Yonsei University Wonju College of Medicine
Department of Emergency Medicine

한국형 가이드라인 II:

돌발성 난청

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SSNHL

1. Background on hearing and hearing loss

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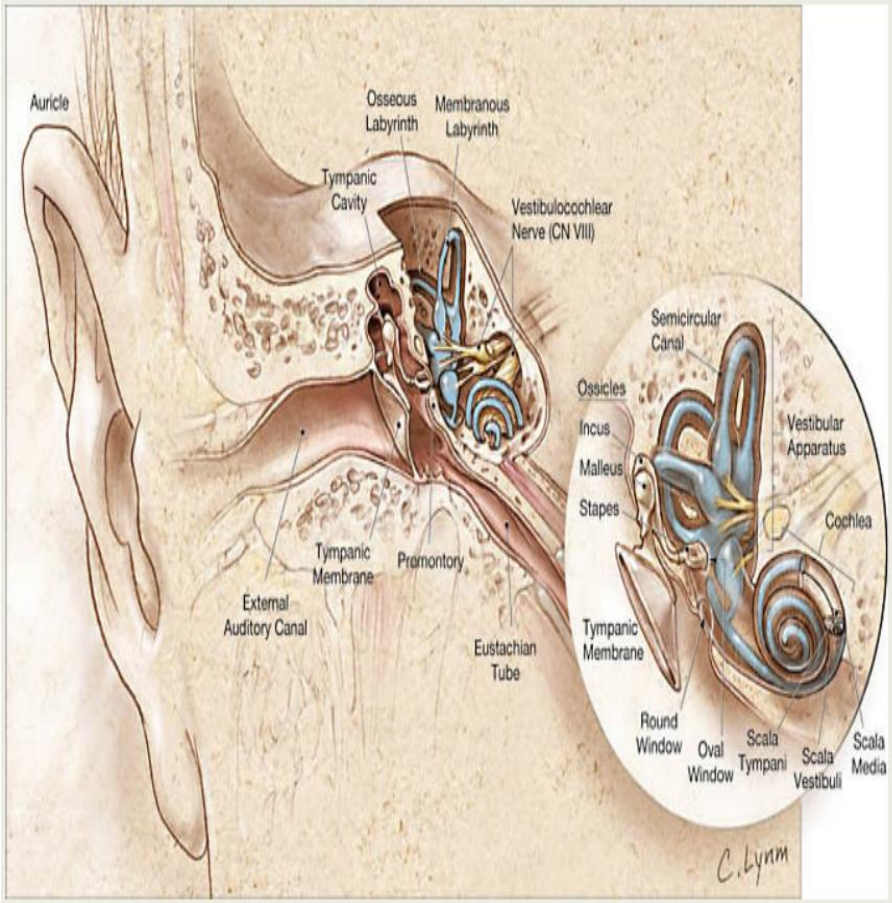
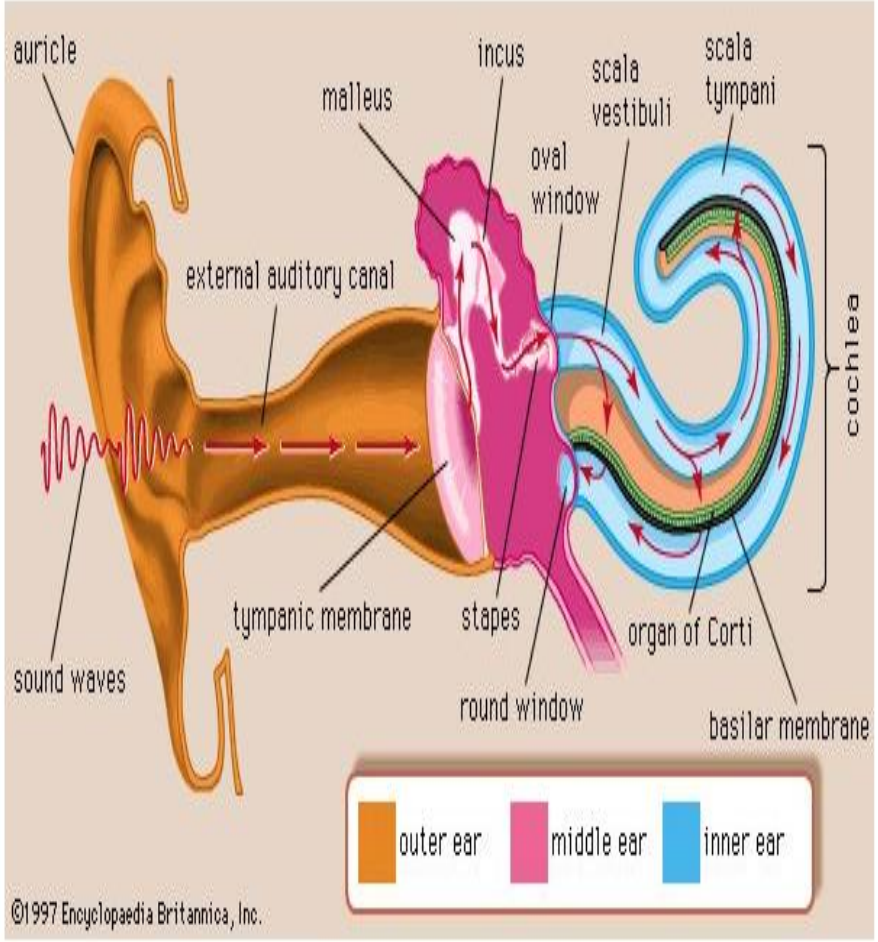
Background

