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Small Nerves, Big Problems

A Comprehensive Patient Guide to Small Fiber Neuropathy

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1 What Is Peripheral Neuropathy?

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Peripheral neuropathy is one of the most common and most complex diseases seen in neurology clinics, and it can be experienced by patients in numerous patterns with a variety of symptoms.

Peripheral neuropathy is defined as a functional disturbance or pathological change in the peripheral nervous system (PNS), which is the system of nerves outside of the brain and spinal cord.

The PNS is that part of the nervous system that connects the central nervous system (CNS)—the brain and spinal cord—with sensory receptors, muscles, blood vessels and glands. The cells that make up the PNS are called neurons. Neurons have long projections, called axons, that are comparable to electrical wires.

An axon can either be **myelinated** or **unmyelinated**. Myelinated fibers are insulated and transmit nerve impulses at a higher speed than unmyelinated fibers, which have no insulation.

The PNS is divided into sensory and motor parts. The **sensory division** transmits information from the skin, body, limbs, and organs to the CNS. Information from the skin, body, and limbs includes the sensations of touch, pressure, vibration, proprioception (awareness of the position of the body), pain, and temperature. Information from organs includes stretch, pain, temperature, chemical changes, and irritation. Special senses such as vision, hearing, taste, smell, and balance are also transmitted through the sensory division of the PNS.



Chapter 1 Summary

Peripheral neuropathy is a functional disturbance or pathological change in the peripheral nervous system. It is one of the most common diseases seen by a neurologist.

The human body's nervous system is made up of three main parts:

- The central nervous system (CNS) is the brain and spinal cord, which control the activities of the body.
- The peripheral nervous system (PNS) connects the central nervous system with sensory receptors, muscles, blood vessels and glands, and is made up of sensory, motor and autonomic nerves.
- The autonomic nervous system (ANS) controls many of the automatic or subconscious functions of the body such as regulating blood pressure or digesting food

Diabetes is the most common cause of neuropathy in the developed world, but other things that can cause neuropathy are autoimmune disease, kidney disease, metabolic and endocrine disorders, nutritional deficiency, cancer, infection, trauma, toxins, and hereditary disease.

Aides (or clues) to help diagnose neuropathy include:

- The timespan of symptoms being noticed by the patient
- Location of symptoms
- By the size of the nerve fibers that are affected
- By the type of nerve cell affected
- Through a skin biopsy and/or other lab tests (see chapter 3)

Common signs and symptoms of small fiber neuropathy may include:

- sensations of burning, cold, tingling, shock-like and itching
- difficulty feeling temperature and pain which could result in burns and an inability to feel cuts on the bottom of the feet
- widespread symptoms of pain throughout the body and its extremities
- physical changes in the skin, hair, joints, and bone
- changes in the patient's emotional well-being and quality of life

Key Terminology

peripheral neuropathy • central nervous system • peripheral nervous system
sensory nerves • motor nerves • autonomic nerves
axon (small vs. large axons) • small fiber neuropathy

2 What Is Small Fiber Neuropathy?

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What is small fiber neuropathy?

This direct question can often be a challenging one to discuss and to address for a physician. The question itself, for a physician who specializes in neuropathy, is a relatively brief answer: Damage to the small unmyelinated or lightly myelinated nerve fibers outside of the brain and spinal cord. Of course, for many people, the answer usually just raises more questions.

There are a number of different types of nerve fibers. Nerves are designed to send information to and from your brain to different parts of your body, but the nerves are designed to only carry a specific type of information, and only in one direction.

Some nerves are located in the brain or the spinal cord, and these are part of the central nervous system. Peripheral nerves refer to nerves that are outside of the brain and the spinal cord. That is where the term “peripheral” comes from when describing “peripheral neuropathy”—the nerves that are involved are outside of the brain and spinal cord.

Some nerves send signals very quickly—these are the *large nerve fibers*. Other nerves send information more slowly—these are the *small nerve fibers*. Large nerve fibers operate like a highway, sending information quickly from one place to another. However, just as there is limited space on the highway, there is limited space for nerves that can send information quickly. In order to understand why these fast-conducting nerves are called “large nerve fibers,” there needs to be an understanding of how nerve fibers are constructed. Large nerve fibers have a coating around them, called myelin, that allows the information to move quickly—think of this as the large clear highway that allows rapid transit. Figure 1 shows the structure

Chapter 2 Summary

TWO

Small fiber neuropathy (SFN) is a disease where damage has occurred to the small unmyelinated or lightly myelinated nerve fibers outside of the brain and spinal cord.

SFN may show symptoms in a number of different ways, which is why it can be difficult for a doctor to diagnose.

SFN can affect the feet or different parts of the body. Each patient's experience may be unique.

SFN may involve sensory nerves or autonomic nerve fibers.

The types of symptoms that are typically associated with small fiber sensory neuropathy can be broken down into two different potential areas:

1. Loss of function (loss of sensation if sensory fibers are involved, or loss of autonomic function if autonomic fibers are involved)
2. Pain

The types of symptoms that are typically associated with small fiber autonomic neuropathy may include any of the following:

- High resting heart rate, or a heart rate that does not increase enough with exercise
- A drop in blood pressure with standing (orthostatic hypotension), causing lightheadedness, dizziness or fainting
- Constipation or diarrhea
- Trouble digesting (*gastroparesis*), vomiting, early fullness
- Going to the bathroom frequently, having to go suddenly, or difficulty emptying the bladder
- Erectile dysfunction in men. Vaginal dryness in women that can cause painful intercourse
- Reduced sweating, or too much sweating. This can cause dry skin or irritated skin
- Trouble regulating body temperature
- Dryness or blurry vision
- Dry mouth

Key Terminology

small fiber neuropathy • small fiber sensory neuropathy

small fiber autonomic neuropathy • myelin

large nerve fibers • small nerve fibers

Glossary

Acceptance and commitment therapy (ACT): a newer type of therapy that is based on changing mindset so that people can begin to live their lives in ways consistent with defined life goals and values.

ARA 290: an erythropoietin derivative, ARA 290 is a small protein engineered to activate the body's own repair process that is triggered by inflammation. ARA 290 looks to be a potential disease-modifying agent for the treatment of SFN caused by sarcoidosis.

Autonomic nerves: these nerves are a type of motor nerve that control the movements of your body that are on "autopilot," such as smooth muscle and cardiac muscle functions.

Autonomic nervous system (ANS): controls many of the automatic or subconscious functions of the body such as going to the bathroom, regulating blood pressure, or digesting food

Autonomic testing: a non-invasive test that takes EKG and blood pressure readings under different situations (such as breathing in a certain way) to look for problems with autonomic function. An example is a tilt-table test, which measures blood pressure and heart rate responses to being upright.

Axon (small vs. large axons): an axon is the long projection attached to a neuron (the type of cells that compose the peripheral nervous system). Small axons convey impulses slowly to the autonomic nervous system and have little to no myelin covering. Large axons convey impulses quickly, are generally found in the somatic (voluntary) nervous system, and have a myelin coating.

Burning mouth syndrome: condition in which patients have a severe burning sensation in their mouth and tongue with no obvious cause.

Celiac disease: an autoimmune disorder that can occur when the ingestion of gluten (a protein found in wheat, rye, and barley) causes an immune response that attacks the small intestine.

Central nervous system (CNS): the brain and spinal cord which control