CHRONIC INFLAMMATORY DEMYELINATING POLYNEUROPATHY (CIDP)

An Autoimmune Neurological Disorder

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Chronic inflammatory demyelinating polyneuropathy (CIDP) is a chronic autoimmune neurological disorder that causes a slowly progressiveness motor sensory weakness and a loss of sensation in the legs and arms.

In CIDP, chronic refers to the persistence of this condition. Demyelinating refers to the specific nervous system injury in which the outer myelin covering of nerves is damaged or destroyed, and polyneuropathy refers to the fact that multiple nerves, both proximal and distal nerves, and multiple locations within the body are affected. CIDP is similar to Guillain-Barre syndrome (GBS), which is an acute condition causing a similar type of neuropathy. Unlike CIDP, GBS generally resolves spontaneously.

At one time, CIDP was considered a chronic or persistent condition of GBS. Today, these two disorders are considered separate and distinct, although they cause similar symptoms. Both conditions are known to cause symptoms of fatigue, tingling, loss of reflexes, pain, tingling in the fingers and toes, numbness, weakness, and paralysis.

**Symptoms**

In both conditions, symptoms vary in severity and respond to treatment, especially early treatment intervention used to reduce inflammation. In CIDP, both sides of the body are equally affected (symmetric), but in conditions of multifocal CIDP or other CIDP variants, only one side of the body, for instance, one leg, is affected. The onset of symptoms in CIDP is variable and ranges from several weeks to patients with relapsing-remitting disease to several weeks to several months in more progressive monophasic conditions. In the relapsing form, episodes of symptoms alternate with periods of remission.

Weakness tends to be the most disabling feature in patients with relapsing-remitting CIDP and weakness tends to be more prominent than muscle atrophy. A mild sensory loss is seen in all types of CIDP, and tremor is a common feature. Pain is more likely to occur in patients with sensory CIDP. Problems with urination may also occur in CIDP.

**Environmental Triggers**

Both CIDP and GBS are known to develop after viral infection, especially hepatitis, and after vaccinations. The autoimmune mechanism is uncertain but molecular mimicry has been suggested. In this case viral components can take on the appearance of the body's
own proteins, causing the immune system to react to them. Both IgM and IgG antibodies to beta-tubulin and heparan sulfate are seen in CIDP.

Who is Affected?

Males are affected twice as often as women and the average age at the time of disease onset is 50 years for patients with progressive CIDP and 27 years for the relapsing-remitting form of CIDP. The prognosis is worse for patients with progressive disease courses with central nervous system involvement and increased axonal loss on imagining studies. Similar to GBS, some patients with CIDP will experience one episode and spontaneously recover. Some of these patients may experience a residual numbness or weakness.

Diagnosis and Pathology

CIDP is diagnosed with electrophysiology studies showing slow nerve conduction velocities, variable velocities among nerves and prolonged latencies of F-waves. Blood tests show the presence of IgM and IgG tubulin antibodies and IgM heparan sulfate antibodies. Levels of M protein are usually seen in spinal fluid samples from patients with CIDP. Tissue studies show increased binding of IgG to Schwann cell processes although nerve biopsies are not usually necessary to diagnose CIDP. Imaging tests, particularly MRI, show nerve hypertrophy.

The demyelination process in CIDP is related to macrophage cell activity. Thin myelin sheaths are typically seen with inflammation occurring as an onion-bulb appearance, and axons devoid of myelin may be seen. The body's attempts to repair myelin in CIDP result in loosely compacted myelin sheaths and Schwann cell changes.

CIDP Variants

A number of variants of CIDP have been reported. The most common variant is multifocal or Lewis-Sumner CIDP, which affects arms more than legs, with asymmetric presentation. The distal nerves of the outer extremities such as the hands are usually affected more than the proximal (inner) extremities such as the shoulders.

Other forms of CIDP include focal upper limb demyelinating CIDP, which is characterized by anti-GM1 ganglioside antibodies; sensory CIDP; and Childhood CIDP, which affects children from early childhood to teens and is rarely seen in infancy. CIDP may also develop in patients with insulin dependent type 1 diabetes mellitus, systemic lupus erythematosus, or Sjogren's syndrome. In some cases it can be difficult to distinguish CIDP from multifocal motor neuron disease or POEMS syndrome.

Treatment
Treatment for CIDP consists of corticosteroids such as prednisone and non-steroidal immunosuppressants such as cyclosporine A and methotrexate. When immunosuppressive treatments are inadequate, patients are treated with intravenous immunoglobulins (IVIg therapy) and plasmapheresis. Regardless of the primary treatment used, patients with CIDP usually benefit from a combination of exercise and massage therapy. IVIg treatment has recently been reported as showing success when used intermittently to prevent relapses in patients with CIDP.

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