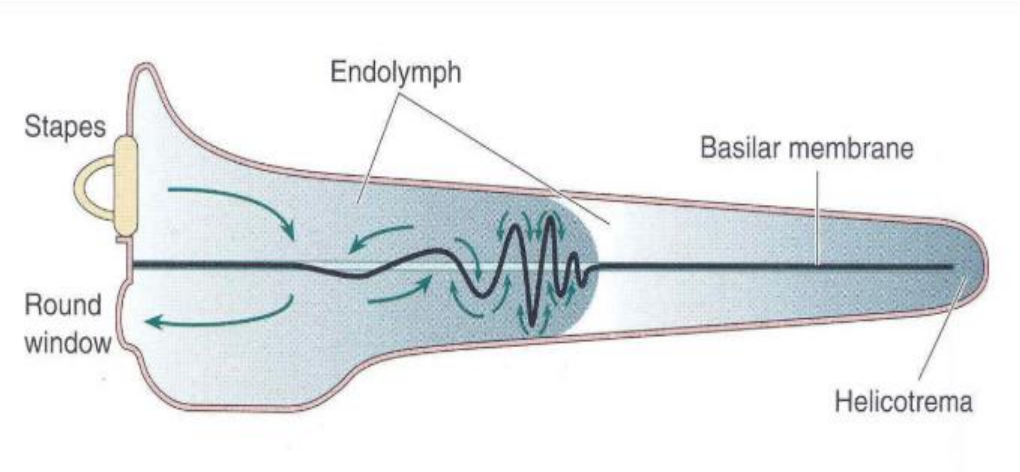


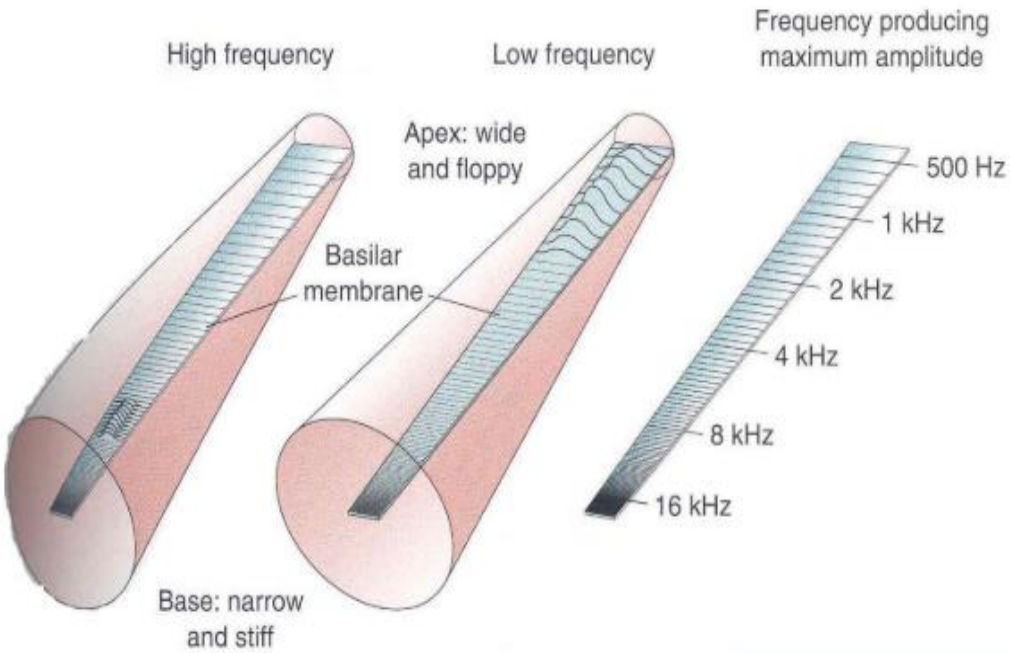
Yueh, B. et al. JAMA 2003



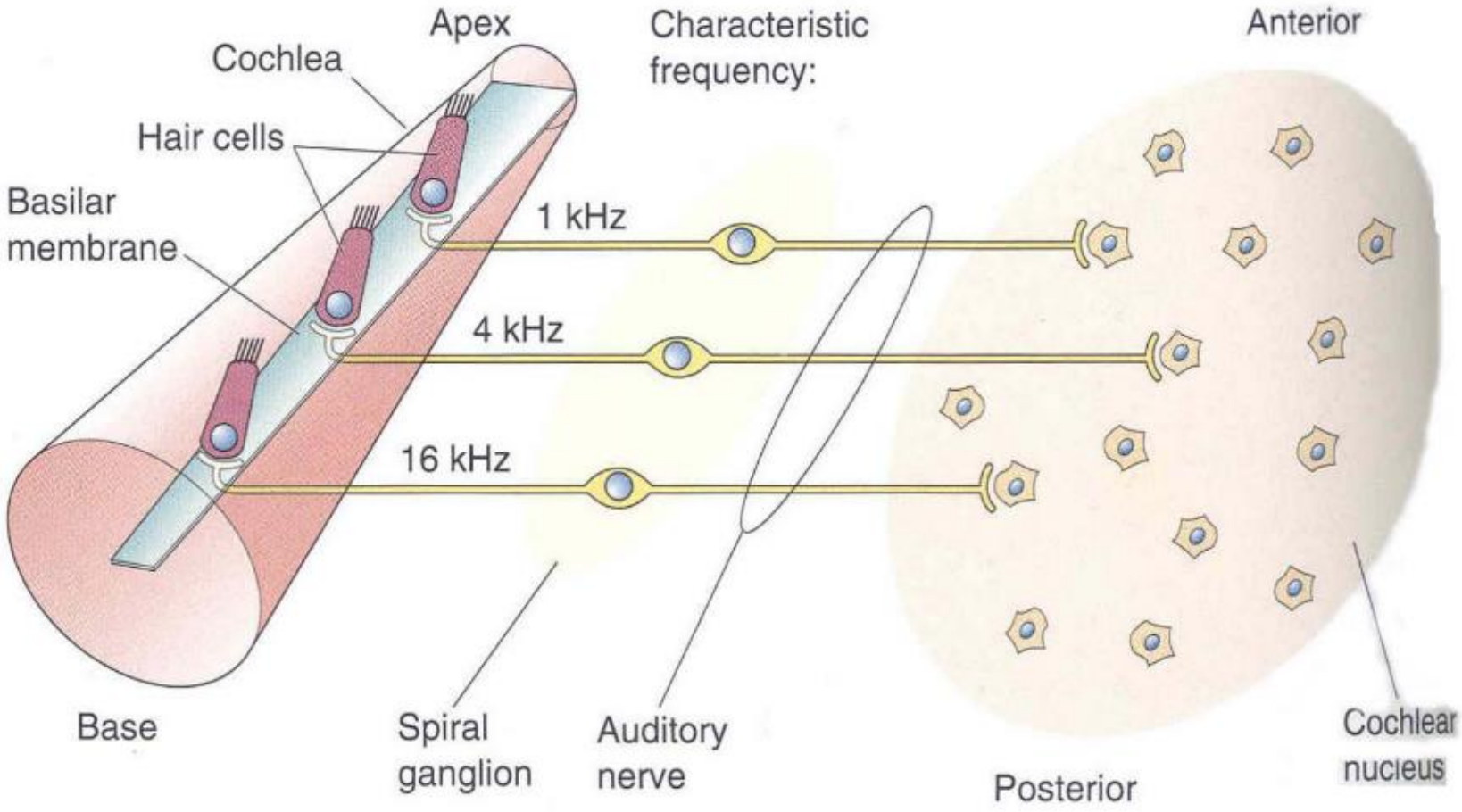
Basilar membrane

- Stiff at base
- Loose at apex
- Tonotopic
  - 각각의 주파수에 따라 구별해 감지
  - 청각세포들이 한쪽은 고음 반대편으로 갈수록 저음을 인식





