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Breaking the Silence: Exploring the Link to Autoimmune Disorders and Sudden Hearing Loss

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- No Disclosures

Outline

- Sudden Sensorineural Hearing Loss (SSNHL)
- Autoimmune Inner Ear Disease (AIED)
- Systemic Autoimmune Diseases (SAID) that are associated with higher SSNHL risk when compared to general population
- Work up
- Management/Treatment

Learning Objectives

- Distinguish between Sudden SNHL (SSNHL) and Autoimmune Inner Ear Disease (AIED) presentations

- Recognize Systemic Autoimmune Diseases (SAID) associated with SSNHL presentation in AIED

- SSNHL diagnosis requires prompt Otolaryngology referral

Sudden Sensorineural Hearing Loss

- Sudden SNHL was first described by De Kleyn et al in 1944

- ***Sudden*** Sensorineural Hearing Loss (SSNHL) definition: **sensorineural hearing decline ≥ 30 dB or more across at least three contiguous audiometric frequencies within a 72- hour period**

- SSNHL annual incidence: 5 - 30 cases per 100,000 individuals

- 30% of SSNHL Etiologies: vascular injury, membrane rupture, viral or bacterial infections, and immune-mediated mechanisms

Sudden Sensorineural Hearing Loss

- Unilateral ear but can be bilateral ears (30%)
- Exact cause is still unknown (Idiopathic)
  - Genetic and environmental contributing factors
- Histological studies: expansion of the endolymphatic compartment within the inner ear
  - Endolymphatic Hydrops may cause Meniere's Disease

Inner Ear and Immune System

- In 1958, German ENT Ernst Lehnhardt was the first to suggest the possibility of sudden or rapidly progressive hearing loss to be the result of an autoimmune process against the inner ear.
- In 1979, American ENT Brian McCabe reported cases of **sudden** progressive SNHL loss ***successfully treated*** by immune suppressive therapy and introduced the clinical entity: **autoimmune SN hearing loss**
- Circulating antibodies against several cochlear antigens in patients with idiopathic, progressive bilateral SNHL (Harris and Sharp, 1990)
  - Heat shock protein-70
  - Anti-HSP70 antibodies have been reported to occur in 10.5-19% of patients with SSHL

Inner Ear and Immune System

- Immune-mediated SNHL usually manifests as a rapidly progressive sensorineural loss, bilateral and asymmetrical
  - often associated with tinnitus and vestibular symptoms
- Important feature: **fast progression** leads to **severe bilateral SNHL** in a few days or weeks
- Rare cases: **unilateral sudden** SNHL and manifests only in the contralateral ear after a variable number of days up to years
- **Sudden** SNHL with ***recurrences***
- *Clinical diagnosis*

Inner Ear and Immune System

- Sudden SNHL – an isolated clinical disorder or presenting symptom of Systemic Autoimmune Diseases (SAID)

- Patients with immune-mediated disease had **increased 4.27 fold risk** of Sudden SNHL than in control group (Rossini BAA et al. 2017)

AIED Diagnostic Challenge

- **Rare** condition immune-mediated damage to inner ear
  - Rapidly progressive SNHL (high frequency) +/- vestibular symptoms: vertigo and tinnitus
  - Estimated yearly incidence: < 5 cases/100,000 people
  - < 1% of all hearing loss cases
  - Prevalence rate: 15/100,000
  - True prevalence may be underestimated

- Immune-mediated damage to inner ear
  - Lack of specific diagnostic tests
    - No specific markers
    - Frequency of antibodies (**anti-68-kDa autoantibody**) against HSP-70 did not differ between AIED patients and controls
    - Access to inner ear is limited

AIED Diagnostic Challenge

- Lack of definitive diagnosis
  - Diagnosis based on clinical presentation
  - Diagnosis of exclusion
- Result in treatment of patients not affected with the disease, resulting in no response
- Lack of standardized criteria
  - Significant variability/heterogeneity in Otolaryngology practices

Autoimmune Inner Ear Disease (AIED)

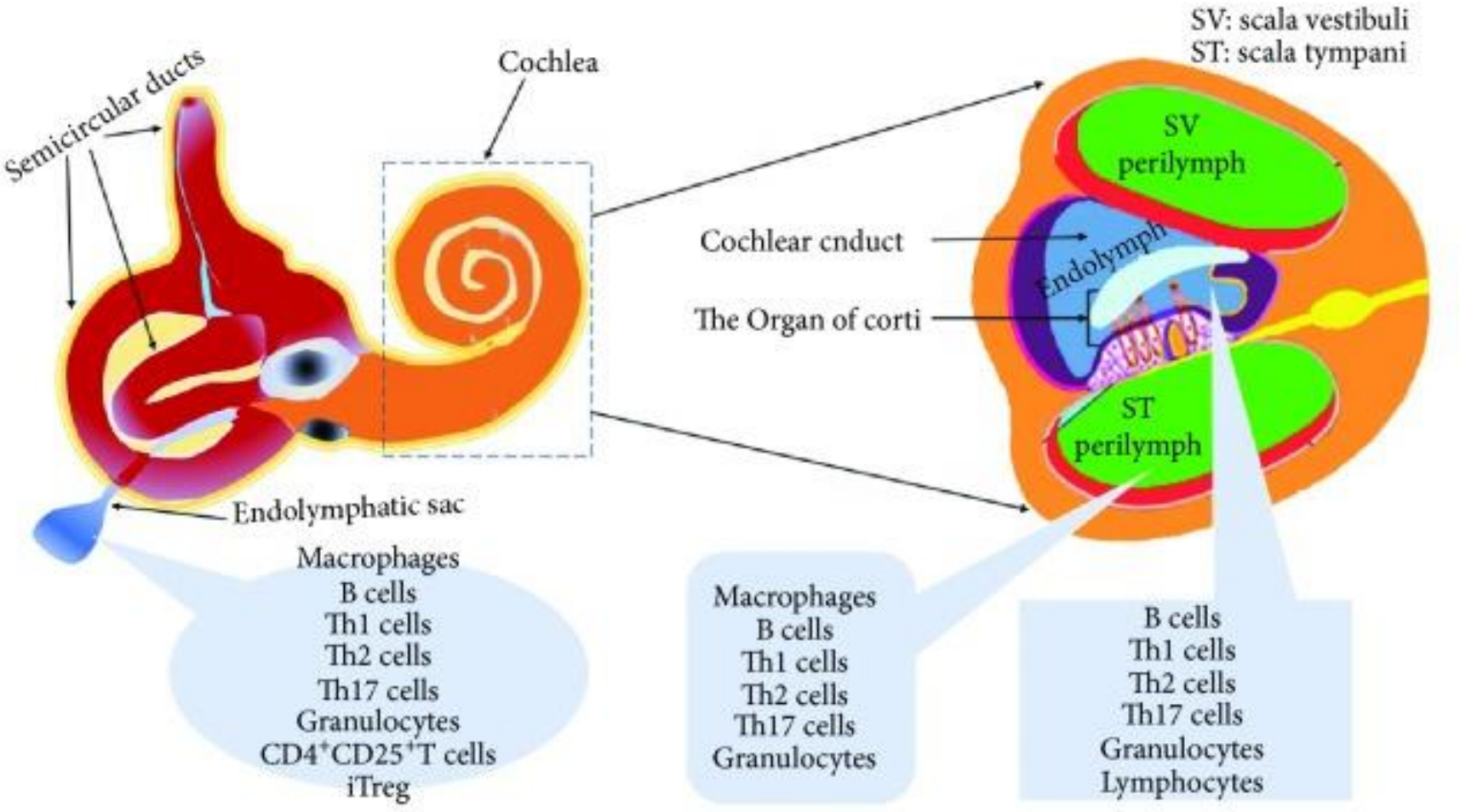
Two categories:

1) **Primary**:
   - Confined to the inner ear

2) **Secondary (15-30%)**:
   - Associated with systemic autoimmune disease
     - Systemic Lupus Erythematosus (SLE)
     - Antiphospholipid syndrome (APS)
     - Rheumatoid arthritis (RA)
     - Wegner's granulomatosis
     - Sjogren syndrome (SS)
     - Behcet's syndrome
     - Sarcoidosis
     - Cogan's syndrome

AIED symptoms

- Variable presentations
  - Sudden, rapid progressive and fluctuating hearing loss
- **Bilateral** ear involvement
- Accompanying symptoms: tinnitus, aural fullness, vestibular symptoms
- Immune mediated hearing loss differential dx:
  - Meniere's disease, sudden SNHL, AIED
  - Perilymphatic fistula (unilateral)



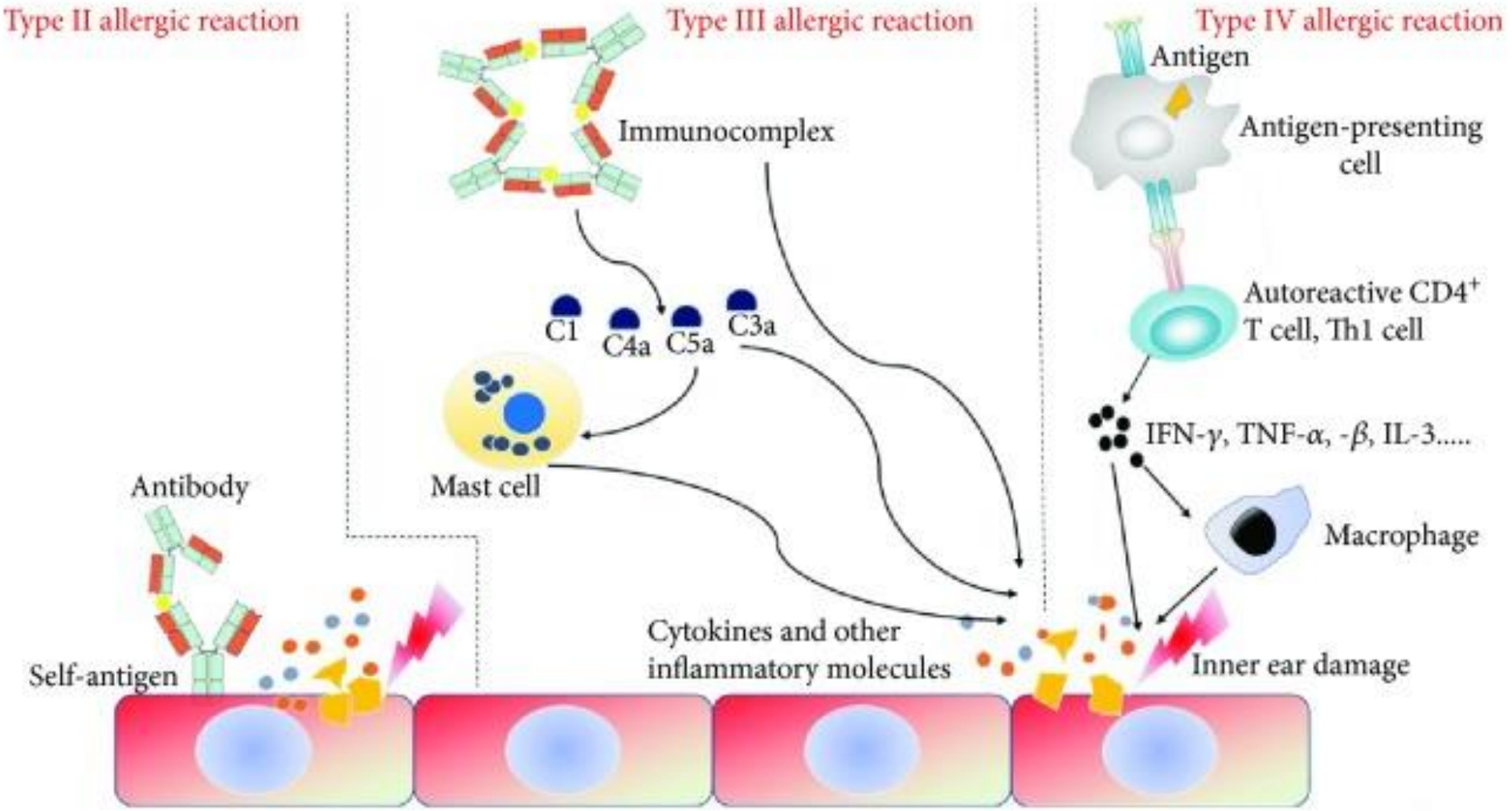
Distribution of immune cells in the inner ear when the immune response is initiated. **Not an immune-privileged site**.

Li G et al. Neural Plast. 2018

Immune Pathogenesis of Hearing Loss

- **Inner ear's immune system (Type II)**
  - **endolymphatic sac** generating and sustaining **an immune response** against **self-antigens** following the initial injury or inflammatory insult (Tomiyama S, Harris JP. Laryngoscope 1986)
  - Examples: Anti-cochlin IgG and Anti-HSP70 IgG
  - **Antibod**y-mediated tissue damage in the inner ear
- **Molecular mimicry**
  - **Antibodies** developed against pathogens with antigenic similarity to the inner ear proteins cross-react with host tissues (Cusick A et al. Int J Immunopathol Pharmacol 2018)
- **Bystander effect**
  - cytokines (IL-1) and tumor necrosis factor (TNF) released from adjacent cells promote local immune activation, immune tolerance breakdown, wherein concealed antigens become accessible to the immune system upon tissue damage, provoking an autoimmune response

    (Zomer A et al. J Extracell Vesicles 2014)



The mechanisms of inner ear damage by Type II – IV allergic reactions: Autoantibodies or T-cells.

Diagnostic Challenge

- Bilateral sudden SNHL: 31% prevalence rate + antinuclear antibody (ANA)

- Blood tests:
  - CBC, serum fasting glucose, lipid profile, renal function, thyroid function, erythrocyte sedimentation rate, serology for syphilis

- Specific serological test:
  - rheumatoid factor (RF), ANA, antiphospholipid antibodies, ANCA, lupus anticoagulant, cryoglobulin levels

Diagnostic Challenge

Specific Antibodies that target inner ear antigens:

- Collagen type II and IX
- Cochlin – an inner ear protein for maintaining vestibule homeostais
- Supporting Cell protein (DEP-1/CD148): Cogan's syndrome
- Anti-68-kD anti-cochlear antibody
- Anti-HSP-70 antibody: cochlear and vestibular cells
  - High sensitivity: 54.5%
  - High specificity: 42.9%

  (Matsukoa et al. Audiol Neurootol.2013)

Limited sensitivity and no efficacy in diagnosing AIED!

