



ORL

KLINIKA OTORINOLARYNGOLÓGIE  
A CHIRURGIE HLAVY A KRKU  
LFUK A UNB

# Sudden Sensorineural Hearing Loss State-of-the-art

***Milan Profant***

*Dept ORL HNS*

*Medical School, Comenius University, Bratislava, Slovakia*

*IFOS*

*Otology, Audiology course, Lima, Nov. 11-13, 2018*

# Sudden Sensorineural Hearing Loss SSNHL

- Incidence: 5-20/100.000
- Definition („**3x3**“):
  - Sudden onset (in 72h) – **3 days**
  - Hearing threshold **30dB** and more
  - At least **3** neighboring frequencies
- If there is no audiogram before the SSNHL onset the other side threshold is accepted as referential threshold
- In everyday clinical practice we accept for treatment also SSNHL that does not reach 30dB in 3 frequencies



# SSNHL – additional symptoms

- In 2/3 of patients tinnitus, in 1/3 vertigo
- Unilateral >95 %,
- Bilateral <5 %
- Bilateral is always more serious condition!
  - More severe HL, less chance for recovery, higher morbidity (mortality?)
  - Vestibular symptoms less frequent
  - Associated with toxic, genetic, autoimmune, neoplastic and vascular ethio-pathology
  - Requires complex diagnostic approach

Category	Condition or cause
Toxic	Alcohol
	Cocaine, heroin, ecstasy
	Opioid
	Benzodiazepine
	Pegylated interferon
	Antiviral agents
	Alkalisising agents
	Synthetic prostacyclin PGI <sub>2</sub> analogue
	Retinoid
	Chemotherapeutic agents
	NSAIDs
	Immunosuppressive drugs
	Bisphosphonates
	Skeletal muscle relaxants
	Insecticides
	Gentamycin
Neoplastic	CPA or petrous meningioma
	CPA or petrous apex metastasis
	Neurolymphomatosis
	Leptomeningeal carcinoma
	MDS-associated hypercoagulability
	Vestibular schwannoma
	Acoustic neurofibroma
Vascular	Meningeal carcinoma
	Cerebrovascular accident
	Migraine-associated vasospasm
	Sickle cell disease
Autoimmune	Autoimmune inner ear disease
	Cogan's disease
	Kawasaki disease
	Guillain-Barré syndrome
	Scleroderma
	Anti-phospholipid syndrome
	Crohn's disease
Infectious	Polychondritis
	Mumps
	HIV
	HSV
	Cryptococcal meningitis
	Bacterial meningitis
Iatrogenic	Viral URTI
	Micro-embolic surgical complications
	GA haemodynamic complication
	GA ototoxicity

SNHL = sensorineural hearing loss; PGI<sub>2</sub> = prostaglandin I<sub>2</sub>; NSAIDs = nonsteroidal anti-inflammatory drugs; CPA = cerebello-pontine angle; MDS = myelodysplastic syndrome; HIV = human immunodeficiency virus; HSV = herpes simplex virus; URTI = upper respiratory tract infection; GA = general anaesthetic

Age	Likely aetiology	Assessment tasks
Younger*	Infective	Sx on presentation Ix: WCC, CRP, viral serology
	Toxic	Sx on presentation Hx: exposure to drugs shown in Table I
	Autoimmune	Sx on presentation FMHx: autoimmune conditions Ix: autoimmune blood panel
Older <sup>†</sup>	Vascular	Sx on presentation Hx: cardiovascular risk factors Ix: CT or MRI
	Neoplastic	Constitutional Sx Ix: WCC, MRI
	Iatrogenic	Hx of recent surgery Hx of exposure to anaesthetic agents

\*30–50 years; <sup>†</sup>>50 years. SNHL = sensorineural hearing loss; Sx = symptoms, Ix = investigations; WCC = white cell count; CRP = C-reactive protein; Hx = history; FMHx = family history; CT = computed tomography; MRI = magnetic resonance imaging

Sara *et al.* 2014, J Laryngol Otol.

# SSNHL Etiopathogenesis



- In 90 % of cases unclear (ISSNHL),
- Exceptions:
  - Acutraumata
  - PCA tumor
  - Labyrinthitis from concurrent otitis
- Hypothesis:
  - Vascular
  - Infectious (parainfectious)
  - Autoimmune
  - Mechanical inner ear damage (membrane rupture)



# Vascular hypothesis



- Pros

- *A. labyrinthi* – terminal artery
- Coincidence with cardiovascular diseases and risk factors
- Clinical development similar to heart attack or stroke

- Cons

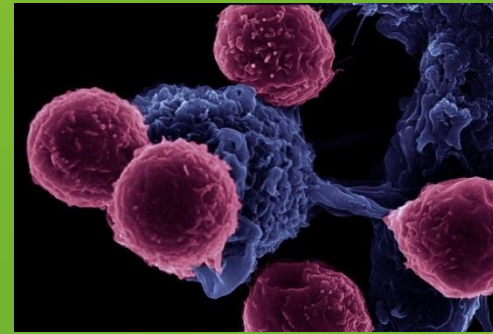
- Young healthy generation involved frequently
- Absence of vestibular symptomatology
- Questionable effect of vasoactive treatment
- Missing histopathologic proof
- Frequent reversibility

# Viral hypothesis



- Pros
  - Common cold prior to SSNHL in many patients (but common cold is very frequent condition)
- Cons
  - Missing the proof of virus to complete Robert Koch postulates
  - Majority of patients suffer from unilateral SSNHL
  - Vaccination did not reduce incidence of ISSNHL

# Autoimmunne hypothesis



- Pros
  - Rapidly developing condition
  - Respond to corticosteroid treatment
- Cons
  - Unknown specific antigenes or lab markers



# Hypothesis of inner ear membranes rupture

- Pros:

- Clinical development
- Unilaterality



- Cons:

- Problematic proof (beta-2 transferín, kochlín-tomoproteín),
- Absence of additional ear pathology
- Missing pressure (RW membrane 2 atm, Ear drum 0,39 atm) Kringlebotn 2000, Ann Otol Rhinol Laryngol

# Diagnosis

- Pure Tone Audiometry
- HRCT (brain, TB, PCA)?,
- MRI PCU?
- USG of carotid arteries?
- RTG C-spine?
- Lab tests?
- Serology?
- ???



