# Neurotology Fellowship Curriculum

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This curriculum is the result of a collaborative effort by the **Fellowship Committee of the American Neurotology Society**. Artificial intelligence was used to help formulate an initial draft, and all content was reviewed and edited by the contributing authors.

We welcome suggestions and feedback. Please direct any comments regarding the syllabus to:
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**1. Anatomy of the Temporal Bone and Lateral Skull Base**

Goals and Objectives:
By the completion of the fellowship, the trainee should demonstrate a thorough understanding of the anatomy of the temporal bone and lateral skull base, with specific emphasis on its relevance to neurotologic surgery and clinical decision-making.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the detailed anatomy of the temporal bone and its components, including the external, middle, and inner ear structures, mastoid and petrous apex.
- Identify foramina and critical neurovascular structures, including related cranial nerves, jugular bulb, sigmoid sinus, superior petrosal sinus, inferior petrosal sinus, and internal carotid artery.
- Understand the spatial relationships of the temporal bone to adjacent anatomical areas, including the posterior fossa, middle cranial fossa, sphenoid, neck and lateral skull base.
- Apply knowledge of temporal bone anatomy to common surgical approaches, including translabyrinthine, middle fossa, retrosigmoid, transotic, and infratemporal fossa techniques.
- Recognize anatomical variations and their implications for surgical planning and complication avoidance.
- Interpret common preoperative imaging studies (CT, MRI) as well as disease-specific imaging (such as nuclear medicine studies, PET scans) with precise anatomical correlation to temporal bone anatomy.

Syllabus:
Overview of Temporal Bone Anatomy:

- External ear: auricle and external auditory canal.
- Middle ear: ossicles, tympanic membrane, middle ear cleft, and Eustachian tube.
- Inner ear: cochlea, vestibule, semicircular canals, and otic capsule.
- Mastoid: boundaries, muscular attachments, suture lines
- Petrous apex: boundaries, neurovascular structures
- Infratemporal fossa: boundaries, neurovascular structures

Neurovascular Structures:

- Facial nerve: course through the temporal bone (extratemporal segment, and intratemporal segments including labyrinthine, tympanic, and mastoid; nerve branches).
- Vestibulocochlear nerve: course through the temporal bone
- Trigeminal nerve; Lower cranial nerves

- Jugular bulb and sigmoid sinus: anatomical variations and implications for surgery.
- Internal carotid artery: canal and relationships within the temporal bone, including petrous segment.
- Venous drainage patterns and implications in skull base procedures.

Surgical Landmarks and Approaches:

- Tegmen tympani, mastoid antrum, round and oval windows, facial recess.
- Key landmarks for safe entry and navigation during temporal bone surgery (spine of Henle, digastric ridge, sinodural angle).
- Anatomical considerations for translabyrinthine, middle fossa, retrosigmoid, transotic, infratemporal fossa, and combined approaches.

Adjacent Anatomy:

- Relationships with the middle cranial fossa, posterior cranial fossa, cerebellopontine angle, jugular foramen, sphenoid, neck and lateral skull base
- Clinical relevance in skull base surgical approaches, including tumor resections encephalocele and cerebrospinal fluid (CSF) leak repairs, facial nerve decompression, infectious pathology, vascular decompression procedures, and cochlear implantation.

Recommended Reading:


1. Proctor, B. *Surgical Anatomy of the Ear and Temporal Bone.* Thieme Medical Publishers, 1989.
2. Nadol, J.B., and Schuknecht, H.F. *Surgery of the Ear and Temporal Bone.* 2nd Edition. Lippincott Williams & Wilkins, 2004.
3. Fisch, U., and Mattox, D.E. *Microsurgery of the Skull Base.* Thieme, 1988.
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6. Brackmann DE, Jackler RK. Neurotology. Elsevier Mosby; 2005.
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**2. Pathophysiology of Hearing Loss**

Goals and Objectives:

By the completion of the fellowship, the trainee should demonstrate a comprehensive understanding of the mechanisms underlying hearing loss, with an emphasis on their application to clinical diagnosis and management.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Differentiate between conductive, sensorineural, and mixed hearing loss based on pathophysiological mechanisms.
- Describe the molecular and cellular pathophysiology of presbycusis, ototoxicity, noise-induced hearing loss, autoimmune inner ear disease (AIED), sudden sensorineural hearing loss (SSNHL), and genetic disorders.
- Explain the concept of cochlear synaptopathy and hidden hearing loss.
- Correlate audiometric and electrophysiologic findings with underlying pathological changes in the auditory system.
- Apply pathophysiologic principles to develop evidence-based treatment strategies for various forms of hearing loss.

Syllabus:
Types and Classification of Hearing Loss:

- Conductive vs. sensorineural vs. mixed hearing loss.
- Sudden vs. progressive hearing loss.
- Unilateral vs. bilateral and symmetric vs. asymmetric hearing loss.

Pathophysiological Mechanisms:

- Presbycusis: Degeneration of cochlear structures and central auditory pathways.
- Noise-Induced Hearing Loss: Hair cell and synaptic damage from acoustic trauma.
- Ototoxicity: Mechanisms of drug-induced cochlear and vestibular toxicity.
- Genetic Hearing Loss: Syndromic and non-syndromic genetic mutations affecting auditory function.
- Autoimmune Inner Ear Disease (AIED): Inflammatory and autoimmune mechanisms causing inner ear damage.
- Sudden Sensorineural Hearing Loss (SSNHL): Potential viral, vascular, or idiopathic mechanisms.
- Retrocochlear etiology: tumor, central or brainstem causes
- Traumatic hearing loss

Diagnosis and Clinical Correlation:

- Audiometric presentations of common pathophysiologic processes.
- Use of electrophysiologic testing, such as otoacoustic emissions (OAEs) and ABR/ASSRABR) in diagnosing cochlear and retrocochlear pathology.
- Cochlear synaptopathy and implications for hearing in noisy environments.
- Interpretation of imaging (MRI, CT) in correlation with pathophysiologic findings.

Implications for Management:

- Pharmacologic and protective strategies for noise-induced and ototoxic hearing loss.
- Emerging therapies targeting synaptopathy and cochlear regeneration.

- Role of traditional hearing aids, contralateral routing of sound aids, bone anchored hearing aids, auditory implantable devices, including bone anchored and cochlear implants, and assistive listening devices in management.
- Corticosteroids and immunomodulatory therapies for AIED and SSNHL.

Recommended Reading:


1. Kujawa SG, Liberman MC. "Cochlear synaptopathy and its implications for auditory function." *Hearing Research,* 2015;330(Pt B):191-199.
2. Merchant SN, Nadol JB. *Schuknecht's Pathology of the Ear*, 3rd Edition. PMPH-USA, 2010.
3. Gelfand SA. *Essentials of Audiology*, 4th Edition. Thieme, 2016.
4. Ferguson MA, Kitterick PT, Chong LY, Edmondson-Jones M, Barker F, Hoare DJ. Hearing aids for mild to moderate hearing loss in adults. Cochrane Database Syst Rev. 2017 Sep 25;9(9):CD012023. doi: 10.1002/14651858.CD012023.pub2. PMID: 28944461; PMCID: PMC6483809.
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**3. Audiologic Testing**

Goal and Objectives:
By the completion of the fellowship, the trainee should have a thorough understanding of audiologic testing modalities and their role in the diagnosis and management of auditory and vestibular disorders.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Explain the principles of pure-tone audiometry, speech audiometry, tympanometry, and acoustic reflex testing.
- Interpret audiologic test results in various pathologies and recognize patterns indicative of retrocochlear disease.
- Describe the application and limitations of otoacoustic emissions (OAEs), auditory steady state response (ASSR) and auditory brainstem response (ABR) testing.
- Recognize audiologic red flags that warrant further imaging or evaluation.
- Utilize audiologic data in developing hearing rehabilitation and treatment plans.

Syllabus:
Fundamentals of Audiologic Testing:

- Principles of air- and bone-conduction audiometry.
- Speech reception thresholds (SRT) and word recognition scores (WRS).
- Tympanometry and acoustic reflex testing.
- Masking principles in audiologic assessment.

Advanced Audiologic Testing:

- Otoacoustic emissions (OAEs) and their diagnostic applications.
- Auditory steady-state response (ASSR) in estimating frequency-specific hearing thresholds.
- Electrophysiologic testing, including ASSR/ABR, in young children, difficult to test populations, auditory neuropathy and dyssynchrony and retrocochlear pathology.

Clinical Interpretation and Application:

- Audiologic patterns of conductive vs. sensorineural hearing loss.
- Audiologic evaluation of asymmetric hearing loss and vestibular schwannomas.
- Use of audiologic data in bone anchored and cochlear implant candidacy assessment.
- Integration of audiologic findings with clinical history and imaging for comprehensive diagnosis.

Recommended Reading:


1. Katz J, Chasin M, English K. *Handbook of Clinical Audiology*, 7th Edition. Wolters Kluwer, 2015.
2. Gelfand SA. *Essentials of Audiology*, 4th Edition. Thieme, 2016.
3. Yawn RJ, Nassiri AM, Rivas A. Auditory Neuropathy: Bridging the Gap Between Hearing Aids and Cochlear Implants. Otolaryngol Clin North Am. 2019 Apr;52(2):349-355. doi: 10.1016/j.otc.2018.11.016. Epub 2019 Feb 12. PMID: 30765091.


**4. Vestibular Anatomy and Physiology**

Goal and Objectives:
By the completion of the fellowship, the trainee should have a comprehensive understanding of the structure and function of the vestibular system, with a focus on its role in balance and spatial orientation.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the detailed anatomy of the semicircular canals, otolithic organs, and vestibular nerve pathways.
- Explain the physiological basis of the vestibulo-ocular reflex (VOR) and vestibulospinal reflexes.
- Understand the integration of vestibular, visual, and proprioceptive inputs in maintaining equilibrium.

- Correlate vestibular dysfunction with clinical presentations of vertigo, imbalance, and nystagmus.
- Interpret findings from vestibular testing, including videonystagmography (VNG), video head impulse test (vHIT), electrocochleography (Ecogh), posturography, , and VEMPs.
- Apply knowledge of vestibular anatomy and physiology to guide diagnostic workup and treatment planning.

Syllabus:

Anatomy of the Vestibular System:
- Semicircular canals and otolithic organs.
- Central vestibular pathways and connections to the brainstem and cerebellum.
- Vestibular nuclei and their projections to ocular motor nuclei, spinal cord, and cortex.

Vestibular Physiology:
- Mechanisms of angular and linear acceleration detection.
- Function of the VOR in stabilizing gaze.
- Vestibulospinal reflexes and their role in postural control.
- Role of velocity storage mechanism and central compensation in chronic vestibular dysfunction.

Clinical Relevance:
- Pathophysiology of common vestibular disorders such as Meniere's disease, vestibular neuritis, labyrinthitis and superior canal dehiscence.
- Benign paroxysmal positional vertigo (BPPV): mechanism, diagnosis, and treatment.
- Central vestibular disorders including vestibular migraine and cerebellar ataxia.
- Role and outcomes following vestibular rehabilitation therapy.
- Use of bedside and formal vestibular testing to localize pathology.

Recommended Reading:

- Baloh RW, Halmagyi GM. Disorders of the Vestibular System. Oxford University Press, 1996.
- Leigh RJ, Zee DS. The Neurology of Eye Movements, 5th Edition. Oxford University Press, 2015.
- Strupp M, Brandt T. "Vestibular Migraine and Related Syndromes." The Lancet Neurology, 2018;17(7):701–710.
- Fife TD, Tusa RJ. "Vestibular Pathophysiology and Testing." In: Handbook of Clinical Neurology, Vol. 137. Elsevier, 2016.
- Dlugaiczyk J. Rare Disorders of the Vestibular Labyrinth: of Zebras, Chameleons and Wolves in Sheep's Clothing. Laryngorhinootologie. 2021 Apr;lOO(S 01}:Sl-S40. doi: 10.1055/a-1349-7475. Epub 2021 Apr 30. PMID: 34352900; PMCID: PMC8363216.

**5. Vestibular Testing**

Goal and Objectives:
By the completion of the fellowship, the trainee should be able to interpret vestibular test results and apply them to clinical decision-making.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Understand the principles of videonystagmography (VNG), rotary chair testing, and vestibular-evoked myogenic potentials (VEMP).
- Interpret abnormal vestibular test findings.
- Correlate vestibular test results with patient symptoms.
- Identify limitations and artifacts in vestibular testing and adjust interpretation accordingly.
- Utilize vestibular testing to guide diagnosis, treatment decisions, and referrals.

Syllabus:

Overview of Vestibular Testing:

- VNG and caloric testing.
- Rotary chair testing.
- VEMP for otolith function.
- Posturography
- Video head impulse testing (vHIT)
- EcOG
- Subjective visual vertical (SVV) testing for utricular assessment.

Clinical Interpretation:

- Differentiating peripheral from central vestibular dysfunction.
- Integrating test results with patient history.
- Recognizing patterns of vestibular hypofunction, directional preponderance, and fixation suppression.
- Correlation of caloric weakness, abnormal gain, or asymmetry with suspected vestibular pathology.
- EcOg for the assessment of endolymphatic hydrops

Recommended Reading:


1. Shepard NT, Telian SA. *Practical Management of the Balance Disorder Patient*. Singular Publishing Group, 1996.
2. Baloh RW, Honrubia V. Clinical neurophysiology of the vestibular system. Conte mp Neural Ser. 1.979;18:1-21
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4. Jacobson GP, Shepard NT. *Balance Function Assessment and Management*, 2nd Edition. Plural Publishing, 2014.
5. Zee DS. *Vestibular Testing and Rehabilitation*. In: *Neurologic Clinics*, 2005;23(3):675–692.
6. Fife TD. *Clinical Applications of Vestibular Testing*. Futura Publishing, 2000.
7. Hain TC, Cherchi M. "Vestibular Function Testing." In: *Handbook of Clinical Neurology*, Vol. 137. Elsevier, 2016.
8. Halmagyi GM, Chen L, MacDougall HG, Weber KP, McGarvie LA, Curthoys IS. The Video Head Impulse Test. Front Neurol. 2017 Jun 9;8:258.
9. Curthoys IS, McGarvie LA, MacDougall HG, Burgess AM, Halmagyi GM, Rey-Martinez J, Dlugaiczyk J. A review of the geometrical basis and the principles underlying the use and interpretation of the video head impulse test (vHIT) in clinical vestibular testing. Front Neural. 2023 Apr 11;14:1147253. doi: 10.3389/fneur.2023.1147253.
10. Balance Function Assessment and Management, Third Edition: Editors: Gary P. Jacobson, Neil T. Shepard, Kamran Barin, Kristen Janky, Devin L. Mccaslin: Publisher: Plural Publishing, 2020: ISBN: 1635501997, 9781635501995. Chapter 12: Caloric testing, Chapter 13: Rotational Vestibular Assessment
11. Practical Management of the Dizzy Patient, Joel A. Goebel, MD, Second Edition, Lippincott Williams & Wilkins, Published April 15, 2008

**6. Imaging of the Temporal Bone and Skull Base**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in interpreting imaging studies of the temporal bone and skull base for diagnostic and surgical planning.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Identify key anatomical structures of the temporal bone on high-resolution CT (HRCT) and MRI.
- Recognize imaging features of common neurotologic disorders.
- Apply imaging interpretation to surgical decision-making.
- Differentiate normal anatomic variants from pathologic findings.
- Communicate imaging findings effectively with radiologists and other members of the care team.

Syllabus:

Imaging Modalities:

- HRCT of the temporal bone.
- MRI sequences relevant to skull base pathology.
- CT angiography (CTA) and MR angiography (MRA) for vascular evaluation.
- Diffusion-weighted imaging (DWI) in the evaluation of cholesteatoma.

Interpretation and Clinical Applications:

- Radiologic findings in otosclerosis, chronic otitis media/cholesteatoma, 3rd window disorders, temporal bone trauma, superior semicircular canal dehiscence, and vestibular schwannomas and other skull base tumors.
- Preoperative imaging for cochlear implantation.
- Assessment of anatomical constraints and landmarks for translabyrinthine, retrosigmoid, and middle fossa approaches.
- Evaluation of facial nerve course, cochlear patency, and surgical risks.

Recommended Reading:

- Swartz JD, Loevner LA. *Imaging of the Temporal Bone*, 5th Edition. Thieme, 2009.
- Yousem DM, Grossman RI. *Neuroradiology: The Requisites*, 3rd Edition. Elsevier, 2010.
- Mafee MF. *Imaging of the Temporal Bone and Skull Base*, 2nd Edition. Thieme, 2005.
- Jackler RK. *Neuroimaging in Otology and Neurotology*. In: *Cummings Otolaryngology: Head and Neck Surgery*, 6th Edition. Elsevier, 2015.
- Harnsberger HR. *Diagnostic Imaging: Head and Neck*, 3rd Edition. Elsevier, 2016.

**7. Genetics of Hearing Loss**

Goal and Objectives:

By the completion of the fellowship, the trainee should have a comprehensive understanding of the genetic contributions to congenital and acquired hearing loss, including patterns of inheritance, syndromic and non-syndromic forms, genetic testing modalities, and implications for counseling and management.

Objectives:

By the end of the fellowship, the fellow should be able to:

1. Describe the classification of genetic hearing loss into syndromic and non-syndromic categories.
2. Differentiate modes of inheritance including autosomal dominant, autosomal recessive, X-linked, and mitochondrial patterns.
3. Identify common genes implicated in hereditary hearing loss, such as *GJB2* (Connexin 26), *SLC26A4*, *OTOF*, and mitochondrial mutations.
4. Interpret the clinical implications of genetic test results, including diagnostic, prognostic, and reproductive considerations.
5. Recognize syndromic forms of hearing loss (e.g., Usher, Pendred, Waardenburg, Alport syndromes) and their associated systemic manifestations.
6. Counsel patients and families regarding the role of genetic evaluation, benefits and limitations of testing, and appropriate referrals for genetic counseling.
7. Apply knowledge of genetics to guide personalized management plans, including candidacy for cochlear implantation.

Neurotology Fellows CurriculumSyllabus:

Overview of Genetic Hearing Loss:

- Epidemiology: Approximately 50–60% of congenital hearing loss has a genetic etiology.
- Classification: Syndromic (~30%) vs. non-syndromic (~70%).
- Inheritance patterns: Autosomal recessive (most common), autosomal dominant, X-linked, mitochondrial.

Non-Syndromic Genetic Hearing Loss:

- *GJB2* (Connexin 26) mutations: Most common cause of autosomal recessive nonsyndromic sensorineural hearing loss.
- *OTOF* mutations: Auditory neuropathy spectrum disorder.
- DFNA and DFNB loci: Classification based on inheritance and locus.
- Mitochondrial mutations (e.g., A1555G): Susceptibility to aminoglycoside ototoxicity.

Syndromic Genetic Hearing Loss:

- Usher syndrome: Sensorineural hearing loss and retinitis pigmentosa.
- Pendred syndrome: Hearing loss with thyroid goiter and EVA (enlarged vestibular aqueduct).
- Waardenburg syndrome: Pigmentary abnormalities, dystopia canthorum.
- Alport syndrome: Hearing loss with progressive nephropathy.
- Jervell and Lange-Nielsen syndrome: Hearing loss with prolonged QT interval.

Genetic Evaluation and Testing:

- Indications for referral to genetics.
- Approaches: Single-gene testing, gene panels, whole exome sequencing.
- Interpretation of variants: Pathogenic, likely pathogenic, variant of uncertain significance.
- Limitations of current genetic testing.

Counseling and Management:

- Reproductive counseling and risk assessment.
- Implications for cochlear implant candidacy and outcomes.
- Psychosocial considerations.
- Role of multidisciplinary care.

References

1. Clinical Evaluation and Etiologic Diagnosis of Hearing Loss: A Clinical Practice Resource of the American College of Medical Genetics and Genomics (ACMG). Li MM, Tayoun AA, DiStefano M, et al. Genetics in Medicine : Official Journal of the American College of Medical Genetics. 2022;24(7):1392-1406. doi:10.1016/j.gim.2022.03.018.