Diagnostic Challenge

1994 National AIED Conference:

- Rapidly progressive history
- Fluctuating bilateral or unilateral SNHL
- Vestibular symptoms: dizziness or tinnitus
- Autoimmune serological markers

Differential diagnoses to exclude:

- sudden SNHL
- ototoxicity
- presbycusis
- Meniere's disease

AIED Diagnostic Criteria

- **Major criteria**
  - Bilateral hearing loss
  - Systemic autoimmune disease
  - ANA > 1:80
  - Decreased native T cells
  - Hearing recovery rate > 80%
- **Minor criteria**
  - Unilateral hearing loss
  - Young or middle age
  - Female gender
  - Hearing recovery rate < 80%
- Definitive diagnosis requires meeting 3 major criteria or 2 major and 2 minor criteria

Secondary AIED

- Sudden SNHL – a common audiovestibular symptom in systemic autoinflammatory diseases

Rates:

- SLE: 6 – 70% (Paraschou et. al, 2021)
- Sjogren: 22 – 46% (Ralli et al, 2018)

- **Pathogenesis of inner ear:**
  - SS and RA: vasculitis of vasa nervorum or vasa vasorum
  - SLE: antibody-antigen reactions, cell mediated cytotoxic damage to vestibular organs or immune complex deposition in micro-vessels
  - APS: micro thrombosis in inner ear
  - Psoriatic arthritis (PsA): inflammation affecting vascular damage neurodegeneration of cochlear and vestibular organs

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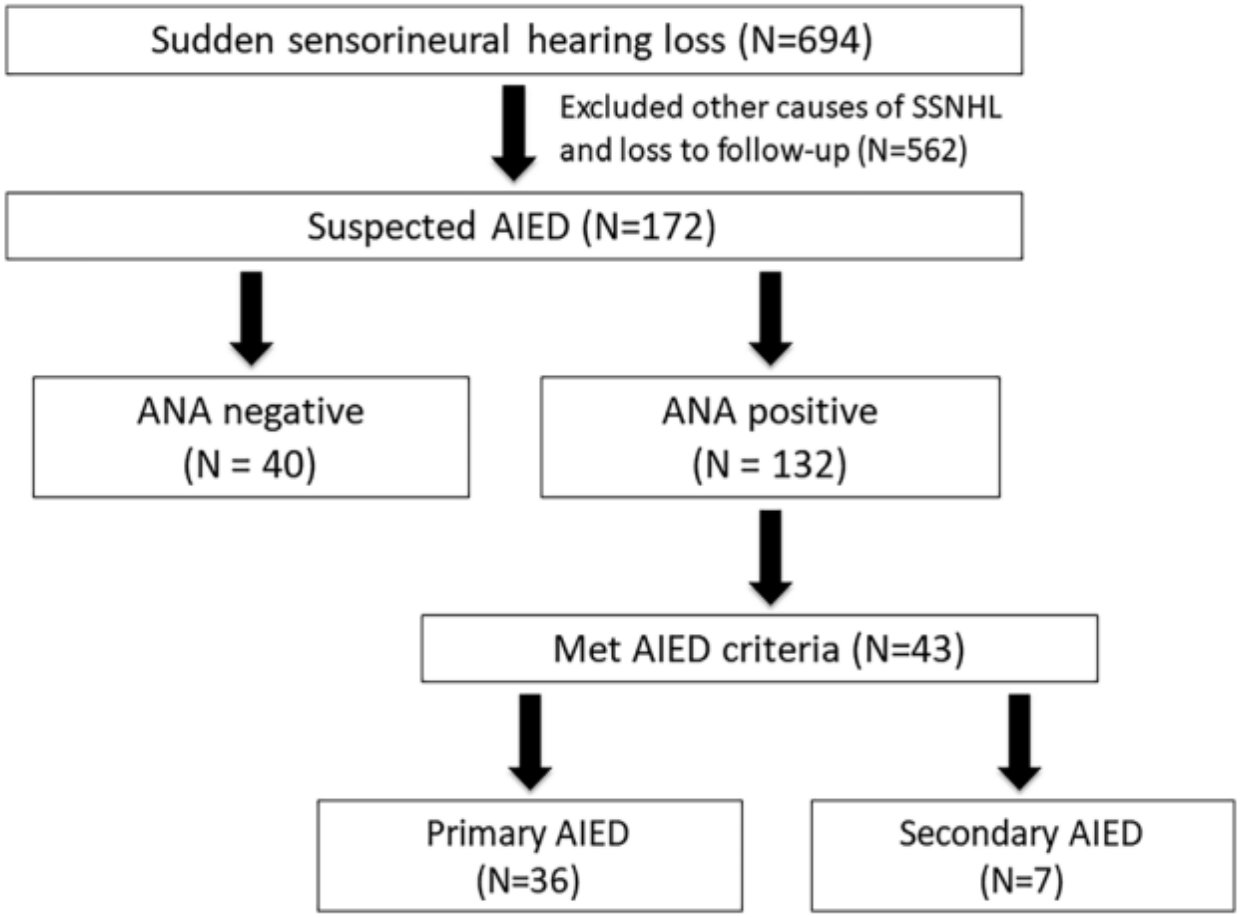
"Prevalence, Clinical profiles, Treatment response of AIED in SSNHL"



Arunthanachaikul T, et al. Otol Neurotol. 2025 Feb 1;46(2):215-220

TABLE 1. *Comparison of clinical characteristics, immune, and treatment features between primary and secondary AIED*

Variables	Overall (N = 43)	PrimaryAIED (N = 36)	SecondaryAIED (N = 7)	p^a
Demographics				
Age, y (SD)	56.9 (11.4)	56.7 (11.6)	57.8 (11.7)	0.81
Female, n (%)	29 (67.4)	24 (67.7)	5 (71.4)	1
Initial hearing level				
Degree of hearing loss, dB (SD)	53.5 (16.1)	54.5 (16.3)	48.3 (14.8)	0.36
Ear affected first, n (%)				
Right	10 (23.3)	9 (25)	1 (14.3)	0.78
Left	13 (30.2)	11 (30.6)	2 (28.6)	
Both	20 (46.5)	16 (44.4)	4 (57.1)	
Associated symptoms, n (%)				
Hearing loss fluctuation	26 (60.5)	21 (58.3)	5 (71.4)	0.68
Tinnitus	11 (25.6)	11 (30.6)	0	0.16
Aural fullness	27 (62.8)	25 (69.4)	2 (28.6)	0.82
ANA titer, n (%)				
1:80	27 (62.8)	22 (61.1)	5 (71.4)	0.7
≥1:160	16 (37.2)	14 (38.9)	2 (28.6)	
ANA pattern, n (%)				
Homogeneous	12 (28)	10 (27.8)	2 (28.6)	1
Speckled	23 (53.5)	18 (50)	5 (71.4)	0.42
Nucleolar	11 (25.6)	10 (27.8)	1 (14.3)	0.66
Cytoplasm	9 (20.9)	8 (22.2)	1 (14.3)	1
Others	10 (23.3)	9 (25)	1 (14.3)	1
Treatment, n (%)				
Steroids only	10 (23.3)	10 (27.8)	0	<0.001
IT steroids only	3 (7)	3 (8.3)	0	
Steroids + IT steroids	15 (34.9)	14 (38.9)	1 (16.7)	
Steroids + IS	8 (18.6)	2 (5.6)	6 (85.1)	
Cochlear implantation	1 (2.3)	1 (2.8)	0	
Outcome				
Hearing gain after treatment,[b] dB (SD)	5.3 (13.5)	4.8 (13.7)	7.7 (13.6)	
Hearing level after treatment, dB (SD)	48.3 (16.1)	50.2 (15.1)	37.5 (18.4)	
Response to therapy,[c] n (%)				0.51
Complete recovery	5 (13.9)	3 (8.3)	2 (28.6)	
Partial improvement	5 (13.9)	4 (11.1)	1 (14.3)	
Slightly improvement	2 (5.5)	2 (5.6)	0	
No improvement	24 (66.7)	20 (55.6)	4 (57.1)	