Tonotopy preserved to cortex



Benefits of two ears (Binaural hearing)

- Head Shadow effect
  - One ear always in optimal position for a sound
- Binaural processing
  - Sounds are perceived differently when heard in both ears
- Sound localization
  - Identifying the source of a sound
- Hearing in background noise

2. Explain the impairment caused by sudden loss of hearing

Sudden hearing loss

- Defined as ≥ 30dB sensorineurial loss in at least 3 contiguous frequencies over 72 hours
  - Wilson et al. 1980
- 7-15% have identifiable cause – may allow for specific treatment
- Majority “idiopathic”
- Etiology, work-up, and care still controversial
- > 400 articles on Pubmed

Epidemiology

- Incidence 5-20/100,000
- 60,000 new cases annually in US
- Many affected individuals who recover never present
- Peak incidence 5th-6th decade
- Men = women
- < 2% bilateral, typically sequential
- Symptoms
  - Tinnitus (roaring) 40-90%
  - Dizziness 30-56%
  - Aural fullness (40-50%)
  - Ear ‘popping’

- Byl, 1984
- Fetterman et al, 1996
- Xenellis et al, 2006
- Chau et al, 2010

Causes – Infectious 13%

- Lyme disease (Borrelia burgdorferi)
  - Early –erythema migrans
  - Up to 20% SSNHL with positive Lyme titers
  - Rash often absent (90%), 40% without risk factors
  - Hearing recovery similar between Lyme- and Lyme+ pts

- Syphilis (Treponema pallidum)
  - Neurosyphilis – usually late stage, can be early
  - Consider in immunosuppressed or high-risk patients
  - Consider in bilateral SNHL

- Lorenzi et al, 2003
- Heman-Ackah et al, 2010
- Mishra et al, 2008
- Marra et al, 2009

Causes – Otologic 5%

- Up to 5%
- Meniere's Disease
  - 15/100,000
  - Typically low frequency hearing loss
  - Recurrent
- Autoimmune inner ear disease
  - 5/100,000
  - Bilateral SNHL over weeks to months
- Vestibular Schwannoma (acoustic neuroma)
  - Incidence 1-4/100,000
- Neoplastic causes (2%)
  - Spontaneous return or return of hearing after treatment does not rule out

Causes – Systemic disease

- Autoimmune disease
  - Cogan's syndrome:
    non-syphilitic interstitial keratitis and audiovestibular disease
  - SLE
  - Wegener's granulomatosis
- Thyroid disease
  - 1-15% with SSNHL
  - TSH work-up

- Narozny et al, 2006
- Heman-Ackah et al, 2010

Causes – Vascular (3%)

- Cochlear blood flow – 2 end arteries, lack collateral
  = High risk
- Sudden onset of SNHL similar to ischemic events
- Vascular risk factors (tobacco, HTN, hyperlipidemia, DM) may be related to increased risk of SSNHL

- Ballesteros et al, 2009
- Capaccio et al, 2007

Causes – Viral

- Infection/reactivation causes inner ear inflammation or damage
- Antibodies to CMV, HSV, HZV, influenza B, mumps, enterovirus, rubeola all isolated from SSNHL patients
- Temporal bones in patients with SSNHL show similar findings to viral labyrinthitis
- Cochlear/labyrinthine enhancement on MRI in 4-9%
- Inoculation of animals with HSV induced SNHL

- Mentel et al 2004
- Wilson et al, 1983
- Chon et al, 2003

Natural history

- Discoverable causes often cause permanent hearing loss
- Some with SSNHL regain hearing
  - Recovery without treatment = 32-65%
  - Typically recovery within 2 weeks of onset
  - Complete recovery – 36%

- Xenelis et al, 2003
- Byl, 1984

SSNHL

3. The 2019 Clinical Practice Guideline

Check for updates

Guidelines Executive Summary

AMERICAN ACADEMY OF OTOLARYNGOLOGY–HEAD AND NECK SURGERY FOUNDATION

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Clinical Practice Guideline: Sudden Hearing Loss (Update) Executive Summary

