canal skin and the graft and ideally an anterior tympanic membrane remnant is present to avoid placement of the graft directly over the bone. Finally, placement of gelfoam packing over the graft at the anterior sulcus may help prevent blunting in that area. The goal should be to preserve an angle of 90 degrees or less at the anterior sulcus.

Blunting is a potential complication of the lateral graft tympanoplasty technique. Risk of blunting can be decreased by removing the anterior canal wall bone, avoiding placement of the graft over anterior wall bone, and packing over the graft at the anterior sulcus.

1. An 86 year old female has been followed for five years for a right sided acoustic neuroma. She had moderate sensorineural hearing loss and grade II House-Brackmann facial nerve function. On recent MRI the lesion was noted to have increased 5mm over the past 12 months with a size of 2.5cm by 3.2cm. You explain that risks and benefits of surgery versus gamma-knife radiation given the significant increase in tumor size. What is a contraindication to gamma knife radiation in this patient?
2. age >75
3. serviceable hearing
4. grade II facial nerve function
5. **size of tumor**

**Difficulty Level: Moderate**

**Explanation:** There are few contraindications to gamma knife radiosurgery but large cerebellopontine tumors (>3cm) may swell following radiation which can lead to post-treatment obstructive hydrocephalus. Absolute contraindications includes inferior spread of the tumor that is not adequately addressed by the collimator helmet. There is also some controversy over the utility of gamma knife surgery for patients with neurofibromatosis type II as traditionally they have been thought to respond poorly to treatment, though at present, gamma knife surgery remains a viable option for this patient population. When discussing treatment with gamma knife radiation it should also be explained that gamma knife radiation does not cure the tumors but rather helps to halt growth thereby preventing further symptoms. Surgery may still be required if tumors progress despite radiation.

Gamma knife radiosurgery is typically reserved for tumors <3 cm.

1. A 37 year old male with no significant past medical history presents to the ED after being assaulted. A high resolution CT scan of the head reveals a left sided otic capsule sparing temporal bone fracture. The patient is evaluated and is found to have a complete facial nerve paralysis on the left. Where is the most likely site of facial nerve injury?
2. The cerebellopontine angle
3. Within the internal auditory canal
4. **The perigeniculate region**
5. The vertical segment
6. At the stylomastoid foramen

**Difficulty Level: Easy**

**Explanation:** The perigeniculate region (geniculate ganglion and tympanic segment) was the location of the injury 93.3% of the time.

The perigeniculate region is the most common site of involvement in traumatic facial nerve injuries.

1. What is the mode of inheritance of branchio-oto-renal syndrome?
2. Autosomal recessive
3. **Autosomal dominant**
4. X-linked recessive
5. Mitochondrial
6. Sporadic

**Difficulty Level: Moderate**

**Explanation:** Autosomal dominant conditions of congenital hearing loss include Waardenburg syndrome, Apert syndrome, Neurofibromatosis type 2, Treacher Collins syndrome, Crouzon’s syndrome, Branchio-Oto-Renal syndrome, and Stickler syndrome (WANT CBS).

Branchio-oto-renal syndrome follows an autosomal dominant inheritance pattern.

1. A 5 year old child presents with a large pulsatile mass in the posterior mesotympanum. The internal jugular vein and the carotid artery are normal, but there is noted to be obliteration of the foramen spinosum on the CT scan. What is the most likely diagnosis?
2. A caroticotympanic artery
3. **Persistent stapedial artery**
4. Tympanic paraganglioma
5. Jugular foramen paraganglioma
6. Petrous apex cholesterol granuloma

**Difficulty Level: Moderate**

**Explanation:** The persistent stapedial artery courses through the crura of the stapes. There is an association with an absent foramen spinosum, with aplasia of the middle meningeal artery.

A persistent stapedial artery should be suspected in cases of pulsatile middle ear masses and absence of a foramen spinosum on imaging. Diagnosis may be confirmed with a CT or angiography.

1. A 23-year-old with late-onset congenital syphilis presents to your clinic after a hearing test. The audiogram shows bilateral symmetric moderate sensorineural hearing loss. He reports that he experienced an episode of vertigo when he was next to the stage at a rock concert the previous week. In regard to middle ear anatomy, which of the following have to be present for him to experience this phenomenon?
2. Intact tympanic membrane, disrupted ossicular chain, and mobile footplate
3. Perforated tympanic membrane, disrupted ossicular chain, and mobile footplate
4. Intact tympanic membrane, intact ossicular chain, and fixed footplate
5. **Intact tympanic membrane, intact ossicular chain, and mobile footplate**
6. Perforated tympanic membrane, intact ossicular chain, and mobile footplate

**Difficulty Level: Moderate**

**Explanation:** Tullio phenomenon is often seen in patients with congenital syphilis and consists of vertigo and nystagmus on stimulation with high-intensity sound. This phenomenon is most commonly due to superior canal dehiscence, however the pathophysiology for otosyphilis is different. Inflammatory lesions of the labyrinth (gummas) lead to labyrinthine fistulas, endolymphatic hydrops with eventual degeneration of the membranous labyrinth. Because it is a hydropic process, otosyphilis has many of the same features as Meniere's disease: Episodic vertigo associated with tinnitus, aural pressure, and hearing loss. There is also a loss of type II hair cells in the semicircular canals and otoliths in otosyphilis.

An intact sound transmission mechanism (intact tympanic membrane, ossicular chain, and mobile footplate) must be present to test for the Tullio phenomenom.

1. A patient undergoes acoustic reflex threshold testing as part of a possible retrocochlear pathology workup. A 70 decibel click is applied to activate the afferent limb of the reflex. The efferent limb is measured by:
2. listening with a microphone in the external ear for a return click
3. **assessing changes in the tympanic membrane compliance**
4. visualizing eye movements related to noise exposure
5. assessing electrical activation of the sternocleidomastoid muscle via an electrode pad
6. visualizing rotational nystagmus with Frenzel lenses

**Difficulty Level: Moderate**

**Explanation:** The acoustic reflex is a contraction of the bilateral stapedius muscles, innervated by the facial nerve, upon exposure to a loud noise, typically within the 70-100 decibel range. The contraction of the stapedius muscle decreases the tympanic membrane compliance as a protective measure. This reflex is measured either with acoustic reflex threshold testing or via acoustic reflex decay. It may be useful for assessing the neural network between cranial nerves seven and eight and their connective pathway in the brainstem.

The acoustic reflex relies upon an intact eigth cranial nerve to sense a loud noise and an intact seventh cranial nerve to contract the stapedius muscles. Since contraction of the stapedius muscles will lead to a decrease in the compliance of the tymapnic membrane, assessing this change in response to a loud noise will test the integrity of this reflex pathway.

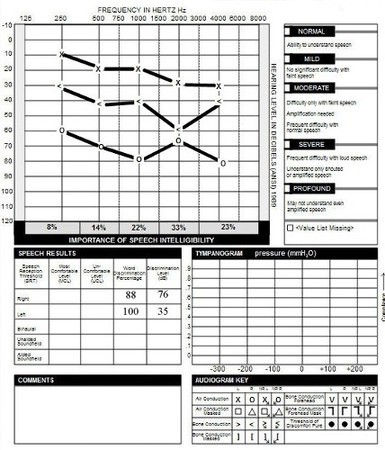
1. The cochlear nerve auditory pathway involves all of the following sites except:
2. Cochlear nucleus
3. Inferior colliculus
4. **Superior colliculus**
5. Lateral lemniscus
6. Auditory cortex
7. Superior olivary complex

**Difficulty Level: Easy**

**Explanation:** The cochlear nerve contains afferent fibers transmitting information from the inner and outer hair cells to the cochlear nucleus in the brainstem. There are ventral and dorsal nuclei representing different tonotopic maps. Projections from the cochlear nucleus travel via the acoustic stria via the superior olivary complex (SOC) and inferior colliculus. The SOC sends fibers to the lateral lemniscus, inferior colliculus and auditory cortex. There are multiple interconnected pathways amongst these structures evidenced by the complexity of auditory processing and signaling. The superior colliculus is described as a visual reflex center without participation in auditory signaling.

The basic pathway for cochlear afferent fibers is from the inner and outer hair cells of the cochlea to the cochlear nerve, cochlear nuclei, superior olivary complex, lateral lemniscus, inferior colliculus and auditory cortex.

1. A 39 year old female presents complaining of hearing loss. She has a brother who had hearing loss in his early 40s. She has a history of recurrent bilateral ear infections as a child and underwent placement of a single pair of pressure equalization tubes bilaterally. On physical exam her bilateral tympanic membranes are sclerotic and you are unable to visualize beyond the tympanic membrane. She has the audiogram shown below, and stapedial reflex is absent. What is the most likely diagnosis?



1. Cholesteatoma
2. Tympanosclerosis
3. Ossicular chain discontinuity
4. **Otosclerosis**
5. Malingering

**Difficulty Level: Easy**

**Explanation:** This patient shows a mixed hearing loss in her right ear with a gap between air and bone conduction, indicating conductive hearing loss, which closes at 2,000 Hz. This is known as Carhart's notch, which is an artificial drop in the bone line due to disruption of ossicular resonance. The Carhart notch is consistent with otosclerosis, although can occur in other disorders including ossicular chain discontinuity. These patients may also experience the phenomenon of paracusis of Willis. This is where a patient hears better in noise due to the dampening of background noise by the conductive hearing loss, and because the speaker generally talks louder in a situation with background noise. A family history of otosclerosis is found in 50% of patients with the disorder, so otosclerosis is suggested by the family history of early onset hearing loss in her brother. There is also presumed to be a hormonal component contributing to otosclerosis as there is a female predominance, and often symptoms are noted around a pregnancy or menopause.

A patient with conductive hearing loss with a Carhart notch and family history of hearing loss in mid adulthood should raise suspicion for otosclerosis.

1. The classic inheritance pattern of otosclerosis is:
2. **autosomal dominant**
3. autosomal recessive
4. X-linked
5. mitochondrial disorder

**Difficulty Level: Easy**

**Explanation:** Multiple studies have supported this autosomal dominant inheritance pattern. Penetrance is in the range of 20 to 40, and multiple gene loci have been implicated. It is estimated that roughly 10% of adult Caucasians have some degree of otosclerosis. Otosclerosis involves abnormal bone growth, commonly around the stapes footplate leading to slowly progressive hearing loss that most often begins during the third decade of life. A low frequency conductive hearing loss is a common early finding on audiogram. Treatment ranges from observation to amplification to surgery. Sodium fluoride has been shown in some patients to slow disease progression. The surgical management of the disease includes stapedectomy which has an excellent success rate approaching 90% for hearing improvement.

Otosclerosis displays an autosomal dominant mode of inheritance with variable penetrance.

1. A patient has an external auditory canal neoplasm with full thickness bone involvement, soft tissue involvement of 0.3 cm, and a House-Brackmann score of VI on the involved side. According to the modified University of Pittsburgh Staging System, what is the T stage?
2. T1
3. T2
4. T3
5. **T4**

**Difficulty Level: Hard**

**Explanation:** The modified University of Pittsburgh Staging System is the most commonly used for temporal bone neoplasms. The T stage (which is different from the AJCC T staging) is as follows:

T1: Tumor limited to the external auditory canal without bony erosion or evidence of soft-tissue extension

T2: Tumor with limited external auditory canal bony erosion (not full thickness) or radiographic finding consistent with limited (<0.5 cm) soft-tissue involvement

T3: Tumor eroding the osseous external auditory canal (full thickness) with limited (<0.5 cm) soft-tissue involvement, tumor involving middle ear and/or mastoid.

T4: Tumor eroding the cochlea, petrous apex, medial wall of the middle ear, carotid canal, jugular foramen or dura, with extensive (>0.5 cm) soft-tissue involvement, or with facial paresis/paralysis.

A temporal bone neoplasm eroding the cochlea, petrous apex, medial wall of the middle ear, carotid canal, jugular foramen or dura, with extensive (>0.5 cm) soft-tissue involvement or facial paresis/paralysis is classified as a T4 tumor under the modified University of Pittsburgh Staging System.

1. A 7-year old male child presents to the emergency room following a penetrating injury to the left external auditory canal with a pencil while playing with his sister. His parents inform you that he had bloody drainage following the injury. Upon questioning, he is asking you to repeat your questions frequently and is complaining that his ‘head is really spinning badly’. On otoscopic exam you note a perforation to the posterior superior quadrant on his left tympanic membrane with some evidence of clear fluid exiting the tympanic membrane. What is the appropriate treatment for this child?
2. Discharge home with instructions for bed rest and stool softeners
3. **Surgery with middle ear exploration**
4. Vestibular suppressants
5. Obtain audiogram for baseline hearing level, discharge home on empiric antibiotics followed by serial audiograms

**Difficulty Level: Moderate**

**Explanation:** This child exhibits symptoms and signs of a traumatic perilymphatic fistula. Management of a presumed perilymphatic fistula typically initially involves bed rest and observation for 24-72 h. This allows sufficient time for all membrane leaks (labyrinthine concussion) to heal. Preoperative hearing loss in traumatic perilymphatic fistula indicates a poor hearing prognosis. Patients with a high index of suspicion for PLF following traumatic injury or those with severe symptoms in the form of intractable vertigo, progressive severe sensorineural hearing loss, radiologic evidence of PLF or meningitis should undergo more prompt exploration. Exploratory tympanotomy is the only management that is both diagnostic and therapeutic for PLF and continues to be the gold standard for diagnosis.

In patients with penetrating ear trauma, middle ear exploration is indicated in instances of intractable vertigo, progressive SNHL, radiographic evidence of a PLF or meningitis. Less severe cases may undergo a trial of conservative management with bed rest and observation.

1. A 12 month old male is found to have bilateral profound sensorineural hearing loss. After a complete medical evaluation the patient is found to be a cochlear implant candidate. After routine pre operative CT imaging the patient is found to have a cochlear anomaly. What is the most likely CT finding?
2. Cochlear aplasia
3. Common cavity
4. Complete labrynthine aplasia
5. Cochlear hypoplasia
6. **Incomplete partition**

**Difficulty Level: Hard**

**Explanation:** Incomplete partition is the most common malformation of the cochlea. In incomplete partition the cochlea only completes only 1 to 1.5 turns as compared to the normal 2.5 turns. When a cochlear anomaly is present, incomplete partition is the cause 42% of the time followed by common cavity (19.4%), cochlear hypoplasia (11.2%), cochlear aplasia (2%), and complete labrynthine aplasia (1%). Of note, cochlear implant surgery may be planned around most of these malformations, the exception being complete labrynthine aplasia.

Incomplete partition is the most common malformation of the cochlea. This occurs due to abnormal development of the cochlea during week 7, and results in 1 to 1.5 turns instead of 2.5 turns in the cochlea.

1. The superficial sensation for the external ear, ear canal, and tympanic membrane is supplied by which of the following nerves?
2. Great auricular nerve
3. Facial nerve
4. Vagus nerve
5. **A, B & C**
6. A and B only

**Difficulty Level: Easy**

**Explanation:** The sensory innervation of the external ear is provided by the following nerve/nerve branches:

CN V: The auriculotemporal branch of V3 innervates the tragus, anterior pinna, anterior lateral surface of the tympanic membrane, and anterosuperior external auditory canal wall.

CN VII: The facial nerve innervates the lateral concha and antihelix, lobule, mastoid, posterior external auditory canal, and posterior portion of the tympanic membrane.

CN IX: The tympanic branch of the glossopharyngeal nerve (Jacobson's nerve) innervates the eustachian tube, mastoid air cells, medial surface of the tympanic membrane, and mucosa of the middle ear.

X: The auricular branch of the vagus nerve (Arnold's nerve) innervates the concha, inferoposterior external auditory canal, tympanic membrane, and postauricular skin.

The sensory innervation of the external ear is provided by CN V, VII, IX, X and branches of the cervical plexus.

1. The chorda tympani nerve passes through the middle ear in which of the following locations?
2. Medial to the captiulum of the stapes.
3. Between the crura of the stapes.
4. Between the stapes and the incus.
5. **Between the malleus and the incus.**
6. Lateral to the malleus.

**Difficulty Level: Moderate**

**Explanation:** The chorda tympani nerve carries both efferent parasympathetic fibers (from the superior salivatory nucleus) and special sensory fibers (from the nucleus tractus solitarius). The fibers from these nuclei exit the brainstem with other parasympathetic fibers as the nervus intermedius (nerve of Wrisberg). The nervus intermedius travels alongside the facial nerve within the internal auditory canal to the geniculate ganglion, where some fibers will pass through the ganglion and form the greater superficial petrosal nerve, and others join the motor portion of the facial nerve. In the vertical segment of the facial nerve, the chorda tympani branches off of the facial nerve and travels anterior to the vertical segment before entering the midddle ear. The chorda tympani passes between the manubrium of the malleus and long process of the incus, and then exits the middle ear through the petrotympanic fissure (Hugier canal). It then enters the infratemporal fossa, where it joins the lingual nerve (CN V3). After traveling with the lingual nerve, special sensory afferents will relay taste sensation from the anterior 2/3 of the tongue and parasympathetic efferent fibers will synapse in the submandibular ganglion with post ganglionic fibers innervating the submandibular and sublingual glands.

Within the middle ear, the chorda tympani passes between the manubrium of the malleus and long process of the incus before exiting the ear through the petrotympanic fissure.

1. A 73-year-old male presents with a 6-month history of left-sided otalgia and recurrent purulent otorrhea. He states that antibiotic otic drops clear the drainage but the pain has become persistently worse over the past few months. The canal is debrided and an exophytic erythematous mass is seen from the posterior EAC wall. In-office biopsy reveals this to be squamous cell carcinoma. Staging computed tomography reveals full-thickness bony erosion of the medial posterior canal with thickening of the tympanic membrane but no involvement of the middle ear. No cervical adenopathy is noted. What is this patients stage per the Pittsburgh staging system?
2. T1N0
3. T2N0
4. **T3N0**
5. T4N0
6. Insufficient information to accurately determine stage

**Difficulty Level: Hard**

**Explanation:** The lesion described above is best classified as a T3N0 squamous cell carcinoma of the temporal bone. The most widely accepted staging system is the Pittsburgh staging system:

- T1 – limited to the external auditory canal (EAC) without bony erosion or soft tissue involvement

- T2 – limited bony erosion of the EAC (not full thickness), limited soft tissue involvement

- T3 – full thickness bony EAC erosion, involvement of the middle ear or mastoid

- T4 – involvement of the carotid canal, jugular bulb, medial middle ear, petrous apex, or facial nerve

The Pittsburgh staging system is used to stage squamous cell carcinomas primary tumors of the temporal bone. Nodal and distant metastasis is the same as for other head and neck malignancies.

1. Which of the following would be expected in a patient with Alport syndrome?
2. **Sensorineural hearing loss**
3. White cell casts
4. Thrombocytopenia
5. Retinitis pigmentosa
6. More prevalent in females

**Difficulty Level: Easy**

**Explanation:** Alport syndrome is an X-linked defect of type IV collagen. Type IV collagen is mostly found in basement membranes of the kidneys, cochlea and eyes, and these are the areas of the body most affected in Alport syndrome. Patients will develop glomerulonephritis, hematuria and proteinuria, and will eventually progressive to renal failure. Hearing is often normal at birth and progressively worsen, typically worse in the high frequencies, in young adulthood. Sight is typically not threatened, though various vision abnormalities have been described.

Alport syndrome is an X-linked disease leading to high frequency SNHL, vision changes and renal failure.

1. The cochlear aqueduct contains which of the following?
2. The cochlear artery
3. **The periotic duct**
4. The singular nerve
5. The cochlear nerve
6. None of the above

**Difficulty Level: Hard**

**Explanation:** The cochlear aqueduct contains the periotic duct (perilymphatic duct). It connects the scala tympani at the basal turn of the cochlea to the subarachanoid space.

The cochlear aqueduct connects the scala tympani to the subarachnoid space. The scala tympani contains perilymph, which has a similar composition to the CSF within the subarachnoid space.

1. Which of the following is true regarding Gradenigo's Syndrome?
2. It is associated with sensorineural hearing loss
3. **It is associated with infection within the petrous apex**
4. It is characterized by involvement of CN VII
5. It is associated with otalgia.

**Difficulty Level: Easy**

**Explanation:** The classic triad of Gradenigo’s syndrome includes otorrhea, abducens palsy, and retro-orbital pain. These are due to petrous apicitis and the inflammatory effects on adjacent structures. The retro-orbital pain is related to irritation in the area of ophthalmic division of the trigeminal nerve, and the abducens palsy is due to inflammation in the region of the Dorello canal. Gradenigo's syndrome is typically a complication of acute otitis media. Initial treatment is similar to that of acute otomastoiditis with appropriate antibiotics. However, surgery is essential in the long term management of this syndrome to establish drainage of the apical air cells. Surgical approaches are variable based on the patient’s anatomy, specifically the jugular bulb and superior semicircular canal.

Gradenigo's syndrome is characterized by otorrhea, abducens palsy and retro-orbital pain. It is due to petrous apicitis, likely secondary to acute otitis media.

1. A 7-year-old boy with a history of allergic rhinitis is referred to clinic for ear pressure and fullness. Otoscopy reveals bilateral tympanic membrane retractions. The patient is diagnosed with Eustachian tube dysfunction. From which branchial structure does the Eustachian tube originate?
2. First branchial cleft
3. **First branchial pouch**
4. Second branchial cleft
5. Second branchial pouch
6. Third branchial pouch

**Difficulty Level: Hard**

**Explanation:** The first branchial pouch becomes elongated and is incorporated into the temporal bone and forms the tubotympanic recess, which eventually becomes the middle ear and eustachian tube. The most lateral portion of the pouch, along with the closing plate of the first branchial cleft, forms the inner layer of the tympanic membrane.

The Eustachian tube and middle ear derived from the first pharyngeal pouch.

1. Which of the following is the most common cause of neonatal facial paralysis?
2. Mobius Syndrome
3. neonatal CMV infection
4. Goldenhar Syndrome
5. **birth trauma**

**Difficulty Level: Easy**

**Explanation:** Approximately 88% of cases of facial paralysis in live births are due to difficult labor, and the majority of those are associated with a history of forceps delivery.

The majority of cases of neonatial facial paralysis are due to birth trauma.

1. Preganglionic parasympathetic fibers carried in the greater superficial petrosal nerve synapse in which ganglion?
2. otic ganglion
3. submandibular ganglion
4. **sphenopalatine ganglion**
5. ciliary ganglion

**Difficulty Level: Hard**

**Explanation:** The pterygopalatine ganglion is commonly referred to as the sphenopalatine ganglion. The preganglionic parasympathetic fibers in the GSPN synapse here then travel to supply the lacrimal glands and seromucinous glands within the nasal cavity.

Preganglionic parasympathetic fibers carried in the greater superficial petrosal nerve synapse in the sphenopalatine ganglion.

1. Scheibe’s anomaly is best described as:
2. Absence of the utricle and semicircular canals
3. Complete labyrinthine aplasia
4. Aplasia of the cochlear aqueduct
5. A confluent cochlea and vestibule
6. **Dysplasia of the cochlea and saccule**

**Difficulty Level: Hard**

**Explanation:** Cochleosaccular dysgenesis (Scheibe aplasia) is a malformation limited to the membranous portion of the pars inferior. This is the most commonly observed temporal bone dysplasia in cases of congenital profound sensorineural hearing loss. There is a range of histopathologic findings which often includes a partially or fully missing Organ of Corti, collapse of the cochlear duct, and degeneration of the stria vascularis. Radiographic findings are typically normal. Audiogram shows a profound sensorineural hearing loss. Amplification is helpful for many patients.