Summary

- No correlation between ANA titer and degree of hearing loss
- Different treatment approaches:
  - **Primary AIED:** variable, oral steroid only, IT steroid, combination, steroids with immunosuppressants and cochlear implants
  - **Secondary AIED:** steroids with immunosuppressants
- Final hearing levels showed no significant difference between Primary vs. Secondary AIED
- **Secondary AIED** patients demonstrated a trend toward **better treatment outcomes with higher rates of complete or partial improvement** compared to Primary AIED patients
- Secondary AIED: bilateral ears with hearing loss fluctuation

Summary

- Prevalence of AIED and Sudden SNHL: 6.2%

Common clinical features:

- Females @ 40s – 60s
- Sudden rapid progressive and fluctuating hearing loss
- Bilateral ear involvement
- Associated symptoms: hearing loss fluctuation and aural fullness

Patients with Sudden SNHL and SAID (systemic autoimmune diseases):

- More severe initial hearing impairment
- Higher percentage of bilateral, lower response to treatment
- Worse prognosis when compared with patients without this association

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"High risk of Sudden SNHL in several Autoimmune Diseases according to a Population-Based National Sample Cohort Study"

Years: 2006 - 2015

Table 1. Demographic characteristics of the autoimmune-disease and control groups

	Autoimmune-disease group (n = 13,250)	Control group (n = 66,250)	p value
SSNHL			<0.001
No events	13,105 (98.91)	65,766 (99.27)	
Events	145 (1.09)	484 (0.73)	
Age range			1.000
20–29 years	1,452 (10.96)	7,260 (10.96)	
30–39 years	2,315 (17.47)	11,575 (17.47)	
40–49 years	2,849 (21.50)	14,245 (21.50)	
50–59 years	2,715 (20.49)	13,575 (20.49)	
60–69 years	2,161 (16.31)	10,805 (16.31)	
70–79 years	1,468 (11.08)	7,340 (11.08)	
≥80 years	290 (2.19)	1,450 (2.19)	
Sex			1.000
Male	4,419 (33.35)	22,095 (33.35)	
Female	8,831 (66.65)	44,155 (66.65)	
Residence			1.000
Seoul (capital)	2,725 (20.57)	13,625 (20.57)	
Metropolitan	3,288 (24.80)	16,430 (24.80)	
City (small and medium-sized)	5,549 (42.63)	28,245 (42.63)	
County	1,590 (12.00)	7,950 (12.00)	
Income level			1.000
1st (lowermost) quintile	2,116 (15.98)	10,580 (15.98)	
2nd quintile	2,217 (16.73)	11,085 (16.73)	
3rd quintile	2,916 (22.01)	14,580 (22.01)	
4th quintile	2,238 (16.87)	11,175 (16.87)	
5th (uppermost) quintile	3,766 (28.42)	18,830 (28.42)	

Values are expressed as n (%). SSNHL, sudden sensorineural hearing loss.

Jeong J, et al. Audiol Neurootol. 2019;24(5):224-230

Table 2. Risk of SSNHL in individual autoimmune diseases based on diagnostic codes compared with the control group

	N	A risk of SSNHL	HR (CI)	*p* value
Autoimmune-disease group	13,250[1]	151		
Antiphospholipid syndrome	4	0	19.916 (1.22–325.062)	0.0358*
Sarcoidosis	33	0	2.25 (0.139–36.319)	0.5676
Multiple sclerosis	52	3	13.335 (4.646–38.277)	<0.0001*
Rheumatoid arthritis	8,233	98	1.519 (1.204–1.917)	0.0004*
Polyarteritis nodosa	10	0	7.146 (0.451–113.133)	0.1629
Wegener's granulomatosis	47	0	1.24 (0.077–19.962)	0.8794
Systemic lupus erythematosus	279	4	1.538 (0.534–4.43)	0.4254
Dermatopolymyositis	46	0	1.51 (0.094–24.316)	0.7712
Systemic sclerosis	37	0	1.856 (0.116–29.593)	0.6615
Connective-tissue diseases including Sjögren syndrome and Behçet disease	890	11	1.878 (1.044–3.378)	0.0354*
Ankylosing spondylitis	1,417	11	1.132 (0.594–2.157)	0.7066
Psoriasis	2,876	24	1.307 (0.852–2.005)	0.2205
Control group	66,250	484	1	

SSNHL, sudden sensorineural hearing loss; HR, hazard ratio; CI, confidence interval. * $p < 0.05$.

[1] Total number in the autoimmune-disease group (13,250) is less than the sum of patients with each autoimmune disease (13,924) because some patients had ≥2 autoimmune diseases.

Table 3. Risk of SSNHL in patients with individual autoimmune diseases based on the "Special Exception of Assessment" codes compared with the control group

Individual autoimmune disease	Patients with a disease, *N* (matched control group, *N*)	Patients with a risk of SSNHL for a disease, *n* (matched controls with a risk of SSNHL, *n*)	HR (CI)	*p* value
Multiple sclerosis	26 (130)	1 (2)	4.12 (0.256–66.213) 1	0.3177
Rheumatoid arthritis	1,112 (5,560)	19 (46)	2.229 (1.297–3.829) 1	0.0037*
Systemic lupus erythematosus	229 (1,145)	4 (13)	2.843 (0.696–11.617) 1	0.1458
Connective-tissue disease[a]	394 (1,970)	5 (13)	2.328 (0.815–6.647) 1	0.1144
Ankylosing spondylitis	367 (1,835)	5 (14)	1.767 (0.582–5.367) 1	0.3153
Psoriasis	37 (185)	0 (2)	1.543 (0.031–76.415) 1	0.8275

SSNHL, sudden sensorineural hearing loss; HR, hazard ratio; CI, confidence interval. * $p < 0.05$.
[a] Including Sjögren syndrome and Behçet disease.

