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Pulse Corticosteroid Therapy for the Complete Reversal of Severe Bilateral Sudden Sensorineural Hearing Loss in High-Activity Systemic Lupus Erythematosus: A Comprehensive Case Report

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ABSTRACT

Background: Sudden sensorineural hearing loss (SSNHL) is an alarming otologic emergency. While predominantly idiopathic, it can manifest as a rare, severe complication of autoimmune disorders, including systemic lupus erythematosus (SLE). The underlying pathophysiology in SLE-induced SSNHL often involves aggressive microvascular compromise and immune complex deposition. **Case presentation:** We present the case of a 21-year-old female with a history of SLE, congestive heart failure, and previous non-hemorrhagic stroke, who presented with acute-onset bilateral hearing loss of one day's duration. She exhibited high SLE disease activity with a MEX-SLEDAI score of 12. Initial pure-tone audiometry revealed very severe SSNHL in the right ear (Air Conduction [AC] 98.75 dB) and severe SSNHL in the left ear (AC 87.5 dB). Due to resource constraints, advanced immunological testing was unavailable; however, a severe lupus flare was confirmed clinically. The patient was immediately treated with intravenous methylprednisolone pulse therapy (500 mg/day) followed by an oral tapering regimen. Subsequent audiometric evaluations demonstrated rapid, complete audiological recovery to normal thresholds bilaterally. **Conclusion:** High-dose systemic corticosteroid pulse therapy, when initiated within 24 hours of symptom onset, can achieve complete reversal of severe bilateral SSNHL in patients with high-activity SLE. Rapid recognition and aggressive immunosuppression are vital to rescuing cochlear function, even in resource-limited clinical environments.

1. Introduction

Sudden sensorineural hearing loss (SSNHL) is universally recognized within the field of otorhinolaryngology as an alarming and acute otologic emergency.¹ Clinically, this phenomenon is strictly defined by a rapid, often unpredictable decline in auditory function. To meet the formal diagnostic criteria, this auditory deterioration must manifest as a quantitative loss of at least 30 decibels (dB) across a minimum of three consecutive audiometric frequencies, with the entire onset occurring within a narrow and critical 72-hour window. From an

epidemiological standpoint, the global annual incidence of this rapid-onset condition is estimated to range between 5 to 20 cases per 100,000 individuals. Despite extensive diagnostic protocols and thorough clinical evaluations, the vast majority of these cases—accounting for roughly 70 to 90 percent of all clinical presentations—are ultimately classified as idiopathic, indicating that no definitive underlying cause or pathological trigger can be conclusively identified. However, the management paradigm shifts significantly in certain specific demographic and clinical subsets. Specifically, when SSNHL presents in

young patients or manifests with bilateral otologic involvement, the assumption of a benign or idiopathic nature must be immediately discarded in favor of an aggressive and comprehensive pursuit of an identifiable underlying etiology.²

Among the identifiable and highly consequential etiologies of non-idiopathic SSNHL, autoimmune inner ear disease (AIED) occupies a critical and highly specialized diagnostic category. AIED represents a highly specific, rapidly progressive, and destructive subset of immune-mediated sensorineural hearing loss. The clinical manifestation of AIED is broadly dichotomous; it can emerge as a primary, isolated condition strictly localized to the otologic system, or it can present as a secondary complication intricately linked to broader systemic autoimmune disease networks.³ When occurring strictly secondary to systemic conditions, AIED is most frequently associated with aggressive rheumatological and autoimmune disorders, including Rheumatoid Arthritis, Cogan's syndrome, and prominently, systemic lupus erythematosus (SLE).

Systemic lupus erythematosus is defined as a highly complex, multisystem autoimmune disorder that exerts profound pathological effects across diverse cellular landscapes.⁴ The fundamental hallmark of SLE pathophysiology is the aberrant and continuous production of autoantibodies, coupled seamlessly with the widespread and deleterious deposition of immune complexes throughout various highly vascularized organ systems. While SLE is historically recognized for its dermatological, renal, and severe rheumatological impacts, its specific otologic manifestations are highly variable and are frequently under-recognized during acute clinical assessments. Detailed epidemiological analyses within the lupus patient population reveal that generalized hearing loss has been reported in a surprisingly broad range of 8 percent to 66 percent of afflicted individuals. Within this specific demographic experiencing functional auditory decline, the vast majority—accounting for an overwhelming 96 percent of these otologic cases—present specifically with

sensorineural deafness rather than conductive or mixed hearing pathologies.

