Review

Vestibular Involvement in Systemic Autoimmune and Rheumatologic Diseases: A Systematic Review and GRADE-Based Assessment

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Review

Vestibular Involvement in Systemic Autoimmune and Rheumatologic Diseases: A Systematic Review and GRADE-Based Assessment

Running Title: Vestibular Involvement in Autoimmune Diseases

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Abstract

Background: Vestibular symptoms and objective vestibular dysfunction have been reported in patients with autoimmune and rheumatologic diseases, but available evidence remains fragmented and methodologically heterogeneous. Previous studies have often addressed audiovestibular involvement as a combined entity, limiting disease-specific interpretation of vestibular outcomes. **Methods:** A PRISMA 2020–based systematic review was conducted using predefined eligibility criteria targeting vestibular outcomes in autoimmune and systemic rheumatologic diseases. Observational studies reporting vestibular symptoms and/or objective vestibular test results were included. Vestibular data were extracted even when studies reported combined audiovestibular outcomes. Certainty of evidence was assessed using the GRADE approach. **Results:** Twenty-nine studies were included in the qualitative synthesis, comprising 18 primary observational studies and 11 reviews. Vestibular involvement was reported across multiple diseases, including systemic sclerosis, giant cell arteritis, ankylosing spondylitis, psoriatic arthritis, Behçet disease, primary Sjögren syndrome, rheumatoid arthritis, systemic lupus erythematosus, antiphospholipid syndrome, and vasculitic disorders. Objective vestibular abnormalities were most frequently identified using caloric testing, balance integration measures, videonystagmography, and video head impulse testing. Systemic sclerosis and giant cell arteritis showed more consistently reported vestibular findings, although heterogeneity in assessment methods precluded quantitative synthesis. **Conclusions:** Vestibular involvement occurs across autoimmune and systemic inflammatory diseases, but overall certainty of evidence remains limited. Standardized vestibular assessment and longitudinal studies are needed to better define disease-specific vestibular phenotypes.

Keywords: vestibular dysfunction; autoimmune diseases; rheumatologic diseases; systemic sclerosis; giant cell arteritis; benign paroxysmal positional vertigo; balance disorders; vestibular assessment; systematic review; GRADE

1. Introduction

Vestibular symptoms and objective vestibular dysfunction have been increasingly reported in patients with systemic autoimmune and rheumatologic diseases. Clinical manifestations such as vertigo, dizziness, imbalance, and benign paroxysmal positional vertigo have been described across a wide range of conditions, suggesting that immune-mediated mechanisms may affect vestibular structures either directly or indirectly. However, the available evidence remains fragmented and methodologically heterogeneous, largely due to differences in study design, patient selection, disease activity, and vestibular assessment strategies [1].

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Most previous investigations have addressed audiovestibular involvement as a combined entity, frequently prioritizing hearing-related outcomes and relegating vestibular findings to secondary or descriptive observations. As a result, vestibular outcomes are often underrecognized, inconsistently reported, or assessed using non-standardized methods, limiting disease-specific interpretation and cross-study comparability. This lack of focused vestibular evaluation hampers the ability to define consistent vestibular phenotypes and to accurately assess the strength of evidence supporting vestibular involvement in systemic immune-mediated diseases [1,2].

Autoimmune involvement of the inner ear has long been recognized as a clinical entity, classically described as autoimmune inner ear disease. Early clinical reviews reported that vestibular symptoms—including imbalance, ataxia, and episodic or positional vertigo—may be present in up to half of affected patients and frequently coexist with systemic autoimmune diseases [3]. These observations provided an initial clinical framework supporting immune-mediated mechanisms affecting both cochlear and vestibular structures, although objective vestibular testing was not systematically applied at that time [3].

A further limitation is that most available syntheses have addressed inner ear involvement using a combined audiovestibular approach, analyzing hearing and balance disorders together, often pooling heterogeneous diseases and outcomes. This has been useful to map the overall otologic spectrum but has also diluted vestibular-specific information, particularly when objective vestibular testing was variably applied or inconsistently reported [1,2,5,6]. Consequently, comparisons across diseases remain difficult and are rarely supported by standardized vestibular endpoints.

