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NOTES FROM THE FIELD

Recommendations for Aligned Nomenclature of Peripheral Nervous System Disorders Across Rheumatology and Neurology

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Introduction

Effective multidisciplinary care for patients with complex conditions such as Sjögren's disease (SjD) is often hindered by inconsistent nomenclature across medical specialties. The Sjögren's Foundation guidelines development panel encountered this challenge when rheumatologists and neurologists came together to formulate guidelines for the care of patients with peripheral nervous system (PNS) disorders with SjD. To address this barrier, a standardized nomenclature was defined to improve communication across specialties for patient care and collaboration in implementing evidence-based medicine.

Evidence-based clinical practice guidelines

Since its emergence in the 1990s, evidence-based medicine has integrated the latest research findings into clinical practice by combining the best available evidence with clinicians' expertise and patients' needs and preferences, making evidence-based guidelines essential resources for informed clinical decisions, especially in areas in which data may be lacking or insufficient.[1,2] Central to the development of evidence-based clinical practice guidelines (CPGs) is the concept of implementability, which emphasizes creating guidelines that are practical and straightforward to implement.[3] Factors influencing a guideline's uptake include its intrinsic implementability, defined by characteristics that predict and promote its use in health care systems.[4,5] Language and nomenclature play a significant role; vague and unclear CPGs can hinder implementation, whereas unambiguous ones can enhance it.[5] A guideline's intrinsic implementability is particularly challenging when recommendations require alignment of language across multiple medical specialties. Mutual understanding across specialties is crucial for providing care to patients with complex conditions involving comorbidities or multisystem manifestations. SjD exemplifies this challenge.

PNS manifestations of SjD

SjD is a systemic autoimmune disease in which almost all body organs can be affected, including the PNS. In addition, SjD is characterized by exocrine gland dysfunction, resulting in dry eyes and dry mouth.[6] PNS involvement in SjD significantly impacts patient quality of life,[7] and a coordinated approach

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among different subspecialties is essential in the care of these patients.[8]

PNS involvement in SjD frequently poses significant diagnostic challenges to clinicians: it is often the first manifestation of the disease, preceding SjD diagnosis.[9–11] Certain patterns of PNS involvement, such as small fiber neuropathy (SFN), are more commonly observed in patients who are seronegative (ie, those who are negative for anti-SSA),[12] in whom the diagnosis cannot be established without a positive minor salivary gland biopsy (MSGB). This can be a diagnostic challenge in practices in which an MSGB is not routinely performed as part of the clinical evaluation.

In addition to the challenge of establishing the diagnosis of SjD, there is the challenge of characterizing the nature of PNS dysfunction. Neuropathies can be classified based on a variety of features: symptomology (ataxic or nonataxic, painful or nonpainful), anatomic pattern of involvement (polyneuropathy vs mononeuropathy vs multiple mononeuropathies, length dependent vs non–length dependent), anatomic localization (peripheral nerve, nerve root, dorsal root ganglia), nerve fiber type affected (large fiber vs small fiber, sensory and/or motor, autonomic), electrophysiology (axonal vs demyelinating), or pathophysiology/etiology (vasculitic, toxic, immune mediated). This has resulted in heterogeneous classification systems and nonuniform nomenclature that interfere with communication in the clinical care of patients and in apprising the literature, as outlined in Table 1.

Regardless of the classification method used, a basic knowledge of anatomy and pathophysiology is essential to understand the spectrum of clinical presentations of neuropathies. Damage to or dysfunction of large nerve fibers (myelinated Aβ axons mediating the sensations of proprioception, vibration, and touch) typically leads to paresthesia and ataxia. Motor nerve involvement leads to clinical or subclinical muscle weakness. Damage to the small nerve fibers (ie, thinly myelinated Aδ and unmyelinated C fibers) leads to SFN, which affects pain and temperature sensations. Therefore, SFN causes sensory symptoms, usually pain, numbness, and tingling but not muscle weakness or ataxia.

Table 1. Examples of disparate neuropathy classification in Sjögren's Disease

Study	Neuropathy patterns identified	Comments

Delalande 2004: 51 patients with SjD with peripheral nervous system involvement (more than one pattern observed in some patients)[11]	• Distal axonal sensorimotor neuropathy (19 patients)
	• Pure sensory neuropathy (nine patients)
	  ○ Symmetric axonal sensory neuropathy without motor involvement (five patients)
	  ○ Ganglionopathy with severe ataxia (four patients)
	• Multiple mononeuropathy (seven patients)
	• Chronic polyradiculoneuropathy (myeloradiculitis; one patient)
	• Cranial neuropathy (16 patients)
		The first group with 19 patients had predominantly sensory symptoms (ie, the "motor" component was observed in electromyographic studies but was not clinically apparent).