# Treatment



- Corticosteroids?
- Virostatics?
- Vasoactive drugs (what kind?),
- Antiagregance/  
thrombolytics?
- Vitamíny?
- Minerals?
- Nutrition supplements?
- Local injection?
- Infusions?
- Hyperbaric  
oxygenation chamber  
(HBOC)?
- Surgery?
- Laser? Magnet?
- Acupuncture?
- ???

# Dept ORL HNS Bratislava (coverage of territory 500.000)

- **50-70 patients/y with SSNHL**
- **Previous management:**
  - No guidelines
  - Different approach of different colleagues
  - Patients always in-ward treatment, infusions of vasoactive drugs and corticosteroids for 7-14days
  - In case of treatment failure long term Cavinton, Agapurin, Ginkgo biloba,...

## Inward patients

**HCT 500-400-300-200-100 mg i.v., Inf. FR + 1A 10 % MgSO4 + 1A Cavinton + 1A Oxantil + 1A Mesocain i.v., 1A Agapurin i.v., Quamatel 2x1**

MD

## Outward patients

- |         |  |
|---------|--|
| 1       | Prednison 20mg: <b>3dni 3x2, následne 2-2-1 až po 0,5-0-0, ex, Quamatel 2x1, Cavinton 3x1, p.p. Betaserc 24 3x1</b>  |
| 2       | Prednison 20 mg : <b>5 dní 3x2, 5 dni 3x1, 5 dni 2x1, 5 dni 1x1, 5 dni 1/2 tbl , Cavinton 3x2 alebo Betaserc 3x2 tbl</b>   |
| 3       | Prednison 20mg <b>2-2-1, 2-2-0, 2-1-0, 1-1-0, 1-0-0 dobrať</b> (+gastroprotekt.), with Cavinton/Trental/Betahistín or their combination  |
| 4       | After discharge Agapurin + Cavinton, follow up after 1 month, if no improvement no additional treatment<br>No treatment if infusion therapy improved hearing                                 |
| 5       | „99%“ inward.; „1%“ oral <b>vasoactibe, Betahistin, Vinpocetin, EGB</b>  |
| 6       | Infusions, hospitalization   |
| 7,<br>8 | Prednison 2-2-2, 2-2-1, 2-2-0, 2-1-0, 2-0-0 3d, 1-0-0 3d, 1/2-0-0 3d, Quamatel 2x1, Cavinton 3x1, Tanakan 3x1, susp. Menier + Betaserc 24 mg 2x1   |
| 9       | 5 days Prednison 20 mg tbl. /+ Helicid /: <b>2-2-2, 2-2-1,5, 2-2-1, 2-2-0,5, 2-2-0.</b> Followed by audiometry, continues Cavinton + Agapurin, resp. Cavinton + Tanakan 2-3 mes., audiometry |

# Development of recommendations

290 Leitlinie

## Die aktuell gefasste Leitlinie „Hörsturz“ (Akuter idiopathischer sensorineuraler Hörverlust)\*

The Revised Version of the German Guidelines “Sudden Idiopathic  
Sensorineural Hearing Loss”

Autor

O. Michel<sup>1, \*\*</sup>

Institute

<sup>1</sup> Universitair Ziekenhuis Brussel, Vrije Universiteit Brussel UZB-VUB, B-1090 Brüssel

<sup>\*\*</sup> für die Mitglieder der Konsensuskonferenz: Prof. Dr. W. Arnold, München; Prof. Dr. F. Bootz, Bonn; Prof. Dr. T. Brusic, Köln; Prof. Dr. G. Hesse, Bad Arolsen; Prof. Dr. E. Klemm, Dresden; Prof. Dr. O. Michel, Brüssel; Prof. Dr. R. Mösges, Köln; Prof. Dr. S. Plontke, Halle/S.; Prof. Dr. M. Suckfüll, München (Vorsitz)

Michel *et al.* 2011, Laryngorhinootologie

**Corticosteroids** (Prednisolon 250 mg in 3 days)  
**+ rheologic treatment**

# Stachler et al. 2012: Clinical practice guideline: Sudden hearing loss.

Otolaryngology-Head and Neck Surgery, 146 (Suppl. 1): 1-35.