Sujana S. Chandrasekhar, MD[1,2,3], Betty S. Tsai Do, MD[4], Seth R. Schwartz, MD, MPH[5], Laura J. Bontempo, MD, MEd[6], Erynne A. Faucett, MD[7], Sandra A. Finestone, PsyD[8], Deena B. Hollingsworth, MSN, FNP-BC[9], David M. Kelley, MD[4], Steven T. Kmucha, MD, JD[10], Gul Moonis, MD[11], Gayla L. Poling, PhD, CCC-A[12], J. Kirk Roberts, MD[11], Robert J. Stachler, MD[13], Daniel M. Zeitler, MD[5], Maureen D. Corrigan[14], Lorraine C. Nnacheta, MPH, DrPH[14], Lisa Satterfield, MS, MPH[14], and Taskin M. Monjur[14]

Clinical practice guideline update: 2019 (2012)

- New RCTs and Strong recommendations included
- Increased emphasis on urgency of evaluation and treatment

Table 1. Definitions of Common Terminology.

Term	Definition
Sudden hearing loss (SHL)	A rapid-onset subjective sensation of hearing impairment in 1 or both ears
Sensorineural hearing loss (SNHL)	Hearing loss resulting from abnormal function of the cochlea, auditory nerve, or higher aspects of central auditory perception or processing
Conductive hearing loss (CHL)	Hearing loss resulting from a problem conducting sound waves anywhere along the route through the outer ear, tympanic membrane, or middle ear
Mixed hearing loss (MHL)	Hearing loss resulting from both SNHL and CHL occurring in the same ear
Sudden sensorineural hearing loss (SSNHL)	A subset of SHL that (a) is sensorineural in nature, (b) occurs within a 72-hour window, and (c) consists of a decrease in hearing of greater than or equal to 30 decibels affecting at least 3 consecutive frequencies
Idiopathic sudden sensorineural hearing loss (ISSNHL)	SSNHL with no identifiable cause despite adequate investigation
Salvage therapy	Any therapy offered after 2 weeks from symptom onset (even if initial therapy was observation)

Clinical practice guideline

Table 3. Strength of Action Terms in Guideline Statements and Implied Levels of Obligation.

Strength	Definition	Implied Obligation
Strong recommendation	A strong recommendation means the benefits of the recommended approach clearly exceed the harms (or, in the case of a strong negative recommendation, that the harms clearly exceed the benefits) and that the quality of the supporting evidence is high (grade A or B).[a] In some clearly identified circumstances, strong recommendations may be made based on lesser evidence when high-quality evidence is impossible to obtain and the anticipated benefits strongly outweigh the harms.	Clinicians should follow a strong recommendation unless a clear and compelling rationale for an alternative approach is present.
Recommendation	A recommendation means the benefits exceed the harms (or, in the case of a negative recommendation, that the harms exceed the benefits), but the quality of evidence is not as high (grade B or C).[a] In some clearly identified circumstances, recommendations may be made based on lesser evidence when high-quality evidence is impossible to obtain and the anticipated benefits outweigh the harms.	Clinicians should also generally follow a recommendation but should remain alert to new information and sensitive to patient preferences.
Option	An option means that either the quality of evidence is suspect (grade D)[a] or that well-done studies (grade A, B, or C)[a] show little clear advantage to one approach vs another.	Clinicians should be flexible in their decision making regarding appropriate practice, although they may set bounds on alternatives; patient preference should have a substantial influencing role.

[a]Adapted from the American Academy of Pediatrics classification scheme[31] (see **Table 2** for definitions of evidence grades).