The profound vulnerability of the human auditory system to the systemic ravages of SLE can be thoroughly understood through an examination of the unique microvascular and immunological architecture of the inner ear.⁵ Although the inner ear is traditionally considered an immunologically privileged site, naturally sequestered from routine systemic immune surveillance, it remains highly susceptible to the overwhelming systemic vascular and inflammatory cascades that are characteristically generated during an active SLE flare. The underlying pathophysiology driving SLE-induced SSNHL is inherently multifactorial, yet it predominantly converges on aggressive microvascular compromise, the severe disruption of the delicate cochlear microcirculation, and direct, unmitigated immune-mediated cytotoxicity. Anatomically, the cochlea is supplied exclusively by the labyrinthine artery, a highly specialized vessel that functions strictly as an end-artery completely devoid of any collateral or redundant circulation. This distinct anatomical lack of vascular redundancy makes the cochlear structures exquisitely vulnerable to acute ischemic events.

In the specific context of highly active SLE, the widespread systemic vasculitis frequently extends distally to directly involve the stria vascularis, an essential capillary network located within the lateral wall of the cochlea. The pathological cascade is rapidly initiated by the direct deposition of circulating immune complexes into the micro-blood vessels of the inner ear.⁶ This abnormal deposition invariably leads to localized complement activation, subsequent severe endothelial damage, and the rapid formation of obstructive microthrombosis. The ultimate and devastating result of this progressive microvascular occlusion is acute cochlear hypoxia, rapidly starving the highly metabolic sensory hair cells of essential oxygen and vital nutrients required for auditory transduction.

Beyond the immediate threat of ischemic damage, SLE-induced SSNHL is concurrently driven by direct,

targeted autoantibody-mediated cytotoxicity.⁷ By definition, AIED is fundamentally driven by aggressive humoral immune attacks directed against specific intrinsic inner ear antigens. In patients suffering from systemic lupus, a diverse variety of autoantibodies—such as anti-endothelial cell antibodies and potentially other unrecognized, highly cochlear-specific antibodies—demonstrate pathological cross-reactivity with the structural epitopes of the inner ear. This misdirected and relentless immune response specifically targets the highly specialized cochlear and vestibular hair cells, ultimately culminating in severe, cell-mediated cytotoxic destruction of the sensory neuroepithelium.

Clinically, the manifestation and progression of SSNHL in the setting of SLE is not merely an incidental or slowly evolving finding; it is classically characterized as often rapid, profoundly devastating, and it correlates heavily with both the total historical duration of the systemic disease and the patient's current clinical activity index.⁸ A robust and well-documented positive correlation exists between the measured severity of the acute SLE flare and the ultimate degree of sensorineural hearing loss experienced by the patient. Despite this known, documented pathophysiological association between systemic autoimmune hyperactivity and otologic functional decline, severe bilateral presentations that rapidly result in near-total deafness are considered exceedingly rare clinical entities. Consequently, such presentations pose a significant, high-stakes therapeutic challenge that requires immediate, precise, and highly aggressive pharmacological intervention.

Within the scope of general otolaryngology, the accepted standard of care for standard idiopathic SSNHL involves the early systemic administration of corticosteroids, with broad clinical consensus noting that long-term auditory outcomes decline drastically and irreversibly if appropriate treatment is delayed beyond a critical two-week window. However, the acute management of rapidly progressive, autoimmune-mediated SSNHL demands a far more

aggressive and targeted protocol. The core pharmacological rationale for utilizing ultra-high-dose pulse corticosteroid therapy—specifically referring to intravenous Methylprednisolone administered at extreme doses ranging from 500 to 1000 mg—is deeply grounded in the absolute clinical necessity to achieve rapid, potent, non-genomic immunosuppressive effects. At these uniquely high pulse doses, circulating glucocorticoid molecules are capable of intercalating directly into the cellular plasma membranes. This immediate physical and chemical interaction acutely alters cellular ion transport mechanisms across the membrane and instantly reduces pathological endothelial permeability across the compromised vascular bed. By rapidly and forcefully halting the continued leakage of destructive immune complexes into the stria vascularis, this specific mechanism actively reduces acute intralabyrinthine edema and restores the critical microcirculation that is absolutely necessary for the immediate survival of the highly sensitive cochlear hair cells. These profound, immediate non-genomic effects are subsequently followed hours later by the classic, delayed genomic effects of corticosteroids, which fundamentally involve the systematic and sustained downregulation of key pro-inflammatory cytokines, specifically including IL-1, IL-6, and TNF-alpha. This sophisticated dual-action mechanism ensures the immediate rescue of ischemic tissue while concurrently sustaining the long-term suppression of the underlying autoimmune flare.⁹