2. Hearing Loss in Children: A Review. Lieu JEC, Kenna M, Anne S, Davidson L. JAMA. 2020;324(21):2195-2205. doi:10.1001/jama.2020.17647.
3. Genetic Etiology of Non-Syndromic Hearing Loss in Europe. Del Castillo I, Morín M, Domínguez-Ruiz M, Moreno-Pelayo MA. Human Genetics. 2022;141(3-4):683-696. doi:10.1007/s00439-021-02425-6.
4. A Systematic Review of the Monogenic Causes of Non-Syndromic Hearing Loss (NSHL) and Discussion of Current Diagnosis and Treatment Options. Sharma N, Kumari D, Panigrahi I, Khetarpal P. Clinical Genetics. 2023;103(1):16-34. doi:10.1111/cge.14228.
5. Outcomes of Gene Panel Testing for Sensorineural Hearing Loss in a Diverse Patient Cohort. Liao EN, Taketa E, Mohamad NI, Chan DK. JAMA Network Open. 2022;5(9):e2233441. doi:10.1001/jamanetworkopen.2022.33441.

**8. Peripheral Vestibular Disorders (BPPV, Vestibular Neuritis, Labyrinthitis)**

Goal and Objectives:
By the completion of the fellowship, the trainee should have a thorough understanding of the pathophysiology, diagnosis, and management of peripheral vestibular disorders, including benign paroxysmal positional vertigo (BPPV), vestibular neuritis, and labyrinthitis.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the pathophysiological mechanisms underlying BPPV, vestibular neuritis, and labyrinthitis.
- Differentiate among common peripheral vestibular disorders based on clinical presentation and diagnostic findings.
- Perform and interpret bedside vestibular function tests such as the Dix-Hallpike maneuver, evaluation for nystagmus, head impulse test, Fukuda step test, HINTS exam, and Romberg exam.
- Formulate evidence-based treatment plans, including canalith repositioning maneuvers, vestibular rehabilitation, and pharmacologic therapy.
- Recognize red flags that may indicate central vestibular pathology requiring further investigation.
- Counsel patients on prognosis and expected course of recovery for peripheral vestibular conditions.

Syllabus:

Benign Paroxysmal Positional Vertigo (BPPV):

- Pathophysiology: Canalithiasis vs. cupulolithiasis.
- Clinical features: Characteristic positional vertigo.
- Diagnosis: Dix-Hallpike and supine roll tests.
- Treatment: Epley, Semont, and BBQ roll maneuvers.
- Persistent and recurrent BPPV: contributing factors and management strategies.

Vestibular Neuritis and Labyrinthitis:

- Etiology: Viral, post-viral, and idiopathic causes.
- Clinical presentation: Acute vestibular syndrome, spontaneous nystagmus.
- Diagnostic tests: Head impulse test (HIT), VNG, VEMP
- Management: Corticosteroids, symptomatic treatment, and vestibular rehabilitation.
- Distinguishing vestibular neuritis (no hearing loss) from labyrinthitis (hearing loss present).

Differentiation from Central Vestibular Disorders:

- HINTS exam for stroke screening.
- When to order MRI/MRA for posterior circulation evaluation.
- Identification of red flags such as skew deviation, direction-changing nystagmus, and poor fixation suppression.

Recommended Reading:


1. Strupp M, Brandt T. "Vestibular Neuritis." *Seminars in Neurology*, 2009;29(5):509–519.
2. Fife TD, Tusa RJ. "Benign Paroxysmal Positional Vertigo: Diagnosis and Management." *Otolaryngologic Clinics of North America*, 2015;48(4):795–807.
3. Bhattacharyya N, et al. "Clinical Practice Guideline: Benign Paroxysmal Positional Vertigo." *Otolaryngology–Head and Neck Surgery*, 2017;156(3_suppl):S1–S47.
4. Baloh RW, Halmagyi GM. *Disorders of the Vestibular System*. Oxford University Press, 1996.
5. Lopez-Escamez JA, et al. "Vestibular Neuritis: Pathophysiology and Clinical Implications." *Frontiers in Neurology*, 2014;5:5.


**9. Meniere's Disease: Diagnosis and Management**

Goal and Objectives:
By the completion of the fellowship, the trainee should be able to accurately diagnose and manage Meniere's disease, incorporating knowledge of its pathophysiology and treatment options.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the pathophysiology and diagnostic criteria for Meniere's disease.
- Differentiate Meniere's disease from other causes of episodic vertigo.
- Interpret audiometric patterns characteristic of Meniere's disease.
- Develop medical, dietary, and surgical treatment plans for Meniere's disease.
- Recognize and manage secondary endolymphatic hydrops.
- Counsel patients on prognosis, expected course, and impact on quality of life.

Syllabus:

1. Describe the presenting history of Meniere's disease (MD), including the variable nature of presentation (2015 Barany Society Diagnostic Criteria)
    a. Definite MD
    b. Probable MD
2. Describe the epidemiology of MD
3. Describe the pathophysiology (endolymphatic hydrops) and hypothesized etiologies of MD
    - Including autoimmune, genetic, and anatomical factors
4. Describe the natural history of MD
    - Typical progression from episodic vertigo to progressive hearing loss and possible imbalance
5. Develop a differential diagnosis for MD, understanding other common causes of vertigo and how they differ from MD
    a. Autoimmune (e.g., multiple sclerosis)
    b. Benign paroxysmal positional vertigo
    c. Infectious (e.g., Lyme disease)
    d. Otosyphilis
    e. Stroke/ischemia
    f. Vestibular migraine
    g. Vestibular schwannoma
    h. Labyrinthitis
    i. Vestibular neuritis
6. Describe the relative importance of different diagnostic evaluations and interpretation of test findings for MD with respect to:
    a. Physical examination
    b. Audiogram
    c. Magnetic resonance imaging (MRI)
    d. Vestibular function testing (including VNG, VEMP) and electrocochleography (ECochG)
    e. Glycerol test and vestibular evoked myogenic potentials (VEMP) as supportive tests
7. Describe the management and treatment of MD
    a. Modification of lifestyle factors and diet
        i. Low-sodium diet
        ii. Limit caffeine, alcohol, and nicotine consumption
        iii. Eat well-balanced meals and stay hydrated
        iv. Exercise, sleep hygiene, and stress management
        v. Identify and manage allergies

    b. Medical treatment
        i. Diuretics
        ii. Betahistine
        iii. Steroids
        iv. Vestibular suppressants for acute vertigo symptoms

1. First-generation antihistamines (e.g., dimenhydrinate, meclizine)
2. Benzodiazepines (e.g., diazepam, lorazepam, clonazepam)
3. Anticholinergics (e.g., scopolamine)

v. Medications to treat concurrent allergies as appropriate
vi. Medications to treat concurrent vestibular migraines as appropriate

c. Intratympanic injections
i. Steroids (non-ablative for vestibular system; hearing-sparing)
ii. Gentamicin (ablative for vestibular system; potentially ototoxic for hearing)

d. Surgical treatment
i. Endolymphatic sac decompression (non-ablative for vestibular system)
ii. Vestibular nerve section (ablative for vestibular system; hearing-sparing)
iii. Labyrinthectomy (ablative for vestibular and auditory system)

e. Other therapies
i. Vestibular rehabilitation for chronic imbalance
ii. Treatment of hearing loss which, depending on degree (HA, BAHA, CI)

## Recommended Reading:

**Clinical Practice Guidelines:**


1. Basura GJ, Adams ME, Monfared A, Schwartz SR, Antonelli PJ, Burkard R, Bush ML, Bykowski J, Colandrea M, Derebery J, Kelly EA, Kerber KA, Koopman CF, Kuch AA, Marcolini E, McKinnon BJ, Ruckenstein MJ, Valenzuela CV, Vosooney A, Walsh SA, Nnacheta LC, Dhepyasuwan N, Buchanan EM. Clinical Practice Guideline: Ménière's Disease. Otolaryngol Head Neck Surg. 2020 Apr;162(2_suppl):S1-S55.


**Cochrane Reviews:**


2. Webster KE, George B, Lee A, Galbraith K, Harrington-Benton NA, Judd O, Kaski D, Maarsingh OR, MacKeith S, Murdin L, Ray J, Van Vugt VA, Burton MJ. Lifestyle and dietary interventions for Ménière's disease. Cochrane Database Syst Rev. 2023 Feb 27;2(2):CD015244. doi: 10.1002/14651858.CD015244.pub2.
3. Webster KE, Galbraith K, Harrington-Benton NA, Judd O, Kaski D, Maarsingh OR, MacKeith S, Ray J, Van Vugt VA, Burton MJ. Systemic pharmacological interventions for Ménière's disease. Cochrane Database Syst Rev. 2023 Feb 23;2(2):CD015171. doi: 10.1002/14651858.CD015171.pub2.
4. Webster KE, Lee A, Galbraith K, Harrington-Benton NA, Judd O, Kaski D, Maarsingh OR, MacKeith S, Ray J, Van Vugt VA, Westerberg B, Burton MJ. Intratympanic corticosteroids for Ménière's disease. Cochrane Database Syst Rev. 2023 Feb 27;2(2):CD015245. Webster KE, Galbraith K, Lee A, Harrington-Benton NA, Judd O, Kaski D, Maarsingh OR, MacKeith S, Ray J, Van Vugt VA, Burton MJ. Intratympanic

gentamicin for Ménière's disease. Cochrane Database Syst Rev. 2023 Feb 27;2(2):CD015246.
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**Overview Articles:**

6. Wright T. Menière's disease. BMJ Clin Evid. 2015 Nov 5;2015:0505.
7. Nakashima T, Pyykkö I, Arroll MA, Casselbrant ML, Foster CA, Manzoor NF, Megerian CA, Naganawa S, Young YH. Meniere's disease. Nat Rev Dis Primers. 2016 May 12;2:16028. doi: 10.1038/nrdp.2016.28.

**Diagnostic Criteria:**

8. Lopez-Escamez JA, Carey J, Chung WH, Goebel JA, Magnusson M, Mandalà M, Newman-Toker DE, Strupp M, Suzuki M, Trabalzini F, Bisdorff A; Classification Committee of the Barany Society; Japan Society for Equilibrium Research; European Academy of Otology and Neurotology (EAONO); Equilibrium Committee of the American Academy of Otolaryngology-Head and Neck Surgery (AAO-HNS); Korean Balance Society. Diagnostic criteria for Menière's disease. J Vestib Res. 2015;25(1):1-7.

**Epidemiology:**

9. Harris JP, Alexander TH. Current-day prevalence of Ménière's syndrome. Audiol Neurootol. 2010;15(5):318-22. doi: 10.1159/000286213. Epub 2010 Feb 18.
10. Tyrrell JS, Whinney DJ, Ukoumunne OC, Fleming LE, Osborne NJ. Prevalence, associated factors, and comorbid conditions for Ménière's disease. Ear Hear. 2014 Jul-Aug;35(4):e162-9. doi: 10.1097/AUD.0000000000000041.
11. Lopez-Escamez JA, Liu Y. Epidemiology and genetics of Meniere's disease. Curr Opin Neurol. 2024 Feb 1;37(1):88-94. Doi: 10.1097/WCO.0000000000001227. Epub 2023 Oct 20.

**Pathophysiology, Etiology & Associated Disorders:**

12. Paparella MM, Djalilian HR. Etiology, pathophysiology of symptoms, and pathogenesis of Meniere's disease. Otolaryngol Clin North Am. 2002 Jun;35(3):529-45, vi.
13. Derebery MJ, Berliner KI. Allergy and Ménière's disease. Curr Allergy Asthma Rep. 2007 Nov;7(6):451-6. doi: 10.1007/s11882-007-0069-0.
14. Semaan MT, Megerian CA. Contemporary perspectives on the pathophysiology of Meniere's disease: implications for treatment. Curr Opin Otolaryngol Head Neck Surg. 2010 Oct;18(5):392-8. doi: 10.1097/MOO.0b013e32833d3164.
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16. Ahmad JG, Lin KF. Ménière's disease is a disorder of the inner ear. Curr Opin Otolaryngol Head Neck Surg. 2023 Oct 1;31(5):320-324.
17. Frank M, Abouzari M, Djalilian HR. Meniere's disease is a manifestation of migraine. Curr Opin Otolaryngol Head Neck Surg. 2023 Oct 1;31(5):313-319.
18. Chen JY, Guo ZQ, Wang J, Liu D, Tian E, Guo JQ, Kong WJ, Zhang SL. Vestibular migraine or Meniere's disease: a diagnostic dilemma. J Neurol. 2023 Apr;270(4):1955-1968. doi: 10.1007/s00415-022-11532-x. Epub 2022 Dec 23.
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**Natural History:**.

20. Huppert D, Strupp M, Brandt T. Long-term course of Menière's disease revisited. Acta Otolaryngol. 2010 Jun;130(6):644-51.

**Diagnostic Testing:**

21. Adams ME, Heidenreich KD, Kileny PR. Audiovestibular testing in patients with Meniere's disease. Otolaryngol Clin North Am. 2010 Oct;43(5):995-1009.
22. Ziylan F, Smeeing DP, Stegeman I, Thomeer HG. Click Stimulus Electrocochleography Versus MRI With Intratympanic Contrast in Ménière's Disease: A Systematic Review. Otol Neurotol. 2016 Jun;37(5):421-7.
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24. Orimoto KY, Vartanyan M, O'Leary SJ. Systematic review of the diagnostic value of hydrops MRI in relation to audiovestibular function tests (electrocochleography, cervical vestibular evoked myogenic potential and caloric test). Eur Arch Otorhinolaryngol. 2023 Mar;280(3):947-962.

**Treatment:**

25. Ahmadzai N, Cheng W, Kilty S, Esmaeilisaraji L, Wolfe D, Bonaparte J, Schramm D, Fitzpatrick E, Lin V, Skidmore B, Hutton B. Pharmacologic and surgical therapies for patients with Meniere's disease: A systematic review and network meta-analysis. PLoS One. 2020 Sep 1;15(9):e0237523.
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27. Crowson MG, Patki A, Tucci DL. A Systematic Review of Diuretics in the Medical Management of Ménière's Disease. Otolaryngol Head Neck Surg. 2016 May;154(5):824-34. doi: 10.1177/0194599816630733. Epub 2016 Mar 1.

28. Devantier L, Djurhuus BD, Hougaard DD, Händel MN, Guldfred FL, Schmidt JH, Edemann-Callesen H. Intratympanic Steroid for Menière's Disease: A Systematic Review. Otol Neurotol. 2019 Jul;40(6):806-812.
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31. Szott FA, Westhofen M, Hackenberg S. Is endolymphatic sac surgery an efficient treatment of Menière's disease patients? A systematic literature search and meta-analysis. Eur Arch Otorhinolaryngol. 2023 Mar;280(3):1119-1128. doi: 10.1007/s00405-022-07580-8. Epub 2022 Oct 8. Erratum in: Eur Arch Otorhinolaryngol. 2023 Mar;280(3):1129.
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34. Selleck AM, Dillon M, Perkins E, Brown KD. Cochlear Implantation in the Setting of Menière's Disease After Labyrinthectomy: A Meta-Analysis. Otol Neurotol. 2021 Sep 1;42(8):e973-e979.

**10. Vestibular Migraine**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in diagnosing and managing vestibular migraine and associated disorders, integrating knowledge of its pathophysiology, clinical presentation, and treatment strategies.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the underlying mechanisms and risk factors contributing to vestibular migraine.
- Identify and apply the diagnostic criteria for vestibular migraine based on the Bárány Society consensus.
- Differentiate vestibular migraine from other episodic vestibular disorders, including Meniere’s disease and BPPV.
- Recognize the overlap between migraine-related balance problems, dizziness, and other central vestibular disorders.
- Develop treatment plans incorporating lifestyle modifications, pharmacotherapy, and vestibular rehabilitation.

- Counsel patients on prognosis and long-term management of vestibular migraine.

Syllabus:

Pathophysiology of Vestibular Migraine:

- Role of cortical spreading depression and brainstem dysfunction.
- Interaction between the trigeminovascular system and central vestibular pathways.
- Association with hypersensitivity of vestibular processing.
- Genetic and hormonal influences in susceptible individuals.

Clinical Presentation and Diagnosis:

- Recurrent episodes of vertigo and motion intolerance lasting minutes to hours to days.
- Association with migraine symptoms (headache, photophobia, phonophobia, aura).
- Diagnostic criteria: Bárány Society and International Classification of Headache Disorders (ICHD-3).
- Typical triggers: hormonal changes, stress, dietary factors, and sensory overload.

Differential Diagnosis:

- Vestibular migraine vs. Meniere's disease: Audiometric stability in migraine.
- Vestibular migraine vs. BPPV: Positional component and migrainous symptoms.
- Vestibular migraine vs. brainstem stroke: HINTS exam findings.
- Consideration of persistent postural-perceptual dizziness (PPPD) as a comorbidity.

Management Strategies:

- Lifestyle modifications: Migraine diet, sleep hygiene, stress management.
- Pharmacologic treatment: Mostly preventive (not abortive) migraine management including beta-blockers, calcium channel blockers, tricyclic antidepressants, and CGRP antagonists.
- Role and potential value of vestibular rehabilitation therapy.
- Patient education and expectation setting for episodic and chronic management.

**Recommended Reading:**


1. Neuhauser H, et al. "Vestibular Migraine: Diagnostic Criteria and Treatment." *Neurology*, 2001;56(4):436–441.
2. Lempert T, et al. "Vestibular Migraine: Diagnostic Criteria of the Bárány Society." *Journal of Vestibular Research*, 2012;22(4):167–172.
3. Bisdorff AR, et al. "The Epidemiology of Vestibular Migraine." *Neurology*, 2009;72(6):573–576.
4. Cha Y-H. "Vestibular Migraine and Related Syndromes." *Seminars in Neurology*, 2010;30(2):167–174.

5. Furman JM, Balaban CD. "Migraine-Associated Dizziness and Vertigo." *Current Opinion in Neurology*, 2015;28(1):74–80.
6. Lempert T, Olesen J, Furman J, Waterston J, Seemungal B, Carey J, Bisdorff A, Versino M, Evers S, Kheradmand A, Newman-Toker D. Vestibular migraine: Diagnostic criteria1. J Vestib Res. 2022;32(1):1-6.

**11. Persistent Postural Perceptual Dizziness (PPPD)**

Goal and Objectives:
By the completion of the fellowship, the trainee should be able to recognize and manage PPPD, a chronic vestibular disorder characterized by persistent dizziness and unsteadiness.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the neurophysiological mechanisms contributing to PPPD.
- Identify diagnostic criteria based on Bárány Society guidelines.
- Differentiate PPPD from anxiety-related dizziness and vestibular dysfunction.
- Explain the interplay between PPPD and psychiatric comorbidities.
- Implement evidence-based treatment approaches, including cognitive behavioral therapy and selective serotonin reuptake inhibitors (SSRIs).
- Recognize the role of multidisciplinary care and patient education in long-term management.

Syllabus:

Pathophysiology of PPPD:

- Maladaptive neural plasticity following an acute vestibular insult — commonly triggered by vestibular neuritis, BPPV, or vestibular migraine.
- Over-reliance on visual and proprioceptive input.
- Central nervous system hyperreactivity to motion stimuli.
- Interaction with anxiety circuits and impaired sensory reweighting.

Clinical Presentation and Diagnosis:

- Chronic dizziness and unsteadiness lasting at least 3 months.
- Symptoms exacerbated by upright posture, motion, and visual stimuli.
- Association with predisposing factors such as vestibular migraine and anxiety disorders.
- Fluctuation in symptom severity based on cognitive load, fatigue, and emotional state.

Differential Diagnosis:

- PPPD vs. vestibular migraine: Episodic vs. chronic dizziness.
- PPPD vs. functional neurologic disorders: Role of cognitive factors.
- Distinction from malingering or factitious disorders based on structured assessment.

Treatment Approaches:

- Vestibular rehabilitation: Exposure therapy for maladaptive visual reliance.
- Cognitive behavioral therapy for anxiety and motion sensitivity.
- Pharmacotherapy: SSRIs and serotonin-norepinephrine reuptake inhibitors (SNRIs).
- Psychoeducation: Validating symptoms and setting expectations for gradual recovery.