- Guideline for adults (18+y)
- Based on evidence (EBM)
- Criteria of NIDCD (National Institute on Deafness and other Communication Disorders)
- **13 recommendations**



Guideline

**Clinical Practice Guideline:  
Sudden Hearing Loss**

Robert J. Stachler, MD<sup>1</sup>, Sujana S. Chandrasekhar, MD<sup>2</sup>, Sanford M. Archer, MD<sup>3</sup>, Richard M. Rosenfeld, MD, MPH<sup>4</sup>, Seth R. Schwartz, MD, MPH<sup>5</sup>, David M. Barrs, MD<sup>6</sup>, Steven R. Brown, MD<sup>7</sup>, Terry D. Fife, MD, FAAN<sup>8</sup>, Peg Ford<sup>9</sup>, Theodore G. Ganiats, MD<sup>10</sup>, Deena B. Hollingsworth, RN, MSN, FNP<sup>11</sup>, Christopher A. Lewandowski, MD<sup>12</sup>, Joseph J. Montano, EdD<sup>13</sup>, James E. Saunders, MD<sup>14</sup>, Debara L. Tucci, MD, MS<sup>15</sup>, Michael Valente, PhD<sup>16</sup>, Barbara E. Warren, PsyD, MEd<sup>17</sup>, Kathleen L. Yaremchuk, MD, MSA<sup>18</sup>, and Peter J. Robertson, MPA<sup>19</sup>

FOUNDATION  
Otolaryngology—  
Head and Neck Surgery  
146(1S) S1–S35  
© American Academy of  
Otolaryngology—Head and Neck  
Surgery Foundation 2012  
Reprints and permission:  
sagepub.com/journalsPermissions.nav  
DOI: 10.1177/014599812436449  
http://otojournal.org  
SAGE

# Level of recommendations in relation to EBM

Code	Quality of Evidence	Definition
A	High	<ul style="list-style-type: none"><li>•Clinicians should follow recommendations</li><li>•<b>Strong recommendation (SR)</b></li></ul>
B	Moderate	<ul style="list-style-type: none"><li>•Clinicians should follow recommendations</li><li>•<b>Recommendation (R)</b></li></ul>
C	Low	<ul style="list-style-type: none"><li>•One or more studies with severe limitations</li><li>•Clinicians should be flexible in decisions, requirements of patients are important</li><li>•<b>Option (Op)</b></li></ul>
D	Very Low	<p>Any estimate of effect is very uncertain.</p> <ul style="list-style-type: none"><li>•Expert opinion</li><li>•No direct research evidence</li><li>•<b>No recommendation (NoR)</b></li></ul>



# 1. Exclusion of conductive hearing loss in SHL

## 1. Strong recommendation (SR)

- **The most frequent reason for HL**

- Cerumen

- Tubal dysfunction in common cold

- **History**

- **Otoscopy**

- **Tuning forks**

- **Audiometry**

- **Tympanometry**



## **2. Special attention to:**

Bilateral SSNHL

Recurrent SSNHL

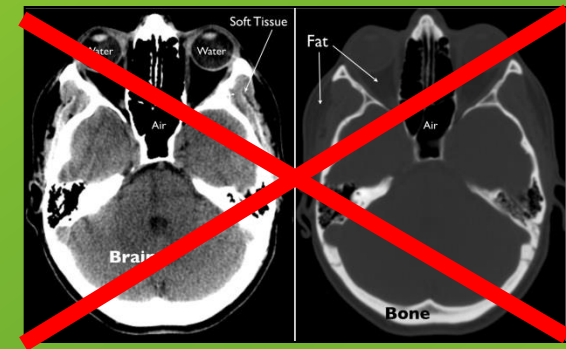
Focal neurologic symptoms

## **Recommendation (R)**

- Significant possibility of other reasons for SSNHL
  - Autoimmune
  - Metabolic
  - M. Ménière
  - Stroke
- **Detailed history**
- **General medical investigation**
- **Neurologic investigation**

# 3. CT imaging

- Strong recommendation (SR) against !



- CT has no sense in initial diagnosis of SSNHL  
(low information, irradiation, price)
- **Exceptions** – clinical suspicion for stroke, head trauma, otitis

## 4. Audiometry to confirm ISSNHL (R)

- HL  $\geq$  30 dB na 3 neighboring frequencies
- Onset in 72h
- Unknown reason for HL according to history and investigations
  
- If audiometry is not available diagnosis is based on tuning fork tests
- Speech audiometry is also recommended

# 5. Lab tests

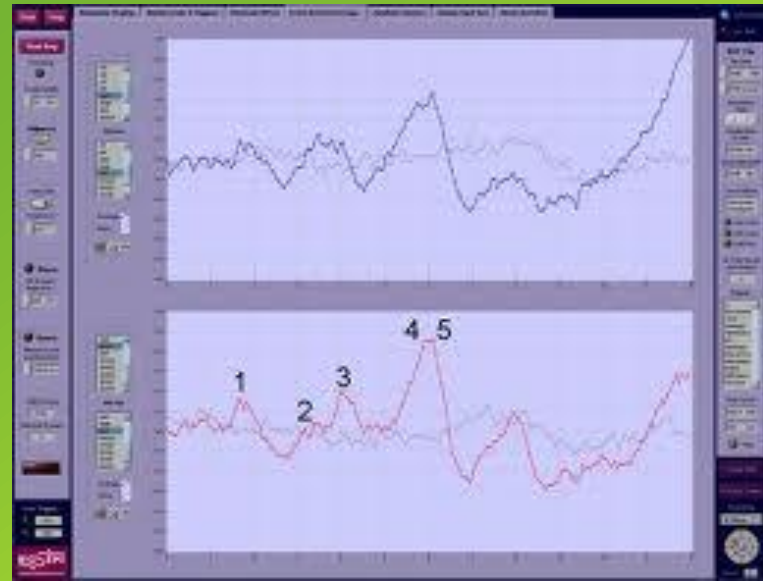
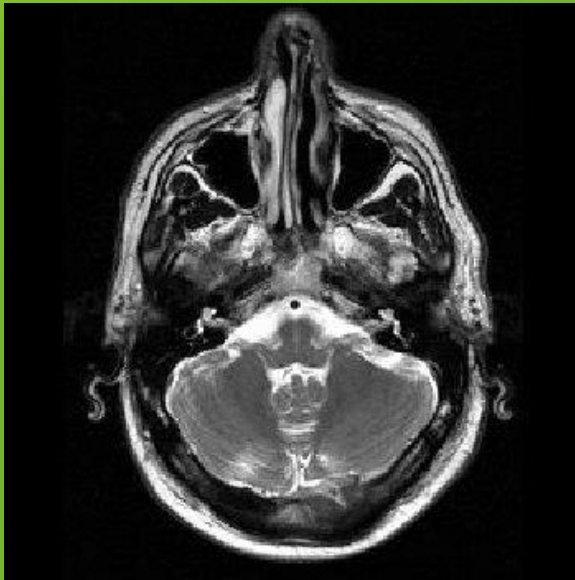
- **SR – against!**