Terrier 2007: 40 patients with SjD with neuropathy who underwent muscle and nerve biopsies[13]	Classification based on types of neuropathy: • Polyneuropathy (25 patients) • Multiple mononeuropathy (11 patients) • Ganglionopathy (five patients) • Trigeminal neuropathy (one patient) Classification based on symptoms: • Pure superficial sensory • Profound with or without superficial sensory • Sensorimotor patterns Classification based on electromyographic patterns: • Sensorimotor involvement • Axonal impairment • Axonal and demyelinating impairment	Different strategies to classify these patients were used based on anatomy, symptoms, and electromyographic findings.
Sireesha 2019: 21 patients with SjD with peripheral nervous system involvement[9]	• Mononeuritis multiplex (mononeuropathy multiplex; seven patients) • Ganglionopathy with sensory ataxia (four patients) • Length-dependent sensorimotor neuropathy (two patients) • Painful small fiber neuropathy (one patient) • Autonomic neuropathy (two patients) • Trigeminal neuropathy (two patients) • Cranial neuropathy (two patients)	• Authors described different "phenotypic patterns of neuropathy." • Unexpectedly, mononeuritis multiplex was the most common phenotype observed.
Mori 2005: 92 patients with SjD with associated neuropathy[10]	• Sensory ataxic neuropathy (36 patients) • Painful sensory neuropathy without sensory ataxia (18 patients) • Multiple mononeuropathy (11 patients) • Radiculoneuropathy (four patients) • Autonomic neuropathy (three patients) • Trigeminal neuropathy (15 patients) • Multiple cranial neuropathy (five patients)	• The term "sensorimotor polyneuropathy" was not used in this series despite common usage in related literature. • The term ganglionopathy was not used in classification but was discussed in detail in the Discussion section.