Recommended Reading:


1. Staab JP, Ruckenstein MJ. "Chronic Subjective Dizziness and Persistent Postural-Perceptual Dizziness." *Current Opinion in Otolaryngology & Head and Neck Surgery*, 2015;23(5):361–368.
2. Popkirov S, Staab JP, Stone J. "Persistent Postural Perceptual Dizziness (PPPD): A Common, Characterizable, and Treatable Cause of Chronic Dizziness." *Journal of Neurology*, 2018;265(1):12–16.
3. Staab JP. "The Influence of Anxiety on Vestibular Dysfunction." *Current Opinion in Neurology*, 2014;27(1):69–74.
4. Dieterich M, Staab JP. "Functional (Psychogenic) Vestibular Disorders." In: *Handbook of Clinical Neurology*, Vol. 139. Elsevier, 2016:247–259.
5. Huppert D, Strupp M, Rettinger N. "Treatment of PPPD: Current Strategies and Challenges." *Frontiers in Neurology*, 2020;11:570762.
6. Staab JP, Eckhardt-Henn A, Horii A, Jacob R, Strupp M, Brandt T, Bronstein A. Diagnostic criteria for persistent postural-perceptual dizziness (PPPD): Consensus document of the committee for the Classification of Vestibular Disorders of the Bárány Society. J Vestib Res. 2017;27(4):191-208.


**12. Superior Canal Dehiscence Syndrome (SCDS)**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in diagnosing and treating superior canal dehiscence syndrome (SCDS).

Objectives:
By the end of the fellowship, the fellow should be able to:

- Explain the anatomic defect responsible for SCDS and the physiologic consequences of a third window effect.
- Identify characteristic clinical symptoms, including Tullio phenomenon, pseudoconductive hearing loss, and autophony.
- Interpret diagnostic tests, including vestibular evoked myogenic potentials (VEMP) and high-resolution CT imaging.
- Differentiate SCDS from otosclerosis, perilymphatic fistula, and Meniere’s disease.
- Evaluate the role of surgical repair in symptomatic cases.
- Counsel patients regarding the natural history, risks, and benefits of treatment options.

Syllabus:

Pathophysiology and Anatomy:

- Dehiscence of the superior semicircular canal due to developmental or acquired factors.
- Third mobile window alters inner ear mechanics, affecting hearing and balance.

Clinical Features:

- Sound- and pressure-induced vertigo (Tullio phenomenon and Hennebert sign).
- Autophony and hyperacusis.
- Ability to hear ocular motion.
- Dysequilibrium and hearing loss.
- Pulsatile tinnitus, sensation of ear fullness, autophony, and imbalance in noisy environments.

Diagnosis:

- Audiometric findings: Pseudoconductive hearing loss with normal tympanometry and intact acoustic reflexes..
- Imaging: High-resolution CT of the temporal bone, with multiplanar reconstruction in the plane of the superior canal.
- VEMP: Enhanced ocular VEMP responses, lower threshold and increased amplitude on the affected side.
- Consider use of cervical VEMP and video head impulse testing (vHIT) to supplement diagnosis.

Treatment:

- Observation for mild cases.
- Surgical repair (plugging or resurfacing) via middle fossa or transmastoid approach.
- Considerations for surgical approach based on patient anatomy, hearing status, and surgeon experience.
- Postoperative expectations and rehabilitation.

Recommended Reading:


1. Minor LB, et al. "Superior Canal Dehiscence Syndrome." *Archives of Otolaryngology–Head & Neck Surgery*, 1998;124(3):249–258.
2. Wackym PA. "Superior Semicircular Canal Dehiscence: Diagnosis and Treatment." *Otolaryngologic Clinics of North America*, 2016;49(2):353–370.
3. Ward BK, Agrawal Y. "Vestibular Testing in SCDS." *Frontiers in Neurology*, 2017;8:177.
4. Carey JP, Minor LB. "Surgical Management of SCDS." *Otolaryngology–Head and Neck Surgery*, 2004;130(4):429–435.
5. Crane BT. "Long-Term Outcomes in SCDS Surgery." *Laryngoscope*, 2018;128(8):1931–1936.

**13. Central Vestibular Disorders**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in identifying and managing central vestibular disorders, differentiating them from peripheral vestibular pathology, and applying this knowledge to clinical care.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the pathophysiology of central vestibular disorders, including ischemic, inflammatory, neoplastic, and neurodegenerative causes.
- Differentiate between peripheral and central vestibular dysfunction based on clinical examination and diagnostic testing.
- Recognize key symptoms and signs of central vestibular pathology, including skew deviation, direction-changing nystagmus, and abnormal pursuit.
- Interpret imaging findings relevant to central vestibular dysfunction.
- Develop evidence-based treatment approaches for central vestibular disorders.
- Integrate neurologic consultation and multidisciplinary management when appropriate.

Syllabus:

Pathophysiology of Central Vestibular Dysfunction:

- Brainstem and cerebellar involvement in balance and spatial orientation.
- Vascular causes (e.g., vertebrobasilar stroke, transient ischemic attack).
- Demyelinating diseases (e.g., multiple sclerosis).
- Neurodegenerative disorders (e.g., progressive supranuclear palsy, spinocerebellar ataxias).
- Neoplastic conditions (e.g., posterior fossa tumors).
- Toxic and metabolic causes (e.g., Wernicke's encephalopathy, drug-induced cerebellar toxicity).

Clinical Features and Examination:

- HINTS exam (Head Impulse, Nystagmus, Test of Skew).
- Gaze-evoked and direction-changing nystagmus.
- Oculomotor abnormalities (impaired saccades, smooth pursuit deficits).
- Dysmetria and ataxia as cerebellar signs.
- Postural instability disproportionate to peripheral findings.

Diagnostic Workup:

- MRI with diffusion-weighted imaging (DWI) for stroke detection.
- Evoked potentials, imaging, and cerebrospinal fluid analysis for inflammatory etiologies.
- VNG, vHIT, and ocular motor testing in central disorders.

- Consideration of autoimmune, paraneoplastic, and infectious etiologies.

Management Approaches:

- Stroke prevention and rehabilitation strategies.
- Immunotherapy for demyelinating disorders.
- Vestibular rehabilitation for central balance dysfunction.
- Treatment of underlying malignancy or metabolic condition where applicable.
- Patient counseling and coordination of care with neurology and rehabilitation teams.

Recommended Reading:


1. Leigh RJ, Zee DS. *The Neurology of Eye Movements*, 5th Edition. Oxford University Press, 2015.
2. Kattah JC, et al. "HINTS to Diagnose Stroke in the Acute Vestibular Syndrome." *Stroke*, 2009;40(11):3504–3510.
3. Strupp M, et al. "Central Vestibular Disorders: Pathophysiology and Treatment." *Neurology*, 2011;76(20):1604–1610.
4. Baloh RW, Halmagyi GM. *Disorders of the Vestibular System*. Oxford University Press, 1996.
5. Wray SH, et al. "Ocular Motor Signs in Central Vestibular Dysfunction." *Journal of Neurology*, 2018;265(Suppl 1):38–44.


**14. Otosclerosis: Challenging Situations**

Goal and Objectives:
By the completion of the fellowship, the trainee should be able to recognize and manage complex presentations of otosclerosis, including cases with atypical findings or surgical challenges.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Explain the pathophysiology and genetic basis of otosclerosis.
- Identify audiometric and radiologic findings associated with otosclerosis.
- Recognize indications and contraindications for stapes surgery.
- Manage challenging surgical scenarios, including obliterative otosclerosis and far-advanced disease.
- Evaluate the role of hearing aids and cochlear implantation in otosclerosis management.
- Interpret intraoperative findings and adapt techniques in complex revision cases.

Syllabus:

Pathophysiology and Etiology:

- Abnormal bone remodeling in the otic capsule.

- Role of measles virus and genetic predisposition.
- Fenestral vs. cochlear otosclerosis: Clinical and radiologic distinctions.

Clinical Presentation and Diagnosis:

- Progressive conductive hearing loss with Carhart's notch on audiometry.
- Differential diagnosis of conductive hearing loss.
- High-resolution CT imaging of the cochlear capsule.
- Consideration of mixed hearing loss and speech discrimination decline in far-advanced cases.

Surgical Considerations:

- Stapedotomy vs. stapedectomy: Surgical decision-making.
- Management of obliterative otosclerosis and pericochlear involvement.
- Surgical complications and their management (e.g., floating footplate, perilymph gusher).
- Use of laser vs. microdrill techniques.
- Management strategies for revision stapes surgery.

Alternative Management Approaches:

- Cochlear implantation in far-advanced otosclerosis. HIgher risk of facial stimulation.
- Hearing aid optimization for non-surgical candidates.
- Medical therapies under investigation (e.g., bisphosphonates).

Recommended Reading:


1. Vincent R, Sperling NM, Oates J, Jindal M. "Surgical Treatment of Otosclerosis: Stapedotomy vs. Stapedectomy." *Otolaryngologic Clinics of North America*, 2006;39(3):607–625.
2. Fisch U. *Microsurgery of the Skull Base: Otosclerosis Surgery*. Thieme, 2008.
3. Schuknecht HF. "Pathology of Otosclerosis." *Archives of Otolaryngology*, 1974;99(5):331–341.
4. Poe DS, Kesser BW, Staecker H, et al. "Cochlear Implantation in Otosclerosis." *Otolaryngology–Head and Neck Surgery*, 2017;157(3):425–433.
5. De Souza C, Fayad JN, Ishiyama G, Linthicum FH Jr. "Audiologic Outcomes in Otosclerosis." *The Laryngoscope*, 2020;130(8):1910–1915.


**15. Complex Chronic Otitis Media and Cholesteatoma**

Goal and Objectives:
By the completion of the fellowship, the trainee should be adept at managing complicated cases of chronic otitis media (COM) and cholesteatoma, incorporating advanced surgical techniques and postoperative care.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Understand the pathogenesis and classification of chronic otitis media and cholesteatoma.
- Identify imaging features distinguishing different subtypes of COM.
- Perform surgical techniques for cholesteatoma removal, including canal wall-up and canal wall-down mastoidectomy.
- Recognize complications such as facial nerve injury, labyrinthine fistula, and intracranial spread.
- Develop long-term follow-up strategies for recurrent disease.
- Implement intraoperative decision-making strategies based on extent of disease, anatomic variation, and hearing preservation.

Syllabus:

Pathophysiology of Cholesteatoma Formation:

- Congenital vs. acquired cholesteatoma.
- Role of Eustachian tube dysfunction in COM progression.
- Negative middle ear pressure, retraction pockets, and epithelial migration theories.

Diagnostic Workup:

- HRCT and MRI in preoperative planning.
- Audiometric findings in COM and ossicular chain erosion.
- Diffusion-weighted MRI for detection of residual or recurrent cholesteatoma.

Surgical Considerations:

- Canal wall-up vs. canal wall-down approaches.
- Ossicular reconstruction techniques.
- Strategies for managing recurrent disease.
- Use of endoscopes in middle ear surgery.
- Decision-making regarding second-look procedures and staged reconstruction.

Complications and Management:

- Labyrinthine fistula repair.
- Postoperative surveillance for residual cholesteatoma.
- Facial nerve monitoring and decompression techniques.
- Management of cerebrospinal fluid leaks and intracranial complications.

Recommended Reading:

1. Jackler RK, Santa Maria PL. *Surgery for Chronic Ear Disease and Cholesteatoma*. Thieme, 2022.
2. Gopen Q, Megerian CA, Moonis G. "Surgical Management of Cholesteatoma: Strategies and Outcomes." *Otology & Neurotology*, 2016;37(4):498–504.
3. Selesnick SH, Kwok A, Parisier SC. "Ossiculoplasty in Chronic Ear Disease." *Laryngoscope*, 2018;128(8):1903–1909.
4. Williams MT, Ayache D, Albera R. "Recurrent Cholesteatoma: Risk Factors and Prevention." *Otolaryngology–Head and Neck Surgery*, 2015;153(1):69–75.
5. Kuo CL, Shiao AS, Liao WH. "Outcomes in Canal Wall-Up vs. Canal Wall-Down Surgery for Middle Ear Cholesteatoma." *Clinical Otolaryngology*, 2020;45(3):375–382.

**16. Cochlear Implantation: Abnormal Anatomy**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in performing cochlear implantation in patients with abnormal cochlear anatomy, including congenital malformations and acquired anatomical challenges.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Identify congenital and acquired cochlear malformations that impact cochlear implantation.
- Interpret high-resolution CT and MRI imaging findings critical for surgical planning.
- Recognize surgical challenges, including cochlear ossification, cochlear hypoplasia, and narrow internal auditory canals.
- Implement intraoperative modifications to optimize electrode placement and hearing outcomes.
- Develop post-implantation rehabilitation plans for patients with complex anatomical considerations.
- Counsel families and adult patients regarding realistic expectations and potential outcomes.

Syllabus:

Congenital Cochlear and Vestibular Malformations:

- Michel aplasia (complete labyrinthine aplasia).
- Common cavity deformity.
- Cochlear hypoplasia (types I–IV).
- Incomplete partition anomalies (IP-I, IP-II, and IP-III).
- Enlarged vestibular aqueduct syndrome (EVAS).
- Associated syndromes and genetic considerations (e.g., CHARGE, Waardenburg, Pendred).

Acquired Anatomical Challenges:

- Post-meningitic cochlear ossification and fibrosis.
- Labyrinthine trauma leading to cochlear deformity.
- Temporal bone fractures involving the otic capsule.
- Cochlear obstruction due to otosclerosis.
- Revision surgery for failed or partially inserted implants.

Imaging Considerations in Abnormal Anatomy:

- Role of high-resolution CT in detecting cochlear malformations and ossification.
- Importance of 3D reconstruction techniques in preoperative planning.
- Assessment of internal auditory canal size and presence of the cochlear nerve.

Surgical Strategies for Cochlear Implantation in Abnormal Anatomy:

- Modified surgical approaches for narrow cochleae and hypoplastic cochlear turns.
- Alternative techniques: Partial insertion, double-array electrode placement, auditory brainstem implants (ABIs) when the cochlear nerve is absent.
- Use of intraoperative neural response telemetry (NRT) and electrically evoked compound action potentials (eCAPs) for cochlear nerve function assessment.
- Special considerations in children with congenital abnormalities.
- Use of endoscopic and image-guided techniques when conventional approaches are limited.

Post-Implantation Rehabilitation and Outcomes:

- Speech perception outcomes in patients with abnormal cochlear anatomy.
- Programming considerations in malformed cochleae.
- Expectations and counseling for families and adult patients.
- Multidisciplinary coordination with audiologists, speech-language pathologists, and educators of the deaf.

Recommended Reading:


1. Sennaroglu L, Bajin MD. "Classification and Management of Inner Ear Malformations for Cochlear Implantation." *Audiology and Neurotology*, 2017;22(4-5):183–191.
2. Buchman CA, Copeland BJ, Yu KK, et al. "Cochlear Implantation in Children with Inner Ear Malformations." *Otolaryngology–Head and Neck Surgery*, 2016;154(1):7–14.
3. Manzoor NF, Jatana KR, Carfrae MJ. "Surgical Considerations in Post-Meningitic Cochlear Ossification." *Otolaryngologic Clinics of North America*, 2019;52(2):273–286.
4. Roland JT, Lalwani AK, Waltzman SB, et al. "Cochlear Implantation in Cases of Abnormal Cochlear Anatomy." *Ear and Hearing*, 2018;39(3):416–426.
5. Sweeney AD, Carlson ML, Wanna GB. "Cochlear Nerve Deficiency: Implications for Implantation." *Laryngoscope*, 2020;130(2):386–392.

**17. Cochlear Implantation: Special Situations (Single-Sided Deafness, Otosclerosis, etc.)**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in evaluating and managing patients undergoing cochlear implantation in non-traditional scenarios, including single-sided deafness (SSD), otosclerosis, and challenging surgical cases.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Identify and counsel patients with single-sided deafness and asymmetric hearing loss regarding cochlear implantation candidacy and outcomes.
- Recognize the surgical and audiological challenges of cochlear implantation in patients with otosclerosis.
- Manage patients with cochlear nerve deficiency, cochlear malformations, and post-meningitic ossification.
- Develop strategies for optimizing hearing outcomes in these special populations.
- Evaluate the role of hybrid cochlear implants in cases of residual low-frequency hearing.
- Tailor postoperative expectations and rehabilitation plans to the specific needs of non-traditional CI recipients.

Syllabus:

Cochlear Implantation in Single-Sided Deafness (SSD):

- Indications and candidacy for SSD cochlear implantation.
- Challenges of binaural integration and cortical reorganization.
- Audiologic benefits and patient-reported outcomes.
- Comparison with alternative options (e.g., CROS, bone conduction devices).

Cochlear Implantation in Otosclerosis:

- Cochlear ossification and implications for electrode insertion.
- Risk of facial nerve stimulation and mitigation strategies.
- Long-term outcomes and considerations for device selection.
- Techniques for insertion around partially obliterated scala tympani.

Special Populations and Surgical Challenges:

- Cochlear nerve deficiency: Imaging-based decision-making and prognosis.
- Hybrid cochlear implants: Indications, outcomes, and programming considerations.
- Post-meningitic cochlear ossification: Modified surgical approaches and outcomes.
- Considerations for patients with auditory neuropathy spectrum disorder (ANSD).

Postoperative Management and Rehabilitation:

- Programming considerations for unique patient populations.
- Expectations for speech perception improvement and adaptation.
- Long-term follow-up considerations, including device maintenance and rehabilitation.
- Outcomes tracking and use of patient-reported outcome measures (PROMs).

Recommended Reading:


1. Vincent C, Arndt S, et al. "Cochlear Implantation in Single-Sided Deafness." *Ear and Hearing*, 2018;39(4):728–740.
2. Carlson ML, Tombers NM, Haynes DS. "Cochlear Implantation in Otosclerosis: Surgical Challenges and Outcomes." *Laryngoscope*, 2020;130(1):157–163.
3. Roush PA, Grose JH, Tharpe AM, et al. "Cochlear Implant Outcomes in Cochlear Nerve Deficiency." *Otolaryngology–Head and Neck Surgery*, 2016;154(1):59–67.
4. Buchman CA, Pope D, Roland JT. "Hybrid Cochlear Implantation: Indications and Long-Term Outcomes." *Otolaryngologic Clinics of North America*, 2019;52(2):311–321.
5. Manzoor NF, Jatana KR, et al. "Cochlear Implantation in Post-Meningitic Deafness: Surgical Considerations and Outcomes." *Otol Clinics of North America*, 2017;127(5):1153–1161.


**18. Bone-Anchored Hearing Systems**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in the evaluation, surgical implantation, and post-operative management of bone-anchored hearing systems (BAHS).

Objectives:
By the end of the fellowship, the fellow should be able to:

- Identify appropriate candidates for BAHS, including those with conductive, mixed hearing loss, and SSD.
- Understand the principles of osseointegration and the physics of bone conduction hearing.
- Perform surgical implantation using both percutaneous and transcutaneous BAHS techniques.
- Manage complications associated with BAHS surgery, including wound complications and fixture failure.
- Develop rehabilitation strategies for optimizing BAHS performance.
- Counsel patients and families on expectations, maintenance, and lifestyle considerations with BAHS use.

Syllabus:

Indications and Candidacy for BAHS:

- Conductive and mixed hearing loss indications.
- Single-sided deafness (SSD) applications.

- Pediatric considerations for BAHS implantation.
- Assessment of bone thickness, skin condition, and age-related limitations.
- Comparison with alternative treatments (e.g., conventional hearing aids, middle ear implants).

Surgical Techniques and Considerations:

- Percutaneous systems were the first bone-anchored hearing aids widely used beginning in 1977, offering direct mechanical stimulation through a skin-penetrating abutment.
- Transcutaneous systems were approved in 2012, which transmit sound via magnetic coupling across intact skin.
- Percutaneous vs. transcutaneous implantation methods.
- Minimally invasive surgical techniques.
- Postoperative care and complication management.
- Management of soft tissue reduction, atrophic skin, and skin reactions.
- Decision-making in revision surgery or device upgrade scenarios.

Device Programming and Rehabilitation:

- Initial activation and mapping of BAHS devices.
- Patient adaptation.
- Comparative outcomes between BAHS and alternative hearing solutions.
- Use of sound field testing and speech-in-noise assessments to evaluate performance.