Scheibe's anomaly describes a malformation of the membranous portion of the pars inferior.

1. Which of the following audiogram patterns correlates to complete ossicular discontinuity?
2. Air-bone gap greater in higher frequencies that lower frequencies
3. Air-bone gap greater in lower frequencies than higher frequencies
4. Depression in bone conduction thresholds, more significantly at 2000 Hz
5. **Air-bone gap consistent across all frequencies**
6. Air conduction thresholds better in mid frequencies

**Difficulty Level: Easy**

**Explanation:** With complete ossicular discontinuity, the air-bone gap is consistent across all frequencies. There is near maximum conductive hearing loss (assuming an intact tympanic membrane) and air-conduction and bone-conduction thresholds look like parallel lines 50 to 60 dB apart across the frequencies.

With complete ossicular discontinuity, the air-bone gap is consistent across all frequencies and approximately 50-60 dB apart if the tympanic membrane is intact.

1. A 77-year-old man with type 2 diabetes presents with a two week history of progressively worsening left ear pain. He has been treated with two courses of oral antibiotics without significant improvement. On examination, he has intense otalgia and drainage from his ear. His tympanic membrane cannot be visualized due to edema and granulation tissue in the ear canal. He is also noted to have a dysphagia and facial droopiness. Which of the following is true of this condition?
2. Cranial nerve involvement has no significant correlation with worse mortality
3. Streptococcus pneumoniae is the single most likely pathogen
4. **Biopsy is recommended to rule out carcinoma**
5. CT and technetium scans are used to follow the progress of this condition
6. The cranial nerves of the jugular foramen (CN IX, X, XI) are most frequently affected

**Difficulty Level: Moderate**

**Explanation:** The patient has evidence of necrotizing (malignant) otitis externa, which has overlapping features with temporal bone carcinomas. MOE is seen almost exclusively in the elderly, the immunocompromised, and diabetic patients. It is far more serious than simple otitis externa due to the risk of spread of infection to the mastoid, the dural sinuses, and the meninges. Early, mild cases may be treated with outpatient fluoroquinolones active against Pseudomonas, but most cases will require admission for IV antibiotics, aural toileting and possible surgical debridement.

Carcinomas of the ear canal may mimic many features of necrotizing otitis externa and biopsies are required to rule this out.

1. A 25 year old male comes to the ED after being involved in a ATV accident. The patient is a GCS of 3 and is intubated. A head CT shows a subdural hematoma and a skull base fracture that is shown below. No facial nerve exam is able to be done due to the patient'™s GCS. Otoscopic exam reveals blood in the EAC and hemotympanum. There is no obvious leakage of clear fluid. What is this patient'™s overall risk of developing meningitis?



1. **1%**
2. 10%
3. 30%
4. 50%
5. 75%

**Difficulty Level: Hard**

**Explanation:** In a review of 547 temporal bone fractures at the University of California- Davis there was a 1% incidence of meningitis when there was no cerebrospinal fluid fistula.

The incidence of meningitis in patients with temporal bone fractures without a CSF leak is very low.

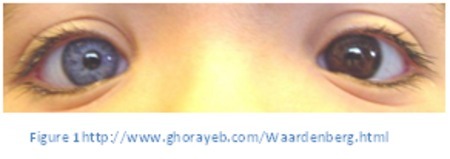
1. A 35 year old male with no significant past medical history presents to the ENT clinic with a 6 month history of a right sided subjective hearing loss, right sided otorrhea, and vertigo. On physical exam there is a tympanic membrane perforation on the right with obvious cholesteatoma. The patient has nystagmus with pneumatic otoscopy on the right and a labrynthine fistula is suspected. What is the most likely location of the labyrinthine fistula?
2. **Horizontal semicircular canal**
3. Posterior semicircular canal
4. Superior semicircular canal
5. Cochlea
6. Saccule

**Difficulty Level: Easy**

**Explanation:** A labrynthine fistula is an erosive loss of endochondral bone without the loss of perilymph and is almost exclusively caused by cholesteatoma. Clinical symptoms of a labrynthine fistula include vertigo, disequilibrium, and imbalance. The vestibular symptoms however are intermittent and patients tend to have normal vestibular function most of the time. Most labrynthine fisutlas involve the horizontal semicircular canal. Rarely erosions into the superior semicircular canal, posterior semicircular canal, cochlea, and/or vestibule have been reported.

Most labyrinthine fistulas involve the horizontal semicircular canal and are due to erosion from a cholesteatoma.

1. A 10 year old boy presents with facial features as seen below. What distinguishes Waardenburg type 1 from Waardenburg type 2 syndromes?



1. Presence of a white forelock
2. **Displacement of the inner canthi**
3. Synophrys
4. Pigment changes in the iris
5. Severity of hearing loss

**Difficulty Level: Hard**

**Explanation:** There are 4 types of Waardenburg syndrome, and 3/4 are inherited in autosomal dominant fashion. Major criteria for diagnosis include congenital SNHL, iris pigment abnormalities, hair hypopigmentation, dystopia canthorum, and a first degree relative with Waardenburg. Minor criteria include congenital leukodermia, synophrys, broad/high nasal root, hypoplastic nasal alae, or premature graying of hair. Diagnosis requires either 2 major or 1 major and 1 minor criteria being met. Type 2 Waardenburg is similar to Type 1 but without dystopia canthorum. Type 3 is associated with limb hypoplasia. Type 4 is either autosomal dominant or recessive, and associated with Hirschsprung disease or other neurologic abnormalities.

Type 2 Waardenburg syndrome is similar to Type 1 Waardenburg, but without dystopia canthorum.

1. A 14 year-old patient is referred to you for evaluation of unilateral hearing loss. The patient undergoes audiometric testing. Which of the following statements is true regarding masking?
2. **The risk of crossover is much lower with insert earphones, often precluding the need for masking**
3. Masking should be performed whenever there is a > 20 dB air-bone gap in the test ear
4. Interaural attenuation with bone conduction begins at 10 dB
5. Masking should be performed whenever there is a > 10 dB difference between the test ear air conduction threshold and non-test ear air conduction threshold with the use of circumaural earphones.

**Difficulty Level: Hard**

**Explanation:** Crossover is a phenomenon in which a sound stimulus presented to the test ear directly stimulates the contralateral (non-test) ear. Because the sound has to travel through the skull, some degree of interaural attenuation can occur. With standard circumaural earphones, interaural attenuation is approximately 40 dB. Insert earphones have much lower likelihood of crossover, and a stimulus up to 70 dB can be presented to the test ear without masking. Thus, masking should be instituted whenever the air-conduction threshold in the test ear exceeds the non-test ear by >40 dB for circumaural earphones or >70 dB for insert earphones.

The threshold for the crossover phenomenon is significantly higher with insert earphones compared to circumaural earphones.

1. A patient who sustained a temporal bone fracture has a Sunderland class V injury 1 mm distal to the nerve to stapedius. Which of the following would you expect the patient to have?
2. Loss of lacrimation
3. Presence of hyperacusis
4. Loss of parasympathetic innervation of the nasal mucosa
5. **Decreased taste sensation**

**Difficulty Level: Moderate**

**Explanation:** The Sunderland classification is as follows:

Grade 1: Neuropraxia

• Conduction disruption with intact axon and preserved supportive structures.

Grade 2: Axonotmesis

• disruption of the myelin sheath and axon The epineurium and perineurium remain intact. Grade

3: Neurotmesis with preservation of perineurium

• Endoneurium is disrupted. Grade

4: Neurotmesis with preservation of epineurium Grade

5: Neurotmesis with complete transection of nerve trunk Grade

IV and V injuries require nerve grafting.

The patient in this vignette sustained a complete transection of the nerve distal to the nerve of stapedius. This would obviously involve loss of all nerve function distal to the injury, which would include the chorda tympani and the extratemporal branches of the facial nerve. Disruption of the fibers of the chorda tympani nerve would result in loss of taste sensation of the ipsilateral anterior 2/3 of the tongue.

Disruption of the facial nerve just distal to the nerve to stapedius would result in dysfunction of the chorda tympani nerve and extratemporal facial nerve branches.

1. The footplate of the stapes is developmentally derived from?
2. First branchial arch
3. Second branchial arch
4. Third branchial arch
5. Fourth branchial arch
6. **Otic mesenchyme**

**Difficulty Level: Moderate**

**Explanation:** The stapes footplate arises from otic mesenchyme.

The stapes superstructure is derived from the second branchial arch, but the footplate arises from the otic mesenchyme.

1. Which of the following is LEAST likely to be a feature of Pendred syndrome?
2. Thyroid goiter
3. Bilateral dilation of vestibular aqueduct
4. Defective ion transport
5. **Conductive hearing loss**
6. Mondini malformation

**Difficulty Level: Moderate**

**Explanation:** Pendred syndrome usually results in a sensorineural or mixed hearing loss, but not a purely conductive loss. The conductive component usually occurs if an enlarged vestibular aqueduct is present, which can cause a third window effect.

Thyroid goiter, Mondini malformations and sensorineural hearing loss are common features of Pendred syndrome. Mixed hearing loss can occur with Pendred syndrome due to the third window effect of an enlarged vestibular aqueduct.

1. While performing stapes surgery for the treatment of otosclerosis, you encounter a persistent stapedial artery occupying the majority of the obturator foramen of the stapes. What is the most appropriate step in management?
2. Proceed with laser coagulation of the artery and then continue with the surgery as planned.
3. Mobilize and displace the artery sufficiently to expose the footplate and attempt the surgical procedure.
4. **Abort the procedure and recommend amplification.**
5. Proceed with arterial embolization.
6. Perform a trans-cervical ligation of the external carotid artery and proceed with the surgery.

**Difficulty Level: Easy**

**Explanation:** A persistent stapedial artery is a rare finding, seen usually on only 1 in 5000 cases. If this artery is mostly a small vestigial remnant, it is appropriate to proceed with laser or bipolar cauterization and subsequent procedure completion. A larger vessel occupying less than 50% of the obturator space could allow for sufficient room for prosthesis placement in experienced hands. A large persistent stapedial artery occupying the majority of the obturator foramen cannot be safely cauterized with laser or bipolar cautery and would necessitate procedure abortion.

A persistent stapedial artery occupying the majority of the obturator foramen is best left alone. Smaller vessels may be amenable to bipolar or laser cauterization.

1. A 65 year old male with a history of diabetes type II presents to the ENT clinic with 1 week of right ear pain and otorrhea. On physical exam the ear canal is edematous and erythematous. After debriding the right external auditory canal the tympanic membrane is intact with a middle ear effusion. The patient is sent home with neomycin otic drops. The patient returns to clinic 1 week later with a maculopapular rash involving the right conchal bowl and right external auditory canal. The right canal is significantly less edematous and erythematous. What is the next best step in management?
2. Continue the neomycin otic drops and add oral ciprofloxacin
3. Admit to hospital for IV anti- pseudomonal antibiotics and blood glucose control
4. **Stop the neomycin drops and start a corticosteroid otic drop**
5. Continue neomycin otic drops and have patient follow up in 1 week
6. MRI to evaluate for necrotizing otitis externa

**Difficulty Level: Easy**

**Explanation:** The patient has an right otitis externa which is diagnosed by history and physical exam alone. The most common pathogen causing otitis externa is Pseudomonas aeruginosa. Neomycin is an appriopriate topical agent to treat otitis externa. When the patient returns with a maculopapular rash and near resolution of the otitis externa the patient is having an allergic reaction to the topical neomycin. The most appropriate management is to stop the neomycin otic drops and start a corticosteroid otic drop.

Corticosteroid otic drops are indicated in the treatment of otic allergic reactions.

1. The superior olivary nucleus is the primary generator of which wave(s) in an auditory brainstem response (ABR)?
2. I, II
3. II, III
4. **III, IV**
5. IV, V
6. V, VI

**Difficulty Level: Hard**

**Explanation:** Auditory brainstem response (ABR) testing functions by using electrodes placed on the vertex and mastoid to detect electrical signal changes in response to unilateral auditory ‘clicks’. These clicks are administered as 100 microsecond bursts of sound that excite the neurons along the auditory path. Up to 7 waves correlating to different intracranial neurologic structures are produced at 1ms intervals after the click is administered. Waves I-VII have been correlated with the following neurologic structures:

I: Distal eighth nerve (vestibulocochlear nerve)

II: Proximal eighth nerve/cochlear nucleus

III: Cochlear nucleus/superior olivary complex

IV: Superior olivary complex V: Lateral lemniscus

VI, VII: Inferior colliculus

However, ABR waves are much more complex and nuanced than this. The early waves (waves I and II), have single generator sites, however later waves often have multiple generator sites and may not exactly correspond with a single location.

ABR testing is primarily used to either elucidate audiometric thresholds or for otoneurologic applications. Earlier than age 2 the five waves within an ABR are not fully formed and are less reliable. Typically, waves I, III, and V are present at birth with an increased I-V time.

A useful mnemonic to help aid in the interpretation of waveforms in the auditory brainstem response is E COLI: I Distal

1. A 53 year-old male presents to your clinic for evaluation of chronic unilateral otorrhea, hearing loss, and occasional vertigo for approximately 2 years. The patient denies prior ear surgery. On examination an attic retraction pocket cholesteatoma is noted. CT-scan reveals scutum erosion and thinning of the lateral semicircular canal wall. The patient is adamant about not undergoing more than one surgery and would like his problem addressed fully upon the first surgery. During the procedure, cholesteaoma matrix is identified over the lateral semicircular canal with an underlying bluish coloration. Which of the following procedures would be the most appropriate in this patient?
2. Atticotomy.
3. Cortical (simple) mastoidectomy.
4. Modified radical mastoidectomy (Bondy procedure).
5. **Canal-wall down mastoidectomy.**
6. Radical mastoidectomy.

**Difficulty Level: Easy**

**Explanation:** This patient has evidence of attic retraction cholesteatoma with erosion of the lateral semicircular canal. Because of the bluish coloration underlying the cholesteatoma matrix over the lateral semicircular canal, this matrix should not be removed, as it can expose an underlying fistula and risk sensorineural hearing loss. As a result, this focal disease should be left behind, which subsequently warrants a 6-month second-look procedure or a canal wall down mastoidectomy. This patient has stated that he does not want to undergo multiple surgeries. As a result, the best option would be to perform a canal wall-down mastoidectomy (CWD), occasionally referred to (incorrectly) as a modified radical mastoidectomy. A CWD procedure removes the posterior canal wall, which allows for future debridement of any cholesteatoma as well as monitoring of disease and potentially avoids the need for a second-look surgery.

Canal wall down mastoidectomies obviate the need for second look procedures, though they do require periodic mastoid bowl debridements.

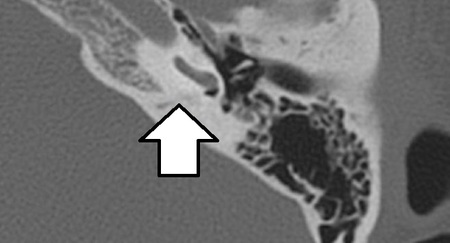
1. Besides the internal acoustic meatus, which of the following is the narrowest segment of the fallopian canal?
2. Meatal
3. Labyrinthine
4. Tympanic
5. Vertical

**Difficulty Level: Easy**

**Explanation:** The labyrinthine segment is the most narrow as well as the shortest. It contains the geniculate ganglion. This area is vulnerable to the effects of edema and nerve compression in cases of trauma or inflammatory disorders.

The labyrinthine portion is the most narrow segment of the fallopian canal. The facial nerve is particularly vulnerable to nerve compression in this area due to the small, bony confines of the canal at this location.

1. The structure identified in the image below is responsible for which of the following actions?



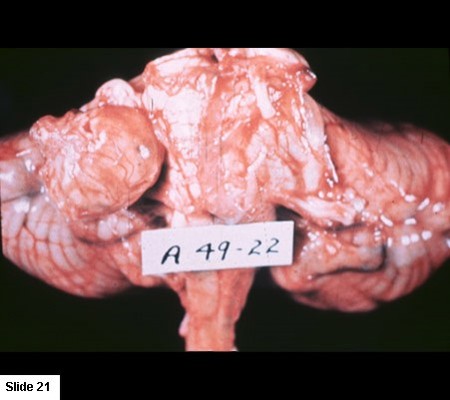
1. **Connecting the perilymph of the cochlea with the subarachnoid space**
2. Regulating the volume and pressure of endolymphatic fluid
3. Marking the bony interface between the petrous and mastoid portions of the temporal bone
4. Transmitting the vestibulcochlear and facial nerves through the temporal bone
5. Providing arterial blood supply to the cochlea

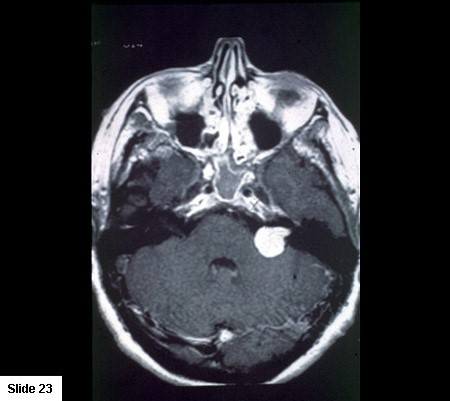
**Difficulty Level: Moderate**

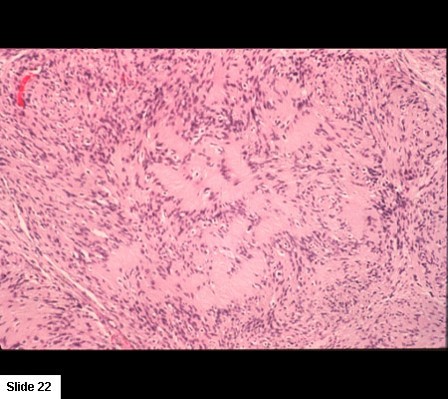
**Explanation:** The structure identified above is the cochlear aqueduct which serves to connect the perilymphatic space of the cochlea with the subarachnoid space. The cochlear aqueduct typically runs parallel to the internal auditory canal.

The cochlear aqueduct runs from the perilymphatic space of the cochlea to the subarachnoid space. Unlike the vestibular aqueduct, the cochlear aqueduct is parallel to the internal auditory canal.

1. Which nerve or nerves does the tumor depicted below most commonly originate from?







1. Facial nerve
2. Cochlear nerve
3. Vestibular nerve
4. Singular nerve
5. Vestibular and cochlear nerves equally

**Difficulty Level: Easy**

**Explanation:** The most common site of origin of acoustic neuromas is both the superior and inferior divisions of the vestibular nerve. The frequency of involvement of these two divisions is approximately equal with some studies citing slightly higher rates in the superior division. Acoustic neuromas, or vestibular schwannomas, commonly envelope the facial and cochlear nerves but do not often arise from them.

Vestibular schwannomas most commonly arise from the vestibular nerves.

1. A 25 year-old male is brought to the emergency room for evaluation following cranial trauma due to a motorcycle accident this morning. The patient suffered impact trauma to the occiput as he fell backwards trying to “pull a wheelie” without wearing a helmet. The patient admits to loss of consciousness and memory loss as well as sudden right hearing loss and intense vertigo. On examination he has a House-Brackmann III right facial nerve paralysis, a right hemotympanum, and a right external auditory canal laceration. Which of the following statements is correct?
2. Transverse temporal bone fracture is most likely to occur after a blow to the temporal parietal bone
3. Given his described injuries, he should be unable to completely close his eye with maximal effort
4. The most likely facial nerve region injured is the mastoid segment
5. **Facial paralysis is the complication of temporal bone trauma which is most likely to dictate the need for early surgical intervention**
6. In temporal bone trauma, hearing loss when associated with tinnitus is associated with a decreased likelihood of hearing recovery

**Difficulty Level: Moderate**

**Explanation:** Each temporal bone is divided into five components: Squamous, tympanic, styloid, mastoid, and petrous. The squamous portion lies anterior and superior in relation to the remainder of the temporal bone. The zygomatic process arises from the squamous part and connects the temporal bone to the zygomatic bone of the face through the zygomatic arch. It also provides the superior articulation of the temporomandibular joint.

Compared to all other complications of temporal bone trauma, complaints of facial paresis or paralysis and facial asymmetry are indicative of injury cranial nerve VII and are most likely to dictate the need for early surgical intervention. Decompression may be warranted in instances where imaging indicates a severely disrupted or displaced nerve and/or testing demonstrates severe dysfunction. It is important to note that facial nerve dysfunction can worsen over time, and so even patients who initially present with fairly symmetric facies should have repeat examinations in the acute phase to ensure a progression of dysfunction is not missed.

Facial paralysis or paresis is the complication of temporal bone trauma which is most likely to dictate the need for early surgical intervention.

1. What is the loss of gain resulting from impedance mismatching between the air in the ear canal and fluid in the cochlea that must be overcome by the mechanism of the tympanic membrane and ossicles?
2. 5 dB
3. 10 dB
4. 15 dB
5. **30 dB**
6. 50 dB

**Difficulty Level: Hard**

**Explanation:** Ossicular coupling is the mechanism of transmission of sound pressure through the tympanic membrane and ossicular chain via the oval window. There is a natural state of impedance mismatch between the air in the ear canal (low impedance) and the fluid within the cochlea (high impedance) and results in a loss of gain in the realm of 30 dB. The coupling effect of the tympanic membrane and ossicles overcomes this loss. The ratio of the surface area of the tympanic membrane to the oval window accounts for about 26 dB of this gain. The malleoincudal lever effect results in an additional 2 dB of gain. The final factor is called the catenary lever effect that is generated by the elastic stretching of the tympanic membrane from the annulus to the malleus manubrium resulting in another 6 dB in gain.

The loss of gain resulting from impedance mismatching between the air in the ear canal and fluid in the cochlea is about 30 dB. The three mechanisms for impedance matching are the ratio of the surface area of the tympanic membrane to the oval window, malleoincudal lever and tympanic membrane buckling.

1. A 58-year-old female presents with complaints of facial asymmetry. On physical exam she is found to have a complete left facial nerve paralysis (House Brackman 6/6). Magnetic resonant imaging confirms the presence of a cerebello-pontine angle tumor. Given the sensory innervation of the ear, where would the patient likely be insensate?
2. Mastoid skin
3. Anterior osseous external auditory canal skin
4. Tragal skin
5. Floor of the external auditory canal skin
6. **Lateral concha**

**Difficulty Level: Hard**

**Explanation:** The ear has very complex sensory innervation from multiple nerves, and there is often some degree of overlap. The lateral concha, part of the posterior external auditory canal/meatus and tympanic membrane are supplied by the facial nerve. This is specifically supplied by the nervus intermedius portion which also carries the parasympathetic portions of VII.

The lateral concha, part of the posterior external auditory canal/meatus and tympanic membrane are supplied by the facial nerve

1. Which of the following is true regarding the genetics of hearing loss?
2. Pendred’s syndrome is linked to a COL2A1 gene mutation
3. Neurofibromatosis 2 (NF2) is more common than neurofibromatosis 1 (NF1)
4. **Autosomal recessive hereditary hearing impairment is usually more severe than autosomal dominant hereditary hearing impairment**
5. The majority of hereditary hearing loss is inherited in an autosomal dominant fashion

**Difficulty Level: Hard**

In autosomal recessive hereditary hearing impairment, hearing loss is usually profound and prelingual in onset. In contrast, autosomal dominant hereditary hearing impairment is usually post lingual, less severe and progresses with age.

