## What is Chronic Inflammatory Demyelinating Polyneuropathy (CIDP)?

Chronic Inflammatory Demyelinating Polyneuropathy is an acquired chronic, but treatable, condition affecting both motor and sensory peripheral nerves that connect the spinal cord with the muscles. The damage to these nerves by a patient's immune system results in muscle weakness and loss of sensation in the legs that can progress in a symmetrical, chronic fashion to include the arms. Over time, if left untreated, it can lead to major disability. The clinical course can be progressive over many years, progress and remit over one to three years or occur in repeated episodes separated by months.

## Mission Statement

To improve the quality of life for individuals and families worldwide affected by GBS, CIDP and variants by:

- Providing a network for all patients, their caregivers and families so that GBS or CIDP patients can depend on the Foundation for support and reliable, up-to-date information.
- Providing public and professional educational programs worldwide designed to heighten awareness and improve the understanding and treatment of GBS, CIDP and variants.
- Expanding the Foundation's role in sponsoring research and engaging in patient advocacy.

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# CIDP

## Chronic Inflammatory Demyelinating Polyneuropathy

Support
Education
Research
Advocacy

*Working for a future when no one afflicted with Guillain-Barré syndrome (GBS), chronic inflammatory demyelinating polyneuropathy (CIDP) or variants suffers alone and every patient has a full recovery.*

## What are the Symptoms of CIDP?

The number of new cases of CIDP each year is estimated to 1.5 to 8.5 people per million and, because of the chronic nature of the disease, may affect up to 40,000 patients in the U.S. at any one time. This disorder can affect children and adults of any age.

The peak period of life during which patients typically develop CIDP is between 50 to 60 years of age. The disorder is more common in men than in women.

CIDP is characterized by progressive symmetrical weakness and sensory loss in a patient's legs and arms that develop in a steady or stepwise fashion over more than eight weeks and can continue for years. Weakness is symmetric, occurring on each side of the body about equally, with loss of deep tendon reflexes, such as those at the knee and ankle, in affected extremities. If the disease is severe and long term over years, muscle wasting can occur in the hands and feet. Muscles used for breathing usually are not involved. Patients also have sensory loss that is most obvious in the hands and feet. Lost sensation includes pain, light touch, vibration and proprioception - the ability of a person to know the position of a body part relative to the rest of his or her body.

This longer progression of the condition distinguishes CIDP from acute neuropathies, such as Guillain-Barré Syndrome (GBS) in which patients develop their peak weakness within one-to-four weeks. In GBS, 85 percent of patients will recover their strength and walk after three months. Unlike GBS, CIDP frequently is not self-limiting and, if left untreated, about 30 percent of patients will progress to wheelchair dependence. About 60 percent of patients are able to continue to work, but with progressing disability. Early recognition and treatment can prevent disability from progressing to a significant degree.

Not all patients follow this typical pattern. A small number of patients have variants, including the Lewis-Sumner syndrome, which is multifocal and affects motor and sensory nerves in some areas of the arms greater than legs in an asymmetric manner (i.e., one arm more than the other). Another form primarily affects sensory nerves and results in poor balance, falls and pain.

## How is CIDP Diagnosed?

Treatment of CIDP early in the clinical course is very important as it is then more likely that patients will respond to treatment and some patients will be more likely to go into remission. Unfortunately, there is no reliable diagnostic test for CIDP. Instead, the diagnosis is based on a combination of the clinical history provided by the patient, the examination performed by a neurologist and supporting diagnostic studies, including nerve conduction studies and a spinal fluid examination. Nerve conduction velocities study the ability of a nerve to carry an electrical impulse. Studies on the patient's blood samples are also done to rule out other conditions such as diabetes, infections, toxin exposure and inflammatory disease of blood vessels. Rarely, a nerve biopsy also will be done to either confirm the diagnosis of CIDP or rule out other diseases or conditions.

## How Is CIDP Treated?

Up to 80 percent of CIDP patients respond to one or more therapies that modify the immune system. Several of these have been tested in randomized, controlled trials. As an initial measure, patients can be treated with corticosteroids, plasma exchange or intravenous immunoglobulin (IVIG), all of which are considered to be equally effective.
Early and adequate treatment is capable of limiting the disease and preserving function and quality of life. Patients with extensive nerve damage prior to beginning treatment may have a poor or incomplete response to treatment.

IVIG-C was shown to be beneficial in randomized and a placebo-controlled trial that extended out 48 weeks. This study resulted in the only drug indication for IVIG-C in CIDP by the U.S. Food and Drug Administration (FDA). In patients who do not respond to these measures, other immune-suppressing drugs can be used, including cyclophosphamide, azathioprine and cyclosporin, but these drugs have not been tested in controlled trials. In addition, side effects such as cancer of the white blood cells and kidney or liver damage associated with the long-term use of these drugs limit their benefit with chronic disorders.

Physical and occupational therapy, as well as aids that facilitate specific function - such as a hook to facilitate buttoning a shirt, can help patients to maintain activities of daily living through exercise. However, excessive exercise may result in increased muscle pain and poorer function, if not monitored properly by a trained and certified professional.

## What Causes CIDP?

Evidence suggests that the patient's own immune system - including both white blood cells and antibodies - is stimulated to damage the myelin layer that surrounds the motor and sensory nerves. Destruction of the myelin results in the loss of the ability of a nerve to conduct or carry an electrical impulse and leads to muscle weakness and decreased ability to feel different sensations. With time and severity of the disease, the nerve fiber, or axon, itself can be damaged leading to poor recovery even with good treatment, particularly in older patients. Early information suggests that the immune response may reflect overlapping gene mutations. Regardless, this disease is most likely not inherited from parent to child.

**Although CIDP can affect children and adults of any age, the peak period of life during which patients typically develop this disorder is between 50 to 60 years of age. It is more common in men than women.**