- No recommendations for routine lab tests in patients with SSNHL
- Reasons:
  - False positivity/negativity
  - Low value
  - No influence on treatment outcome
  - Price

## 6. Exclusion of retrocochlear pathology (SR)

- 1. **MRI** (gold standard) - every patient with idiopathic SSNHL should have MRI !!!
- 2. **BERA**
- **Pathologic finding in MRI in SSNHL is 7-14 % -**  
*The most effective method to identify etiopathology in SSNHL*



# 7. Patient education (SR)

- Type of diagnosis
- Benefits and risks of medical interventions
- Limits of medical proofs and their effectivity



# 8. Initial Corticosteroid treatment (Op)

- Oral CS:

- Suppression of hypothalamo-pituitar-adrenal axis
- Risk of Cushing like sy
- Treatment duration 10-14 days
- Low price

- Intratympanic CS:

- Minimal general effect
- Local reaction
  - Pain
  - Ear drum perforation
  - Dizziness
- High price
- Frequent visits



- **Oral corticosteroids:**

Best chance to improve hearing during first 2 weeks,  
low chance after 4-6 weeks

Doses:

- Maximal effect in **Prednisone 1mg/kg/d**,
- Usually the daily doses not more than 60mg
- Single shot doses
- 7-14 days of treatment followed by reducing the doses in the same time period
  - 200-300 mg HCT ~ Prednison 60mg ~ Dexametazon 10 mg  
(Prednison is 4x, Metylprednisolon 5x, Dexamethason 25x more effective than HCT)

**Contraindications:** insulin dependent DM (or decomp DM),  
Hypertension, TBC, peptic ulcer, some psychiatric dg

# 9. Hyperbaric Oxygenotherapy (Op)

- To be applied before 3 months of SSNHL
- Statistically the effect of treatment does not differ from CS
- Price?
- Availability?
- Eustachian tube function (barotrauma)
- Repeated sessions (5-10X)



# 10. Other pharmacologic treatment

- R – against!



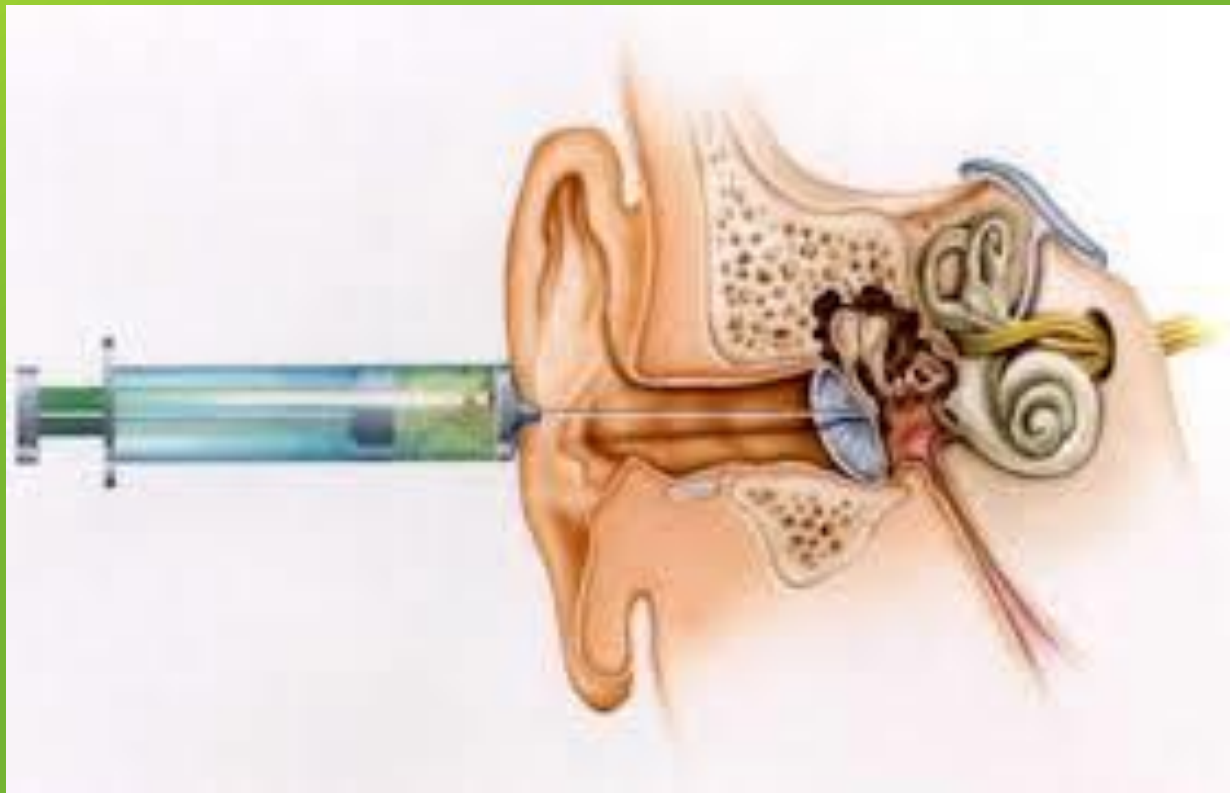
- Antiviral drugs, thrombolytics, vasodilators, vasoactive drugs should not be routinely administered
- Reasons:
  - Nonsignificant results against placebo or corticotherapy,
  - Price,
  - Risk of sideeffect is overweighting benefit (overtreatment)
- Individual needs may indicate such a treatment