Table 2. Peripheral neuropathies in Sjögren's disease*

Neuropathy type	Description
Mononeuropathy	
Other nomenclature	Neuropathy; focal neuropathy
Definition	Mononeuropathy refers to dysfunction or disorder of a single nerve. This is in contrast to the more diffuse dysfunction seen in polyneuropathy.
Symmetry	Involvement of only one nerve would result in an asymmetric presentation.
Presentation	Patients will present with sensory and/or motor symptoms and signs in the distribution of a single nerve. Sensory symptoms can include negative (numbness) or positive (tingling paresthesias or pins and needles) symptoms or neuropathic pain. Motor symptoms would be weakness or loss of muscle bulk. On examination, sensory loss should be restricted to the cutaneous distribution of the single nerve, and weakness or atrophy should be found in muscles innervated by the affected nerve. If the nerve mediates a deep tendon reflex, that reflex may be reduced or absent. A Tinel sign may be elicited by tapping on the affected nerve, which would result in electric or pins and needles paresthesias in the cutaneous distribution of the nerve. Signs and symptoms can affect the face if a cranial nerve, such as the facial or trigeminal, is involved.
Etiologies/differential diagnosis	Mononeuropathy can be mechanical due to compression or entrapment, such as in median neuropathy at the wrist seen in carpal tunnel syndrome, which is the most common mononeuropathy.[14] Infection, inflammation, trauma, ischemia, or vasculitis are other etiologies.
Pathophysiology	This varies based on the etiology and can be axonal or demyelinating.
Evaluation	Electrodiagnostic studies (nerve conduction studies [NCS] and electromyography [EMG]) can confirm the clinical suspicion of a mononeuropathy and help to localize the lesion along the course of the nerve. Imaging, with ultrasound or magnetic resonance imaging (MRI), may be helpful to evaluate for a structural cause.
Large fiber (axonal) neuropathy	
Other nomenclature	Sensory polyneuropathy (pure sensory axonal neuropathy of the distal nerves); sensory motor polyneuropathy (axonal sensorimotor polyneuropathy)
Definition	Large fiber neuropathy is a peripheral neuropathy primarily affecting sensory nerves. Motor nerves, which are also large fiber, can be involved, resulting in a sensory and motor (or sensorimotor) polyneuropathy. In large fiber neuropathy, there is dysfunction of Aβ fibers, which are myelinated fibers involved in proprioception, vibration, and touch sensations. Motor neuropathies or motor neuronopathies can rarely be seen in Sjögren's disease.
Symmetry	Usually symmetric[15]
Presentation	Large fiber neuropathy results from the dysfunction or damage of Aβ fibers, which mediate the sensations of proprioception, vibration, and touch. Abnormal proprioception may result in problems with balance and an ataxic gait (wide-based, unsteady).
Etiologies/differential diagnosis	Large fiber neuropathy may occur idiopathically or due to immune-mediated, metabolic, hereditary, infectious, or toxic etiologies.
Pathophysiology	Damage to Aβ axons
Evaluation	Electrodiagnostic studies (NCSs and EMG) should be performed. Consider the following: • Fat pad biopsy can assess for an amyloid or other infiltrative process. • Nerve biopsy can assess for vasculitis, neoplasm, amyloid, or other infiltrative process if the index of suspicion is high for these processes. • Lumbar puncture should be reserved for assessing suspected cases of inflammatory demyelinating polyradiculoneuropathy (including AIDP and CIDP), ganglionopathy, neoplastic diseases, or infection.
Small fiber neuropathy[16]	
Other nomenclature	Small fiber polyneuropathy; small fiber sensory neuropathy
Definition	Small fiber neuropathy is a peripheral neuropathy affecting small nerve fibers—thinly myelinated Aδ and unmyelinated C nerve fibers.
Symmetry	Small fiber neuropathy is usually symmetric and length dependent but can present in a patchy or asymmetric manner.[15]
Presentation	Small fiber neuropathy typically presents with pain, burning, numbness, and tingling in a stocking-glove distribution. Symptoms usually begin starting in the feet and can ascend. Examination will show diminished pain and temperature sensations in the distal limbs. Less frequently, there can be early proximal or patchy evolution. Because large fibers that mediate proprioceptive (balance) and motor functions are not involved, these patients should not have ataxia or muscle weakness.[17] Fibers of the peripheral autonomic nervous system are also small caliber fibers, and small fiber neuropathy can affect these autonomic fibers, leading to autonomic dysfunction. This will result in autonomic symptoms that otherwise can be difficult to localize.[18]
Etiologies/differential diagnosis	A clear etiologic explanation is commonly not identified even after extensive laboratory testing. However, testing is aimed at uncovering potential immune-mediated, metabolic, hereditary, infectious, or toxic etiologies.
Pathophysiology	Small fiber neuropathy is due to either dysfunction or loss of small Aδ and C fibers. These fibers convey pain and temperature sensations. Injury thus results in either sensory loss/impairment or

(Continued)

Table 2. (*Cont'd*)