Within this heterogeneous field, vestibular involvement has been reported in multiple autoimmune and systemic inflammatory conditions, including systemic sclerosis, giant cell arteritis, ankylosing spondylitis, psoriatic arthritis, Behçet disease, primary Sjögren syndrome, rheumatoid arthritis, systemic lupus erythematosus, antiphospholipid syndrome, Anti-Neutrophil Cytoplasmic Antibodies (ANCA)-associated vasculitis, sarcoidosis, and Cogan syndrome [7–19,22,24–30]. However, the strength and consistency of available evidence vary widely between diseases, and the majority of primary studies are observational.

Among systemic autoimmune diseases, systemic sclerosis has emerged as the condition with the most consistent evidence of vestibular involvement. Observational studies have demonstrated objective vestibular abnormalities in systemic sclerosis, including altered caloric responses, impaired sensory integration on balance testing, and an increased prevalence of benign paroxysmal positional vertigo [7,8]. A recent systematic review and meta-analysis confirmed that vertigo is significantly more prevalent in patients with systemic sclerosis compared with control populations, although objective vestibular outcomes could not be quantitatively pooled due to methodological heterogeneity [9].

Vestibular involvement has also been reported in other systemic inflammatory and rheumatologic diseases. In giant cell arteritis, independent studies have described a strong association with benign paroxysmal positional vertigo, supporting a vascular or ischemic mechanism affecting vestibular end organs [10,22], with partial symptom improvement after corticosteroid therapy reported in some cases [24]. In ankylosing spondylitis and psoriatic arthritis, case–control studies have demonstrated impaired vestibular sensory integration and an increased prevalence of positional vertigo compared with healthy controls [11,12].

More recently, objective vestibular abnormalities have been identified in diseases traditionally considered less frequently associated with vestibular dysfunction. In Behçet's disease, video head impulse testing and head-shake maneuvers have revealed horizontal canal dysfunction and increased rates of head-shake–induced nystagmus [13]. In primary Sjögren syndrome, subclinical vestibular involvement has been demonstrated using vestibular evoked myogenic potentials and video head impulse testing [14]. Similarly, in rheumatoid arthritis, large case–control studies using videonystagmography and caloric testing have reported both central-type oculomotor abnormalities and peripheral vestibular hypofunction [15].

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Vestibular manifestations have additionally been described in systemic lupus erythematosus, antiphospholipid syndrome, ANCA-associated vasculitis, sarcoidosis, and Cogan syndrome, although the evidence in these conditions remains heterogeneous and largely observational [16–19]. In thrombotic autoimmune conditions such as antiphospholipid syndrome, recurrent vertigo has been linked to thrombotic and microvascular mechanisms involving the inner ear circulation, a finding supported by recent systematic reviews [17,30]. Finally, autoimmune comorbidity has also been described in Ménière's disease cohorts, raising questions about immune contributions to recurrent vertigo syndromes in selected patients. An association between Ménière's disease and systemic autoimmune disorders has been supported by large observational studies. In a multicenter cohort including nearly 700 patients with Ménière's disease, Gázquez et al. reported a significantly increased prevalence of systemic autoimmune diseases, particularly rheumatoid arthritis, systemic lupus erythematosus, and ankylosing spondylitis, compared with the general population. Importantly, autoimmune diseases were more frequently observed in patients with Ménière's disease and migraine, suggesting the existence of a distinct immune-mediated clinical phenotype. These findings support an autoimmune background in a subset of patients with Ménière's disease and provide a broader context for immune-mediated vestibular disorders [23].

The aim of the present systematic review was to synthesize and critically appraise the available evidence on vestibular involvement in the most prominent and highest scientific quality of studies in systemic autoimmune, autoinflammatory, and rheumatologic diseases, focusing specifically on vestibular outcomes rather than combined audiovestibular manifestations. Secondary objectives were to compare vestibular findings across diseases and to assess certainty of evidence using the GRADE framework.

2. Materials and Methods

This systematic review was conducted and reported in accordance with the PRISMA 2020 statement [20].

2.1. Search Strategy

A comprehensive literature search was performed in PubMed/MEDLINE, Embase, Web of Science, and the Cochrane Library from database inception to the final search date (December 2025). The search strategy combined controlled vocabulary and free-text terms related to vestibular involvement and autoimmune diseases, including: *vestibular*, *vertigo*, *dizziness*, *balance*, *autoimmune*, *rheumatologic*, *systemic sclerosis*, *vasculitis*, *giant cell arteritis*, *ankylosing spondylitis*, *psoriatic arthritis*, *Behçet*, *Sjögren*, *rheumatoid arthritis*, and *systemic lupus erythematosus*. Reference lists of relevant reviews and included articles were manually screened to identify additional eligible studies [1,2,6].