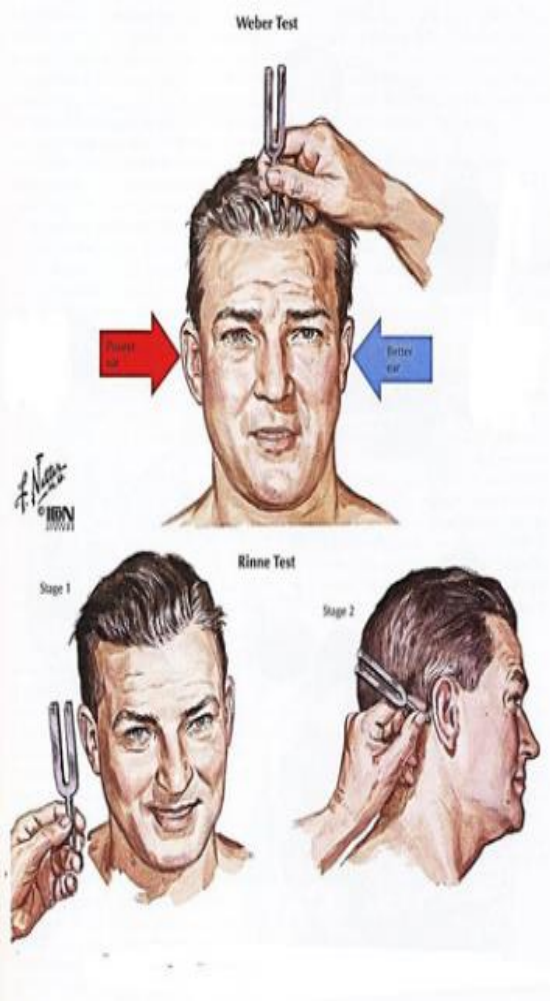
Clinical practice guideline

Table 4. Summary of Guideline Key Action Statements.

Statement	Action	Strength
1. Exclusion of conductive hearing loss	Clinicians should distinguish sensorineural hearing loss (SNHL) from conductive hearing loss (CHL) when a patient first presents with SHL	Strong recommendation
2. Modifying factors	Clinicians should assess patients with presumptive SSNHL through history and physical examination for bilateral SHL, recurrent episodes of SHL, and/or focal neurologic findings.	Recommendation
3. Computed tomography	Clinicians should *not* order routine computed tomography (CT) of the head in the initial evaluation of a patient with presumptive SSNHL.	Strong recommendation against
4. Audiometric confirmation of SSNHL	In patients with SHL, clinicians should obtain, or refer to a clinician who can obtain, audiometry as soon as possible (within 14 days of symptom onset) to confirm the diagnosis of SSNHL	Recommendation
5. Laboratory testing	Clinicians should *not* obtain routine laboratory tests in patients with SSNHL.	Strong recommendation against
6. Retrocochlear pathology	Clinicians should evaluate patients with SSNHL for retrocochlear pathology by obtaining magnetic resonance imaging or auditory brainstem response (ABR).	Recommendation
7. Patient education	Clinicians should educate patients with SSNHL about the natural history of the condition, the benefits and risks of medical interventions, and the limitations of existing evidence regarding efficacy.	Strong recommendation
8. Initial corticosteroids	Clinicians may offer corticosteroids as initial therapy to patients with SSNHL within 2 weeks of symptom onset.	Option
9a. Initial therapy with hyperbaric oxygen therapy	Clinicians may offer, or refer to a clinician who can offer, hyperbaric oxygen therapy (HBOT) combined with steroid therapy within 2 weeks of onset of SSNHL.	Option
9b. Salvage therapy with hyperbaric oxygen therapy	Clinicians may offer, or refer to a clinician who can offer, HBOT combined with steroid therapy as salvage within 1 month of onset of SSNHL.	Option
10. Intratympanic steroids for salvage therapy	Clinicians should offer, or refer to a clinician who can offer, intratympanic steroid therapy when patients have incomplete recovery from SSNHL 2 to 6 weeks after onset of symptoms.	Recommendation
11. Other pharmacologic therapy	Clinicians should *not* routinely prescribe antivirals, thrombolytics, vasodilators, or vasoactive substances to patients with SSNHL.	Strong recommendation against
12. Outcomes assessment	Clinicians should obtain follow-up audiometric evaluation for patients with SSNHL at the conclusion of treatment and within 6 months of completion of treatment.	Recommendation
13. Rehabilitation	Clinicians should counsel patients with SSNHL who have residual hearing loss and/or tinnitus about the possible benefits of audiological rehabilitation and other supportive measures.	Strong recommendation

Guideline Key Action Statements -1

- Statement 1
  - Exclusion of Conductive hearing loss: SR
    - Outer ear or middle ear problem
- Statement 2
  - Modifying factors: R
  - Clinicians should assess patients with presumptive SSNHL for bilateral SHL, recurrent episodes of SHL, or focal neurologic findings
- Statement 3
  - CT: SR against
  - Clinicians should not order routine CT of the head in the initial evaluation
  - HRCT temporal bones if needed