1. Which of the following is a contraindication to stapes surgery for otosclerosis?
2. History of frequent middle ear infection.
3. History of benign paroxysmal positional vertigo
4. An air-bone gap less than 25 dB across 3 frequencies
5. Mixed conductive/sensorineural hearing loss
6. **Profound sensorineural hearing loss in the contralateral ear**

**Difficulty Level: Moderate**

**Explanation:** Because of the risk of profound sensorineural hearing loss with stapes surgery, it is contraindicated in any patient where the surgical ear is the only-hearing ear. Exceptions to this would be in a patient with severe hearing loss where there is minimal benefit from amplification.

Conditions that represent a contraindication to stapes surgery include the presence of Meniere disease, active vertigo or balance problems, proposed surgery is in the only hearing or the better hearing ear, active middle or external ear infection.

1. A 64 year-old-male presented to your office for evaluation of unilateral hearing loss. An audiogram demonstrated mild to moderate sensorineural hearing loss on the right and moderate to severe sensorineural hearing loss on the left. Speech discrimination was 80% on the left. An MRI demonstrates a 1.2 cm intracanalicular vestibular schwannoma. He has no personal or family history suggestive of neurofibromatosis type 2. After discussing his diagnosis and treatment options, he inquires about his chances of being able to benefit from hearing aids over the next several years. Which of the following is the strongest predictor of long-term serviceable hearing in patients with vestibular schwannomas?
2. Location
3. **Speech discrimination**
4. Patient age
5. Size
6. Rate of growth

**Difficulty Level: Moderate**

**Explanation:** Multiple studies have demonstrated that speech discrimination at presentation strongly predicts long-term serviceable hearing. For VS patients with 100% speech discrimination, the prognosis for serviceable hearing even as long as 10 years following diagnosis is excellent. However, for patients with small reductions in speech discrimination at the time of presentation, the prognosis for long-term serviceable hearing is significantly worse.

Speech discrimination score at presentation is a strong predictor of long-term serviceable hearing in patients with vestibular schwannoma.

1. While performing a tympanoplasty to address a 40 % posterior perforation on a 21 year old male with chronic eustachian tube dysfunction it is discovered that the ossicles are fixated due to diffuse tympanosclerosis. The incus is not in continuity with the stapes and the lenticular process is eroded. After removal of tympanosclerosis, the stapes remains immobile. What is the most appropriate decision in the management of this situation?
2. Reattach incus to stapes and place graft for repair of tympanic membrane
3. **Remove incus, place graft for repair of tympanic membrane, and return for second stage stapedectomy and ossiculoplasty**
4. Perform stapedectomy and ossiculoplasty, and return for second stage tympanoplasty
5. Perform stapedectomy, and return for second stage tympanoplasty with ossiculoplasty
6. Perform stapedectomy and ossiculoplasty, and place graft for repair of tympanic membrane

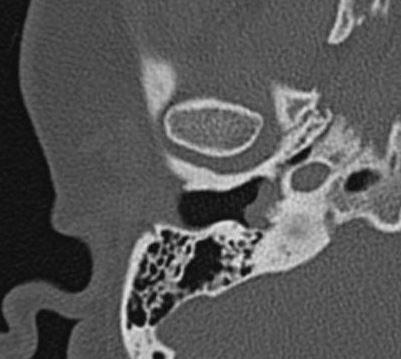
**Difficulty Level: Moderate**

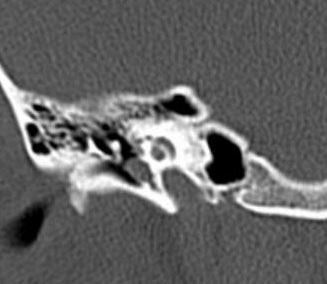
**Explanation:** In this case, the patient has an unstable middle ear space that requires repair of a large perforation. With diffuse tympanosclerosis, a fixed stapes, and a discontinuous ossicular chain, he will eventually need a stapedectomy and ossiculoplasty. It is not appropriate to attempt to reattach the incus to the stapes as the incus is nonviable and the stapes is fixed; if this were to occur, he would still have a conductive hearing loss. A stable middle ear space should be established in this case before proceeding with stapedectomy and ossiculoplasty. Stapedectomy should not be performed in a potentially infected ear with poor Eustachian tube dysfunction due to risk of labyrinthitis and profound hearing loss.

Tympanic membrane perforation is a contraindication to stapedectomy as an intact tympanic membrane would prevent some pathogens from entering the vestibule and reduce the risk for labyrinthitis and/or sensorineural hearing loss post operatively. Repair of the tympanic membrane should occur prior to stapedectomy, and then a stapedectomy with ossiculoplasty can proceed once the repair has demonstrated success.

1. A 57-year-old male with no significant past medical history presents to the clinic for evaluation of right-sided pulsatile tinnitus that has been present for 6 months. An audiogram shows normal hearing on the left and a moderate conductive hearing loss on the right. Otoscopy on the right is shown in the picture below. CT images are also shown below. What is the most reasonable next step in management of his ear mass?







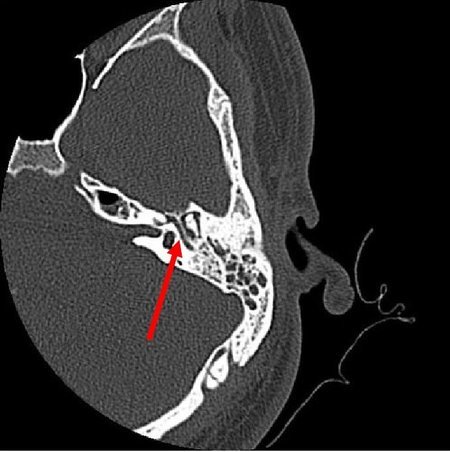
1. PET scan
2. Biopsy of middle ear mass
3. Gallium-68 Dotatate scan
4. **MRI**
5. Ciprodex and dry ear precautions

**Difficulty Level: Moderate**

**Explanation:** Tympanic paraganglioma tumors (glomus tympanicum) arise from the glomus body of the promontory along Jacobson's nerve. These tumors are typically confined to the mesotympanum. In this case, the diagnosis of a tympanic paraganglioma has been made clinically, and the next step is further imaging with MRI, which provides superior soft tissue and vascular resolution. The Endocrine Society guidelines recommend MRI to manage paragangliomas of the head and neck. Subsequent management may include observation, stereotactic radiosurgery, transcanal or endoscopic middle ear exploration with removal of the tumor. The extent of resection should be tailored to the patient, as leaving a limited adherent tumor remnant on the facial nerve or carotid artery should be considered with the advanced infiltrative disease to prevent unnecessary morbidity. Remnant tumors or small tumors can also be treated with radiation therapy. Microsurgery or endoscopic surgery is often preferred due to high success rates with minimal morbidity.

Tympanic paragangliomas are of neuroendocrine origin that are highly vascular. It is common to obtain multiple imaging modalities, including CT, CTA, MRI, MRA, and angiography, to determine the extent of the neoplasm before resection.

1. A 27-year-old male with a long-standing history of Eustachian tube dysfunction has received multiple sets of pressure equalization tubes. He has recently been having foul smelling otorrhea from his left and a white mass is visualized behind the tympanic membrane. The patient is scheduled for surgical intervention. Based on the computed tomography below, if the indicated structure were damaged in surgery, what would be the likely outcome?



1. **Facial paralysis**
2. Conductive hearing loss
3. Sensorineural hearing loss
4. Vertigo
5. No expected consequences

**Difficulty Level: Easy**

**Explanation:** This patient has signs and symptoms most consistent with a cholesteatoma. Part of the workup for a cholesteatoma involves obtaining a computed tomography of the temporal bone without contrast. This is an axial, non-contrast computed tomography of the left temporal bone. The indicated structure is the tympanic segment of the facial nerve. If it were damaged, this would lead to facial paralysis. Other structures visible on this slice include the malleus and incus (the white structure that appears like an ice cream cone up and to the right of the arrow) which if they were damaged would lead to a conductive hearing loss. The beginning of the semicircular canals are seen up and to the left of the arrowhead. Not visible on this slice but the most commonly damaged portion of the vestibular system is the lateral semicircular canal which if it were damaged would lead to vertigo. Damage to the cochlea would lead to a sensorineural hearing loss. The ear has many important structures within a narrow area and damage to any of them may have significant consequences, which is why the patient should be consented of all of the risks and benefits prior to surgical intervention.

The structure indicated in this CT is the tympanic segment of the facial nerve.

1. Concerning management of malignant otitis externa, which of the following is true?
2. Antibiotic duration is often 2-4 weeks
3. **An elevated CRP is the most characteristic laboratory abnormality and is a useful way of monitoring disease activity**
4. Oral ciprofloxacin has not been shown to be successful as a single agent
5. Gallium citrate (Ga-67) scanning is the best test for the assessment of bone erosion
6. Adjuvant hyperbaric oxygen therapy has not been shown to be successful in advance and refractory cases

**Difficulty Level: Hard**

Although nonspecific, an elevated ESR or CRP is the most characteristic laboratory abnormality and is a useful way of monitoring disease activity. Other laboratory parameters are generally normal.

1. A 45 year old female presents with gradual hearing loss over the last 5 years. An ophthalmologist recently noted that her visual acuity difficulties were due to interstitial keratitis. What is the most likely diagnosis?
2. **Cogan syndrome**
3. Usher syndrome
4. Meniere’s disease
5. Vogt Koyanagi Harada syndrome
6. Presbycusis

**Difficulty Level: Easy**

**Explanation:** Cogan syndrome is an autoimmune condition leading to fluctuating sensorineural hearing loss, peripheral vestibulopathy and interstitial keratitis of the eyes. Systemic symptoms may also be present and typically involve the cardiopulmonary system. Initial treatment is with corticosteroid, and maintenance therapy typically consists of azathioprine or cyclophosphamide.

Cogan syndrome is an autoimmune condition leading to fluctuating sensorineural hearing loss, peripheral vestibulopathy and interstitial keratitis of the eyes.

1. Which of the following statements is true regarding the lateral graft tympanoplasty technique?
2. The graft is placed lateral to the fibrous layer of the tympanic membrane and the malleus handle
3. Lateral graft technique does not require complete removal of the squamous epithelium of the tympanic membrane
4. The trimmed anterior canal skin should cover 5 to 6 mm of the anterior end of the graft
5. An advantage of the lateral graft technique compared to the medial graft technique is shorter healing time
6. **The risk of iatrogenic cholesteatoma after lateral graft technique is higher than the medial graft technique**

**Difficulty Level: Hard**

**Explanation:** The lateral grafting technique is less often employed compared to the medial grafting technique, however is useful for very large perforations and complicated revision cases. Disadvantages of the lateral grafting technique for tympanoplasty include post operative blunting, increased risk of cholesteatoma, greater technical demands, longer healing times, and lateralization. The risk of iatrogenic cholesteatoma after lateral graft technique is higher than the medial graft technique due to incomplete squamous epithelium removal. This highlights the importance of meticulous removal of any remaining epithelium, and may necessitate the use of bone currettes or small diamond burs (aside from their use during the canalplasty portion of the procedure) to removed any trapped epithelium.

Disadvantages of the lateral grafting technique for tympanoplasty include post operative blunting, increased risk of cholesteatoma, greater technical demands, longer healing times, and graft lateralization.

1. A 28-year-old female presents with progressive hearing loss over the past year. After a complete history and physical, the patient is diagnosed with otosclerosis. The patient is not interested in surgery at this time and asks if there are any non-surgical treatments. Which of the following supplements has demonstrated efficacy in reversing otosclerosis?
2. Zinc
3. Magnesium
4. Fluoride
5. Vitamin B12
6. **None of the above**

**Difficulty Level: Moderate**

**Explanation:** None of the above answer choices have been shown to reverse otosclerosis.

There is some evidence that suggests fluoride treatment slows the progression of otosclerosis, but no medical management has demonstrated efficacy in reversing the pathology.

1. A 75-year-old male presents a three-day history of worsening ear pain and vesicular eruptions associated with progressive facial weakness. He had chicken pox when he was five years old. He has a history of hypertension and hyperlipidemia and is borderline diabetic. On examination, he has multiple vesicular lesions and shallow ulcerations of the left ear canal. He has mild weakness of the left forehead, and his smile is asymmetric. He can fully close his eyes with mild effort, and he can purse his lips. Which statement below about his condition is most accurate?
2. 20% of patients will demonstrate a House-Brackmann score IV-VI
3. **The prognosis for recovery from Ramsay-Hunt-associated facial paralysis is worse than that of idiopathic Bell palsy**
4. Only the motor divisions of CN VII can be affected
5. Decompression of the facial nerve in the labyrinthine segment is helpful in improving House-Brackmann score
6. Standard treatment is Valacyclovir and prednisone for 5 days

**Difficulty Level: Moderate**

**Explanation:** This patient has Ramsay-Hunt syndrome, an infection caused by varicella zoster virus leading to vesicular eruptions in the conchal bowl and rapidly progressive facial palsy that is House-Brackmann score IV-VI in 50% of patients. The prognosis for facial paralysis recovery is poorer than that of idiopathic Bell palsy.

Patients with Ramsay-Hunt syndrome have a worse prognosis for recovery of facial paralysis compared to patients with idiopathic Bell's palsy, and surgical decompression options are more limited in efficacy.

1. A 57 year old male with a neurofibromatosis type 2 is 24 months status post left sided microsurgical resection of a vestibular schwannoma via a retrosigmoid approach. The patient is found to have non serviceable hearing on the side of resection. The patient had non serviceable on the opposite side prior to the resection. A recent MRI confirms the patient to have an intact cochlear nerve and no residual disease. What is the patient'™s best option for optimal audiometric performance?
2. Hearing aid
3. Auditory brainstem implantation
4. **Cochlear implantation**
5. BAHA
6. Patient will receive no audiometric benefit from any of the above

**Difficulty Level: Moderate**

**Explanation:** Cochlear implantation is an alternative to auditory brainstem implantation in patients with neurofibromatosis type 2. Approximately 70% of patients receive open set speech discrimination and score at the ceiling of audiometric testing. Cochlear implantation should be considered for NF2 patients with non serviceable hearing and an intact cochlear nerve. Auditory brainstem implantation is an option for those NF2 patients with non serviceable hearing and cochlear nerve loss.

Cochlear implantation should be considered for NF2 patients with non serviceable hearing and an intact cochlear nerve.

1. A 43-year-old gentleman presents with complaints of ear pain and drainage and retrobulbar pain. On physical exam, you note purulent fluid in the middle ear and an abducens nerve palsy. These symptoms indicate an infection in which location?
2. Cavernous sinus
3. Infratemporal fossa
4. Posterior cranial fossa
5. Mastoid
6. **Petrous apex of the temporal bone**

**Difficulty Level: Easy**

**Explanation:** This is the presentation of Gradenigo’s syndrome (petrous apicitis). This is often a complication of a middle ear infection, and so patients usually have symptoms of pain and drainage from the ear. Symptoms can be explained by the location of the petrous apex. Irritation of the trigeminal in Meckel’s cave explains the retro-orbital pain. The VI nerve (abducens) travels through Dorello’s canal, which also abuts the petrous apex, explaining the palsy of this nerve. The signs and symptoms of Gradenigo's syndrome can easily be remembered by the mnemonic "EAR:"

Gradenigo's syndrome is characterized by otorrhea, retrofacial pain, and abducens nerve palsy. It is caused by an infection at the petrous apex.

1. What is the most common complication of myringotomy tube insertion?
2. Granulation tissue
3. Persistent perforation
4. Cholesteatoma
5. Obstructed myringotomy tube
6. **Transient otorrhea**

**Difficulty Level: Easy**

**Explanation:** Transient otorrhea occurs in 16%. It is important that this be included as part of the pre-operative discussion and informed consent process, even though it is typically a benign process. It is also worth counseling parents that otorrhea does not necessarily mean an infection is present or that the child needs antibiotics.

Otorrhea is a common "complication" of tympanostomy tube insertion.

1. An 18 month child presents with acute otitis media and acute onset right sided facial weakness. What is the most appropriate management?
2. Admit for IV antibiotics and perform urgent mastoidectomy
3. **Admit for IV antibiotics and perform urgent myringotomy tube**
4. Treat as outpatient with PO antibiotics and scheduled myringotomy tube in 48-72 hours
5. Treat as an outpatient with PO antibiotics and close observation
6. Admit for IV antibiotics and IV steroids

**Difficulty Level: Moderate**

**Explanation:** IV antibiotics are necessary. Urgent myringotomy tube should be placed to allow the suppurative and toxic effects of the infection a route to drain.

Cases of complicated acute otitis media can generally be managed with IV antibiotics and tympanostomy tubes.

1. A 20 year-old male presents for evaluation of left-sided non-pulsatile tinnitus for 6 months. Ear examination is unremarkable. Audiologic testing reveals a notched moderate high-frequency sensorineural hearing loss on the left at 4 KHz. Further testing shows that the speech reception threshold (SRT) is 30 dB. During testing of the patient's speech discrimination score (SDS), speech stimulus is presented at 40 dB above the patient's SRT. Which of the following statements is correct?
2. The stimulus was presented at 40 dB sound pressure level (SPL).
3. The stimulus was presented at 40 dB intensity level (IL).
4. **The stimulus was presented at 70 dB hearing level (HL).**
5. The stimulus was presented at 70 dB sensation level (SL).
6. None of the above

**Difficulty Level: Moderate**

**Explanation:** The patient has a speech reception threshold of 30 dB HL. The speech reception threshold (SRT) is the lowest decibel level at which a patient can correctly repeat 50% of test words. The SDS was tested using a stimulus 40 dB SL or 70 dB HL.

Sound pressure level (SPL) is a logarithmic measure of the effective pressure of a sound relative to a reference value. It is measured in micropascals. Intensity level (IL) is the sound power per unit area,, usually measured in the air at a listener's location and expressed in watts/cm2. SPL and IL units do not lend themselves to routine utilization in audiometry.

Sensation level (SL) is the number of dB above a person's threshold.

1. A 33-year-old female is referred to your clinic for dizziness. She notes that her symptoms began yesterday and have not subsided today. She describes her dizziness “as if the world is spinning.” She has never had anything like this before. Her symptoms do not improve while she lays flat. She denies any changes in her hearing or aural fullness. What is the most likely diagnosis?
2. **Vestibular neuritis**
3. Labyrinthitis
4. Endolymphatic hydrops
5. Benign paroxysmal positional vertigo
6. Migraine variant vertigo

**Difficulty Level: Easy**

**Explanation:** Key points for determining the etiology of vertigo include the timing and associated symptoms. In this patient, she has never had anything like this before, there is a fairly acute onset of severe vertigo, it is not relieved by staying still, and there are no other neurological symptoms, including hearing loss. The exact etiology of vestibular neuritis has been debated, and most practitioners believe it is either a small stroke of the vessels supplying the vestibular nerve (the superior division in particular), or viral induced inflammation of the nerve. Exam findings supportive of a diagnosis of vestibular neuritis include horizontal rotatory nystagmus with a fast phase to the unaffected ear, nystagmus that improves with fixation, and catch up saccades with head thrust testing to the affected ear. Imaging may not be necessary if the history is highly suggestive of vestibular neuritis, but if other symptoms are present or if the patient is unable to stand without assistance, imaging is typically warranted. Treatment is supportive with antiemetics and hydration. Antivirals and steroids have also been employed with varying reported successes. After the acute phase, patients will often experience chronic imbalance and BPPV-like symptoms. This highlights the importance of vestibular rehabilitation and need for early exercise/return to function in these patients.

Vestibular neuritis typically presents as acute onset and often severe rotatory vertigo that does not improve with positional changes, and is not associated with other symptoms.

1. A 3-year-old male patient is evalulated for a cerebellopontine angle (CPA) tumor seen on a recent head CT obtained for head trauma. He does not have any facial weakness, hearing loss, or balance issues.
2. **Vestibular schwannoma**
3. Epidermoid
4. Glioma
5. Meningioma
6. Ependymoma

**Difficulty Level: Hard**

**Explanation:** Cerebellopontine angle tumors are very rare in young children. Extra-axial tumors arising out of the CPA include vestibular schwannomas, meningiomas and epidermoids, with vestibular schwannomas being the most common among these.

Exophytic intra-axial tumors like ependymomas or gliomas (pilocytic astrocytoma, ganglioglioma) are also encountered relatively frequently, depending on the inclusion criteria of the study.

1. A 32 year-old male presents to you for a 6-month follow-up regarding a prior history of left tympanic membrane rupture following a workplace accident 1 year ago. The patient had demonstrated a left moderate conductive hearing loss at the time of initial evaluation and a small central tympanic membrane perforation. On today'™s visit, the patient reports a persistent left hearing loss. Examination reveals a well-healed tympanic membrane on the left. Pure tone audiometry shows a moderate flat left sensorineural hearing loss with a left pure tone average of 50 dB. The speech reception threshold (SRT) is 30 dB on the left and 10 dB on the right. The speech discrimination score (SDS) is 50% at 60 dB HL on the left and 100% at 40 dB HL on the right. Tympanometry reveals bilateral type A curves. Bilateral acoustic reflexes are present at 75 dB HL. Which is the next most appropriate diagnostic test?
2. Otoacoustic Emissions (OAE).
3. Electrocochleography.
4. **Stenger test.**
5. MRI brainstem.

**Difficulty Level: Moderate**

**Explanation:** This patient's presentation and audiologic testing results are suspicious for pseudohypoacusis. To begin, the patient demonstrates a new-onset moderate SNHL while he had a pure conductive loss on the previous evaluation, which represents a significant change in type of hearing loss. Next, the patient's left-sided PTA (50 dB) differs significantly from his SRT (30 dB) and his SDS score of 50% at 60 dB HL (30 dB SL) is unexpectedly low. Finally, bilateral acoustic reflexes are present at 75 dB (only 25 dB SL on the left compared to PTA). The patient's normal examination and tympanometry also raise suspicion for a non-organic cause. The ideal next diagnostic step would be performing the Stenger test given its informative applicability in this case and low-cost. The Stenger test takes advantage of the fact that all individuals, when presented with a binaural sound stimulus with asymmetric loudness, will experience the sound as originating from the side receiving the loudest perceived stimulus. A Stenger test for this individual would present a stimulus of 45 dB on the left (5 dB below threshold for the bad ear) and 10 dB on the right (5 dB above threshold for the good ear). Normally, a person with a hearing loss similar to this patient's would report hearing a sound stimulus on the good ear only (unable to hear the sound on the bad ear). A person with pseudohypoacusis, in contrast, would not respond because, being able to hear on both sides, the sound would present itself as being louder on the bad ear (45 dB on the left, 10 dB on the right).

Stenger and ABR testing can be used to assess for malingering/pseudohyperacusis.

1. A patient seen below presents with the noted ear abnormality. Which of the following is an absolute contraindication to repair of ear canal atresia?



1. Laterally displaced CN VII
2. Thick atretic plate
3. Ossicular abnormalities
4. **Malformed inner ear with abnormal sensorineural hearing**
5. Presence of canal cholesteatoma

**Difficulty Level: Hard**

**Explanation:** If there is a malformed inner ear noted on pre-operative imaging with sensorineural hearing loss on audiometry then repair of the canal atresia would be futile as this patient may not have serviceable hearing.