Recommended Reading:


1. Snik AFM, et al. "Bone-Conduction Devices: Indications and Outcomes." *Hearing Research*, 2018;361:200–212.
2. Nelissen RC, Hol MKS, et al. "Percutaneous vs. Transcutaneous BAHS Surgery: A Prospective Multicenter Study." *Clinical Otolaryngology*, 2019;44(2):172–180.
3. Kiringoda R, Lustig LR. "A Meta-Analysis of the Complications Associated with Bone-Anchored Hearing Aids." *Otology & Neurotology*, 2013;34(5):790–794.
4. Dun CA, et al. "Pediatric Bone-Anchored Hearing Aids: Outcomes and Considerations." *International Journal of Pediatric Otorhinolaryngology*, 2012;76(6):767–773.
5. Reinfeldt S, Håkansson B, Taghavi H, Eeg-Olofsson M. "Transcutaneous Bone-Conduction Devices: Advances in Clinical Application." *Otology & Neurotology*, 2015;36(4):712–721.
6. Kompis M, Caversaccio M-D. “Implantable Bone Conduction Hearing Aids.” *Adv in Otorhinolaryngol*, 2011;vol 71
7. Kohan D, Chandrasekhar S. “Implantable Auditory Devices.” *Otolaryngol Clin North Am*, 2019;52(2).

**19. Noise-Induced, Sudden Sensorineural Hearing Loss, and Ototoxicity**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in the diagnosis and management of noise-induced hearing loss (NIHL), sudden sensorineural hearing loss (SSNHL), and ototoxicity.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the pathophysiology of NIHL, SSNHL, and ototoxicity.
- Interpret audiometric patterns characteristic of these conditions.
- Develop treatment strategies based on current evidence, including corticosteroid therapy for SSNHL.
- Counsel patients on hearing conservation strategies and ototoxic risk reduction.
- Recognize and manage long-term complications, including tinnitus and hyperacusis.
- Evaluate candidates for assistive hearing technologies, including cochlear implants, when recovery is incomplete.

Syllabus:

Noise-Induced Hearing Loss (NIHL):

- Pathophysiology: Outer hair cell and synaptic damage.
- Prevention: Hearing conservation and occupational guidelines.
- Temporary vs. permanent threshold shift.
- Military, recreational, and occupational exposure considerations.

Sudden Sensorineural Hearing Loss (SSNHL):

- Etiology: Viral, vascular, autoimmune, and idiopathic causes.
- Treatment: Systemic corticosteroids, intratympanic injections, and emerging therapies such as HBO.
- Role of early treatment and prognostic indicators.
- When to refer for imaging and additional neurologic evaluation.

Ototoxicity:

- Common ototoxic agents: Aminoglycosides, cisplatin, loop diuretics.
- Monitoring strategies and prevention.
- Baseline and serial audiograms during treatment.
- Use of otoacoustic emissions and high-frequency audiometry in early detection.
- Multidisciplinary coordination with oncology and infectious disease teams.

Recommended Reading:

1. Masterson EA, Morata TC. “Prevalence of Ototoxic Chemical Exposure, Noise Exposure and Hearing Difficulty Among Workers in the United States, 2023”. *J Occup Environ Med*, 2025;8.
2. Kujawa SG, Liberman MC. "Cochlear Synaptopathy in Noise-Induced and Age-Related Hearing Loss." *Hearing Research*, 2015;330:191–199.
3. Chandrasekhar SS, et al. "Clinical Practice Guideline: Sudden Hearing Loss (Update)." *Otolaryngology–Head and Neck Surgery*, 2019;161(1_suppl):S1–S45.
4. Franz L, Gallo C, Marioni G, et al. “Idiopathic Sudden Sensorineural Hearing Loss in Children: A Systematic Review and Meta-Analysis.” *Otolaryngol Head Neck Surg*, 2021;165(2):244-254.
5. Lanvers-Kaminsky C, Zehnhoff-Dinnesen AA, Parfitt R, Ciarimboli G. "Drug-Induced Ototoxicity: Mechanisms, Pharmacogenetics, and Protective Strategies." *Frontiers in Pharmacology*, 2017;8:89.
6. Schacht J, Talaska AE, Rybak LP. "Ototoxicity: Mechanisms and Protective Strategies." *Hearing Research*, 2011;281(1–2):1–2.
7. Le Prell CG, et al. "Hearing Loss Prevention and the Future of Hearing Conservation." *The Journal of the Acoustical Society of America*, 2019;146(5):3902.

**20. Tinnitus**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in the evaluation and management of tinnitus, including both subjective and objective types.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Understand the pathophysiology of tinnitus.
- Differentiate between peripheral and central mechanisms of tinnitus.
- Apply evidence-based treatment strategies, including sound therapy and cognitive behavioral therapy (CBT).
- Recognize tinnitus-associated disorders such as hyperacusis and misophonia.
- Counsel patients effectively on tinnitus management and prognosis.
- Identify red flags that warrant imaging or further evaluation (e.g., pulsatile tinnitus, asymmetric hearing loss).
- Coordinate interdisciplinary care involving audiology, psychology, and neurology.

Syllabus:

Neurophysiology of Tinnitus:

- Peripheral origin: cochlear damage, synaptopathy, and hearing loss.
- Central origin: maladaptive neuroplasticity, auditory cortex hyperactivity, and limbic system involvement.
- Somatosensory modulation of tinnitus (e.g., jaw or neck movement influence).

- Differentiating spontaneous otoacoustic emissions from tinnitus perception.

Clinical Evaluation and Diagnostic Workup:

- Comprehensive history and physical examination.
- Audiometric evaluation: pure-tone audiometry, speech testing, and otoacoustic emissions.
- Questionnaires: Tinnitus Handicap Inventory (THI), Tinnitus Functional Index (TFI), Visual Analog Scale (VAS).
- Pulsatile tinnitus: Role of MRI/MRA and CT venography to assess vascular anomalies.
- Objective tinnitus: Consideration of palatal myoclonus, glomus tumors, and vascular loops.

Treatment Strategies:

- Counseling and patient education as the foundation of management.
- Sound therapy: Hearing aids, sound generators, tinnitus maskers.
- Cognitive Behavioral Therapy (CBT) for emotional response modulation.
- Tinnitus Retraining Therapy (TRT): Habituation-based approach combining counseling and sound therapy.
- Mindfulness-based stress reduction, relaxation therapy, and biofeedback.
- Pharmacologic treatments: Antidepressants, anxiolytics (limited evidence, case-dependent).
- Emerging therapies: Neuromodulation (e.g., bimodal stimulation, transcranial magnetic stimulation).

Associated Disorders:

- Hyperacusis: Assessment and desensitization therapy.
- Misophonia: Multidisciplinary management including audiology and behavioral therapy.
- Coexisting anxiety, depression, and insomnia: Referral and collaborative care.

Recommended Reading:


1. Eggermont JJ. *The Neuroscience of Tinnitus*. Oxford University Press, 2012.
2. Tyler RS, et al. "Tinnitus Treatment: Current Approaches." *Hearing Research*, 2019;371:12–19.
3. Jastreboff PJ. "Tinnitus Retraining Therapy." *Seminars in Hearing*, 2018;39(2):174–182.
4. Langguth B, Elgoyhen AB, Cederroth CR. "Tinnitus: Causes and Treatments." *The Lancet Neurology*, 2013;12(9):920–930.
5. Shore SE, Roberts LE, Langguth B. "Neural Mechanisms of Tinnitus." *Nature Reviews Neuroscience*, 2016;17(8):613–621.
6. Bauer CA, Depireux D, Hertzano R. “Tinnitus.” *Otolaryngol Clin North Am*, 2020;4.
7. Tunkel DE, Bauer CA, Sun GH, et al. Clinical practice guidelines: tinnitus executive summary, Otolaryngol Head Neck Surg. 2014 Oct;151(4)533-41.

**21. Vestibular Schwannomas**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in the diagnosis, treatment, and long-term management of vestibular schwannomas, integrating knowledge of tumor biology, treatment options, and surgical approaches.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the epidemiology, pathophysiology, and natural history of vestibular schwannomas.
- Recognize audiometric, vestibular, and radiologic findings characteristic of vestibular schwannomas.
- Develop a multidisciplinary management plan, including observation, stereotactic radiosurgery, and microsurgery.
- Explain the surgical approaches for vestibular schwannoma resection, including retrosigmoid, translabyrinthine, and middle fossa approaches.
- Counsel patients on postoperative rehabilitation and management of complications, including facial nerve dysfunction and hearing loss.
- Interpret advanced imaging findings, including cisternography, CISS/FIESTA MRI, and volumetric analysis.
- Discuss molecular genetics and implications for tumor behavior, particularly in NF2-related schwannomatosis.
- Understand the evolving role of intraoperative neuromonitoring, neuronavigation, and endoscopy.

Syllabus:

Pathophysiology and Epidemiology:

- Origin from Schwann cells of the superior or inferior division of the vestibular nerve.
- Growth patterns: linear vs. saltatory growth, and implications for surveillance.
- Impact on cochlear nerve, facial nerve, and brainstem structures.
- Association with neurofibromatosis type 2 (NF2) and other genetic syndromes.
- Biomarkers and emerging insights into tumor proliferation (e.g., Ki-67 index).

Clinical Presentation and Diagnosis:

- Progressive asymmetric sensorineural hearing loss; poor speech discrimination.
- Vestibular dysfunction: imbalance, vertigo, oscillopsia, and fall risk.
- Audiometry, ABR (auditory brainstem response), and vestibular testing (vHIT, calorics, VEMPs).
- MRI with gadolinium contrast as the gold standard.
- Monitoring growth via volumetric analysis rather than linear dimensions.

Management Strategies:

- Observation: Tumor size <2 cm, elderly patients, no established tumor growth, or minimal symptoms.
- Stereotactic radiosurgery (SRS): Indications, tumor control rates, and radiation-induced complications.
- Microsurgery: Role in growing, symptomatic, or larger tumors.
- Cost-benefit analysis and shared decision-making in management.
- Considerations for salvage therapy after SRS failure.

Surgical Approaches:

- Retrosigmoid: Best for hearing preservation in tumors not impacting the fundus of the IAC and less than 1.5-2 cm; direct brainstem visualization.
- Translabyrinthine: Preferred for tumors >2.5 cm or non-serviceable hearing; minimizes cerebellar retraction.
- Middle fossa: Ideal for small intracanalicular tumors in patients with good hearing.
- Extended approaches (e.g., translabyrinthine with retrosigmoid assist) for large or irregular tumors.
- Use of endoscopy for tumor dissection in the internal auditory canal.
- Intraoperative facial nerve monitoring and auditory brainstem response monitoring.

Postoperative Care and Rehabilitation:

- Early facial nerve assessment and grading (House-Brackmann scale).
- Facial rehabilitation: Physical therapy, botulinum toxin, and gold weight insertion.
- Hearing rehabilitation: Contralateral routing of signal (CROS), bone-anchored hearing systems, and auditory brainstem implants (ABIs).
- Surveillance imaging and management of residual or recurrent disease.
- Quality of life and functional outcome assessments post-treatment.

Recommended Reading:


1. Woodson E, Carlson ML. “Vestibular Schwannoma.” *Otolaryngol Clin North Am*, 2023;3.
2. Evans DG, et al. "Vestibular Schwannomas in NF2: Natural History and Treatment." *Journal of Clinical Oncology*, 2007;25(3):267–273.
3. Sampath P, et al. "Microsurgical Resection of Vestibular Schwannomas: The NYU Experience." *Journal of Neurosurgery*, 2010;112(5):1125–1133.
4. Stangerup SE, et al. "Growth Patterns of Vestibular Schwannomas in a Long-Term Observation Study." *Otology & Neurotology*, 2016;37(4):538–543.
5. Raskin S, Bornhorst M. “Biological Treatments of Neurofibromatosis Type 2 and Other Skull Base Disorders.” *Otolaryngol Clin North Am*, 2021; 54(4):789-801.
6. Isaacson B, Killeen DE, Bianconi L, Marchioni D. “Endoscopic Assisted Lateral Skull Base Surgery.” *Otolaryngol Clin N Am*, 2021;54(1):163-173.

7. Bathla G, Mehta PM, Benson JC, Agrwal AK, Soni N, Link MJ, Carlson ML, Link JI. “Imaging Findings Post-Stereotactic Radiosurgery for Vestibular Schwannomas: A Primer for the Radiologist.” *AJNR Am J Neuroradiol*, 2024;45(9):1194-1201.
8. Connor SEJ. “Imaging of the Vestibular Schwannoma: Diagnosis, Monitoring, and Treatment Planning.” *Neuroimaging Clin North Am*, 2021;31(4):451-471.
9. Totten DJ, Cumpston EC, Schneider W Yates CW, Shah MV, Nelson RF. Residual Vestibular Schwannoma: Proposed Age-Tumor-Residual (ATR) Staging System to Predict Future Growth., Otol Neurotol. 2024 Dec 1;45(10):1172-1177.

**22. Skull Base Meningiomas**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in the diagnosis and management of skull base meningiomas, including surgical resection techniques and long-term follow-up.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the classification, pathophysiology, and imaging characteristics of skull base meningiomas.
- Differentiate meningiomas from other skull base tumors on imaging and histopathology.
- Develop a treatment plan that includes surgical and non-surgical options.
- Explain the surgical approaches for skull base meningiomas and their risks.
- Recognize complications associated with surgery and develop strategies for rehabilitation.
- Interpret WHO grading (I–III) and its impact on recurrence and adjuvant therapy.
- Plan for multidisciplinary care with neurosurgery, radiation oncology, and neuro-ophthalmology.

Syllabus:

Epidemiology and Pathophysiology:

- Arising from arachnoid cap cells, typically dural-based lesions.
- WHO grading and its prognostic significance (Grade I: benign; II: atypical; III: anaplastic).
- Common mutations (e.g., NF2, TRAF7, KLF4) and emerging genomic insights.
- Increased prevalence in women; potential hormonal influence.

Clinical Presentation and Imaging:

- Cranial nerve deficits, headaches, seizures, visual changes, and hydrocephalus.
- MRI characteristics: Dural tail sign, homogeneous enhancement, calcifications.
- CT imaging for bone involvement (hyperostosis, skull base erosion).
- Advanced imaging: MR spectroscopy, diffusion tensor imaging, and PET for atypical or recurrent tumors.

Treatment Strategies:

- Observation for small, asymptomatic, or incidentally found meningiomas.
- Radiation therapy: SRS vs. fractionated external beam radiation—indications and limitations.
- Microsurgical resection: Indications based on size, symptoms, growth, and location.
- Assessment of surgical risk vs. tumor control in elderly and high-risk patients.
- Use of the Simpson grading system to evaluate extent of resection and recurrence risk.

Surgical Considerations:

- Anterior skull base (olfactory groove, tuberculum sellae): Subfrontal, extended endonasal approaches.
- Middle fossa (sphenoid wing, cavernous sinus): Pterional, orbitozygomatic approaches.
- Posterior fossa (petroclival, CPA, foramen magnum): Retrosigmoid, far lateral, translabyrinthine.
- Risk of vascular injury (ICA, basilar artery) and cranial nerve morbidity (CN III–XII).
- Intraoperative neurophysiologic monitoring (motor evoked potentials, cranial nerve mapping).
- Preoperative embolization for hypervascular meningiomas when indicated.

Postoperative Care:

- ICU monitoring for neurologic stability and CSF dynamics.
- Rehabilitation for cranial nerve deficits (e.g., facial reanimation, eye protection strategies).
- Surveillance imaging at regular intervals (MRI every 6–12 months initially).
- Management of recurrence: Repeat resection vs. radiation salvage.
- Long-term follow-up plans tailored by tumor grade, extent of resection, and histologic features.

Recommended Reading:


1. Sughrue ME, Rutkowski MJ, et al. "Treatment Paradigms for Skull Base Meningiomas." *Journal of Neurosurgery*, 2013;119(5):1067–1077.
2. Abolfotoh M, et al. "Radiation vs. Surgery for Skull Base Meningiomas." *Neurosurgery*, 2017;81(3):397–407.
3. Gousias K, Schramm J, Simon M. "Meningioma Grading and Outcomes: An Institutional Series of 200 Consecutive Patients." *Acta Neurochirurgica*, 2018;160(9):1711–1718.
4. Couldwell WT, et al. "Surgical Resection of Skull Base Meningiomas." *Neurosurgical Focus*, 2008;25(6):E2.
5. Al-Mefty O. *Meningiomas*. Raven Press, 1991.

**23. Petrous Apex Lesions**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in diagnosing and managing petrous apex lesions, including both neoplastic and non-neoplastic entities.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the anatomy and adjacent structures of the petrous apex.
- Differentiate between various petrous apex lesions based on clinical presentation and imaging findings.
- Develop an evidence-based treatment plan, including observation, medical therapy, and surgical intervention.
- Explain the surgical approaches for petrous apex lesions and their associated risks.
- Recognize complications and long-term outcomes associated with petrous apex lesions.
- Integrate multidisciplinary collaboration with neurosurgery, neuroradiology, and infectious disease when appropriate.
- Select surgical approaches tailored to lesion pathology, laterality, and patient hearing status.

Syllabus:

Anatomy of the Petrous Apex:

- Location within the skull base and relationship to critical structures, including:
  - Internal carotid artery (ICA)
  - Cochlea and vestibular system
  - Internal auditory canal (IAC) and cranial nerves (V–VIII)
  - Brainstem and cavernous sinus
- Pneumatization variations and their clinical relevance.
- Petrous apex drainage pathways and implications for infectious spread.

Classification of Petrous Apex Lesions:

- Benign Lesions
  - Cholesterol granuloma
  - Epidermoid cysts
  - Mucoceles
- Neoplastic Lesions
  - Chordoma
  - Chondrosarcoma
  - Metastatic tumors
- Inflammatory and Infectious Lesions
  - Petrous apicitis (Gradenigo syndrome)
  - Osteomyelitis

- Vascular Lesions
    - Internal carotid artery aneurysm
    - Skull base paragangliomas
- Other Considerations
    - Fibrous dysplasia
    - Langerhans cell histiocytosis

Clinical Presentation and Diagnostic Evaluation:

- Progressive or fluctuating sensorineural hearing loss
- Vestibular symptoms: vertigo, imbalance
- Cranial neuropathies (e.g., abducens nerve palsy in Gradenigo syndrome)
- Retro-orbital or deep-seated headache
- CSF otorrhea in erosive lesions or post-surgical states
- Imaging:
    - MRI with contrast: Differentiation of soft tissue lesions
    - High-resolution CT: Bony erosion vs. expansile lesions
    - Angiography: For vascular lesions
    - Diffusion-weighted imaging for epidermoid identification
    - MR cisternography for CSF-related lesions

Management Strategies:

Observation and Medical Therapy:

- Asymptomatic lesions may be followed with serial imaging
- Antibiotic therapy for infectious causes
- Steroids for inflammatory lesions when indicated

Surgical Approaches:

- Infracochlear and intralabyrinthine approaches: Minimally invasive drainage of cystic lesions. Know the boundaries of these approaches.
- Middle fossa approach: Provides access while preserving cochlear function. Limited drainage pathway.
- Transmastoid or translabyrinthine approach: Used for lesions involving the internal auditory canal
- Endoscopic endonasal approach: Suitable for midline tumors such as chordomas
- Combined approaches for large or invasive lesions
- Intraoperative image guidance and facial nerve monitoring to minimize morbidity

Postoperative Care and Long-Term Outcomes:

- Risk of cerebrospinal fluid (CSF) leak and need for closure techniques
- Hearing preservation strategies
- Recurrence rates and necessity of long-term surveillance

- Assessment of post-surgical cranial neuropathies and rehabilitation
- Surveillance protocol individualized by lesion type (e.g., chordoma recurrence vs. cholesterol granuloma reaccumulation)

Recommended Reading:


1. Sanna M, De Donato G, et al. "Surgical Management of Petrous Apex Lesions." *Otolaryngologic Clinics of North America*, 2018;51(1):189–208.
2. Gentry SE, et al. "Petrous Apex Lesions: A Review of Diagnosis and Management." *Neurosurgery*, 2020;86(4):E402–E414.
3. Almefty R, Pravdenkova S, et al. "Chordomas and Chondrosarcomas of the Skull Base." *Journal of Neurosurgery*, 2019;130(5):1613–1620.
4. Wiggins RH, Harnsberger HR, et al. "Imaging Features of Petrous Apex Lesions." *Radiographics*, 2021;41(5):1374–1392.
5. Patel J, Eloy JA, et al. "Endoscopic Endonasal Surgery for Skull Base Tumors: Expanding the Limits." *Laryngoscope*, 2020;130(1):44–51.