# 11. (Salvage therapy) (R) (parallel with primary CS?)

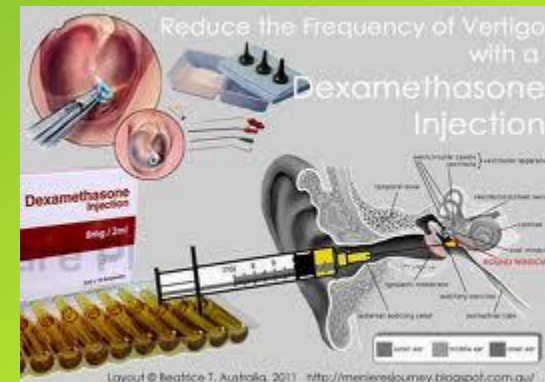


- After unsuccessful primary treatment salvage therapy should be recommended
  - **intratympanic** corticosteroids
- Higher concentration in the target organ can be reached than in general CS administration
- Method:
  - 5 days - 3 months after primary unsuccessful treatment
  - 3-5 sessions during 1-2 weeks or every 2 days

- Hearing improvement can be expected in 50% of patients



- **Intratympanic corticotherapy:**
- **Different schemes: at least 3-4 administrations during 1-2 weeks**
- **Myringotomy or tube**
- **15-30 min. in otologic position**
- **Audiometry before each administration, after treatments and during follow ups**

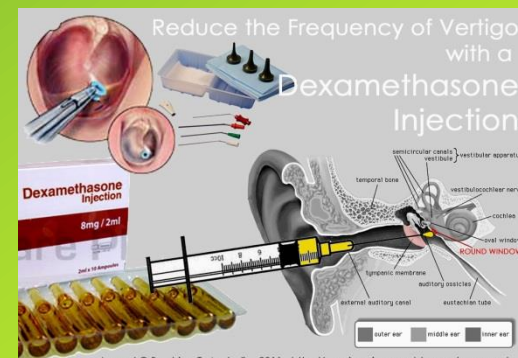


# 12. Evaluation of treatment results (R)

- 97 % pac. have stabilized hearing 3 months after treatment
- No additional improvement can be expected
- Minimal follow up should be 6 mmonths after treatment completion
- Follow up includes PTA a Speech Audiometry

# IT corticotherapy

- Outpatient procedure
- Spinal needle, 2 ml syringe,
- Dexamethason or Methylprednisolon (for i.v. administration)
- Local anesthesia
- IT administration of cca 0,5 ml corticoid through posterior inferior quadrant
- 15-20 min. otologic position, avoid swallowing
- Audiogram before each administration