A malformed inner ear with abnormal sensorineural hearing is a contraindication to atresiaplasy, though repair of the microtia may proceed (for cosmetic purposes).

1. A 7 year old is found to have a white mass behind the tympanic membrane on the left side during routine physical exam. Audiogram confirms a conductive hearing loss in the left ear. He has no history of previous ear surgery and you do not appreciate any retraction of the tympanic membrane. What is your diagnosis?
2. **Congenital cholesteatoma**
3. Primary acquired cholesteatoma
4. Secondary acquired cholesteatoma
5. Glomus tympanicum
6. Tympanosclerosis

**Difficulty Level: Easy**

**Explanation:** In general, there are 3 types of cholesteatoma: Congenital, primary acquired, and secondary acquired. A white mass behind the tympanic membrane in an otherwise asymptomatic child is most consistent with a congenital cholesteatoma. The exact etiology remains controversial (ectoderm formation from the 1st pharyngeal cleft vs epithelial migration through a patent eustachian tube early in development) but if it grows to a large enough size it can lead to a conductive hearing loss.

A congenital cholesteatoma is thought to be the result of epithelium trapped in the middle ear space early in embryogenesis.

1. What is Brown's sign?
2. Sudden or fluctuating hearing loss accompanied by dizziness or vertigo which worsens with changes in middle ear pressure and elicited by pneumatic otoscopy when the patient is asked to Valsalva
3. **blanching of a middle ear mass with application of pressure using a pneumatic otoscope**
4. a reddish hue on the promontory
5. splaying of the internal and external carotid arteries

**Difficulty Level: Moderate**

**Explanation:** This is classically seen in cases of tympanic paragangliomas (glomus tympanicum tumors) within the middle ear.

The trend in recent years has been to reduce the use of eponyms both in clinical practice and on board examinations. However, understanding the answer choices and explanations presented here will help you when presented with similar information within a questions stem on the boards.

Brown's sign describes blanching of a middle ear mass with application of pressure. This is classically associated with vascular masses, such as a tympanic paraganglioma.

1. A 28-year-old female presents with photophobia, lacrimation, blurry vision and decreased hearing. Further questioning reveals she has been having a sensation that the world is spinning around her. She denies any history of similar problems. What is the likely diagnosis?
2. Meniere's disease
3. Autoimmune hearing loss
4. **Cogan's syndrome**
5. Goldenhar's syndrome
6. Alport syndrome

**Difficulty Level: Easy**

**Explanation:** Cogan’s syndrome is an autoimmune disease with episodic vertigo, interstitial keratitis, and bilateral fluctuating sensorineural hearing loss. It tends to present in the 20s to 30s. Treatment is with high dose steroids and possibly immunosuppresants including cyclophosphamide or azathioprine.

Characteristics of Cogan's syndrome include episodic vertigo, interstitial keratitis and bilateral fluctuating hearing loss. Of note, very few entities cause sensineural hearing loss and interstitial keratitis. Syphilis and Cogan's syndrome are among these.

1. What is the most common reason a child who passed newborn hearing screening presents for evaluation of hearing loss?
2. Failed school hearing test
3. **Parental concerns**
4. Failed hearing test at the primary care provider’s office
5. Speech and language delay
6. Incidental imaging findings

**Difficulty Level: Hard**

**Explanation:**  According to a recent study, parental concerns were the most common reason for suspicion of hearing loss prompting ENT referral, followed by failed school hearing screens, speech and language delay, failed primary care provider screen, audiogram after tympanostomy tube placement, and incidental scan findings.

Parental concern for hearing loss is a common reason for ENT referral for pediatric patients, even when the chid has passed a newborn hearing screen and has no objective evidence showing hearing loss.

1. A 53 year old male presents with a left longitudinal temporal bone fracture after falling off his motorcycle. The fracture is otic capsule sparing and travels just anterior to the geniculate ganglion. Facial strength is intact and symmetric. Based on the location of his fracture, what is the most likely sign or symptom he will have?
2. midface hypoesthesia
3. palatal numbness
4. dilated pupil
5. **dry eye**
6. altered taste sensation

**Difficulty Level: Moderate**

**Explanation:** Anterior to the geniculate ganglion the facial nerve gives off the greater superficial petrosal nerve (GSPN). This nerve contains parasympathetic innervation to the lacrimal gland, which if damaged would result in dry eyes as seen in this patient. The course of these nerve fibers is from the superior salivary nucleus, via the facial nerve, through the geniculate ganglion without synapsing, via the GSPN, and then joins with the deep petrosal nerve to form the vidian nerve. The vidian nerve synpases in the pterygopalatine fossa which completes the preganglionic pathway. Postganglionic axons then travel from the pterygopalatine fossa to the lacrimal gland to provide parasymphatetic innervation.

A fracture anterior to the geniculate ganglion may disrupt the greater superficial petrosal nerve. This nerve caries the parasympathetic innervation to the lacrimal gland via the pterygopalatine ganglion, and so injury to this nerve may result in dry eye symptoms.

1. All of the following nerves provide sensory innervation to the auricle except?
2. **C1**
3. C2
4. C3
5. Cranial nerve VII
6. Cranial nerve X

**Difficulty Level: Hard**

**Explanation:** C1 does not contribute sensory innervation to the auricle.

1. A 32 year-old Caucasian female patient is referred to you by the emergency room for evaluation of sudden bilateral facial nerve paralysis with onset 2 days ago. The patient reports having had fatigue, myalgias and arthralgias for the last 2 weeks as well as chills and malaise. She denies any prior medical or surgical history or prior episodes of Bell'™s palsy. She reports having worked as a summer camp counselor in Connecticut 2 months ago. Examination reveals bilateral House-Brackmann IV facial paralysis but an otherwise normal examination. Which of the following is true regarding this patient'™s most likely diagnosis?
2. Serologic testing for Babesia Microti should be initiated
3. ACE levels are likely elevated in this patient.
4. ELISA testing for anti-Borrelia burgdorferi IgG & IgM would likely be negative.
5. **This patient is showing signs of and symptoms of Stage II Lyme disease.**
6. Intravenous Immunoglobulin (IVIg) will likely result in complete symptom resolution within one week.

**Difficulty Level: Easy**

**Explanation:** This patient is showing symptoms of Stage II Lyme disease. Cardiac abnormalities, like AV block, meningoencephalitis, and multiple annular rashes may also occur in stage II. Treatment with a tetracycline (e.g. doxycycline 100 mg BID) for 21 days is the therapy of choice in these patients. There are very few entities that can cause bilateral Bell's palsy, and so if this is observed, Lyme disease can be presumed until proven otherwise.

Stage II of Lyme disease can manifest as multiple annular rashes, AV conduction block, meningoencephalitis and Bell's palsy, which may be bilateral. In general, it is uncommon to have bilateral Bell's palsy, and so the presence of bilateral Bell's palsy is highly suggestive of Lyme disease.

1. A 70-year-old male presents with a 3-month history of right pulsatile tinnitus with hearing loss, unstable gait, and autophony. ECOG demonstrates an SP/AP ratio of 0.6. A diagnosis of superior semicircular canal dehiscence is suspected. Which of the following is a good indicator test that would give support to this diagnosis?
2. Otoacoustic emissions
3. Tympanometry
4. **Cervical vestibular evoked myogenic potential**
5. Auditory brainstem response
6. Acoustic reflexes

**Difficulty Level: Easy**

**Explanation:** Superior semicircular canal dehiscence (SSCD) usually demonstrates an elevated SP/AP ratio above 0.4 on ECOG. An additional test that can be a good diagnostic indicator is cervical vestibular evoked myogenic potential (cVEMP). This tests the function of the saccule and the inferior vestibular nerve by measuring evoked potentials in the sternocleidomastoid muscle. In SSCD the threshold of the cVEMP is abnormally low (65 dB or less) and amplitudes are high, making this test a good indicator of SSCD. Occular vestibular evoked myogenic potential (oVEMP) tests the function of the superior vestibular nerve by measuring evoked potentials in the inferior oblique muscle. Some studies have shown superiority of oVEMP compared to cVEMP testing in the evaluation of SSCD, however cVEMP tends to be performed more often, likely because it is more practical, and there is generally more consensus regarding standards adn thresholds for cVEMP compared to oVEMP. The gold standard for diagnosis is a dedicated temporal bone CT.

A high SP/AP ratio on ECOG, and low threshold/high amplitude on cVEMP is very sensitive for SSCD.

1. A 53-year-old patient with bilateral profound SNHL undergoes cochlear implantation. During the post-operative period testing, there is spasm of the ipsilateral face. What is the appropriate management?
2. Turn off the implant and implant the other side
3. Reposition the electrode
4. **Reprogram the electrode**
5. Botox to the ipsilateral face

**Difficulty Level: Moderate**

**Explanation:** Facial nerve stimulation (FNS) is considered a minor complications of the after cochlear implantation (CI). FNS can occur immediately upon CI activation but more frequently can be delayed. The management of FNS requires familiarity with programming techniques and, in some cases, surgical explantation or re-implantation. Generally, FNS can be resolved minimally by reprograming the electrode.

In instances of facial nerve stimulation following cochlear implantation, attempts should be made to reprogram the electrode as this is a safe and fairly simple method that is typically successful.

1. The scutum represents:
2. **Lateral wall of the epitympanum**
3. Area between the chorda tympani and facial nerve
4. Area between the subiculum and the ponticulus
5. Medial wall of the mesotympanum
6. None of the above

**Difficulty Level: Easy**

**Explanation:** The scutum is a wedge shaped piece of bone at the lateral border of the epitympanum. Blunting of the scutum seen on CT scan is often a sign of cholesteatoma.

The scutum represents the lateral wall of the epitympanum.

1. A 57 year old male presents with complaints of brief episodes of ‘room spinning’. He describes these episodes as lasting 10-30 seconds and are typically triggered when turning his head while driving or rolling over in bed to the left only. He had several episodes of this last year but these resolved without relapse until the past week. He has no hearing loss or other significant otologic history. What finding on exam would support your diagnosis?
2. Positive head thrust testing
3. Rotary nystagmus lasting less than 60 seconds during a right sided Dix-Hallpike maneuver
4. Rotary nystagmus lasting more than 60 seconds during a right sided Dix-Hallpike maneuver
5. **Rotary nystagmus lasting less than 60 seconds during a left sided Dix-Hallpike maneuver**
6. Rotary nystagmus lasting more than 60 seconds during a left sided Dix-Hallpike maneuver
7. Normal physical exam

**Difficulty Level: Easy**

**Explanation:** This patient displays classic symptoms of benign paroxysmal positional vertigo including: brief episodes of vertigo, vertigo precipitated by head movement, previous self-resolving episodes, and lack of hearing loss. The classic finding for BPPV is a rotary nystagmus (with a vertical component) on ipsilateral Dix-Hallpike testing that lasts for less than 60 seconds. As this patient has symptoms when rolling over in bed to the left only the canal most likely to be affected is the left posterior semicircular canal. The posterior semicircular canal is by far the most commonly affected site for BPPV. The horizontal semicircular canal is affected in a minority of patients, and it is unclear whether or not superior semicircular canal BPPV truly exists.

Posterior semicircular canal BPPV would be expected to produce a rotary nystagmus lasting less than 60 seconds during an ipsilateral Dix-Hallpike maneuver.

1. A 24 year old male patient presents for evaluation of right sided unilateral hearing loss and vertigo. He states that he first noted the hearing loss several months ago, and over the past several weeks, has had intermittent episodes of vertigo. He denies any significant otologic history or head trauma. On exam, you note a white mass behind the tympanic membrane without any retraction or perforation. An audiogram demonstrates a mixed hearing loss on the right with normal hearing on the left. The audiologist tells you that the patient became very vertiginous when she performed tympanometry on the right side. A CT demonstrates a soft tissue density mass in the right middle ear, with erosion of the ossicles and lateral semicircular canal. A middle ear exploration is performed, and a pearly mass with keratin debris is encountered, consistent with a cholesteatoma. With regards to congenital cholesteatoma, which of the following is true?
2. There is a female to male preponderance of 2:1
3. Most congenital cholesteatomas are visualized as a white mass in the anterior-inferior quadrant
4. A history of otorrhea must precede the diagnosis of congenital cholesteatoma
5. **The mean age of presentation is around the age of 5**

**Difficulty Level: Moderate**

**Explanation:** Cholesteatomas are differentiated into congenital, primary acquired and secondary acquired. Congenital cholesteatomas arise out of keratinizing epithelium within the middle ear clefts. Congenital cholesteatomas tend to behave differently than acquired cholesteatomas, with slower growth and overall less destructive capacity. It is not uncommon for patients with a congenital cholesteatoma to not have hearing loss until the cholesteatoma becomes fairly large and begins to encroach on the ossicles. Physical exam most often shows that the cholesteatoma is in the anterior superior quadrant, and typically the remainder of the visualized middle ear space and tympanic membrane are unremarkable. The mean age of presentation is at the 4.5 years, and there is a male to female preponderance. In contrast to patients with congenital cholesteatomas, patients with acquired cholesteatomas tend to be older and have a history of chronic ear disease (eustachian tube dysfunction, recurrent acute otitis media) or a history of ear surgery (usually tympanostomy tubes). Surgical outcomes are generally better for patients with congenital cholesteatomas.

Although patients with congenital cholesteatomas present at a younger age, their disease is generally less severe and outcomes are better compared to those with acquired cholesteatomas.

1. A 67 year old male undergoes left sided cochlear implantation for profound bilateral deafness. During drillout of the facial nerve the patient bucks violently. After anesthesia is deepened it is noted that the facial nerve was completely transected with a 1mm diamond burr along the midportion of the mastoid segment. The frayed edges of the nerve are in contact.What is the best choice for repair of this facial nerve injury?
2. place the cut edges of the nerve in contact and abort the procedure
3. **trim the frayed edges of the nerve and drillout the rest of the facial nerve canal as needed to relieve tension for reapproximation**
4. trim 1cm superiorly and inferiorly from the site of injury and reconstruct with ipsilateral great auricular nerve
5. tag the distal end of the nerve injury with a clip, abort procedure, and plan for a future hypoglossal facial jump graft
6. perform concomitant upper eyelid gold weight placement, lower lid canthoplasty, and static facial sling for the expected complete facial paralysis

**Difficulty Level: Easy**

**Explanation:** After trimming the frayed edges the facial nerve can be further released by decompressing the adjacent facial nerve from bony canal. This should provide a small amount of laxity to the nerve allowing a tension free approximation without sutures. Sutures may be used but this should be under a completely tension free closure and the fewest number of sutures should be used to provide stable coaptation.

In repairing a transected nerve, the goal is to obtain a primary, tension free closure.

1. A patient undergoes middle ear exploration for suspected spontaneous perilymphatic fistula (PLF). Upon thorough evaluation of the middle ear through a transcanal approach, no anatomic abnormalities are noted. The ossicles appear normal and there is a bony overhang over the round window. The scutum is taken down to obtain full view of the stapes footplate. Gentle palpation of the incus elicits a small amount of clear fluid around the stapes. This is repeated to identify the site of fistula but the source cannot be determined. You decide to proceed with a temporalis fascia repair of the fistula. Based on the most common site of PLF where should the graft be placed?
2. Between the stapes crura
3. **Anterior to the anterior crus**
4. Posterior to the posterior crus
5. Along the inferior aspect of the stapes
6. Over the round window niche

**Difficulty Level: Hard**

**Explanation:** This patient has a confirmed PLF of the oval window. In the absence of congenital abnormalities of the ossicles (such as a perforated footplate or absent posterior crus) the most common sites of fistula (in order) are: 1) anterior to the anterior crus, 2) anteroinferior to the anterior crus, 3) anterosuperior to the anterior crus. Fistulas of the round window are less frequent and should not be grafted unless a leak can be demonstrated.

The most common site of a spontaneous perilymphatic fistula is anterior to the anterior crus.

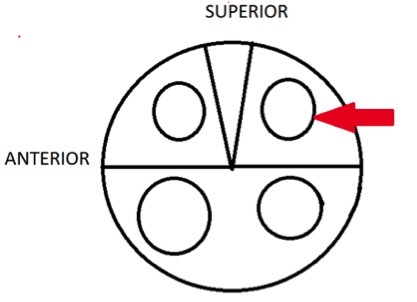
1. Which of the following represents the most common patterns of spread from Prussak's space of cholesteatomas in order of decreasing frequency?
2. Posterior epitympanum, anterior epitympanum, posterior mesotympanum
3. **Posterior epitympanum, posterior mesotympanum, anterior epitympanum**
4. Anterior epitympanum, posterior mesotympanum, posterior epitympanum
5. Posterior mesotympanum, anterior epitympanum, posterior epitympanum
6. Anterior epitympanum, posterior epitympanum, posterior mesotympanum

**Difficulty Level: Hard**

**Explanation:** Epitympanum cholesteatomas originate in a shallow pocket, called Prussak space, which lies between the pars flaccida and the neck of the malleus. Cholesteatomas most commonly exit Prussak space by the posterior route and penetrates the superior incudal space lateral to the body of the incus. From there, a cholesteatoma can transverse the aditus ad antrum to enter the mastoid.

The most common patterns of spread from Prussak's space of cholesteatomas in decreasing frequency are the posterior epitympanum, posterior mesotympanum, and the anterior epitympanum.

1. A depiction of the internal auditory canal is displayed below. If a lesion involved the structure displayed by the arrow, which of the following will be the pathophysiologic result?



1. Facial asymmetry at rest
2. Hearing loss
3. Posterior semicircular canal deficit on impulse testing
4. **Superior semicircular canal deficit on impulse testing**
5. External auditory meatus numbness

**Difficulty Level: Easy**

**Explanation:** Knowing the orientation of structures in the internal auditory canal is important in localizing small pathology and correlating to clinical signs and symptoms. The internal auditory canal is broken into quandrants essentially by Bill’s bar vertically and the falciform (or transverse) crest horizontally. The facial nerve is in the anterior-superior quadrant. The cochlear nerve is in the anterior-inferior quadrant. The superior vestibular nerve is the the posterior-superior quadrant. The inferior vestibular nerve is in the posterior-inferior quadrant. The superior vestibular nerve controls function of the superior and lateral semicircular canals, as well as the utricle. The inferior vestibular nerve controls function of the posterior semicircular canal and saccule. Thus a lesion involving the superior vestibular nerve would cause a deficit of superior semicircular canal function seen on impulse testing.

The superior vestibular nerve runs in the posterior superior quadrant of the internal auditory canal, and receives sensory information from the utricle, superior and lateral semicircular canals.

1. Which of the following aminoglycosides is the most vestibulotoxic:
2. Gentamicin
3. **Streptomycin**
4. Amikacin
5. Neomycin
6. Kanamicin

**Difficulty Level: Hard**

**Explanation:** The primarily vestibulotoxic aminoglycosides include gentamicin and streptomycin. Of the two, streptomycin is more vestibulotoxic. The primarily cochleotoxic aminoglycosides include amikacin, kanamycin, and neomycin.

Streptomycin is the most vestibulotoxic aminoglycoside, followed by gentamicin.

1. A 21 year-old-male presents to the ED after being involved in a motor vehicle accident. The patient is admitted to the trauma service for his other injuries. During his hospital course, the patient is noted to have right-sided bloody otorrhea and a right House Brackman score of VI. After reviewing the records, the patient is documented to have facial paralysis on arrival in the ED. A CT of the temporal bone shows an otic capsule sparing right-sided temporal bone fracture. To help guide your surgical decision-making, you order electroneuronography (ENoG). What is the latest time point following the onset of acute facial paralysis that ENoG is considered helpful?
2. 3 days
3. 1 week
4. **2 weeks**
5. 3 weeks
6. 6 weeks

**Difficulty Level: Hard**

**Explanation:** ENoG is only useful to predict the prognosis between 3 days and 2 weeks after the onset of acute facial nerve paralysis. An ENoG should not be performed before 72 hours post-injury as Wallerian degeneration has not occurred yet. Accurate prediction using ENoG is difficult after two weeks when regeneration begins at the proximal end. A collateral neural circuit can develop from a healthy nerve around a degenerated nerve. With nonacute injury facial nerve, ENoG results are reliable for up to 3 weeks, although optimal reliability occurs up to 2 weeks. After 3 weeks, a desynchronization of electrically evoked facial nerve discharge can occur, preventing a single unified firing of all neurons. It is no longer possible to compare the diseased (asynchronous) side with the normal (synchronous) side. The comparison between the two sides (diseased versus normal) gives the results of ENoG. Surgery may be considered if ENoG is done within 2 weeks and shows a greater than 90% degeneration on the affected side compared to the normal side.

An ENoG should be performed between 3-14 days after acute facial nerve paralysis, and a greater than 90% degeneration on the affected side is an indication for surgery. For nonacute facial nerve paralysis, an ENoG can be performed between 3-21 days.

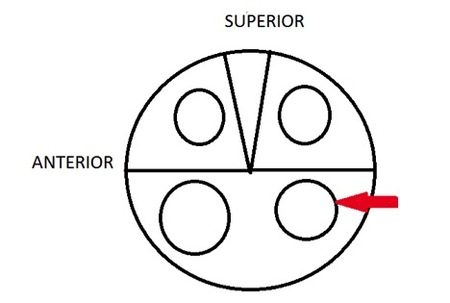
1. A 42 year old male with HIV presents to the emergency room with 1 month of left ear pain and drainage. The patient has not been on antibiotics and is non compliant with his HIV medications. The patient'™s vital signs are HR 102 BP 110/86 RR 17 O2 99% and Tmax 101.2. On physical exam the patient'™s left external auditory canal significantly edematous and there is granulation tissue seen at the bony cartilaginous junction. A CT scan is obtained showing significant inflammation of the left external auditory canal soft tissues and bony erosion of the bony external auditory canal. What is the most common fungal pathogen causing this disease process?
2. Candida
3. **Aspergillus**
4. Mucor species
5. Histoplasmosis
6. Cryptococcus

**Difficulty Level: Moderate**

**Explanation:** Malignant otitis externa (aka necrotizing otitis externa) presents as long standing otalgia (typically >4 weeks), otorrhea, and the hallmark finding of granulation tissue at the bony cartilaginous junction. In its early stages the infection is confined to the soft tissues but can progress to involve the bone leading to cranial nerve palsies. Most cases of malignant otits externa are caused by Pseudomonas aeruginosa. The question asks for the most common fungal pathogen which is Aspergillus sp.

In cases of malignant otitis externa, Pseudomonas aeruginosa is the most common bacterial pathogen and Aspergillus sp is the most common fungal pathogen.

1. A depiction of the internal auditory canal is displayed below. If a lesion involved the structure displayed by the arrow, which of the following will be the pathophysiologic result?



1. Facial asymmetry at rest
2. Hearing loss
3. **Posterior semicircular canal deficit on impulse testing**
4. Superior semicircular canal deficit on impulse testing
5. Superior semicircular canal deficit on impulse testing

**Difficulty Level: Easy**

**Explanation:** Knowing the orientation of structures in the internal auditory canal is important in localizing small pathology and correlating to clinical signs and symptoms. The internal auditory canal is broken into quandrants essentially by Bill’s bar vertically and the falciform (or transverse) crest horizontally. The facial nerve is in the anterior-superior quadrant. The cochlear nerve is in the anterior-inferior quadrant. The superior vestibular nerve is the the posterior-superior quadrant. The inferior vestibular nerve is in the posterior-inferior quadrant. The superior vestibular nerve controls function of the superior and lateral semicircular canals, as well as the utricle. The inferior vestibular nerve controls function of the posterior semicircular canal and saccule. Thus a lesion involving the inferior vestibular nerve would cause a deficit of posterior semicircular canal function seen on impulse testing. A facial nerve lesion would result in facial asymmetry. A cochlear nerve lesion would result in hearing loss. The nervus intermedius runs alongside the facial nerve and contributes sensory input to the external auditory meatus.