2.2. Eligibility Criteria

Studies were included if they met the following criteria:

(1) observational design (cohort, case–control, or cross-sectional);

(2) evaluation of patients with autoimmune, autoinflammatory, or systemic rheumatologic diseases;

(3) reporting vestibular symptoms and/or objective vestibular test results;

(4) availability of extractable vestibular data.

Studies were excluded if they:

(1) focused exclusively on auditory outcomes;

(2) were case reports, conference abstracts, editorials, or animal studies;

(3) lacked sufficient methodological detail;

(4) reported overlapping patient cohorts without novel vestibular data.

When studies reported combined audiovestibular outcomes, only vestibular-related data were extracted and analyzed, in line with previous methodological recommendations [1,2].

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2.3. Study Selection

After duplicate removal, titles and abstracts were independently screened for eligibility. Full-text articles were retrieved and assessed when abstracts suggested potential relevance. Discrepancies were resolved by consensus. The study selection process is summarized in the PRISMA flow diagram (Figure 1) [20].

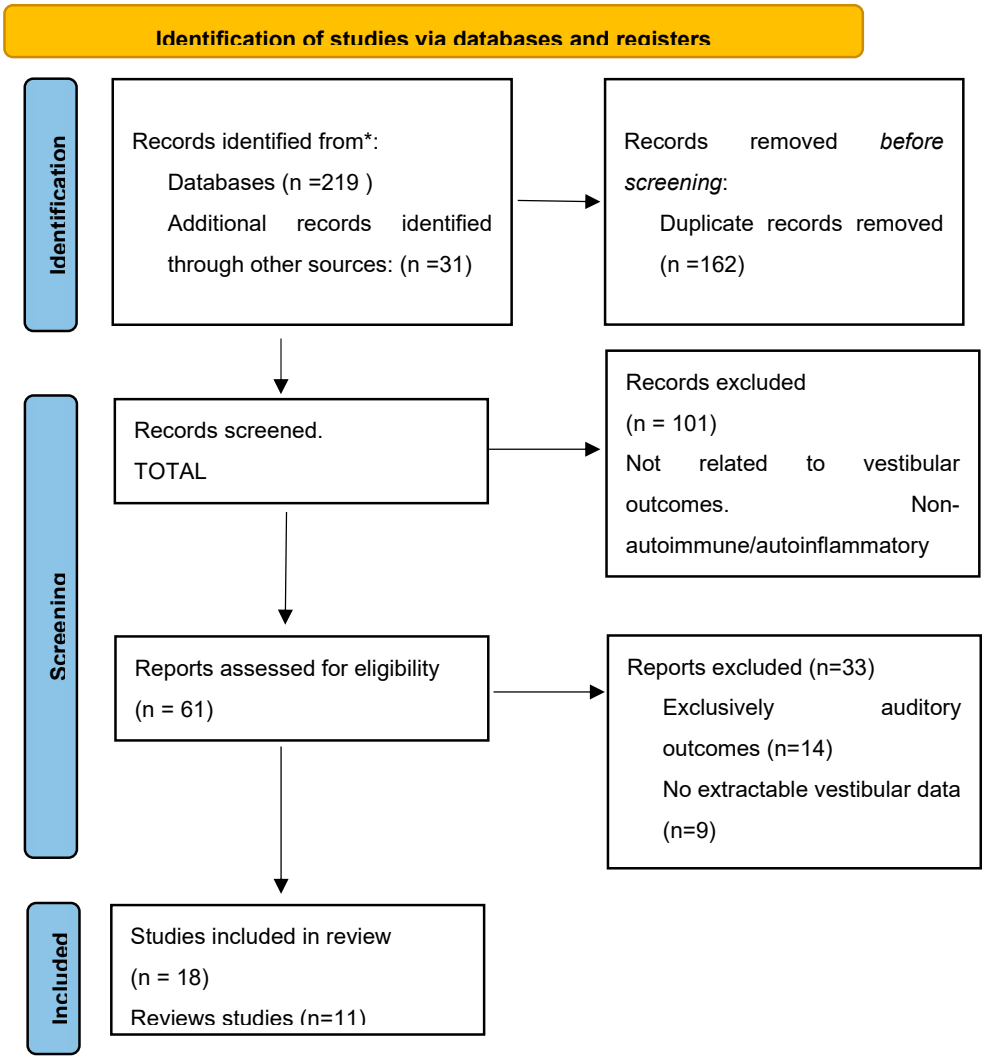

Figure 1. PRISMA 2020 flow diagram illustrating study selection for the systematic review of vestibular involvement in autoimmune and systemic rheumatologic diseases.

2.4. Data Extraction

For each included primary study, the following data were extracted: first author, year of publication, disease category, study design, sample size, vestibular assessment methods, vestibular outcomes, and main findings. Reviews were analyzed separately to contextualize the primary evidence. Data extraction was performed using a standardized form and cross-checked for accuracy.

2.5. Risk of Bias Assessment

Risk of bias was qualitatively assessed for primary studies based on study design, sample size, selection criteria, and completeness of vestibular outcome reporting, following principles commonly applied in observational studies of vestibular disorders [5,20].

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2.6. Certainty of Evidence