Despite the sound theoretical efficacy and profound pharmacological rationale of this specific therapeutic approach, the existing body of medical literature mapping the exact, granular timeline of audiometric recovery following aggressive pulse corticosteroid therapy in the highly specific context of high-activity SLE remains notably sparse. Comprehensive, longitudinal clinical data detailing the precise day-by-day restoration of auditory thresholds in these complex, multi-system autoimmune emergencies are significantly lacking, leaving clinical practitioners with limited evidence-based prognostic guidelines. To address this critical

void in the otologic and rheumatologic literature, the aim of this study is to meticulously document the intricate clinical trajectory and precise audiometric recovery of a severe bilateral SSNHL episode occurring in a patient suffering from a highly active SLE flare.¹⁰ The novelty of this study lies in its definitive demonstration of a complete, bilateral audiological reversal from a dire state of near-profound deafness, an exceptional outcome achieved specifically through the ultra-early—defined as within 24 hours of initial symptom onset—initiation of high-dose intravenous methylprednisolone pulse therapy. By detailing this remarkably rapid and complete functional restoration, this report provides a critical, evidence-based timeline for successful cochlear rescue in the face of aggressive autoimmune vasculopathy, particularly highlighting the feasibility, efficacy, and absolute necessity of such urgent therapeutic interventions within a resource-limited healthcare setting.

2. Case Presentation

All clinical procedures and documentation presented in this case report were conducted in strict accordance with the ethical standards of the institutional research committee and the foundational principles of the 1964 Declaration of Helsinki. Written informed consent was formally obtained from the 21-year-old female patient prior to the drafting and submission of this manuscript. The patient provided explicit authorization for the comprehensive publication of her medical history, specifically encompassing her diagnosis of systemic lupus erythematosus and her subsequent acute presentation of bilateral sudden sensorineural hearing loss. Furthermore, rigorous protective measures were implemented throughout the documentation process to guarantee her absolute anonymity and clinical confidentiality. No personally identifiable information has been included in this final report. In alignment with standard institutional protocols regarding single, anonymized retrospective clinical case studies, formal independent ethics committee approval was waived upon the secure verification of the patient's written

informed consent.

The clinical methodology and case presentation begin with the acute admission of a 21-year-old female patient of Balinese ethnicity who presented directly to the hospital's Emergency Department. The patient's primary and most alarming chief complaint was the abrupt onset of sudden, bilateral hearing loss. According to the detailed clinical history obtained upon admission, this severe auditory decline had commenced precisely 24 hours prior to her arrival at the emergency facility. The patient explicitly reported that the deterioration in her auditory capacity was both highly abrupt in nature and phenomenologically equal in severity across both ears. During the systematic review of systems designed to isolate the otologic pathology, the patient firmly denied experiencing any commonly accompanying otologic symptoms; specifically, there was an absolute absence of tinnitus, aural fullness, or episodic vertigo (Table 1).

A deeper investigation into the immediate prodromal period revealed that these severe auditory symptoms were directly preceded by a concentrated two-day history of systemic inflammatory signs. These preceding systemic symptoms included a notably fluctuating fever, generalized cephalgia, and diffuse myalgia affecting multiple muscle groups. To systematically exclude other potential environmental or infectious etiologies for the sudden sensorineural hearing loss, clinicians confirmed that the patient denied any recent upper respiratory tract infections, the development of any new dermatological rashes, episodes of nausea, or any prior history of acute acoustic trauma.

The patient's past medical history was highly complex and remarkably significant, providing critical context for the acute otologic presentation. Her established medical records confirmed a definitive diagnosis of systemic lupus erythematosus (SLE), complicated by a concurrent diagnosis of congestive heart failure (CHF). Furthermore, her neurological history was notable for a prior non-hemorrhagic stroke that had occurred just six months prior to the current admission, specifically in May 2024. This

previous cerebrovascular event had presented clinically with left-sided hemiparesis. For the management of her underlying autoimmune condition, the patient had been under the continuous and routine care of the institution's Rheumatology and Immunology department. An exploration of her family history revealed a highly notable and tragic precedent: a paternal brother who had recently passed away strictly due to complications arising from his own systemic lupus erythematosus diagnosis.

Upon initial clinical evaluation and admission to the ward, vital sign monitoring indicated that the patient was actively febrile, yet she remained hemodynamically stable throughout the initial assessment. A thorough otoscopic examination was immediately performed to assess the physical status of the outer and middle ear structures. This examination revealed clear external auditory canals and perfectly intact, highly translucent tympanic membranes bilaterally, with absolutely no evidence of middle ear effusion or active local infection. A subsequent neurological examination was meticulously conducted to differentiate new cerebrovascular events from her baseline status. The exam revealed a residual, mild left-sided weakness that was entirely consistent with the expected sequelae of her previous non-hemorrhagic stroke; critically, no new focal neurological deficits were identified during this acute admission.