The inferior vestibular nerve, which runs in the posterior inferior quadrant in the internal auditory canal, receives sensory information from the posterior semicircular canal and saccule.

1. Which of the following best describes the correct anatomic relationship of the facial nerve and its branches to adjacent structures?
2. **Parallel and superior to the cochleariform process and just superior to the oval window**
3. Posteromedial to the styloid process at the stylomastoid foramen
4. Deep to the mentalis and depressor anguli oris muscles
5. Superior to the pyramidal eminence
6. Inferior to the the vestibulocochlear nerve in the internal auditory canal

**Difficulty Level: Hard**

**Explanation:** Within the internal auditory canal, the facial nerve is found in the anterosuperior quadrant immediately above the cochlear nerve. The superior and inferior vestibular nerves travel in the posterosuperior and posteriorinferior quadrants, respectively. After the first genu, the tympanic segment of the facial nerve runs parallel and superior the cochleariform process and just superior to the oval window. It then turns inferiorly (second genu) and travels vertically through the mastoid passing posterior to the pyramidal eminence. It exits out of the stylomastoid foramen, which lies posterolateral to the styloid process. The extratemporal branches of the facial nerve run deep to the SMAS in the lower face and innervate most mimetic muscles on their deep surfaces. The only exceptions to this rule are the mentalis, levator anguli oris and buccinators muscles, which are innervated on their superficial surface.

The tympanic segment of the facial nerve runs parallel and superior the cochleariform process and just superior to the oval window. It is important to understand this relationship when performing a stapedectomy, clearing cholesteatoma or other disease near the footplate, or other procedures in this area.

1. A 44 year old construction worker presents for evaluation of hearing loss. He has noted steady, progressive hearing loss in both ears for many years, and now has had trouble hearing lower volumes of noise, particularly at the end of his work day. He also frequents rock concerts. He wears ear protection at work, but notes he did not use any when he was younger and does not wear any ear plugs when he goes to concerts. Which of the following daily occupational noise exposure levels meets OSHA standards for the work place?
2. **85 dB continuous exposure for 8 hours**
3. 4 hours of exposure to 100 dB and 20 minutes exposure to 110 dB
4. 3 separate 1 hour exposures to 100 dB
5. 1 hour of exposure to 110 dB
6. 5 minutes of exposure to 130 dB

**Difficulty Level: Moderate**

**Explanation:** The Occupational Safety and Health Administration (OHSA) sets limits on the permissible amounts of noise exposure in the workplace to help prevent noise-induced hearing loss. The maximum allowable daily noise exposure limit has been specified as an average of 90 dB over an 8 hour period. Sound intensity greater than 90 dB can be experienced, but for shorter periods of time. For example, 2 hours of 100 dB exposure or 30 minutes of 110 dB exposure is permissible. As one may extrapolate every five dB increase in noise level halves the allotted exposure time. This can thus be extrapolated to 7.5 minutes for 120 dB and <2 minutes for 130dB. OSHA also mandates that there be no exposure to 140dB sound whatsoever.

An easy way to remember the OSHA limits for daily occupational noise exposure is to remember that the scale begins at 16 hours of 85 dB noise exposure, and for every 5 dB increase in the sound level, the permissible number of hours at that level is halved. So, 8 hours at 90 dB, 4 hours at 95 dB, 2 hours at 100 dB, etc.

1. Which of the following is true regarding otosclerosis?
2. There is a 3:1 male:female predilection
3. **It can present with a sensorineural hearing loss**
4. It is an autosomal recessive inheritance pattern
5. It is more common in Black/African-American than Caucasian patients
6. The incidence of histopathologic disease is much lower than the incidence of clinical otosclerosis

**Difficulty Level: Moderate**

**Explanation:** Although it most commonly presents with a conductive hearing loss, otosclerosis can rarely present as a sensorineural loss if the otospongiotic process is primarily around the cochlea rather than around the conductive apparatus.

Otosclerosis traditionally presents as a conductive hearing loss with a Carhart notch at 2k Hz, however it can present as a mixed or sensineural hearing loss in a minority of patients.

1. What Is the most common complication of cochlear implantation in children?
2. **Flap infection**
3. Device failure
4. Facial nerve palsy
5. Displaced electrode
6. Facial nerve stimulation

**Difficulty Level: Hard**

**Explanation:** Flap infection was the most common complication in the cited studies. It occurred in 2.6% of patients.

Complication rates in pediatric cochlear implantations are low. The most common complication is a flap infection.

1. What is the most common intracranial complication of otitis media?
2. Brain abscess
3. Petrous apicitis
4. **Meningitis**
5. Lateral sinus thrombosis
6. Labyrinthine apoplexy

**Difficulty Level: Moderate**

Although intracranial complications from acute otitis media are a rare event due to widespread use of antibiotics, meningitis is the most common intracranial complication.

1. The majority of congenital cholesteatomas are seen in what portion of the middle ear?
2. **Anterior-superior**
3. Anterior-inferior
4. Posterior-superior
5. Posterior-inferior
6. Within the tympanic membrane

**Difficulty Level: Moderate**

**Explanation:** Two-thirds of middle ear congenital cholesteatomas are seen as a white mass in the anterior-superior quadrant. As such, they do not tend to cause a conductive hearing loss until they are big enough to inferfere with or begin to erode with the ossicles. They may also be found within the tympanic membrane and in the petrous apex. The mean age at presentation is 4.5 years, with a male to female predominance of 3:1.

The majority of congenital cholesteatomas are found within the anterior-superior quandrant.

1. What is the most common inherited or developmental form of congenital facial palsy?
2. Birth trauma
3. Moebius syndrome
4. Albers-Schonberg syndrome
5. **Congenital unilateral lower lip palsy**
6. Ramsay-Hunt syndrome

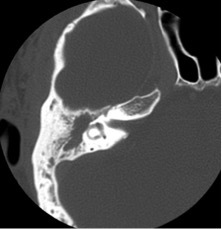
**Difficulty Level: Hard**

**Explanation:** CULLP occurs in about 1/160 live births, making it the most common inherited form of congenital facial palsy. It is due to hypoplasia/absence of the depressor anguli oris or depressor labii inferioris muscles. Patients will have drooping of the lower lip, most obvious when laughing or crying, but the rest of the facial function is normal. A significant minority of these patients will have cardiac defects and/or be on the 22q11 deletion syndrome spectrum.

Congenital unilateral lower lip palsy is the most common inherited form of congenital facial palsy.

1. A 32 year old male with severe to profound left side sensorineural hearing loss and mild right side sensorineural hearing loss presents with concern for cholesteatoma in the right ear. He describes having vertigo in environments with loud noises. High resolution CT is pictured below.

Which of the following is the appropriate surgical management of this patient?



1. Canal wall up mastoidectomy with removal of all cholesteatoma matrix
2. Canal wall up mastoidectomy and leave matrix intact over fistula
3. Canal wall down mastoidectomy with removal of all cholesteatoma matrix
4. **Canal wall down mastoidectomy and leave matrix intact over fistula**
5. Radical mastoidectomy

**Difficulty Level: Moderate**

**Explanation:** This patient has a labyrinthine fistula as evidenced by CT imaging and Tullio phenomenon (vertigo induced by loud noise). The procedure of choice in a labyrinthine fistula case is a modified canal wall down mastoidectomy. This avoids leaving residual disease concealed in the mastoid cavity. Management of the cholesteatoma matrix covering the fistula depends on several factors, including the infection status of the ear, the degree of sensorineural hearing loss in the involved ear as well as the opposite ear, the size and location of the fistula, and the surgeon’s skill. Because this patient has profound hearing loss in the left ear, care should be taken to avoid worsening the hearing in the right ear. A canal wall down procedure should be performed and the matrix left intact over the fistula. Attempting to remove the matrix places the patient at risk for permanent total sensorineural hearing loss. A radical mastoidectomy is too aggressive at this point.

Although canal wall down procedures are more aggressive and require ongoing maintenance, they are preferable to canal wall up procedures in certain instances. These include instances where known disease is being left behind, as the mastoid cavity becomes externalized and can be cleaned periodically, and particularly in instances that require matrix to be left over an eroded inner ear structure.

1. A 48 year old presents to your clinic complaining of unilateral tinnitus. Further questioning reveals it is pulsatile in nature and present all of the time. On physical exam you see a reddish mass behind the tympanic membrane which blanches with pneumatic otoscopy. What is the most appropriate next step in management?
2. Perform trans tympanic biopsy
3. Watchful waiting
4. Order an ultrasound
5. Prescribe empric amoxicillin/clavulanic acid
6. **Order a computed tomography with contrast**

**Difficulty Level: Easy**

**Explanation:** Unilateral tinnitus is more concerning than bilateral tinnitus. The fact that his is pulsatile in nature indicates it is likely vascular in nature. "Brown's sign" is what occurs when the discoloration blanches with pneumatic otoscopy, indicative of a vascular tumor. The most common middle ear vascular tumors are tympanic paragangliomas (glomus tympanicum) which are neuroendocrine tumors that develop from the paraganglion cells. These lesions can be visualized with MRI, computed tomography with contrast, or a computed tomographic angiogram.

If a vascular tumor of the middle ear is suspected, further imaging with an MRI, CT with contrast, or CTA is indicated as part of the evaluation.

1. A full term female is found to have an external ear deformity and aural atresia during a newborn examination. The ear is essentially a nubbin of tissue with no recognizable landmarks. You diagnose her with congenital aural atresia and a type III microtia.

The antihelical crura of the ear is derived from which Hillock of His?



1. First
2. Second
3. Third
4. **Fourth**
5. Fifth

**Difficulty Level: Hard**

**Explanation:** The mesoderm from the first and second branchial arches form the Hillocks of His, which give rise to the structures of the external ear. There are 6 hillocks of His: The first three hillocks develop from the first branchial arch into the tragus (1), helical crus (2), and helix (3). The second three hillocks develop from the second branchial arch into the antihelical crus(4), antihelix (5), and antitragus/lobule (6). There are some variations in the literature regarding which structures are derived from certain hillocks, and more current resources acknowledge that there is likely overlap. The hillocks of His are fused by 12 weeks gestation and reach a final shape around 20 weeks gestation. Innappropriate fusion may result in the development of a fistula or sinus tract.

The fourth Hillock of His gives rise to crura of the antihelix.

1. An 8 year old girl presents to your clinic after failing a school hearing screen test. She has bilateral severe high frequency sensorineural hearing loss on audiologic evaluation. On physical exam she has a diffusely enlarged thyroid gland. Which of the following should be ordered to identify the cause?
2. EKG
3. Renal ultrasound
4. **Genetic testing for mutation in SLC26A4**
5. TSH and T4
6. CT Scan

**Difficulty Level: Easy**

**Explanation:** This child has Pendred syndrome, an autosomal recessive syndrome characterized by thyroid goiter and profound sensorineural hearing loss. The hearing loss is associated with abnormal iodine metabolism resulting in a euthyroid goiter, which usually becomes clinically detectable at about 8 years of age. Currently, testing for mutations in the Pendrin gene, SLC26A4 (also termed PDS), is diagnostic. A mutation results in defects in thyroid hormone biosynthesis, specifically in transport across the apical membrane where it functions as a chloride/iodide pump and is responsible for transporting iodine out of the cell and into the follicular colloid, and in the cochlea, where it results in a sensorineural hearing loss. The perchlorate discharge test was previously the gold standard for diagnosis. It would demonstrate abnormal organification of nonorganic iodine in these patients. Specific inborn errors of thyroid metabolism can be identified by thyroid radionuclide imaging or ultrasonography, radioiodine uptake followed by measurement of perchlorate discharge in patients with elevated uptake, or measurements of serum thyroglobulin and iodothyronines. The results will determine which gene may be involved. A CT scan is indicated after diagnosis, to look for associated anomalies (such as enlarged vestibular aquaduct or mondini malformation).

A genetic test showing a mutation in the Pendrin gene, SLC26A4, is diagnostic for Pendred syndrome.

1. An elevated SP/AP ratio is a useful value determined by using which of the following diagnostic tests?
2. Tympanometry
3. auditory brainstem response
4. **electrocochleography**
5. distortion product otoacoustic emissions

**Difficulty Level: Easy**

Endolymphatic hydrops affects the basilar membrane elasticity which can be seen as increased amplitude of the SP (summating potential) relative to that of the AP (whole-nerve action potential). This testing requires either a transtympanic electrode at round window or tympanic membrane surface electrode.

Electrocochleography measures the cochlear microphonic, summation potential and action potential of the cochlea. An SP/AP ratio >0.4 is suggestive (though not diagnostic) of a hydropic process.

1. Which of the following statements regarding schwannomas of the temporal bone is true?
2. The most common schwannoma of the temporal bone is the facial nerve schwannoma
3. The most common presenting symptom of vestibular schwannoma is facial numbness
4. Facial nerve schwannomas usually present with acute facial nerve weakness
5. **Sensorineural hearing loss is more common in patients with vestibular schwannoma and conductive hearing loss is more common in patients with facial nerve schwannoma**
6. Histologic sections of schwannoma demonstrate spindle shaped cells arranged in nests and psammoma bodies

**Difficulty Level: Hard**

**Explanation:** Schwannomas can arise from all cranial nerves except the optic nerve. They are slow-growing, benign, encapsulated neoplasms that arise from the nerve sheath. They consist of Schwann cells in a collagenous matrix. Schwannomas also occur in spinal nerve roots, flexor surfaces of the upper and lower extremities, and the tongue. Sensorineural hearing loss is more common in patients with vestibular schwannoma and conductive hearing loss is more common in patients with facial nerve schwannoma. Audiometric evaluation is helpful in differentiating the two. Conceptually this should make sense since the 8th cranial nerve is not present within the middle ear space where the structures are that make up the conductive hearing mechanism, but the 7th cranial nerve's tympanic segment does pass through.

Sensorineural hearing loss is more common in patients with vestibular schwannoma and conductive hearing loss is more common in patients with facial nerve schwannoma.

1. A 65 year old male with a history right sided herpes zoster oticus two years ago, comes in to clinic with complaints of chronic right ear pain. He complains of burning pain in right ear. Exam does not show any abnormalities of ear canal skin, tympanic membrane or middle ear. What is the most appropriate therapy?
2. NSAID
3. **Gabapentin**
4. Varicella zoster vaccine
5. Valacyclovir
6. Prednisone

**Difficulty Level: Easy**

**Explanation:** Patient exhibits symptoms of post herpetic neuralgia. Postherpetic neuralgia can be a significant problem, and it is more common in patients over 60 years of age. Gabapentin has been shown to be the preferred agent to treat this pain. Universal vaccination of infants against VZV and the U.S. Food and Drug Administration approval of a zoster vaccine for older adults may result in a decreased incidence of zoster infections in the future; however, these vaccines are not indicated for treatment of active infection or postherpetic neuralgia.

Gabapentin is the preferred medication for treatment of post herpetic neuralgia.

1. A 7 year old male is brought to your clinic by his parents because he is having difficulty at school. An audiogram shows severe to profound sensorineural hearing loss bilaterally. He also has diffuse symmetrical neck swelling which moves with deglutition. Which of the following test would be most useful to establish a diagnosis?
2. Perchlorate discharge test
3. **Mutations in the SLC26A4 gene**
4. Repeat audiogram
5. Thyroid stimulation hormone levels
6. Ultrasound of the neck

**Difficulty Level: Moderate**

**Explanation:** This patient has symptoms and signs most consistent with Pendred syndrome. This is a syndrome associated with congenital and usually progressive sensorineural hearing loss. The majority of these patients will also have a thyroid goiter after the age of 10. It is caused by a homozygous or compound heterozygous mutation in the SLC26A4 gene on chromosome 7q and is usually inherited in an autosomal recessive manner. These patients frequently have an enlarged vestibular aqueduct and Mondini malformation which may be seen on a computed tomography of the temporal bone.

Pendred syndrome is an autosomal recessive condition characterized by sensineural hearing loss, Mondini malformation with an enlarged vestibular aqueduct, and euthyroid goiter. It is caused by mutations in the SLC26A4 gene.

1. Select the best answer regarding the earliest age could you perform an auditory brainstem implant for a patient with NF2 and hearing loss
2. 1 year
3. 5 years
4. **12 years**
5. 15 years
6. only over 18 years of age

**Difficulty Level: Hard**

**Explanation:** Trials are being conducted in Europe and the US to examine the utility of implantation at younger ages, but currently this device is FDA approved for patients 12 and older.

Auditory brainstem implants are FDA approved for patients 12 years of age or older who have been diagnosed with neurofibromatosis type 2.

1. A 55 year old female presents with a complaint of left side pulsatile tinnitus and says she hears her footsteps loudly when she walks. Her audiogram reveals a mild conductive hearing loss with air bone gaps greatest in the lower frequencies. Acoustic reflexes are normal. Electrocochleography reveals an SP/AP ratio of 0.6. Based on history and testing alone, what is the most likely diagnosis?
2. Meniere disease
3. **Superior semicircular canal dehiscence**
4. Acoustic neuroma
5. Otosclerosis
6. Tympanic membrane perforation

**Difficulty Level: Easy**

**Explanation:** Superior semicircular canal dehiscence has a range of auditory and vestibular manifestations including autophony, pulsatile tinnitus, aural pressure, hyperacusis, unstable gait, and sound or pressure evoked vertigo. The pulsatile tinnitus occurs due to exposure of the canal to changes in CSF pressure or to the flow of adjacent blood vessels. There is typically normal symmetric hearing, but a conductive hearing loss with larger low-frequency air-bone gaps and supranormal bone conduction thresholds can be seen. Acoustic reflex testing is normal. Electrocochleography (ECOG) will show an elevated SP/AP ratio above 0.4 typically, and VEMP testing will show a low threshold with high amplitudes. If SSCD is suspected, a CT is indicated for further evaluation.

Superior semicircular canal dehiscence often presents with autophony, pulsatile tinnitus, aural pressure, hyperacusis, unstable gait, and sound or pressure evoked vertigo. Audiometry, ECOG, and VEMP testing can all aid in diagnosis, but ultimately the gold standard for diagnosis is a temporal bone CT.

1. A 32 year-old male presents for the evaluation of right hearing loss for several years. He reports a history of recurrent otitis media affecting his right ear for most of his childhood. He denies prior ear surgeries and currently reports no otalgia, otorrhea, or vertigo. Examination reveals a near-total perforation of the right tympanic membrane. You decide to proceed with tympanoplasty via a lateral grafting technique. Which of the following is associated with the lateral grafting tympanoplasty technique compared to other techniques?
2. **Blunting**
3. Shorter operative time
4. Reduced healing time
5. Decreased graft-take rate
6. Less technically demanding

**Difficulty Level: Easy**

**Explanation:** Blunting is a phenomenon characterized by scarring in the anterior sulcus region that bridges the anterior graft and the anterior canal wall. Blunting can occur due to the sharp angulation of the anterior sulcus and can result in a persistent conductive hearing loss postoperatively. Blunting occurs mostly with lateral grafting tympanoplasty techniques, as the graft is free to heal against the anterior sulcus skin/bone. Graft lateralization is a significant risk of the lateral grafting technique, as the graft is free to displace laterally.

Blunting and graft lateralization are complications of the lateral grafting technique for tympanoplasty. The lateral grafting technique also requires meticulous removal of epithelial remnants prior to grafting, as this can cause a cholesteatoma

1. A 29 year-old woman presents with pain in the right ear beginning one week ago. This was followed by the gradual onset of weakness throughout the right side of her face. She also describes an oversensitivity to noise in the right ear and impaired taste. Which of the following is true?
2. The lesion of the facial nerve must be distal to the stylomastoid foramen
3. **The lesion of the facial nerve must affect the chorda tympani**
4. Facial reflexes will be spared
5. Emotional facial responses will be spared, while only voluntary motions will be affected
6. Weakness is unlikely to fully resolve

**Difficulty Level: Easy**

**Explanation:** The patient likely presents with Bell's palsy, an acute, unilateral, infranuclear facial nerve palsy. The onset typically consists of pain in or behind the ipsilateral ear, followed by worsening facial weakness over several days. Patients may also report impaired taste or hyperacusis, depending upon the nerve regions involved. MRI may reveal contrast enhancement of the facial nerve.

Bell’s palsy is often viral in etiology (i.e., herpes simplex). In many cases, the cause remains unknown. Other less common etiologies include sarcoidosis and Lyme disease. The lower motor neuron impairment of Bell’s palsy must be distinguished from upper motor neuron dysfunction. The dorsal aspect of the facial motor nucleus receives input from bilateral cortices, while neurons in the ventral part of the nucleus receive primarily contralateral cortical input. The result is that the upper face muscles are under bilateral cortical control, while the lower facial musculature is under contralateral cortical control. The clinical consequence is that unilateral cortical lesions may spare facial emotional responses and reflexes due to their bilateral cortical control. The more distal lower motor lesions would impair facial nerve function more broadly, voluntary movements, emotional responses, and reflexes equally.

Bell's palsy is an acute, unilateral, infranuclear facial nerve palsy. The onset typically consists of pain in or behind the ipsilateral ear, followed by worsening facial weakness over several days. Patients may also report impaired taste or hyperacusis, depending upon the nerve regions involved. MRI may reveal contrast enhancement of the facial nerve.

1. Following tympanostomy tube insertion, which of the following is true?
2. Routine antibiotic ear drops are recommended for a duration of 5 days
3. Tympanostomy tubes should be evaluated 3-6 months after placement
4. Dry ear precautions are recommended
5. **Intra-operatively, saline washouts and antibiotic/steroid ear drops have comparable efficacy in preventing post-operative otorrhea**

**Difficulty Level: Moderate**

**Explanation:** A Cochrane review from 2013 demonstrated similar efficacy of multiple saline washes and a single application of antibiotic/steroid ear drop in preventing post-operative otorrhea. Comparing the number needed to treat between the two methods, saline washouts had more favorable outcomes. Utilization of saline washouts after tube insertion has several advantages, including avoidance of antibiotics and lower cost. For patients with purulent effusions, antibiotic/steroid ear drops are generally recommended.

Saline washes and antibiotic/steroid ear drops intra-operatively have similar efficacy in preventing post-operative otorrhea, but saline washes avoid unnecessary antibiotic use and have lower cost.