The certainty of evidence for vestibular involvement across diseases was evaluated using the GRADE framework [21]. Evidence derived from observational studies was initially rated as low certainty and subsequently downgraded or upgraded based on risk of bias, inconsistency, indirectness, imprecision, and other considerations.

Given the absence of randomized or longitudinal vestibular studies in this field, highest scientific quality was interpreted as well-designed observational studies and systematic reviews.

3. Results

3.1. Study Selection

The systematic literature search identified a total of 29 eligible studies for qualitative synthesis after removal of duplicates and application of predefined inclusion and exclusion criteria (Figure 1). These comprised 18 primary observational studies addressing vestibular involvement in specific diseases and 11 reviews providing clinical, epidemiological, or mechanistic context. No randomized controlled trials were identified. All included studies were published in peer-reviewed journals.

3.2. Characteristics of Included Studies

The characteristics of the 18 primary observational studies are summarized in Table 1. Most studies employed a case–control or cross-sectional design and included adult patients with established diagnoses of systemic autoimmune or rheumatologic diseases. Sample sizes varied widely across studies, ranging from small cohorts to larger disease-specific populations.

Table 1. Primary studies reporting vestibular involvement in autoimmune and rheumatologic diseases.

Author (Year)	Disease	Study Design	Participants (Patients/Controls)	Vestibular Assessment	Vestibular Outcomes	Main Vestibular Findings
Amor-Dorado et al. (2003)	Giant cell arteritis	Prospective cohort	44 / 44	Positional tests, caloric test	Vertigo, peripheral vestibulopathy	Vestibular dysfunction significantly more frequent than in controls
Amor-Dorado et al. (2004)	Giant cell arteritis	Case–control	44 / 44	Dix–Hallpike maneuver	BPPV	Strong association between GCA and BPPV
Amor-Dorado et al. (2008a)	Limited systemic sclerosis	Case–control	35 / 59	Caloric test, CTSIB, oculography	Chronic imbalance	Objective vestibular abnormalities significantly more frequent
Amor-Dorado et al. (2008b)	Systemic sclerosis	Case–control	42 / 74	Dix–Hallpike, CTSIB	BPPV, postural instability	Increased prevalence of BPPV and abnormal sensory integration
Amor-Dorado	Ankylosing spondylitis	Case–control	59 / 46	Caloric test, CTSIB	Dizziness, imbalance	Peripheral vestibular hypofunction