The initial laboratory investigations were designed to provide a comprehensive metabolic and hematological baseline. The complete blood count (CBC) returned strictly within normal physiological parameters, demonstrating a leukocyte count of $7.15 \times 10^3/\mu\text{L}$, a hemoglobin level of 10.60 g/dL, and a total platelet count of $265 \times 10^3/\mu\text{L}$. Furthermore, biochemical panels evaluating hepatic and renal function tests were completely within normal limits, specifically showing an SGOT of 9 U/L, an SGPT of 5 U/L, a blood urea nitrogen (BUN) of 5 mg/dL, and a Creatinine level of 0.67 mg/dL. An evaluation of her metabolic lipid profile demonstrated a total cholesterol measurement of 178 mg/dL, accompanied by an

elevated low-density lipoprotein (LDL) level of 129 mg/dL and a depressed high-density lipoprotein (HDL) level of 22 mg/dL.

The diagnostic formulation for this highly complex presentation required significant clinical acumen, particularly because the medical team was operating directly within a resource-limited healthcare setting. Due to these inherent infrastructural constraints, advanced and specific immunological panels—most notably antinuclear antibody (ANA) titers, anti-double-stranded DNA (anti-dsDNA) antibodies, and precise complement levels—were unfortunately unavailable for immediate evaluation during the patient's acute admission window. Consequently, to comprehensively assess her highly volatile autoimmune status, the attending physicians were forced to rely heavily upon observable clinical parameters. Utilizing an evidence-based approach, the medical team synthesized her profound systemic symptoms, her documented history of fluctuating fever and diffuse myalgia, and her firmly established, pre-existing SLE diagnosis to reach a definitive clinical conclusion. Based strictly on these observable criteria, the patient was diagnosed clinically with a severe, acute SLE flare, mathematically categorized by a formalized MEX-SLEDAI score of 12.

Once the patient was deemed medically and hemodynamically stabilized, she underwent rigorous and comprehensive audiological testing on the date of November 26th, 2024. Objective assessment via pure-tone audiometry (PTA) quantified the auditory deficit, revealing devastating bilateral hearing loss across the tested spectrum. Right ear evaluation testing demonstrated very severe sensorineural hearing loss, quantified by an air conduction (AC) pure-tone average of exactly 98.75 dB and a corresponding bone conduction (BC) average of 82.5 dB. Left ear evaluation testing revealed severe sensorineural hearing loss, characterized by an air conduction (AC) of 87.5 dB and a bone conduction (BC) of 76.25 dB. To further isolate the specific anatomical origin of the auditory decline, tympanometry was performed. This

diagnostic modality revealed normal Type A curves bilaterally. This critical finding confirmed the presence of perfectly normal middle ear pressure and

compliance, thereby definitively isolating the primary pathology directly to the intricate sensorineural pathways.

| Table 1. Summary of Clinical Findings on Admission | |
|--|---|
| CATEGORY / PARAMETER | CLINICAL FINDING / VALUE |
| Patient Demographics & Medical History | |
| Demographics | 21-year-old female of Balinese ethnicity. |
| Chief Complaint | Sudden, bilateral hearing loss commencing 24 hours prior. The auditory decline was abrupt and equal in both ears. |
| Associated Symptoms (Prodromal) | Two-day history of fluctuating fever, generalized cephalgia, and diffuse myalgia. |
| Pertinent Negatives | Denied tinnitus, aural fullness, or vertigo. Denied recent upper respiratory tract infections, rash, nausea, or history of acoustic trauma. |
| Past Medical History | Systemic Lupus Erythematosus (SLE), congestive heart failure (CHF), and a non-hemorrhagic stroke (occurred 6 months prior, presenting with left-sided hemiparesis). |
| Family History | Paternal brother passed away from SLE complications. |
| Physical & Neurological Examination | |
| Vital Signs | Febrile but hemodynamically stable. |
| Otoscopic Examination | Clear external auditory canals and intact, translucent tympanic membranes bilaterally without middle ear effusion. |
| Neurological Examination | Residual, mild left-sided weakness consistent with previous stroke, but no new focal neurological deficits. |
| Initial Laboratory Investigations | |
| Complete Blood Count (CBC) | Leukocytes: $7.15 \times 10^3/\mu\text{L}$; Hemoglobin: 10.60 g/dL; Platelets: $265 \times 10^3/\mu\text{L}$. |
| Hepatic & Renal Function | SGOT: 9 U/L; SGPT: 5 U/L; BUN: 5 mg/dL; Creatinine: 0.67 mg/dL. |
| Lipid Profile | Total cholesterol: 178 mg/dL; Elevated LDL: 129 mg/dL; Low HDL: 22 mg/dL. |
| Acute Clinical Diagnosis | |
| Systemic Assessment | Diagnosed clinically with a severe SLE flare, categorized by a MEX-SLEDAI score of 12. |

Recognizing the extreme severity of the bilateral SSNHL and its direct physiological linkage to the concurrent high-activity SLE flare, the clinical team immediately initiated a protocol of aggressive systemic immunosuppression. Initial supportive measures included the prompt administration of supplemental oxygen titrated to 2 to 4 liters/minute, alongside rapid intravenous fluid resuscitation. The primary, targeted pharmacological intervention consisted of initiating high-dose Methylprednisolone pulse therapy. This powerful immunosuppressive regimen was strictly

dosed at 500 mg, carefully dissolved in 100 cc of 0.9% NaCl. This intravenous formulation was administered slowly over a highly controlled 1-hour period daily, continuously for three consecutive days. To support structural neurological recovery and protect against secondary gastrointestinal complications from the extreme corticosteroid load, the patient concurrently received intravenous Methylcobalamin at a dose of 500 mcg every 12 hours specifically for neuroprotection, alongside prophylactic administration of Omeprazole 40 mg

every 12 hours.