Neuropathy type	Description
	abnormal sensation. Mechanisms resulting in painless sensory loss vs painful abnormal sensation are an area of ongoing investigation. Distal axonal loss (length dependent) or proximal dorsal root ganglion degeneration (non–length dependent) can both lead to small fiber neuropathy symptoms.
Evaluation	The case definition or diagnostic criteria of small fiber neuropathy is not settled. Electrodiagnostic studies (NCSs and EMG) are helpful to exclude subclinical large fiber involvement. Skin biopsy is helpful to demonstrate a reduction in intradermal nerve fiber density compared to reference populations.[19] The test result provides a statistical diagnosis but does not assess small fiber nerve function. There are a number of other neurophysiologic test modalities, such as quantitative sensory testing, quantitative sudomotor axon reflex test (QSART), and sympathetic skin response, which are used to assess small fiber function, each with advantages and disadvantages.
Demyelinating polyradiculoneuropathy[20]	
Other nomenclature	Demyelinating polyneuropathy (including chronic inflammatory demyelinating polyradiculoneuropathy [CIDP])
Definition	Demyelinating polyradiculoneuropathy is a polyneuropathy that occurs with damage to the myelin sheath. A polyradiculoneuropathy indicates impairment at the level of sensory and motor roots as well as their distal peripheral nerve segments.
Symmetry	Demyelinating polyradiculoneuropathy may be symmetric but can be focal or multifocal.[15]
Presentation	The presentation can be progressive or relapsing and remitting. Sensory symptoms can include numbness, burning pain, throbbing, or dysesthesias. Motor complaints are weakness and loss of muscle bulk. Characteristic features of the examination include motor findings of weakness and muscle atrophy and sensory deficits in vibration and proprioception with loss of reflexes. Patients with demyelinating polyradiculoneuropathy will most commonly have a combination of proximal and distal findings on neurologic examination and electrodiagnostic testing. This proximal involvement often distinguishes demyelinating polyradiculoneuropathy from the more common length-dependent polyneuropathies that have only distal involvement initially.
Etiologies/differential diagnosis	CIDP is immune-mediated.
Pathophysiology	By definition, demyelinating neuropathies affect the large myelinated motor (Aα) and sensory (Aβ) and small myelinated (Aδ) fibers of nerves. Whether the neuropathy is acute or chronic, the demyelinating neuropathies are usually accompanied by damage to the axon. The extent of this axonal loss usually correlates with the degree of fixed clinical deficit. Thus, demyelinating neuropathies are often mixed disorders.
Evaluation	Electrodiagnostic studies (NCSs and EMG) play a crucial role in making this determination. It should be recognized that electrodiagnostic testing performed in the first weeks of presentation may not show all characteristics but can provide useful information to guide treatment early on. Findings of significant slowing indicate a primarily demyelinating process, but the electrodiagnostic picture can sometimes be difficult to interpret. Lumbar puncture is often performed to demonstrate increased protein in the cerebrospinal fluid in CIDP.
Ganglionopathy	
Other nomenclature	Sensory neuronopathy of the dorsal root ganglion
Definition	Ganglionopathy is a pure sensory neuronopathy that is caused by dorsal root ganglia injury or irritation.
Symmetry	This disorder most often presents in a non–length-dependent manner. Ganglionopathies can be symmetric or asymmetric.[15,21]
Presentation	The dorsal root ganglion is a pure sensory structure that contains the cell bodies of large and small fiber sensory neurons. The precise nature of the symptoms is related to which sensory modality-specific nerve cell bodies have been affected. Ataxia or severe incoordination is reflective of large fiber sensory nerve cell body injury. Injury to the cell bodies of small fiber nerves results in impaired pain and temperature sensation. Patients may complain of weakness, but by definition, strength is preserved. The weakness symptom results from proprioceptive deficits. There may be autonomic dysfunction associated because conditions that cause sensory ganglionopathy can also affect the autonomic nervous system.
Etiologies/differential diagnosis	Causes of ganglionopathy are varied; however, the likelihood of identifying an immune or paraneoplastic disorder is higher in ganglionopathies than with most length-dependent neuropathies. Drug-related, nutraceutical toxicity and infectious agents should all be considered in the right clinical context.[21,22]
Pathophysiology	The dorsal root ganglia contain cell bodies and have a fenestrated blood supply, resulting in a relatively leakier blood–nerve barrier, thereby making these cells more susceptible to injury. The exact injury to the cell bodies depends on the etiology, and much remains to be learned about the exact pathomechanisms.[22]
Evaluation	Electrodiagnostic studies (NCSs and EMG) should be performed. Motor NCSs and needle EMG should be normal. Sensory nerve responses in both the upper and lower extremities are typically absent. MRI of the spinal cord may show hyperintense T2-weighted lesions of the posterior

(Continued)

Table 2. (*Cont'd*)