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et al. (2011a)						and impaired balance integration
Amor-Dorado et al. (2011b)	Ankylosing spondylitis	Case–control	59 / 46	Dix–Hallpike, CTSIB	BPPV	Higher BPPV prevalence than in controls
Amor-Dorado et al. (2014)	Psoriatic arthritis	Case–control	60 / 60	Caloric test, CTSIB	Dizziness, imbalance	Significant vestibular dysfunction compared with controls
Amor-Dorado et al. (2017)	Psoriatic arthritis	Case–control	60 / 60	Oculography, CTSIB, CDP	Postural instability	Objective abnormalities in balance control
Gázquez et al. (2011)	Ménière's disease + autoimmune disease	Multicenter cohort	>600 / –	Clinical criteria (AAO–HNS)	Episodic vertigo	Autoimmune comorbidity associated with more persistent vertigo
Karataş et al. (2007)	Systemic lupus erythematosus	Observational	28 / –	Clinical–vestibular assessment	Vertigo, dizziness	High frequency of vestibular symptoms
Ertugrul et al. (2019)	Behçet disease	Prospective case–control	31 / 31	vHIT, head-shake test, DHI	Peripheral vestibular dysfunction	Horizontal canal dysfunction and head-shake–induced nystagmus significantly more frequent
Yazici et al. (2001)	Behçet disease	Observational	29 / –	Clinical assessment	Vertigo	Vestibular symptoms reported as part of systemic involvement
Ziavra et al. (2010)	Rheumatoid arthritis	Case–control	25 / 20	Caloric test	Dizziness	Peripheral vestibular hypofunction more frequent than in controls
Özkırış et al. (2014)	Rheumatoid arthritis	Prospective case–control	81 / 81	VNG, caloric test, positional tests	Central and peripheral vestibular dysfunction	VNG abnormalities in 38.3%; canal paresis in 13.6%

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Ulusoy et al. (2022)	Primary Sjögren syndrome	Case–control	35 / 35	vHIT, cVEMP, oVEMP	Subclinical vestibular dysfunction	Reduced vHIT gain and abnormal VEMP latencies
Kim et al. (2012)	ANCA-associated vasculitis	Cohort	32 / –	Caloric test	Imbalance	Vestibular dysfunction associated with disease activity
Kariya et al. (2014)	Sarcoidosis	Observational	18 / –	Clinical ± VNG	Vertigo	Peripheral inflammatory vestibular involvement
Greco et al. (2016)	Cogan syndrome	Cohort	25 / –	VNG	Severe vertigo	Immune-mediated vestibulopathy
Hupper t et al. (2013)	Antiphospholipid syndrome	Case–control	22 / 22	Clinical assessment	Recurrent vertigo	Vestibular symptoms more frequent than in controls

Risk of bias assessment: Overall, a moderate risk of bias was identified across most primary studies, reflecting their observational design. A high risk of bias was observed in the studies by Karataş et al. [16], Yazici et al. [28], and Kariya et al. [30]. AAO–HNS, American Academy of Otolaryngology–Head and Neck Surgery; BPPV, benign paroxysmal positional vertigo; CDP, computerized dynamic posturography; CTSIB, Clinical Test of Sensory Interaction and Balance; DHI, Dizziness Handicap Inventory; vHIT, video head impulse test; VNG, videonystagmography; VEMP, vestibular evoked myogenic potentials.

The 11 review articles included narrative reviews, systematic reviews, and one systematic review with meta-analysis. These reviews addressed vestibular involvement either directly or as part of broader audiovestibular or immune-mediated inner ear frameworks and are summarized in Table 2.

Table 2. Reviews addressing vestibular involvement in autoimmune and systemic rheumatologic diseases.

Author (Year)	Type of Review	Diseases Included	Vestibular Scope	Main Contribution
Ralli et al. (2018)	Narrative clinical review	Systemic autoimmune diseases (SLE, RA, Sjögren, Behçet, vasculitis, Cogan syndrome, sarcoidosis)	Clinical vestibular symptoms	Highlights frequency and underrecognition of vestibular symptoms in systemic autoimmune diseases
Girasoli et al. (2018)	Narrative clinical review	Autoimmune disorders	Clinical vestibular syndromes	Provides an overview of immune-mediated vertigo and diagnostic considerations
Bovo et al. (2006)	Narrative clinical review	Autoimmune inner ear disease (AIED)	Vertigo, imbalance, episodic and positional vertigo	Classical clinical description of AIED, emphasizing frequent vestibular symptoms and coexistence with systemic autoimmune diseases