Summary

- Sudden SNHL **risk** was significantly **higher in the autoimmune-disease group (1.09%)** than in control group (0.73%)
- Sudden SNHL incidence was significantly higher among patients:
  - Antiphospholipid syndrome
  - Multiple sclerosis (MS)
  - Rheumatoid arthritis (RA):
    - common prevalence
    - *vascular ischemia* due to RA autoantibody
  - Connective-tissue diseases: Sjogren syndrome, Behcet's disease

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"Audiological Patterns in Patients with Autoimmune Hearing Loss"

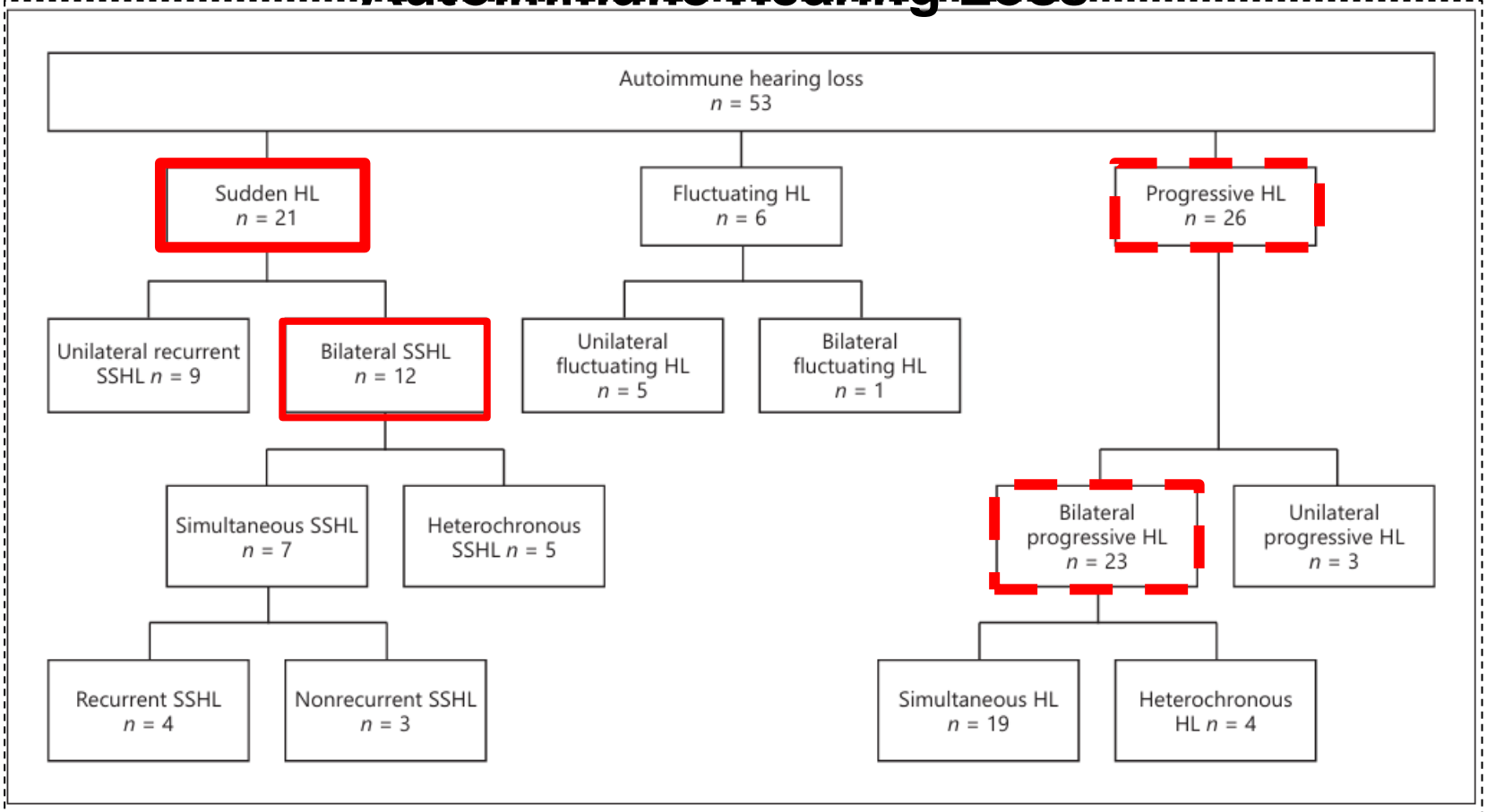


Fig. 1. Distribution of patients according to the onset of autoimmune HL. SSHL, sudden sensorineural hearing loss, HL: hearing loss.

Psillas G. et al, Audiol Neurotol 2022;27: 336-345

Table 1. Characteristics of patients with autoimmune HL, concomitant autoimmune disease, HLA typing and associated audiovestibular symptoms ($N = 53$)

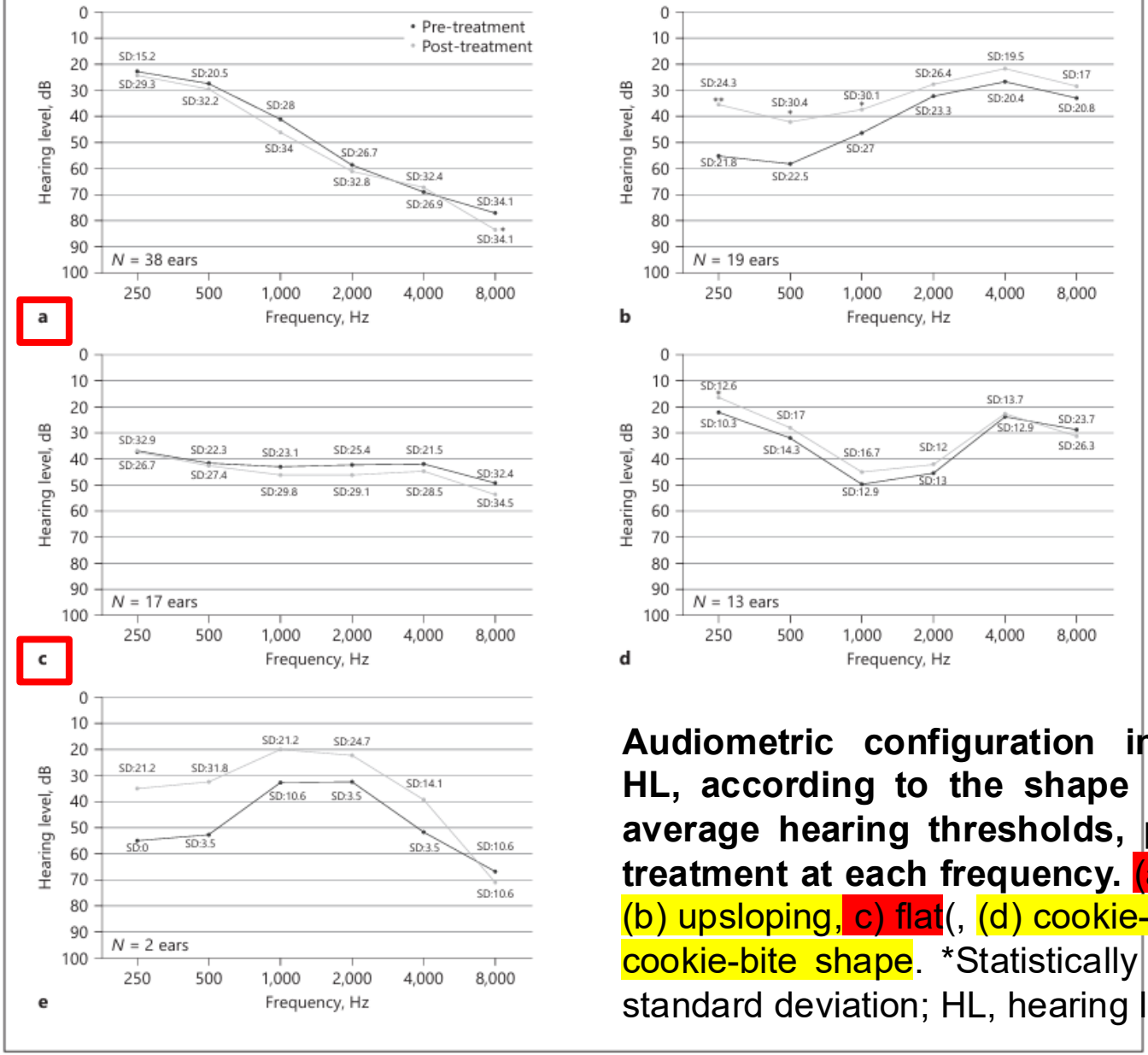
Age (average age ± SD), years	39.3±14
Gender, M/F	10/43
Affected side, R/L	41/48
Unilateral/bilateral HL, *n* (%)	17 (32.1)/36 (67.9)
Antinuclear autoantibodies (ANA), *n* (%)	22 (41.5)
HLA typing (2n alleles), *n* (%)	
HLA B27	2 (1.8)
HLA B35	15 (14.1)
HLA B51	9 (8.4)
HLA C04	17 (16)
HLA C07	15 (14.1)
HLA DRB1*04	11 (10.3)
Concomitant autoimmune disease, *n* (%)	24 (45.2)
HT	13 (24.5)
Rheumatoid arthritis	5 (9.4)
Lupus erythematosus	4 (1.8)
Sjögren's syndrome	1 (1.8)
Idiopathic thrombocytopenic purpura	1 (1.8)
Presence of tinnitus, *n* (%)	41 (77.3)
Presence of vestibular complaints, *n* (%)	21 (39.6)

