

CIDP

EVERYTHING YOU NEED TO KNOW

CIDP (Chronic Inflammatory Demyelinating Polyneuropathy) is a rare disorder of the peripheral nerves characterized by gradually increasing sensory loss and weakness associated with loss of reflexes.

While GBS and CIDP share many features, one that separates them is the onset: in GBS the onset to maximum weakness occurs in under 30 days and in most people in under 14 days, while in CIDP the sensory loss and weakness progress beyond those times. The incidence of new cases of CIDP is about 1-4 per million people but as the disease can be present in any one person for a long time, the prevalence may be as high as 9 per 100,000.

Like GBS, CIDP is caused by damage to the covering of the nerves, called *myelin*. It can start at any age and is more frequent in men than women.

Unlike GBS, CIDP is not self-limiting (with an end to the acute phase). Left untreated, 30% of CIDP patients will progress to wheelchair dependence. Early recognition and proper treatment can avoid a significant amount of disability.

What causes CIDP?

Current theory holds that the body's immune system which normally protects itself perceives myelin as foreign and attacks it. Just what causes this process is not clear.

How is CIDP diagnosed?

Diagnosis of CIDP is based on the combination of symptoms, signs and test results:

- Symptoms such as loss of sensation (numbness), abnormal sensation (tingling and pain) and weakness (difficulty walking, foot drop)
- Signs such as loss of sensation, weakness and loss of reflexes
- Tests such as nerve conduction and EMG (usually showing a demyelinating neuropathy), spinal tap (usually showing elevated protein with normal cell count), blood and urine tests (to rule out other disorders that may cause neuropathy and to look for unusual proteins)

How is CIDP treated?

There are three standard or first line treatments in CIDP:

- Corticosteroids (*Prednisone*, *Prednisolone*) are similar to naturally occurring anti-inflammatory hormones made by the body, and can be used as an initial treatment.

Corticosteroids often improve strength, are conveniently taken by mouth, and are inexpensive. Side effects however can limit long-term use.

- High dose *Intravenous Immune Globulins (IVIG)* is the only drug that has FDA, Canadian and European approval for treatment of CIDP. IVIG contains naturally occurring antibodies obtained from healthy volunteers. IVIG is given through a vein. Newer preparations of higher concentrations that can be given under the skin (subcutaneous) are currently being tested in controlled trials in CIDP patients.
- *Plasma Exchange (PE), or Plasmapheresis (PLEX)*, is a process by which some of the patient's blood is removed and the blood cells returned without the liquid plasma portion of the patient's blood. It may work by removing harmful antibodies contained in the plasma.

There are a large number of so-called second line drugs used to treat CIDP. These are usually used when other treatments fail or have significant side-effects or the clinical response is not optimal. These drugs are largely not tested in randomized controlled trials, but their use is supported by case series from the medical literature.

There are a number of so-called third line treatments, usually chemotherapy drugs, but these should be given only in selected circumstances and by those with extensive experience in their use.

There are also ongoing research studies (see www.clinicaltrials.gov)

Treatment of CIDP is an art. An experienced doctor is more likely to have good outcomes than someone treating their first case as is true throughout medicine. That is why we have set up the Centers of Excellence program. If treated early, most CIDP patients respond well to therapy that can limit the damage to peripheral nerves and contribute to improved function and quality of life and at times cure.

CIDP and its Variants

Typical CIDP is a symmetrical motor and sensory progressive neuropathy affecting proximal and distal muscles with areflexia.

Lewis-Sumner syndrome is a sensory-motor disorder in which there is sensory loss and weakness in the distribution of individual nerves. Diagnostic nerve conduction studies confirm the focal nerve involvement.

Pure sensory CIDP presents with sensory loss, pain and poor balance with abnormal gait or walking. There is no weakness but frequently motor nerve conduction studies are abnormal in addition to sensory conduction studies.

Pure motor CIDP presents with weakness and loss of reflexes without sensory loss.

There are other less well established variants most of which would fall into the category of CIDP.

Living with CIDP

Post-treatment life depends on whether the disease was caught early enough to benefit from treatment options. Patients respond in various ways. The gradual onset of CIDP can delay diagnosis by several months or even years, resulting in significant nerve damage that may limit and delay the response to therapy. The chronic nature of CIDP requires long-term care of patients. Accommodations in the home may be needed to facilitate activities of daily living.