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Amor-Dorado et al. (2009)	Narrative clinical review	Systemic vasculitides (GCA, Takayasu arteritis, PAN, ANCA-associated vasculitis, Behçet disease, Cogan syndrome, RA, SSc, SLE, Sjögren syndrome)	Vertigo, nystagmus, BPPV	Early comprehensive synthesis linking vasculitis and vestibular involvement, particularly in giant cell arteritis
Breslin et al. (2020)	Systematic review	Autoimmune inner ear disease	Vestibular outcomes (secondary)	Focused mainly on hearing outcomes; vestibular data inconsistently reported
Gázquez et al. (2011)	Narrative review / cohort synthesis	Ménière's disease with autoimmune comorbidity	Recurrent vertigo	Demonstrates increased prevalence of systemic autoimmune diseases in Ménière's disease
Athanasopoulos et al. (2024)	Narrative mechanistic review	Autoimmune and autoinflammatory diseases	Mechanistic (non-clinical)	Discusses immune-inflammatory and vascular mechanisms affecting the inner ear
Miwa & Okano (2022)	Narrative mechanistic review	Autoimmune inner ear disease	Experimental / translational (non-clinical)	Demonstrates macrophage-mediated immune mechanisms in the inner ear; supports biological plausibility of vestibular involvement
Li et al. (2025)	Narrative mechanistic review	Autoimmune inner ear disorders	Mechanistic (non-clinical)	Reviews immune microenvironment and inflammatory pathways in inner ear disease
Salvador et al. (2025)	Systematic review and meta-analysis	Systemic sclerosis	Clinical vestibular outcomes	Reports increased prevalence of vertigo in systemic sclerosis; objective vestibular findings highly heterogeneous
Chen et al. (2024)	Systematic review	Antiphospholipid syndrome	Clinical vestibular outcomes	Summarizes evidence linking APS with recurrent vertigo and vestibular dysfunction

AIED, autoimmune inner ear disease; APS, antiphospholipid syndrome; BPPV, benign paroxysmal positional vertigo; GCA, giant cell arteritis; PAN, polyarteritis nodosa; RA, rheumatoid arthritis; SLE, systemic lupus erythematosus; SSc, systemic sclerosis.

3.3. Vestibular Involvement Across Autoimmune and Rheumatologic Diseases

Vestibular symptoms and/or objective vestibular abnormalities were reported across a wide spectrum of diseases, including systemic sclerosis, giant cell arteritis, ankylosing spondylitis, psoriatic arthritis, Behçet disease, primary Sjögren syndrome, rheumatoid arthritis, systemic lupus erythematosus, antiphospholipid syndrome, ANCA-associated vasculitis, sarcoidosis, and Cogan syndrome.

Across studies, vestibular involvement was evaluated using a combination of clinical symptom assessment (vertigo, dizziness, imbalance, and benign paroxysmal positional vertigo) and objective vestibular testing, most commonly caloric testing, videonystagmography (VNG), clinical tests of sensory integration and balance, and video head impulse testing (vHIT). The choice of vestibular assessment methods varied substantially between studies, limiting direct comparisons.

In systemic sclerosis, multiple independent studies reported a higher prevalence of vertigo, balance impairment, and benign paroxysmal positional vertigo compared with control populations.

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Objective abnormalities were frequently identified using caloric testing and balance integration measures. These findings were consistently reported across different cohorts.

In giant cell arteritis, vestibular involvement was predominantly characterized by an increased prevalence of benign paroxysmal positional vertigo confirmed by positional testing. Although objective vestibular testing beyond positional maneuvers was limited, findings were reproducible across independent observational studies.

In ankylosing spondylitis and psoriatic arthritis, vestibular involvement was mainly reflected by impaired balance performance and increased rates of positional vertigo. Studies frequently relied on clinical balance tests and posturography, with fewer studies incorporating semicircular canal–specific vestibular assessments.

In Behçet disease, objective vestibular abnormalities were identified using vHIT and oculomotor or positional maneuvers, suggesting peripheral semicircular canal involvement in selected cohorts. In primary Sjögren syndrome, vestibular abnormalities were often subclinical and detected through physiological vestibular testing rather than overt vestibular symptoms.

In rheumatoid arthritis, vestibular findings were heterogeneous, with studies reporting both peripheral vestibular hypofunction and central-type oculomotor abnormalities. Similar heterogeneity was observed in systemic lupus erythematosus, where vestibular involvement was primarily symptom-based and objective testing was inconsistently applied.

Vestibular involvement in antiphospholipid syndrome, ANCA-associated vasculitis, sarcoidosis, and Cogan syndrome was reported in a limited number of studies, typically small observational cohorts or case series, precluding robust disease-specific comparisons.