Guideline Key Action Statements -2

- Statement 4
  - Audiometric Confirmation: R
  - Clinicians should obtain, or refer to a clinician who can obtain audiometry ASAP to confirm SSNHL within 14 days of symptom onset
- Statement 5
  - Laboratory testing: SR against
  - Clinicians should not obtain routine laboratory tests in patients with SSNHL
  - Cost containment, avoidance of stress and anxiety of patient, avoidance of false positives
  - Specific test may be useful: vascular, Lyme titers, TFTs, immune markers, FTA/VDRL
- Statement 6
  - Retrocochlear pathology: R
  - Clinicians should evaluate patients with SSNHL for retrocochlear pathology by MRI or auditory brainstem response

Guideline Key Action Statements -3

- Statement 7
  - Patient education: SR
  - Clinicians should educate patients with SSNHL about the natural history of the condition, the benefits and risks of medical interventions, and the limitations of existing evidence regarding efficacy.
- Statement 8
  - Initial corticosteroids: option
  - Hearing improvements
  - Only for 2 weeks
- Statement 9a
  - Initial therapy with HBOT: option
  - For 2 weeks of onset with the steroid therapy
- Statement 9b
  - Salvage therapy with HBOT: option
  - For one month of onset with steroid therapy
- Statement 10
  - IT steroids for salvage therapy: R
  - IT steroid therapy when patients have incomplete recovery from SSNHL 2 to 6 weeks after onset
- Statement 11
  - Other pharmacologic therapy: SR against
  - Should not routinely prescribe antivirals, thrombolytic, vasodilators, or vasoactive substances

Guideline Key Action Statements -4

- Statement 12
  - Outcomes assessment: R
  - Should obtain follow-up audiometric evaluation for patients with SSNHL at the conclusion of treatment and within 6 months of completion of treatment.

- Statement 13
  - Rehabilitation: SR
  - Should counsel patients with SSNHL who have residual hearing loss and/or tinnitus about the possible benefits of audiological rehabilitation and other supportive measures.
  - Improve QOL, functionality, hearing and tinnitus and emotional support

Prognosis

- 35-65% regain some hearing even without therapy with average gains of 35dB
- 35-65% have no improvement
- Advanced age (>60) correlated with poor prognosis
- Factors affecting recovery rates:
  - Severity, duration, audiogram 'shape', vertigo, time to treatment.
- Time to presentation correlates with hearing recovery
  - Within 7days = 87%
  - 2 weeks = 52%
  - Longer than 3 months = <10%
- **Otologic emergency**

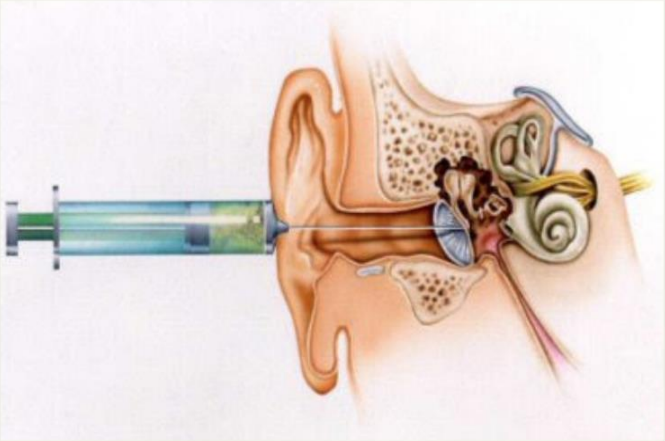
Treatment

	Location	Journal	Articles	Conclusion
Conlin & Parnes, 2007	Ontario, Canada	*Arch Otolaryngol Head Neck Surg*	2	Unclear benefit of steroid vs. placebo
Labus et al. 2010	Brussels, Belguim	*Laryngoscope*	6	Medical therapy > placebo but not significant
Wei et al. 2013	Melbourne, Australia	*Coch Database Sys Rev*	3	Unclear benefit of steroid vs. placebo

Steroid Therapy

- Oral vs. IT
  - 3 RCT's
  - Hong 2009, Dispenza 2011, Rauch 2012
  - Oral were equivalent to IT
- Combined (Oral+IT) vs Oral
  - 6 RCT's
  - 3 no difference
  - 3 combination > oral
    - 88% vs 44% Battaglia
    - 46% vs 21% Zhou
    - 89% vs 61% Gundogan
- Combined (Oral+IT) vs IT
  - 2 RCT's
  - Battaglia et al, 2008 –combined beneficial (87.5% vs 70.5%)
  - Lim et al, 2012 – no difference