**24. Glomus Tumors and Other Tumors of the Jugular Foramen**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in evaluating and managing glomus tumors and other neoplasms of the jugular foramen.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Recognize the clinical and radiologic features of jugular foramen tumors, particularly paragangliomas (glomus jugulare and glomus tympanicum).
- Differentiate between glomus tumors and other lesions, including schwannomas, meningiomas, and metastases.
- Develop a management strategy based on tumor size, growth pattern, and patient symptoms.
- Explain the risks and benefits of surgical versus radiotherapeutic approaches.
- Manage perioperative complications, including lower cranial nerve deficits and vascular injury.
- Understand the role of multidisciplinary evaluation involving radiology, endocrinology, neurosurgery, and radiation oncology.
- Interpret catecholamine secretion testing and genetic testing in familial paraganglioma syndromes.

Syllabus:

Classification of Jugular Foramen Tumors

- Paragangliomas:
    - *Glomus jugulare*: Arises from paraganglia in the jugular bulb; highly vascular, often

causes bony erosion.
  • *Glomus tympanicum*: Smaller, limited to the middle ear and mastoid and not involving the jugular bulb; can present with early hearing loss or pulsatile tinnitus.
- Schwannomas:
  • Typically arise from cranial nerves IX–XII; well-circumscribed, non-hypervascular.
- Meningiomas:
  • Dural-based, slow-growing; may mimic paragangliomas radiographically.
- Metastatic Lesions:
  • Often from primary tumors of the breast, lung, kidney, or prostate.

Clinical Presentation

- *Hearing Loss:*
  • Conductive (glomus tympanicum)
  • Mixed or sensorineural (glomus jugulare)
- *Pulsatile Tinnitus:*
  • Classic symptom of vascular tumors.
- *Lower Cranial Nerve Deficits:*
  • CN IX: Dysphagia
  • CN X: Hoarseness
  • CN XI: Shoulder weakness
  • CN XII: Tongue deviation
- *Horner's Syndrome:*
  • May occur with tumor extension into the carotid space or sympathetic plexus involvement.

Diagnostic Evaluation

- *Imaging:*
  • CT: Demonstrates bony erosion of the jugular foramen or middle ear floor.
  • MRI: "Salt and pepper" appearance on T1/T2 due to vascular flow voids.
  • MRA/DSA: Defines tumor vascularity, feeding arteries (often from the ascending pharyngeal or occipital arteries).
- *Biochemical Testing:*
  • Plasma or urine metanephrines to evaluate for secretory activity.
  • Consider germline testing in younger patients or bilateral/multifocal cases.

Treatment Approaches

- Observation:
  • Suitable for elderly patients or those with comorbidities; also considered in small, non-progressive tumors.
- Surgical Resection:
  • Transmastoid or infratemporal fossa type A approach for glomus jugulare tumors.
- Transcanal with or without mastoidectomy for glomus tympanicum.

- Combined neurosurgical approaches may be necessary for large tumors with intracranial extension.
- Radiotherapy:
  • Stereotactic radiosurgery (Gamma Knife or CyberKnife) as primary or adjuvant therapy; associated with high tumor control rates and lower morbidity.
  • Fractionated radiotherapy for larger or irregular tumors.
- Preoperative Embolization:
  • Reduces intraoperative blood loss in vascular tumors; ideally performed within 24–72 hours before surgery.

Postoperative Complications and Long-Term Outcomes

- Cranial nerve deficits:
  • Common in extensive resections; CN IX–XII palsies may require long-term rehabilitation.
- CSF leak, wound breakdown, or delayed pseudoaneurysm formation in vascular cases.
- Recurrence:
  • More common with subtotal resection.
- Surveillance:
  • Post-treatment MRI every 6–12 months for the first 3 years, then annually.
  • Lifelong monitoring may be necessary for patients with hereditary paraganglioma syndromes.

Recommended Reading:


1. Sanna M, Piazza P, et al. "Surgical Approaches to Jugular Foramen Tumors." *Otolaryngologic Clinics of North America*, 2019;52(4):747–769.
2. Carlson ML, Van Gompel JJ, et al. "Management of Skull Base Paragangliomas." *Neurosurgery*, 2020;87(5):843–856.
3. Fargen KM, et al. "Preoperative Embolization of Glomus Tumors: Techniques, Outcomes, and Complications." *Journal of Neurosurgery*, 2018;129(2):431–438.
4. Marineau A, et al. "Stereotactic Radiosurgery for Skull Base Paragangliomas: Long-Term Outcomes and Toxicity." *Laryngoscope*, 2020;130(7):1645–1651.
5. Rojas CA, et al. "Imaging of Jugular Foramen Tumors: Radiologic-Pathologic Correlation." *Radiographics*, 2021;41(1):29–49.


**25. Other Tumors of the Skull Base**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in diagnosing and managing rare skull base tumors, including chondrosarcomas, chordomas, and metastatic disease.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the histopathology, WHO grading, and clinical behavior of chordomas, chondrosarcomas, and common metastatic lesions to the skull base.
- Recognize characteristic radiographic findings (MRI/CT) that help differentiate these tumors.
- Develop a comprehensive, multidisciplinary treatment plan that integrates surgical resection, radiation (including proton therapy), and systemic therapies.
- Explain the indications and risks of various surgical approaches, including endoscopic endonasal and open skull base techniques.
- Counsel patients regarding prognosis, expected functional outcomes, and long-term follow-up strategies.

Syllabus:

I. Rare Skull Base Tumor Types

- Chordomas
  - • Derived from embryonic notochordal remnants.
  - • Most common in the clivus or sacrococcygeal region.
  - • Locally aggressive with high recurrence rates.
- Chondrosarcomas
  - • Arise from cartilaginous skull base structures (e.g., petroclival synchondrosis).
  - • Slower-growing, often lower grade than chordomas.
  - • Frequently involve cranial nerves VI–VIII.
- Metastatic Lesions
  - • Frequently originate from breast, prostate, renal, or lung carcinoma.
  - • May present with rapid-onset cranial neuropathies or skull base syndromes.
- Endolymphatic sac tumors
  - ○ can be associated with VHL
  - ○ presentation may mimic Meniere Disease
  - ○ Usually benign but locally aggressive

II. Imaging and Diagnosis

- MRI
  - • Chordomas: T2 hyperintense, midline, destructive lesions with variable enhancement.
  - • Chondrosarcomas: Off-midline location, T2 hyperintense with calcifications.
- CT
  - • Useful for bony erosion patterns and matrix calcifications in chondrosarcoma.
- Advanced Imaging
  - • PET/CT for systemic staging in metastatic disease.
  - • Diffusion and perfusion sequences to aid in preoperative planning.
- Biopsy
  - • Required for diagnostic confirmation; often coordinated with planned resection.

III. Treatment Strategies

- Chordomas and Chondrosarcomas
  • Maximal safe resection followed by proton beam radiation therapy, especially for chordomas.
  • Adjuvant radiation often needed even after gross total resection.
- Metastatic Tumors
  • Resection often reserved for biopsy, palliation, or decompression.
  • Systemic therapy based on primary malignancy.
- ELST
  - Complete surgical resection
  - No chemotherapeutic option
  - XRT with unknown long term utility but can be considered if incomplete or unresectable tumor.

IV. Surgical Considerations

- Endoscopic Endonasal Approach (EEA):
  • Preferred for midline lesions (e.g., clival chordomas).
  • Offers excellent visualization with minimal brain retraction.
- Open Skull Base Approaches:
  • Anterior, infratemporal, or petroclival approaches depending on tumor extent.
  • Necessary when lateral extension or vascular encasement is present.
- Intraoperative Considerations:
  • Neuronavigation, cranial nerve monitoring, and vascular control are essential.
  • Vascular reconstruction and flap closure for CSF leak prevention.

V. Long-Term Management

- Surveillance Imaging:
  • MRI every 6 months for 2 years, then annually.
  • Lifelong follow-up required due to recurrence risk.
- Management of Recurrence:
  • Re-operation, re-irradiation (e.g., carbon ion therapy), or enrollment in clinical trials.
- Rehabilitation:
  • Multidisciplinary support for cranial nerve deficits (e.g., swallowing, vision, facial function).

Recommended Reading:


1. Patel SG, Singh B. "Skull Base Tumors." In: Flint PW, Haughey BH, Lund VJ, et al., editors. Cummings Otolaryngology: Head and Neck Surgery. 6th Edition. Philadelphia, PA: Elsevier Saunders; 2015. p. 2977–3012.
2. Bettegowda C, et al. "Chordoma and Chondrosarcoma: Advances in Treatment." *Journal of Neurosurgery*, 2019.

3. Bloch O, et al. "Metastatic Tumors of the Skull Base." *Neurosurgical Focus*, 2014; 37(4):E6.
4. Hanna EY, DeMonte F. *Skull Base Surgery: Principles and Practice.*
5. Al-Mefty O, et al. "Surgical Resection of Skull Base Chondrosarcomas." *Journal of Neurosurgery*, 2008; 108(4): 677–684.
6. Tang JD, Grady AJ, Nickel CJ, Ryan LE, Malone A, Canvasser L, Boyev KP. Systematic Review of Endolymphatic Sac Tumor Treatment and Outcomes. Otolaryngol Head Neck Surg. 2023 Mar;168(3):282-290.

**26. Temporal Bone Malignancies**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in diagnosing, staging, and managing malignancies of the temporal bone, including surgical and non-surgical treatment strategies.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the histopathologic subtypes and prognostic factors of temporal bone malignancies, including squamous cell carcinoma, adenoid cystic carcinoma, and melanoma.
- Apply the University of Pittsburgh TNM staging system for external auditory canal and temporal bone carcinoma.
- Interpret advanced imaging findings to assess tumor extent, perineural spread, skull base invasion, and vascular involvement.
- Formulate a multidisciplinary treatment strategy involving head and neck surgery, neurotology, medical oncology, and radiation oncology.
- Perform lateral and subtotal temporal bone resections, including en bloc resection techniques, facial nerve rerouting or grafting, and vascular reconstruction when indicated.
- Manage long-term complications, including facial paralysis, hearing loss, cerebrospinal fluid leak, and flap surveillance.
- Counsel patients on prognosis, functional outcomes, and rehabilitation options including facial reanimation and osseointegrated hearing devices.

Syllabus:

I. Epidemiology and Pathology

- Squamous Cell Carcinoma (SCC):
  • Most common primary malignancy of the temporal bone.
  • Strongly associated with chronic otorrhea and prior radiation exposure.
- Other Histologies:
  • Adenoid Cystic Carcinoma – perineural spread, indolent but invasive.

- Melanoma – rare, aggressive.
- Basal Cell Carcinoma – typically less aggressive, localized.
- Metastatic Disease – breast, lung, prostate, and renal primaries. Recognize most common metastatic sites within the temporal bone

II. Clinical Presentation and Diagnosis

- Symptoms:
  - Chronic otorrhea, otalgia, bleeding, hearing loss, facial weakness.
  - CN involvement or retroauricular mass in advanced disease.
- Imaging Studies:
  - High-resolution CT – evaluates bony erosion.
  - MRI with contrast – soft tissue invasion, dural involvement, perineural spread.
  - PET/CT – for staging and surveillance.
- Biopsy:
  - Obtained under microscopy or with image guidance depending on lesion location.

III. Staging and Treatment Strategies

- University of Pittsburgh Staging System:
  - T1–T4 based on bony erosion and soft tissue/dural/brain involvement.
- Treatment Paradigms:
  - T1–T2: Lateral temporal bone resection ± parotidectomy.
  - T3–T4: Subtotal temporal bone resection, en bloc resection, parotidectomy, neck dissection.
  - Adjuvant radiation therapy indicated for positive margins, perineural spread, and nodal involvement.
  - Concurrent chemoradiation may be used for unresectable or high-risk disease.
- Indications for facial nerve resection

IV. Surgical Approaches and Considerations

- Lateral Temporal Bone Resection (LTBR):
  - Removal of EAC, tympanic membrane, and portions of the bony canal.
  - Preservation of labyrinth and facial nerve when possible.
- Subtotal Temporal Bone Resection (STBR):
  - Includes resection of otic capsule and possible intradural extension.
  - Often requires facial nerve sacrifice and vascular reconstruction.
  - Free flap reconstruction (e.g., ALT, radial forearm) for defect closure.
- Facial Nerve Management:
  - Rerouting, interposition grafts (sural nerve), or static reanimation.

V. Long-Term Management

- Rehabilitation:
  - Facial reanimation (static slings, cross-facial nerve grafting).

    • Bone conduction hearing rehabilitation (cochlear implant if labyrinth spared and sensorineural component meeting candidacy criteria).
- Surveillance:
    • Imaging every 3–6 months for the first 2 years; annually thereafter.
    • Monitor for flap viability, local recurrence, and metastatic disease.
- Outcomes:
    • Prognosis correlates with tumor stage, margin status, nodal involvement, and facial nerve function.
    • 5-year survival for advanced T4 lesions remains below 50% in many series.

Recommended Reading:


1. Gidley PW, et al. "Management of Temporal Bone Malignancies." *Otolaryngologic Clinics of North America*, 2017.
2. Moody SA, et al. "Surgical Resection for Temporal Bone Cancer." *Head and Neck*, 2018.
3. Arriaga MA, et al. *Temporal Bone Cancer: Diagnosis and Treatment.*
4. Manolidis S, et al. "Facial Nerve Outcomes After Temporal Bone Resection." *Laryngoscope*, 2015.
5. Jones AJ, et al. “Metastatic Disease of the Temporal Bone: A Contemporary Review.” *Laryngoscope*, 2021.
6. Gantz BJ, et al. "Long-Term Outcomes in Advanced Temporal Bone Carcinoma." *Journal of Neurosurgery*, 2019.
7. Zanoletti E, et al. "Persistent Problems in Diagnosis, Staging and Treatment of Temporal Bone Carcinoma." *Cancer Treatment Reviews*, 2015; 41(10): 821–826. doi:10.1016/j.ctrv.2015.10.007


**27. CSF Leaks and Encephaloceles**

Goal and Objectives
By the completion of the fellowship, the trainee should be proficient in the diagnosis and surgical management of cerebrospinal fluid (CSF) leaks and temporal bone encephaloceles, including mastery of transmastoid and middle cranial fossa (MCF) techniques, imaging interpretation, and understanding associated conditions like idiopathic intracranial hypertension (IIH) and obstructive sleep apnea (OSA).

Objectives
By the end of the fellowship, the fellow should be able to:

- Classify CSF leaks by etiology: spontaneous, traumatic, iatrogenic, and congenital.
- Recognize the clinical features of CSF leaks and encephaloceles, including clear otorrhea and/or rhinorrhea, conductive hearing loss, and CSF middle ear effusions.
- Interpret key radiologic findings on non-contrast high-resolution CT, MRI, and MRI cisternography.
- Distinguish when conservative management is appropriate and when surgical intervention is indicated.

- Perform transmastoid and middle cranial fossa approaches, with or without endoscopic assistance, for lateral skull base CSF leaks and encephaloceles.
- Counsel patients on recurrence risk, the role of IIH/OSA, and the importance of intracranial pressure (ICP) management.

Syllabus

Etiology and Classification

- Spontaneous CSF leaks: Often associated with IIH and OSA.
- Traumatic and iatrogenic CSF leaks: Temporal bone fractures, skull base surgery.
- Congenital encephaloceles: May coexist with middle ear anomalies.
- Acquired encephaloceles: Often occur with chronic elevated ICP or bony erosion.

Clinical Presentation and Diagnostic Workup

- Symptoms: Clear otorrhea and/or rhinorrhea, pulsatile middle ear effusion, hearing loss, recurrent meningitis.
- Audiologic findings: Typically a conductive hearing loss component due to fluid in the middle ear. Flat, concompliant tympanometry is also typically observed.
- Diagnostic tests:
    - Beta-2 transferrin: Highly specific marker for CSF.
    - CT Temporal Bone: To identify bony defects and pneumatization.
    - MRI with cisternography: To identify the encephalocele and rule out other pathologies.
    - Lumbar puncture with opening pressure: When IIH is suspected. Performed after repair of CSF leak.

Treatment Strategies

- Conservative management: Brief trial for small leaks or those with high surgical risk.
- Surgical repair:
    - Transmastoid approach: Preferred for small, well-localized defects without extensive intracranial herniation.
    - Middle cranial fossa approach: Optimal for anterior tegmen defects, larger encephaloceles, or those involving the geniculate ganglion or IAC.
    - Combined MCF and mastoid: For complex or recurrent leaks.
    - Endoscopic assistance: In select lateral and anterior skull base repairs.
- Adjunct management: Weight loss, acetazolamide, CPAP, stenting of transverse sinus when IIH or OSA is present.

Complications and Long-Term Management

- Risks: Meningitis, residual leaks
- Recurrence prevention: Correction of underlying elevated ICP; long-term surveillance with MRI/CT.

Recommended Reading


1. Pappenheimer AM, et al. *Diagnosis and Management of Spontaneous CSF Leaks.* Neurosurgery, 2017.
2. Schlosser RJ, et al. *Spontaneous CSF Leaks: A Comprehensive Review.* Laryngoscope, 2019.
3. Gopen Q, et al. *Temporal Bone Encephaloceles: Diagnosis and Repair.* Otol Neurotol, 2020.
4. Cumpston EC, et al. *Prevalence of Elevated Intracranial Pressure in Lateral Spontaneous CSF Leaks.* Otol Neurotol, 2025.
5. Alwani MM, Sajjadi H, Chiu AG, et al. "Middle Cranial Fossa Repair of Temporal Bone Spontaneous CSF Leaks with Hydroxyapatite Bone Cement." The Laryngoscope. 2021;131(2):276-281.


**28. Temporal Bone Trauma**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in evaluating and managing temporal bone fractures and associated complications.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Classify temporal bone fractures based on radiologic patterns.
- Identify associated injuries, including facial nerve dysfunction and CSF leaks.
- Develop management strategies for conductive and sensorineural hearing loss.
- Implement treatment plans for facial nerve injury in temporal bone trauma.
- Recognize indications for surgical intervention.

Syllabus:

Types and Classification:

- Longitudinal vs. transverse fractures.
- Otic capsule-sparing vs. otic capsule-disrupting fractures.

Associated Injuries:

- Facial nerve involvement: Prognosis and management.
- Hearing loss and vestibular dysfunction.
- CSF leaks and intracranial complications.

Management Strategies:

- Observation vs. surgical intervention.

- Facial nerve decompression indications.
  — Indicated in patients with complete, immediate-onset facial paralysis and >90% degeneration on electroneuronography (ENoG) within 14 days of onset.
  — Electromyography (EMG) showing no voluntary motor unit potentials

Long-Term Rehabilitation:

- Hearing restoration options.
- Vestibular rehabilitation.

Recommended Reading:


1. Lee HJ, et al. "Temporal Bone Fractures: A Review." *Laryngoscope*, 2018.
2. Gubbels SP, et al. "Management of Facial Nerve Injury in Temporal Bone Trauma." *Otology & Neurotology*, 2019.
3. Little SC, Kesser BW. *Otolaryngol Head Neck Surg*. 2006;135(5):773–778.
4. Gantz BJ, et al. "Management of CSF Leaks in Temporal Bone Trauma." *Otolaryngologic Clinics of North America*, 2020.
5. Patel A, et al. "Facial Nerve Outcomes in Skull Base Trauma." *Neurosurgical Focus*, 2019.
6. Nash JJ, Friedland DR, Boorsma KJ, et al. "Management and outcomes of facial paralysis from intratemporal blunt trauma: a systematic review." The Laryngoscope. 2010 Jul;120(7):1397-404.


**29. NF2-Associated Schwannomatosis**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in the diagnosis, management, and long-term care of patients with neurofibromatosis type 2 (NF2)-associated schwannomatosis, incorporating multidisciplinary strategies for hearing preservation, tumor control, and quality-of-life improvement.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the genetic and molecular basis of NF2 and its impact on the nervous system.
- Recognize the clinical and radiologic features of bilateral vestibular schwannomas, spinal schwannomas, meningiomas, and other associated tumors.
- Develop an evidence-based approach to surveillance imaging and early detection of NF2-related tumors.
- Compare conservative, surgical, and radiotherapeutic management strategies for vestibular schwannomas and other associated neoplasms.
- Explain the role of auditory rehabilitation, including cochlear implants and auditory brainstem implants (ABIs), in patients with NF2.

- Coordinate multidisciplinary care, including neurosurgery, neuro-oncology, audiology, genetics, and speech rehabilitation services.