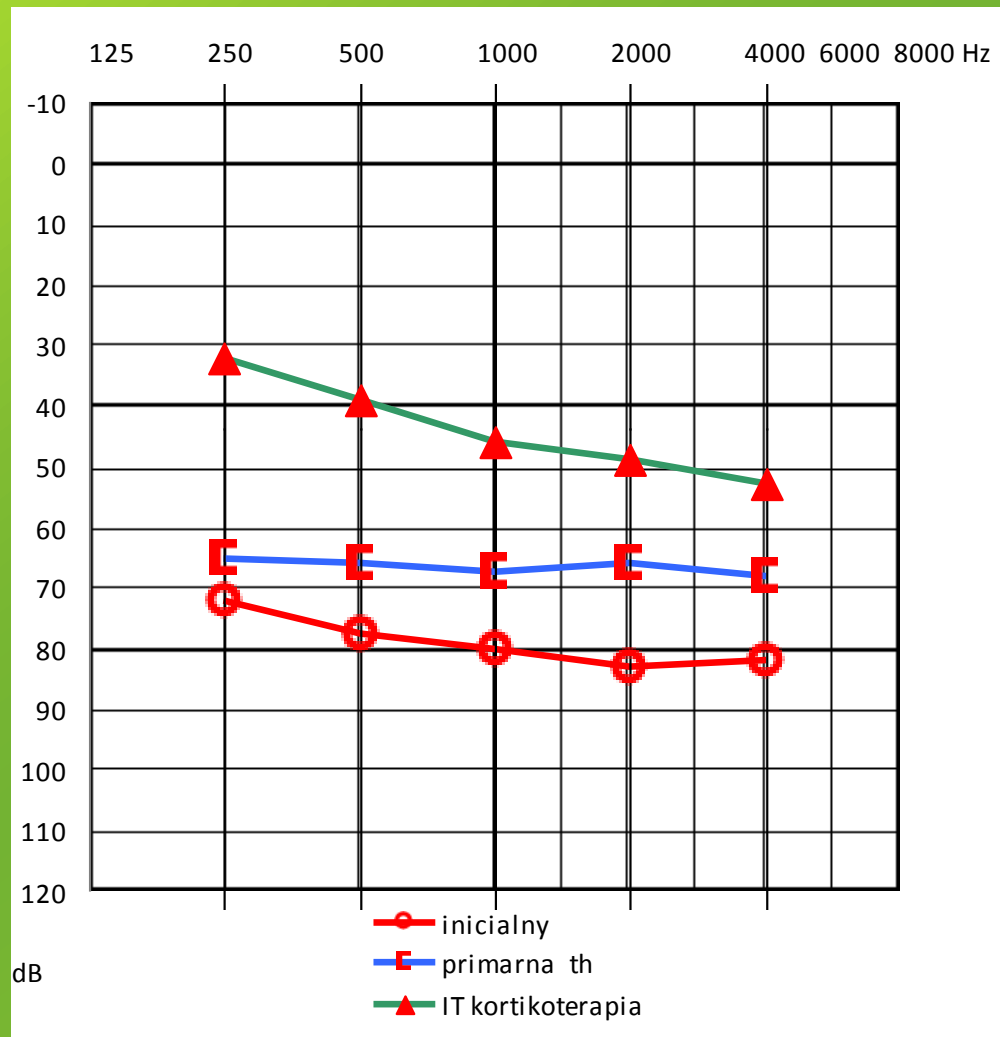
# Complications

- **Caloric vertigo** (2-3 min.) – (solution should have a body temperature)
- **Leak into pharynx** aftertaste in mouth (avoid swallowing)
- **Pain**
- **Permanent perforation (10-15%)**

→ Patient counselling!