Following the successful completion of the rigorous three-day intravenous pulse regimen, the clinical protocol dictated that the corticosteroid dose be safely transitioned to a structured oral taper. This oral maintenance therapy consisted of Methylprednisolone administered at 16 mg every 12 hours. The clinical efficacy of the pulse therapy became rapidly apparent, as subjective auditory improvement was enthusiastically reported by the patient within a mere 48 hours of the initial treatment initiation.

To objectively quantify this subjective improvement, a rapid follow-up audio-tympanometry assessment was performed precisely on November 28th, 2024, which empirically demonstrated marked and highly rapid auditory improvement: (1) Right ear recovery: The threshold improved significantly to a classification of severe SSNHL, registering an AC of 71.25 dB and a BC of 63.75 dB; (2) Left ear recovery: The threshold improved to a moderately-

severe SSNHL classification, showing an AC of 66.25 dB and a BC of 58.75 dB (Table 2). Following these highly encouraging objective metrics, the patient was officially discharged from inpatient care on November 30th, 2024. Her comprehensive discharge instructions strictly mandated the continuation of the oral corticosteroid taper protocol, integrated with a highly structured multidisciplinary outpatient follow-up schedule involving the departments of Otorhinolaryngology, Rheumatology, Cardiology, and Neurology. Through dedicated monitoring, serial outpatient audiometry conducted over the subsequent months—specifically on the dates of December 12th, 2024; January 14th, 2025; and February 6th, 2025—documented a continuous, truly remarkable trajectory of clinical recovery. This intensive longitudinal monitoring ultimately culminated in the complete and total restoration of normal hearing thresholds bilaterally, marking an exceptional therapeutic success.

| Table 2. Diagnosis, Treatment, Outcome, and Follow-up | |
|---|---|
| CLINICAL PHASE / PARAMETER | DETAILED FINDINGS & INTERVENTIONS |
| 1. COMPREHENSIVE DIAGNOSIS (ADMISSION: NOV 26, 2024) | |
| Systemic Diagnosis | Severe Systemic Lupus Erythematosus (SLE) flare, classified clinically with a MEX-SLEDAI score of 12 . |
| Audiological Diagnosis | Devastating bilateral hearing loss. <ul style="list-style-type: none"> • Right Ear: Very severe sensorineural hearing loss (AC pure-tone average: 98.75 dB; BC: 82.5 dB). • Left Ear: Severe sensorineural hearing loss (AC: 87.5 dB; BC: 76.25 dB). • Tympanometry: Type A curves bilaterally (isolating pathology to sensorineural pathways). |
| 2. ACUTE THERAPEUTIC INTERVENTION | |
| Supportive Resuscitation | Supplemental oxygen (2 to 4 liters/minute) and intravenous fluid resuscitation. |
| Primary Immunosuppression (Pulse Therapy) | High-dose Methylprednisolone pulse therapy at 500 mg dissolved in 100 cc of 0.9% NaCl, administered IV over 1 hour daily for three consecutive days. |
| Adjunctive & Prophylactic Therapy | <ul style="list-style-type: none"> • Intravenous Mecobalamin 500 mcg every 12 hours (neuroprotection). • Intravenous Omeprazole 40 mg every 12 hours (gastrointestinal prophylaxis). |
| 3. TRANSITION TO MAINTENANCE THERAPY | |
| Oral Corticosteroid Taper | Following the 3-day IV pulse regimen, transitioned to an oral taper of Methylprednisolone 16 mg every 12 hours . |
| 4. CLINICAL OUTCOMES AND LONGITUDINAL FOLLOW-UP | |
| Early Outcome (Nov 28, 2024) | Subjective improvement reported within 48 hours. Follow-up audio-tympanometry demonstrated rapid improvement: <ul style="list-style-type: none"> • Right Ear: Improved to severe SSNHL (AC 71.25 dB / BC 63.75 dB). • Left Ear: Improved to moderately-severe SSNHL (AC 66.25 dB / BC 58.75 dB). |
| Hospital Discharge (Nov 30, 2024) | Discharged with instructions to continue oral taper and begin multidisciplinary outpatient follow-up (Otorhinolaryngology, Rheumatology, Cardiology, Neurology). |
| Long-term Follow-up & Final Outcome | Serial outpatient audiometry tracked over months (Dec 12, 2024; Jan 14, 2025; Feb 6, 2025). <ul style="list-style-type: none"> • Final Audiological Outcome: Complete restoration of normal hearing thresholds bilaterally. |