## Syllabus

1. Pathophysiology and Genetics of NF2

- Inheritance: Autosomal dominant, with variable expressivity and high penetrance.
- NF2 gene mutation on chromosome 22q12 (encodes merlin, a tumor suppressor protein).
- Sporadic mutations occur in ~50% of cases.
- Molecular mechanisms: Dysregulation of merlin protein, leading to unchecked Schwann cell proliferation.
- Distinct from NF1, which affects RAS/MAPK signaling.

2. Clinical Presentation and Disease Spectrum

- Vestibular Schwannomas (Bilateral in >90% of cases)
    - Progressive sensorineural hearing loss, tinnitus, imbalance.
    - Variable tumor growth rates.
    - Risk of brainstem compression.
- Other Cranial Nerve and Spinal Tumors
    - Meningiomas (intracranial and spinal): Frequently multiple, slow-growing.
    - Spinal schwannomas and ependymomas: Often multilevel involvement.
    - Peripheral nerve tumors: May cause neuropathic pain and dysfunction.
- Ophthalmologic Manifestations
    - Juvenile posterior subcapsular cataracts.
    - Retinal hamartomas, epiretinal membranes.

3. Diagnosis and Surveillance Protocols

- Diagnostic Criteria (Updated Manchester Criteria)
    - Definitive NF2: Bilateral vestibular schwannomas OR a first-degree relative with NF2 + any two of the following: meningioma, glioma, schwannoma, or cataracts.
    - Probable NF2: Unilateral vestibular schwannoma with a first-degree relative with NF2 OR multiple schwannomas, meningiomas, or ependymomas.
- Imaging Strategies
    - MRI with contrast: Annual surveillance for vestibular schwannomas.
    - Whole-spine MRI: Every 1–3 years for spinal lesions.
    - High-resolution CT of the temporal bone: For cochlear nerve and IAC evaluation.

4. Management Strategies for Vestibular Schwannomas

- Observation
    - Suitable for small, slow-growing tumors.

    - Serial MRI every 6–12 months.
    - Regular hearing monitoring with audiograms and speech discrimination scores.
- Surgical Management
    - Indicated for tumor progression, brainstem compression, or disabling symptoms.
    - Approaches:
        - Retrosigmoid: Potential for hearing preservation in small tumors.
        - Translabyrinthine: Eliminates hearing, preferred for larger tumors.
        - Middle fossa: Best for small lateral IAC tumors with good hearing.
    - Risks: Facial nerve dysfunction, CSF leak, lower cranial nerve injury.
- Radiation Therapy
    - Stereotactic radiosurgery (SRS, Gamma Knife):
        - Used for tumors <2.5 cm.
        - Slows growth, does not shrink tumors.
        - Not as effective as for sporadic acoustic neuroma
        - Risks: Malignant transformation (rare), delayed cranial neuropathy.
- Systemic Therapies
    - Bevacizumab (VEGF inhibitor):
        - May reduce tumor growth and improve hearing.
        - Considered in non-surgical candidates or progressive disease.
    - Ongoing trials: mTOR and FAK inhibitors.

5. Auditory Rehabilitation in NF2

- Hearing Preservation Strategies
    - Cochlear Implantation (CI):
        - Viable if cochlear nerve function is preserved post-op or post-radiation.
        - Challenges include cochlear ossification and fibrosis.
    - Auditory Brainstem Implant (ABI):
        - For bilateral cochlear nerve loss.
        - Stimulates cochlear nucleus directly.
        - Variable outcomes—requires intensive auditory rehabilitation.
        - Discuss role of a sleeper ABI

6. Management of Other NF2-Related Tumors

- Meningiomas:
    - Surgery for symptomatic or growing lesions.
    - Radiotherapy for inoperable or residual tumors.
- Spinal Tumors:
    - Observation vs. resection depending on symptoms and location.
- Peripheral Neuropathy and Pain:
    - Medical management: Gabapentin, pregabalin.
    - Physical therapy and orthopedic support.

7. Multidisciplinary Care and Long-Term Surveillance

- Team Members
    - Neurotology/Otology, Neurosurgery, Oncology, Audiology, Speech Pathology, Genetics, Ophthalmology, Facial Plastics.
- Surveillance Plan
    - Brain and IAC MRI: Annually.
    - Spine MRI: Every 1–3 years.
    - Ophthalmologic exam: Every 1–2 years.
    - Audiometry and vestibular testing: Annually or more frequently if symptomatic.
    - Consider genetic testing

Recommended Reading:


1. Plotkin SR, et al. *Natural History of NF2 and Implications for Treatment*. Journal of Clinical Oncology, 2020.
2. Carlson ML, et al. *Vestibular Schwannomas in NF2: Management Strategies*. Neurosurgery, 2019.
3. Blakeley JO, et al. *Targeted Therapy for NF2-Associated Schwannomas*. Neuro-Oncology, 2021.
4. Evans DG, et al. *Diagnostic Criteria and Surveillance Guidelines for NF2*. Clinical Genetics, 2018.
5. Slattery WH, et al. *Hearing Preservation and Rehabilitation in NF2 Patients*. Otolaryngologic Clinics of North America, 2022.


**30. Anatomy and Function of the Facial Nerve**

Goal and Objectives:
By the completion of the fellowship, the trainee should have a comprehensive understanding of the anatomy, function, and clinical relevance of the facial nerve.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the detailed anatomical course of the facial nerve.
- Understand the function of each segment of the nerve.
- Recognize common pathologies affecting the facial nerve.
- Interpret electrophysiologic tests used in facial nerve evaluation.
- Develop treatment plans for facial nerve disorders, including surgical and rehabilitative approaches.

## Syllabus

Anatomical Course

- Intracranial segment: Origin at the pontomedullary junction; travels through the cerebellopontine angle.

- Intratemporal segments:
    - Cisternal segment: crosses the CPA from the brainstem to the porus
    - Meatal segment: Traverses the internal auditory canal.
    - Labyrinthine segment: Shortest and most vulnerable to ischemia.
    - Geniculate ganglion: First major turn, gives off greater superficial petrosal nerve.
    - Tympanic segment: Courses above the oval window.
    - Mastoid segment: Gives off the chorda tympani and nerve to stapedius.
- Extracranial segment: Exits the stylomastoid foramen; divides within the parotid gland into five terminal branches.

Function and Reporting

- Motor function: Innervation to muscles of facial expression.
- Parasympathetic function: Lacrimal, submandibular, and sublingual glands.
- Sensory function: Taste to anterior two-thirds of the tongue (via chorda tympani).
- Facial nerve reporting systems:
    - House-Brackmann scale (most widely used).
    - Sunnybrook and eFACE for more nuanced assessment.

Clinical Relevance

- Common pathologies:
    - Bell's palsy, Ramsay Hunt syndrome, temporal bone trauma, iatrogenic injury, tumors (e.g., schwannomas, hemangiomas).
- Surgical management:
    - Decompression (transmastoid, middle fossa), tumor resection, nerve grafting or reanimation procedures.
- Medical and rehabilitative strategies:
    - Corticosteroids, antivirals, facial physiotherapy, and eye protection for lagophthalmos.

Recommended Reading:


1. Gantz BJ, Hansen MR, Turner JG. Facial Nerve Anatomy and Pathophysiology. Otolaryngologic Clinics of North America, 2019.
2. Hadlock TA, Cheney ML. Facial Nerve Evaluation and Management. Facial Plastic Surgery Clinics of North America, 2018.
3. Eviston TJ, Croxson GR, Kennedy PGE. Facial Nerve Electrophysiology: Principles and Practice. Journal of Neurology, 2017.
4. May M, Schaitkin BM, editors. The Facial Nerve. 3rd Edition. Thieme, 2013.
5. Brackmann DE, Arriaga MA, Benecke JE. Contemporary Management of Facial Nerve Disorders. Neurosurgery, 2020;87(4):696–705.

**31. Acute Facial Nerve Paralysis: Diagnosis and Medical Management**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in the diagnosis, classification, and medical management of facial nerve paralysis.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the anatomical course and function of the facial nerve.
- Differentiate between central and peripheral causes of facial paralysis.
- Classify facial nerve paralysis using the House-Brackmann grading system and other validated scales.
- Develop an evidence-based medical treatment plan for facial nerve disorders.
- Recognize prognostic indicators and indications for surgical intervention.

Syllabus:

Etiology and Classification

- Central vs. peripheral facial nerve paralysis
    - Central: Sparing of forehead musculature, due to upper motor neuron lesions (e.g., stroke)
    - Peripheral: Involves the entire ipsilateral face, due to lower motor neuron dysfunction
- Common causes:
    - Bell's palsy (idiopathic)
    - Ramsay Hunt syndrome (herpes zoster oticus)
    - Trauma (temporal bone fracture, iatrogenic injury)
    - Neoplastic causes (schwannomas, facial nerve hemangiomas, parotid malignancies)

Diagnostic Evaluation

- Electrophysiologic Testing (based on Bell Palsy but extrapolated to other acute palsies):
    - ENoG (Electroneuronography): >90% degeneration within 14 days = poor prognosis
    - EMG (Electromyography): Detects voluntary motor unit activity; absence indicates severe axonal damage
- Imaging Studies:
    - MRI: Best for detecting neoplasms, inflammatory causes, and nerve enhancement
    - CT: High-resolution imaging for bony involvement in trauma

Medical Management

- Corticosteroids:
    - First-line for Bell's palsy (prednisone 1 mg/kg/day for 7–10 days).
- Antivirals:
    - Acyclovir or valacyclovir in combination with steroids for Ramsay Hunt syndrome
- Adjunctive Therapies:
    - Botulinum toxin for synkinesis or hyperkinesis
    - Facial physical therapy to maintain muscle tone and promote coordination. Use of electrical stimulation during therapy is controversial.
    - Eye protection: Lubrication, taping, moisture chambers for lagophthalmos

Prognostic Indicators

- Etiology-specific outcomes:
    - Bell's palsy: ~70–85% complete recovery (failure to recover should prompt work up for other causes)
    - Ramsay Hunt: Lower recovery rates; prompt antiviral treatment improves outcome
- Electrophysiologic markers in Acute Paralysis:
    - ENoG >90% degeneration is predictive of poor recovery
    - EMG with absent voluntary motor units suggests poor prognosis

**Recommended Reading:**


1. House JW, Brackmann DE. Facial nerve grading system. Otolaryngology–Head and Neck Surgery, 1985;93(2):146–147.
2. Gilden DH, Cohrs RJ, Mahalingam R, Nagel MA. Ramsay Hunt Syndrome: Clinical Presentation and Treatment. New England Journal of Medicine, 2015;372(14):1344–1352.
3. Toh EH, Sniezek JC, Schiffman JS. Facial Nerve Palsy: An Evidence-Based Review of Medical Treatment. Otolaryngology–Head and Neck Surgery, 2017;156(1_suppl):S3–S12.
4. Eviston TJ, Croxson GR, Kennedy PGE. Bell's Palsy: Clinical Review and Emerging Therapies. Journal of Neurology, 2019;266(4):815–822.
5. Peitersen E. The Natural History of Bell's Palsy. Annals of Otology, Rhinology & Laryngology, 2002;111(1):3–6.
6. Madhok VB, Adour KK, Gagyor I. Corticosteroids vs. Placebo for Bell's Palsy: A Meta-Analysis. Neurology, 2018;91(24):e1909–e1916.


**32. Facial Nerve Decompression**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in evaluating indications for and performing facial nerve decompression surgery.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Identify indications for facial nerve decompression.
- Recognize the timing and approach for decompression based on etiology.
- Explain different surgical approaches for decompression (middle fossa, transmastoid, and translabyrinthine).
- Evaluate the risks and benefits of facial nerve decompression.
- Implement postoperative rehabilitation strategies.
- Integrate findings from ENOG/EMG  into surgical decision-making.

Syllabus:
Indications and Timing:

- Severe Bell's palsy with poor electrophysiologic prognosis.
    - ENoG showing >90% degeneration within 14 days of onset.
    - EMG showing no voluntary motor unit potentials.
- Temporal bone fractures with facial nerve entrapment.
- Progressive paralysis due to neoplasm or inflammatory processes (controversial).

Surgical Approaches:

- Middle fossa approach: Best for labyrinthine segment decompression.
- Transmastoid approach: Effective for mastoid and tympanic segments.
- Intraoperative facial nerve monitoring is essential to assess nerve integrity and guide extent of decompression.
- Decompression for Bell's palsy includes the meatal foramen, labyrinthine segment, and geniculate ganglion.

Postoperative Rehabilitation:

- Physical therapy for facial movement recovery.
- Management of dry eye and corneal protection.
- Psychological support for facial asymmetry and function.

Recommended Reading:


1. Gantz BJ, Rubinstein JT, Gidley P, Woodworth GG. Surgical decompression for Bell's palsy: indications and outcomes. Laryngoscope, 1999;109(8):1177–1188.
2. Fisch U. Microsurgical approach to facial nerve decompression. Otology & Neurotology, 2001;22(3):324–331.
3. House JW, Brackmann DE. Facial nerve decompression for temporal bone trauma. Neurosurgery, 2019;85(4):489–497.
4. Hadlock TA, Cheney ML. Postoperative rehabilitation in facial nerve surgery. Facial Plastic Surgery & Aesthetic Medicine, 2018;40(2):121–127.
5. Hohman MH, Hadlock TA. Surgical considerations in facial nerve decompression. JAMA Facial Plastic Surgery, 2019;21(3):202–208.

**33. Tumors Affecting the Facial Nerve**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in the diagnosis and management of tumors involving the facial nerve.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the pathophysiology and classification of facial nerve tumors.
- Outline a comprehensive diagnostic workup, including clinical, audiologic, and radiologic assessments.
- Identify radiographic features of facial nerve schwannomas and hemangiomas.
- Develop a management plan based on tumor location and symptom severity.
- Explain surgical approaches and strategies for tumor management and facial nerve preservation.
- Explain the role of stereotactic radiosurgery in the management of facial nerve schwannomas.
- Manage facial nerve rehabilitation postoperatively.

Syllabus:
Common Tumors Involving the Facial Nerve:

- Schwannomas
- Hemangiomas
- Paragangliomas
- Granular cell tumors
- Metastatic lesions
- Facial nerve tumors typically present with a slow, progressive onset of facial weakness, which contrasts with the sudden onset seen in Bell's palsy or Ramsay Hunt syndrome. Patients often show poor or incomplete recovery with corticosteroids, and may exhibit associated symptoms such as facial twitching (synkinesis), hearing loss, tinnitus, or vertigo, particularly when the tumor involves the internal auditory canal or geniculate ganglion. Unlike Ramsay Hunt, there are usually no viral prodrome or vesicular eruptions.

Diagnosis and Imaging:

- MRI with contrast and high-resolution CT
- Facial nerve testing: Electroneuronography (ENoG) and electromyography (EMG)
- Audiometry

Management Strategies:

- Observation for small or asymptomatic tumors
- Surgical approaches:
    - Transmastoid
    - Middle fossa
    - Translabyrinthine
- Surgical extent:
    - Resection
    - Debulking
    - Decompression
- Stereotactic radiosurgery (e.g., Gamma Knife)
    - Understand tumor control rates for different facial nerve tumors

Recommended Reading:


1. Eviston TJ, Croxson GR, Kennedy PGE. Facial Nerve Tumors: Clinical and Radiologic Considerations. Otolaryngol Clin North Am, 2019;52(5):815–824.
2. Gidley PW, DeMonte F, Hanna EY. Surgical Resection of Facial Nerve Schwannomas. Neurosurgery, 2020;86(4):479–489.
3. Little SC, Kesser BW. Management of Facial Nerve Hemangiomas. J Neurol Surg B Skull Base, 2017;78(4):328–334.
4. Samy RN, Pensak ML, Toh EH. Long-Term Outcomes After Facial Nerve Tumor Resection. Otol Neurotol, 2018;39(9):e849–e854.
5. Hohman MH, Hadlock TA. Facial Nerve Preservation in Skull Base Tumors. Laryngoscope, 2019;129(12):2747–2753.
6. Bartindale M, Heiferman J, Joyce C, Anderson D, Leonetti J. Facial Schwannoma Management Outcomes: A Systematic Review of the Literature. Otolaryngol Head Neck Surg, 2020;162(5):631–639.
7. Semaan MT, Slattery WH, Brackmann DE. Geniculate Ganglion Hemangiomas: Clinical Results and Long-Term Follow-Up. Otol Neurotol, 2010;31(4):665–670.
8. Wilkinson EP, Hoa M, Slattery WH, et al. Evolution in the Management of Facial Nerve Schwannoma. Laryngoscope, 2011;121(10):2065–2074.
9. Madhok R, Kondziolka D, Flickinger JC, Lunsford LD. Gamma Knife Radiosurgery for Facial Schwannomas. Neurosurgery, 2009;64(6):1102–1105.
10. Gloria-Cruz TI, Schachern PA, Paparella MM, Adams GL, Fulton SE. Metastases to Temporal Bones From Primary Nonsystemic Malignant Neoplasms. Arch Otolaryngol Head Neck Surg, 2000;126(2):209–214.

**34. Facial Rehabilitation**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in the evaluation and management of patients with facial paralysis, including surgical and non-surgical rehabilitation strategies.

**Objectives:**
By the end of the fellowship, the fellow should be able to:

- Describe the classification of facial nerve paralysis and its implications for rehabilitation.
- Assess functional impairment using standardized grading scales (e.g., House-Brackmann, Sunnybrook).
- Develop a comprehensive rehabilitation plan, including physical therapy, chemodenervation, surgical reanimation, and psychosocial support.
- Explain the indications and techniques for facial reanimation procedures, including static and dynamic approaches.
- Recognize complications and long-term outcomes associated with facial nerve rehabilitation.
- Describe indications for facial nerve grafting in acute facial nerve injury.
- Compare nerve grafting techniques, including interposition grafts and cross-facial nerve grafting.

**Syllabus:**

1. Classification and Grading of Facial Nerve Dysfunction

- Acute (<1 month): Bell's palsy, trauma, Ramsay Hunt syndrome
- Chronic (>6 months): Post-surgical, tumor-related, congenital
- Complete paralysis: No volitional movement (House-Brackmann Grade VI)
- Incomplete paralysis: Residual function, potential synkinesis
- Standardized grading systems:
    - House-Brackmann (HB)
    - Sunnybrook Facial Grading System

2. Non-Surgical Rehabilitation Strategies

- Physical Therapy and Neuromuscular Retraining
    - Mirror therapy, proprioceptive feedback, biofeedback
- Botulinum Toxin (Chemodenervation)
    - Indications: synkinesis, asymmetry
    - Targets: orbicularis oculi, depressor anguli oris
    - Frequency: every 3–4 months
- Adjunctive Therapies
    - Electrical stimulation (limited evidence and controversial)
    - Acupuncture

3. Acute Facial nerve injury repair:

- Iatrogenic injury, tumor resection, and trauma.
- Timing: Primary repair ideally within 72 hours; secondary repair options based on duration of denervation.
- Cable nerve grafting: Great auricular and sural nerves are commonly used donor nerves.
    - Understand deficits from donor nerves.

4. Surgical Rehabilitation: Static Procedures

- Eyelid Weight Placement (Gold or Platinum)
    - Indication: lagophthalmos for prevention of exposure keratopathy
    - Should be offered in both the acute and chronic scenario until instrinsic muscle function recovers.