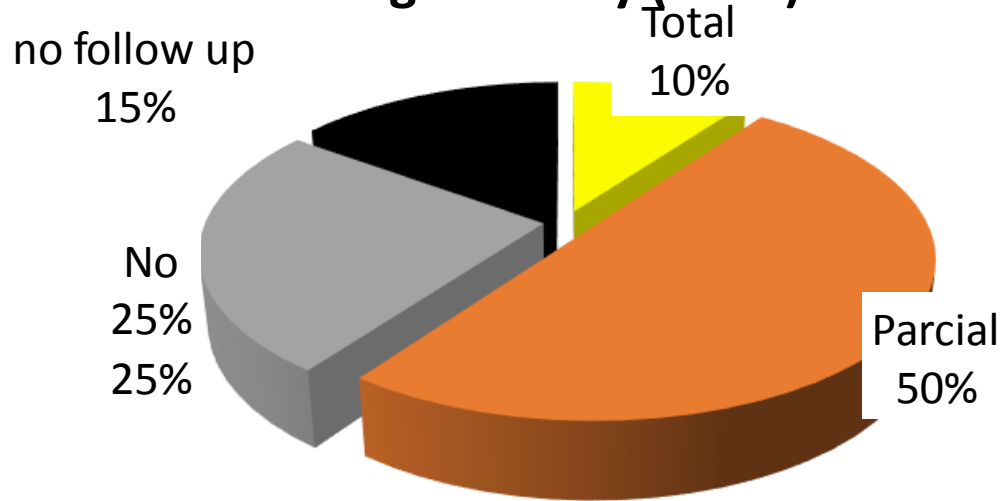


# Audiogram after IT corticotherapy (N=20)

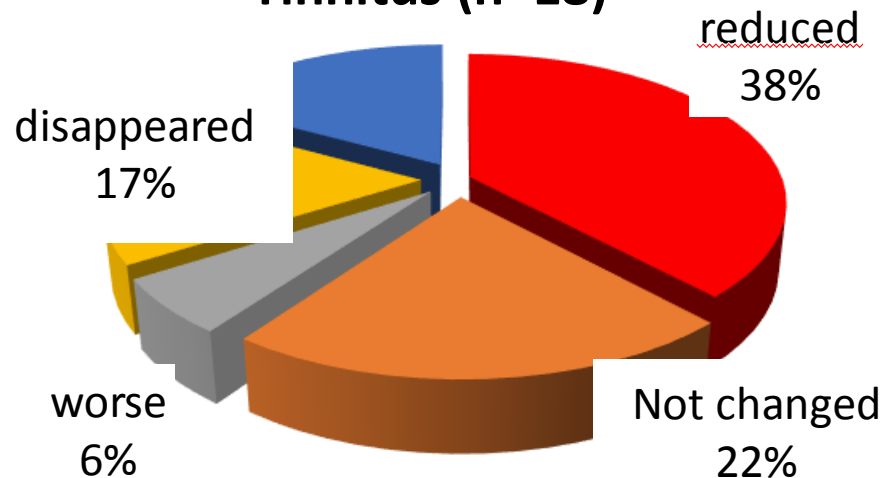


# Salvage IT corticotherapy - results

## Hearing recovery (n=20)

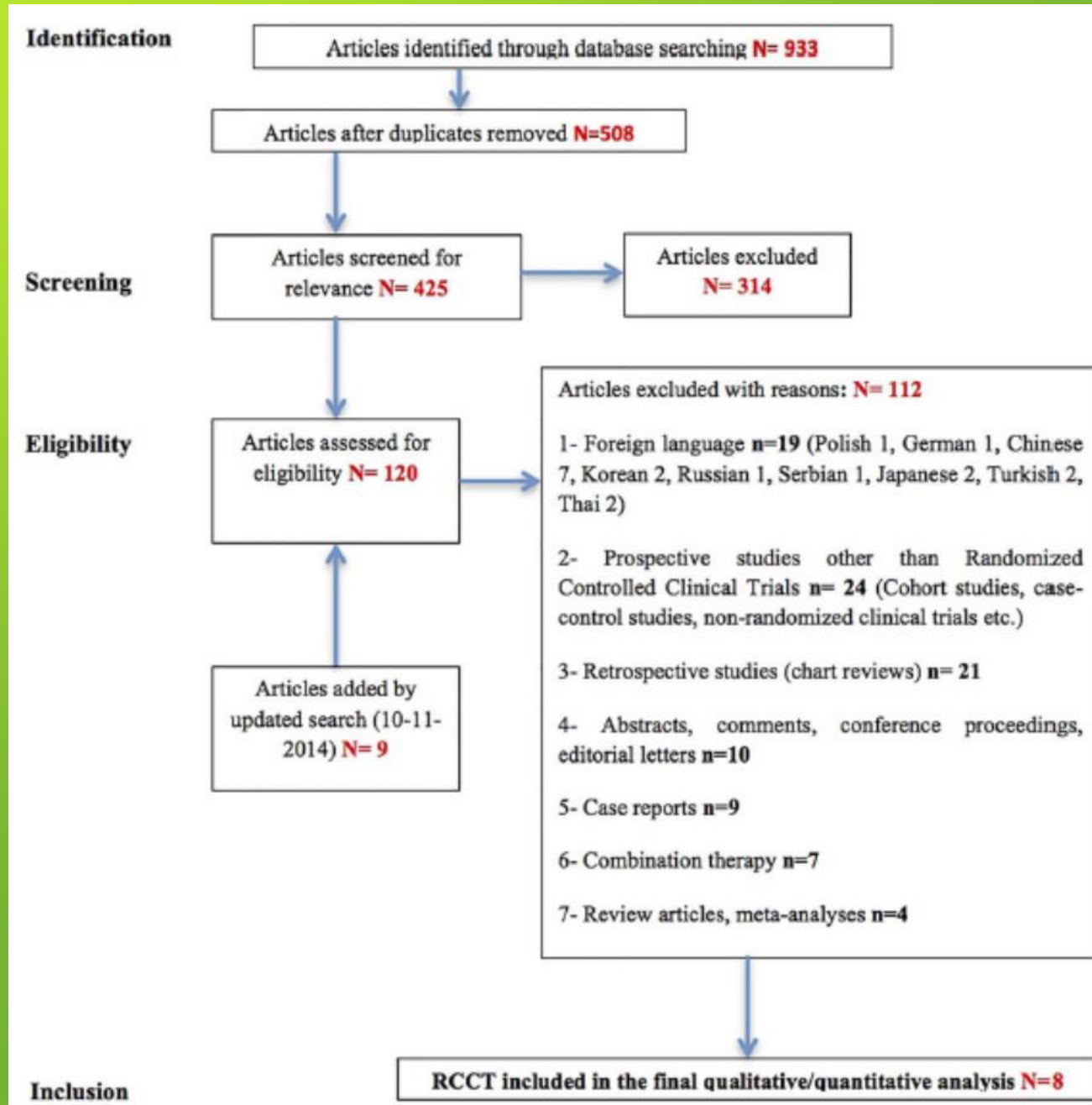


## Tinnitus (n=18)



# Literature 2012-2017

- Too many papers
- Small series
- Non-univocal outcomes
- Metaanalysis – problems with heterogeneity of methodology
- Nowadays simultaneous administration of general and IT treatment



El Sabbagh et al. 2016, Laryngoscope

# Contemporary algorithm

- **Primary th:**
  - Prednison 60 mg 7-14 dní (equivalent HCT 300 mg i.v. inf.),
  - Reducing the dosage during the same period (+gastroprotectives)
- **Salvage therapy:**
  - IT corticotherapy
  - +/- HBOT
- **Simultaneous general and local (IT) corticotherapy**

# New possibilities



How long should patients stay in position  
after intratympanic steroid injection?

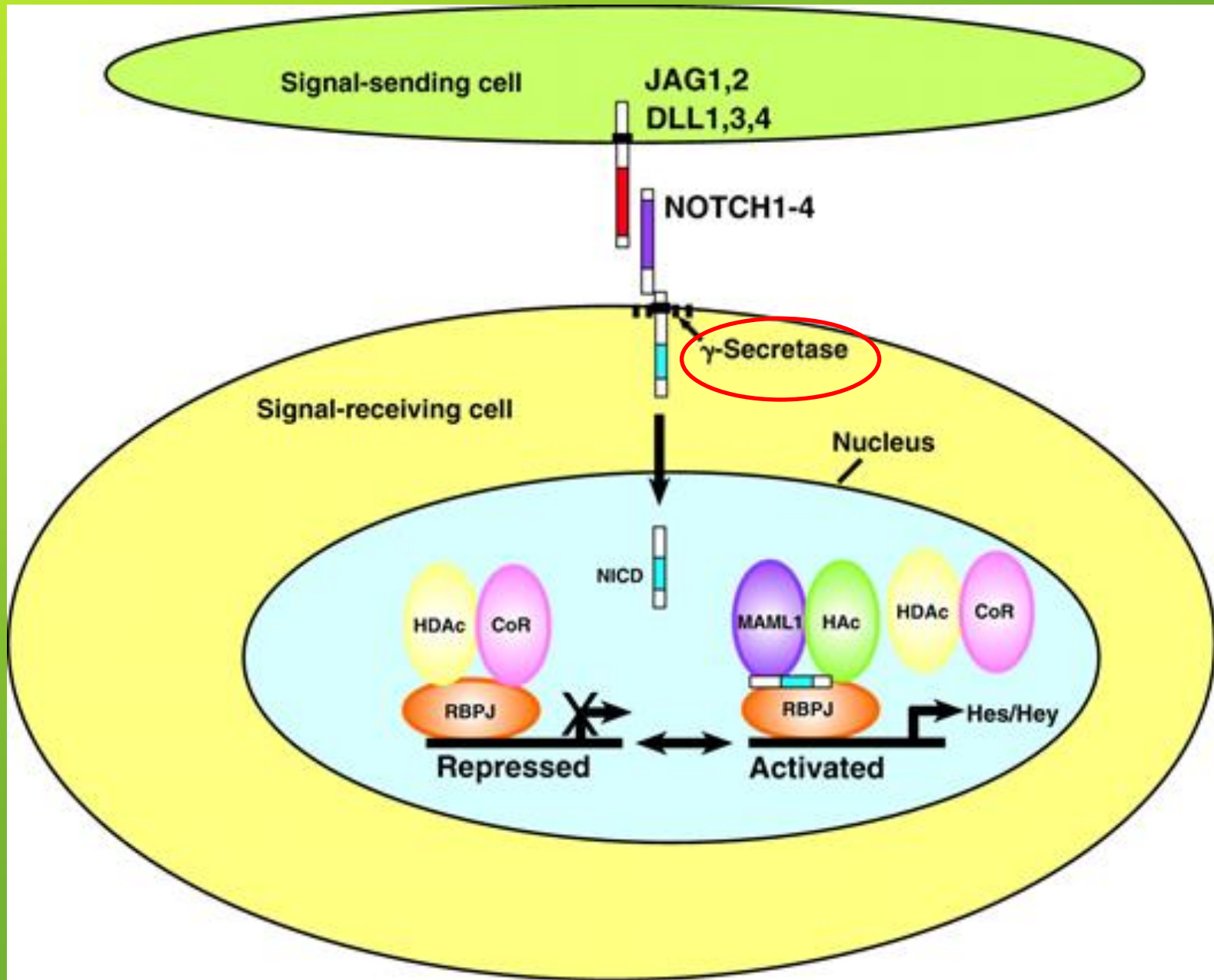
Soon Hyung Park, Jin Young Seo, In Seok Moon  
Keimyung University School of Medicine  
Yonsei University School of Medicine