1. A 42 year old overweight female presents to your office complaining of vertigo lasting 30-60 seconds when she is exposed to loud noises over the past several months. Additionally, she occasionally experiences this when ‘using the restroom’. On exam her ears appear normal and she does not have spontaneous nystagmus. Head thrust testing is compensatory. When she is asked to perform a Valsalva maneuver against a closed glottis she develops vertigo and nystagmus. When she looks to the left and performs the maneuver the slow phase of the nystagmus is vertical. When she looks to the right the slow phase is torsional. On audiometric exam she has 20 dB hearing loss in the left ear compared to the right. What disease process is she suffering from?
2. Benign paroxysmal positional vertigo
3. Meniere’s disease
4. **Superior semicircular canal dehiscence**
5. Tympanic paraganglioma
6. Labrynthitis

**Difficulty Level: Easy**

**Explanation:** Superior semicircular canal dehiscence creates another portal (‘third window’) of pressure release within the labyrinthine system. Because of this third window effect, it is often associated with a conductive or mixed hearing loss, with supranormal bone conduction thresholds, Tullio phenomenon (noise induced vertigo) and Hennebert sign (pressure induced vertigo). When a Valsalva maneuver is performed the intracranial pressure is raised which causes an increased pressure within the unilateral semicircular canal thereby affecting the superior semicircular canal (left in the example above). This elicits nystagmus in the direction of the canal as above. An elevated SP/AP ratio on electrocochleography and a cVEMP with decreased thresholds and increased amplitudes is consistent with SSCD. Diagnosis is most often confirmed with a CT with thin cuts through the tegmen mastoideum.

The third window effect caused by superior semicircular canal dehiscence manifests as a mixed or conductive hearing loss, Tullio phenomenon, and Hennebert sign.

1. A 68 year old male presents for evaluation of the lesion pictured below. It is very tender to palpation, and is very bothersome to him as he often uses the phone on that ear. A biopsy does not demonstrate any sign of malignancy. What is the most likely diagnosis?



1. Acrochordon
2. Seborrheic keratosis
3. **Chondrodermatitis nodularis helicis**
4. Nevus sebaceous
5. Pseudocyst

**Difficulty Level: Easy**

**Explanation:** A painful, tender lesion on the auricle of a middle aged to elderly male with a history of chronic irritation to that ear is a classic presentation for chondrodermatitis nodularis helicis, or Winkler nodule. It is a benign lesion that appears as a punched out crater found most commonly in older men at the rim of the helix or antihelix. The nodule usually enlarges rapidly to its maximum size and then remains stable. It is theorized that the lesion is due to prolonged or excessive pressure resulting in cartilage and skin necrosis from a localized vasculitis. The tenderness and pain often help to distinguish chondrodermatitis nodularis helicis from other benign or malignant lesions but ultimately a biopsy should be performed to rule out malignancy. Treatment is focused on eliminated the source of pressure on the lesion, and may require surgical excision or cryotherapy to remove the abnormal tissue.

Chondrodermatitis nodularis helicis typcially presents as a tender nodule of the helix or antihelix as the result of chronic pressure on the area. These lesions are benign and typically are treated by reducing pressure to the area and excising abnormal tissue.

1. A 15 year old patient reports 3 episodes in last year of facial swelling, particularly around the lip, with temporary facial weakness. What other exam finding may be present?
2. CN 12 weakness
3. **Fissured tongue**
4. Abducens palsy
5. Absence of corneal reflex
6. Hearing loss

**Difficulty Level: Moderate**

**Explanation:** Melkersson-Rosenthal syndrome is characterized by the triad of orofacial swelling, facial paralysis, and a fissured tonuge/angular cheilitis. The etiology is unknown, and steroids and surgical decompression have had limited success in treatment. ACE levels have been noted to be elevated during attacks.

Melkersson-Rosenthal syndrome presents in adolescence/early adulthood with recurrent facial edema accompanied by facial paresis. It characteristically is accompanied by angular cheilitis as well as a fissured tongue.

1. A 44 year old male presents with complaint of waking up this morning unable to hear well from the right ear. He has no significant medical history and is not taking medications or supplements. An audiogram immediately prior to his appointment shows hearing loss of 30 dB over three test frequencies . He is very anxious and wishes to receive some form of treatment for this problem. Which of the following is the next best step in management to offer this patient?
2. Intramuscular dexamethasone injection now
3. Valacyclovir for 14 days starting now
4. High dose oral prednisone taper if hearing loss persists for 4 weeks
5. **High dose oral prednisone starting now, then followed by a taper**
6. Pentoxifylline starting now

**Difficulty Level: Easy**

**Explanation:** Spontaneous improvement is common for sudden sensorineural hearing loss (SSNHL) and the data is contradictory on the effectiveness of glucocorticoid therapy. However, most experts suggest initial treatment with a 10- to 14-day course of high-dose glucocorticoids. Intratympanic (IT) glucocorticoids may be used as initial therapy for patients intolerant to high-dose steroids.

A high dose steroid taper or intratympanic steroid injection may be offered to patients with idiopathic sudden sensorineural hearing loss after a discussion of the risks and benefits of these treatment options. The patient should have an MRI brain/internal auditory canals at some point as some patients with retrocochlear pathology will initially present with sudden sensorineural hearing loss.

1. Donaldson’s line helps to identify the boundary of which structure in the inner ear?
2. The superior semicircular canal
3. **The superior aspect of the endolymphatic sac**
4. The inferior aspect of the endolymphatic sac
5. The superior aspect of the vestibule
6. The inferior aspect of the vestibule

**Difficulty Level: Moderate**

**Explanation:** Donaldson’s line is a hypothetical line that runs parallel to the horizontal semicircular canal and bisects the dome of the posterior semicircular canal. It marks the superior boundary of the endolymphatic sac. It is an important landmark in endolymphatic sac surgery.

1. A 42 year-old male presents complaining of noise-induced vertigo and that his voice sounds distorted and too loud.

Further clinical testing including audiogram, acoustic reflexes, and cervical vestibular evoked myogenic potential (cVEMP) are ordered to help with diagnosis. Physical exam reveals a normal tympanic membrane.

Which of the following represents the expected results of the testing for the disorder he most likely has?

1. Conductive hearing loss, absent acoustic reflexes, lower threshold on cervical vestibular evoked myogenic potential (cVEMP).
2. Conductive hearing loss, present acoustic reflexes, normal threshold on cervical vestibular evoked myogenic potential (cVEMP).
3. Sensorineural hearing loss, absent acoustic reflexes, lower threshold on cervical vestibular evoked myogenic potential (cVEMP).
4. **Sensorineural hearing loss, present acoustic reflexes, normal threshold on cervical vestibular evoked myogenic potential (cVEMP).**

**Difficulty Level: Moderate**

**Explanation:** This patient has a history and symptoms consistent with superior semicircular canal dehiscence (SSCD). This can result from head trauma if the bone overlying the superior semicircular canal is thin, or may be idiopathic in nature. Manifestations can include hearing loss, disequilibrium, vertigo, oscillopsia (the apparent motion of objects that are known to be stationary) evoked by loud noises and/or by maneuvers that change middle-ear or intracranial pressure (such as coughing, sneezing, or straining).

SSCD can present with a conductive hearing loss that mimics otosclerosis and could explain some cases of persistent conductive hearing loss after uneventful stapedectomy. Audiometric testing with attention to absolute bone-conduction thresholds, acoustic reflex testing, VEMP testing, laser vibrometry of the umbo, and computed tomograph scanning can help to identify patients with SSCD presenting with conductive hearing loss without vertigo.

Auditory manifestations of the syndrome include autophony (increased resonance of one’s own voice), hypersensitivity to bone-conducted sounds, and an apparent conductive hearing loss revealed on audiometry. Some patients have exclusively vestibular symptoms and signs; some have both auditory and vestibular manifestations; and still other patients have exclusively auditory complaints.

Testing will help differentiate this disorder from other causes of hearing loss and vestibular symptoms. An audiogram will show an apparent conductive hearing loss. The bone conduction thresholds can be less than 0 dB hearing level (ie supranormal bone conduction thresholds). The air-bone gap will be greatest in the lower frequencies. Acoustic reflexes will still be present which distinguishes SSCD from a conductive hearing loss due to middle ear pathology. Patients with SSCD typically have a lowered threshold on cVEMP due to a “third mobile window” which increases the sensitivity of the vestibular receptors to sound and pressure stimuli, and amplitudes will be higher.

In patients with SSCD, testing typically shows a conductive hearing loss (worse in the lower frequencies and not uncommonly with supranormal bone conduction), acoustic reflexes, and lower thresholds and higher amplitudes on cVEMP testing.

1. A 4 year old female presents with right sided congenital aural atresia. Her parents are interested in surgical correction of the canal and middle ear reconstruction. On exam she has a well formed right moderately microtic auricle and external auditory canal atresia. On computed tomography the mastoid is poorly pneumatized, there is a prominent malleus-incus complex, and the incus does not appear to be in continuity with the stapes. The stapes, round window, oval window, middle ear space and facial nerve course appear to be otherwise normal. Based on exam and imaging criteria what type of surgical candidate for atresia reconstruction is she?
2. Ideal surgical candidate
3. **Moderate surgical candidate**
4. Poor surgical candidate
5. Surgery is contraindicated
6. Cannot determine candidacy for reconstruction until she undergoes surgical exploration

**Difficulty Level: Moderate**

**Explanation:** Congenital aural atresia (CAA) is a rare congenital abnormality that is more often unilateral and associated with deformities of the pinna. CAA can be either syndromic or spontaneous. CAA reconstruction is largely based on CT findings according to the Jahrsdoerfer scoring system. This scoring system is highly correlative with postoperative hearing improvement. The scale is from 0-10 with candidates scoring >6 being surgical candidates. All factors (oval window open, aerated middle ear space, normal facial nerve, malleus-incus complex present, mastoid pneumatization, incus-stapes connection, normal round window, and appearance of the external ear) are worth one point except the presence of a stapes is counted as two points. Over the past few decades, there have been several modifications to Jahrsdoerfer’s original scoring system however none are widely accepted. Other factors that should be considered when undergoing atretic repair include incudostapedial joint angle, the height of the tegmen, the position of the facial nerve, and the size and position of the malleus-incus complex. The candidate above would receive a score of 7 given her IS joint discontinuity, microtia, and poor mastoid pneumatization. A score of 10 would indicate an excellent surgical candidate and a score of 7 would suggest a ‘good’ or moderate surgical candidate. A score of six or less would suggest that the patient cannot be adequately aided with atresia repair and a bone-anchored hearing aid may be the more optimal choice of hearing augmentation.

The Jahrsdoerfer scoring system is used to predict postoperative hearing improvement and has been used to determine a patient's suitability for repair. The scale is from 0-10 with candidates scoring >6 being surgical candidates. All factors (oval window open, aerated middle ear space, normal facial nerve, malleus-incus complex present, mastoid pneumatization, incus-stapes connection, normal round window, and appearance of the external ear) are worth one point except the presence of a stapes is counted as two points. A score of <6 suggests that a patient is not a candidate for atresia repair. Other factors to consider are incudostapedial joint angle, the height of the tegmen, position of the facial nerve, and size and position of the malleus-incus complex.

1. A 17-year-old male presents with a history of Eustachian tube dysfunction and left side hearing loss. He underwent a tympanomastoidectomy for a cholesteatoma 6 months prior. Otoscopy reveals a 30% anterior dry perforation. On Rinne testing, air conduction was greater than bone conduction with a 512 Hz fork but bone conduction was greater than air conduction with a 256 Hz fork. What is the expected level of hearing loss on the audiogram that will be seen in this patient?
2. 5-15 dB
3. **15-25 dB**
4. 25-35 dB
5. 35-45 dB
6. 45-60 dB

**Difficulty Level: Hard**

**Explanation:** An ear that can hear normally should have air conduction (sound waves which travel to the tympanic membrane and are then converted into sound in the inner ear) that is louder than bone conduction (sound transmitted through the vibration of the skull and then into the cochlea). The tuning fork test not only aids the examiner in determining the type of hearing loss present but also can indicate the degree of hearing loss. If a patient has a positive Rinne test with a 512 Hz fork but a negative test with a 256 Hz fork, hearing loss on that side is approximately 15-30 dB. A systematic review and meta-analysis examining the accuracy of tuning fork testing demonstrated a transition point (ie when Rinne went from AC>BC to BC>AC) of 20.5 dB (range 13-40 dB) for the 256 Hz fork and a transition point of 26 dB (range 17-40 dB) for the 512 Hz fork. The 1024 Hz fork was not included as part of this study, but a negative test at this frequency has been associated with a >45 dB hearing loss. Of note, some older texts will cite the following and this seems to be perpetuated despite the findings from the cited study being seemingly the most accurate to date:

A 256 Hz tuning fork has a transition point around 20.5 dB and a 512 Hz tuning fork has a transition point around 26 dB. The 1024 Hz tuning fork appears to have a much higher transition point; at least 45 dB.

1. What is the relative size of the semicircular duct of the membranous labyrinth compared with the semicircular canal of the osseous labyrinth within which it lies?
2. 10%
3. **25%**
4. 50%
5. 75%
6. 100%

**Difficulty Level: Hard**

**Explanation:** The semicircular ducts occupy a small portion of the semicircular canals of the bony labyrinth; the fluid in the ducts is termed endolymph and the duct is surrounded by the perilymph of the osseous canal.

1. A 50-year-old female is being treated in your clinic for left-sided hearing loss due to otosclerosis. Which of the following findings is most consistent with advanced otosclerosis without cochlear involvement?
2. **Weber testing which lateralizes to the left ear**
3. Supranormal bone conduction thresholds
4. Patient report of being able to hear conversation better in quiet environments
5. Family history is rarely contributory

**Difficulty Level: Moderate**

**Explanation:**  Hearing loss in otosclerosis classically begins as a conductive loss in the lower frequencies which progresses to the higher frequencies as well. It is also important to note that some cases can have cochlear involvement and therefore may have findings consistent with mixed or sensorineural hearing loss.

In patients with advanced otosclerosis regarding acoustic reflexes, the diseased ear may lose all of these reflexes, however the contralateral ear (with presumed normal hearing) would retain an ipsilateral acoustic response since the stapes/tendon are unaffected. An otosclerotic stapes is fixed in the oval window to the point where it will not respond to contraction of the stapedius muscle when loud noise is introduced to the contralateral (unaffected) ear. Similarly, loud noise presented to the otosclerotic ear may not produce a stapedial reflex in the contralateral ear because the stapes fixation in the affected ear effectively dampens the intensity of the presented sound. A “diphasic reflex pattern” can be seen in early otosclerosis where there is an increase in compliance at the onset and cessation of a sound stimulus. Eventually, this gives way to complete absence of reflexes.

A Weber test lateralizing to the affected ear supports a diagnosis of otosclerosis.

1. Which of the following otologic procedures accounts for the highest number of iatrogenic facial nerve injuries?
2. Stapedectomy
3. Canalplasty
4. Ossicular chain reconstruction
5. **Tympanoplasty and mastoidectomy**

**Difficulty Level: Moderate**

**Explanation:** In a 10 year retrospective review of iatrogenic facial nerve injuries, 17% of cases were attributed to otologic surgery. Of these cases, 82% were due to mastoid surgery (tympanoplasty/mastoidectomy, cochlear implantation, mastoid oblitaration, labyrinthectomy, endolymphatic sac shunt, glomus tumor resection), of which tympanoplasty and mastoidectomy accounted for almost half.

Tympanoplasty with mastoidectomy accounts for the highest number of iatrogenic facial nerve injuries.

1. A 75-year-old patient with a long history of diabetes mellitus presents with a 2-week history of progressive left ear pain and drainage, particularly worse at night. On physical exam, the patient has significant exudate of the external ear canal (depicted below). Otoscopy is significant for a mound of granulation tissue at the bony cartilaginous junction of the external auditory canal. A culture of the drainage will most likely grow which of the following organisms?



1. Staphylococcus aureus
2. Staphylococcus epidermidis
3. Proteus mirabilis
4. Aspergillus fumigates
5. **Pseudomonas aeruginosa**

**Difficulty Level: Easy**

**Explanation:** This patient depicts the classic presentation of malignant otitis externa (MOE). MOE is most often seen in the elderly and patients with diabetes. Symptoms include otorrhea, otalgia, and deficits of cranial nerves VII-XII. Otalgia is often worse at nighttime. A finding of mounds of granulation tissue at the isthmus of the external auditory canal (EAC) is pathognomonic for MOE.

Pseudomonas aeruginosa is the most common bacterial isolate in malignant otitis externa.

1. A 25 year-old-female presents with a unilateral vestibular schwannoma and has a family member diagnosed with Neurofibromatosis type 2. Which finding could help confirm the diagnosis?
2. COL2A1 abnormality
3. Presence of dystopia canthorum
4. Abnormal levels of treacle protein
5. **Abnormal production of merlin protein**
6. Presence of retinitis pigmentosa

**Difficulty Level: Easy**

**Explanation:** Neurofibromatosis type 2 (NF2) is an autosomal dominant condition caused by a mutation in a 17 exon gene that codes for a 595-amino acid protein named merlin on chromosome 22q12. Merlin is a tumor suppressor that regulates the actin cytoskeleton. Although its mechanism of action is not entirely understood, microarray analysis has identified numerous other genes that become deregulated during tumorigenesis.

NF2 is characterized by bilateral vestibular schwannomas, schwannomas of other cranial, spinal, and cutaneous nerves, and cranial and spinal meningiomas. Although the classic presentation is a patient with bilateral vestibular schwannomas, patients may present with unilateral schwannomas. In one study, 18% of subjects with NF2 presented with a unilateral vestibular schwannoma. Many subjects, particularly those with familial NF2, eventually developed bilateral disease.

1. Aminoglycoside ototoxicity is most commonly associated with damage to which of the following sites within the inner ear?
2. **Outer hair cells**
3. Inner hair cells
4. Stria vascularis
5. Reissner’s membrane

**Difficulty Level: Moderate**

**Explanation:** Aminoglycosides primarily damage the outer hair cells first at the basal cochlea with progression towards the apex causing high frequency sensorineural hearing loss (SNHL). Inner hair cell damage occurs at a later stage. Damage occurs when the drug forms a complex with iron forming free radicals that damage the outer hair cells and vestibular cells. Tinnitus is the most common symptoms of aminoglycoside toxicity and precedes SNHL. The overall incidence of hearing loss is 10-15%. Cisplatin causes permanent, high frequency SNHL that is bilateral & symmetric in 20% of patients by damaging the stria vascularis and basal outer hair cells. Carboplatin causes damage primarily to the inner hair cells. Diuretics, such as lasix and ethacrynic acid cause SNHL, tinnitus and vertigo by causing injury to the stria vascularis.

Aminoglycosides cause sensorineural hearing loss due to their toxic effects on the outer hair cells.

1. In the Sunderland nerve injury classification, when does Wallerian degeneration first develop?
2. Grade 1
3. **Grade 2**
4. Grade 3
5. Grade 4
6. Grade 5

**Difficulty Level: Hard**

**Explanation:** Grade 2 is axonotmesis, where the axon is transected and there is subsequent Wallerian degeneration. The endoneurial sheath is preserved, however, and each regenerating axon is confined to its original sheath.

Wallerian degeneration occurs in Sunderland grade 2 injuries and higher.

1. What structure can be found inferior to the subiculum and posteroinferior to the promontory?
2. Oval window
3. **Round window**
4. Lateral semicircular canal
5. Tensor tympani
6. Eustachian tube

**Difficulty Level: Moderate**

The round window is posterinferior to the promontory.

1. A 32-year-old male patient presents for evaluation of right-sided hearing loss. Several months ago, he was involved in a car accident and sustained an otic capsule sparing temporal bone fracture. At that time, he had a documented tympanic membrane perforation, and subsequent audiometric testing showed a conductive hearing loss without any sensorineural component. He now complains that his hearing is worse, and on examination, you noted that his perforation has healed. His audiogram today demonstrates a maximum conductive hearing loss and the tympanogram shows hyper compliance. Which of the following aspects of the physiology of the middle ear allow for the greatest amount of impedance matching?
2. Lever action of the ossicular chain
3. Phase difference between the oval and round windows
4. **Area effect of the tympanic membrane compared to the area of the stapes footplate**
5. Weight and stiffness of the ossicles

**Difficulty Level: Moderate**

**Explanation:** Impedance-matching ensures that acoustic energy is not lost as it transitions from air to a fluid wave. Were it not for this system, up to 99% of acoustic energy would be lost after arriving in the cochlea. The greatest amount of impedance matching is accomplished by the differences in the area between the tympanic membrane and the oval window. The area of the tympanic membrane is approximately 60 mm and that of the oval window Is only 3.2 mm . This leads to an approximately 20:1 increase in sound energy (26 dB). Other mechanisms for impedance matching are described below. For optimal hearing, it is certainly advantageous to have all components functioning, however in some instances, if one component is not functioning, it will severely impact hearing. For example, the patient in the vignette has a maximum conductive hearing loss likely because of ossicular discontinuity and an intact tympanic membrane. His hearing was actually better before the perforation healed because some sound was able to bypass the tympanic membrane and non-functioning ossicular chain and be transmitted directly to the inner ear through either the oval or round windows.

The ratio of the surface areas of the tympanic membrane to the oval window, the lever action of the ossicles, and the buckling of the tympanic membrane are the main mechanisms for impedance matching. Of these three, the first mechanism provides the largest contribution.

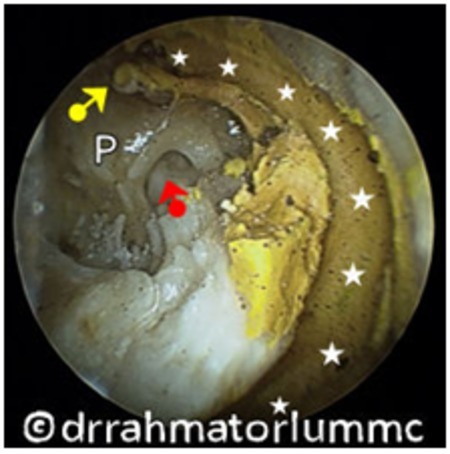
1. A 25 year old male presents with otorrhea for the last few months as demonstrated in the picture below. What is the most common microorganism source of otitis externa?
2. **Pseudomonas aeruginosa**
3. Staphylococcus auerus
4. Candida albicans
5. Actinomyces israelli
6. Enterococcus Faecalis

**Difficulty Level: Easy**

**Explanation:** According to the cited study, Pseudomonas aeruginosa accounted for 38% of all cases of otitis externa.

Pseudomonas aeruginosa is the most common bacterial cause of otitis externa.

1. Which inner ear structure terminates at the round window? (Image shows the middle ear as seen just beyond the tympanic annulus showing the opened horizontal and descending segment of left facial canal. [P- promontory, yellow arrow- capitulum of stapes head, red arrow- round window niche, stars facial tunnel])



1. **The scala tympani**
2. The scala media
3. The scala vestibule
4. The endolymphatic duct
5. The cochlear aqueduct

**Difficulty Level: Moderate**

The scala tympani which is the lower compartment of the cochlea ends at the round window.

1. A 12 month old male child presents to your office with maternal concern for hearing loss. The history is remarkable for meningitis at the age of 4 weeks. Which of the following options is true regarding hearing loss related to bacterial meningitis:
2. **It results from damage to the cochlea and spiral ganglia by purulent exudate entering through the cochlear aqueduct**
3. Cochlear ossification is a contraindication for cochlear implant
4. The incidence of post-meningitis deafness is <1%
5. Corticosteroids are not effective in reducing hearing loss in the acute phase.

**Difficulty Level: Hard**

**Explanation:** Damage to the cochlea and spiral ganglia from purulent exudate is observed. The route of spread is presumed to be through a patent cochlear aqueduct. It enters from the subarachnoid space to enter perilymph in the basal turn of the scala tympani, or by extension of inflammation into the internal auditory canal reaching the cochlear modiolus. The cochlear aqueduct is often patent in small children.

Hearing loss as a result of bacterial meningitis is due to extension of infection from either the subarachnoid space to the scala tympani via a patent cochlear aqueduct or inflammation extending to the internal auditory canal.

