



Your Questions Answered

NEUROPATHY

Q What is autonomic neuropathy? How did I get it, and what can I expect as the disease progresses?



DR. LOUIS H. WEIMER RESPONDS:

A Autonomic neuropathy involves damage to certain nerves that run through the peripheral nervous system. Peripheral nerves transmit electrical signals from the brain and spinal cord to and from the rest of the body. Each nerve fiber serves a dedicated function: motor nerves control movement; sensory nerves control physical sensation; and autonomic nerves transmit signals for the “automatic” functions, such as heart rate and blood pressure, which require no conscious control.

Because autonomic nerves help control virtually every organ in the body, the symptoms of autonomic neuropathy can be highly varied and widespread. Common complaints include dry eyes and mouth, reduced or excessive sweating, bloating, nausea, or episodes of diarrhea. Autonomic functions decline to some degree with normal aging.

Autonomic neuropathy can worsen, improve, or continue unchanged depending on the severity and progression of the underlying cause. The most common cause is diabetes. Severe cases of autonomic neuropathy can result from autoimmune or bone marrow disorders, certain toxins and medications, and rare genetic conditions. In some

cases, no cause can be identified.

Some degree of autonomic neuropathy occurs in the majority of neuropathy cases. Patients with neuropathy affecting their sensory nerves typically have more severe foot and lower leg involvement; the condition is often aggravated by additional autonomic impairment in affected areas—for example, coldness, loss of sweating and hair, and susceptibility to foot ulcers.

Louis H. Weimer, M.D., is co-director of Columbia Neuropathy Research Center and associate clinical professor of neurology at Columbia University in New York, NY.

APHASIA

Q Three years ago I suddenly couldn't talk. My diagnosis was primary progressive aphasia (PPA). What causes PPA? What are the best ways to treat and manage it?



DR. MARSEL MESULAM RESPONDS:

A PPA is caused by diseases that impair the function of nerve cells in language centers of the brain. In most right-handers, these centers are on the left side of the brain. The symptoms—which include impaired word-finding, spelling, and word comprehension—emerge very slowly. If the symptoms appear suddenly, other diagnoses should be considered.

In contrast to typical forms of Alzheimer's disease, patients with PPA may have intact memory for recent events and pursue complex activities of daily life. The memory problems are only related to language. PPA is usually caused by a disease called frontotemporal lobar degeneration, but in about 30 percent of patients, the cause may be an atypical form of Alzheimer's disease. This is why many doctors will prescribe drugs that are also used in Alzheimer's. Speech therapy may be useful for some patients. PPA has many different forms and it is crucial to individualize patient care.

Marsel M. Mesulam, M.D., is the Dunbar Professor of Neurology and Psychiatry and director of the Cognitive Neurology and Alzheimer's Disease Center at Northwestern University in Chicago, IL.

DO YOU HAVE A QUESTION TO ASK THE EXPERTS?
Send it to neurologynow@lwwny.com



A patient with a painful neuropathic foot.

PICK'S DISEASE

Q My wife has Pick's disease. Is it similar to Alzheimer's? Is there any hope for a cure?



**DR. WILLIAM
SEELEY
RESPONDS:**



**A brain with
Pick's disease
showing atrophy
of the frontal and
temporal lobes**

A Pick's disease, also known as frontotemporal dementia (FTD), is a progressive neurodegenerative disease that shares many features with Alzheimer's. Both illnesses result from misfolded proteins that interfere with brain cell function and cause those cells to slowly die over time. However, the genes and proteins implicated in FTD and Alzheimer's disease are different. Patients with FTD usually develop symptoms in their 50s or 60s, which is signifi-

cantly earlier in life than most patients with Alzheimer's. Also, whereas Alzheimer's disease begins with memory loss, FTD usually begins with behavioral changes or problems with speech and language.

Treatments for FTD remain limited. A placebo-controlled trial of a drug called memantine is actively enrolling patients; for more information, visit tinyurl.com/cfy9w. Other trials likely to begin in the next year are listed on the Web site clinicaltrials.gov: search by typing "frontotemporal dementia."

Although potential cures remain years away, recent advances in the understanding of basic FTD biology, including the discovery of new causative genes and proteins, have fueled a surge of optimism among FTD researchers.

William Seeley, M.D., is assistant professor of neurology at University of California, San Francisco Memory & Aging Center.

ATAXIA

Q What causes adult-onset ataxia, and how can it be treated?



**DR. WILLIAM
WEINER
RESPONDS:**

A The word "ataxia" is used to describe a symptom—lack of coordination—which can be associated with injuries or degenerative changes in the central nervous system. Examples of such injuries and changes include stroke, multiple sclerosis, head injury, or alcoholism. This is known as acquired ataxia.

Ataxia also indicates a group of specific degenerative and progressive diseases of the nervous system called the hereditary and sporadic ataxias. These diseases damage parts of the nervous system that control movement. Often the first apparent symptom of these disorders is difficulty with balance and walking. Symptoms of hereditary ataxias commonly begin in childhood, but one type—Friedreich's ataxia—has an adult onset in some cases. People with Friedreich's ataxia develop weakness in the muscles of the feet, lower legs, and hands. They often rely on

a wheelchair within 15 years of the appearance of symptoms. As the disease progresses, patients may experience slow, slurred speech; rapid, involuntary eye movements; spinal curvature (scoliosis); and heart disease and heart failure.

People with sporadic ataxia have symptoms that usually begin in adulthood but no known family history of the disease. Sporadic ataxias may result from a new abnormality of the gene or as a result of an underlying disease, including thyroid disease, chronic hypoglycemia, stroke, and vitamin deficiencies.

The type of ataxia and the age of symptom onset indicate how severe the disability will become and whether the disease will lead to death. There is no specific treatment for ataxia and no cure for the hereditary ataxias, but adaptive devices (such as canes or walkers) and therapies (such as speech therapy to improve speech and aid swallowing) can help. To identify an ataxia, your physician should take your medical and family history and conduct a complete neurological evaluation, including an MRI scan of the brain. Genetic blood tests are now available to confirm a diagnosis for some types of hereditary ataxias.

William Weiner, M.D., is chairman of the department of neurology at the University of Maryland School of Medicine and chief of Neurology at the University of Maryland Medical Center in Baltimore, MD.