Neuropathy type	Description
	columns due to the degeneration of the dorsal root ganglia's central projections in the gracile and cuneate fasciculi. Excisional biopsy of dorsal root ganglion with histologic analysis is rarely performed. Sural nerve biopsy may show axon loss, but this finding is not specific to ganglionopathy and typically does not clarify etiology. Lumbar puncture could be considered.
Vasculitic neuropathy	
Other nomenclature	Multiple mononeuropathy; mononeuritis multiplex
Definition	Vasculitic neuropathies refer to neuropathies associated with histologic evidence of a vasculitic process involving the peripheral nerves, leading to immune-mediated injury of nerve blood vessels and subsequent ischemic injury, and are associated with several clinical and electrophysiologic presentations. These neuropathies can occur in isolation as well as part of systemic vasculitis. Multiple mononeuropathy is the pattern most closely associated with vasculitic neuropathy. Vasculitic neuropathy is a peripheral neuropathy that affects large and small fibers in sensory and motor nerves.
Symmetry	Typically, vasculitic neuropathies are asymmetric at the onset, though with time, patients who do not receive treatment can become confluent, giving the impression of a symmetric polyneuropathy.
Presentation	Vasculitic neuropathy is typically acute or subacute and usually painful. Asymmetric foot or wrist drop is a common initial presentation.
Etiologies/differential diagnosis	By definition, vasculitic neuropathy implies an immune-mediated disorder. Other causes of asymmetric neuropathies not due to immune causes should be excluded.
Pathophysiology	Immune-mediated inflammation involving the vasa nervorum leads to ischemic injury of the peripheral nerves and occurs as part of a systemic inflammatory process or as an isolated process confined to the nerves.
Evaluation	Electrodiagnostic testing of right and left motor and sensory nerves should demonstrate markedly asymmetric responses. Nerve and muscle biopsy should be obtained to provide pathologic evidence of blood vessel inflammation.[13,23] Serologic testing can be used to support evidence of systemic involvement.
Autonomic nervous system neuropathy	
Other nomenclature	Autonomic neuropathy; autonomic ganglionopathy
Definition	Autonomic nervous system neuropathy is a form of polyneuropathy that affects the autonomic nervous system, and its regulation of functions is mediated by the parasympathetic and sympathetic nervous systems.
Symmetry	N/A
Presentation	Organ systems involved include the cardiovascular, gastrointestinal, genitourinary, thermoregulatory, and secretory. Symptoms can include hypotension, tachycardia, constipation, bloating, early satiety, nocturnal diarrhea, sexual dysfunction, bladder dysfunction, photosensitivity, impaired vision, anhidrosis, and sicca symptoms.[24,25]
Etiologies/differential diagnosis	Acute etiologies include autoimmune disorders (Sjögren's disease, celiac disease), paraneoplastic syndromes, autoimmune autonomic ganglionopathy, Guillain–Barré syndrome, infection, toxins, and medications/chemotherapy.[26–28] Chronic etiologies include diabetes, amyloidosis, and being hereditary.
Pathophysiology	The pathophysiology remains unclear; some cases appear to be due to an autonomic ganglionopathy, others from peripheral autonomic nerve damage, and there is a report of a T cell–mediated process.
Evaluation	Screening questions and questionnaires should be performed, and the following should be considered: • Tilt table test • The QSART • Measures of heart rate variability • Assessment of blood pressure changes, including the Valsalva maneuver • Gastrointestinal motility testing • Urodynamic testing • Skin biopsy to assess sweat gland nerve fiber density • Laboratory testing, such as vitamin B12, hemoglobin A1c, serum immunofixation, dysautonomia autoantibody panel (ganglionic acetylcholine receptor and paraneoplastic autoantibodies), to exclude other potential causes.

* Peripheral nervous system involvement in Sjögren's disease is common, occurs in several forms, and may be underdiagnosed.[29] In a series of 92 patients with Sjögren's disease–related neuropathies, 39% had sensory neuropathy, 20% had SFN, 16% had trigeminal neuropathy, 12% had multiple mononeuropathies, 5% had multiple cranial neuropathies, 4% had polyradiculoneuropathies, and 3% had autonomic neuropathies.[10,30] Some authors estimate that among all patients with Sjögren's disease, 5% have sensory neuropathy and 5% to 10% have an SFN.[30] Although less frequent than other forms of peripheral neuropathies, sensory ganglionopathy tends to be fulminant, resulting in greater disability early on. Patients with Sjögren's disease can develop more than one type of peripheral neuropathy, and peripheral neuropathies stemming from other etiologies that are common in the general population can also occur. Therefore, involvement of a neurologist in the care of patients with Sjögren's disease early when neuropathy is suspected is ideal. N/A, not applicable; SFN, small fiber neuropathy. AIDP. Acute inflammatory demyelinating polyradiculoneuropathy. CIDP. Chronic inflammatory demyelinating polyradiculoneuropathy. *Source*: Reprinted with permission from the Sjögren's Foundation.

Electrodiagnostic studies are invaluable tools in characterizing neuropathy, but one must understand their utility and limitations. Nerve conduction studies and electromyography can reveal patterns of neuropathy and nerve fiber type involvement as well as demonstrate axonal or demyelinating pathophysiology, but electrophysiologic abnormalities may not entirely correspond to clinical findings, which can be confusing to practitioners. For example, most patients with SjD with electrophysiologic evidence of sensorimotor polyneuropathy have predominantly sensory symptoms with minimal or no clinical muscle weakness.[11] Another point of confusion is that routine nerve conduction studies do not detect SFN, and a skin biopsy or specialized neurophysiologic testing may be necessary to confirm the diagnosis. Similarly, specialized autonomic nervous system testing is needed to demonstrate autonomic neuropathy but may not be available in all locations. Despite the challenges, electrophysiologic studies complement the clinical examination to better assess and classify PNS disorders.