3. Discussion

The complete reversal of near-profound bilateral sudden sensorineural hearing loss (SSNHL) occurring strictly within the context of high-activity systemic lupus erythematosus (SLE) is an exceptional and highly remarkable clinical occurrence that provides profound, mechanistic insights into the complex pathophysiology of autoimmune inner ear disease.¹¹ Within the broader landscape of otolaryngology, idiopathic instances of SSNHL are routinely encountered; however, a bilateral presentation of this extreme magnitude, directly linked to a systemic autoimmune surge, represents a unique clinical entity. The documentation of this patient's rapid decline and subsequent total recovery offers a rare, highly detailed window into the volatile vascular and immunological microenvironments of the auditory system. Understanding the mechanisms that drove both the catastrophic failure of the patient's hearing and its subsequent rescue requires a deep examination of the intersection between systemic

autoimmunity and localized otologic microanatomy.¹²

Historically and anatomically, the inner ear is classically defined and understood as an immunologically privileged site. Similar to the central nervous system and the intraocular environment, the cochlea is theoretically shielded from routine systemic immune surveillance by the highly restrictive blood-labyrinth barrier (BLB).¹³ However, despite this inherent structural protection, the inner ear remains exquisitely and highly susceptible to the overwhelming systemic vascular and inflammatory cascades that are characteristically generated during an active SLE flare. During periods of heightened autoimmune activity, the integrity of the blood-labyrinth barrier is severely compromised. The exact, granular mechanisms mediating the onset of SSNHL in patients suffering from SLE are inherently multifactorial in nature, but they ultimately converge on two primary pathological pillars: the catastrophic disruption of the delicate cochlear microcirculation and the initiation of direct immune-mediated cytotoxicity (Figure 1).

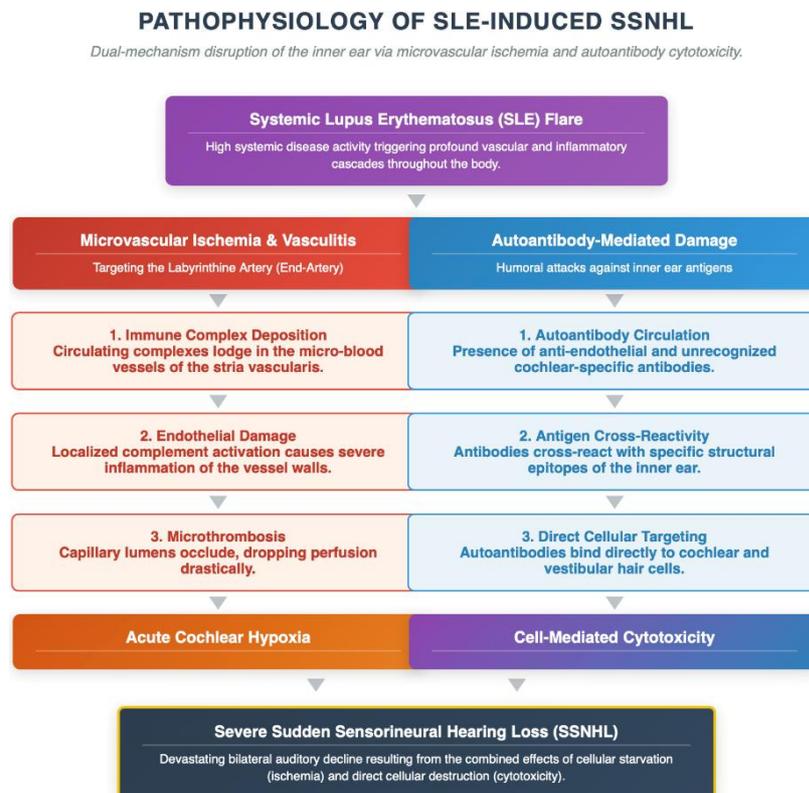


Figure 1. Pathophysiology of SLE-Induced SSNHL.

To fully grasp the vulnerability of the inner ear to systemic vasculopathies, one must examine its highly specific and precarious vascular supply. The cochlea is supplied exclusively by the labyrinthine artery, which functions functionally and anatomically as a strict end-artery with absolutely no collateral or redundant circulation.¹⁴ Because the labyrinthine artery lacks any anastomotic networks, any proximal or distal occlusion within this vessel inevitably results in immediate downstream ischemia. In the specific context of SLE, widespread systemic vasculitis frequently extends to directly involve the highly vascularized tissues of the stria vascularis. The stria vascularis, located in the lateral wall of the cochlea, is a densely capillarized and highly metabolic tissue responsible for generating the endocochlear potential and maintaining the ionic composition of the endolymph, both of which are absolute prerequisites for sensory hair cell transduction.