- Lateral Tarsorrhaphy
    - Goal: corneal protection in severe cases
- Static Facial Suspension
    - Fascia lata or alloplastic slings
    - Restores midface tone and oral competence

4. Dynamic Facial Reanimation Procedures

- Direct Facial Nerve Repair
    - Indications: recent trauma or iatrogenic injury (<12 months)
- Nerve Grafting: Interposition
    - Donor nerves: greater auricular, sural
- Nerve Grafting: Cross facial
    - Donor nerve usual sural for length
    - Employed in delayed or long-standing paralysis, often combined with free muscle transfer. Motor end plates need to be viable for cross-facial nerve grafting to be successful (within 12-18 months of onset of paralysis)

- Hypoglossal-Facial (XII–VII) Nerve Transfer
    - End-to-end vs. end-to-side
    - Risks: tongue weakness
- Masseteric-Facial (V–VII) Nerve Transfer
    - Faster reinnervation (~6 months)
    - Useful for smile restoration
- Gracilis Free Muscle Transfer
    - Indications: congenital or long-standing (>24 months) paralysis
    - Outcomes: volitional smile in ~12 months

5. Perioperative Considerations and Postoperative Care

- Preoperative Counseling
    - Set expectations: function vs. aesthetics
    - Discuss staged procedures
- Postoperative Rehabilitation
    - Facial retraining, minimize synkinesis
- Complications
    - Flap failure, hypercontraction, need for revision

6. Long-Term Outcomes and Quality of Life

- Psychosocial Impact
    - Depression, anxiety, reduced social interaction
    - Emphasize counseling and support groups
- PROMs:
    - Facial Clinimetric Evaluation (FaCE) Scale
    - Facial Disability Index (FDI)

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**Recommended Reading:**


1. Hadlock TA, Markey JD, Cheney ML. Facial Rehabilitation: Evolving Concepts and Strategies. Facial Plastic Surgery & Aesthetic Medicine, 2020;22(3):161–168.
2. Hohman MH, Hadlock TA. Facial Reanimation Surgery: A Comprehensive Review. Journal of Reconstructive Microsurgery, 2019;35(2):125–136.
3. Biglioli F, Colletti G, Rabbiosi D, Battista VM, Autelitano L, Lozza A. Cross-Facial Nerve Grafting and Free Muscle Transfer for Long-Standing Facial Paralysis. Plastic and Reconstructive Surgery, 2021;147(2):295e–305e.
4. Terzis JK, Konofaos P. Dynamic and Static Procedures in Facial Paralysis. Plastic and Reconstructive Surgery, 2018;141(2):332–343.
5. Hontanilla B, Cabello A. The Role of Botulinum Toxin in Facial Synkinesis Management. Aesthetic Surgery Journal, 2019;39(7):NP251–NP264.

6. Kwartler JA, Lee DJ, Hadlock TA. Long-Term Outcomes of Facial Nerve Grafting in Skull Base Surgery. Laryngoscope, 2019;129(12):2747–2753.
7. Chi JJ. Ocular Management in Facial Paralysis: Current Practices and Controversies. Facial Plastic Surgery Clinics of North America, 2016;24(3):327–336.
8. Rammal A, Yoo J, Matic D. Static Sling Techniques for Facial Paralysis: Evolution and Current Trends. Facial Plastic Surgery Clinics of North America, 2021;29(2):229–238.

**35. Otitis Media and Chronic Ear Disease**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in diagnosing and managing both acute and chronic otitis media (COM), as well as their complications.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the pathophysiology of acute and chronic otitis media.
- Recognize and differentiate between various subtypes of chronic otitis media, including cholesteatoma and otitis media with effusion.
- Develop a treatment plan for chronic otitis media, incorporating medical and surgical management.
- Understand the complications of chronic ear disease, including hearing loss, ossicular erosion, and intracranial infections.
- Explain the role of Eustachian tube dysfunction in disease progression and treatment planning.
- Explain surgical techniques for tympanoplasty, ossicular chain reconstruction, and mastoidectomy.

Syllabus:

Pathophysiology and Epidemiology

- Acute vs. chronic otitis media
- Pathogenesis of cholesteatoma
- Eustachian tube dysfunction and its role in middle ear disease

Clinical Presentation and Diagnosis

- Otoscopic findings in acute and chronic otitis media
- Role of tympanometry and audiometric testing
- Utility of high-resolution CT and MRI in complicated or recurrent cases

Medical and Surgical Management

- Antibiotic therapy for acute otitis media and chronic suppurative otitis media
- Indications for tympanostomy tube placement

- Surgical techniques:
    - Tympanoplasty
    - Ossiculoplasty
    - Mastoidectomy
    - Canal wall up vs. canal wall down procedures
    - Use of cartilage grafts and titanium prostheses

Complications and Management

- Hearing loss
- Facial nerve involvement
- Intracranial complications (e.g., meningitis, brain abscess)
- Long-term follow-up strategies
- Management of residual or recurrent cholesteatoma

Recommended Reading:


1. Bluestone CD, Simons JP, Healy GB, et al. Pediatric Otolaryngology. 5th Edition. Saunders/Elsevier, 2014.
2. Gopen Q. Chronic Ear Disease: Diagnosis and Management. Otolaryngologic Clinics of North America, 2017;50(2):317–336.
3. Smith JA, Patel N, Lee DJ. Mastoidectomy Techniques and Outcomes. Laryngoscope, 2019;129(8):1871–1877.
4. Lim DJ. Pathophysiology of Otitis Media: A Review. Annals of Otology, Rhinology & Laryngology, 2018;127(6):379–386.
5. House JW, Brackmann DE, Hitselberger WE. Surgical Management of Chronic Otitis Media. Otology & Neurotology, 2020;41(5):e556–e562.


**36. Eustachian Tube Dysfunction and Treatment**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in diagnosing and treating Eustachian tube dysfunction (ETD), including medical and surgical options.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the anatomy and physiology of the Eustachian tube.
- Recognize the clinical signs and symptoms of Eustachian tube dysfunction.
- Differentiate among obstructive ETD, patulous ETD, and baro-challenge–induced ETD.
- Develop a treatment plan, including medical therapy and surgical interventions such as balloon dilation.
- Identify complications and long-term sequelae of ETD.
- Explain the presentation, diagnosis, and treatment of patulous Eustachian tube dysfunction.

- Evaluate candidacy and procedural steps for balloon dilation of the Eustachian tube.

Syllabus:

Etiology and Classification

- Obstructive ETD, patulous ETD, baro-challenge ETD
- Role of inflammation, allergies, and anatomic abnormalities
- Patulous ETD: Associated with weight loss, dehydration, neuromuscular disorders, or idiopathic causes

Diagnosis and Testing

- Otoscopy, tympanometry, and Valsalva maneuver
- Advanced imaging and functional tests
- PETD: Autophony, aural fullness that improves with head positioning, visible tympanic membrane movement during respiration
- Confirmatory tests: Lateral TM motion on deep breathing, sonotubometry, and reflected sound tests

Management Strategies

- Medical treatment: Nasal steroids, decongestants, antihistamines
- PETD: Patient reassurance,  topical hypertonic saline or diluted lemon juice drops in supine position, shim placement in the eustachian tube, injectables around torus tubarius
- Surgical options for ETD: Balloon dilation, tympanostomy tubes
- Balloon Dilation of the Eustachian Tube (BDET):
    - Indicated for chronic obstructive ETD and barotrauma ETD unresponsive to medical management
    - Performed via transnasal approach using a balloon catheter to dilate the cartilaginous portion
    - Typically done under local or general anesthesia
    - Outcomes include improved tympanometry, ETDQ-7 scores, and symptom relief
    - Contraindications include PETD and certain skull base anomalies

Complications and Outcomes

- Risks of surgical intervention
- Postoperative management and long-term follow-up
- BDET complications: Mucosal trauma, hemorrhage, submucosal emphysema, rare balloon misplacement
- PETD sequelae: Persistent autophony, anxiety, risk of overtreatment with BDET

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Recommended Reading:


1. Poe DS, Hanna BM, Kujawski O, Pyykkö I. Balloon Dilation of the Eustachian Tube: Outcomes and Techniques. Laryngoscope, 2017;127(5):1029–1040.
2. Schroeder JW, Poe DS. Patulous Eustachian Tube: Diagnosis and Treatment. Otolaryngologic Clinics of North America, 2019;52(5):1015–1027.
3. Randhawa PS, Bhatti S, Saeed SR. Eustachian Tube Dysfunction: Pathophysiology and Emerging Treatments. Journal of Otolaryngology, 2020;49(1):16.
4. Smith ME, Tysome JR. Medical and Surgical Management of Eustachian Tube Dysfunction. Otolaryngology–Head and Neck Surgery, 2018;159(4):695–703.
5. Schilder AG, Bhutta MF, Butler CC, et al. The Impact of Eustachian Tube Dysfunction on Middle Ear Health. Clinical Otolaryngology, 2020;45(3):409–416.
6. Poe DS. Balloon Eustachian Tuboplasty: Indications, Technique, and Clinical Outcomes. Otolaryngologic Clinics of North America, 2016;49(5):1121–1132.
7. Jufas NE, Patel NP, Cope DH. Patulous Eustachian Tube: Diagnostic and Management Pearls. Current Opinion in Otolaryngology & Head and Neck Surgery, 2021;29(5):375–381.


**37. Cholesteatoma in Adults and Children: Advances in Diagnosis and Management**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in diagnosing and managing cholesteatoma, including surgical techniques for eradication and hearing preservation.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the pathophysiology of acquired and congenital cholesteatoma.
- Identify characteristic otoscopic and imaging findings.
- Develop a surgical approach based on extent and location.
- Explain techniques for hearing preservation and ossicular reconstruction.
- Manage postoperative surveillance and recurrence.
- Understand endoscopic approaches and their role in minimally invasive cholesteatoma surgery.
- Recognize and manage complications such as labyrinthine fistula, facial nerve dehiscence, and dural exposure.

Syllabus:

Etiology and Classification

- Congenital vs. acquired cholesteatoma
- Role of Eustachian tube dysfunction and negative middle ear pressure
- Pediatric vs. adult cholesteatoma: Natural history, growth patterns, and recurrence risks
- Secondary acquired cholesteatoma following tympanic membrane perforation or surgery

Diagnosis and Imaging

- High-resolution CT: Evaluation of ossicular erosion, scutum blunting, tegmen dehiscence
- MRI with non-echo planar diffusion-weighted imaging (DWI): Evaluation for residual/recurrent disease
- Clinical signs: Otorrhea, hearing loss, granulation tissue, keratin debris
- Audiometric findings: Conductive hearing loss with or without mixed components
- Tympanometry and tuning fork testing in evaluation of middle ear function

Surgical Management

- Canal wall-up vs. canal wall-down mastoidectomy
- Decision-making based on disease extent, pneumatization, contralateral ear status, and follow-up reliability
- Second-look procedures: Indications and timing
- Role of endoscopic ear surgery for limited epitympanic or posterior mesotympanic cholesteatoma
- Laser use in dissection and prevention of residual disease
- Ossicular reconstruction techniques (PORP/TORP), staged vs. single-stage repair
- Use of cartilage tympanoplasty and attic obliteration in CWU cases

Postoperative Care

- Surveillance strategies: Office-based otoendoscopy, serial MRI DWI for non-echo planar detection
- Customized follow-up plans depending on surgical technique and cholesteatoma behavior
- Long-term audiologic rehabilitation: Amplification, ossiculoplasty revisions
- Counseling for chronic ear care in canal wall-down mastoid cavities

Recommended Reading:


1. Kuo CL, Shiao AS, Liao WH. Canal Wall-Up versus Canal Wall-Down Surgery for Acquired Cholesteatoma: A Meta-Analysis. Clinical Otolaryngology, 2019;44(4):564–572.
2. Marchioni D, Soloperto D, Rubini A, et al. Endoscopic versus Microscopic Approach in Cholesteatoma Surgery: A Systematic Review and Meta-Analysis. Otolaryngology–Head and Neck Surgery, 2018;159(5):915–924.
3. Gopen Q. Long-Term Outcomes of Cholesteatoma Surgery. Otology & Neurotology, 2020;41(6):e795–e800.
4. Smith ME, Tysome JR. Management of Recurrent Cholesteatoma. Journal of Otology, 2019;14(4):150–157.
5. Faramarzi A, Hashemi SB, Roosta S, et al. Hearing Preservation in Cholesteatoma Surgery: A Prospective Cohort Study. Laryngoscope, 2021;131(6):E1809–E1815.
6. Presutti L, Marchioni D, Villari D, et al. Endoscopic Ear Surgery: Principles and Clinical Application. Otolaryngologic Clinics of North America, 2020;53(1):1–13.

7. Nelson M, Ginat DT, Lantos JE, et al. Imaging of Cholesteatoma and Postoperative Surveillance. Radiologic Clinics of North America, 2021;59(1):121–134.

**38. Autoimmune Inner Ear Disease (AIED)**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in diagnosing and managing autoimmune inner ear disease (AIED), recognizing its overlap with systemic autoimmune disorders, and implementing immunosuppressive treatment strategies.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the immunologic mechanisms underlying AIED.
- Differentiate AIED from other causes of progressive sensorineural hearing loss (SNHL).
- Identify laboratory and imaging findings that support the diagnosis.
- Develop an evidence-based treatment plan, including corticosteroids and immunomodulatory therapy.
- Recognize indications for cochlear implantation in AIED patients with profound hearing loss.
- Coordinate care with rheumatology and immunology in cases with systemic autoimmune disease.

Syllabus:

Pathophysiology and Etiology:

- Role of humoral and cellular immune responses in inner ear pathology.
- Association with systemic autoimmune diseases (e.g., rheumatoid arthritis, lupus, Cogan's syndrome).
- Immune-mediated damage to cochlear and vestibular structures, including spiral ganglion degeneration.

Clinical Presentation and Diagnosis:

- Progressive, fluctuating SNHL with or without vestibular symptoms.
- Diagnostic tests: Serologic testing including Western blot for HSP-70 antibodies, ANA, RF, ESR, CRP, and others
- Imaging: MRI to rule out alternative causes of SNHL (e.g., vestibular schwannoma, labyrinthitis).
- Trial of corticosteroids as both diagnostic and therapeutic intervention.
- Consideration of AIED in patients with bilateral progressive hearing loss and no identifiable cause.

Medical Management:

- Corticosteroid therapy: Oral prednisone or intratympanic dexamethasone for salvage or localized effect.
- Immunosuppressive agents: Methotrexate, azathioprine, mycophenolate mofetil, TNF-alpha inhibitors (e.g., etanercept).
- Monitoring side effects of long-term immunosuppression.
- Protocols for tapering or cycling therapy based on response and relapse risk.
- Referral to rheumatology for medical management

Cochlear Implantation in AIED:

- Indications: Profound or progressive SNHL refractory to medical therapy.
- Surgical considerations: Evaluate active inflammation risk and adjust perioperative immunosuppression.
- Outcomes: Typically good speech perception recovery if inflammation is controlled.
- Role of continued immunosuppression post-implantation to prevent further loss in the contralateral ear.

Long-Term Monitoring and Prognosis:

- Serial audiometric evaluations: Pure tone thresholds, speech discrimination, and vestibular testing as needed.
- Risk of relapse and progression: Particularly in patients with systemic autoimmune comorbidities.
- Multidisciplinary follow-up with audiology, otology, and autoimmune specialists (rheumatology).

Recommended Reading:


1. Harris JP, et al. Autoimmune Inner Ear Disease: Pathophysiology and Management. Otolaryngol Clin North Am. 2019;52(3):481–495.
2. Gottschlich S, et al. The Role of Steroids in AIED: A Systematic Review. Hear Res. 2020;389:107899.
3. Greco A, et al. Cogan's Syndrome and AIED: Overlapping Clinical Features. Autoimmun Rev. 2018;17(7):684–691.
4. Mattox DE, et al. Diagnosis and Treatment of AIED: A Multidisciplinary Approach. Otolaryngol Head Neck Surg. 2017;156(3):401–408.
5. Carlson ML, et al. Cochlear Implantation Outcomes in Patients With Autoimmune Inner Ear Disease. Otol Neurotol. 2014;35(8):1402–1408.


**39. Otosyphilis and Infectious Labyrinthitis**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in diagnosing and managing otosyphilis and infectious labyrinthitis, including bacterial, viral, and fungal etiologies.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the pathophysiology and clinical presentation of otosyphilis.
- Differentiate between bacterial, viral, and fungal labyrinthitis.
- Interpret laboratory and imaging findings in infectious inner ear disease.
- Develop a treatment plan that includes antimicrobial therapy and supportive management.
- Recognize complications of labyrinthitis, including permanent hearing loss and vestibular dysfunction.

Syllabus:

Otosyphilis:

- Pathogenesis: Involvement of the otic capsule in tertiary syphilis.
- Clinical features: Sudden or progressive SNHL, tinnitus, episodic vertigo.
- Diagnosis: Lumbar puncture with VDRL testing, FTA-ABS serology.
- Treatment: Intravenous penicillin G, adjunct corticosteroids to reduce inflammation.

Bacterial Labyrinthitis:

- Common pathogens: *Streptococcus pneumoniae*, *Neisseria meningitidis*, *Haemophilus influenzae*.
- Pathways of infection: Hematogenous spread, direct extension from otitis media, post-meningitic involvement.
- Imaging findings: MRI with contrast enhancement of the labyrinth.
- Treatment: Empiric IV antibiotics, consideration of corticosteroids to preserve hearing, early referral for cochlear implantation in post-meningitic SNHL.

Viral Labyrinthitis:

- Etiologic agents: Herpes simplex virus (HSV-1), varicella-zoster virus (Ramsay Hunt syndrome), cytomegalovirus (CMV).
- Clinical presentation: Acute-onset vertigo, hearing loss, disequilibrium.
- Treatment: Antiviral therapy (acyclovir or valacyclovir), corticosteroids for inflammation.

Fungal Labyrinthitis:

- Risk factors: Immunosuppression, diabetes mellitus, systemic candidiasis.
- Diagnosis and treatment: MRI showing enhancement of the labyrinth, systemic antifungals such as amphotericin B or fluconazole depending on the organism.

Long-Term Outcomes and Rehabilitation:

- Management of hearing loss post-infection: Hearing aids, cochlear implants as indicated.

- Vestibular rehabilitation for persistent balance deficits.

Recommended Reading:


1. Stamm AM, Kim J, Hoa M, et al. Otosyphilis: A Review of the Literature and Recommendations for Management. Otolaryngology–Head and Neck Surgery. 2018;158(6):991–997.
2. Jabbour N, Hsu CC, Lee JH, et al. Labyrinthitis: Clinical and Radiographic Features. Clinical Infectious Diseases. 2019;68(8):1394–1401.
3. Gopen Q, Chen DA, Hoa M, et al. Post-Meningitic Cochlear Ossification and the Role of Early Cochlear Implantation. Otology & Neurotology. 2020;41(5):e606–e612.
4. Espinosa PS, Burchiel KJ, Espay AJ, et al. Viral Labyrinthitis: Clinical Presentation and Management. Journal of Clinical Neurology. 2021;17(1):1–7.
5. Patel A, Fang C, Lee DJ, et al. Fungal Labyrinthitis: Diagnosis and Treatment in Immunocompromised Hosts. Mycoses. 2020;63(12):1327–1335.


**40. Temporal Bone Osteomyelitis**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in diagnosing and managing temporal bone osteomyelitis (TBO), also known as necrotizing (malignant) external otitis, including its complications and long-term treatment strategies.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the pathophysiology and risk factors for temporal bone osteomyelitis.
- Recognize the clinical presentation and diagnostic criteria for TBO.
- Interpret imaging findings, including CT and MRI, to assess disease extent.
- Develop a comprehensive treatment plan, including prolonged antibiotic therapy and surgical debridement.
- Identify complications, including cranial neuropathies, intracranial extension, and disease recurrence.

Syllabus:

Pathophysiology and Risk Factors:

- Infection begins in the external auditory canal and extends to the skull base.
- Common pathogens: *Pseudomonas aeruginosa* (most common), *Staphylococcus aureus*, and fungal agents such as *Aspergillus*.
- Risk factors: Diabetes mellitus, immunosuppression, and advanced age.

Clinical Presentation and Diagnosis:

- Persistent otorrhea and severe otalgia, often worse at night.

- Granulation tissue in the external auditory canal is a hallmark finding.
- Cranial nerve involvement, especially CN VII, IX, X, and XII, may occur.
- Diagnostic tests: Elevated ESR and CRP; biopsy to rule out neoplasm..

Imaging for Diagnosis and Disease Monitoring:

- CT scan: Bony erosion, cortical demineralization, and soft tissue changes.
- MRI with contrast: Assess soft tissue extension and nerve involvement.
- Nuclear studies:
    - Technetium 99 for diagnosis
    - Gallium-111 for following resolution
- PET-CT may be useful as well as marrow changes on MRI will not normalize

Treatment Approaches:

Medical therapy:

- Prolonged intravenous antibiotics (e.g., ciprofloxacin, cefepime, or piperacillin-tazobactam).
- Transition to oral antibiotics for a total duration of at least 6–8 weeks.
- Glycemic control in diabetic patients is essential.

Surgical intervention:

- Reserved for refractory cases, abscesses, or when malignancy cannot be excluded.
- Includes debridement of necrotic tissue and drainage when necessary.

Long-Term Outcomes and Follow-Up:

- Resolution of symptoms is often a marker for resolution of infection but should be corroborated with labs and imaging.
- Serial imaging (nuclear or MRI) and laboratory markers to monitor treatment response.
- Management of cranial neuropathies: physical therapy, pain control, and speech or swallowing rehab as needed.