Disparate definitions of neuropathies

The Sjögren's Foundation convened an interdisciplinary panel of experts to develop evidence-based CPGs for PNS manifestations of SjD. This panel, including neurologists and rheumatologists, followed an evidence-based process during which it became clear that the medical literature contains disparate definitions of types of neuropathies. Examples are provided in Table 1.

Which type is the most common in SjD? As shown in Table 1, different studies suggested different frequencies, in part due to the small sample size in most studies. Together, heterogeneity of data and definitions often leads to difficulty in understanding and reconciling the data.

Aligned nomenclature of the PNS neuropathies

This stark lack of standardized nomenclature highlights a broader issue impacting effective communication across medical specialties. To address this need, the Sjögren's Foundation PNS guideline panel undertook an effort to create shared definitions of the types of neuropathies that occur in SjD. The project aimed to define specific PNS manifestations that could be understood across disciplines and interdisciplinary teams. We initially identified differences in terminology and definitions used by both specialties as well as areas of overlap. We then developed an aligned nomenclature and defined the terminology used to describe specific peripheral neurologic manifestations of SjD to ensure agreement among rheumatologists, neurologists, and other medical specialties (Table 2). Terms describing seven PNS categories were delineated, with descriptions of clinical presentations, possible etiologies, diagnoses, pathologies, and evaluation. This included commonly used synonymous terms for clarification.

Conclusions

Clear communication among specialists involved in the care of patients with complex conditions, such as SjD, is essential. The communication gap identified by the Sjögren's Foundation PNS CPGs panel highlights a challenge for multispecialty collaboration in clinical management and clinical research. The development of shared definitions for PNS manifestations bridges this gap by harmonizing the terminology used by rheumatologists and neurologists. This alignment of nomenclature is an attempt to enhance communication across different medical specialties with the goal of improving the multidisciplinary management of autoimmune-mediated peripheral neuropathies, ultimately leading to better patient outcomes and a higher quality of care.

AUTHOR CONTRIBUTIONS

All authors contributed to at least one of the following manuscript preparation roles: conceptualization AND/OR methodology, software, investigation, formal analysis, data curation, visualization, and validation AND drafting or reviewing/editing the final draft. As corresponding author, Ms. Hammitt confirms that all authors have provided the final approval of the version to be published, and takes responsibility for the affirmations regarding article submission (eg, not under consideration by another journal), the integrity of the data presented, and the statements regarding compliance with institutional review board/Declaration of Helsinki requirements.

REFERENCES

1. Thoma A, Eaves FF III. A brief history of evidence-based medicine (EBM) and the contributions of Dr David Sackett. Aesthet Surg J 2015;35(8):NP261–NP263.
2. Djulbegovic B, Guyatt G. Evidence vs consensus in clinical practice guidelines. JAMA 2019;322(8):725–726.
3. Gagliardi AR, Brouwers MC, Palda VA, et al. How can we improve guideline use? A conceptual framework of implementability. Implement Sci 2011;6:26.
4. Shiffman RN, Dixon J, Brandt C, et al. The GuideLine Implementability Appraisal (GLIA): development of an instrument to identify obstacles to guideline implementation. BMC Med Inform Decis Mak 2005;5:23.
5. Gupta S, Rai N, Bhattacharrya O, et al. Optimizing the language and format of guidelines to improve guideline uptake. CMAJ 2016; 188(14):E362–E368.
6. Patel R, Shahane A. The epidemiology of Sjögren's syndrome. Clin Epidemiol 2014;6:247–255.
7. Lackner A, Ficjan A, Stradner MH, et al. It's more than dryness and fatigue: the patient perspective on health-related quality of life in primary Sjögren's syndrome - a qualitative study. PLoS One 2017; 12(2):e0172056.
8. Liampas A, Parperis K, Erotocritou MF, et al. Primary Sjögren syndrome-related peripheral neuropathy: a systematic review and meta-analysis. Eur J Neurol 2023;30(1):255–265.
9. Sireesha Y, Kanikannan MA, Pyal A, et al. Patterns of peripheral neuropathy in Sjögren's syndrome in a tertiary care hospital from South India. Neurol India 2019;67(Supplement):S94–S99.
10. Mori K, Iijima M, Koike H, et al. The wide spectrum of clinical manifestations in Sjögren's syndrome-associated neuropathy. Brain 2005; 128(Pt 11):2518–2534.