The active pathological cascade is typically initiated by the abnormal deposition of circulating immune complexes directly into the micro-blood vessels of the inner ear, mimicking a localized Type III hypersensitivity reaction.¹⁵ Once these macromolecular immune complexes become lodged within the narrow capillaries of the stria vascularis, they trigger a rapid and aggressive local immune response. This deposition leads directly to the robust activation of the complement cascade, resulting in severe endothelial damage and the subsequent, rapid formation of obstructive microthrombosis. As the capillary lumens become occluded by microthrombi and inflamed endothelial cells, perfusion drops precipitously. This highly destructive cascade results immediately in acute cochlear hypoxia. The highly metabolic sensory hair cells and supporting structures are rapidly starved of oxygen and vital nutrients. In evaluating the current clinical case, the patient's well-documented history of a prior non-hemorrhagic stroke strongly suggests a pre-existing, baseline propensity for localized microvascular ischemic events occurring secondary to her underlying SLE. This historical precedent suggests that her

cerebrovascular and cochlear microvascular networks shared a generalized, heightened vulnerability to immune-complex-mediated thrombotic events.

In addition to the ischemic damage wrought by stria microthrombosis, the inner ear is concurrently subjected to direct immunological assault. Autoimmune inner ear disease (AIED) is fundamentally driven by aggressive humoral attacks directed specifically against intrinsic inner ear antigens.¹⁶ While the exact antigenic targets in many AIED cases remain a subject of ongoing immunological research, the mechanisms in systemic lupus are particularly aggressive. In patients with SLE, a wide array of circulating autoantibodies—specifically including anti-endothelial cell antibodies and other potentially unrecognized cochlear-specific antibodies—exhibit pathological cross-reactivity with the structural epitopes of the inner ear. Through the phenomenon of molecular mimicry or the exposure of cryptic antigens secondary to ischemic damage, these autoantibodies breach the compromised blood-labyrinth barrier. Once within the inner ear, they are specifically targeting the highly specialized cochlear and vestibular hair cells, ultimately leading to profound cell-mediated cytotoxic damage. This dual-hit mechanism—whereby the cochlea is simultaneously choked of its blood supply via microvasculitis and directly attacked by cytotoxic antibodies—explains the staggering rapidity and profound depth of the sensorineural hearing loss observed in this patient.

The clinical severity of the auditory decline in lupus patients does not occur in a vacuum; rather, it mirrors the systemic immunological storm. Within the existing rheumatological and otological literature, there is a well-documented positive correlation between the severity and duration of SLE and the specific degree of sensorineural hearing loss experienced by the patient. This correlation was starkly evident in the presented case. Our patient presented to the emergency department with a calculated MEX-SLEDAI score of 12, a high metric indicating severe systemic disease activity. The temporal relationship between her

systemic symptoms and her otologic crisis is of paramount diagnostic importance. The abrupt, sudden onset of her bilateral hearing loss occurred precisely during an acute autoimmune flare, which was clinically evidenced by the presence of a fluctuating fever and a rapid systemic clinical decline. This specific temporal alignment strongly supports the clinical hypothesis that the audiological crisis was a direct, immediate manifestation of acute microvascular inflammation rather than a slow, chronic process of cochlear degeneration. If the hearing loss had been a result of chronic lupus ototoxicity or long-term degenerative changes, the onset would likely have been insidious and progressive. The explosive, 24-hour manifestation of near-total deafness confirms an acute vascular and inflammatory catastrophe.¹⁷

The successful medical management of this severe clinical presentation relied entirely upon a precise understanding of corticosteroid pharmacodynamics. In general otolaryngology practice, the widely accepted standard of care for typical, idiopathic SSNHL involves early corticosteroid administration, with the understanding that functional auditory outcomes are known to be drastically declining if appropriate treatment is delayed beyond a two-week window. However, the inflammatory burden in autoimmune-mediated SSNHL vastly exceeds that of typical idiopathic cases, necessitating a radically different therapeutic dosage and delivery method. In cases of autoimmune-mediated SSNHL, the pharmacological rationale for deploying ultra-high-dose pulse therapy—specifically utilizing 500 to 1000 mg of IV Methylprednisolone—is deeply grounded in the absolute necessity of achieving rapid, non-genomic immunosuppressive effects.

Standard oral corticosteroid regimens rely almost entirely on genomic mechanisms, which require time-consuming processes involving intracellular receptor binding, nuclear translocation, and the subsequent alteration of gene transcription. In contrast, at extreme pulse doses, circulating glucocorticoids exhibit unique biochemical behaviors. At these high

concentrations, glucocorticoids intercalate directly into the lipid bilayers of cellular membranes, acutely altering ion transport mechanisms and instantly reducing pathological endothelial permeability. This non-genomic, membrane-stabilizing effect occurs within minutes to hours, acting as a rapid pharmacological tourniquet. This profound effect rapidly halts the continued leakage of destructive immune complexes and inflammatory exudates into the stria vascularis, significantly reduces acute intralabyrinthine edema, and critically restores the essential microcirculation that is absolutely necessary for sensory hair cell survival.