# New possibilities

- Inhibitor of gamma secretase
  - Influencing of Notch signal pathway that in mammals switches of possibility of hair cells regeneration (Phase II study)
- IGF-1 – influencing supporting cells of organ of Corti to inhibit hair cells apoptosis (Yamahara *et al.* 2015, Hear Res).





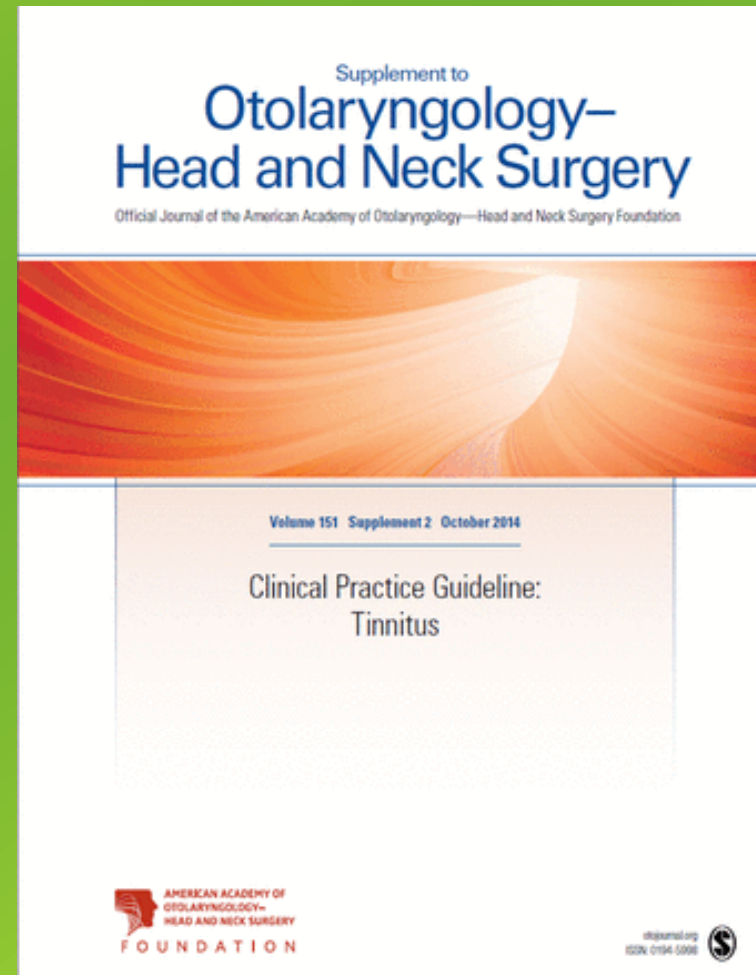
# Tinnitus



- Tinnitus: The perception of sound when there is no external source of the sound
- Primary tinnitus: Tinnitus that is idiopathic and may or may not be associated with SNHL
- Secondary tinnitus: Tinnitus that is associated with specific underlying cause (other than SNHL) or an identifiable condition
- Acute tinnitus less than 6 months
- Chronic tinnitus more than 6 months

**Tunkel et al. Clinical Practice Guideline:  
Tinnitus.** *Otolaryngology - Head  
and Neck Surgery* October 2014

- To deal mmostly with chronic tinnitus
- **Acute tinnitus associated with SSNHL  
to be managed the same way as SSNHL**
- *G. biloba medication or any other medical  
treatment is not recommended*



# Take home message in SSNHL

- Diagnosis:
  - Typical history
  - Otoscopy normal
  - Tuning fork tests, tympanometry, audiometry
  - MRI PCU
- Treatment
  - As soon as possible oral and/or intratympanic corticosteroids in sufficient dosis on outpatient basis
- Other drugs and HBOT unclear effect, not recommended
- Delayed treatment (after 3 months) non-legitimate
- Audioprosthetic rehab when treatment fails

**Thank you**