11. Delalande S, de Seze J, Fauchais AL, et al. Neurologic manifestations in primary Sjögren syndrome: a study of 82 patients. Medicine (Baltimore) 2004;83(5):280–291.
12. Sene D, Cacoub P, Authier FJ, et al. Sjögren syndrome-associated small fiber neuropathy: characterization from a prospective series of 40 cases. Medicine (Baltimore) 2013;92(5):e10–e18.
13. Terrier B, Lacroix C, Guillevin L, et al. Diagnostic and prognostic relevance of neuromuscular biopsy in primary Sjögren's syndrome-related neuropathy. Arthritis Rheum 2007;57(8):1520–1529.
14. England JD, Asbury AK. Peripheral neuropathy. Lancet 2004; 363(9427):2151–2161.
15. Barohn RJ, Amato AA. Pattern-recognition approach to neuropathy and neuronopathy. Neurol Clin 2013;31(2):343–361.
16. Cascio MA, Mukhdomi T. Small Fiber Neuropathy. StatPearls; 2023.
17. Small fiber sensory neuropathy. Johns Hopkins Medicine. Accessed November 6, 2023. https://www.hopkinsmedicine.org/neurology-neurosurgery/specialty-areas/peripheral-nerve/small-fiber-sensory-neuropathy
18. Birnbaum J, Lalji A, Saed A, et al. Biopsy-proven small-fiber neuropathy in primary sjögren's syndrome: neuropathic pain characteristics, autoantibody findings, and histopathologic features. Arthritis Care Res (Hoboken) 2019;71(7):936–948.
19. Hays AP. Utility of skin biopsy to evaluate peripheral neuropathy. Curr Neurol Neurosci Rep 2010;10(2):101–107.
20. Jani-Acsadi A, Lewis RA. Evaluation of a patient with suspected chronic demyelinating polyneuropathy. In: Said G, Krarup C, eds. Handbook of Clinical Neurology. Elsevier; 2013:253–264.
21. Pereira PR, Viala K, Maisonobe T, et al. Sjögren sensory neuronopathy (Sjögren ganglionopathy): long-term outcome and treatment response in a series of 13 cases. Medicine (Baltimore) 2016;95(19): e3632.
22. Gwathmey KG. Neuromuscular notes: sensory neuronopathies. Practical Neurology 2020:34–37.
23. Vital C, Vital A, Canron MH, et al. Combined nerve and muscle biopsy in the diagnosis of vasculitic neuropathy. A 16-year retrospective study of 202 cases. J Peripher Nerv Syst 2006;11(1):20–29.
24. Golden EP, Vernino S. Autoimmune autonomic neuropathies and ganglionopathies: epidemiology, pathophysiology, and therapeutic advances. Clin Auton Res 2019;29(3):277–288.
25. Davies K, Ng WF. Autonomic nervous system dysfunction in primary Sjögren's syndrome. Front Immunol 2021;12:702505.
26. Vernino S, Low PA, Fealey RD, et al. Autoantibodies to ganglionic acetylcholine receptors in autoimmune autonomic neuropathies. N Engl J Med 2000;343(12):847–855.
27. Kimpinski K, Iodice V, Vernino S, et al. Association of N-type calcium channel autoimmunity in patients with autoimmune autonomic ganglionopathy. Auton Neurosci 2009;150(1–2):136–139.
28. McKeon A, Lennon VA, Lachance DH, et al. Ganglionic acetylcholine receptor autoantibody: oncological, neurological, and serological accompaniments. Arch Neurol 2009;66(6):735–741.
29. Seeliger T, Prenzler NK, Gingele S, et al. Neuro-Sjögren: peripheral neuropathy with limb weakness in Sjögren's syndrome. Front Immunol 2019;10:1600.
30. Birnbaum J. Peripheral nervous system manifestations of Sjögren syndrome: clinical patterns, diagnostic paradigms, etiopathogenesis, and therapeutic strategies. Neurologist 2010;16(5):287–297.