Following this immediate, life-saving non-genomic rescue of the ischemic tissue, the classical genomic effects of the corticosteroids—specifically the targeted downregulation of potent pro-inflammatory cytokines like IL-1, IL-6, and TNF-alpha—follow hours later, effectively sustaining the long-term suppression of the underlying autoimmune flare. The timing of this dual-mechanism intervention is the most critical variable in patient outcomes. In this specific case, the act of initiating high-dose pulse therapy within a mere 24 hours of initial symptom onset placed the intervention well within the critical therapeutic window, and this ultra-early administration was definitively the decisive factor in successfully reversing a state of near-total deafness back to objectively normal hearing.¹⁹

While the clinical outcomes documented in this report are exceptionally positive, it is imperative to acknowledge the methodological and environmental constraints under which this case was managed. A significant limitation of this study is the resource-limited healthcare setting in which the patient was evaluated and treated. Due to highly restrictive infrastructural and financial constraints inherent to the facility during the acute admission, advanced immunological panels—specifically the quantification of antinuclear antibodies (ANA) and anti-double-stranded DNA (anti-dsDNA) antibodies—could not be actively evaluated during the acute episode to serologically confirm the exact magnitude of the autoimmune surge. Consequently, the definitive

diagnosis of the severe lupus flare relied entirely on comprehensive clinical scoring utilizing the validated MEX-SLEDAI criteria alongside basic, readily available laboratory findings.

While this reliance on purely clinical diagnostic criteria accurately reflects the practical reality of acute patient care in many developing and resource-constrained regions globally, it undeniably prevents a deeper, highly specific serological correlation with the observed audiometric recovery. The inability to map the exact trajectory of antibody titers against the daily improvements in decibel thresholds leaves a gap in the serological understanding of the event. Furthermore, from an epidemiological and statistical standpoint, the inherent single-case design of this report strictly precludes any broad statistical generalization regarding the universal efficacy of pulse steroids across the many diverse and highly variable SLE phenotypes. Additionally, the unavoidable lack of pre-morbid baseline audiometry for this specific patient makes it exceedingly difficult to ascertain if subclinical, high-frequency auditory deficits pre-existed the acute ischemic attack.²⁰

To address these gaps in the current literature, future multi-center, prospective cohort studies are urgently needed. These future investigations should focus on utilizing serial high-frequency audiometry combined with advanced inner ear MRI protocols, such as heavily T2-weighted 3D-FLAIR imaging, which are specifically required to accurately map the precise incidence and anatomical distribution of subclinical inner ear vasculopathy in broader populations of SLE patients. Additionally, ongoing clinical research should deliberately aim to formalize and standardize specific pulse corticosteroid protocols based entirely on observable clinical indicators for use in situations when advanced serological biomarkers are completely unavailable in low-resource environments. Establishing these clinically based protocols will ensure that life-altering therapies are not delayed while awaiting slow or inaccessible laboratory results.

4. Conclusion

Systemic lupus erythematosus is a highly destructive systemic disease that possesses the confirmed capacity to precipitate devastating, sudden bilateral sensorineural hearing loss, a catastrophic event driven primarily by acute microvasculitis and the pathological deposition of immune complexes directly within the highly sensitive vascular networks of the inner ear. This comprehensive case report unequivocally demonstrates that, even in the alarming presence of very severe to profound bilateral deafness that is concurrently associated with high systemic disease activity—specifically quantified by a MEX-SLEDAI score of 12—complete and total audiological recovery is biologically and clinically attainable.

The critical determinant of therapeutic success in these dire neurotologic emergencies is not simply the choice of drug, but the speed of its application. The ultra-early recognition of the otologic crisis, paired with the immediate administration of high-dose intravenous corticosteroid pulse therapy, is absolutely required to rapidly halt progressive cochlear ischemia and suppress localized tissue inflammation before irreversible hair cell death occurs. Moving forward, it is essential that both otorhinolaryngologists and rheumatologists alike must maintain a perpetually high index of suspicion for the development of Autoimmune Inner Ear Disease in all lupus patients presenting with sudden, unexplained hearing changes. Cultivating this high level of clinical vigilance will ensure rapid, targeted, and aggressive intervention, requiring clinicians to act decisively by relying heavily on robust clinical assessment tools, especially when advanced diagnostic resources are geographically or financially limited. Through such rapid clinical reasoning and decisive pharmacological action, profound functional losses can be successfully reversed, preserving the patient's crucial sensory connections to the world.

5. References

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