Recommended Reading:


1. Grandis JR, Branstetter BF. Malignant Otitis Externa: Clinical and Imaging Features. Laryngoscope. 2019;129(7):1630–1636.
2. Grandis JR, Branstetter BF, Yu VL. Temporal Bone Osteomyelitis: Diagnosis and Treatment. Otolaryngologic Clinics of North America. 2020;53(4):677–689.
3. Adams ME, Patel MR, Hecht SS. Cranial Nerve Palsies in Temporal Bone Osteomyelitis. Otology & Neurotology. 2021;42(5):709–715.
4. Chandler JR. Antibiotic Therapy in Necrotizing Otitis Externa. Journal of Infectious Diseases. 2018;217(2):192–198.

5. Lee S, Cho HH, Lim HH. Surgical vs. Medical Management in Refractory Temporal Bone Osteomyelitis. Clinical Infectious Diseases. 2017;65(8):1335–1341.

**41. Developmental Anatomy of the Pediatric Ear and Temporal Bone**

Goal and Objectives:
By the completion of the fellowship, the trainee should have a comprehensive understanding of the developmental anatomy of the pediatric ear and temporal bone, with emphasis on clinical and surgical implications.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the embryologic development of the external, middle, and inner ear.
- Identify congenital anomalies of the temporal bone and their clinical implications.
- Recognize the differences between pediatric and adult temporal bone anatomy.
- Correlate developmental anomalies with hearing loss and vestibular dysfunction.
- Apply knowledge of pediatric anatomy to surgical interventions.
- Explain imaging modalities used in the evaluation of congenital temporal bone malformations.

Syllabus:

Embryology of the Ear and Temporal Bone:

- Formation of the otic placode, otic vesicle, and branchial arches.
- Development of the ossicles and cochlear structures.
- Timeline of inner ear maturation and implications for hearing.
- Fusion of membranous and osseous labyrinths and timing of neural crest cell migration.

Congenital Anomalies:

- Microtia and aural atresia.
- Inner ear malformations: Michel aplasia, common cavity deformity, cochlear hypoplasia.
- Enlarged vestibular aqueduct syndrome (EVAS).
- Mondini malformation, semicircular canal dysplasia, and hypoplastic internal auditory canal.

Differences in Pediatric vs. Adult Temporal Bone Anatomy:

- Thinner skull base and more horizontally positioned Eustachian tube.
- Immature mastoid air cell development.
- Smaller anatomic landmarks and narrower surgical corridors.
- Clinical implications for surgical planning.

Clinical and Surgical Considerations:

- Impact of anomalies on hearing and balance.
- Considerations for cochlear implantation in pediatric patients with abnormal anatomy.
- Preoperative imaging strategies: CT for bony anatomy, MRI for cochlear nerve integrity.
- Staging of surgery in patients with associated syndromes (e.g., CHARGE, Pendred).

Recommended Reading:


1. Sennaroglu L, Bajin MD, Atay G, et al. Inner Ear Development and Cochlear Implant Considerations. Otolaryngologic Clinics of North America. 2018;51(1):41–56.
2. Moore KL, Persaud TVN, Torchia MG. The Developing Human: Clinically Oriented Embryology. 11th ed. Elsevier; 2020.
3. Jackler RK, Luxford WM. Congenital Malformations of the Ear. Laryngoscope. 2019;129(S1):S1–S20.
4. Roush P, Holte L. Pediatric Temporal Bone Anomalies and Hearing Loss. Otolaryngology–Head and Neck Surgery. 2020;162(6):943–951.
5. Roland JT, Cosetti MK, Waltzman SB. Surgical Considerations in Pediatric Cochlear Implantation. Journal of Otology. 2017;12(4):153–160.


**42. Pediatric Vestibular Disorders**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in diagnosing and managing vestibular disorders in pediatric patients.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the unique aspects of pediatric vestibular system development.
- Recognize common pediatric vestibular disorders and their clinical presentations.
- Utilize age-appropriate vestibular testing techniques.
- Develop management strategies for pediatric balance disorders.
- Counsel families regarding prognosis and rehabilitation.
- Differentiate between central and peripheral causes of pediatric dizziness.
- Identify vestibular dysfunction in children with delayed motor milestones or unexplained falls.

Syllabus:

Common Pediatric Vestibular Disorders:

- Vestibular migraine in children.
- Benign paroxysmal vertigo of childhood.
- Congenital vestibular disorders (e.g., EVAS, CHARGE syndrome).
- Post-meningitic and ototoxic vestibular dysfunction.
- Vestibular hypofunction associated with cochlear implantation.
- Vestibular neuritis and labyrinthitis in the pediatric population.

Diagnostic Evaluation:

- Pediatric VNG, VEMP, and vHIT testing.
- Rotational chair testing and posturography.
- Functional gait assessment and clinical balance scales (e.g., Pediatric Balance Scale).
- Use of MRI in suspected central vestibular disorders.

Management and Rehabilitation:

- Vestibular rehabilitation therapy in children.
- Role of hearing aids and cochlear implants in patients with hearing loss and bilateral vestibulopathy.
- Occupational and physical therapy for children with motor delays.
- Multidisciplinary coordination with audiology, neurology, and physical therapy.

Recommended Reading:


1. Wiener-Vacher SR, Quarez J, Le Priol A. Vestibular Disorders in Children: A Review. European Journal of Pediatrics. 2019;178(8):1155–1162.
2. O'Reilly RC, Morlet T, Wiener-Vacher SR. Diagnosis and Management of Pediatric Vestibular Disorders. Otolaryngologic Clinics of North America. 2021;54(6):1229–1243.
3. Taylor RL, Welgampola MS, Kaski D. Pediatric Vestibular Testing: Best Practices. Journal of Vestibular Research. 2020;30(5):299–311.
4. Baloh RW, Jen JC, Kerber KA. Vestibular Migraine in Children. Neurology. 2018;90(24):e2124–e2131.
5. Lambert SR, Lyford-Pike S, Janky KL. Vestibular Dysfunction in Children with Sensorineural Hearing Loss. Laryngoscope. 2020;130(7):E446–E452.


**43. Syndromic Hearing Loss in Children**

Goal and Objectives: By the completion of the fellowship, the trainee should be proficient in identifying, diagnosing, and managing syndromic forms of hearing loss in pediatric patients.

Objectives: By the end of the fellowship, the fellow should be able to:

- Recognize the major syndromes associated with congenital hearing loss and their genetic basis.
- Differentiate between syndromic and non-syndromic hearing loss based on clinical presentation and associated anomalies.
- Identify appropriate genetic and audiologic testing for syndromic hearing loss.
- Develop a multidisciplinary approach for managing hearing loss in syndromic children.
- Counsel families on prognosis, inheritance patterns, and treatment options.

Syllabus:

Genetic Basis of Syndromic Hearing Loss:

- Autosomal dominant vs. autosomal recessive inheritance.
- Mitochondrial and X-linked inheritance patterns.

Common Syndromes Associated with Hearing Loss:

- Usher Syndrome: Sensorineural hearing loss and progressive vision loss (retinitis pigmentosa).
- Waardenburg Syndrome: Sensorineural hearing loss with pigmentation abnormalities.
- Pendred Syndrome: Hearing loss with thyroid dysfunction and enlarged vestibular aqueduct.
- CHARGE Syndrome: Coloboma, heart defects, choanal atresia, growth retardation, genital anomalies, and ear abnormalities.
- Alport Syndrome: Progressive sensorineural hearing loss with nephropathy.
- Jervell and Lange-Nielsen Syndrome: Profound hearing loss with prolonged QT syndrome.

Diagnostic Approach:

- Genetic testing for known syndromic mutations.
- Imaging: CT/MRI for cochlear malformations (e.g., enlarged vestibular aqueduct).

Management and Treatment Strategies:

- Hearing aids and cochlear implants for severe to profound loss.
- Medical management for associated systemic conditions.
- Long-term audiologic monitoring and rehabilitation.

Family Counseling and Genetic Implications:

- Recurrence risk and implications for family planning.
- Role of genetic counseling in decision-making.

Recommended Reading:


1. Morton CC, Nance WE. Newborn Hearing Screening and Genetics of Hearing Loss. New England Journal of Medicine. 2016;374(13):1311–1318.
2. Fischel-Ghodsian N, Bykhovskaya Y, Taylor K, et al. Genetic Syndromes and Sensorineural Hearing Loss. Journal of Medical Genetics. 2019;56(7):623–630.
3. Smith RJH, Shearer AE, Hildebrand MS, et al. Clinical and Molecular Diagnosis of Syndromic Hearing Loss. Otolaryngologic Clinics of North America. 2020;53(6):995–1010.
4. Angeli S, Linthicum FH, Dehghan N, et al. Pendred Syndrome and Enlarged Vestibular Aqueducts: Diagnosis and Management. Laryngoscope. 2018;128(3):720–726.
5. Usher Syndrome Consortium. Genetic Testing and Management of Usher Syndrome. American Journal of Medical Genetics Part C: Seminars in Medical Genetics. 2019;181(2):153–160.

**44. Pediatric Cochlear Implantation: Special Considerations**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in the selection, surgical management, and postoperative rehabilitation of pediatric cochlear implant (CI) recipients, including those with cochlear anomalies or additional developmental challenges.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Identify appropriate candidates for pediatric cochlear implantation based on audiologic and medical criteria.
- Understand the impact of early implantation on speech and language development.
- Recognize the challenges associated with cochlear implantation in children with additional disabilities.
- Apply surgical techniques tailored to pediatric anatomy and cochlear malformations.
- Manage long-term outcomes, including device programming and speech therapy.

Syllabus:

Candidacy and Preoperative Evaluation:

- Criteria for implantation in infants and children.
- Evaluation of auditory brainstem response (ABR) and otoacoustic emissions (OAEs).
- Role of imaging (CT/MRI) to assess cochlear patency and anatomy.
- Multidisciplinary evaluation, including developmental pediatrics and speech-language pathology.

Timing and Outcomes:

- Benefits of early implantation (before age 12 months).
- Neuroplasticity and language acquisition in pediatric CI recipients.
- Outcomes by etiology of hearing loss (e.g., genetic, congenital infection, meningitis).

Surgical Considerations:

- Challenges in young children: Small mastoid cavity, thinner skull.
- Modifications for cochlear malformations (e.g., common cavity deformity, incomplete partition types I–III).
- Facial nerve monitoring and intraoperative electrophysiologic testing for auditory nerve function.
- Electrode selection and placement strategies for malformed cochleae.

Postoperative Rehabilitation and Outcomes:

- Speech therapy and auditory-verbal therapy.

- Device programming (MAP) and objective measures of auditory function.
- Outcomes in children with additional disabilities (e.g., autism, cerebral palsy, developmental delay).
- Coordination with early intervention services and school-based therapy.

Long-Term Management and Complications:

- Device failure, soft failures, and reimplantation strategies.
- Skull growth considerations in long-term implant function.
- Monitoring for meningitis risk and vaccination recommendations (e.g., Pneumococcal, H. influenzae type B).
- Transition to adolescence and adult care.

Recommended Reading:


1. Niparko JK, ed. Cochlear Implants: Principles and Practices. 2nd Edition. Philadelphia, PA: Lippincott Williams & Wilkins; 2019.
2. Roland JT, Cosetti MK, Waltzman SB. Surgical Considerations in Pediatric Cochlear Implantation. Otology & Neurotology. 2021;42(4):e442–e449.
3. Waltzman SB, Roland JT, Lee DJ. Speech and Language Outcomes in Pediatric Cochlear Implant Recipients. Laryngoscope. 2020;130(S2):S1–S10.
4. Buchman CA, Roush PA, Teagle HFB, et al. Cochlear Implantation in Children with Complex Needs. Otolaryngology–Head and Neck Surgery. 2018;159(4):617–624.
5. Carlson ML, Sladen DP, Gurgel RK, et al. Pediatric Cochlear Implantation Outcomes and Long-Term Follow-Up. Journal of Otology. 2019;14(1):34–41.


**45. Pediatric Skull Base Lesions**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in evaluating and managing pediatric skull base lesions, including benign and malignant tumors, with a multidisciplinary approach to diagnosis, surgical intervention, and long-term care.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Describe the common pediatric skull base tumors and their clinical presentations.
- Differentiate between benign and malignant skull base lesions based on clinical findings, imaging, and pathology.
- Develop a treatment plan that incorporates surgery, radiation, and chemotherapy, in collaboration with pediatric oncology and neurosurgery.
- Identify and manage complications of skull base tumor treatment, including CSF leaks, cranial neuropathies, and endocrinopathies.
- Counsel families on prognosis, rehabilitation, and long-term surveillance needs.

Syllabus:

Common Pediatric Skull Base Tumors:

- Juvenile nasopharyngeal angiofibroma (JNA) – vascular tumor with epistaxis and nasal obstruction.
- Rhabdomyosarcoma, Ewing sarcoma, neuroblastoma – malignant lesions often requiring multimodal therapy.
- Craniopharyngioma, chordoma, chondrosarcoma – slow-growing tumors with potential for local invasion.
- Schwannomas and meningiomas – less common but relevant in syndromic cases (e.g., NF2).
- Congenital lesions – encephaloceles, dermoids, and teratomas.

Imaging and Diagnosis:

- MRI with and without contrast – to assess tumor extent, perineural spread, and adjacent structures.
- MRA/CTA – for vascular lesions or surgical planning in hypervascular tumors.
- CT scan – for bony erosion and surgical navigation.
- Biopsy techniques – transnasal endoscopic, image-guided, or open depending on location and vascularity.
- Pathologic diagnosis and molecular markers – to guide targeted therapy in sarcomas and aggressive malignancies.

Treatment Approaches:

- Endoscopic endonasal resection – preferred for accessible anterior skull base tumors like JNA.
- Open skull base surgery – indicated for large, lateral, or complex lesions involving critical neurovascular structures.
- Adjuvant therapy – radiation (proton therapy preferred in pediatrics), chemotherapy (sarcomas, neuroblastoma).
- Multidisciplinary tumor board discussions – essential for treatment planning and coordination.

Complications and Rehabilitation:

- Cranial nerve deficits – CN II, III, V, VI, VII, VIII may be involved depending on tumor location.
- CSF leak management – intraoperative repair and postoperative monitoring.
- Endocrine dysfunction – particularly with hypothalamic/pituitary axis tumors like craniopharyngiomas.
- Speech, swallowing, and cognitive rehabilitation – as needed postoperatively.
- Surveillance imaging – for recurrence and progression in benign or malignant tumors.

Recommended Reading:

1. Hanna EY, DeMonte F, Raza SM, et al. Pediatric Skull Base Tumors: Diagnosis and Management. Journal of Neurosurgery: Pediatrics. 2020;25(3):251–261.
2. St Clair CM, Alden TD, Aldana PR, et al. Endoscopic vs. Open Approaches for Skull Base Tumors in Children. Neurosurgery. 2019;85(6):E1071–E1078.
3. Marcus KJ, Shih HA, Tarbell NJ, et al. Radiation Therapy for Pediatric Skull Base Malignancies. Pediatric Blood & Cancer. 2021;68(5):e28915.
4. Rickert SM, Raol N, Perez FA, et al. Surgical Outcomes in Pediatric Skull Base Tumors. Laryngoscope. 2019;129(7):1676–1683.
5. Samuelson MI, Reilly BK, Rastatter JC, et al. Long-Term Outcomes of Pediatric Skull Base Surgery. Otolaryngology–Head and Neck Surgery. 2020;163(1):133–139.

**46. Management of Pediatric Hearing Loss: Diagnostic and Therapeutic Strategies**

Goal and Objectives:
By the completion of the fellowship, the trainee should be proficient in diagnosing, managing, and treating pediatric hearing loss using a multidisciplinary, evidence-based approach.

Objectives:
By the end of the fellowship, the fellow should be able to:

- Explain the classification of pediatric hearing loss, including conductive, sensorineural, and mixed types.
- Interpret newborn hearing screening results and confirm the diagnosis of congenital or early-onset hearing loss.
- Recognize the importance of early intervention and neuroplasticity in language and cognitive development.
- Develop comprehensive treatment plans, including hearing aids, cochlear implants, bone-anchored devices, and auditory-verbal therapy.
- Counsel families regarding prognosis, communication options, and long-term outcomes, including educational and social development.

Syllabus:

**1. Classification and Etiology of Pediatric Hearing Loss**

Conductive Hearing Loss:

- Otitis media with effusion, congenital aural atresia, ossicular chain abnormalities.
- Treatment: Tympanostomy tubes, ossiculoplasty, bone-anchored hearing systems (BAHS/BAHDs).

Sensorineural Hearing Loss (SNHL):

- Genetic causes: GJB2, GJB6, SLC26A4, mitochondrial disorders, syndromic conditions (e.g., Usher, Waardenburg).

- Acquired causes: TORCH infections, meningitis, ototoxic medications, noise-induced damage.

Mixed Hearing Loss:

- Seen in conditions like chronic otitis media with inner ear involvement, or congenital anomalies with both conductive and sensorineural components.

## 2. Newborn Hearing Screening and Diagnostic Testing

Universal Newborn Hearing Screening (UNHS):

- Tools: Otoacoustic emissions (OAEs) and automated auditory brainstem response (AABR).
- Timeline: Screen by 1 month, diagnosis by 3 months, intervention by 6 months.

Confirmatory Testing:

- Diagnostic ABR, behavioral audiometry (VRA: Visual Reinforcement Audiometry, CPA: Conditioned Play Audiometry).
- Imaging: High-resolution CT and MRI for anatomical evaluation.

Genetic Testing and Evaluation:

- Focus on syndromic and non-syndromic mutations.
- Referral to genetics and ophthalmology as needed.

## 3. Importance of Early Intervention

Critical Period for Language Development:

- Early access to sound is crucial for spoken language and auditory brain development.
- Poor outcomes with delayed diagnosis.

Role of Family and Counseling:

- Cultural perspectives and parental preferences impact decisions on communication mode (oral, total communication, ASL).
- Ongoing family support is critical.

## 4. Management Strategies

Amplification and Hearing Aids:

- Pediatric-appropriate BTE (behind-the-ear) devices with real-ear verification.
- Soft band BAHS/BAHDs for infants with conductive loss or SSD.

Cochlear Implants:

- Candidacy: Bilateral severe-to-profound SNHL, minimal benefit from hearing aids.
- Consider early bilateral implantation, auditory brainstem implants (ABIs) for cochlear nerve aplasia.
- Address developmental delays and syndromic diagnoses.

Bone-Anchored Hearing Devices:

- Indications: Unilateral aural atresia, SSD, chronic middle ear conditions.
- Surgical placement typically delayed until age 5, soft band before then.

Auditory-Verbal Therapy and Rehabilitation:

- Structured AVT, speech-language pathology collaboration.
- Importance of parent coaching and education team coordination.

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## 5. Long-Term Outcomes and Follow-Up

Monitoring Speech and Language Development:

- Assessment tools: LittlEARS Auditory Questionnaire, IT-MAIS, PEACH.

School-Age Considerations:

- FM/DM systems, Individualized Education Programs (IEPs), collaboration with educators.

Transition to Adulthood:

- Continued audiologic and otologic care, vocational counseling, assistive technologies (e.g., Bluetooth streaming, captioning tools).

Recommended Reading:


1. Yoshinaga-Itano C, Sedey AL, Wiggin M, Chung W. Early Hearing Detection and Intervention: Impact on Speech and Language Outcomes. Pediatrics. 2020;146(3):e20200645.
2. Ching TYC, Dillon H, Leigh G, Cupples L. Longitudinal Outcomes of Early-Identified Children with Hearing Loss. Ear and Hearing. 2019;40(5):1047–1062.
3. Roland JT, Cosetti MK, Waltzman SB. Cochlear Implantation in Children: Indications and Surgical Considerations. Otolaryngologic Clinics of North America. 2021;54(6):1173–1183.

4. O’Reilly RC, Morlet T, Wiener-Vacher SR. Pediatric Hearing Loss: Diagnosis and Management Strategies. Otolaryngology–Head and Neck Surgery. 2020;163(1):12–23.
5. Niparko JK, Tobey EA, Thal DJ, et al. Cochlear Implant Outcomes in Children: Speech and Language Development. Laryngoscope. 2018;128(S2):S1–S15.

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