1. Which of the following statements regarding osteomata and exostoses of the external auditory canal is true?
2. Exostoses are typically solitary pedunculated osseous lesions that originate on the tympanosquamous or tympanomastoid suture line
3. Exostoses contain cancellous bone that contains marrow spaces and fibrovascular tissue, while osteomata have no fibrovascular channels
4. Osteomata are broad-bases osseous lesions that occur around the circumference of the medial aspect of the external auditory canal
5. **Osteomata is strongly correlated with exposure to cold water**

**Difficulty Level: Hard**

**Explanation:** Surgical removal of exostoses is usually more challenging than removal of osteomata. Generally, a postauricular approach is required while osteomata can be removed through a transcanal approach.

Osteomata, which are typically solitary and pedunculated, are much easier to remove compared to exostoses, which are more broad based and tend to occur as multiple lesions.

1. A patient with severe vertigo is referred for vestibular testing. Which of the following pathologies is associated with increased amplitudes during cervical vestibular evoked myogenic potential (cVEMP)?
2. Menieres disease
3. **Superior semi-circular canal dehiscence**
4. Otosclerosis
5. Acoustic neuroma
6. Vestibular neuronitis

**Difficulty Level: Moderate**

**Explanation:** cVEMPs are used to assess the function of the saccule and/or inferior vestibular nerve. High-intensity acoustic stimuli are used to produce electromyograms recorded from surface electrodes over the sternocleidomastoid muscles. The amplitude of response and threshold asymmetries between the right and left sternocleidomastoid muscles are assessed. VEMP thresholds are elevated (and therefore associated with diminished amplitudes) in multiple diseases, including Meniere’s disease, otosclerosis, acoustic neuromas of the inferior vestibular nerve, and vestibular neuronitis. Superior semi-circular canal dehiscence is associated with lowered cVEMP thresholds and increased amplitudes.

cVEMP testing in a patient with superior semicircular canal dehiscence should demonstarte a decreased threshold with increased amplitude.

1. A 45 year-old female undergoes a right stapedotomy for the treatment of moderate conductive hearing loss secondary to otosclerosis. Intra-operatively, the surgeon utilizes a micro-drill to perform the stapedotomy. During this part of the procedure, however, the surgeon notes that the footplate becomes fully mobile and sinks into the vestibule and is poorly visualized. What is the most appropriate next step in management?
2. Abort the procedure.
3. Pack the oval window with soft tissue and wait 6 months before returning for second attempt.
4. **Leave the footplate in the vestibule and place a prosthesis and tissue graft.**
5. Attempt to dislodge the depressed footplate with a hook.
6. Use the drill to fracture the footplate and then proceed with prosthesis and soft tissue graft placement.

**Difficulty Level: Hard**

**Explanation:** Although the management of a floating footplate during stapedotomy has become more easily manageable since the advent of laser and microdrill techniques, a depressed footplate that has sunken into the vestibule is still a challenging problem. A mere floating footplate can still be puncuted with the laser, allowing for the creating of a large enough hole to fit the prosthesis. If no laser is available, a drill can be used to create a small bur hole inferior to the anular ligament. A hook can then be used to elevate and remove the footplate followed by placement of the prosthesis and a soft tissue seal. If the footplate fractures into multiple pieces, small superficial floating fragments can be retrieved. Fragments that sink into the vestibule should be left alone. Finally, a completely subluxed footplate that has fallen into the vestibule and is poorly visualized and reachable should be left alone as attempt to retrieve it carry a high risk of sensorineural hearing loss. The surgeon should place the prosthesis as usual followed by the graft. Such a patient is more likely to have post-operative vertigo and potential re-fixation of the footplate.

If a depressed footplate is created during a stapedectomy and the footpalte is well visualized and accessible, an attempt can be made to perform a stapedotomy or remove the footplate. If it is not well visualized, it should be left alone but placement of a prosthesis with a tissue graft can still take place.

1. A 55 year-old-male patient presents to your office to evaluate hearing loss. He complains of bilateral, symmetric hearing loss over several weeks. He has no vestibular symptoms and no prior otologic history, including ear surgery, significant noise exposure, or head trauma. After obtaining his history of present illness and review of systems, you get his medical history and medication use. Which of the following statements regarding ototoxicity is true?
2. Ototoxic drugs preferentially affect inner hair cells of the apex of the cochlea
3. Aspirin ototoxicity occurs independent of dose
4. **Recovery from aspirin toxicity usually occurs 24 to 72 hours after cessation of the drug**
5. Cisplatin is preferentially toxic to inner hair cells
6. Chlorhexidine is safe to use to prep for surgery in patients with tympanic membrane perforations

**Difficulty Level: Hard**

**Explanation:** With aspirin and other salicylate use, tinnitus and reversible sensorineural hearing loss can occur. Hearing loss can be severe; however, it is expected to recover within 72 hours.

Aspirin causes reversible hearing loss, and recovery occurs within days of cessation.

1. What nerve palsies are present in Mobius syndrome?
2. CN V and VI
3. **CN VI and VII**
4. CN III, IV, V1 and VI
5. CN IX, X, and XI
6. CN X, XI and XII

**Difficulty Level: Easy**

**Explanation:** Mobius syndrome is rare congenital disorder of the rhomboencephalon (hindbrain). Cranial nerve palsies of VI and VII are core features of the syndrome, but otherwise the presentation of this syndrome can be very variable and include other cranial nerve palsies, craniofacial and limb abnormalitis, and other neurologic dysfunction. A 2019 systematic review of Mobius syndrome cases proposed 2 subtypes of Mobius syndrome: 1) Micrognathia, limb anomalies, dysphagia, 2) radiologically detectable neurologic abnormalities, developmental delay.

CN VI and VII palsies are core features of Mobius syndrome. It is also noteworthy that Mobius syndrome can also present with additional neurological, craniofacial and limb abnormalities.

1. Which of the following is true regarding retrocochlear lesions?
2. The most common retrocochlear lesion is a meningioma
3. Retrocochlear refers to lesions distal to the cochlea
4. The absence of tone decay suggests a retrocochlear lesion
5. **The absence of recruitment in the setting of an ear with sensorineural hearing loss suggests a retrocochlear lesion**

**Difficulty Level: Hard**

**Explanation:** Recruitment refers to increases in perceived loudness of sound in an ear with sensorineural hearing loss. The theory is that damaged or ineffective hair cells “recruit” adjacent hair cells with normal function, which then respond to that signal. This results in a narrowing range between the softest sound a person can hear and the loudest sound he or she can tolerate. Because the defect is with the hair cells, recruitment is a sign of cochlear pathology. Absence of recruitment in the setting of an ear with sensorineural hearing loss can be suggestive of a retrocochlear site of lesion.

The absence of recruitment in a patient with sensorineural hearing loss suggests a retrocochlear pathology. The presence of recruitment suggests a cochlear pathology.

1. Approximately what increase in dB is required to double sound intensity?
2. 2dB
3. **3 dB**
4. 6 dB
5. 10 dB
6. 50 dB

**Difficulty Level: Hard**

**Explanation:** Sound intensity is defined as the sound power per unit area. It is an objective measurement of sound intensity in the air at a listener's location, usually in watts/cm^2. Many sound intensity measurements are made relative to a standard threshold of hearing intensity( I0). Sound intensity is usually described in decibels, which measure the ratio of a given intensity I to the threshold of hearing intensity, which takes the value 0 dB. Intensity differs from loudness because the sensitivity of the ear is taken into account when measuring loudness. The intensity of a sound wave depends on the pressure of the wave, the density of the medium and the speed of sound in the medium. Higher density and higher sound speed both give a lower intensity.

The methods to calculate decibels for sound intensity and sound pressure are similar in that they are based on a logarithmic scale, however because sound pressure is related ot the square root of sound intensity, the formulas do have important differences. To determine the decibels for sound intensity, the formula is X dB=10log

1. What structure receives the crura from the semicircular canals?
2. Saccule
3. **Vestibule**
4. Endolymphatic sac
5. Scala vestibule
6. None of the above

**Difficulty Level: Moderate**

The vestibule receives the crura of the semicircular canals.

1. Which abnormal finding is associated with the most common infectious cause of congenital hearing loss in the US?
2. sores around the mouth and eyes
3. Hennebert's sign
4. **cerebral calcifications**
5. cataracts

**Difficulty Level: Moderate**

**Explanation:** Cytomegalovirus (CMV) is the most common infectious cause of congenital hearing loss in the United States. Periventricular calcifications, hepatosplenomegaly, jaundice, and intellectual disability are common findings.

Cytomegalovirus is the most common infectious cause of congenital hearing loss in the United States. Cerebral calcifications on CT are a common finding.

1. Where is the facial nerve most commonly stimulated in patients who develop hemifacial spasm after cochlear implantation?
2. Chorda tympani
3. **Labrynthine segment**
4. Tympanic segment
5. Pes anserinus

**Difficulty Level: Hard**

Facial nerve stimulation is a potential complication of cochlear implantation that can produce hemifacial spasms and facial pain. The risk of facial nerve stimulation following implantation is generally higher in patients with congenital cochlear abnormalities and otosclerosis. In the cited study, patients who developed facial nerve stimulation post implantation had radiographic evidence showing that the labyrinthine portion of the facial nerve was in closer proximity to the basal turn of the cochlea compared to patients without facial nerve stimulation. Most cases of facial nerve stimulation can be resolved with reprogramming of the device, with a minority of patients requiring explantation or replacement of the electrode.

1. A 43-year-old male with a history of chronic otitis media presents to the ED with persistent right-sided otalgia and fevers, despite antibiotic therapy. His vital signs on arrival are HR 103, BP 119/92, SaO2: 100%, T 101.7 F. On physical exam, he has purulent otorrhea and a right-sided lateral rectus palsy. Fundoscopic exam is normal. A CT scan confirms your suspicion. Which of the following syndromes is present?
2. Orbital apex syndrome
3. Vernet syndrome
4. Lemierre syndrome
5. Villaret syndrome
6. **Gradenigo'™s syndrome**

**Difficulty Level: Easy**

**Explanation:** Gradenigo’s syndrome is the name given to a triad of symptoms classically associated with petrous apicitis. The symptoms are deep facial pain, otitis media, and ipsilateral abducens nerve palsy. The etiology is thought to be through direct extension from the mastoid or middle ear through pneumotized air cell tracts. The petrous apex is divided into anterior and posterior. The posterior petrous apex is pneumotized in 30% of patients and the anterior petrous apex is pneumotized in 10% of patients. Treatment of petrous apicitis is aimed at eradicating the infection. First line management is topical and systemic antibiotics. If the patient fails medical management surgical drainage of the infection may be necessary.

Gradenigo’s syndrome describes the triad of symtpoms of deep facial pain, otitis media, and ipsilateral abducens nerve palsy. It is due to petrous apicitis, and usually a complication of otitis media.

1. A 41-year-old woman presents for management of progressive hearing loss. An audiogram shows a moderate mixed hearing loss with 25 dB drop in the bone conduction line at 2000 Hz. After discussion, the patient elects to proceed with a middle ear exploration. Which of the following is considered a relative contraindication to stapedectomy?
2. Active Menieres disease
3. Tympanic membrane perforation
4. **Prior surgery without perceived benefit**
5. Only hearing ear
6. Active otitis media

**Difficulty Level: Hard**

**Explanation:** This patient likley has otosclerosis and may benefit from a stapedectomy or stapedotomy. An experienced surgeon can expect to be successful at closing the air-bone gap to within 10 dB 95% of the time in primary cases. Revision cases have a success rate of 50-80% in experienced hands and prior surgery alone should not be considered an absolute contraindication. The remaining answer choices are absolute contraindications to surgery.

A prior unsuccessful stapedectomy is a relative, but not absolute, contraindication for future attempts at a stapedectomy.

1. Which of the following surgical techniques would definitely fail to resolve vestibular symptoms in a patient with Meniere’s Disease?
2. Transtympanic aminoglycoside injections
3. Endolymphatic sac surgery
4. Vestibular nerve section
5. **Occlusion of superior semicircular canal**
6. Labyrinthectomy

**Difficulty Level: Easy**

**Explanation:** Occlusion of the superior semicircular canal has been described for patient’s with superior semicircular canal dehiscence, but would not help a patient with Meniere’s Disease.

Intratympanic gentamicin injection, endolymphatic sac decompression/shunting, vestibular nerve sectioning and labyrinthectomy are all acceptable ablative therapies for recalcitrant Meniere's disease.

1. A 47-year-old male patient is complaining that his voice is louder in the left ear. He also notes that during severe storms he feels like the world is spinning. Dix-hallpike is negative. His tympanic membrane is intact without effusions. On pneumatic otoscopy he becomes acutely vertiginous. Audiometric testing shows a conductive hearing loss in the left ear as well as decreased vestibular evoked myogenic potential (VEMP) threshold. What is the likely diagnosis?
2. **Superior canal dehiscence**
3. Benign paroxysmal positional vertigo (BPPV)
4. Endolymphatic hydrops
5. Malingering
6. Vestibular schwannoma

**Difficulty Level: Easy**

**Explanation:** This patient’s symptoms are classic for superior semicircular canal dehiscence syndrome (although it was only described in 1998). The pathology involves bony dehiscence overlying the semicircular canal. This creates a third window (with the round and oval windows being the normal two) allowing for pressure waves to pass through the vestibular organ. The normal path of least resistance is around the cochlea with pressure waves leaving through the round window. In this syndrome, the path of least resistance is through the third window which causes stimulation of the vestibular system. Tullio’s phenomenon is sound induced vertigo which is what is happening with the thunder during violent thunderstorms. Hennebert sign is pressure-induced vertigo which is what is happening with the pneumatic otoscopy induced vertigo. These patients also have a conductive hearing loss from the third window and VEMPs with a low threshold but high amplitude because of the lower amount of energy required to stimulate the vestibular system. They may also have supranormal bone conduction thresholds, and have complaints like they can hear their eyes move or hear their own quiet breathing.

Superior semicircular canal dehisence often presents with conductive hearing loss and noise and pressure induced vestibular symptoms. Audiometric testing may show supranormal bone conduction and VEMPs characteristically show a low threshold and high amplitude.

1. A 58 year-old male with bilateral moderate sloping to severe sensorineural hearing loss (SNHL) presents for follow-up. As you enter the room, you greet the patient and ask, “how was your vacation?” The patient responds “I’m sorry I couldn’t understand you, could you speak louder?” As you repeat the question at a moderately higher volume the patient responds, “Ok, you don’t have to shout though!” Which type of phenomenon is the patient experiencing?
2. Diplacusis
3. **Recruitment**
4. Rollover
5. Paracusis Willisii
6. Hyperacusis

**Difficulty Level: Moderate**

**Explanation:** Loudness recruitment is a phenomenon in which patients with hearing loss experience a much more rapid increase in subjective loudness of a stimulus. A patient with a 50 dB hearing loss can barely hear a sound at 55 dB but may experience a 100 dB sound as loud as a person with normal hearing. Recruitment is indicative of a cochlear pathology.

Recruitment describes an abnormal, rapid increase in sound perception, and is indicative of a cochlear pathology.

1. A 2-year-old female is referred to clinic for evaluation of hearing loss. An audiogram shows a left side 15 dB conductive hearing loss with type A tympanograms. Otoscopy is unremarkable however imaging suggests an abnormality of the stapes. What branchial arch forms the stapes superstructure?
2. First
3. **Second**
4. Third
5. Fourth
6. Fifth

**Difficulty Level: Easy**

**Explanation:** The Reichert cartilage of the second branchial arch forms bony structures proximally and distally. Its central portion withers, leaving a fibrous band—the stylohyoid ligament. Proximally, it forms the styloid process and stapes suprastructure. Distally (anteroinferiorly), the second arch cartilage forms the superior portion of the body and the lesser cornu of the hyoid bone. The manubrium of the malleus and long process of the incus may also be 2nd arch derivatives however this is controversial (ie dual arch vs classical theory).

The stylohyoid ligament, stapes superstructure, body, and lesser cornu of the hyoid are second branchial arch structures.

1. A child presents to the ED with fever and a left posterior periauricular painful mass for three days. Her mother reports that she has been complaining of hearing loss, and feeling dizzy. Other than well-child checks, she has never been to the doctor. The ear is shown below. Which of the following is true regarding the patient's suspected diagnosis?



1. The organism most often isolated is Pseudomonas aeruginosa
2. It is most commonly encountered in children older than two years of age
3. When imaging is indicated the test of choice is a computed tomography (CT) without contrast
4. **The initial antimicrobial regimen should include intravenous vancomycin**
5. It is mostly commonly occurs as a complication of acute otitis externa

**Difficulty Level: Hard**

**Explanation:** This patient has the characteristic findings of mastoiditis. The diagnosis of acute mastoiditis is usually made clinically in most patients with characteristic findings. The most frequently used diagnostic criteria include fever, otalgia, post-auricular erythema, swelling, fluctuance, or mass, and proptosis of the ear. In children with the classic clinical findings of acute mastoiditis, imaging is not necessary to make the diagnosis. However, if there are not characteristic findings imaging is helpful in determining the extent of disease and in surgical planning for children with coalescent mastoiditis. In addition, imaging should be performed if there are clinical findings suggestive of extracranial complications (such as postauricular mass, mass in the neck, cranial nerve deficits, nystagmus, retro-orbital pain, hearing loss, tinnitus, or vertigo).

Treatment of mastoiditis requires intravenous antimicrobial therapy. The choice of initial agent is dependent upon whether there is a preceding history of recurrent acute otitis media or recent antibiotic therapy. For children without a history of recurrent episodes of acute otitis media or recent antibiotics the recommended first-line agents are vancomycin or linezolid (one or the other, not both). An antibiotic active against P. aeruginosa should be added if the patient has had recurrent episodes of otitis media, or there has been recent antibiotic use.

Treatment of mastoiditis without a history of AOM should include vancomycin or linezolid, and in patients with a history of AOM, and anti-psuedomonal antibiotic should be added.

1. Which of the following statements with regards to microtia is correct?
2. Females are more commonly affected than males
3. Bilateral involvement is more common than unilateral involvement
4. **The right ear is more commonly affected than the left**
5. In the US, the incidence is highest among Caucasians
6. Approximately 10% of patients with microtia will have associated congenital anomalies

**Difficulty Level: Hard**

**Explanation:** Microtia is a congenital deformity of the external ear and ranges in severity from complete absence of an auricle (stage IV microtia or anotia) to mild diminution in auricular size (stage I microtia). For reasons unknown, the right ear is more commonly affected than the left by a 3:2 ratio.

The right ear, males, Latin or Native Americans and unilateral involvement are most common in cases of microtia.

1. Cisplatin ototoxicity is related most to which of the following?
2. rate of infusion
3. age of the patient
4. frequency of dosing
5. overall dose
6. **both the age of the patient and overall dose**

**Difficulty Level: Hard**

**Explanation:** Cisplatin is a common chemotherapeutic agent used to treat a number of malignancies. Its mechanism of action is to crosslink DNA, thereby interfering with DNA replication and inducing DNA repair mechanisms, which then induce apoptosis. Common side effects include ototoxicity, nephrotoxicity, and neurotoxicity. Ototoxicity is more likely in patients younger than 5 years. It is also related to the cumulative dose of cisplatin delivered.

Ototoxicity from cisplatin is related to the age of the patient, with patients younger than 5 years being affected more, and the cumulative dose of cisplatin delivered.

1. During an endolymphatic sac decompression surgery you complete the mastoidectomy portion of the procedure and identify all of your major landmarks. You then use Donaldson’s line to estimate the position of the endolymphatic sac during the transmastoid approach. What major landmark is Donaldson’s line based off of?
2. sigmoid sinus
3. Koerner’s septum
4. Incus
5. **horizontal semicircular canal**
6. malleus

**Difficulty Level: Easy**

**Explanation:** Donaldson’s line is an imaginary line drawn posteriorly through the plane of the horizontal semicircular canal. The endolymphatic sac is generally found along this line or just inferior to it. It lies posterior to the mastoid facial nerve, anterior to the sigmoid sinus, and inferior to the antrum. Once the retrofacial air cells have been removed the sac should be visible in the posterior fossa. The main indication for endolymphatic sac decompression is persistent ongoing vertigo in Meniere’s disease despite medical therapy.

Donaldson’s line is an imaginary line drawn posteriorly through the plane of the horizontal semicircular canal, and approximates the location of the endolymphatic sac.

1. With regards to the diagnosis and management of tinnitus, which of the following is true?
2. Gingko biloba should be considered as a treatment option for persistent, bothersome tinnitus
3. **Audiologic evaluation should be performed for patients with unilateral tinnitus, or persistent tinnitus greater than 6 months**
4. Imaging studies should be routinely obtained in patients even in those without localization to one ear or nonpulsatile features
5. Anxiolytic medication should be considered as a treatment option for persistent, bothersome tinnitus.

**Difficulty Level: Easy**

**Explanation:** Per Clinical Practice Guidelines published by the American Academy of Otolaryngology Head and Neck Surgery: Strong recommendations were made against obtaining imaging studies of the head and neck in patients with tinnitus, specifically to evaluate tinnitus that does not localize to 1 ear, is nonpulsatile, and is not associated with focal neurologic abnormalities or an asymmetric hearing loss. The guideline makes the following recommendations: Clinicians should (a) perform a targeted history and physical examination at the initial evaluation of a patient with presumed primary tinnitus to identify conditions that if promptly identified and managed may relieve tinnitus; (b) obtain a prompt, comprehensive audiologic examination in patients with tinnitus that is unilateral, persistent (≥ 6 months), or associated with hearing difficulties; (c) distinguish patients with bothersome tinnitus of recent onset from those with persistent symptoms (≥ 6 months) to prioritize intervention and facilitate discussions about natural history and follow-up care; (d) educate patients with persistent, bothersome tinnitus about management strategies; (e) recommend a hearing aid evaluation for patients who have persistent, bothersome tinnitus associated with documented hearing loss; and (f) recommend cognitive behavioral therapy to patients with persistent, bothersome tinnitus. The panel recommended against (a) antidepressants, anticonvulsants, anxiolytics, or intratympanic medications for the routine treatment of patients with persistent, bothersome tinnitus; (b) Ginkgo biloba, melatonin, zinc, or other dietary supplements for treating patients with persistent, bothersome tinnitus; and (c) transcranial magnetic stimulation for the routine treatment of patients with persistent, bothersome tinnitus.

Audiometric evaluation is a key part of the evaluation of the patient with tinnitus, and should be obtained for patients with unilateral or persistent tinnitus lasting 6 months or more and patients with other hearing related complaints. Imaging is not indicated for tinnitus that does not localize to 1 ear, is nonpulsatile, or is not associated with focal neurologic abnormalities or an asymmetric hearing loss.

1. A 52 year old female presents with a 4 month history of gradually worsening right sided hearing loss. Audiogram confirms a severe sensorineural hearing loss. An MRI demonstrates a right sided cerebellopontine angle mass that is isointense to brain on T1 and T2 and moderately enhances with contrast. The mass has a broad base of dural enhancement. What is the most likely diagnosis?
2. Cholesteatoma
3. **Meningioma**
4. Cholesterol granuloma
5. Dermoid cyst
6. Vestibular schwannoma

**Difficulty Level: Easy**

**Explanation:** Meningiomas are isointense on T1 and T2 and enhance with contrast, though typically not as strongly as vestibular schwannomas. They also have a characteristic broad based dural attachment.