SD, standard deviation; HL, hearing loss; HT, Hashimoto thyroiditis.

Table 3. Shape of pretreatment pure-tone audiogram according to the affected side in patients with autoimmune HL

	Both ears affected	Right ear affected	Left ear affected	Ears ($N = 89$) (%)
Downsloping	16	2	4	38 (42.6)
Upsloping	4	4	7	19 (21.3)
Flat	8	–	1	17 (19.1)
Cookie-bite	6	1	–	13 (14.6)
Inverse cookie-bite	–	–	2	2 (2.2)

HL, hearing loss.



Audiometric configuration in autoimmune HL, according to the shape and based on average hearing thresholds, pre- and post-treatment at each frequency. (a) downsloping, (b) upsloping, c) flat(, (d) cookie-bite, (e) inverse cookie-bite shape. *Statistically significant, SD, standard deviation; HL, hearing loss.

Summary

- AIHL onset: progressive SNHL, Sudden SNHL, fluctuating HL
- Most common Pure-tone audiogram: down sloping pattern
- Bilateral progressive AIHL was more frequently simultaneous

- ***Recurrent* bilateral Sudden SNHL** vs. recurrent unilateral Sudden SNHL had *statistically* ***negative*** *effect* on hearing recovery (*OR = 0.042, p < 0.05*)
- Heterochronous bilateral Sudden SNHL may have better prognosis than simultaneous bilateral Sudden SNHL (*OR = 10, p = 0.099*)

- Gender, age, concomitant autoimmune disease, high ANA, HLA alleles, tinnitus and vestibular symptoms have no statistical effect on favorable outcome of AIHL

Summary

Limitations

- Small sample size
- Lack a detailed individual analysis of the immunologic profile for a thorough description of autoimmune comorbidities
- Genetic characteristics and clinical presentation
- Needs longer follow up period

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AIED Treatment

- **Shorter** disease duration
- **Less severe** hearing loss at presentation
- Timely **EARLY** immunosuppressive therapy

AIED Treatment

Steroid

- Positive *initial* response up to 70%
- Oral 60 mg (1 mg/kg/d) x 4 weeks
- If hearing improves, treatment should be continued and gradually reduced over 6 months
- Intra-tympanic steroid
- Combination: Oral and IT steroid injection
- Variable treatment response rate

AIED Treatment

Immunosuppressive drugs for secondary AIED

- **Methotrexate** (Response rate: 0 - 80%)
  - acts primarily as an antimetabolite that inhibits dihydrofolate reductase (DHFR), crucial for DNA synthesis, repair, and cellular replication, effectively halting the S-phase of the cell cycle.
  - increases adenosine levels, providing potent anti-inflammatory effects by inhibiting T-cell activation and cytokine production
- **Azathioprine**
  - prevents organ rejection in kidney transplants and treat autoimmune conditions like rheumatoid arthritis, IBD, and lupus
  - inhibits DNA synthesis, reducing immune cell activity.
- **Mycophenolate mofetil** (CellCept)
  - prevents organ rejection in kidney, heart, or liver transplants by inhibiting T and B lymphocyte proliferation