3.4. Certainty of Evidence (GRADE Assessment)

Certainty of evidence for vestibular involvement was assessed using the GRADE framework, considering the overall body of evidence available for each disease (Table 3). For systemic sclerosis and giant cell arteritis, certainty of evidence was rated as moderate at best, within the limitations inherent to observational study designs and heterogeneous vestibular assessment methods.

Table 3. Certainty of evidence for vestibular involvement assessed using the Grading of Recommendations Assessment, Development and Evaluation (GRADE) approach. Certainty ratings were assigned by disease, based on the overall body of relevant evidence available for each autoimmune or rheumatologic condition.

Disease	Vestibular Outcomes	Evidence Base	Main Limitations	Certainty of Evidence (GRADE)
Systemic sclerosis	Vertigo, balance impairment, BPPV	Multiple observational case–control studies; systematic review and meta-analysis	Observational designs; heterogeneous vestibular testing; lack of longitudinal data	**Moderate at best, within observational limitations**
Giant cell arteritis	BPPV, vertigo	Observational cohort and case–control studies	Small sample sizes; limited objective vestibular testing	**Moderate at best, within observational limitations**
Ankylosing spondylitis	Balance impairment, BPPV	Observational case–control studies	Non-specific balance endpoints; heterogeneous methods	**Low**
Psoriatic arthritis	Balance impairment	Observational case–control studies	Small cohorts; limited vestibular specificity	**Low**

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Behçet disease	Vestibular hypofunction, abnormal vHIT	Observational studies	Limited number of studies; inconsistent outcomes	**Low**
Primary Sjögren syndrome	Subclinical vestibular abnormalities	Single observational study	Small sample size; lack of replication	**Low**
Rheumatoid arthritis	Peripheral and central vestibular abnormalities	Observational studies	Conflicting findings; heterogeneous testing	**Low**
Systemic lupus erythematosus	Vertigo, vestibular dysfunction	Observational studies	Symptom-based outcomes; limited objective data	**Low**
Antiphospholipid syndrome	Recurrent vertigo	Observational studies; systematic review	Sparse primary data; indirect vestibular outcomes	**Low**
ANCA-associated vasculitis	Vestibular dysfunction	Small observational series	Very limited evidence	**Very low**
Sarcoidosis	Vestibular involvement	Case series	Rare condition; non-comparative data	**Very low**
Cogan syndrome	Vestibular dysfunction	Observational studies	Rare disease; mixed audiovestibular outcomes	**Very low**

BPPV, benign paroxysmal positional vertigo; GRADE, Grading of Recommendations Assessment, Development and Evaluation; vHIT, video head impulse test.

For ankylosing spondylitis, psoriatic arthritis, Behçet disease, primary Sjögren syndrome, rheumatoid arthritis, systemic lupus erythematosus, and antiphospholipid syndrome, certainty of evidence was rated as low, primarily due to small sample sizes, methodological heterogeneity, and limited use of standardized vestibular testing. For ANCA-associated vasculitis, sarcoidosis, and Cogan syndrome, certainty of evidence was rated as very low, reflecting sparse and predominantly descriptive data.

4. Discussion

This systematic review shows that vestibular involvement has been reported across a broad range of systemic autoimmune and rheumatologic diseases, although the evidence remains heterogeneous and largely observational. By focusing on vestibular outcomes, this work addresses an important limitation of prior audiovestibular syntheses, where vestibular findings were frequently embedded within combined outcomes and assessed using non-uniform diagnostic strategies [1,2,5,6].

Across the included conditions, systemic sclerosis (SSc) and giant cell arteritis (GCA) were the diseases in which vestibular findings were most consistently reported across multiple independent primary studies, rather than supported by high-level causal evidence. In SSc, case–control studies reported higher rates of vertigo and balance impairment, as well as an increased prevalence of benign paroxysmal positional vertigo (BPPV), compared with controls [7,8]. A recent systematic review and meta-analysis supports a higher frequency of vertigo in SSc. However, objective vestibular outcomes could not be quantitatively pooled because of heterogeneous testing protocols and outcome definitions [9]. Collectively, these data suggest a reproducible association between SSc and vestibular symptoms, while emphasizing the need for standardized vestibular endpoints to enable robust comparisons and pooling.