Intratympanic (IT) steroids



Summary

- Determine if CHL or SNHL
- Evaluate with urgent audiogram
- Offer steroids
  - IT or high dose PO 1mg/kg/day up to 70mg/day for minimum 7 days
- If audiogram unavailable and SNHL – treat and refer
- Refer to Otolaryngology for HBOT +/- IT steroids

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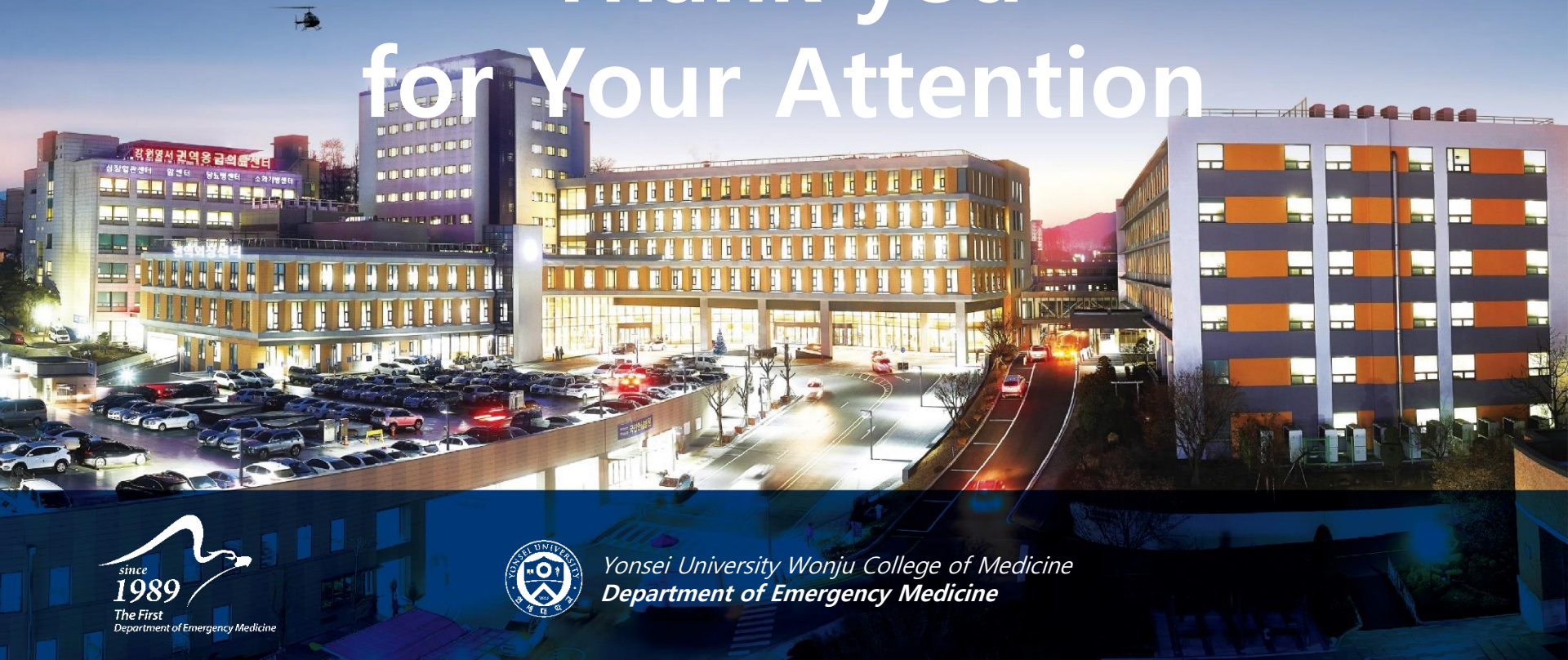
Thank you for Your Attention



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Discussion point!

1. 폐쇄공포증, 이과적, 내과적 과거력상 합병증 발생의 위험성이 있는 환자에서 어느 선까지 HBOT를 강조하여 치료 해야할지?
2. 현재 고압산소치료의 적응 기준 80dB이 적절한가?
3. 현재 고압산소치료 회수 14회 적절한가?
4. Primary treatment로서의 HBOT와 salvage treatment 목적으로서의 HBOT 효과가 있을까?
5. 초기에 HBOT를 받지 못한 경우 늦어도 언제부터는 HBOT 시작해야 할까? / 언제 까지는 받는게 의미 있을까?