Tinnitus – sensitive predictor of relapse

AIED Treatment

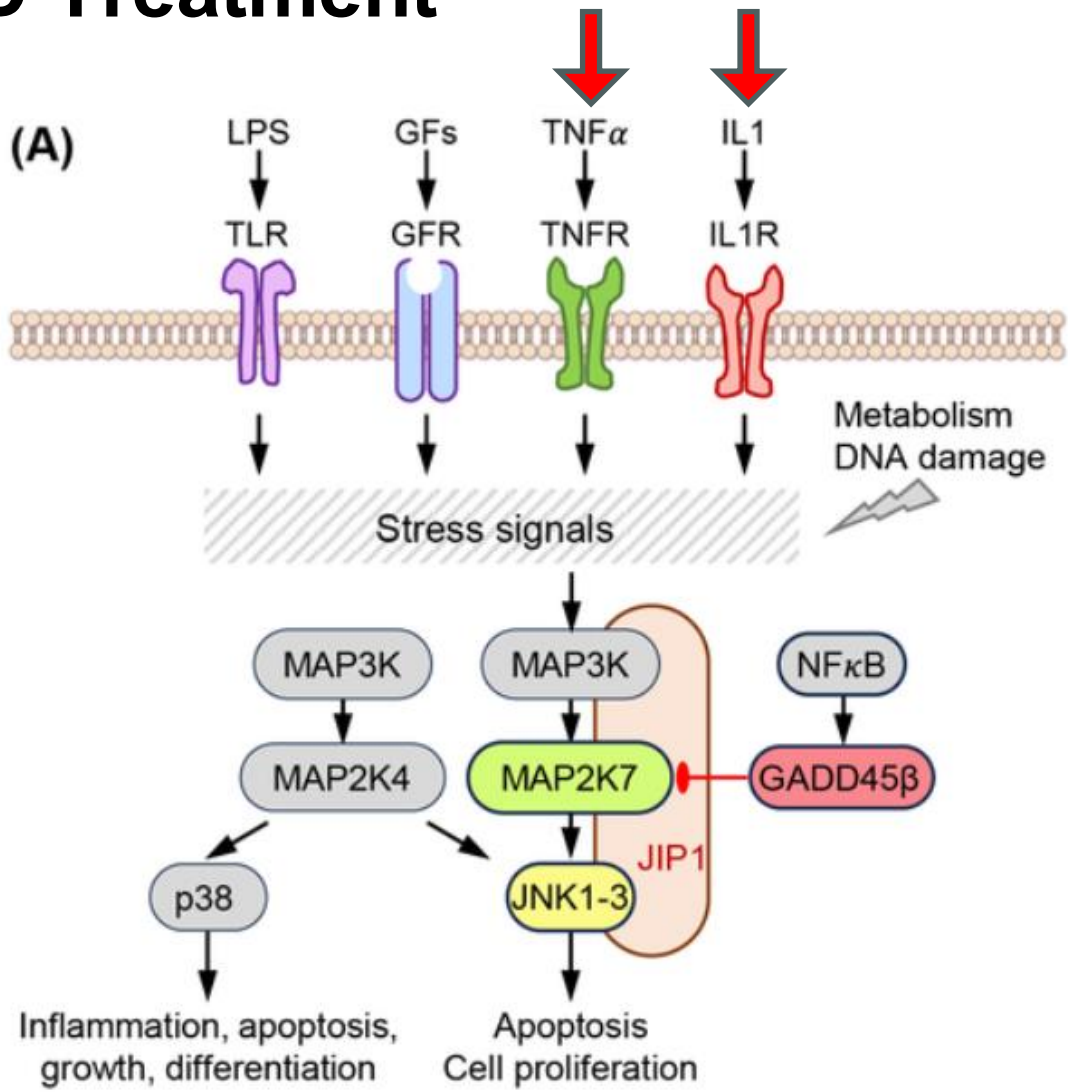
Biologics for refractory cases

- Of the 60% of patients who initially respond to steroid treatment, **only 14% remain responsive after 34 months**, and those patients are at risk for side effects that can range from osteoporosis to psychosis.
  - **Tumor Necrosis Factor (TNF) blocker** (infliximab, Remicade)
  - **CD20 monoclonal antibody** (Rituximab, Rituxan): targets and destroys B-cells
  - **Interleukin IL-6 receptor antagonists** (tocilizumab, Actemra)
  - **Interleukin IL-1 receptor antagonist (IL-1Ra)** (Anakinra)
    - Anakinra is FDA-approved to treat moderate to severely active rheumatoid arthritis

AIED Treatment

- Either the **TNF pathway**, which is either blocked by corticosteroids, N-acetyl cysteine, or a TNF blockade OR **IL-1 pathway**

- Question: which pathway is at play at the time of the patient's hearing loss presentation?
- Either pathway signals on to JNK pathway signaling and cochlear organ destruction:
  - Vascular atrophy
  - Loss of Hair cells and Spiral Ganglion neurons



Lacorazza HD. Front Oncol. 2024 Dec 9;14:1486756

AIED Treatment

Corticosteroid-Resistant and Corticosteroid-Dependent AIED Patients Treated With Anakinra: A Decade of Experience

Vambutas, Andrea*,†; Mullooly, Virginia*; Weber, Sharon*; Pathak, Shresh*,†

Author Information

Otology & Neurotology 46(9):p 1059-1069, October 2025. | **DOI:** 10.1097/MAO.0000000000004553

- 46% of corticosteroid-resistant patients derived benefit from anakinra,
- 73% of corticosteroid-dependent patients achieved a substantial reduction in corticosteroids while maintaining or improving hearing,
- 78% of patients who previously participated in the anakinra trial achieved lasting benefit from ongoing therapy

- **Working hypothesis:**
  - if patient have high TNF levels and low IL-1, then steroid sensitive
  - if patient have high IL-1 beta levels and low TNF, then steroid resistant

AIED Treatment

Autoimmune or Autoinflammatory?

- Understanding autoimmune and autoinflammatory disease requires knowing the difference between the ***innate*** and the ***adaptive*** immune response.
- When the innate immune system works, the macrophages come in first and present antigen to the adaptive immune response.
- **Autoimmune** disease occurs when the adaptive immune response isn't working.
- **Autoinflammatory** disease occurs when the innate immune response isn't working.

- Several autoimmune diseases are now being reclassified as having features of **both *autoimmune* disease and *autoinflammatory* disease**

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Audiologist Questions:

- A patient with recurrent sudden SNHL, treated with steroids and hearing would improve
- ***Recurrence***
- Resistant MRI

- Can autoimmune conditions cause fluctuating hearing loss?
- Is this an autoimmune issue ?
- Symptoms that audiologists look for?
- Proper referrals

Autoimmunity suspected:

- **Bilateral** and **progressive** hearing loss in patients with diagnosed autoimmune disease or positive autoimmune laboratory tests

- Not bilateral progressive hearing loss
- Immune or inflammatory mechanism could also underlie **unilateral** Sudden SNHL
- Unilateral Sudden SNHL could be an initial aspect of autoimmune bilateral SNHL and association with autoimmune disease

AIED/SSNHL Management

Multi-disciplinary approach

- Otolaryngologist
- Rheumatologist
  - Collaboration between otolaryngology and rheumatology is essential to navigate the intricate relationship between AIED and its associated systemic autoimmune diseases (SAIDs)

- **Audiologist**
  - Diagnostic audiogram
  - Serial audiograms to monitor treatment
  - Hearing aids
  - Cochlear implant

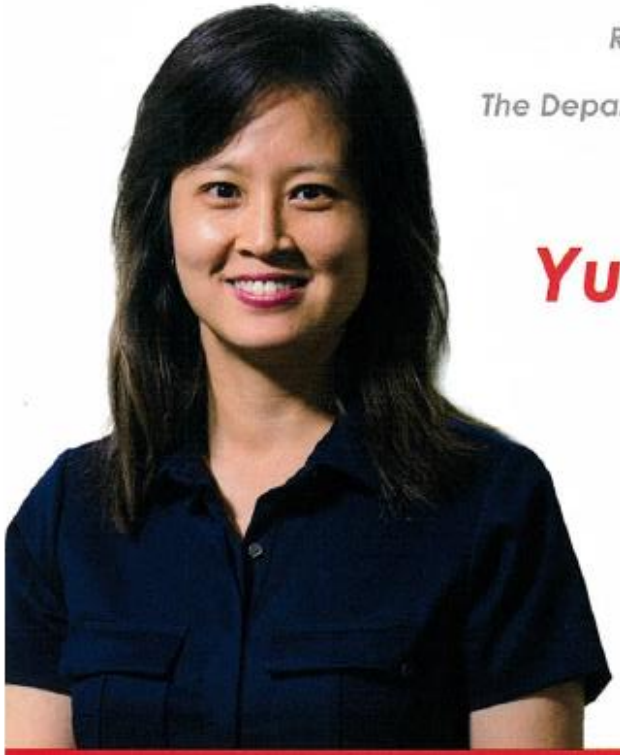
Conclusions

- **Complexity of managing AIED and SSNHL**
  - No single antibody/diagnostic marker associated with AIED, severity of disease and how it should be treated or for how long
  - Unclear whether AIED is one disease or many
    - Animal studies to identify the molecular mechanisms of this disease have been confounding
  - AIED and SSNHL may have similar immunologic responses to **disparate antigens**