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In GCA, observational studies reported a higher prevalence of BPPV confirmed by positional testing [10,22]. Although these findings do not establish causality, they align with narrative syntheses of systemic vasculitis proposing that ischemic involvement of labyrinthine circulation may contribute to vestibular syndromes in this setting [24]. Importantly, the certainty of evidence should be interpreted within the limitations of observational designs, and mechanistic explanations should be framed as plausible rather than definitive [24].

Evidence from spondyloarthritis-related diseases suggests a tendency toward vestibular/balance involvement. In ankylosing spondylitis and psoriatic arthritis, studies using sensory integration and balance measures, and in some cases posturography, reported higher rates of balance impairment and positional vertigo compared with controls [11,12,25,26]. While these findings are clinically relevant, differences in vestibular assessment strategies and the frequent reliance on non-specific balance endpoints limit disease-specific phenotyping and cross-study comparability. More recent studies have expanded objective vestibular assessment to additional autoimmune diseases. In Behçet disease, abnormalities detected using video head impulse testing and oculomotor/positional maneuvers suggest peripheral semicircular canal involvement in selected cohorts [13,27]. In primary Sjögren syndrome, vestibular abnormalities were often subclinical and identified through physiological testing [14]. In rheumatoid arthritis, vestibular findings were heterogeneous across studies, with both peripheral hypofunction and central-type oculomotor abnormalities reported, likely reflecting differences in patient selection and testing protocols [15,28]. Vestibular involvement has also been described in systemic lupus erythematosus, antiphospholipid syndrome, ANCA-associated vasculitis, sarcoidosis, and Cogan syndrome, but the evidence base remains sparse and methodologically diverse [16–19,21,29,30].

Ménière's disease was not considered a systemic autoimmune disorder in this review. However, it was included as a related clinical model of recurrent vertigo with documented autoimmune comorbidity. Large observational data indicate an increased prevalence of systemic autoimmune diseases among patients with Ménière's disease, supporting an autoimmune background in a subset and highlighting potential immune contributions to vertigo persistence [23]. These findings provide contextual support for immune-mediated vestibular symptom chronicity, while remaining distinct from disease-specific vestibular testing evidence.

The clinical concept of immune-mediated inner ear involvement predates modern vestibular testing and was historically framed as autoimmune inner ear disease (AIED). Classical reviews described frequent vestibular symptoms and a relevant coexistence with systemic autoimmune disorders [3]. More recently, mechanistic reviews have emphasized the role of resident macrophages and innate immune signaling within the inner ear, providing biological plausibility for immune-mediated effects on vestibular end organs, even though these data are not confirmatory of clinical vestibular dysfunction [31]. In parallel, biomarker-focused scoping work highlights the ongoing lack of sensitive and specific diagnostic immune biomarkers for routine clinical use, which may partly explain the heterogeneity of clinical phenotypes and outcomes reported across studies [32]. Recent narrative syntheses on AIED pathogenesis and treatment similarly underscore that therapeutic responses to corticosteroids or immunosuppressive agents are variable and largely supported by observational evidence, reinforcing the need for standardized vestibular phenotyping and longitudinal designs [33].

Overall, applying the GRADE framework indicates that the certainty of evidence is moderate at best for a limited subset of conditions and low to very low for most diseases, primarily due to observational study designs, limited sample sizes, and inconsistency in vestibular assessment methodologies [20]. Future studies should prioritize standardized vestibular testing protocols (including clearly defined symptom definitions and objective endpoints), longitudinal follow-up, and transparent reporting of disease activity and treatments to clarify disease-specific vestibular phenotypes and their clinical implications.

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5. Conclusions

Vestibular involvement has been reported across a wide spectrum of systemic autoimmune and rheumatologic diseases. However, the overall certainty of evidence remains limited and is predominantly based on observational data. Among the conditions reviewed, systemic sclerosis and giant cell arteritis show the most consistently reported vestibular findings across independent studies, although the certainty of evidence should be interpreted as **moderate at best and within the limitations inherent to observational designs.** For most other diseases, evidence remains low or very low due to heterogeneous methodologies, small sample sizes, and inconsistent vestibular assessment strategies. Future research should prioritize standardized vestibular testing protocols, longitudinal designs, and transparent reporting of disease activity and treatment exposure to better define disease-specific vestibular phenotypes and their clinical implications.

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