Meningiomas are isointense on T1 and T2, enhance with contrast, and have a broad based dural attachment.

1. Which of the following statements regarding management of tinnitus is correct?
2. **Clinicians should obtain prompt, comprehensive audiologic examination in patients who present with tinnitus that is unilateral, persistent, or associated with hearing difficulties**
3. Clinicians should obtain imaging studies in a patient with bilateral nonpulsatile tinnitus and symmetric sensorineural hearing loss
4. Clinicians should recommend a hearing aid evaluation for patients with intermittent tinnitus
5. Clinicians should always recommend antidepressants or anxiolytics for treating tinnitus

**Difficulty Level: Easy**

**Explanation:** Per Clinical Practice Guidelines published by the American Academy of Otolaryngology Head and Neck Surgery: Strong recommendations were made against obtaining imaging studies of the head and neck in patients with tinnitus, specifically to evaluate tinnitus that does not localize to 1 ear, is nonpulsatile, and is not associated with focal neurologic abnormalities or an asymmetric hearing loss. The guideline makes the following recommendations: Clinicians should (a) perform a targeted history and physical examination at the initial evaluation of a patient with presumed primary tinnitus to identify conditions that if promptly identified and managed may relieve tinnitus; (b) obtain a prompt, comprehensive audiologic examination in patients with tinnitus that is unilateral, persistent (≥ 6 months), or associated with hearing difficulties; (c) distinguish patients with bothersome tinnitus of recent onset from those with persistent symptoms (≥ 6 months) to prioritize intervention and facilitate discussions about natural history and follow-up care; (d) educate patients with persistent, bothersome tinnitus about management strategies; (e) recommend a hearing aid evaluation for patients who have persistent, bothersome tinnitus associated with documented hearing loss; and (f) recommend cognitive behavioral therapy to patients with persistent, bothersome tinnitus. The panel recommended against (a) antidepressants, anticonvulsants, anxiolytics, or intratympanic medications for the routine treatment of patients with persistent, bothersome tinnitus; (b) Ginkgo biloba, melatonin, zinc, or other dietary supplements for treating patients with persistent, bothersome tinnitus; and (c) transcranial magnetic stimulation for the routine treatment of patients with persistent, bothersome tinnitus.

Imaging studies should only be obtained in patients with tinnitus that localizes to on ear, pulsatile tinnitus, asymmetric hearing loss, or focal neurologic deficits. Clinicians should recommend a hearing aid evaluation for patients with hearing loss and persistent, severe tinnitus. Clinicians should not routinely recommend antidepressants or anxiolytics for treating tinnitus.

1. A 15 year old male presents with a 2 week history of progressively worsening left ear pain and drainage. Symptoms started shortly after returning home from a weekend trip to a lake. Otoscopy reveals an erythematous and edematous ear canal that is tender to manipulation. Which of the following statements about ototopical therapies to treat this problem is true?
2. Neomcyin and polymyxin B plus hydrocortisone is more effective than ciprofloxacin plus dexamethasone
3. Acetic acid alone is just as effective as acetic acid plus corticosteroid
4. **The addition of a steroid to flouroquinolone agents decreases the symptomatic period**
5. Acetic acid alone has a similar recurrence rate as an antibiotic plus corticosteroid

**Difficulty Level: Easy**

**Explanation:** The addition of a steroid to fluoroquinolone agent decreases the symptomatic period, albeit shortly by 0.8 days.

The addition of a steroid to a fluoroquinolone agent slightly shortens the symptomatic period.

1. A 40-year-old female undergoes a stapedectomy and placement of a titanium piston prosthesis for conductive hearing loss due to otosclerosis. She is initially pleased with the results of her surgery and reports a subjective improvement in her hearing, confirmed with tuning fork testing. Four weeks after surgery, the patient returns complaining of vertigo and progressive hearing loss in the same ear. Tuning fork testing suggests a sensorineural hearing loss. Which of the following is the most likely cause of her symptoms?
2. Obliterative otosclerosis of the round window
3. Formation of middle ear adhesions
4. **Reparative granuloma of the oval window**
5. Necrosis of the long process of the incus
6. Displacement of the prosthesis

**Difficulty Level: Hard**

**Explanation:** Reparative granuloma is an uncommon complication of stapes surgery. In cases of reparative granuloma, hearing after stapes surgery is improved, but gradually or suddenly becomes worse in the 1st to 6th postoperative week. The hearing loss associated with reparative granuloma is classically sensorineural (but can be mixed or conductive), sudden onset, and is frequently associated with vertigo and tinnitus. Treatment consists of steroids and antibiotics, or revision surgery.

A reparative granuloma following a stapedectomy is associated with hearing loss (usually sensorineural but can be mixed or conductive), vertigo, and tinnitus. It is a rare complication, and typically occurs 1-6 weeks after surgery.

1. Which of the following syndromes is inherited in an autosomal recessive pattern?
2. Apert Syndrome
3. Stickler Syndrome
4. **Jervell / Lange-Nielsen Syndrome**
5. Waardenburg Syndrome
6. Treacher Collins Syndrome

**Difficulty Level: Moderate**

**Explanation:** Jervell/Lange-Nielsen syndrome is inherited in autosomal recessive fashion. It is associated with long QT syndrome and bilateral severe sensorineural hearing loss due to mutations in potassium channel genes. The most common mode of transmission for hereditary deafness is autosomal recessive fashion, with connexin 26 mutations accounting for the majority of non syndromic cases and Usher syndrome accounting for the majority of syndromic cases.

Jervell/Lange-Nielsen syndrome is an autosomal recessive cause of sensorineural hearing loss.

1. A 42-year-old female with a history of type 2 diabetes presents to the clinic with 3 months of decreased hearing on both sides. Patient denies vertigo and fevers. She feels like there is ringing in her ears, and she is "hearing underwater." On tuning fork exam Weber is midline and bone conduction is greater than air conduction bilaterally. Otoscopy is shown in the picture below. Which of the following is true regarding this type of effusion?



1. A bacterial pathogen is identified in the majority of middle ear effusions of those with OME
2. When patients are symptomatic, the predominant symptom is vertigo
3. Tympanometry is helpful in differentiating an infectious from a non-infectious cause.
4. Acoustic reflectometry is helpful in differentiating an infectious from a non-infectious cause.
5. **Bacterial biofilms have a major role in the pathogenesis**

**Difficulty Level: Moderate**

**Explanation:** This patient´s presentation is consistent with otitis media with effusion (OME), sometimes referred to as serous otitis. The pathogenesis of OME is not well understood but appears to be the result of a variety of inciting events involving inflammation and tissue hyperplasia. Bacterial biofilms are thought to play a major role in the pathogenesis of OME. Biofilms consist of bacterial cells within a biopolymer matrix. This matrix provides a potential means of antimicrobial resistance, and can potentially create a communal system by sharing nutritents and other biological materials. Despite commonly being culture-negative at myringotomy, immunofluorescence studies have demonstrated bacterial DNA (most commonly from Haemophilus influenzae).

Bacterial biofilms are significant factors in the pathogenesis of otitis media with effusion, and promote antimicrobial resistance.

1. Regarding the blood supply to the vestibular end organs, which of the following is accurate?
2. The posterior cerebellar artery is a major source of blood supply
3. The posterior vestibular artery supplies the utricle
4. **The anterior vestibular artery supplies the utricle**
5. The posterior vestibular artery does not supply the majority of the saccule

**Difficulty Level: Hard**

**Explanation:** The anterior vestibular artery supplies most of the utricle, superior and horizontal semicircular canals and a small portion of the saccule.

1. A 42-year-old female presents to the emergency room for evaluation of acute-onset vertigo. The vertigo is described as an intense spinning sensation associated with nausea and vomiting and started the night before presentation. If present on exam, which of the following would support a diagnosis of left vestibular neuritis?
2. Intense vertigo lasting several minutes with head rotation.
3. Associated sudden tinnitus and hearing loss with vertigo.
4. **Spontaneous nystagmus with fast-phase component towards the right.**
5. MRI brain demonstrating lesion involving the left vestibular nucleus.
6. Inability to stand and dysdiadochokinesia.

**Difficulty Level: Moderate**

**Explanation:** Patients with vestibular neuritis often have severe spontaneous vertigo that lasts days to weeks. Unlike labyrinthitis, there is no associated hearing loss. The visible nystamus is typically horizontal and rotatory and directed with a fast-phase away from the affected side. As the patient recovers from the acute phase, it is not uncommon for persistent symptoms of disequilibrium or positional vertigo to persist over the long term.

An acute episode of unilateral vestibular neuritis typically presents with the fast-phase of nystagmus directed away from the affected side. Other key differentiating factors include acute onset, typically preceded by an upper respiratory infection, severe/unremitting symptoms, and absence of hearing loss.

1. A 52 year old male is taken to the operating room for stapedotomy for otosclerosis. He is otherwise healthy and has a 40dB preoperative unilateral conductive hearing loss. Intraoperatively, during placement of a bucket handle prosthesis the incus becomes hypermobile. It is reset into its original position however manipulation shows it to be moderately displaced with any manipulation. How should this be managed?
2. Proceed with stapedotomy and prosthesis placement as planned
3. Switch to a nitinol wire prosthesis
4. Remove the incus and malleus and place a total ossicular replacement prosthesis
5. Use bone cement to fix the incus to the malleus and proceed with the stapedotomy procedure
6. **Abort the procedure and return in six months**

**Difficulty Level: Hard**

**Explanation:** Completion of the ossicular chain reconstruction may occur in 4-6 months once the incus has reattached to the malleus. Placement of any prosthesis may result in permanent dislocation of the incus requiring revision ossiculoplasty with poorer hearing outcomes.

Incus dislocation precludes routine stapedotomy and if encountered should be cause for aborting the procedure.

1. A 3-year-old male presents with concern for hearing loss from his parents. His speech is significantly impaired compared to his peers and he has developed significant behavioral problems at his day care. He has a history of recurrent acute otitis media with tympanostomy tube placement. The tubes are confirmed to be in good position and patent. A hearing test is attempted but he is noncompliant and no useful information can be gathered from the testing. He subsequently undergoes auditory brainstem response testing which reveals an abnormal interaural difference at wave VI. What structure does this wave correlate to?
2. Proximal eighth nerve
3. **Inferior colliculus**
4. Cochlear nuclei
5. Superior olivary nucleus
6. Medial geniculate body

**Difficulty Level: Moderate**

**Explanation:** Auditory brainstem response (ABR) testing functions by using electrodes placed on the vertex and mastoid to detect electrical signal changes in response to unilateral auditory ‘clicks’. This clicks are administered as 100 microsecond bursts of sound that excite the neurons along the auditory path. Up to seven waves correlating to different intracranial neurologic structures are produced at 1ms intervals after the click is administered. The following waves are correlated with the following neurologic structures:

A useful mnemonic to help aid in the interpretation of waveforms in the auditory brainstem response is E COLI: I Proximal Eighth nerve (vestibulocochlear nerve), II distal Eigth nerve/Cochlear nucleus, III Cochlear nucleus/superior Olivary complex, IV superior Olivary complex, V Lateral lemniscus, and VI-VII Inferior colliculus.

1. The utricle senses which of the following?
2. **linear acceleration in a horizontal plane**
3. linear acceleration in a vertical plane
4. rotational acceleration in the plane of the posterior semicircular canal
5. rotational acceleration in the plane of the superior semicircular canal

**Difficulty Level: Moderate**

**Explanation:** The otolithic organs (utricle and saccule) sense linear movements.

The utricle senses linear acceleration in a horizontal plane.

1. A 28-year-old woman is referred to her otologist for right-sided hearing loss. Audiogram shows a mixed hearing loss. Acoustic reflexes are shown below. What is the most likely diagnosis?
2. Meniere’s disease
3. Otosclerosis
4. Superior semi-circular canal dehiscence
5. Vestibular schwannoma
6. **Otosyphillis**

**Difficulty Level: Moderate**

**Explanation:** Classically, early otosclerosis demonstrates a pathognomonic “on-off” pattern, whereby a brief increase in compliance is noted at the onset and termination of the stimulus. As the disease progresses, thresholds increase ipsilaterally, then contralaterally, before disappearing completely. For most patients, acoustic reflexes are absent at presentation. Due to stiffness at the oval window, stimulus presented to the affected ear would not produce a reflex ipsilaterally or contralaterally. Similarly, ossicular fixation would not permit reflexes to be measured when the contralateral ear is stimulated (though the reflexes in the stimulated ear should be present, assuming the disease is unilateral).

Acoustic reflexes are frequently disrupted by otosclerosis, and depending on the severity of disease, several different findings may be observed.

1. Which of the following is part of the pathway of sympathetic innervation to the nerve of the pterygoid canal proximal to the sphenopalatine ganglion?
2. Superior salivatory nucleus
3. Nervus intermedius
4. Greater superficial petrosal nerve
5. **Deep petrosal nerve**
6. Zygomaticotemporal nerve

**Difficulty Level: Hard**

**Explanation:** Sympathetic fibers from the carotid plexus form the deep petrosal nerve, which then joins the greater superficial petrosal nerve (GSPN) to form the nerve of the pterygoid canal.

The general visceral efferents that join the deep petrosal nerve as the GSPN originate from the superior salivatory nucleus in the pons, travel through the nervus intermedius to the geniculate ganglion which gives rise to the GSPN. The GSPN joins the deep petrosal nerve to form the nerve of the pterygoid canal, which travels to the sphenopalatine ganglion. The parasympathetic fibers then travel with the zygomaticotemporal nerve up to the lacrimal gland as well as send fibers to the nasal mucosa.

The nerve of the pterygoid canal receives its sympathetic fibers from the deep petrosal nerve arrising from the carotid plexus.

1. A 42 year old male presents with a newly diagnosed acoustic neuroma. He has mild ipsilateral hearing loss, no facial nerve weakness, and no history of other otologic disorders. On MRI the tumor is <1cm and is 90% intracanalicular. Upon review of treatment options he desires to undergo surgical excision. What is the best approach to the tumor in this patient?
2. Transcochlear
3. Translabyrinthine
4. Retrosigmoid
5. **middle fossa**
6. suboccipital

**Difficulty Level: Moderate**

**Explanation:** There are a variety of approaches to the cerebellopontine angle (CPA) tumors and preoperative status and patient age weigh heavily into decision making as to the most appropriate approach for each patient. The middle fossa approach is best used for patients with good preoperative hearing and small primarily intracanalicular tumors. This approach ensures adequate exposure to the internal auditory canal but not the cerebellopontine angle, as well as providing an attempt at hearing preservation. Caution with this approach in the geriatric population should be used given the fragility of the dura and need for temporal lobe retraction.

The middle cranial fossa approach for CPA tumor excision is best used for patients with serviceable hearing and primarily intracanalicular tumors.

1. A 42 year old female with a vestibular schwannoma asks about risk of long term dizziness. Which of the following is a risk factor of having long term dizziness in patients with vestibular schwannoma?
2. History of seizures
3. **History of pretreatment dizziness**
4. Male gender
5. Small tumor size
6. Young age at diagnosis

**Difficulty Level: Easy**

**Explanation:** In vestibular schwannoma patients, pretreatment variables associated with greater dizziness handicap included female sex, older age, larger tumor size, preexisting diagnosis of headache or migraine, and symptoms of dizziness predating treatment. Significant post-treatment features strongly associated with poor long-term Dizziness Handicap Inventory scores included frequency and severity of ongoing headache. On multivariable analysis, treatment modality (observation, stereotactic radiosurgery, microsurgery) did not influence long-term dizziness handicap.

Female sex, older age, larger tumor size, preexisting diagnosis of headache or migraine, and symptoms of dizziness predating treatment are all associated with a greater risk of long term dizziness in patients with vestibular schwannoma.

1. A 45 year old male presents to your clinic with a complaint of progressive right sided hearing loss. History, physical exam, and audiometric testing are consistent with right sided otosclerosis with a 35 dB air-bone gap. He is offered stapedectomy for correction of his hearing loss. He desires to undergo the procedure but inquires as to whether he should undergo local or general anesthesia for the procedure. The main advantage of local over general anesthesia is:
2. Decreased postoperative pain
3. Decreased incidence of severe sensorineural hearing loss
4. Greater ease of surgery
5. Higher rates of air-bone gap closure
6. **Early detection of vestibular symptoms**

**Difficulty Level: Easy**

**Explanation:** To date there have been three studies and one systematic review evaluating local versus general anesthesia during primary stapes surgery and their effect on hearing outcomes and complications. All three studies concluded almost equivalent rates of air-bone gap closure to within 10 dB and no significant risk of severe sensorineural hearing loss with either method. Postoperative pain has not been evaluated between the two methods but it stands to reason the postoperative pain would be similar given virtually identical procedures. Surgery is technically easier under general anesthesia because analgesia and patient movement are removed from the equation. The greatest benefit is seen when patients experience vertigo following prosthesis placement under local anesthesia. In these situations if the vertigo does not resolve during the operation the prosthesis can be adjusted until symptoms abate.

The main advantage of performing a stapedectomy under local anesthesia as opposed to general anesthesia is the ability to detect vestibular symptoms intra-op and make adjustments to the position of the prosthesis accordingly.

1. Endolymphatic sac tumors are associated with which of the following conditions?
2. Neurofibromatosis
3. Sturge-Weber syndrome
4. **von Hippel-Lindau disease**
5. Osler-Weber-Rendu disease
6. Churg-Strauss syndrome

**Difficulty Level: Moderate**

**Explanation:** von Hippel-Lindau disease is characterized by multiple hemangioblastomas of the retina and central nervous system accompanied by renal cysts, clear cell renal cell carcinoma, pheochromocytoma, and endolymphatic sac tumor. Around 10% of patients with von Hippel-Lindau disease have endolymphatic sac tumor. Therefore it is prudent to screen for this in these patients.

Approximately 10% of patients with von Hippel-Lindau disease will have an endolymphatic sac tumor.

1. Directional preponderance is calculated from which of the following?
2. Electrocochleography
3. Auditory brainstem response
4. Speech detection threshold
5. **Caloric testing**

**Difficulty Level: Moderate**

**Explanation:** Caloric testing is an essential part of the battery of tests performed during a formal vestibular evaluation, and is one of the few tests that can evaluate each side independently. In caloric testing the , horizontal semicircular canal is either stimulated or inhibited (warm water/air or cold water/air, respectively), and eye movements are recorded before, during and after irrigation. Computer algorithms are then able to calculate and graph values for the quick and slow components of the resulting nystagmus. This data can then be used to calculate the directional preponderance and unilateral weakness. The directional preponderance is a measure of the difference in total eye speeds between slow phases on each side. Unilateral weakness is the absolute value of the difference in total eye speeds for each side divided by the overall total.

Caloric testing data can be used to generate directional preponderance and unilateral weakness measures.

1. A 38-year-old male returns to the ENT clinic 6 weeks after undergoing a left-sided stapedectomy for otosclerosis. He reports that initially after the surgery his hearing on the left was great. Now, however, the patient states his hearing is bad as it has ever been. The tuning fork exam reveals bone conduction is greater than air conduction on the left and Weber lateralizes to the left. What is the most common cause of failure following stapes surgery?
2. Granulation tissue
3. Incus erosion
4. Bony regrowth.
5. **Displaced prosthesis**
6. Perilymphatic fisula

**Difficulty Level: Moderate**

**Explanation:** The House Ear Group revealed an overall rate of air-bone gap closure to within 10 db and 20 db, 60%, and 78% of the time respectively. The most common cause of primary stapes surgery failure is displaced prosthesis (53%) followed by incus erosion (26%) and bony regrowth (14%). The latter 2 causes often occur well after surgery, and would not be likely 6 weeks past this patient's procedure.

A displaced prosthesis is the most common cause of primary stapes surgery failure.

1. Which diagnostic tool may be helpful in the diagnosis of Usher syndrome?
2. **Electroretinography**
3. EKG
4. Perchlorate test
5. Serum BUN
6. Presence of cleft palate

**Difficulty Level: Easy**

**Explanation:** Usher syndrome is an autosomal recessive condition presents with hearing loss, decreased vestibular function, and retinitis pigmentosa, which can be assessed with electroretinography. There are 4 recognized phenotypes of Usher syndrome with the following features:

Usher syndrome is characterized by hearing loss, vestibular dysfunction and retinitis pigmentosa. Retinitis pigmentosa can be diagnosed with electroretinography.

1. A 48 year old female presents with a 6 month history of progressive left side hearing loss and tinnitus. An MRI with contrast is performed which reveals a lateral temporal bone lesion that is hyperintense on T1 and T2. What is the most likely diagnosis?
2. Vestibular schwannoma
3. Meningioma
4. Cholesteatoma
5. Lipoma
6. **Cholesterol granuloma**

**Difficulty Level: Easy**

Cholesterol granulomas are the only lesions that are hyperintense on both T1 and T2.

1. On an audiogram what is the speech reception threshold (SRT)?
2. **level at which a patient can repeat 50% of the speech material**
3. level at which a patient is aware of a speech signal 50% of the time
4. amount of monosyllabic words repeated after hearing them at 50 dB above threshold
5. amount of times a patient with perceived unilateral hearing loss responds to a stimulus 10 dB above threshold in the good ear with a stimulus 10 dB below threshold in the bad ear

**Difficulty Level: Moderate**

The speech reception threshold is the level at which a patient can repeat 50% of the speech material.

1. A mother is known to have Neurofibromatosis type II (+,-). What is the risk for her offspring to inherit the disease?
2. 0%
3. 25%
4. **50%**
5. 75%
6. 100%

**Difficulty Level: Easy**

**Explanation:** Because Neurofibromatosis types 1 and 2 are transmitted in an autosomal dominant fashion, the risk of transmission, assuming the father is not afflicted, is 50%.

Neurofibromatosis types 1 and 2 are autosomal dominant conditions.

1. Which of the following is true of the cochlear afferent nerves?
2. The first synapse is in the inferior colliculus
3. The majority synapse on the outer hair cells
4. **The majority are bipolar neurons**
5. They innervate the stria vascularis
6. Each nerve fiber synapses on 1 outer hair cell

**Difficulty Level: Hard**

**Explanation:** 2 types of nerve cells comprise the cochlear nerve afferents: Type I and type II. 95% of cochlear nerve afferents are type I cells, and 5% are type II. Type I cells are bipolar, and synapse directly on the body of a single inner hair cell. Each inner hair cell has approximately 20 type I fibers synapsing at its terminal. Type II nerve cells are pseudounipolar, and branch to synapse onto multiple outer hair cells.

The majority of cochlear nerve afferent fibers are bipolar neurons that synapse directly onto individual inner hair cells (type I cells). A minority are pseudounipolar and synapse onto multiple outer hair cells (type II cells).

1. A mother that is congenitally deaf is found to have positive heterozygous EYA1 mutation. What is the chance she will pass deafness via the EYA1 mutation on to her child?
2. 100%
3. 75%
4. **50%**
5. 25%
6. 5%

**Difficulty Level: Moderate**

**Explanation:** EYA1 mutation is found in those with branchio-oto-renal syndrome, which is transmitted in an autosomal dominant fashion. An understanding of Mendelian genetics is key to answering this question. Since she has a 50% chance of passing the affected allele on to her child, the child will have a 50% chance of inheriting the trait.

Branchio-oto-renal syndrome is transmitted in autosomal dominant fashion, and the EYA1 mutation is responsible for this syndrome.