Future directions:

- Multicenter studies to better explore the genetic background and environmental factors (allergies, acoustic trauma) of AIED/SSNHL patients
- Immune-mediated mechanism linked to audiometric patterns

Thank You



Rutgers, The State University of New Jersey,
New Jersey Medical School and
The Department of Otolaryngology - Head and Neck Sugery

are pleased to announce
the appointment of

Yu-Lan Mary Ying, MD

Neurotology and Otology

RUTGERS
New Jersey Medical School

Yu-Lan Mary Ying, MD 90 Bergen Street, Suite 8100, Newark, NJ 07103 Phone: (973) 972-2548 Fax: (973) 972-8567

RUTGERS

Patient JC

- 42 year old male
- Hx right Meniere's disease
- Experienced left sudden SNHL (12/2023)
  - Left CI
- Experienced right acute SNHL (3/12/25)
- Rheumatology consultation

  + HLA-B27 antigen

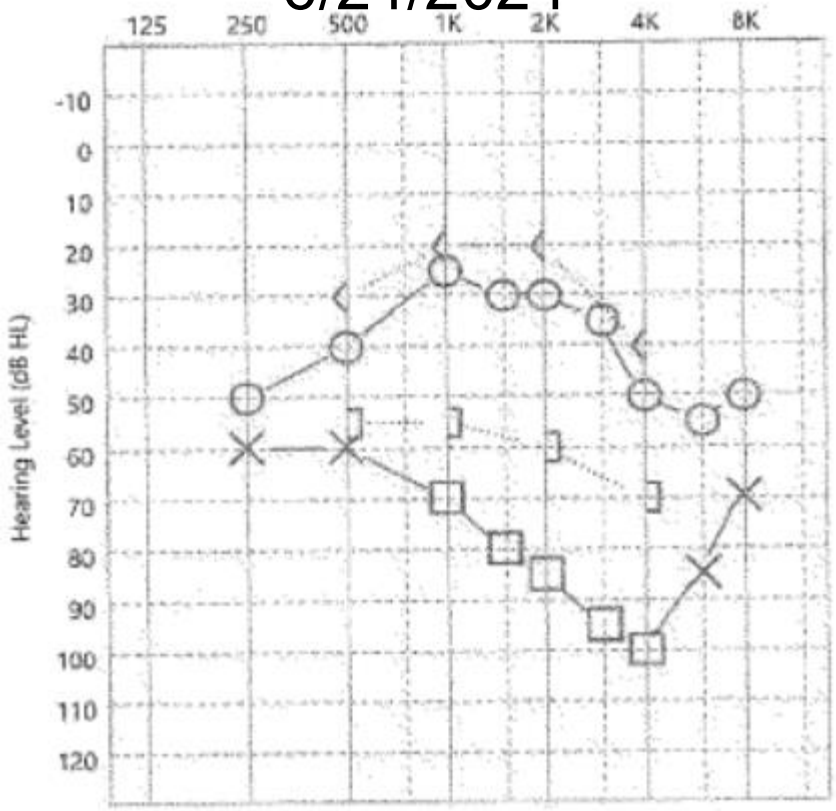
  Psoriasis

- Right progressive moderate SNHL

SRT = 65 dB HL, WRS = 80%. Type C

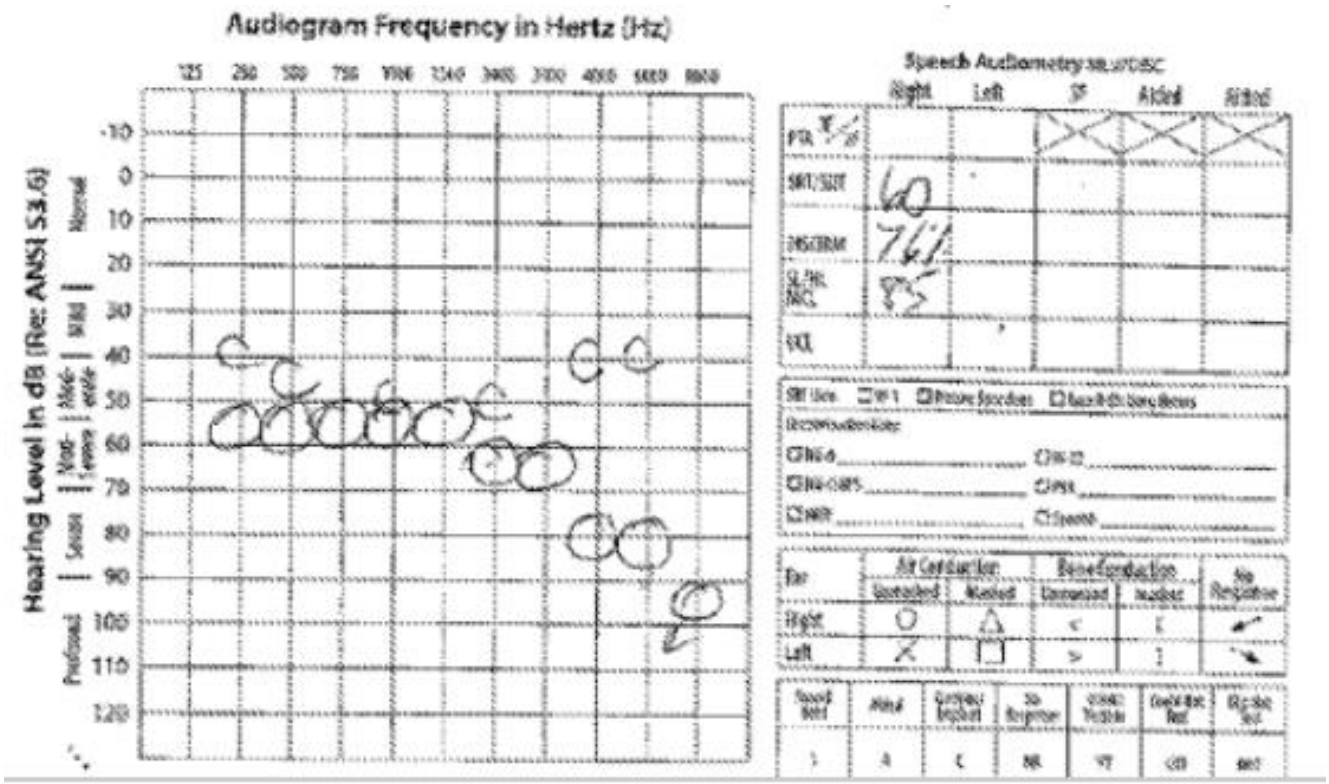
- Medications: folic acid and methotrexate

6/24/2024



Ear	Test Type	Int Ext Mic	Word List	Aided	%	dB HL	dB EM
R	SRT	Microphone	N/A			35	
L	SRT	Microphone	N/A			75	55
R	WRS	Int	NU-6 LIST 2A		100	70	
L	WRS	Int	NU-6 LIST 2A		4	100	60

11/19/2025


Audiogram Frequency in Hertz (Hz)
Hearing Level in dB (Re: ANSI S3.6)
Speech Audiometry
Right
Left
Aided
Unaided